# Chapter 158: Complications of Temporal Bone Infection

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## General Considerations

Complications resulting from acute or chronic suppurative disease within the pneumatized spaces of the temporal bone are unusual; however, they are often serious, and to prevent incipient complications and to diagnose and treat existing ones properly at the earliest possible moment, the physician needs a high level of suspicion and a thorough understanding of pathogenesis.

This chapter clarifies some of the existing controversy and confusion about these complications by reviewing definitions, pathogenesis, and principles of diagnosis and treatment, so that otolaryngologists - head and neck surgeons in training as well as experienced practitioners can make accurate specific diagnoses earlier. Better management and patient outcomes should follow.

## Definitions

The dictionary defines *itis* as "a word termination denoting inflammation of the part indicated by the word stem to which it is attached"; this suffix does not indicate etiology.

*Otitis media, mastoiditis,* and *petrositis* merely indicate the site of inflammatory lesions. *Otitis media* refers to the middle ear, *mastoiditis* refers to the mastoid region of pneumatization within the temporal bone, and *petrositis* refers to the petrous apex portion of the temporal bone. The inflammation involves the lining of those spaces - that is, the mucosa - with its epithelial layer and lamina propria adjacent to the periosteum of the underlying bone. The lumen of the pneumatized space may contain an effusion. The presence or absence of an effusion within the lumen does not alter the definition of inflammation if the mucosa is involved with an inflammatory process (Bluestone and Klein, 1983a).

*Suppurative* means "producing crust or associated with suppuration". *Suppuration* is "the formation of pus; the act of becoming converted into and discharging pus". *Suppurative* therefore is a qualitative statement defining the gross observation of the effusion within the lumen of the space.

*Acute, subacute,* and *chronic* refers exclusively to time. By convention, *acute* refers to the presence of an event or disease for 3 weeks or less, *subacute* is between 3 weeks and 3 months, and *chronic* is for a time greater than 3 months (Bluestone and Klein, 1983a).

The five regions of pneumatization (Allam, 1969) within the temporal bone are the following:

1. Middle ear region

2. Mastoid region

3. Perilabyrinthine region

4. Petroux apex region

5. Accessory region.

To some degree, inflammation in the middle ear region is transmitted by contiguity to all other regions of pneumatization. Hence, otitis media is invariably also mastoiditis and petrositis if the petrous apex is pneumatized (Bluestone and Klein, 1983a; Neely, 1978). Description diagnoses of the types of otitis media (otitis media and mastoiditis) must include consideration of the following three things:

1. State of the mucosa

2. Content of the pneumatized spaces

3. Presence or absence of a tympanic membrane perforation.

If state of the mucosa is inflamed, one must consider the duration of that inflammation: acute, subacute, or chronic.

If the pneumatized space contains something other than air, an effusion exists if the tympanic membrane is intact, and a discharge exists if there is a perforation. The effusion or discharge is properly described as serous, mucoid, or suppurative (Bluestone and Klein, 1983a).

A *complication* from suppuration in the pneumatized spaces of the temporal bone is defined as the extension of inflammation beyond the confines of the lumen and its mucosa or the products of that inflammation (Mawson, 1974; Schuknecht, 1974; Shambaugh, 1967). The structures immediately adjacent to the mucosa of the pneumatized spaces are bone and the tympanic and round window membranes. When bone is involved, complications occur from bone destruction by either enzymatic reabsorption or osteitis. Even though inflammation of the bony periosteum can induce abnormal new bone growth and is frequently seen as a result of acute or chronic infection, it is not considered a complication. By definition, then, *otitis media, mastoiditis*, and *petrositis* refer to mucosal inflammation of those specific regions; they are not complications. But when bone destruction can be identified in these regions, a complication exists. I therefore suggest that the terms *mastoiditis with bone destruction* and *petrositis with bone destruction* be used to denote the complications, and *mastoiditis* and *petrositis* per se to refer to the noncomplicated inflammation associated with otitis media.

## Classification

Classification of complications varies slightly among authors, depending on whether acute mastoiditis with bone destruction (acute coalescent mastoiditis) and chronic mastoiditis with bone destruction (chronic mastoiditis with osteitis) are included. The following classification combines those of Mawson, Schuknecht, and me. Ossicular and tympanic membrane destruction are also complications. They are not reviewed here but are discussed in Part A of this chapter. The complications are the following:

Aural Complications

--> Mastoiditis with bone destruction

--> Subperiosteal abscess

--> Petrositis with bone destruction

--> Facial paralysis

--> Labyrinthitis (serous, suppurative, or chronic)

Intracranial complications

--> Extradural abscess

--> Lateralsinus thrombophlebitis or thrombosis

--> Subdural abscess

--> Meningitis

--> Brain abscess

--> Otitic hydrocephalus.

## Epidemiology

Complications can result directly from either acute otitis media and mastoiditis with suppurative effusion or chronic otitis media and mastoiditis with suppurative discharge. They may also arise from their first-order complications: acute otitis media (AOM) and mastoiditis with suppurative effusion and bone destruction or chronic otitis media and mastoiditis with suppurative discharge and bone destruction. It is helpful to relate each specific complication to the suppurative middle ear and mastoid conditions that commonly precede it. This is done in tabular form (Table 158-1) and in an algorithm (Fig. 158-1).

**Table 158-1.** Complications relative to type of infection

| Complication | AOM | ACMASTOM | COM |
|---|---|---|---|
| Mastoiditis with bone destruction | | X | |
| Petrositis with bone destruction | | X | |
| Facial paralysis | X | | |
| Labyrinthitis, serous | X | | X |
| Labyrinthitis, suppurative | | | X |
| Labyrinthitis, chronic | | | X |
| Subperiosteal abscess | | X | |
| Extradural granulation/effusion or abscess | X | X | |
| Sigmoid sinus thrombophlebitis | X | X | |
| Subdural effusion or abscess | X | | |
| Meningitis | X | | |
| Brain abscess | X | X | |
| Otitic hydrocephalus | X | X. | |

Incidence and prevalence data for complications of otitis media are as sketchy and incomplete as they are for acute and chronic otitis media. Acute or chronic otitis media with effusion is remarkably common in children and occurs at least once in 75% of children by the age of 3 years (Teele et al, 1983). The prevalence is much lower in children over 10 and adults.

Gower and McGuirt's excellent study (1983) analyzed 100 intracranial complications after reviewing more than 300.000 admissions to a single hospital over a 20-year period. The complication distribution was as follows:

| | |
|---|---|
| Meningitis | 76 |
| Subdural effusion or abscess | 8 |
| Brain abscess | 6 |
| Otitis hydrocephalus | 5 |
| Lateral sinus thrombophlebitis | 5. |

Of the complications, 73 resulted from AOM (63 meningitis, 8 subdural effusions, and 2 brain abscesses); 2 of the 73 had preexisting bone destruction as a first complication of the AOM and mastoiditis; 46 of the complications (39 meningitis and 7 subdural effusions or abscesses) occurred in infants 2 years of age or younger.

The approximate prevalence of chronic otitis media with discharge in children is only 9 in 1000 (Mawson and Ludman, 1979), and the annual incidence of cholesteatomas has been calculated to be 0.6 in 1000 patients, with the peak incidence occurring in the second and third decades of life (Harker, 1977).

The incidence and prevalence of aural complications are less clear. *Mastoiditis with bone destruction* and *petrositis with bone destruction* may result from AOM and mastoiditis with suppurative effusion or chronic otitis media and mastoditis with suppurative discharge, but bone destruction appears more likely to occur from chronic rather than from acute

infections. Juselius and Kaltiokallio (1972) discovered that only 29 cases of acute mastoiditis with bone destruction were reported in Finland during a 15-year period. Conversely, at the University of Michigan over 6 years Ritter (1977) discovered that 152 ears were operated on for cholesteatoma; of those, 40 cases had evidence of pathologic bone destruction, such as lateral semicircular canal fistulas, facial-nerve exposure, or pathologic dural exposure. Sheehy et al (19770, over 10 years, found that 104 of 949 patients with a cholesteatoma had a labyrinthine fistula.

In summary, the most common complications appear to be meningitis, serous labyrinthitis (see later discussion), and chronic otitis media and mastoiditis with bone destruction. All other complications are decidedly less common.

## Conditions and factors influencing development of complications

The two most important factors in the development of complications are (1) an acute infection in an anatomically susceptible ear, and (2) persistence of infection and the development of granulation tissue. Meningitis and facial paralysis arise early in the course of a single, isolated, suppurative AOM in young children and occur because the ear's peculiar anatomy provides an opportunity for the spread of infection.

Granulation tissue is a natural result of epithelial ulceration and is directly proportional to the persistence of infection. Its anatomic bulk reduces drainage, and its enzymatic production erodes bone and creates new anatomic pathways extending beyond the confines of the pneumatized spaces of the temporal bone. Suppurative (rather than serous or mucinous) effusion or discharge is far more likely to be associated with mucosal ulceration and granulation tissue production.

Five factors make up the fine detail of these aforementioned major determinates of the development of a complication: (1) bacteriology, (2) antimicrobial therapy, (3) host resistance, (4) anatomic routes of and barriers against spread, and (5) drainage.

### *Bacteriology*

The bacteria involved in the primary site and the complication predominantly depend on the presence or absence of a tympanic membrane perforation. Without perforation the involved organisms tend to be those that initiate the original acute infection. With perforation, there is contamination and subsequent colonization of organisms from the external auditory canal. Organisms frequently cultured from ears with suppurative AOM are predominantly *Streptococcus pneumoniae*, with the next most common being *Haemophilus influenzae*. Group A streptococcus is a distant third. *Staphylococcus aureus* and gram-negative bacillus are cultured infrequently (Bluestone and Klein, 1983a).

Intraoperative cultures of the ear with cholesteatoma (Harker and Koontz, 1977) revealed 70% of the 30 ears to have aerobes and 67% to have anaerobes; 50% of the ears contained both aerobes and anaerobes. By far the most common aerobe was *Pseudomonas aeruginosa.* The most common anaerobes cultured were *Bacteroides* and *Peptococcus, Peptostreptococcus;* the third most common anaerobe was *Propionobacterium acnes.* In 13% no organisms were cultured, in 30% only 1 organism was cultured, and in 57% multiple

organisms were cultured. In the cases in which only 1 organism was cultured, the predominant anaerobe was *P. acnes.* In the 57% of ears in which multiple organisms were cultured, the range was 2 to 11 organisms, with the average being slightly greater than 3 organisms per ear. The foul-smelling ears usually contained 5 to 11 organisms, and in all of the foul-smelling ears anaerobes and aerobes existed.

Cultures of the external auditory canal, taken when the tympanic membrane was intact, show a variety of anaerobic, aerobic, gram-negatiive, and gram-positive organisms similar to cultures from skin in other body areas. The number and ratio of bacteria vary widely, however, from one skin area to another and from patient to patient. The predominant organism in a dry external auditory canal is *Staphylococcus;* however, with any degree of moisture and mild diffuse external otitis, *Pseudomonas* organisms begin to predominate rapidly (Senturia et al, 1980).

Neither pathogenicity, the capacity of a general group of organisms to cause disease, nor virulence, the degree of pathogenicity of a given organism for a particular host under specific conditions (MacLeod and Bernheimer, 1965), have been systematically studied in patients with complications of suppurative ear disease.

### *Antimicrobial therapy*

In vitro sensitivies and in vivo peak and trough blood levels of antimicrobials help to determine the appropriate choice and dosage of the drug. However, sufficient duration of therapy is of equal importance, and knowledge of it is derived only by inference from experiences with similar cases. Even with an appropriate antibiotic and blood levels given for what apppears to be a sufficient duration, the therapy may be compromised by alterations in the metabolic state of the organism, antagonism by other drugs or metabolites from infection, and unusual barriers to drug penetration (Goodman and Gilman, 1975). Thus, despite adequate antibiotic therapy, the organisms might persist, and a "masked" infection might later result in a complication when the bony barriers give way. Careful monitoring of the patient's clinical course and radiographic findings is important in confirming the resolution of an acute infection. The resolution of a chronic infection is harder to discern.

### *Host resistance*

Even bactericidal antibiotics require host defense mechanisms for the eradication of infection. The primary local host response is known as the inflammatory reaction (Thorn et al, 1977). The inflammatory reaction itself may add to the disease process by causing bone resorption and obstruction to drainage. In classifying the five stages of suppurative AOM (hyperemia, exudation, suppuration, coalescence, and complication), Shambaugh (1967) described the inflammatory reaction, which takes the following course:

1. Constriction followed by dilatation of small blood vessels (hyperemia).

2. Leukocytic adhesion to the endothelium.

3. Increaed vessel permeability from damaged mast cell release of histamine and serotonin (exudation).

4. Chemotactic leukocytic infiltration of extra-vascular tissue and spaces (suppuration).

5. Dilatation of lymph vessels in response to tissue edema.

6. Fiber and clot formation with the formation of barriers against the spread of infection.

7. Proliferation of fibrous and vascular tissue known as granulation tissue. The proliferating capillaries in the granulation tissue are surrounded by pericytes, some of which transform into histiocytes. The histiocytes predominate in the marginal zone close to bone (Thomsen et al, 1977). This zone of granulation tissue has beenshown to have marked acid phosphatase activity associated with bone resorption (coalescence).

Granulation tissue is much more responsible for bone resorption than is squamous epithelium. In cases of cholesteatoma, Sadé et al (1977) never found cholesteatoma matrix lying on bone lesion without granulation tissue, connective tissue, or both between the cholesteatoma matrix and the bone. He also found that infection enhances the bone-destructive capabilities of the granulation tissue.

Other host resistances to bacterial infection are anatomic barriers, antibodies, phagocytosis, and the chemical microenvironment adjacent to the organisms. Factors that may alter host resistance are hormones, systemic diseases, nutrition, and the presence or absence of competitive normal flora (Hirsch and Dubos, 1965). Although patients wiith complications from suppurative middle ear disease are usually not compromised hosts, patients with diabetes mellitus or leukemia are much more likely to incur a complication if they acquire a middle ear infection.

### *Anatomic routes of and barriers against spread of infection*

Anatomic barriers are very important to prevent spread of infection beyond the pneumatized spaces of the temporal bone and its attendant mucosa. This is particularly true in acute suppurative infections because additional barriers, such as hyperostotic bone, have not developed. Organisms tend to spread rapidly through preformed pathways, whether they originate congenitally, traumatically, or from bone destruction from previous or persistent infections. Bone is the best anatomic barrier. Blood vessels with their surrounding adventitia unfortunately compromise the osseous barriers to some degree. Osseous anatomic barriers also may be reduced by persistent or chronic infection; hence, an acute exacerbation or chronic otitis media with suppurative discharge more commonly results in a complication that does a single AOM with suppurative effusion or chronic otitis media with serous or mucinous discharge.

The three routes of spread of infection are (1) direct extension, (2) thrombophlebitis, and (3) hematogenous dissemination.

Bone-destructive lesions, whether in the mastoid or petrous region, occur by direct extension, as do all of the aural complications. Intracranial complications may occur from any the three routes. Extracranial subperiosteal abscesses, intracranial extradural abscesses, and sigmoid sinus thrombophlebitis are almost invariably from direct extension, either by direct

bone erosion or by penetration along vessels. Brain abscesses are usually a result of distant thrombosis of cerebral veins; however, direct extension can also create brain abscess. Meningitis, particularly in young children, is most often a result of hematogenous dissemination, although direct extension through perilymphatic fistulas in patients with Mondini deformity does occur. Direct extension along soft tissue planes through the petromastoid canal to the posterior fossa or along the petrosquamous suture line to the middle fossa has been implicated. Subdural effusion or abscess is more likely to occur as a separate complication of meningitis in young children than as a direct extension of a middle ear infection.

A rich network of veins exists in and around the temporal bone in which there is direct complication between extracranial, intracranial, and cranial diploic veins (Fig. 158-2). The extracranial venous system enters the sigmoid sinus via the mastoid emissary vein. The sigmoid sinus, one of the three dural venous sinuses immediately adjacent to the temporal bone (superior petrosal sinus, inferior petrosal sinus, and sigmoid sinus), receives tributaries either directly or indirectly from the supratentorial cerebral vein andthe infratentorial cerebellar veins. The diploic spaces and sinusoids within the cranium, particularly the parietal and occipital areas, communicate with these dural venous sinuses. Thrombophlebitis of any of these veins may spread to the others; hence osteomyelitis of the calvaria or brain abscess may occur at some distance from the temporal bone.

### *Drainage*

The mastoid region in an adult usually ranges from 15 to 20 cc (Silbiger, 1950). This relatively large volume must communicate with the middle ear (approximately 0.9 cc) through the aditus ad antrum and two extremely small isthmuses medial to the ossicular chain in the epitympanum (Proctor, 1964). A brisk inflammatory reaction can easily obstruct the isthmuses, the aditus ad antrum, or both, creating a potential abscess cavity of the whole mastoid region. It is important to note, however, that any single pneumatized space or group of spaces may be obstructed. This is particularly true of the large peripheral mastoid cells about the sigmoid sinus, lateral to Körner's septum and the mastoid tip cells.

## Diagnosis

Impending complications should be suspected whenever any sign or symptom of infection recurs 2 to 3 weeks after an initial episode of suppurative AOM has been treated. Frequent "recurrences" occurring regularly between 2 and 4 weeks after antibiotic therapy should be considered as a persistent infection with recurrent intermittent exacerbations rather than true de novo infections. Impending complications in chronic suppurative otitis media should be suspected when there is an acute excerbation of purulent discharge, particularly when the discharge is fetid.

Persistence of a purulent, particularly fetid, discharge despite treatment is a significant clinical sign that bone destruction is occurring. The development of vestibular symptoms in a patient with a chronic draining ear is another symptom suggestinga possible complication. Patients who have tolerated persistent drainage for long periods of time rarely develop otalgia concomitant with reduction or cessation of drainage. This usually indicates blockage of the normally patent drainage channel to the exterior and may presage a significant complication.

Facial paralysis, meningitis, subperiosteal abscess, and suppurative labyrinthitis are clinically obvious when they exist. Serous labyrinthitis slowly affecting high-frequency hearing is not clinically obvious; however, a serous labyrinthitis of reasonably sudden onset resulting in vestibular symptoms is usually clinically obvious. Chronic labyrinthitis, subdural abscesses, extradural abscesses, sigmoid sinus thrombophlebitis, brain abscesses, and otitic hydrocephalus are not obvious unless one specifically looks for them.

Fig. 158-1 shows groups of associated complications and the primary infection from which they most often arise. These associations are diagnostically useful. A patient with AOM with suppurative effusion may rapidly develop facial paralysis or meningitis, both of which would be clinically obvious. The young patient who develops meningitis subsequently may have a subdural effusion or abscess, which may not be clinically obvious and which may remain undetected unless computed tomography (CT) scanning shows the lesion.

A persistence of AOM with suppurative effusion may result in acute coalescent mastoiditis. If the acute coalescent mastoiditis then results in a subperiosteal abscess, it becomes clinically obvious. On the other hand, if it does not result in a subperiosteal abscess, sigmoid sinus thrombophlebitis, otitic hydrocephalus, and/or a bran abscess. Each may be clinically silent for variable lengths of time or, in the case of an extradural abscess or granulation tissue, until properly performed surgery uncovers the pathology.

Chronic otitis media with serous, mucinous, or suppurative discharge may over time result in serous labyrinthitis and a high-frequency sensorineural hearing loss, which can be easily overlooked. Clinically obvious suppurative labyrinthitis can result from bacterial penetration of the round window membrane or from a labyrinthine fistula in cases of chronic otitis media, cholesteatoma, or both. If, however, a labyrinthine fistula exists and bacteria or their toxins do not penetrate into the perilymph, the chronic labyrinthitis can be completely silent. The chronis presence of granulation tissue with its bone-destructive qualities can insidiously extend into the extradural space and create a noninfected, nonocclusive intraluminal mural thrombus in the sigmoid sinus, which would not be clinically obvious. An acute exacerbation of infection can infect the already-compromised extradural space and the sigmoid sinus to result in a fulminant course of sigmoid sinus thrombophlebitis with coexisting otitis hydrocephalus. This fulminant thrombophlebitis may result in a distant brain abscess, which may remain completely silent during its formation and development.

CT scanning with infusion is the single most useful test in diagnosing intracranial complications. With it, large extradural abscesses, sigmoid sinus thrombophlebitis, subdural abscesses, cerebritis, and brain abscesses can all be identified with an appropriate study. Patients without signs and symptoms of meningitis should not have a lumbar puncture. Patients with meningeal irritative signs and symptoms should have a CT scan before lumbar puncture to assess the presence or degree of hydrocephalus. The venous phase of cerebral angiography can occasionally detect a sigmoid sinus thrombophlebitis that may be unrecognized by CT scanning, but this is unusual (Venzio et al, 1982).

Investigating every chronic otitis media with CT testing is not appropriate; however, a high index of suspicion and a sensitivity to persistent acute or chronic otitis media with suppurative effusion or discharge and the presence of granulation tissue may lead to an earlier diagnosis of these complications.

## Management

Complications of suppurative middle ear disease result from two problems at the primary site: infection and poor drainage. Treatment of both the ear and the secondary site of complication involves eradication or infection (by the use or properly administered antibiotics), establishment of adequate drainage, and excision of infected tissue when necessary.

Complications resulting from AOM with suppurative effusion are usually satisfactorily managed by a myringotomy and adequate parenteral antibiotics. Complications involving bone destruction and complications resulting from chronic otitis media with mastoiditis usually require some type of mastoidectomy, parenteral antibiotics, and special management techniques specific for the complication(s) involved. The details on management of each complication are considered separately later in this chapter.

## Mastoiditis with Bone Destruction

### Pathophysiology

The mastoid is not a bone; it is one of five regions of pneumatization within the temporal bone. Each of the regions are contiguous, and when one is involved with inflammation or infection, all are variably involved (Schuknecht, 1974).

Two types of mastoiditis are associated with bone destruction: acute coalescent mastoiditis and chronic mastoiditis with osteitis. Acute coalescent mastoiditis derives its name and is diagnosed from the observation of enzymatic resorption of the thin bony septa separating air cells in well-developed mastoids where an acute infection persists. The absorption of the septa makes the air spaces appear to coalesce. Despite what may appear to be adequate antibiotic treatment, a persistent acute infection may result in an inflammatory reaction sufficient to obstruct the aditus ad antrum, epitympanum, and the lateral peripheral cells next to the sigmoid sinus.

Inadequate treatment of AOM and mastoiditis may result in a clearing oof the middle ear portion of the infection, with persistence of infection somewhere within the adjoining pneumatized spaces. This "masked mastoiditis" occurred in 15% of cases in the early antibiotic era (Smeraldi, 1947), and although uncommon, it still occurs today (Holt and Gates, 1983).

The organisms associated with acute coalescent mastoiditis are the same as for AOM with suppurative effusion. If the tympanic membrane is perforated, however, a culture of the external auditory canal discharge may fail to reveal the proper pathogens in as high as 25% of the cases (Rudberg, 1948).

### Diagnosis

Controversy exists concerning the association of subperiosteal abscesses and acute coalescent mastoiditis, and concerning the diagnosis and treatment of acute coalescent mastoiditis with or without subperiosteal abscess.

Hawkins and Dru (1983) emphasized that subperiosteal abscess was the single indication for surgery in 50 patients with acute mastoiditis. They defined mastoiditis as the presence of acute or subacute otitis media with postauricular swelling, erythema, tenderness, and a protruding ear with clouding of the ipsilateral mastoid air cells on mastoid radiographs. The presence of a subperiosteal abscess was defined by the aspiration or incision and drainage of purulent material from that locus. Of the 50 patients with "acute mastoiditis" but *without* subperiosteal abscess, 31 recovered without mastoidectomy, 2 of the 19 patients *with* subperiosteal abscess recovered with the use of antibiotics and only incision and drainage of the abscess. The other 17 of the 19 patients *with* abscess underwent mastoidectomy. One patient with subperiosteal abscess had two abscesses 2 years apart, both requiring a mastoidectomy. They emphasized that mastoid radiographs were not a factor in the decision to operate and that 31 of the 50 patients with mastoiditis and without subperiosteal abcess recovered.

In another journal 2 months later, Holt and Gates (1983) reported 9 patients with masked mastoiditis, none of whom had a subperiosteal abscess but all of whom had a significant intracranial or neural complication; the duration of otologic symptoms in these cases was from 1 week to 3 months. Opheim and Willie (1948) described a case of acute coalescent mastoiditis treated medically with resolution, and 2.5 months later the child developed meningitis. Dolowitz and Wakefield (1949) reported a case of subperiosteal abscess occurring 3 years after the suppurative AOM.

The resolution of these controversies is apparent in the literature and in my experience: Persistent infection and granulation tissue in the mastoid is capable of creating bone-destructive lesions *without* extending through the subperiosteal space. The clinical and radiographic identification of this persistent infection with ultimate bone destruction can be difficult. It is well known that greater than 50% of subperiosteal abscesses result from soft tissue and vascular extensions of infection rather than from a mastoid cortical bone defect (Hawkins and Dru, 1983); this fact is important in the problem-solving diagnostic and therapeutic decision process. Acute infections within the mastoid can cause a periosteal inflammation with resultant edema, erythema, and tenderness. This alone is not pathognomonic of an uncontrolled persistent bone-destructive lesion in the mastoid and is not an absolute indication of mastoidectomy (Bluestone and Klein, 1983b); however, the greater the obvious infection in the mastoid region, the greater the index of suspicion should be for impending complication, and the more vigorously the case should be treated.

Chronic mastoiditis with bone destruction is even harder to identify. Remembering the definition of terms is particularly helpful. Acute, subacute, or chronic otitis media and mastoiditis refer to the state of the mucosa in these pneumatized regions. The serous, mucinous, or suppurative effusion or discharge is a related but separate physical entity. As the discharge progresses from serous to mucinous to suppurative, the probabilities increase that the mucosa is being replaced by granulation tissue. As the suppuration increases in quantity or duration, the probabilities increase that the granulation tissue is irreversibly infected. As the purulent discharge becomes fetid, the probabilities increase that anaerobic organisms exist, suggesting poor aeration and the possibility of osteitis and bone destruction. Careful attention to the physical signs and history thus help to establish the presence or absence of a persistent infection with granulation tissue and the increasing probability of osteitis. The greater the duration of irreversible suppuration, the greater the probability for

complication (Sheehy et al, 1977).

Radiography is of little value because the chronic infection already has periosteal irritation and hyperosteosis, and the density of the bone makes the detection of underlying lytic areas difficult. Bone-destructive lesions of the scutum and periantral cells can sometimes be seen radiographically in cases of cholesteatoma, but this observation adds little new information to the physical examination, at least with regard to establishing the presence or absence of a cholesteatoma. Unless the bone-destructive lesion is extensive, radiographic identification of bony labyrinthine fistulas is difficult or impossible. But when bone-destructive lesions are found adjacent to the sigmoid sinus in chronic disease, they are an important finding, as they are often filled with granulation tissue and are dangerous (Neely, 1979).

## Management

Patients with acute coalescent mastoiditis and an associated subperiosteal abscess should have the abscess drained and a complete transcortical mastoidectomy performed, with attention to establishing good communication from the mastoid into the middle ear space; this frequently requires a facial-recess approach to the middle ear and a large myringotomy. When there is acute coalescent mastoiditis without a subperiosteal abscess, and a mastoidectomy is not performed, careful and repeated clinical and radiographic evaluation is indicated, until it is absolutely certain that the ear and mastoid have returned to normal. It should be remembered that a single pneumatized space or small groups of them may be obstructed and contain smoldering infection while the remainder of the middle ear and much of the mastoid, particularly medial to Körner's septum, is clear. My preference therefore is to threat every case of acute coalescent mastoiditis surgically.

Chronic mastoiditis with bone destruction is usually treated exactly like chronic mastoiditis without bone destruction unless unusually extensive osteitis and osteomyelitis exist. In that case, long-term parenteral antibiotics are used. This situation occurs with necrotizing, or "malignant", external otitis and is not discussed in this chapter. It should be noted, however, that such an extensive necrotizing process can originate in the middle ear rather than in the external ear.

## Subperiosteal Abscess

A subperiosteal abscess is a collection of pus adjacent to the mastoid that results from acute or chronic otitis media with mastoiditis and bone destruction. It occurs most commonly over the mastooid cortex at Macewen's triangle but may occasionally occur over the root of the zygoma or in the upper neck (Bezold's abscess) when it has penetrated the periosteum of the medial surface of the mastoid tip.

The exact incidence and prevalence of subperiosteal abscesses are unknown both because of their infrequency and because they are often not recorded separately from acute opalescent mastoiditis. They are much more likely to be associated with a case of acute coalescent mastoiditis than with a case of chronic otitis media and mastoiditis with bone destruction, because there are many more cases of chronic otitis media with mastoiditis and bone destruction than acute coalescent mastoiditis; but subperiosteal abscesses can be seen

with both bone-destructive processes.

Subperiosteal abscesses appear as inflamed tender fluctuant masses in the locations just listed and are usually associated with a draining ear, although previously administered antibiotics may obscure the presentation.

When overlying the mastoid, they can be mimicked by postauricular suppurative adenitis from external otitis, but a Towne's view radiograph reveals no opacification or bone destruction in this adenitis.

Pathogenesis and management are discussed in the section on mastoiditis with bone destruction.

## Petrositis with Bone Destruction

### Pathophysiology

The definition of the terms, organisms, and pathophysiology of petrositis with bone destruction are similar to those described for mastoiditis with and without bone destruction. Important distinguishing features include the following:

1. Only approximately 30% of petrous bones are pneumatized into the petrous apex (Mawson, 1974). Because of its shape and its location anteromedial to the internal auditory meatus and otic capsule, the petrous apex has the following three other distinguishing features:

2. Drainage is more restricted.

3. Proximity of the apical air cells to diploic spaces predisposes to osteomyelitis.

4. Proximity to intracranial structures and poor drainage predispose to intracranial extension.

The most common complications of petrositis with or without bone destruction are meningitis, extradural abscess, brain abscess, and suppurative labyrinthitis (Lindsay, 1938). Allam and Schuknecht (1968) emphasized that infection from the middle ear and mastoid can directly extend into a nonpneumatized petrous apex; thus petrositis with bone destruction can occur in a pneumatized or diploic petrous apex.

Kopetzky and Almour (1930)) emphasized that there is a latency period between petrositis and intracranial complications, and the symptoms are subtle. These facts were reemphasized during the antiobiotic era (Shambaugh, 1967). The pathophysiology discussed in the section on mastoiditis applies to petrositis as well.

## Diagnosis

The predominant symptom of a persistent infection deep in the petrous bone is a deep-boring pain. It is the only symptom that leads a careful observer to a diagnosis before an intracranial complication develops. Perilabyrinthine infection in the petrous bone refers pain to the occipital, parietal, or temporal area; and infection in the petrous apex refers pain to the deep retroorbital area (Shambaugh, 1967). The predominant sensory innervation of the mastoid is via the glossopharyngeal nerve (CN IX). Innervation of the middle ear is via CN XI, the trigeminal nerve (CN V), or both. Sensory innervation of the middle cranial fossa, dorsal aspect of the tentorium cerevelli, superior petrosal sinus, and transverse sinus is via CN V, with the cells of origin in the ophthalmic division of the trigeminal ganglion. Sensory innervation of the posterior fossa dura is from CN IX, the vagus nerve (CN X), and the first three cervical nerves, which reach the meninges via recurrent branches of CN X and the hypoglossal nerve (CN XII) (Hollinshead, 1968).

Continued aural discharge, even after radical mastoidectomy, is the most common sign of uncontrolled infection in the petrous bone (Shambaugh, 1967). Gradenigo (1904) described a classic triad of abducens nerve (CN VI) paralysis, retroorbital pain, and ispilateral aural discharge. This triad, known as Gradenigo's syndrome, was thought to be pathognomonic of a petrous apex abscess. In actual fact, petrous apicitis frequently exiists without the full triad; Kopetzky and Almour (1930) emphasized this fact in the preantibiotic era, and it remains true.

Radiographic evidence of petrous infection is quite difficult to establish. High-resolution CT, gallium-67 and technetium-99m bone scanning are helpful, but observation of the clinical signs and symptoms and a high index of suspicion remain the most useful tools for diagnosis.

## Management

Effective management requires removal of irreversible mucosal and bone infection, establishment of adequate drainage, and appropriate antibiotic therapy. Surgically the first course of action is usually a radical mastoidectomy and dissection along the cell tracts to the petrous regions; three of these operations have the following proper names (Mawson, 1974; Shambaugh, 1967):

1. Frechner's approach - following the subarcuate tract.

2. Thornvaldt's approach - following the supralabyrinthine portion of the perilabyrinthine tracts.

3. Lempert-Ramadier's approach - following the peritubal cells to the petrous apex between the cochlea and the carotid artery.

If infection continues or if the middle ear has responded to treatment but the apex has not, a middle fossa approach to exenterate the pneumatized spaces of the temporal bone completely may be necessary (Hendershot and Wood, 1973). For a further discussion see Chapter 169.

## Facial Paralysis

### Pathophysiology

In children, facial paralysis from infection is more likely to result from AOM and mastoiditis with suppurative effusion than from acute or chronic suppurative bone-destructive lesions or chronic infections without bone destruction. In adults facial paralysis results from infection less commonly than in children. When it does occur, the cause is more likely to be an acute exacerbation of chronic otitis media and mastoiditis with suppuration and cholesteatoma (May, 1983; Sheehy et al, 1977). It can also occur in a patient with a central perforation, chronic otitis media, and mucoid discharge caused by an occult cholesteatoma or granulation tissue (Miehlke, 1973).

Lower motor - neuron facial paralysis is clinically obvious when present, as is the presence of acute or chronic otitis media. The assumption that the facial paralysis is secondary to the inflammatory process is based on strict temporal and spatial diagnostic criteria: the inflammatory process must be on the same side as the facial paralysis, and the onset of the acute infection or exacerbation of the chronic infection must be temporally associated with the onset of the paralysis. In most instances this assumption is correct. However, the diagnosis can be more difficult in immunocompromised patients, such as those taking chemotherapeutic agents for leukemia. Accurate diagnosis depends on a strict attention to detail in following the course and resolution of the disease.

Because very few specimens have been available for histologic study, pathogenesis is based primarily on assumption. Infection may involve the facial nerve at any point in its course. The most common site is thought to be the tympanic segment of the fallopian canal just proximal to the pyramidal genu because this portion of the canal is most frequently eroded by cholesteatoma, and congenital incomplete closure of the canal in this region occurs in 57% of temporal bones. Even when the fallopian canal is eroded and the cholesteatoma matrix lies adjacent to the nerve, significant compression is usually not documented at surgery, in my experience. Furthermore, paralysis occurs in acute infections even when penetration of the fallopian canal does not exist (Shambaugh, 1967).

It is generally held that venous congestion, tissue edema, and perhaps direct neural toxicity are the major factors involved with paralysis from infection and that subacute or chronic infections are more likely to erode bone and allow nerve damage. Schuknecht (1974) presented histologic evidence of this theory by demonstrating that granulation tissue and osteitis adjacent to the facial nerve's mastoid segment resulted in an inflammatory cell infiltrate that caused distal degeneration of the nerve. Probably facial paralysis from middle ear suppuration occurs more often from acute than from chronic infections because there are many more cases of acute than chronic infections, not because each case of acute infection is more likely to cause facial paralysis than is each case of chronic otitis media.

Extrinsic neural compression secondary to edema, intracanal accumulation of granulation tissue, or a cholesteatoma is frequently difficult to differentiate from intraneural infiltration. Initially, compression or toxicity should result in a conduction block without neural destruction, whereas intraneural inflammatory invasion is more likely to cause fiber destruction. Compression or entrapment neuropathies can be so severe as to invaginate the

nodes of Ranvier at the margins of the compression area and ultimately result in segmental demyelination in the compressed area; this may proceed to a reduction of axonal membrane resistance to stretch and distortion and result in nerve degeneration (Aguayo, 1975).

A detailed study of the possible relationship between bacterial exotoxins and nontoxic bacterial metabolites, such as hyaluronidase, streptokinase, deoxyribonuclease, and coagulase, is not available in cases of facial paralysis from acute or chronic inflammation.

The epineurium offers little resistance to the spread of infection; conversely, the perineurium is a very effective barrier to the spread of local inflammation. If the perineurium is broken, infection rapidly spreads within the confines of the fascicle. Development of granulation tissue inside the fascicle may enlarge to create an intrafascicular pressure sufficient to compress the remaining fibers against the perineurium. Incision of the perineurium in such a case usually results in relief; on the other hand, incision of an intact perineurium that is acting as a barrier to infection can have quite severe results (Sunderland, 1972).

The perineurium also acts as an effective barriier to the penetration of macromolecular substances such as toxins, antigens, or even viruses. Reduced perineural permeability is most commonly the result of local trauma. Interestingly, ischemia and mediators of the inflammatory response have no appreciable effect on the perineurial permeability; the perineurium provides an effective barrier even to frank pus (Thomas and Olsson, 1975).

Increased permeability of the vasa nervorum readily occurs in respons to inflammatory diseases of infectious or allergic origin. Altered permeability in the vasa nervorum may result in an increased flow of plasma into the tissue and neural edema; regions of increased permeability show proteinaceous exudates and extravasated mononuclear cells most marked in the area of the severest lesion. Increased permeability of the vasa nervorum occurs early in the vicinity of the lesion. There is also a delayed phase of increased permeability peaking approximately 2 weeks after injury; this phase spreads to involve the distal nerve.

The effect of acute, transient ischemia on increased permeability of the vasa nervorum is graded relative to the duration of the ischemia. There is very little increased permeability after 6 hours of ischemia, but after 8 hours it begins to occur and can be quite marked after 10 hours. The epineurial vessels are much more prone to these effects than are the endoneurial vessels (Olsson, 1975).

Facial paralysis from suppurative AOM may occur suddenly or gradually, usually within the first 2 weeks of infection. The majority of these patients have partial paralysis, or paresis, and usually experience a satisfactory recovery. This recovery is not invariable and should not be taken for granted; as many as 30% of those who exhibit a complete paralysis initially have an unsatisfactory recovery (Silberman and Lewis, 1983).

Facial paralysis occurring later than 2 weeks after the acute onset of infection may be associated with a bone-destructive lesion in the mastoid such as acute coalescent mastoiditis.

Chronic suppurative otitis media may cause a sudden or gradual onset of paralysis that may involve a single branch and progress from branch to branch (Miehlke, 1973).

Lacrimal function is rarely affected; testing for lacrimal function is useful, however, for detecting unexpected proximal lesions.

## Diagnosis

Diagnostic studies focus on determining the extent of infection and the severity of neural involvement in suppurative AOM. Differential diagnosis is between edema with associated conduction block and a bone-destructive lesion with invasion of the fallopian canal by granulation tissue. The latency between the onset of the infection and the onset of the facial paralysis is helpful, but mastoid radiographs are helpful only if they demonstrate bone destruction.

Methods to determine the severity of neural dysfunction are inadequate to identify accurately the nature and degree of the pathophysiologic process causing the facial nerve weakness. Nonetheless, it is important to perform the tests serially and to plot the course of function or dysfunction over time.

Excitability threshold testing, maximal stimulation testing, electroneurography, and electromyography are all useful modalities, and using most or all of them to follow a paralysis helps to determine if the nerve is involved with only a conduction block and is responding to treatment or is worsening (see Chapter 149).

It is useful to test the main trunk near the stylomastoid foramen and each branch separately. Electrodiagnostic tests have been standardized on the totally paralyzed face; however, Silberman and Lewis (1983), in a 20-year retrospective study, determined that only 37% of patients with a facial paralysis associated with AOM presented with a complete facial paralysis. Of those, 30% had an unsatisfactory recovery. They also noted that 1 of 22 patients with partial paralysis had an unsatisfactory recovery and 2 additional cases had good but not excellent recovery. These facts emphasize the need for serial electrodiagnostic tests in the patient presenting with a partial facial paralysis, and the importance of testing each branch.

If electrical testing by the maximal stimulation test begins to show a progressively destructive lesion and this is confirmed by the threshold nerve excitability test, surgical intervention may be necessary. If the threshold nerve excitability test does not change but electroneuronography confirms a major reduction in amplitude, surgical intervention should be considered. The disappearance of voulntary action potentials on electromyography would support this decision.

Except for its prognostic value, electrical testing makes no difference in the management of facial paralysis from chronic infection.

## Management

Management is designed to resoolve the infection in the middle ear and mastoid as rapidly as possible. For patients with suppurative AOM, hospitalization; myringotomy; aspiration for smear, culture, and sensitivity; and parenteral antibiotic treatment are usually sufficient. Facial nerve function should be followed closely, perhaps daily, until recovery is evident.

Facial paralysis occurring 2 weeks or more after the onset of suppurative AOM suggests fallopian canal invasion with a bone-destructive lesion. Myringotomy and parenteral antibiotics are appropriate for initial therapy, if radiographic evidence of coalescence cannot be verified and if the clinical impression is that acute coalescent mastoiditis or a masked mastoiditis is absent.

If there is progression of the degree of injury in the suppurative AOM, or if acute coalescent mastoiditis or masked mastoiditis is diagnosed, then complete transcortical mastoidectomy with the facial-recess approach to the middle ear and a myringotomy is indicated. The fallopian canal need not be opened but may be explored by thinning the bone of the canal with a diamond burr to allow inspection of the contents through the thinned bone. If granulation tissue has entered the canal, the osseous canal should be opened and the granulation tissue carefully removed from the most external aspect of the facial nerve sheath. Extreme care should be taken not to penetrate the perineurium. If granulation tissue is seen to penetrate the perineurium, it is perhaps better to stop at that point and not open the perineurium. This situation is extremely rare.

In cases of facial paralysis occurring from chronic suppurative otitis media, immediate hospitalization, culture and sensitivity of the ear discharge, parenteral antibiotics, local cleansing, and antibiotic care are necessary. Surgical intervention should be undertaken within the next few days. A complete transcortical mastoidectomy and tympanotomy, with exenteration of the hypotympanic cells and exploration of the osseous fallopian canal from the ganglion to the stylomastoid foramen, is necessary. Again, the canal need not be entered unless tissue seems to penetrate into the canal. It is usually best to refrain from a tympanoplasty at this initial surgery, but there is no absolute contraindication.

If a cholesteatoma or granulation tissue enters the fallopian canal, the canal should beopened widely in the area of involvement. When granulation tissue invades the epineurium, it may be resected, with care taken not to penetrate the perineurium. If the nerve appears to be completely surrounded by granulation tissue, it is probably wisest to resect the majority of granulation tissue but not, initially, to resect the nerve. Resection and grafting may be necessary when infection has resolved and fibrosis has matured.

It is important to remember that a coexisting fistula of the horizontal semicircular canal is not uncommon in these cases. It should also be emphasized that adequate smears, cultures, and histologies should be obtained to keep from missing tuberculosis, syphilis, or malignant neoplasias.

## Labyrinthitis

Schuknecht (1974) defined the types of labyrinthitis resulting from acute or chronic suppurative middle ear disease by the substance or tissue that enters the perilymphatic space. Serous labyrinthitis results from toxic products entering the perilymph; suppurative labyrinthitis results from bacteria entering the perilymph; and chronic labyrinthitis results from soft tissue, such as a cholesteatoma matrix, entering the perilymphatic space.

Since the early 1970s, it has become apparent from the work of Paparella et al (1972; 1980) that serous labyrinthitis is much more common than previously recognized. It appears to be the single most common overall complication of acute or chronic middle ear suppuration. Paparella et al (1972) reviewed 344 temporal bones; 75 of these bones had evidence of middle ear inflammation with effusion, 30 had acute purulent effusion, 10 had chronic purulent effusion, and 35 had serous effusion. Control cases were divided into two groups; 31 cases had blood in the middle ear, and 238 were normal ears. They found that the gross pathologic changes in the round window membrane and further serofibrinouos precipitates and inflammatory cells in the scala tympani adjacent to the round window occurred frequently in acute and chronic purulent ears. In 82% of the acute ears and 77% of the chronic purulent ears there were pathologic changes in the scala tympani.

In a subsequent study of temporal bones with chronic otitis media, including granulation tissue, cholesteatomas, cholesterol granulomas, bony changes, and fibrosis, Meyerhoff et al (1978) emphasized the importance of the formation of granulation tissue, with or without a cholesteatoma, in the pathogenesis of complications and sequelae of otitis media. In a previous study an insidious high-frequency sensorineural hearing loss in patients with chronic otitis media was discovered in a significant number of these patients (Paparella et al, 1970). The degree of hearing loss appeared to correlate with the duration of the inflammation. In a subsequent review article the pathologic and the audiologic studies in humans were related, and experimental data in cats were correlated with the human data (Paparella et al, 1980).

These preliminary experimental data suggest that small molecules, such as bacterial toxins (for example, staphylotoxins), and enzymes rapidly diffuse through the round window in otitis media. Toxic products may also penetrate into the perilymphatic fluids through labyrinthine fistulas; hence, serous labyrinthitis may occur in a patient with chronic labyrinthitis, such as a labyrinthine fistula from cholesteatoma.

Suppuratiive labyrinthitis as a direct complication from middle ear disease is more likely to occur from chronic otitis media and mastoiditis with suppurative discharge than from acute middle ear infection, with or without bone destruction. However, the most common entry of bacteria into the inner ear is through the cerebrospinal fluid (CSF) in patients with meningitis; the meningitis may or may not have been a complication of suppurative middle ear disease. The incidence of suppurative labyrinthitis is unknown. In 96 cases of complications from acute or chronic otitis media, only 5 cases of suppurative labyrinthitis were discovered, each of these due to a labyrinthine fistula from a cholesteatoma (Juselius and Kaltiokallio, 1972). Existing suppurative labyrinthitis is clinically obvious (see Fig. 158-1).

Chronic labyrinthitis results when soft tissue enters the perilymphatic space. This event is almost always a result of a labyrinthine fistula from a cholesteatoma; however, other bone-destructive lesions, such as a gumma or carcinoma, may enter the labyrinth. By far the most common site of injury is the lateral semicircular canal. Sheehy et al (1977) found 104 patients with labyrinthine fistulas in 949 patients with cholesteatomas. Ritter (1977) found 16 cases of lateral semicircular canal fistula in 152 cases of cholesteatoma.

## Pathophysiology

Labyrinthitis is an inflammatory process of the labyrinth. The term does not specify whether the site of involvement is the endolymph, perilymph, or bony labyrinth. For symptoms to occur, the membranous labyrinth and its contents must ultimately be involved. The initially affected space is significant. Entities, such as viral infections, that initially affect the membranous labyrinth (presumably by hematogenous routes) are usually not lethal, although they may destroy all vestibular or auditory function of the involved ear. Conversely, entities that initially affect the otic capsule and extend into the periotic labyrinth (perilymphatic space), such as syphilis, tuberculosis, and other bacterial infections, may be lethal. Hence, it is important to ascertain whether the initial involvement of the labyrinthitis is from the perilymphatic system (Mawson, 1974; Schuknecht, 1974).

Only labyrinthitis associated with acute or chronic bacterial infection of the pneumatized spaces of the temporal bone is considered in this chapter. The following classification of labyrinthitis in this chapter is that of Schuknecht (1974), which is based on invading noxious agents:

1. Acute toxic (serous) labyrinthitis, in which bacterial toxins or chemical products invade the perilymph.

2. Suppurative labyrinthitis, in which bacteria invade the perilymph.

3. Chronic labyrinthitis, in which soft tissue invades the periotic labyrinth.

The cytoarchitectural changes resulting from acute toxic (serous) labyrinthitis depend on the quality and quantity of the invading noxious agent and the system involved. The auditory system tends to be more severely involved than the vestibular system is (Schuknecht, 1974). The progression of disease is as follows:

1. The round window membrane is infiltrated by acute and/or chronic inflammatory cells.

2. The perilymph of the scala tympani develops a fine serofibrinous precipitate just medial to the round window membrane. This precipitate may spread longitudinally up the scala tympani through the helicotrema to the scala vestibuli.

3. Acute and/or chronic inflammatory cells may penetrate the round window membrane and follow the precipitate.

4. Toxins and/or inflammatory cells may penetrate the basilar membrane from the scala tympani, invade the endolymph at the basal turn, and progress toward the apex.

5. Neuroepithelial degeneration may follow, beginning with the derangement of the smooth endoplasmic reticulum and mitochondria and progressing to total cellular destruction. In the cochlea the outer hair cells are most vulnerable; those of the outer hair cells are most vulnerable; those of the basal turn are affected first. The stria vascularis is reasonably resistant. The cytoarchitectural changes are similar to those from ototoxic drugs and tend to

support the hypothesis that bacterial toxins, rather than bacteria, are involved.

6. Chemical alterations may also occur so that lactic and malic dehydrogenase are increased in the perilymph (Paparella et al, 1972).

It should be emphasized that the locus of entry determines the initial area affected and thus the presenting symptoms.

When considering inflammation of the labyrinth associated with infections of the pneumatized spaces of the temporal bone, perilabyrinthitis may be tympanogenic or meningogenic. The term *tympanogenic* implies that the noxious agent enters the perilymph directly from the pneumatized spaces of the temporal bone. *Meningogenic* implies that the noxious agents enter from the meninges and CSF. An example of tympanogenic labyrinthitis is the serous labyrinthitis from window membrane penetration in cases of chronic suppurative otitis media. An example of the meningogenic labyrinthitis associated with bacterial ear infection is acute suppurative labyrinthitis secondary to a middle ear infection immediately following a stapedial fracture from head trauma, and the subsequent development of bacterial meningitis from bacteria migrating through the perilymph into the CSF, and ultimately a bacterial labyrinthitis of the opposite ear as a result of the spread of bacteria from the meninges into the opposite labyrinth. This actual case is an example of both the tympanogenic and the meningogenic origin of labyrinthitis from ear infections. Meningogenic suppurative labyrinthitis is usually bilateral, in contrast to tympanogenic suppurative labyrinthitis, which is almost always unilateral.

The spread of infection from the labyrinth to the CSF occurs along (1) the vestibular nerve and (2) the cochlear nerve (Benitez, 1977; Matz et al, 1968). When one considers only the topic of acute or chronic suppurative ear disease, the most common cause of suppurative labyrinthitis is a horizontal semicircular canal fistula from a cholesteatoma (Paparella and Sugiura, 1967). Fulminating and rapidly fatal infections, however, have resulted from suppurative AOM in a post-stapedectomy ear. My primary experience with acute suppurative labyrinthitis, which invariably results in meningitis, is from acute middle ear infections in cases of traumatic stapes footplate fracture or congenital oval window and vestibular dehiscences associated with Mondini malformations (Neely, 1985).

Endolymphatic hydrops is seen early in serous labyrinthitis and suppurative labyrinthitis (Paparella and Sugiura, 1967; Schuknecht, 1974). The basilar membrane is vulnerable to penetration by leukocytes and bacteria, which increase the protein content in the endolymph and allow destruction of the organ of Corti. Conversely, Reissner's membrane is resistant to infection, and its presence allows development of a differential osmotic pressure gradient across it, thus resulting in the hydrops.

Chronic labyrinthitis occurs when soft tissue enters the perilymphatic space, most commonly as a result of osteitis or cholesteatoma bone resorption. More than 75% of fistulas are in the horizontal semicircular canal; however, the superior or posterior semicircular canals, vestibule, cochlea, or complete vestibular labyrinth may be involved with the fistula (McCabe, 1978). The soft tissue entering the labyrinth may be squamous epithelium, fibrous tissue, granulation tissue, or all three. If the tissue remains predominantly extra-luminal with an intact endosteum beneath the fistula, the patient may have few or no symptoms or have

symptoms only when the fistula is manipulated. Hence, a fistula may exist without any vestibular symptoms. This situation, however, occurs in only about 10% of the fistula cases (McCabe, 1978). Labyrinthine fistulas are usually covered with a protective cholesteatoma matrix. The development of a serous or suppurative labyrinthitis depends on a breakdown in this protective epithelial barrier.

A single fistula can be very small and shallow or quite large and deep; in fact, the complete vestibular labyrinth can be destroyed and replaced with a cholesteatoma. Interestingly, in such cases the cochlear reserve, measured by bone-conduction audiometry, is good to excellent. Bumsted et al (1977) found four of these cases. I have had experience with only one such case, in which the cholesteatoma additionally expanded laterally from the vestibule and compressed the tympanic segment of the facial nerve. A lower motor-neuron facial paralysis, the patient's presenting symptom, occurred in a radical mastoidectomy cavity, which was performed many year previously.

Labyrinthine sclerosis is the replacement of all or part of the labyrinth with fibrous or osseous tissue and is sometimes called *labyrinthitis ossificans.* Labyrinthine sclerosis represents a healing process, which most commonly follows suppurative labyrinthitis. The origin of the new intralabyrinthine bone is theoretically from (1) the endosteum, (2) metaplasia of connective tissue, (3) granulation tissue, or (4) undifferentiated perivascular mesenchymal cells (Schuknecht, 1974). There is reasonably good supportive evidence that the bone originates from the endosteum.

## Diagnosis

The diagnosis of labyrinthitis from acute or chronic ear infection is based on signs and/or symptoms of inner ear dysfunction. Most commonly, serous or toxic labyrinthitis results in a slowly progressive, insidious, high-frequency sensorineural hearing loss, of which the patient may not be aware. A more rapid onset of serous labyrinthitis results in a partial or complete sensorineural hearing loss and vestibular symptoms that may range from a feeling of unsteadiness, to constant or intermittent positional vertigo, to episodes of spontaneous vertigo suggesting endolymphatic hydrops.

Suppurative labyrinthitis is much more fulminant in its onset in both the auditory and vestibular systems. However, there are no effective ways to differentiate prospectively severe toxic labyrinthitis from suppurative labyrinthitis (Schuknecht, 1974). Any patient with an acute or chronic ear infection who presents with a sudden or rapidly progressive sensorineural hearing loss and significant vertigo should be hospitalized and treated immediately, with the concern that suppurative labyrinthitis is present or imminent and that meningitis is soon to follow. It should be noted that fever is not a symptom of suppurative labyrinthitis (Shambaugh, 1967). The existence of fever in a patient having a sudden onset of labyrinthine dysfunction should immediately suggest the possibility of meningitis.

The signs and symptoms associated with chronic labyrinthitis are quite variable. Fistulas tend to become symptomatic if they are manipulated or if there is local or diffuse serous labyrinthitis. Even though 90% of patients with labyrinthine fistulas have vestibular symptoms, missing these symptoms is very easy unless one asks the right questions. In every patient with a cholesteatoma, a careful check for a history of labyrinthine symptoms is

important, and the performance of a fistula test is essential.

McCabe (1978) emphasized that the technique and the interpretation of the fistula test are not well recorded in the literature; because of this, authors have reported that only 40% of patients with labyrinthine fistulas have a positive fistula test. Even with the attachment of electronystagmography leads to enhance the interpretation of the fistula test, Shirabi (1975) found atypical results in 50% of the cases. Conversely, McCabe reports 72% positive fistula tests in patients with labyrinthine fistulas. His results suggest that a truly positive fistula test is infrequently false positive; however, a negative fistula test is often false negative.

The fistula test McCabe described in detail is as follows: With a pneumatic otoscope maintaining a good seal in the external auditory canal, positive pressure is applied for 5 seconds; then, pulsed positive pressures are delivered. The patient is given an opportunity to rest for a moment and the negative pressure is applied for 5 seconds. This is again followed by pulsed negative pressures. The eyes are observed, preferably by a second observer, for *conjugate deviation* of the eyes when pulsed positive or negative pressure is applied. The eyes are also observed for *nystagmus* when the pressure is maintained.

It is emphasized that by definition the criterior for a positive test is conjugate deviation of the eyes. When positive pressure is applied, the eyes conjugately deviate to the opposite side. If the pressure is held for 5 seconds, nystagmus occurs and the fast component is toward the involved ear. This is true if the fistula is in the horizontal semicircular canal and suggests that positive pressure deflects the cupula toward the utricle. The utriculopedal deflection results in deviation of the eyes away from the stimulating canal, and the fast component of nystagmus is to the stimulating canal. McCabe made a point that some patients may not respond to positive pressure but do respond to negative pressure; a response with conjugate deviation of the eyes to positive or negative pressure is considered a positive fistula test. Fistulas in the vertical canals, vestibule, or cochlea may result in deviation of the eyes in directions different from those described relative to the horizontal canal.

## Management

Management of labyrinthitis is first directed toward control of the middle ear, mastoid, and, if present, meningeal infection. Surgical intervention may also become necessary to eradicate the middle ear and mastoid infection. Draining the labyrinth is rarely necessary.

Any patient with acute or chronic suppurative ear disease resulting in sudden or rapidly progressive inner ear symptoms should be hospitalized immediately and treated vigorously. If there are any signs or symptoms of meningitis, a lumbar puncture should be performed after ascertaining that increased intracranial pressure does not exist.

Suppurative AOM complicated by labyrinthitis should be treated with a myringotomy and parenteral antibiotics. If the hearing loss remains stable, the vertigo improves, the organism is determined to be sensitive to the antibiotics, and the infection in the middle ear is clearing, then the condition may continue to be treated much like an uncomplicated suppurative AOM. If, however, the hearing loss is total and the vertigo severe, suppurative labyrinthitis should be considered to be present and meningitis imminent. In this case, the condition should be treated like meningitis. Only retrospectively can it be determined whether

suppurative labyrinthitis had indeed occurred. Serous labyrinthitis can result in a permanent profound hearing loss but usually does not; suppurative labyrinthitis always does. Mastoidectomy is not necessary unless masked mastoiditis or acute coalescent mastoiditis exists.

The patient presenting with vertigo, nystagmus, and sensorineural hearing loss with chronic suppurative otitis media, with or without a cholesteatoma, should be hospitalized and treated with local cleansing and topical antibiotic solutions to the affected ear as well as parenteral antibiotics capable of penetrating the blood-brain barrier. Appropriate surgery for the chronic ear disease should be instituted within several days of admission.

Two difficult and controversial treatment problems are (1) when to perform a labyrinthectomy in cases of suppurative labyrinthitis, and (2) how to manage a labyrinthine fistula.

Much available clinical information suggests that meningogenic suppurative labyrinthitis is satisfactorily treated when the meningitis is satisfactorily treated. There is very little evidence, one way or another, to suggest whether tympanogenic suppurative labyrinthitis itself is adequately controlled when the treatment is sufficient to control the meningitis resulting from it. A labyrinthectomy might be indicated if meningitis recurred despite vigorous medical treatment, or if it failed to respond. However, my experience is that acute or chronic suppurative ear disease associated with resistant or recurrent meningitis is more likely to result from unrecognized posterior fossa epidural abscesses with dural perforation, or from congenital direct communications with the CSF. In operated cases with Mondini malformations, suppurative labyrinthitis, and recurrent meningitis, no intralabyrinthine abscess was found (Neely, 1985). In chronic suppurative otitis media with a cholesteatoma completely obliterating the vestibular labyrinth, a complete vestibular labyrinthectomy can and should be performed; frequently hearing can be maintained (McCabe, 1978).

The management of the labyrinthine fistula is perhaps less controversial than it would appear at first view. It is clear that the removal of disease from a fistula involves risk; it is also known that allowing disease to remain on the fistula involves risk. A logical, graded approach to the problem lessens the controversy. The following facts help in the decision process:

A. Evidence of serous or suppurative labyrinthitis in chronic otitis media suggests a break in the protective epithelium and perhaps the endosteum over the fistula. It also indicates the presence of an abnormal and sometimes unusually sensitive inner ear.

B. A pressure-sensitive fistula suggests that the soft tissue is reasonably intimately involved with, and may well be adherent to, the membranous labyrinth.

C. The closer a fistula is to the auditory system, the more devastating it can be to the inner ear. Hence, fistulas over the vestibule or cochlea probably should not have the disease removed from them.

D. Large or deep fistulas, identified by cleaning the debris from the cholesteatoma and observing the fistula through the intact matrix, should not be disturbed.

E. Fistulas that occur over the semicircular canals and that are not large and deep may be approached in one of three ways:

1. The cholesteatoma matrix is carefully removed at the time of chonic ear surgery, with caution taken not to penetrate the endosteum. If the endosteum appears to have been violated by the disease, the procedure is immediately stopped.

2. The matrix over the fistula is left, and the fistula is exteriorized by construction of a cavity (Guilford, 1969).

3. The matrix is left over the fistula, and an intact canal wall procedure is performed with the intention of reentering the ear in 6 months to remove the cholesteatoma in a "sterile" environment (Sheehy, 1985).

The delicate technique required to evaluate the depth and extent of otic capsule erosion properly is gained over many years of experience. It should be emphasized, however, that several hours in the laboratory dissecting the otic capsule and advancing the dissection toward the periotic labyrinth can equal years of experience. These types of dissections are mandatory for anybody doing chronic ear surgery. It is as necessary to know how deep the dissection or disease has gone into the otic capsule as it is to know the detailed anatomy of the mastoid and middle ear. All of the reports on the proper handling of fistulas cannot replace a personal ability to comprehend the anatomy.

## Extradural Abscess

### Pathophysiology

Infection in the extradural space is manifest by pus, granulation tissue, or infected effusion on the lateral surface of the dura just medial to the bone. These collections are frequently asymptomatic and are discovered incidentally during mastoidectomies for acute or chronic disease. In my experience this complication occurs far more frequently than is reported. Holt and Gates (1983) reported three epidural collections in nine cases of masked mastoiditis presenting with vague or nonclassic symptoms from the mastoid disease. Significant aural and intracranial complications were present on admission in seven of the patients, and in two others an unsuspected epidural abscess was found during surgery. The third epidural abscess was found in one of the patients with meningitis. Of 152 patients with cholesteatoma Ritter (1977) found 12 patients with pathologic dual exposure, the forerunner of epidural abscess. Epidural collections are usually in direct continuity with the suppurative process in the pneumatized spaces; but in chronically infected ears, hyperostotic bone sometimes completely obscures granulation tissue on the dura. If the dura is not explored through thin bone, particularly over the posterior fossa and sigmoid sinus, such accumulations cannot be discovered. Although large amounts of pus may accumulate deep to a bony dehiscence, granulation tissue is much more common. The cause, prognosis, and treatment are the same.

Extradural abscesses or granulation tissue may result from acute coalescent mastoiditis, subacute masked mastoiditis, or chronic otitis media and mastoiditis with suppurative discharge and bone destruction. Sigmoid sinus thrombophlebitis, otitic hydrocephalus, and, rarely, brain abscess are complications that can coexist with or result from extradural infection (see Fig. 154-2).

The dura of the sigmoid sinus or the posterior fossa just posterior to the otic capsule is the site of pathologic dural exposure and granulation tissue most commonly encountered in acute coalescent mastoiditis, masked mastoiditis, and chronic suppurative otitis media without cholesteatoma. But in chronic suppurative otitis media with cholesteatoma the dural involvement is most frequently at the tegmen of the tympanum or mastoid.

It is worth emphasizing that a single acute exacerbation of infection may convert an otherwise silent extradural granulation into a more severe intracranial complication.

## Diagnosis

The diagnosis of extradural granulation tissue and/or abscess is confounded by two factors: They are usually asymptomatic, and a careful intraoperative exploration of the dura through thin bone is not frequently made. Referred or deep local pain or headache with a low-grade fever occasionally may result from an extradural infection and provide a clue to the diagnosis.

The diagnosis of an extradural abscess may be possible by magnetic resonance imaging (MRI); however, failure to identify an abscess my MRI does not mean a small abscess and/or granulation tissue is not present. In large extradural abscesses, a hypointense rim, suggesting displaced dura, may be seen between the abscess and the brain, the abscess being mildly hyyperintense compared to CSF on short TR pulse sequences (Weingarter et al, 1989).

## Management

If extradural granulation tissue is discovered, the bone is removed over the granulation tissue and adjacent dura until normal dura is found circumferentially. The granulationtissue is then removed carefully from the dura with blunt dissection. If an accumulation of pus is found, particular attention is necessary to remove all of the bone lateral to the purulent material. This sometimes requires removing the entire posterior fossa bony plate from an anteromedial margin at the posterior semiciruclar and fallopian canals, to a posterolateral margin 1 or 2 cm behind the sigmoid sinus. Occasionally there is a perforation in the dura allowing the puss to communicate with the CSF; this additional condition cannot be identified without careful inspection of the entire abscess cavity. In my experience, this kind of communication has always been associated with meningitis.

A large accumulation of epidural pus can be missed even after a sculptured mastoid cavity has been constructed, if the surgeon does not make the bone over the sigmoid sinus and posterior fossa plate very, very thin and fails to inspect the dura through the thin bone carefully.

## Sigmoid Sinus Thrombophlebitis

Seid and Sellars (1973) found 13 patients with lateral sinus thrombophlebitis from ear disease in a period of 3 years; 70% had chronic infection. However, in Gower and McGuirt's series (1983), four of five patients with lateral sinus thrombophlebitis had acute oor subacute infection. Ritter (1977) reviewed 152 cholesteatoma cases and found sigmoid sinus thrombosis in only two.

## Pathophysiology

Of the three dural sinuses intimately associated withthe temporal bone (sigmoid, superior petrosal, and inferior petrosal0, the sigmoid is most commonly affected by acute or chronic temporal bone infection. These sinuses are intradural structures with one portion of the circumference adjacent to the arachnoid membrane in the lateral recess of the pontine cistern and the other portion adjacent to a sulcus in the temporalbone. Perisinus inflammation in the extradural space adjacent to the sigmoid sinus results in a localized phlebitis, which promotes formation of a mural thrombus. The mural thrombus enlarges intraluminally and may occlude the lumen and become inflamed or infected (Mawson, 1974; Neely, 1979; Seid and Sellars, 1973). Whether or not infected, the thrombus may propagate in either directiion and become organized. If infected, it becomes necrotic and releases septic emboli, causing septicemia and high-spiking fevers once or twice daily

## Diagnosis

The signs and symptoms associated with sigmoid sinus thrombophlebitis result from inflammation and altered intracranial hydrodynamics.

The classic symptoms (Mawson, 1974) are headache, general malaise, spiking fever, chills, increased intracranial pressure, and Griesinger's sign (postauricular edema secondary to thrombosis of the mastoid emissary vein). Griesinger's sign is described as edema over the mastoid process, but the mastoid emissary vein joins the posterior aspect of the sigmoid sinus posterior to most of the mastoid pneumatization. The edema of Griesinger's sign should not be confused with subperiosteal edema or abscess from acute coalescent mastoiditis; it should be centered more posteriorly.

Headache, irritability, lethargy, and frequently papilledema (often referred to as otitic hydrocephalus) are seen as a result of increased intracranial pressure. Unrelenting spiking fever and chills are a result of uncontrolled intrasinus suppuration or severe cellulitis of the sinus wall resuling in bacteremia. Extension of the thrombophlebitis process posteriorly as far as the torcular herophili rarely occurs; extension along the petrosal sinuses to the cavernouos sinus or into the jugular bulb and internal jugular and subclavian veins is rare (Surkin et al, 1983).

The classic signs and symptoms of sigmoid sinus thrombophlebitis can be significantly altered by antibiotic treatment. Spiking fever and septicemia are frequently absent (Juselius and Kaltiokallio, 1972). Sigmoid sinus thrombophlebitis may be asymptomatic. The earliest symptoms are mild headache, general malaise, and low-grade fever in a patient with acute, subacute, or chronic ear infection.

In cases of sigmoid sinus thrombophlebitis, an extradural abscess is expected, otitic hydrocephalus is frequent, and a brain abscess is occasionally present concomitantly or may soon develop (Venezio et al, 1982).

The Queckenstedt-Stookey and Tobey-Ayer tests performed during lumbar puncture in hopes of identifying a thrombosed sigmoid sinus are not only dangerous but unreliable (Juselius and Kaltiokallio, 1972; Seid and Sellars, 1973).

Today, sigmoid sinus thrombosis, if occluding, may be reliably diagnosed by MRI, which demonstrates an intense signal within the sinus on all planes and all pulse sequences. Normally, the patent sinus is markedly hypodense (Mas et al, 1990) (Nadel et al, 1990) (Fig. 158-3).

## Management

The treatment of sigmoid sinus thrombophlebitis is reasonably straightforward. It is designed to control the infection, not to reestablish blood flow through the sigmoid sinus.

Sufficient doses of the appropriate antibiotic given parenterally, mastoidectomy of the appropriate type relative to the disease process, and exploration of the dura of the sigmoid sinus are mandatory.

Granulation tissue found on the sigmoid sinus should be removed carefully, with blunt dissection and enough bone removed to expose normal dura. If the sinus is found to be normal in color and texture and is easily compressible, nothing else is done.

If, on the other hand, the sinus appears to be abnormal within its lumen, a 20-gauge needle is carefully inserted into the sinus, with extreme care taken not to penetrate its medial wall. If blood is encountered, the operation is complete; if not, the sinus should be opened through a very small incision. Usually a sickle knife is sufficient to make this incision. If blood is encountered, the hole should be occluded immediately with the fingertip, and a piece of Surgicel should be held against the small incision with a cottonoid to stop the hemorrhage. The Surgicel piece can be left in place. I have seen one case of septicemia resuling from Surgicel being placed into the lumen of the sinus that until then was patent. In cases like these, the Surgicel piece should not be placed intraluminally but may be left in place extraluminally when the wound is closed.

If a fibrotic mural thrombus is encountered, there is no need to dissect further (Fig. 158-4). If an abscess or an obviously infected thrombus is encountered, it should be suctioned from the sinus (Seid and Sellars, 19730. These surgical measures and appropriate antibiotic therapy are sufficient to treat the problem.

In the extremely rare case in which septic emboli occur, ligation of internal jugular veins may be necessary (Seid and Sellars, 1973), and intravenous heparinization should be used. An extraordinary case reported by Surkin et al (1983) showed progression of the thrombosis and thrombophlebitis to include the subclavian vein even after internal jugular vein ligation. Appropriate antibiotics and intravenous heparin resolved this clinical problem.

## Subdural Abscess

Gower and McGuirt (1983) described eight pediatric patients who developed a subdural fluid collection - three instances of empyema and five of effusions. Seven of the eight patients were younger than 2 years of age; the other was 6. Seven had acute suppurative otitis media, and one had acute coalescent mastoiditis. *H. influenzae,* type B or nontyped, was cultured from three of five effusions and two of the three empyemas.

## Pathophysiology

A subdural collection may be an abscess, empyema, and/or an effusion. A subdural abscess is a circumscribed collection of pus that is walled off from the remaining subdural spaces. A subdural empyema is purulence that has spread over a large surface area, usually over the convexity of the cerebrum. A subdural effusion is a localized or diffuse fluid collection that is not frankly purulent to gross inspection, although it may be infected (Courville, 1944).

The subdural space is the actual or potential space lined with a one-cell layer of flattened mesothelial cells between the deepest aspect of the dura and the most superficial aspect of the arachnoid. Deep to the arachnoid is the CSF compartment. At the base of the brain and in the posterior fossa the cranial nerves and vessels tether the arachnoid tightly to the dura; the subdural space is only a potential space and is rarely involved with an effusion, empyema, or an abscess. The basilar surface of the frontal lobes and especially the temporal lobes are also closely approximated to the dura and rarely involved with subdural fluid collection (Courviille, 1944). But the subdural space over the convexity of the cerebral hemispheres is a real (rather than potential) space without anatomic interruption. It extends between the two cerebral hemispheres adjacent to the falx cerebri.

Farmer and Wise (1973) reported an important difference in the pathophysiology and the etiologic organisms in infants compared to those of children and adults. In infants, subdural empyema was invariably secondary to meningitis, and the organisms encountered were *H. influenzae, S. pneumoniae,* and *Paracolon escherichia.* Conversely, in children and adults the overwhelming majority were from frontal sinus infections, and only rarely from middle ear or other sites (Farmer and Wise, 1973; Khan and Greibel, 1984). The organisms found in these cases were frequently hemolytic and anaerobic *Streptococci* and also *Staphylococcus aureus* (Khan and Greibel, 1984).

In both children and adults the infection reaches the subdural space through adjacent venous channels. Extension of inflammation and cellulitis to the dura results in thrombophlebitis of venous channels adjacent to the subdural space. The spread of infection along these venous channels iis capable of generating subdural empyema and concomitant subcortical and cortical abscesses, which are not infrequently associated with subdural effusion, particularly over the convexity of the cerebrum (Courville, 1944; Courville and Nielsen, 1934).

Because of the rarity of the association of ear infection with subdural empyema in children and adults, the pathogenesis has not been as well established as it has been for meningitis in infancy or for subdural empyema secondary to frontal sinus infection.

## Diagnosis

The signs and symptoms associated with subdural empyema are stupor or coma, hemiparesis, convulsions, fever, and often a stiff neck. Older children and adults also tend to complain of headache and are twice as likely as infants to have papilledema, although papilledema may exist in only 35% of all cases (Farmer and Wise, 1973). These cases progress very rapidly to coma and death unless treated vigorously and early. Many of the infants in the Gower and McGuirt series (1983) presented with symptoms of meningitis, but they tended to fail to respond to treatment as expected or had focal motor seizures, leading to the suspicion of a subdural effusion or empyema.

MRI is currently the most sensitive and reliable test to diagnose subdural abscesses. These abscesses appear as mildly hyperintense lesions when compared to CSF on short TR pulse sequences and are distributed over the surface of the brain or between the hemispheres (Weingarten et al, 1989). CT scanning with infusion is the next most useful diagnostic tool if MRI is not available. It usually shows the degree and extent of cerebral edema in dural fluid collection. The subdural collection is usually crescent-shaped and of decreased density, with a rimlike crescent of increased density adjacent to the brain parenchyma (Goodman, 1982). If CT scanning is not available, the next most valuable tool is cerebral angiography.

Subdural effusions in infancy are usually suspected clinically and confirmed by MRI or CT, followed by a subdural tap.

## Management

When empyema exists, the basiic principles for treatment involve immediate measures to reduce intracranial pressure and to evacuate the empyema. A craniotomy or multiple burr holes are usually necessary to remove the purulent effusion, which can be quite thick and not easily aspirated through a single burr hole. Intraoperative irrigation with bacitracin, neomycin, and polymyxin and post-operative irrigation through drains that are slowly withdrawn over the course of a week complete the treatment (Goodman, 1982). Then treatment of the suppurative ear disease can be undertaken.

The mortality and significant morbidity rate is as high as 20% to 40% unless diagnosed and treated early (Dodge, 1981). With early diagnosis and treatment, recovery can be quite good, as Khan and Greibel (1984) illustrated. Of their 15 patients, 11 recovered with minimal or no neurologic deficits, 2 patients had major permanent neurologic residual problems, and 2 patients died. In the 13 survivors, 6 had focal or generalized seizures for the first 6 months after recovery. One of these required a cortical cicatrix excision 2 years later to control seizures.

Subdural effusions in infancy can usually be successfully managed by repeated subdural taps and appropriate antibiotics.

It should be noted in passing that "benign" subdural collections of fluid in infancy have been reported. Robertson et al (1979) found six infants with subdural collections of fluid of unknown origin. These patients, except for having large heads, were otherwise asymptomatic and during a brief follow-up pperiod seemed to develop quite normally with

no neurologic sequelae.

## Meningitis

### Pathophysiology

Bacterial meningitis is an inflammation of the meninges and CSF from a bacterial infection. The signs and symptoms associated with meningitis result from irritation of the meninges.

Approximately 90% of the reported cases of bacterial meningitis occur in patients between 1 month and 5 years of age, and approximately 86% of that same age group have at least one episode of suppurative AOM (Dodge, 1981). The frequent association between suppurative AOM, pneumonia, and meningitis suggests that all three diseases may relate to a systemic bacterial infection, with its port of entry in the upper respiratory tract. Clinical and experimental data show that meningitis, even in patients with suppurative AOM, follows bacteremia, and hematogenous dissemination plays a predominant role in bacterial meningitis from suppurative AOM. This is supported by a temporal bone histopathology study from the preantibiotic era (Lindsay, 1938), which failed to disclose the route of spread in 6 of 15 cases of meningitis associated with suppurative AOM.

The next most common route of spread of acute infection in infants and children is through congenitally preformed tissue planes, such as into the posterior fossa along the subarcuate artery in the petromastoid canal, and into the middle fossa along the soft tissue of the petrosquamous suture line. The least common route of spread of acute infection is directly through the inner ear through labyrinthine or stapedial fracture or through congenital dehiscences in the oval or round window.

Chronic suppurative otitis media results in meningitis by direct extension through bone and dura or through the inner ear via a labyrinthine fistula caused by a cholesteatoma.

Bacteria causing the meningitis are similar to those causing the ear infection. There are, however, variances to that general rule. First, the percentage of otogenic *H. influenzae* meningitis is significantly greater than the percentage of *H. influenzae* type B isolates from random middle ear effusions. Of pediatric patients with tympanogenic meningitis 71% had *H. influenzae* cultured from the CSF; and all of the five patients with subdural effusion and positive cultures were also found to have *H. influenzae* in the effusion (Gower and McGuirt, 1983). Based on this information, Gower and McGuirt felt that *H. influenzae* is a much more aggressive pathogen and offers a greater risk of intracranial complications. The other variance to the rule is that three or more microorganisms exist in most chronic ear infections with a cholesteatoma (Harker and Koontz, 1977). One or all of these organisms may ultimately penetrate through the meninges; hence, precise identification of the organism involved in the meningitis, particularly chronic ear infections, is important.

## Diagnosis

Bacterial meningitis has three clinical signs, each of which has a characteristic CSF findings. These clinical stages are the following:

1. The *stage of onset.* This stage is indicated by meningism and increased CSF production, frequently with an increase in intracranial pressure. This state is also known as CSF *serous state.*

2. The *stage of established disease.* This stage is characterized by obvious signs of meningitis, such as fever, headache, meningism, lethargy or irritability, and a marked change of CSF pressure and content. In this stage the protein increases and the glucose decreases. Inflammatory cells also accumulate in the CSF *cellular stage.*

3. The *stage of paralysis.* In this stage the classic signs of meningitis are fulminate and lethargy is predominant. Bacteria are seen in the CSF. This stage is also known as CSF *bacterial stage.*

Headache, fever, neck rigidity, and abnormal reflexes strengthen the clinical impression. One abnormal reflex is Kernig's sign, in which the leg cannot be completely extended when the thigh is flexed on the abdomen. Another abnormal refllex is Brudzinski's sign, in which there is an involuntary flexion of the ankles, knees, and hips when the neck is flexed. Brudzinski's "contralateral sign" is the involuntary flexion of the opposite lower limb when the other limb is passively flexed by an examiner.

Meningitis may precede subdural collections of pus or multiple brain abscesses. if it occurs several weeks after the onset of suppurative AOM or in chronic suppurative otitis media, a bone-destructive lesion is probable; extradural abscess, sigmois sinus thrombophlebitis, otitic hydrocephalus, and a single brain abscess are more probable, in that order. If chronic labyrinthitis is present in an ear containing a cholesteatoma, and that ear becomes profoundly deafened, unilateral suppurative labyrinthitis on that side may be the source of the meningitis.

The most powerful diagnostic tools in these situations are a detailed history, a careful physical examination, CT scanning with infusion, and a lumbar puncture with analysis of the CSF.

In addition to the standard study of CSF for cells, bacteria, glucose, protein, and chloride, analysis of the fluid sample for lactate, lysozyme, C-reactive protein, and serum amyloid-A protein may be particularly helpful to diagnose bacterial meningitis in the early stages. Ponka et al (1983) found that the CSF lactate levels are frequently elevated in bacterial meningitis. The lactate levels above 3.0 micromol/L had a sensitivity, specificity, and positive and negative predictive value of over 90%. The lysozyme levels, C-reactive protein, and serum amyloid-A protein concentration are much less sensitive tests; however, when these tests are positive, the positive predictive value is quite high - greater than that of the lactate.

Recurrent meningitis from middle ear suppurative disease usually involves a CSF fistula, and most commonly *S. pneumoniae* is the responsible organism. A rare cause of recurrent meningitis from *Neisseria meningitidis* is a genetically transmitted defect in complement activation.

## Management

Management of meningitis consists of the appropriate administration of intravenous antibiotics and periodic monitoring of the progress by lumbar puncture.

Surgical intervention is limited to the rapid eradication oof the ear infection and any additional intracranial complications requiring surgery. an immediate myringotomy with suction evacuation of purulent material for smear, culture, and sensitivity is required in cases of suppurative AOM associated with meningitis. Evacuation of more than 0.9 mL of pus through the myringotomy is presumptive evidence of adequate drainage from the mastoid through the middle ear.

If acute coalescent mastoiditis is identified, immediate drainage of any subperiosteal abscess and a myringotomy with culture and sensitivity is necessary. As soon as the patient can withstand surgery, a complete transcortical mastoidectomy with facial-recess approach to the middle ear and a large myringotomy should be performed.

A mastoidectomy should also be considered if acute coalescent mastoiditis cannot clearly be diagnosed but the mastoid radiograpphy fails to clear completely, particularly in the large peripheral cells around the sigmoid sinus.

Chronic suppurative otitis media associated with meningitis with or without obvious bone destruction, with or without cholesteatoma, requires immediate local care to the infected ear and a mastoidectomy as soon as the meningitis is stable and the patient can withstand the surgery. The surgical technique and associated procedures depend on the pathology that is found. A search for fistulas and a very careful inspection of the dura of the middle and posterior fossa through thin bone is absolutely necessary. To be certain that there is no epidural focus of infection, it is often necessary to remove some of the bone and inspect the dura directly.

When AOM has caused a suppurative labyrinthitis that results in meningitis, the entire process is likely to respond to antibiotics. In the more common situation, in which chronic otitis media and mastoiditis result first in suppurative labyrinthitis and then in meningitis, stabilization of the meningitis is followed by a mastoidectomy, which includes opening of the semicircular canals and labyrinth to provide adequate drainage. When meningitis results from suppuratiive AOM and the bacteria gain entrance to the CSF space through an oval or round window membrane fistula, that perilymphatic fistula should be surgically repaired when the meningitis has resolved.

Standard therapy for most cases of acute bacteria meningitis is ampicillin, 200 to 400 mg/kkg/day, plus chloramphenicol, 100 mg/kg/day, in four divided doses, given in 20- to 30-minute IV infusions for at least 5 days. If the patient is eating well by then, the dosage may be taken orally. The treatment duration depends on the organisms: For *N. meningitidis,*

treatment is 7 days; for *H. influenzae* it is 10 days; for streptococci it is 14 days; for all other organisms 21 days of treatment is usually necessary (Steele and Bradsher, 1983)0. Other, new antibiotics, such as ceftriaxone, may be as or more effective. This is particularly true with the escalating numbers of ampicillin- and cloramphenicol-resistant *H. influenzae* organisms.

Monitoring the CSF is extremely important to ensure the adequacy of antibiotic therapy. By convention, lumbar punctures are performed befoore therapy, after 24 hours, after 48 to 72 hours, and after the termination of therapy (Steele and Bradsher, 1983). In subacute or chronic ear infections, more lumbar punctures may be necessary to follow the course of recovery.

In the first 2 to 3 days, the cultures and the direct smears should be converting to negative; however, the protein, glucose, white cell count, and percentage of polymorphonuclear leukocytes remain approximately the same as during pretreatment and later resolve (Blazer et al, 1982).

## Brain Abscess

### Pathophysiology

A brain abscess is an accumulation of pus surrounded by a region of encephalitis within the cerebrum or cerebellum. Brain abscesses predominate in men, particularly in the third decade of life (Harrison, 1982) but they may occur at any age. In children the predominant predisposing factor is cyanotic heart disease (Hirsch et al, 1983).

Reports vary widely as to the number of brain abscesses encountered and the percentage that is otogenic. Foor example, Nielsen et al (1983)0 reported 200 patients with brain abscesses discovered over a 41-year period; conversely, Ayyagarii et al (1983) reported 45 patients with brain abscesses encountered in 21 months and estimated that 42% were otogenic. In children, 35% of brain abscesses originate from ear, nose, and throat infection (Hirsch et al, 1983).

Otogenic brain abscesses predominantly originate from venous thrombophlebitis and not from direct extension through the dura (Hirsch et al, 1983)0. The temporal lobe is most frequently involved and the cerebellum is second. Multiple brain abscesses are uncommon; when present, they usually result from meningitis occurring in neonates or infants (Hirsch, 1983).

The dura is extremely resistant to infection, but persistent infection may locally inflame the dura to such an extent that thrombophlebitis occurs in adjacent cerebral vessels. Retrograde thrombophlebitis in the cerebral or cerebellar veins rapidly extends to the small terminal veins in the white matter, where the defense against infection is minimal, and rapid spread of liquifaction necrosis results in abscess formation. The surrounding septic encephalitis incites a poor and unpredictable encapsulation attempt by brain fibroblast and glial cells. Softening of the surrounding tissue and poor encapsulation sometimes allow the infection to spread toward the ventricles or peripherally toward the cortex, ultimately to rupture into a ventricle or the subarachnoid space.

Aerobic and/or anaerobic organisms are found in pure or mixed cultures within brain abscesses. Multiple organisms are found in at least 55% of these cases (Ayyagari et al, 1983). Aerobic organisms are gram-positive cocci and gram-negative bacilli. The most common gram-positive cocci are *Streptococcus* species followed very closely by *Staphylococcus* species (Table 158-2). By far the most common gram-negative bacilli are *Proteus* species, followed by *Escherichia coli*, *Pseudomonas* species, and *Klebsiella* species. In order of frequency, the three most common anaerobic organisms to cause brain abscesses are *Peptococcus* species, *Peptostreptococcus* species, and *Bacteroides fragilis.*

It is of interest to note that *H. influenzae* and *Pseudomonas* species are very common otologic pathogens but are rarely found in brain abscesses. The high incidence of gram-positive aerobic streptococci and staphylococci and the significant number of anaerobic streptococci and *Bacteroides* organisms are consistent with clinical experience and impression that acute exacerbation in chronic ear disease is an important precursor to the development of brain abscess. A fetid aural discharge is frequently associated with staphylococci and with anaerobic organisms; fetid discharge suggests poor aeration, blockage, and bone destruction.

## Diagnosis

The classic symptoms of a brain abscess are fever, impaired consciousness, headache, vomiting, stiff neck, focal motor seizures, and papilledema (Hirsch et al, 1983). However, for a given individual to manifest all of these clinical signs is unusual. In fact, even within a center to which brain abscesses are characteristically referred from a wide region, the correct diagnosis was frequently not considered on admission (Harrison, 1982).

Brain abscesses develop through four clinical stages over a period of weeks or months. The last two stages are frequently misdiagnosed despite being the most obvious stages (Mawson, 1974). The four clinical stages are:

1. Invasion (initial encephalitis).

2. Localization (latent or quiescent abscess).

3. Enlargement (manifest abscess).

4. Termination (rupture of the abscess).

In stage 1 there is usually a low-grade fever, loss of concentration or drowsiness, and, most important, a headache and feeling of general malaise. Frequently these subtle symptoms are overlooked or remain undiagnosed. After several days these nonspecific symptoms disappear.

Stage 2 is often clinically silent. In this stage the patient has no symptoms and may ultimately forget the initial symptoms that existed during stage 1. This latent stage may last for weeks.

Stage 3 begins as the abscess forms from the previous cerebritis and begins to enlarge. Focal symptoms from the expanding mass may result in visual field defects, aphasia, motor

or sensory paralysis, ataxia, and generalized or focal seizures, depending on the site of lesion. Focal motor deficits may occur in only 38% of the patients and seizures in only 29% (Hirsch et al, 1983). The most common clinical features appearing on admission during this stage are fever in 62%, impaired consciousness or lethargy in 56%, and headache and vomiting in 53%. When looked for, papilledema exists in 70% (Hirsch et al, 1983).

Stage 4 occurs when the abscess ruptures into either the ventricle or the subarachnoid space, resulting in a rapidly progressive, frequently fatal, course.

Electroencephalography is positive in 96% of cases. The most common abnormality is a focus of delta waves (Hirsch et al, 1983). CT scanning with infusion confirms the diagnosis; the abscess appears as a hypodense area encircled by an enhancing ring (Fig. 158-5). CT findings may be subtle or absent in the first two stages. MRI is helpful in identifying brain abscesses and more sensitive in detecting extraparenchymal spread to the subarachnoid space or ventricle than is CT (Haimes et al, 1989; Maniglia et al, 1989; Zimmerman and Haimes, 1989) (Fig. 158-6).

## Management

Treatment is rather straightforward. The patient is admitted to the hospital and, after a diagnosis by CT scan, 4 million units of intravenous penicillin is given every 6 hours, 600 mg of chloramphenicol every 6 hours, and 500 mg of metronidazole every 6 hours. Intravenous dexamethasone, 4 mg every 6 hours, is also initiated (Maurice-Williams, 1983). The ear is suctioned clean, and topical antibiotic drops are begun.

The patient is taken to surgery as soon as possible. Frequently mannitol is given intravenously immediately before surgery. Under general anesthesia the abscess is approached through a sterile field (not the ear); cannulated; and aspirated for smear, culture, and sensitivity, after which the abscess cavity is gently irrigated with saline and antibiotics.

Some controversy exists regarding treatment of the abscess from this point. The following therapeutic alternatives are available:

1. If necessary, repeated aspiration and irrigation of the abscess cavity until resolution.

2. Open incision and drainage and irrigation of the abscess cavity under direct vision, followed by closure. This is performed immediately after cannulation, aspiration, and irrigation, under the same general anesthetic.

3. Immediate or delayed resection of the abscess.

There is a tendency toward repeated aspiration if necessary (Hirsch et al, 1983; Nielsen et al, 1983) or open evacuation of pus without resection (Maurice-Williams, 1983) rather than primary or secondary removal of the abscess. Experience indicates that the incidence of late epilepsy is actually less with these techniques than with resection. Resection does play a reasonably noncontroversial role if evidence of a persistent abscess capsule is discovered, particularly if epilepsy is concomitant. The final decision is best left to the neurosurgical colleague (Hirsch et al, 1983).

As soon as it is suitable in the next several days, the ear is operated on with the appropriate surgical procedure.

In cases treated with repeated aspiration it is not uncommon for there to be a small hyperdense lesion on the CT scan 2 months after the treatment, which is interpreted as inflammatory granuloma. Most of these have cleared 1 year later. A few take more than a year to clear (Hirsch et al, 1983). When pus is openly evacuated, the CT scan frequently shows clearance in 4 to 8 weeks (Maurice-Williams, 1983).

Approximately 73% of survivors have limited, neurologic sequelae or none at all, allowing a normal life, work, and schooling (Hirsch et al, 1983). The mortality rate has been reported to be as low as 0% (Maurice-Williams, 1983) and as high as 80% (Dohrmann and Elrick, 1982). The general trend from the late 1950s to the 1980s has been a gradual reduction of mortality from 57% to, ultimately, 12% (Hirsch et al, 1983). The major factor influencing mortality is the condition of the patient on admission. Hence, the earlier the diagnosis is made, the better the chances of survival with little or no neurologic sequelae.

## Otitic Hydrocephalus

### Pathophysiology

Symonds (1931) coined the term *otitic hydrocephalus* to describe a syndrome of increased intracranial pressure associated with suppurative ear disease, in which no meningitis or brain abscess could be found. Symonds (1927) had earlier discussed the diagnosis and localization of brain abscesses and described three cases in which the signs and symptoms simulated an abscess but no abscess was found. Three cases had suppurative otitis media, focal signs, increased intracranial pressure, normal CSF, and no evidence of cerebral abscess. In one of the cases an acute coalescent mastoditis and an extradural abscess were found compressing but not thrombosing the lateral sinus. Symonds (1937) later hypothesized that superior longitudinal sinus thrombosis and thus obstruction to CSF resorption was the basis for the syndrome.

Williams (19380 reviewed otitic hydrocephalus, considering the hydrodynamics of CSF production and resorption as it relates to inflammation. Very little has been written since on the topic, despite the tremendous accumulation of information and technologic advances for the study of CSF production and intracranial pressure regulation. Although unproven, it seems likely that otitic hydrocephalus results from sudden inadequate venous drainage from the cranial cavity caused by obstruction (thrombophlebitis or phlebothrombosis) of one lateral or sigmoid sinus.

### Diagnosis

The important points to make at this time are not of pathophysiology but of practicality. Known causes of increased intracranial pressure, such as brain edema, extradural and subdural effusions, meningitis, and brain abscess, must be sought. Failing to discover any of these entities preoperatively, one should remember that most if not all cases of otitic hydrocephalus are associated with extradural granulation tissue or abscess and sigmoid sinus thrombophlebitis. It is imperative that surgical attention be directed to these two areas in cases

labeled "otitic hydrocephalus".

Significant papilledema, which can ultimately lead to blindness, is a frequent finding in this disease, as are headache and reduced consciousness or lethargy.

## Management

Management is designed to eradicate the suppurative ear disease rapidly with the appropriate antimicrobial and surgical therapy and to treat the increased intracranial pressure aggressively to prevent the sequelae of severe intracranial pressure. Visual fields are usually affected earlier than is visual acuity. Careful monitoring of the visual fields, acuity, and degree of papilledema is necessary. Spontaneous resolution of intracranial pressure after the eradication of otologic disease usually takes several weeks to months.

The literature on increased intracranial pressure of unknown origin not associated with suppurative ear disease is not discussed in this chapter.

## Summary

A complication from suppurative ear disease is defined as the spread of infection beyond the confines of the pneumatized spaces of the temporal bone and the attendant mucosa. The first such area is the bone surrounding the pneumatized spaces; hence, bone destruction is a sign of complication.

Current terminology, predominantly developed and popularized by Bluestone, is extremely helpful in understanding and properly describing the inflammatory processes involving the pneumatized spaces of the temporal bone. *Otitis media, mastoiditis,* and *petrositis* are site-of-lesion statements and also describe the state of the mucosa. *Acute, subacute,* and *chronic* are time statements identifying the duration of mucosal inflammation: *acute* is less than 3 weeks, *subacute* is 3 weeks to 3 months, and *chronic* is greater than 3 months. The substance occupying the space is an *effusion* if the tympanic membrane is intact and a *discharge* if the tympanic membrane is perforated. *Serous, mucoid,* and *suppuratiive* are qualitative statements describing the gross characteristic of the effusion or discharge.

Precise terminology is of more than academic importance. The identification of an impending complication is singularly dependent on the proper use and recording of appropriate terminology. For example, a "chronically draining ear" may represent a chronic otitis media - mastoiditis with intermittent mucoid discharge, or it may represent chronic otitis media - mastoiditis with persistent fetid suppurative discharge despite treatment. The implications for complication in these two examples are remarkably different.

Some complications are obvious when present, such as facial paralysis, meningitis, suppurative labyrinthitis, and subperiosteal abscess. Some complications are quite subtle, such as serous labyrinthitis, chronic labyrinthitis, subdural abscess, sigmoid sinus thrombophlebitis, otitic hydrocephalus, and brain abscess.

Complications tend to follow predictable patterns. The organisms in the ear and in the complication are predictable, depending on the presence or absence of a tympanic membrane perforation. Suppurative ear disease and a resultant complication with an intact tympanic membrane are more likely to involve *S. pneumoniae* and *H. influenzae.* Suppurative ear disease with a complication and a tympanic membrane perforation is more likely to involve staphylococci, gram-negative bacilli, and anaerobic flora. A fetid discharge frequently contains staphylococci and anaerobic bacteria.

Persistent acute infection or acute exacerbation of a chronic infection are the usual precursors of complications. The principal exceptions are acute bacterial meningitis and acute lower motor-neuron facial paralysis in young children. It should be emphasized that "recurrent" infections occurring at a 2-month or less interval may not be recurrent de novo infections, but recurrent clinical manifestations of a persistent acute infection that is being inadequately eradicated and "masked" by the treatment.

Treatment is focused at the ear to eradicate the infection rapidly and at the complication to eradicate infection and reduce sequelae.

The most important tools leading to the early diagnosis of impending or early complications are precise thinking and careful notation on the patient's records.