Chapter 129: Esophageal Diverticula

Patrick J. Doyle, Holly E. Stevens

The classical, easily diagnosed esophageal diverticulum occurs often enough that it may give the otolaryngologist-head and neck surgeon a false sense of security in the management of all diverticula. However, subtle differences in signs and symptoms allow the physician to differentiate between types of diverticula. Consider the following cases:

The first patient is a 64-year-old man who consults his physician with a complaint of increasing dysphagia over a period of 6 years. He notes that food "sticks", and he must make several attempts to swallow successfully. He has recently had episodes of spontaneous regurgitation of foul, undigested food. Aspiration of food causes episodes of coughing, which are a severe problem at night. He denies all other symptoms of otolaryngeal or pharyngeal disease, and physical examination is normal except for pooling of secretions in the piriform sinuses. A barium swallow examination confirms the physician's provisional diagnosis. This man has Zenker's diverticulum (Fig. 130-1), and surgical treatment successfuly relieves his symptoms.

The second patient is a 60-year-old woman with progressively increasing dysphagia of 3 years' duration. Episodes of substernal pain that come on after she begins her meal make it necessary for her to discontinue eating for 30 to 40 minutes. After this rest period she is able to complete the meal. History and physical examination reveal no other symptoms or signs, but a barium swallow examination demonstrates a large midthoracic diverticulum (Fig. 130-2). This patient requires transthoracic resection of the diverticulum.

To adequately diagnose and threat these and other patients with esophageal diverticula, the surgeon must be able to answer the following questions:

What is a diverticulum?

How are diverticula classified?

What are the relevant anatomy and physiology of the hypopharynx and esophagus?

What causes esophageal diverticula?

What are the signs and symptoms of esophageal diverticula?

How should these patients be investigated?

How should these patients be treated?

Definition and Classification

A diverticulum is defined as a circumscribed pouch or sac created by herniation of the lining mucous membrane through a defect in the muscular coat of a tubular organ. Esophageal diverticula may be congenital or acquired, and the acquired esophageal diverticulum may be single or multiple. Although these diverticula may be classified by mode of development or the composition of the diverticular wall, the most practical and commonly used classification is by location (Fig. 130-3).

Zenker's diverticulum (also referred to as a *hypopharyngeal* or *pharyngoesophageal diverticulum*) consists of a herniation of mucosa and submucosa through a defect in the pharyngeal muscular coat. The diverticulum is referred to as a *pulsion diverticulum*, but the exact cause remains somewhat controversial and is discussed in detail later in the chapter.

An *epiphrenic diverticulum* is located in the distal esophagus and contains some attenuated muscle layer in its wall. It is also a pulsion diverticulum.

A *midthoracic diverticulum* is usually a traction diverticulum produced by continued peristalsis of the esophagus against a fixed esophageal adhesion. It occurs as a result of previous mediastinal inflammatory disease. Pulsion diverticulum also occur in the midthoracic esophagus.

Intramural pseudodiverticulosis consists of tiny, multiple out-pouchings along the course of the esophagus.

Anatomy (Fig. 130-4)

The inferior constrictor muscle of the pharynx arises from the oblique line of the thyroid cartilage and the posterior part of the arch of the cricoid cartilage. It inserts into the pharyngeal raphe in the posterior midline. The thyropharyngeal portion consists of upper fibers that arch upward to overlap a part of the midline constrictor and middle fibers that ascend less obliquely. The cricopharyngeal portion consists of lower fibers that are approximately horizontal or even arch slightly downward. They blend with the thyropharyngeal part above and the circular fibers of the esophagus below. This cricopharyngeal part surrounds the posterior pharynx like a sling, is thicker than the muscle above and below, and has no muscular raphe. It is usually referred to as the *cricopharyngeus muscle*, and it corresponds to the upper esophageal sphincter. Immediately above this sphincter, between it and the thicker upper fibers of the thyropharyngeus muscle, is an area of relative weakness known as *Killian's dehiscence* or *triangle*. This is the generally accepted site of herniation of hypopharyngeal diverticula (Ellis et al, 1969; Gullane et al, 1983; King, 1947).

The esophagus begins at the lower border of the cricoid cartilage. For most of its length it is reinforced with two muscular layers: an inner circular layer and an outer longitudinal layer. The former blends superiorly with the cricopharyngeus muscle above. The latter, however, diverges at the upper end, forming two bands that swing laterally and anteriorly around the esophagus to attach to a common tendon behind the cricoid cartilage

(Hollinshead, 1968). The posterior esophageal wall between these divergent bands is therefore covered with a single layer of circular fibers. This forms a second potentially weak area known as *Laimer's triangle* or the *Laimer-Haeckermann area.* However, the lack of any reports of diverticula arising in this site argues against the theory of congenital weakness as a sole factor in the creation of diverticula.

Hollinshead (1968) includes other potentially weak areas in his summary. The Killian-Jamieson area is a fat-filled lateral dehiscence that transmits the inferior laryngeal nerve and artery forward between the cricopharyngeus muscle above and the upper esophageal fibers below. Diverticula arising in this site explain the close relationship of the neck of the sac to the inferior laryngeal nerve. Hollinshead points out, however, that this relationship necessarily develops with any sac attaining the diameter of the esophagus. Other congenitally weak areas in the region of the cricopharyngeus muscle, such as those allowing passage of branches of the inferior thyroid artery, have also been implicated in the development of hypopharyngeal diverticula.

Zenker's Diverticulum

Etiology

The fact that hypopharyngeal diverticula most commonly arise between the sixth and ninth decade has been regarded as evidence against a purely congenital origin. Most authors attribute their development to a wide spectrum of functional disorders, none of which are irrefutable to date. With the advent of more sophisticated and accurate manometric methods for measuring pharyngoesophageal sphincter function, one might have hoped for a common consensus. On the contrary, even the most popular theories continue to be questioned.

In the normal state the cricopharyngeus muscle is in a state of tonic contraction, with a mean pressure of approximately 40 cm H_2O. During the second phase of swallowing, sequential contraction of the pharyngeal constrictors occurs from above downward, stripping the bolus inferiorly. Synchronous with this wave, the pressure in the cricopharyngeal sphincter drops abruptly for an average of 0.5 to 1.2 seconds. This relaxation allows the bolus to enter the esophagus, after which the pressure abruptly rises again, indicating a contraction wave passing through the sphincter, which is propagated down the esophagus as the peristaltic wave.

Sutherland (1962) and Belsey (1966) both favored the theory that tonic spasm or achalasia of the cricopharyngeus muscle prevents the downward passage of the bolus, and the weakened area of Killian's dehiscence gives way to the continued pressure above the sphincter with eventual herniation. Negus (1950) cited several possible reasons for the achalasia (chronic inflammation preventing relaxation, healed inflammatory lesions leading to secondary stenosis, neurosensory deficits, and idiopathic causes), and he placed particular emphsis on the weak area above the cricopharyngeus muscle as being important. Furthermore, he suggested that during swallowing this muscle gap tends to widen because of elevation of the upper pharynx and larynx while the esophagus, anchored to the diaphragm, exerts a downward pull. With spasm of the cricopharyngeus muscle or incoordination between it and the contracted pharynx above, prolapse of mucosa develops through this widened dehiscence.

Dohlman and Mattsson (1959) stressed the importance of an intact prevertebral fascial layer that normally supports the posterior wall of the larynx. With increasing age, this fascia may weaken, allowing the larynx to fall forward and thus decrease the circumference of the cricopharyngeus muscle, which is normally stretched open during act of swallowing. A functional obstruction would be produced, and with elevation of intrapharyngeal pressure on swallowing, herniation of mucosa through the weakened posterior triangle, now unsupported by prevertebral fascia, would occur.

Ardran et al (1964) used a cineradiographic study to demonstrate that the problem is one of early closure of the cricopharyngeal sphincter. The contracting sphincter is directed forward and slightly upward. Simultaneously the descending peristaltic wave cause backward bowing of the posterior pharyngeal wall. The development of a small dimple marks the site of a pouch where the barium in the pharynx is held back primarily by the shelflike projection formed as a result of early closure of the cricopharyngeus muscle. These patients usually demonstrate weakness or incoordination in the pharyngeal peristaltic wave, further impairing passage of the bolus.

Ellis et al (1969) came to similar conclusions by measuring intraluminal pressures in patients with diverticula and comparing them to those of patients without diverticula. Once again, the resting tone and time of onset of relaxation of the sphincter were normal. The investigators found, however, that the termination of relaxation occurs a fraction of a second before pharyngeal contraction is complete, so that the pharyngoesophageal peristaltic sequence is not coordinated.

Lichter (1978) also supports the incoordination theory. His manometric study showed that although the sphincter relaxes for the normal amount of time, relaxation begins prematurely and therefore ends prematurely. The effect is the same: high pharyngeal pressures conducive to herniation of mucosa through a muscle-deficient area. Lichter also noted the phenomenon of repetitive pharyngeal swallow, thought to be a result of obstruction to the swallow wave at the upper esophageal sphincter because it disappeared following sphincterotomy.

In contrast to Lichter's concept of premature relaxation in an otherwise normal sphincter, Cross et al (1961), also using manometry, found that base pressures were high, the relaxation phase was diminished and delayed, and the contraction phase was exaggerated. The hypertonic cricopharyngeus muscle then required higher pharyngeal pressure for propulsion of the bolus, hence predisposing to a pulsion diverticulum. Their studies were done in patients who did not yet have diverticula, lending support to a cause-rather-than-effect relationship.

Wilson (1962), Delahunty et al (1971), Kodicek and Creamer (1961), Pedersen et al (1973), Knuff et al (1982), and van Overbeek and Hoeksema (1982) have all submitted theories that are modifications of those described above.

Opinion concerning the pathophysiology of hypopharyngeal diverticula is divergent. Part of the disagreement may be ascribed to differences in measuring techniques. The high reactivity of the sphincter to mechanical stimulation makes it difficult to study. Furthermore, the anatomic configuration of the sphincter is such that normal pressure readings vary with the circumferential orientation of the pressure-reading catheters (Winans, 1972). It is

noteworthy as well that although the investigators drew their conclusions from the predominant findings, not all the abnormalities were noted in all patients on all swallows. This individual variability further complicates accurate pressure measurements. The true pathophysiology therefore remains uncertain, although some mechanism incorporating incoordination between the descending peristaltic wave and cricopharyngeal dysfunction is still most widely held as being responsible.

Clinical features

Hypopharyngeal diverticula can occur through adult life but are most commonly seen in the sixth through ninth decades. Men are affected approximately two to three times more often than women.

The chief symptom is dysphagia, often long-standing and of insidious onset. Diverticular retention leads to episodes of spontaneous regurgitation of foul, undigested food that interrupt the patient's meal and cause bad breath. The patient may complain of food sticking and having to swallow several times. A choking sensation may develop. As the sac grows larger, the patient may take longer and longer to eat, and distension of the sac with food and saliva may lead to local pain.

Recurrent aspiration and laryngeal irritation cause coughing. This may occur immediately following ingestion but is often worse at night. Nocturnal aspiration, especially in elderly and debilitated patients, leads to pneumonia or even lung abscess but rarely is respiratory distress the initial symptom (Welsh and Payne, 1973). Benign fistulas between diverticula and the trachea have also been recorded and are another cause of cough (Senders and Babin, 1983).

Hoarseness may occur secondarily to laryngeal irritation or occasionally as a result of compression of the recurrent laryngeal nerve. However, in that instance one must always suspect secondary carcinoma. This suspicion is raised when blood is seen in the regurgitated material. Carcinoma in Zenker's diverticulum is a rare phenomenon. In a retrospective study of 961 surgical cases, only three malignancies were reported, an incidence of 0.31% (Wychulius et al, 1969).

Whereas a long-neglected diverticulum may eventually lead to emaciation and complete esophageal obstruction, small pouches of recent onset are often entirely asymptomatic. Findings on physical examination are most often unremarkable, although a large diverticulum may be palpable as a soft, compressible, doughy, cervical mass just posterior to the lower part of the sternocleidomastoid muscle, usually on the left side. Manual pressure may produce a gurgling noise followed by a cough. On indirect laryngoscopy, pooling of secretions may be seen in the piriform sinuses.

Diagnostic assessment

The diagnosis of hypopharyngeal diverticulum is most often suspected on the basis of the history alone. Lateral soft-tissue films may demonstrate retropharyngeal fullness or an air-fluid level behind the pharyngoesophageal junction. Secondary dilatation of the pharynx may occur. Confirmation by contrast radiography is usually definitive. A barium swallow examination and cine-esophagogram (Fig. 130-1) define the size and orientation of the sac. It usually herniates to the left and may range in size from a 1-cm pouch to a massive lesion extending into the thorax. Radiographic defects in the sac usually result from retained food (Fig. 130-5), but tumor formation must be considered if the defect remains unchanged on repeated examination. Other esophageal disorders such as a hiatus hernia may be demonstrated.

Esophagoscopy is hazardous and is not required to make the diagnosis. Therefore it should be avoided unless evidence of secondary disease such as a carcinoma exists or evacuation of a sac is necessary preoperatively. Should it be necessary, passing the esophagoscope over a previously swallowed thread may enhance safety. Perforation of the thin-walled diverticulum is the chief risk. Likewise, manometry has no place in the usual workup.

The sac wall is composed of mucosa, which may be inflamed, and fibrous tissue with no muscular component. It enlarges both in diameter and length and descends to lie between the esophagus and the vertebral column, most often on the left. Presumably this occurs because the normal cervical esophagus bulges to the left. King (1947) theorized that there is less resistance on the left side because the common carotid is further removed from the trachea and esophagus on that side. Occasionally the sac wall reaches the posterosuperior mediastinum. Distension of the sac with food and secretions causes secondary angulation and compression of the esophagus so that esophageal obstruction increases and food preferentially enters the sac. Compression of the recurrent laryngeal nerve may be caused by a large pouch, especially if secondary neoplasia develops. Carcinomas are uncommon and usually, but not exclusively, occur in patients with long-standing symptoms of diverticula.

Differential diagnosis

The differential diagnosis depends on the stage of the hypopharyngeal diverticulum when it is first examined. In the early stages it must be differentiated from other causes of dysphagia. Structural abnormalities may exist, such as congenital or acquired stenosis, stricturing following caustic ingestion, Plummer-Vinson syndrome, an extrinsic mass lesion causing esophageal compression, and an esophageal carcinoma. A variety of functional disorders causing muscular incoordination of the upper esophageal sphincter must be considered, including achalasia and spasm. Blakeley's classification (1968) of muscular incoordination provides an easy checklist: (1) myopathic, including muscular dystrophy, thyrotoxicosis, myasthenia gravis, and carcinomatosis; (2) central neuropathic, including cerebral vascular accident, bulbar palsy, and trauma; (3) peripheral neuropathic, including iatrogenic trauma, neuritis, and collagen vascular disorders; and (4) idiopathic.

Once the sac enlarges into the neck, it must be differentiated from other causes of neck masses. Cystic lesions such as cystic hygromas, branchial cleft cysts, and (rarely) thyroglossal duct cysts occur in the lateral part of the neck. Laryngoceles, carotid aneurysms, lipomas, neurofibromas, neck abscesses, and adenopathy may also cause lateral neck masses. With the typical history of dysphagia and aspiration, however, differentiation is usually possible on the history alone, and the diagnosis is certainly made obvious once radiographic studies are obtained.

Management

If a hypopharyngeal diverticulum is large enough to cause symptoms, surgery is necessary. As testimony to the various possible causes, many surgical treatments have been proposed. Diverticulectomy with or without myotomy, diverticulopexy with or without myotomy, and myotomy alone are external approaches. Endoscopic procedures include diathermy of the esophageal lumen-diverticulum party wall (Dohlman's procedure) and dilatation.

Von Bergmann reported the first surgical success in 1892 (Payne and Clagett, 1965). Goldman, in 1909, first popularized the two-stage procedure believed to be necessary in the preantibiotic era to avoid problems with wound infection and mediastinitis (Negus, 1950). However, between 1939 and 1948 several authors (Harrington, 1939; King, 1947; Sweet, 1947; Shallow and Clerf, 1948) showed that the one-stage procedure was just as safe with equally good results and actually resulted in less morbidity, as well as a shorter hospital stay. In 1965 Payne and Clagett, in a series of 478 patients, confirmed the safety of the one-stage procedure.

Negus (1950) suggested dilatation of the sphincter with a hydrostatic bag to be repeated every few months as necessary until the sac became large enough so that sufficient tissue would be available for closure of the defect following excision of the diverticulum. It is of interest that he condemned the idea of suspension of the diverticulum with myotomy because he believed that this would lead to air swallowing and ballooning of the esophagus. He also believed that a recurrence would be likely, since the weak area above the cricopharyngeus muscle was still present and not reinforced in any way. However, in 1951 Kaplan described the techniques of cricopharyngeal myotomy for cricopharyngeal achalasia in a variety of other conditions and found it to be satisfactory (Payne and Clagett, 1965).

In 1962 Sutherland proposed cricopharyngeal myotomy without removal of the sac when repeated dilatation alone failed to relieve symptoms. He performed 10 such myotomies over an English 20-gauge esophageal tube passed into the esophagus. Not all patients had pouches, and those present were small, but in every case the patient was completely symptom free or suffered minimal symptoms following this procedure.

In 1966 Belsey introduced the diverticulopexy for diverticula greater than 2 cm in diameter. He suspended the inverted sac by tacking it to the anterior spinal ligament. In all cases a cricopharyngeal myotomy was performed. He reasoned that excision of a diverticulum without relief of the underlying spasm would allow the same pressures that produced the sac in the first place to predispose to leakage and fistulization at the incision line as well as recurrence of the sac. With myotomy and suspension, the underlying problem was removed

and an incision line was avoided completely.

Further support for the myotomy came in 1967 with an article by Einarsson and Hallen (167). They followed 20 patients for up to 12 years who had undergone diverticulectomy without myotomy. Seventeen had radiographic evidence of recurrent pouches.

Although cricopharyngeal myotomy was not always done in the past, it seems to be the accepted method today. Payne and Clagett reported on a large series from the Mayo Clinic in 1965 in which 478 patients underwent excision without myotomy. They followed about half of these patients for an unspecified time period and found radiographic evidence of recurrent diverticula in only six; narrowing at the resection site required dilatation in three. Seven patients who showed no objective signs of disease complained of dysphagia.

This series was expanded and reported again in 1973 (Welsh and Payne) at which time the same one-stage technique was being practiced. Of 164 patients who had been followed for 5 to 14 years, 93% had excellent results (symptom free) or good results (occasional sticking) based on subjective reports of symptoms only. A total of 809 patients were included in the study, and of the remainder, all of whom were followed for at least 4 years, only 3.3% reported recurrence. However, these were subjective reports.

Despite these seemingly excellent results, the same authors reported a third time in 1983 (Payne and King). In the last decade they added cricopharyngeal myotomy to the one-stage excision or did myotomy alone for very small sacs. They found, however, that in 35 patients with diverticulectomy and myotomy, the results were no better than in those patients with diverticulectomy alone. They could therefore not justify myotomy on the basis of their data; however, in those patients who had anatomically proven recurrences, those who had undergone previous myotomy had much less severe symptoms than the other patients. Since the added operating time and morbidity of myotomy are insignificant, Payne and King believe the procedure is warranted. Their recurrence rate with myotomy alone (22% compared with 5% for diverticulectomy with or without myotomy) was not encouraging; however, their series was very small. Presumably in those nine patients the sacs were so small that myotomy alone was the only appropriate treatment, and they therefore support this treatment for symptomatic patients with small sacs.

Gullane et al (1983) reported on 25 patients followed for 42 to 48 months. They noted their results to be much poorer than others reported but pointed out the large discrepancy between subjective and objective reporting. Of 17 patients who underwent myotomy and diverticulectomy, 8 (47%) showed radiographic evidence of recurrence at 42 months. Only four were symptomatic. Six of seven patients (85.7%) treated with myotomy plus diverticulopexy showed recurrent diverticula, but only one was symptomatic. One of three patients who underwent myotomy alone showed radiographic evidence of recurrence. The overall objective recurrence rate was 55.6%, but only 18.6% of patients were symptomatic.

Reviewing the literature does not conclusively prove that the addition of cricopharyngeal myotomy gives better resuilts than diverticulectomy alone. Analysis is difficult because of the varying methods and follow-up times of the postoperative assessments, individual variations among patients, the size of the diverticula, the presence or absence of

associated esophageal disease, and factors related to different surgical techniques. Nevertheless, the trend toward cricopharyngeal myotomy is obvious and seems to be the accepted method today. For a small diverticulum in which excision would prove difficult, myotomy alone is indicated. Excision is preferable to suspension in patients with a large diverticulum and should be combined with myotomy.

Dohlman's procedure is an alternative method of surgical treatment for hypopharyngeal diverticula. In 1906 Mosher first cut the party wall between the sac and esophageal lumen with a knife (Dohlman and Mattsson, 1960). Six patients were treated successfully, but mediastinitis resulted in the seventh case, and the procedure was abandoned. Another successful endoscopic procedure was not reported until about 30 years later. In 1937 Sieffert cut the party wall with scissors. Almost simultaneously, Dohlman began using the endoscopic diathermy method that has since been popularized and bears his name (Dohlman and Mattsson, 1960). In 1960 he reported on a series of 100 patients with recurrence rate of only 7%. He reported "no deaths or severe complications due to the operation". Advocates of this procedure agree that, compared to conventional management, it is a lesser operative procedure with a shorter intraoperative time, shorter convalescence and hospital stay, and quicker return to swallowing and nutrition. All these factors make it ideal for an older patient less able to withstand a surgical procedure. Although recurrence is more likely, the time it would take to develop may exceed the patient's life span. Consequently, recurrence is not a clinical problem.

Dohlman's technique seems to have met greater favor in Europe than in North America. Those reporting on this method have found that the subjective results closely parallel those of the conventional method. Todd (1974) notes relief of symptoms in 84% of patients undergoing diverticulotomy with diathermy as compared with relief in 88% with the conventional method. No follow-up period was given. Complications occurred in 9% versus 58% for the conventional procedure. In a series of 211 patients, most of whom were folloed for longer than 2 years, van Overbeek and Hoeksema (1982) found 91.5% to be highly satisfied and 8% to be fairly satisfied. These results compare favorably with conventional treatment. On follow-up, however, despite their asymptomatic state, residual diverticula were evident radiographically in an unspecified number of patients. The authors stressed that the procedure requires operator experience and expertise.

Holinger and Shild (1969) reported on 160 patients who underwent surgical treatment, 37 of whom had diverticulomies by the diathermy technique. Their follow-up period was as long as 26 years. Although there was radiographic evidence of a residual sac in more of the patients who had undergone Dohlman's procedure than in those who had had an external incision, the authors found that the patients were generally satisfied and experienced only slight hesitation on swallowing certain foods. None had any obstruction. Any sacs that were seen emptied easily on a second swallow, and aspiration was never a problem.

Trible (1975) found in his series of 24 patients that relief of symptoms ranged from "miraculous to moderate". The patients had no complications. He pointed out, however, that the procedure is simply meant to allow better drainage of material into the esophagus and to prevent spillage into the larynx. The postoperative esophagogram resembles the preoperative esophagogram immediately, but should totally clear in 2 to 5 minutes. Trible's patients were seen at yearly intervals. Many reported that they had recurrent symptoms, but none were significant enought to warrant reoperation.

The previous discussion brings out several points. Most authors believe that an experienced physician can perform Dohlman's procedure safely, but because recurrences are more common in the long term, all younger and fit patients should have a conventional operation as the first choice. Patients who are old or medically unfit or those with a generally poor prognosis are more appropriately treated by the endoscopic procedure, which an experienced surgeon can accomplish in a few minutes. Larger diverticula often require more than one procedure to completely cut the party wall and are less likely to have a good result. Harrison (1958) suggested that in the case of larger sacs, hypertrophy of the cricopharyngeus muscle or calcium deposits in the party wall may make endoscopic division difficult. For that reason, whenever possible, larger sacs are treated with diverticulectomy. When large sacs are treated by the endoscopic method, most surgeons cut only one half to one third of the tissue bridge in one sitting; the procedure is repeated a few days or weeks later. Todd (1974) stressed that additional procedures do not result in further improvement and suggested than to more than two be performed. The purposes of the procedure - to shorten the time the patient is anesthetized, to decrease postoperative morbidity, and to shorten the total hospital stay - are defeated if the patient must have repeated procedures.

A final argument against using Dohlman's procedure is that because the sac is not removed, the risk exists of a carcinoma developing in the long-standing sac. Even though stagnation, which is believed to be important in the development of a carcinoma, no longer occurs (at least at first), changes may have already occurred, thereby leaving the patient at risk.

One-stage hypopharyngeal diverticulectomy with cricopharyngeal myotomy

Today most patients with a hypopharyngeal diverticulum seek medical treatment before complications develop, and therefore little preoperative preparation is required. However, if the patient is malnourished and dehydrated as a result of advanced disease or if complications such as pneumonitis or lung abscess have occurred, these factors should be appropriately treated preoperatively whenever possible. Furthermore, if the sac is large, an effort should be made to ensure the sac is emptied of all secretions, food, and barium before induction of anesthesia to avoid aspiration. Antibiotics should be administered in the usual fashion for surgery of the pharynx in which the lumen is entered. The first dose is given with the premedications, and therapy is continued for 24 to 48 hours.

The procedure is carried out with the patient under general anesthesia. In a patient in whom the diverticulum has not been emptied, intubation while the patient is awake may be desired to prevent aspiration. This is followed by induction of general anesthesia.

An incision is made along the anterior border of the left sternocleidomastoid muscle, extending from the hyoid bone above to approximately two fingerbreadths above the clavicle. The sternocleidomastoid muscle is then retracted laterally to reveal the anterior belly of the omohyoid muscle, which can be dissected free and either cut or retracted. Anteriorly the sternohyoid and sternothyroid muscles are exposed and retracted. It may be necessary to divide and ligate the middle thyroid veins, allowing the ipsilateral lobe of the thyroid gland to be turned forward to help expose the operative field.

Blunt and sharp dissection is used to enter the retropharyngeal space just cephalad to the omohyoid muscle. The contents of the carotid sheath are retracted laterally while the pharynx and larynx are retracted to the right. Division and ligation of the inferior thyroid artery may be required. The dissection is then carried posteriorly along the anterior aspect of the prevertebral fascia to the tracheoesophageal groove and the diverticulum. It is good practice to positively identify the recurrent laryngeal nerve, thereby ensuring that it is not accidentally injured. It enters the larynx just inferior to the cricothyroid joint.

The sac is usually identified without difficulty, but an endoscopist can pass an esophagoscope into the sac until identification is positively made, at which time the scope is withdrawn and passed into the lumen of the esophagus proper. This provides a base on which dissection of the sac can be carried out and also prevents excessive excision of the esophageal wall, which could predispose the patient to subsequent stenosis. An esophageal bougie can be used for this purpose; its guidance into the esophageal lumen is facilitated by the surgeon's fingers. A nasogastric tube should also be passed for the purposes of feeding postoperatively.

Once the diverticulum is completely mobilized and held at a right angle to the long axis of the esophagus, its neck is dissected at the point where the mucosal sac protrudes through the muscular defect of the pharynx just above the cricopharyngeus muscle. Often a coat of attenuated muscle and connective tissue 3 to 4 mm thick covers the mucosal sac. Meticulous dissection at the junction of the cricopharyngeus muscle should be performed, and several layers of tissue may require division to expose the neck free of surrounding tissue. Care must be taken to not pull normal mucosa out through the deficiency and make this part of the sac, because division of this mucosa will predispose the patient to stricture postoperatively.

The neck of the sac is then clamped; once again the surgeon is careful to identify the recurrent laryngeal nerve. The sac is excised and can be closed in layers using No. 0-0 chromic catgut. The mucosal suture line is reinforced with vertical closure of the overlying muscle fibers.

If a clinical pharyngeal myotomy is to be perofmed, it is best done before excision of the neck of the sac. The left recurrent laryngeal nerve is again identified. The esophagus is rotated anteromedially and a right-angle forceps can be used to develop an extramucosal dissection plane just inferior to the neck of the diverticulum. The transverse fibers of the cricopharyngeus muscle and the lower fibers of the inferior constrictor muscle are then divided for a total length of about 5 cm, just off the posterior midline, allowing the mucosa to bulge.

A Penrose drain is used for 24 to 48 hours. Fluids by mouth are started shortly thereafter, and as soon as it is established that no fistula exists, the nasogastric tube is withdrawn.

Dohlman's procedure

The preoperative preparation for the endoscopic diathermy technique (Dohlman's procedure) is similar to that discussed previously. It may be useful, however, to have the patient swallow a black thread preoperatively in order to facilitate location of the esophageal

inlet during endoscopy.

The principle of endoscopic treatment is that the sphincter (ie, the cricopharyngeus muscle and the upper part of the septum between the diverticulum and the esophagus) is divided, and at the same time a more ample overflow from diverticulum to esophagus is effected. A bivalved esophageal speculum such as the one first devised by Dohlman (1960) is required. Insertion of the scope and location of the esophageal inlet with proper adjustment of the tissue bridge between the diverticulum and the esophagus require considerable experience and are the most difficult aspects of the procedure. The scope is inserted so that the anterior blade is in the esophagus and the posterior blade enters the pouch. As the instrument is advanced, the septum between the esophageal lumen and the sac forms a bulky horizontal mass containing the cricopharyngeal sphincter. To help alleviate bleeding, which can sometimes be troublesome, crushing the wall with an artery forceps, which is left on for 1 to 2 minutes before division, is useful. The jaws of the diathermy forceps are then applied to the crushed wall, and a coagulating current is applied until the tissues held by the forceps become blanched. The forceps is then withdrawn, and the diathermy knife is used to cut through the center of the coagulated tissue.

Recently van Overbeek et al (1984), strong advocates of the endoscopic technique, reported on the use of a modified esophagoscope in conjunction with the operating microscope to facilitate exposure of the tissue bridge and its subsequent division. They have also begun using the CO_2 laser instead of electrocoagulation, hoping for less tissue necrosis and scar formation. To date, however, they have found no inherent advantage to the laser over the conventional technique.

Complications

Complications may relate to the primary disease or be consequent to surgery. The first category is covered in the discussion on symptomatology. Because most people seek medical assistance before the disease is advanced, these problems rarely occur. The surgical morbidity and mortality are variously reported. The former has been reported to be from 0% to 50%. A realistic overall mortality rate is probably between 0.5% and 1.5%.

The complication most feared in surgical management is mediastinitis caused by leakage at the resection site. Prevention is the best treatment. Preoperative antibiotics, meticulous care in avoiding spillage, accuracy in regard to suture or staple replacement, and adequate drainage are important in avoiding this devastating problem. In the endoscopic method great care is taken to adequately isolate the party wall before diathermy and to ensure that no more than two thirds of it is coagulated. Temperature and pulse rates should be monitored postoperatively to detect evidence of fever or tachycardia. Should mediastinitis develop, after either the external or endoscopic operation, prompt and thorough drainage is indicated.

Other complications are more common but not of such great consequence and can usually be managed conservatively. Hemorrhage is uncommon with the endoscopic method but can be troublesome if it occurs. (Prevention is discussed under surgical technique.) In the external procedure it is important to not clamp blindly at a bleeding site because of the possibility of injury to an important structure such as the recurrent laryngeal nerve. If a

hematoma develops following closure (an uncommon occurrence since the advent of closed suction drainage), the wound is reopened, the hematoma evacuated, and the bleeding site controlled by suture or packing as appropriate.

Leakage is a nuisance but is usually not serious. Signs of excessive wound drainage should alert the surgeon to suspect leakage, or it may be discovered on a postoperative radiographic study. Drains should be left in place and the patient fed nothing by mouth for 7 to 10 days. If the leak persists, some alternative methods of feeding should be instituted. Once a well-formed tract has developed, the patient may resume oral feeding. All fistulas eventually close spontaneously, usually within 1 month, but occasionally closure takes as long as 3 months.

Local infection or abscess at the wound site may develop when a leak occurs. In this situation the wound should be opened and drained as necessary, irrigated, and packed. It is not advisable to attempt secondary repair at this time, nor is it usually necessary. Delayed closure of the wound is performed as appropriate.

Recurrent laryngeal nerve palsy is an occasional complication following the external approach. It is more common with larger sacs where much dissection occurs in the tracheoesophageal groove. For that reason, as previously mentioned, it is best to positively identify the nerve at the time of surgery. In most cases the damage is only temporary, but permanent paralysis occasionally occurs and is reported in almost every series.

Surgical emphysema is common. It is localized in the neck and is rarely a serious problem. Observation is all that is usually required, and resolution occurs within a few days.

Esophagostenosis and recurrent diverticula are two complications that may lead to long-term morbidity. As previously indicated, recurrence that is evident radiologically does not necessarily imply symptoms. Patients with minimal dysphagia can often manage without further treatment. A sizable recurrent diverticulum that is symptomatic and allows retention of food and fluid should be surgically removed. Stenosis can be avoided by using proper surgical technique. Significant dysphagia persisting longer than 3 weeks postoperatively is indicative of some degree of narrowing, and treatment is required. The diagnosis may be confirmed radiographically to rule out persistence or recurrence of the diverticulum. Esophagostenosis is managed by endoscopic dilatation or bougienage as frequently as necessary. Severe stenosis or stricture may require a recontructive procedure.

Epiphrenic Diverticula

Epiphrenic diverticula or supradiaphragmatic diverticula are uncommon and probably constitute less than 10% of esophageal diverticula. The male-to-female ration is 2 to 1. These diverticula are located just above the diaphragm, usually within 10 cm of the cardia, but the exact site is variable.

Etiology

Most epiphrenic diverticula are pulsion in type, but traction and combined forms do occur. Protrusion of the mucosa and submucosa develops through a muscular defect in the esophageal wall. The site is usually posterior with extension of the sac into the right thoracic cavity but it may protrude through the anterior esophageal wall or off to the left. Occasionally multiple diverticula are present.

Epiphrenic diverticula usually occur in conjunction with other esophageal or diaphragmatic disease. This contributes both to their development and symptomatology (Cross et al, 1961; Garcia et al, 1972; Payne and Clagett, 1965). Habien et al (1956) found associated lesions of the esophagus or diaphragm in 50% to 70% of patients. The most common disorders are hiatus hernia, achalasia, diffuse esophageal spasm, and reflux esophagitis. There may be coexistent diverticula at other sites. Eventration of the diaphragm and carcinomas of the esophagus and of the epiphrenic diverticulum occur rarely (Habien et al, 1956; Postlethwait, 1979). It is therefore thought that epiphrenic diverticula represent abnormal esophageal function.

Cross et al (1961) stated that "pulsion diverticula suggest increased intraluminal pressure to which has been added a weak area in the musculature of the esophageal wall for out-pouching of the mucosa". The motility disorders they found included diffuse increase in lower esophageal tone, hyperactive and incoordinate peristalsis, esophageal spasm, delayed opening of the esophageal sphincter, and reflux esophagitis. Whatever the case, the common underlying problem is basically the same: normal or hyperactive peristalsis acting against a functional obstruction, with resulting increase in intraluminal pressure being dissipated by formation of a diverticulum through an esophageal wall that cannot withstand the high pressure.

Clinical features

Patients with epiphrenic diverticula are frequently asymptomatic. When symptoms do occur they are likely to be caused by the associated underlying motility disorder. However, as the sac becomes larger and food preferentially enters the lumen, it may produce additional symptoms that are directly related to the size of the sac. (Epiphrenic diverticula as opposed to hypopharyngeal diverticula may attain considerable size before emptying becomes a problem.) Dysphagia is common but rarely severe, and weight loss is uncommon. Belching and regurgitation of undigested food (which is often described as tasting sweet) is aggravated by positional change and lying down. Because of the distal location of the epiphrenic diverticulum, this problem is much less common that it is with hypopharyngeal diverticula. For the same reason aspiration and pulmonary complications are also lesser threats. Pain is occasionally a significant complaint. It is substernal and referred to the lower chest, epigastrium, or back between the scapulae. Other symptoms include hiccups, heartburn, cough, and bad breath. Bleeding is a rare complication, and although carcinomas in the diverticula and the distal esophagus have rarely been reported, this possibility should be kept in mind if bleeding occurs (Allen and Clagett, 1965; Postlethwait, 1979).

Complications

Complications may occur with a large sac and are usually secondary to aspiration and subsequent pneumonitis. A large diverticulum can also cause complete esophageal obstruction by extrinsic pressure; it may even compromise lung function. As previously mentioned, carcinomas within the diverticulum have been recorded. Benign lesions have also been found and include fibromas and myomas in the sac wall (Postlethwait, 1979). Ulceration of the wall may also occur. A fortunately rare but serious complication is spontaneous perforation (Stalheim, 1978), which leads to mediastinitis and is often fatal.

Diagnostic assessment

The diagnosis may be made incidentally during investigation for associated esophageal dysfunction or because the clinician is suspicious on the basis of history. Radiography is the definitive diagnostic tool. On barium swallow examination the diverticulum will be demonstrated as a distended, barium-filled sac above the diaphragm, usually protruding to the right (Fig. 130-6). Cinefluoroscopy and positional maneuvers are used to demonstrate associated esophageal disease, such as tertiary contractions, spasm, or a hiatus hernia. Pseudodiverticula may develop, but these are differentiated because they are not persistent in location on subsequent examination. Motility studies should be done to determine the exact nature of the underlying motility disorder and as an adjunct to radiography. This information aids in proper treatment of the underlying esophageal disease, which is most often the only treatment necessary. Endoscopy is useful to confirm and assess the degree of esophageal inflammation and obstruction, to exclude a carcinoma or other disease, and, especially if surgery is a consideration, to remove debris from the sac (a previously swallowed thread is a good guide for safe instrumentation).

Management

Management of epiphrenic diverticula is conservative and largely directed toward correcting the underlying esophageal disorder. Postural maneuvers and liquids following meals may help empty the sac, and bland diets, antacids, and histamine (H2) blockers can be used to treat ulceration. Esophageal dilatation may be necessary for spasm or organic obstruction.

Surgery is indicated only for severe progressive symptoms and significant increase in size of the diverticulum. A left thoracotomy approach is recommended because it provides the best access to the diverticulum itself and allows for repair of associated disorders, which must be corrected to prevent recurrences. Even if a motility disorder was not equivocally demonstrated preoperatively and even if the esophageal musculature is not greatly hypertrophied at surgery, a long extramucosal myotomy is recommended. Fewer complications such as leakage at the suture line and fewer recurrences have been clearly demonstrated than if this procedure was omitted (Allen and Clagett, 1965).

Complications of surgery include leakage at the suture line with empyema, an esophagobronchial fistula, and hemorrhage. Since concomitant myotomy has become a routine part of the procedure, the approximate recurrence rate is only 5% (Postlethwait, 1979).

Midthoracic Diverticula

Midthoracic (midesophageal or peribronchial) diverticula are traditionally considered traction diverticula and are composed of all layers of the esophageal wall, including the muscular coat. They are located in the anterior esophageal wall at or near the tracheal bifurcation. They are the most common type of diverticulum but are usually small and asymptomatic.

Etiology

In 1922 Kragh examined 51 traction diverticula by serial section and found the majority demonstrated adherence to tuberculous mediastinal lymph nodes.

In the 1960s cases of pulsion-type diverticula of the mid-esophagus were reported (Belsey, 1966; Cross et al, 1961), and this set the stage for new concepts in the etiology of midesophageal diverticula. Current thinking relates these diverticula, like those in the upper and lower esophagus, to associated esophageal motor disorders in the majority of cases. This is not to say that traction diverticula secondary to tuberculosis or other granulomatous adenitis still cannot occur, but tuberculosis is probably an uncommon cause today. Traction diverticula of the upper esophagus located below the cricopharyngeus muscle have been reported secondary to a cervical esophagostomy (Kaplan et al, 1982). This condition may become more common with frequent use of cervical esophagostomies.

Belsey's radiographic studies (1966) supported the new concept that uncoordinated esophageal contraction and diffuse spasm in the midthoracic esophagus are associated with formation of pulsion diverticula in high-pressure zones between spastic segments. Cross et al (1961), using cinefluoroscopy and intraluminal pressure studies, also concluded that these diverticula represent the end products of severe neuromuscular imbalance. A favorable site and increased luminal pressure are the necessary prerequisites. These investigators failed to find the usual contour ascribed to traction diverticula in most of the cases they studied and introduced the concept of a contributing congenital component. It was thought that a likely mechanism leading to weakness in the esophageal wall is a residual congenital attachment of the esophagus to the tracheobronchial tree in the region of the carina, especially because this is also the site of congenital tracheoesophageal fistulas. Further support for the congenital theories is found in a report by Ishigami et al (1965).

Kaye (1974) also found a high incidence of esophageal motor dyfunction in patients with midthoracic diverticula, but in only one was there evidence of tuberculosis, consisting only of small, scattered, calcific densities without obvious mediastinal involvement.

Clinical features

The neck of the diverticulum is broad, and the esophageal lumen is not compromised. Emptying can occur easily, and the intact muscular wall prevents progressive increase in the size of the sac. Therefore these lesions are usually asymptomatic and often are found incidentally during a routine examination of the esophagus. Even the true traction type may occur in association with other esophageal disease, and symptoms may arise secondary to these other processes. Certainly with the pulsion type, in which a high incidence of

esophageal dysfunction is found, this is likely to be the case. Nevertheless, patients may compain of substernal pain or dysphagia; bleeding occurs rarely. Expectoration of calcium or other symptoms of broncholithiasis is also possible (Payne and Clagett, 1965).

Complications

When tuberculosis was more prevalent, necrosis of inflammatory tissue occasionally cause erosion of blood vessels with bleeding into the esophagus or tracheobronchial tree. When the diverticula are pulled to the left at the aortobronchial level by fibrotis mediastinal nodes, recurrent laryngeal nerve paresis may develop (Lerner and Katz, 1975). Acquired tracheoesophageal or bronchoesophageal fistulas, which rarely close spontaneously, may also occur with subsequent serious suppurative lung disease. Rupture may cause fatal mediastinitis (Lynn, 1975). A pulsion diverticulum has the capacity to balloon out and obtain a dependent position, thereby growing larger and allowing stagnation of food, esophageal obstruction, and secondary inflammatory complications.

Diagnostic assessment

As with all diverticula, the diagnosis is based primarily on radiographic findings. A traction diverticulum shows a wide mouth, usually in a transverse location, and unimpaired emptying (Fig. 130-2). The pulsion type is more globular with a narrower neck. It frequently projects inferiorly, thereby retaining barium and emptying less readily. Traction diverticula extent anteriorly and usually to the right, just below the bifurcation of the trachea, the usual site of granulomatous lymphadenitis. Calcification in the mediastinum or elsewhere in the chest should be looked for. The pulsion type may be based higher, around the aorta, but may occur anywhere in the thoracic segment and may be multiple.

Esophagoscopy may be indicated to assess the status of the esophageal lumen or wall and for the detection of possible complications. In the case of a fistula, bronchoscopy more often identifies at least one end of the tract. Manometry, although not necessary for the diagnosis, identifies associated motility disorders.

Management

Uncomplicated traction diverticula of the midesophagus rarely require treatment, although complications that may be life threatening obviously need surgical intervention. Pulsion diverticula with their greater propensity to a narrow and a dependent sac that does not readily empty should be managed surgically. In most situations a transthoracic approach is used.

Esophageal Intramural Pseudodiverticulosis

Esophageal intramural pseudodiverticulosis (EIPD) is a rare condition of unknown origin characterized by multiple small, flask-shaped out-pouchings in the esophageal wall. These out-pouchings are actually dilated excretory ducts of mucous glands.

The etiology and pathogenesis remain uncertain, although it is generally thought to be an acquired condition. One theory suggests that increased intraluminal pressure around an esophageal stricture initiates the process of ductal dilatation. However, most patients with esophageal strictures do not have EIPD, and a few patients affected with pseudodiverticulosis are not affected with an esophageal stricture. Furthermore, when the two are coincident, the diverticula are often found below the level of the narrowing, which would seem to rule out mechanical pressure as an initial factor.

Lupovitch and Tippins (1974) advocated the "adenosis" theory. An unknown factor leads to proliferation of submucosal glands, which predisposes to recurrent and chronic inflammation. Dilatation causes the glandular structures to eventually reach macroscopic proportions. Secondary hyperplasia and metaplasia of the glandular and ductal epithelium occur, predisposing the patient to further infection, especially fungal infections. The resultant findings of submucosal fibrosis, segmental stricture, and paraesophageal adhesions can well be explained by this sequence. Glandular proliferation has not been demonstrated in every case, however, and therefore the implication of submucosal glands in the pathogenesis of EIPD awaits further documentation based on pathologic studies.

Possibly an increase in viscosity of the duct contents is responsible for ductal obstruction and subsequent dilatation. The fact that at endoscopy or on inspection of specimens a thick, creamy liquid is expressible from the orifices supports this hypothesis. On microscopic examination mucus is replaced either by epithelial cells and leukocytes or pseudohyphae of *Candida albicans* (which has a high association with EIPD).

The ductal orifices may be compressed by periductal infiltrates and/or fibrosis. The end result in all cases is the same: dilatation of the excretory ducts of the mucous glands. Many associated conditions found in esophageal intramural pseudodiverticulosis cause submucosal fibrosis, ductal obstruction, and strictures. In a review of 53 patients esophageal reflux was seen radiographically in 19%, and esophagitis was demonstrated endoscopically in 59% (Bruhlmann et al, 1981). *Candida albicans* is frequently cultured. Whether it is an etiologic factor or an opportunistic infection remains to be determined. Many of the patients also have diabetes or are chronic alcoholics. In a series of 43 patients reported by Cho et al (1981) 20% had associated malignant neoplasms. Several other conditions predisposing to host compromise have been reported less frequently.

The disease has been recorded from the first through the ninth decade, but the peak incidence is in the sixth and seventh decades. Men are affected more often than women in most series, and there seems to be a slightly higher incidence in the black population.

Clinical features

The outstanding clinical feature is dysphagia, which may be either chronic or acute and constant, progressive, or intermittent. Some patients complain of an acute bolus obstruction that usually resolves spontaneously.

Diagnostic assessment

The diagnosis is primarily by radiographic examination. Barium esophagography is excellent in this regard, and computed tomography (CT) scanning is a useful adjunct.

The radiographic findings are quite characteristic but do show great variation in the localization and severity of visible changes. The typical appearance consists of multiple small (1 to 5 mm deep), flask-shaped out-pouchings that are parallel to the long axis of the esophagus (Fig. 130-7). Most have a rounded end, but occasionally they branch in a rootlike fashion, forming bridges with adjacent diverticula. Stenotic and strictures areas are present in up to 90% of cases. These areas are longer and are situated higher in the esophagus than in typical peptic stenosis. The diverticula are usually more numerous at these sites, although the esophagus generally is involved diffusely along its entire length. When only localized segments are involved, however, no predilection for the upper, middle, or lower third is apparent. Additional radiographic findings such as a hiatus hernia, esophageal reflux, webs, and motility disorders may be demonstrated. On barium swallow examination the main differential diagnosis is esophageal candidiasis, which is often coexistent, thereby causing confusion. In the presence of *Candida* alone, however, out-pouchings are larger and mucosal irregularity is more prominent.

CT scanning demonstrates thickening of the esophageal wall consistent with submucosal inflammatin and fibrosis. The irregular and edematous mucosa caused by associated esophagitis is prominently displayed. Such findings, especially in association with loss of soft-tissue planes, may be a feature of extensive esophageal neoplasm, but the many small, intraluminal gas collections that correspond with the pseudodiverticula help make this distinction.

Endoscopy may reveal the orifices of the pseudodiverticula, which are described as pin-sized, yellowish white elevations with thick, creamy liquid expressible from the center. Sometimes the lesions appear as subepithelial cysts without visible openings or as tiny punctate depressions. Most patients have general changes of acute or chronic esophagitis as well as strictures or stenotic segments.

Management

Despite speculation as to the etiology and pathogenesis of esophageal intraluminal pseudodiverticulosis, dilatation of associated strictures, which are present up to 90% of the time, is the treatment of choice. Bruhlmann et al (1981) and Cho et al (1981) produced symptomatic relief in 94% and 96% of patients, respectively, with this therapy. Candidiasis, when present, should be treated with nystatin (Mycostatin). Dietary control and antacids offer symptomatic relief.