Chapter 188: Differential Diagnosis of Neoplasms of the Posterior Fossa

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Cerebellopontine angle (CPA) lesions are the predominant skull base neoplasms that affect the posterior fossa. Although acoustic neuromas (AN) account for the majority of primary neoplasms, a wide array of other lesions must also be considered in the differential diagnosis. Improved diagnostic capabilities facilitate routine diagnosis of such tumors while they are much smaller than previously detectable.

Posterior fossa skull base neoplasms may be grouped into four basic categories: common CPA lesions (including internal auditory canal lesions), petrous apex lesions, rare CPA lesions, and intraaxial lesions. This chapter describes the signs, symptoms, and diagnostic procedures in the differential diagnosis of the tumors within the described categories (see box on p. 3272). The surgical treatment of these lesions is described in Chapter 192.

Box: Differential diagnosis of skull base neoplasms involving the posterior fossa

Common CPA lesions	*Uncommon CPA lesions*
Acoustic neuroma	Metastatic tumors
Meningioma	Lipomas
Epidermoid	Dermoids
Nonacoustic neuroma	Teratomas
Paraganglioma	Chordomas
Arachnoid cyst	Chondrosarcomas
Hemangiomas	Giant cell tumors
Petrous apex lesions	*Intraaxial tumors*
Cholesterol granuloma	Hemangioblastoma
Epidermoid	Medulloblastoma
Asymmetric pneumatization	Astrocytoma
Retained mucus or mucocele	Glioma
Petrous carotid artery aneurysm	Fourth ventricle tumor.

Common Cerebellopontine Angle Neoplasms

In the House Ear Clinic's series of CPA neoplasms, ANs are the most common tumors accounting for over 90%. The remaining primary tumors were meningiomas (3%), primary cholesteatomas (2.5%), and facial nerve schwannomas (1%) with less common tumors composing the remaining lesions (Brackmann and Bartels, 1980). When secondary tumors are also considered, paragangliomas constitute up to 10% of CPA neoplasms (Valvanis et al, 1987). Because AN is the most common lesion, it will be described in detail; only the distinguishing characteristics for the other skull base lesions are discussed.

Acoustic neuroma

AN refers to a benign schwannoma of the eighth nerve. These lesions are relatively common and constitute 8% to 10% of all intracranial tumors. ANs arise principally from the vestibular division of the nerve near Scarpa's ganglia; the superior and inferior divisions are equally affected (Kartush et al, 1987). ANs are slowly growing neoplasms that originate in the nerve sheath and consist of schwann cells in a collagenous matrix. They are typically circumscribed and encapsulated grossly encroaching on and displacing neural structures without direct invasion. Their consistency varies from firm and dense to soft with large cystic spaces.

ANs usually arise near the myelin-glial junction near the porus acusticus. Thus these lesions usually arise within the internal auditory canal (IAC); however, they occasionally develop in the CPA medial to the porus. Because ANs produce symptoms by exerting pressure on surrounding neurovascular structures, auditory and vestibular symptoms develop earlier with tumors of the IAC than with tumors of the CPA.

Of the 2000 to 3000 ANs diagnosed annually in the USA, over 95% occur as unilateral and nonhereditary lesions. The remaining ANs occur as manifestations of the neurofibromatoses, which consist of at least two distinct genetic disorders. Neurofibromatosis (NF) type 1 (von Recklinghausen's disease) is a relatively common autosomal dominant disorder with variable penetrance and an incidence of 1 in 4000 live births. NF type 1 neuromas occur throughout the body, both intracranially and extracranially from the Schwann's cells of any nerve; however, less than 5% of affected individuals develop ANs, and bilateral AN is not a part of the syndrome. NF type 2 is the central form of neurofibromatosis characterized by bilateral AN in up to 96% of patients (Martuza and Eldridge, 1988). The precise frequency of NF type 2 is unknown, but it is far less frequent than NF type 1. Unlike NF type 1, in which the gene deficit is localized to chromosome 17, the gene defect in NF type 2 is on chromosome 22. ANs in NF type 2 are characterized by an onset early in life, often before the age of 21 years, as opposed to unilateral lesions, the majority of which occur between 40 and 60 years. Thus ANs appearing before the age of 30 years mandate particularly close evaluation of the contralateral ear. Although the ANs in NF type 1 and 2 resemble the lesions in nonhereditary ANs, they are technically more challenging to remove because of a tendency to adhere to nearby structures. The clinical presentation of AN in NF is identical to that of unilateral AN.

Malignant schwannomas may rarely occur; 40% to 50% of them are associated with NF, but they may also occur with solitary schwannomas. Another very uncommon variant is a pigmented schwannoma (Brackmann and Gherini, 1986).

Natural history

The growth rate of ANs is extremely variable. They are generally slow-growing tumors, with average growth rates from 0.25 to 0.4 cm/year; however, growth rates in excess of 2 cm/year have been documented. Untreated ANs are potentially lethal tumors. Gradual enlargement can occur, leading to indentation of the brainstem, increased intracranial pressure, and death over a course of 5 to 15 years (Jackler et al, 1990).

Growth of ANs generally occurs in three phases: IAC, cisternal, and brainstem compression. IAC growth results in acoustic and facial nerve compression and attenuation. Displacement of the seventh nerve, acoustic nerve, and anterior inferior cerebellar artery (AICA) occurs just medial to the porus acusticus (cisternal portion). Medial tumor growth augments the blood supply of the tumor with bridging vessels from the brainstem surface. Fourth ventricle shift often occurs when the CPA component reaches 2 to 3 cm and total ventricle obstruction with resulting hydrocephalus occurs with continued tumor growth. Trigeminal compression occurs at about the 3-cm stage, which permits the superior portion of the tumor to abut the fifth nerve.

Signs and symptoms

Sensorineural hearing loss (SNHL), tinnitus, dysequilibrium, and facial hypesthesia are, in order, the most common symptoms of AN. Although progressive unilateral SNHL is the most common symptoms, loss of speech discrimination in particular is characteristic of retrocochlear dysfunction of the cochlear nerve - presumably from pressure on the auditory nerve. The symptoms are usually slowly progressive, with a median duration of 2 years in one series (Kasantikul et al, 1980). However, up to 20% of patients with AN experience sudden SNHL, which may totally recover (Sataloff et al, 1985). Overall, only 1% of patients with a sudden SNHL are found to have AN. On the other hand, up to 5% of patients with AN have normal hearing (Beatty et al, 1987).

The dysequilibrium associated with acoustic tumors is usually a mild balance disturbance, which is often so minor that the patient does not mention it, and only careful questioning elicits it. Rotatory vertigo is far less common.

Diminished facial sensation or corneal reflex results from compression of the fifth cranial nerve, which is more likely with medium and large tumors. As large ANs compress the fourth ventricle and brainstem, long tract signs, ataxia, and even findings of increased intracranial pressure such as headaches and nausea are produced.

Diagnostic studies

Two categories of studies are performed in the diagnosis of ANs. Auditory and vestibular studies assess the functional integrity of the audiovestibular system, whereas imaging studies are performed for the definitive anatomic diagnosis.

Audiometry. Routine air, bone, and speech discrimination studies are the first that may suggest the possibility of an AN. Asymmetric SNHL or impairment of speech discrimination disproportionate to the pure tone loss requires specific investigation for a retrocochlear lesion. In the past the retrocochlear audiologic test battery included short increment sensitivity index (SISI), tone decay tests, and Bekésy audiometry. These tests are not sensitive or specific enough to detect ANs, and auditory brainstem response (ABR) testing and acoustic reflex testing have replaced them as the principal screening tests for ANs.

Auditory brainstem response audiometry. In patients with hearing sufficient to generate an adequate ABR response, this study is the most sensitive to detect tumors, with detection rates ranging between 95% and 100% (Brackmann and Kwartler, 1990). The details

of ABR testing are described elsewhere (Selters and Brackmann, 1977). Briefly, the brainstem response to an 83 dB broad-band click is recorded while the contralateral ear is masked by 78 dB white noise. The latency for detection of wave V for the two ears is compared, and an adjusted interaural latency for wave V greater than 0.2 msec is considered abnormal. There is approximately a 10% false-positive rate for patients with sensorineural hearing impairments who do not have a CPA tumor.

Acoustic reflex testing. When ABR is not available, acoustic reflex testing is a valuable screening tool for ANs. Nearly 88% of patients with confirmed AN will have no acoustic reflex (Johnson, 1979) or acoustic reflex decay (Jerger et al, 1987).

Vestibular tests. Vestibular testing is no longer considered a useful screening test for AN. Electroneurography (ENOG) and infrared (IR) video caloric testing are helpful in defining whether the tumor arises from the superior or inferior vestibular nerve (Linthicum et al, 1988). Such information is valuable when considering hearing preservation surgery for ANs, as the possibility of hearing preservation is higher with superior vestibular nerve tumors.

Imaging studies. Conventional radiography and polytomographic studies of the IAC are no longer routinely used in the diagnosis of CPA tumors, because the sensitivity of these studies is only 90% at best.

Computed tomographic (CT) scans and magnetic resonance imaging (MRI) with the paramagnetic intravenous contrast agent gadolinium (Gd)-DTPA are the principal imaging modalities for CPA lesions.

Table 188-1. CT/MRI differential diagnostic categories of CPA lesions

Location	Incidence	Type
Extraaxial	Most common	Acoustic neuroma
	Common	Meningioma
	Common	Epidermoid (and other cysts: arachnoid, cysticercal, dermoid)
	Rare	Nonacoustic neuromas (V, VII, IX, X, XI, XII)
	Rare	Vascular lesions (loops, aneurysms, malformations)
Extradural	Common	Paraganglioma (glomus jugulare, vagale)
	Rare	Bone lesions (benign or malignant; primary or meta)
Intraaxial	Rare	Astrocytoma, ependymoma, papilloma, hemangioblastoma, metastasis.

Lo (1991) has proposed a scheme for the CT/MRI differential diagnosis of CPA lesions by considering them anatomically (extraaxial, extradural, or intraaxial) and according to incidence (rare or common). Table 188-1 outlines CPA lesions according to those categories. In adults and teenagers, ANs, meningiomas, and epidermoids are the three most common lesions. ANs are very rare in children; instead, brainstem gliomas (which can enlarge the IAC) are the most common CPA lesion in younger children (Segall et al, 1982). Distinguishing among the three most common CPA lesions is based on specific imaging characteristics for these lesions with CT and MRI as summarized in Table 188-2.

Table 188-2. Imaging features of the three most common CPA lesions

Feature	**Acoustic neuroma**	**Meningiomas**	**Epidermoids**
Location	Centered on IAC	Eccentric to IAC	Anterolateral or posterolateral to brainstem
Bone changes	Most enlarge IAC	Occasional hyperostosis	Occasional erosion
Shape	Spherical or ovoid	Acute bone-tumor angle	Hemispherical, rarely plaquelike, may herniate through tentorium, obtuse bone-tumor angle	Variable - tends to dumbbell into middle fossa or contralateral CPA
CT density	Mostly isodense	Slightly hypodense	Some calcified	Mostly hypodense	Occasional peripheral calcium
CT enhancement	Moderate to marked	Often inhomogeneous	Marked and homogeneous	Nonenhancing
T1 MRI	Isointense or hypointense	Isointense or hypointense	Hypointense
Gadolinium enhancement	Marked	Moderate	Nonenhancing

T2 MRI
Isointense or hypointense
Variable
Hyperintense.

Computed tomographic scans. CT scans revolutionized the diagnosis of CPA tumors. Until recently, the most sensitive test for ANs were contrast-enhanced CT scans with iodinated contrast materal for tumors larger than 1.5 cm and oxygen contrast for intracanalicular lesions. The CT finding characteristic of an acoustic tumor is an ovoid lesion centered on the internal canal with moderate enhancing qualities. The tumor is often not homogeneous, exhibiting areas of lesser and greater enhancement (Fig. 188-1). Approximately 85% of ANs show acute angles at the bone-tumor interface, in contrast to meningiomas in which the interface is obtuse in 75% of tumors (Lo, 1991).

Intravenous contrast CT scans can fail to detect ANs smaller than 1.5 cm. Oxygen cisternography was used to diagnose small, mainly intracanalicular lesions. In this technique, 4 cc of oxygen were injected into the subarachnoid space through a lumbar puncture to highlight the structures of the IAC. This technique was very sensitive for small lesions (Fig. 188-2); however, it was limited by a false-positive rate of 5% and the need for a lumbar puncture (Barrs et al, 1984). In 1991, oxygen cysternography was not routinely used for diagnosing small ANs by the members of the House Ear Clinic. Air-contrast CT is still used for the diagnosis of IAC vascular loop compression (Fig. 188-3).

Magnetic resonance imaging. Gd-contrast MRI is sensitive and specific for AN, and it is now the diagnostic imaging technique of choice in the evaluation of AN and other CPA lesions (Press and Hesselink, 1988) (Fig. 188-4). AN images on MRI are isointense or mildly hypointense to brain on T1 images and mildly hyperintense to brain on T2 images (Lo, 1991). MRI scans for possible AN should be performed with intravenous contrast. Gd increases the diagnostic sensitivity of MRI on T1-weighted images (Fig. 188-5), and lesions as small as 3x3 mm have been reported (Press and Hesselink, 1988). In addition to improving sensitivity in diagnosing ANs, MRI is noninvasive, and the patient receives no radiation.

Meningiomas

Meningiomas represent up to 18% of all intracranial tumors and approximately 3% of CPA tumors (Rubenstein, 1972). The cells lining the arachnoid villae are the cells of origin; these cells are distributed throughout the intracranial space predominantly in relation to veins and dural sinuses. Meningiomas are benign but locally aggressive tumors, whcih occur at different anatomic sites in the following order of frequency: parasagittal region, falx, convexity, olfactory groove, tuberculum sellae, sphenoid ridge, petrous face (CPA), tentorium, lateral ventricle, clivus and others (Langman et al, 1990).

The gross appearance is typically a globular mass that is firmly adherent to the dura matter, with characteristic speckles scattered throughout the tumor that correspond to the microscopic psammoma bodies. The tumor displaces but does not invade adjacent neural tissue and has a thin investing capsule. Meningiomas can invade bone without destruction by extension along haversian canals. Adjacent bone is hyperostotic in 25% of cases (Langman et al, 1990).

Many histopathologic classifications have been proposed, but a single, widely used system distinguishes among (1) syncytial, (2) transitional, (3) fibrous, (4) angioblastic, and (5) sarcomatous types. The specific histopathologies and growth characteristics are reviewed elsewhere (Russell and Rubenstein, 1977).

In the posterior fossa, meningiomas usually arise on the posterior surface of the petrous bone, away from or at the edge of the IAC, or along the sigmoid sinus. Because they usually arise outside of the IAC, they may become large before producing signs and symptoms of CN VIII compression. Most eventually do involve CN VIII.

Signs and symptoms

Audiovestibular symptoms are usually the first indication of a posterior fossa meningioma. Among patients presenting to neurosurgeons, a higher proportion first experienced trigeminal symptoms (Sekhar and Jannetta, 1984). The signs and symptoms of meningiomas are similar to those of ANs. Small tumors produce hearing loss, tinnitus, and imbalance. Larger tumors also produce signs and symptoms of other cranial nerve involvement and hydrocephalus.

Diagnostic studies

Audiovestibular testing. It is impossible to distinguish ANs from meningiomas on the basis of audiovestibular testing. As meningiomas cause CN VII compression, auditory and vestibular tests become abnormal in a similar pattern with ANs. Because meningiomas usually do not arise within the IAC, the sensitivity of audiometric and vestibular testing is lower with meningiomas than ANs. For instance, only 75% of patients with meningiomas have an abnormal ABR.

Imaging. Conventional radiography and polytomography is of even less value with meningiomas than ANs, as meningiomas arise outside of the IAC, and petrous pyramid radiographs are normal.

Table 188-2 summarizes features of meningiomas that assist in differentiating them from ANs. Unlike ANs, meningiomas are usually eccentric to the porus. Whereas ANs seldom herniate into the middle fossa, about 60% of meningiomas extend to the middle fossa (Lo, 1991). Meningiomas are usually hemispheric due to their broad-based attachment to the posterior petrous wall, accounting for the obtuse bone-tumor angles found in 75% of meningiomas. Unlike the size of origin of ANs, that of posterior fossa meningiomas is varied (Fig. 188-6).

Computed tomography. On CT approximately two thirds of meningiomas are hyperdense relative to the brain. Unlike ANs, meningiomas are homogeneous and occasionally calcified. They demonstrate homogeneous enhancement with iodine infusion, which usually permits differentiation from ANs (Fig. 188-7). Hyperostosis of adjacent bone is infrequent but characteristic of meningiomas when present.

Magnetic resonance imaging. On MRI, meningiomas are extremely variable in intensity on T2 images and either isointense or slightly hypointense to brain on T1 images (Fig. 188-8). The different signal intensities among meningiomas correspond to different histopathologic subtypes (Elster et al, 1989). Surface flow-voids on MRI correspond to marginal pial blood vessels, and arborizing flow-voids represent active feeders to the tumor. Calcification and cystic foci cause heterogeneity on MRI images of meningiomas.

Primary cholesteatomas (epidermoids)

Primary cholesteatomas consist of stratified squamous epithelial linings surrounding desquamated keratin, which originates from epithelial rests within the temporal bone or CPA. These lesions are usually slow growing, and symptoms often do not become apparent until the second to fourth decade of life. As they expand, compression and irritation of surrounding structures produce the signs and symptoms of this lesion.

Primary cholesteatomas occur adjacent to the brainstem. Because they expand into the area of least resistance, primary cholesteatomas have variable shapes with irregular surfaces. They may burrow into crevices on the surface of the brain or dumbbell into the middle fossa.

Signs and symptoms

Although primary cholesteatomas produce CN VII and VIII nerve pressure with continued enlargement, these lesions can become quite large without producting any symptoms. Facial twitching is a distinguishing feature of primary cholesteatomas. Progressive facial paralysis is more common with these lesions than with schwannomas.

Diagnostic studies

Auditory testing. Auditory testing does not show any distinguishing features. Like other retrocochlear lesions, speech discrimination is poorer than degree of pure-tone loss. ABR is also frequently abnormal in primary cholesteatomas (Brackmann and Anderson, 1979).

Imaging. CT and MRI provide complementary information that is useful to differentiate between primary cholesteatomas and other lesions. Epidermoids originating in the CPA must be distinguished from arachnoid cysts, and epidermoids in the petrous apex must be distinguished from the much more common cholesterol granulomas. This distinction affects therapy because adequate treatment for primary cholesteatoma requires excision, whereas drainage is sufficient for cholesterol granulomas and arachnoid cysts.

Computed tomography. On CT scans primary cholesteatomas are less dense than brain (approximately CSF density) and exhibit no enhancement with intravenous contrast. These lesions have irregular margins and are eccentric to the porus acusticus (Fig. 188-9). Enhancing components suggest an associated malignancy (Garcia et al, 1981).

Magnetic resonance imaging. With MRI, primary cholesteatomas are inhomogeneous and hypointense to brain on T1 images, and homogeneous and isointense or hyperintense to brain on T2 images (Fig. 188-10). Schwannomas, meningiomas, and chondromas are similar to primary cholesteatomas by intensity criteria; however, epidermoids are differentiated because they are nonenhancing.

Arachnoid cysts of the CPA are difficult to distinguish from epidermoids on both CT and MRI. Both lesions are of CSF density and nonenhancing; however, arachnoid cysts have smoother surfaces than primary cholesteatomas, and MRI multiecho sequence studies may differentiate the intensity of the epidermoid from the CSF intensity of the arachnoid cyst.

Facial nerve neuroma (schwannoma)

Facial nerve neuromas are uncommon benign neoplasms of Schwann's cells that may arise anywhere along the course of the facial nerve.

Signs and symptoms

Symptoms associated with these tumors depend on the portion of the nerve affected by the neoplasm. Peripheral involvement can present as a parotid mass; middle ear involvement can produce conductive hearing losses, and IAC or CPA involvement may result in sensorineural hearing loss. Unlike hemangiomas of the facial nerve, schwannomas do not produce facial weakness until the tumors are very large. Sometimes a facial nerve tic is evident, which helps distinguish facial neuromas from ANs but not from primary cholesteatomas. Notably, infratemporal lesions, with the possibility of neural entrapment, are more likely to result in facial paralysis than CPA lesions (Dort and Fisch, 1991).

Diagnostic studies

Auditory testing. The mechanisms for conductive and sensorineural hearing impairment have already been described. Impedance testing may reflect motor fiber impairment on ipsilateral reflex testing or CN VIII involvement on contralateral reflex testing. ABR testing of tumors arising in the IAC demonstrates abnormalities similar to those seen with AN (Selters and Brackmann, 1977).

Electroneurography. Electroneurography (ENOG) measures the muscle response to a maximal bipolar stimulation of the facial nerve near the stylomastoid foramen (see Chapter 149). ENOG potentials may be reduced in facial nerve neuromas even when there is no facial weakness or tic, whereas ENOG remains normal in AN until the tumor becomes very large.

Imaging. Intratemporal facial nerve lesions may produce bone destruction. Because the clinical presentation and audiometric studies in facial neuroma are not distinct, CT and MRI are the mainstays of diagnosis.

Computed tomography. Because facial nerve neuromas are histologically identical to ANs, they have the same enhancement characteristics. It is not usually possible to distinguish these lesions with the IAC. Anterosuperior erosion of the IAC or erosion of the labyrinthine facial nerve canal if present may be the only diagnostic clue (Lo, 1991). More distal tumors

enlarge the geniculate ganglion and fallopian canal (Fig. 188-11).

Magnetic resonance imaging. Just as with CT, MRI of facial neuroma produces imaging characteristics identical to those of ANs. The preoperative diagnosis of intracranial facial neuroma is difficult (Fig. 188-12 and 188-13). Early facial nerve symptoms are an obvious warning that a CPA lesion may rarely be a facial nerve neuroma. At the House Ear Clinic all patients diagnosed with CPA neuromas are warned preoperatively of a 1% risk of facial nerve neuroma. This possibility is emphasized particularly if preoperative ENOG is abnormal on the tumor side.

Other cranial nerve neuromas (schwannomas)

Neuromas may arise on any of the other cranial nerves in the posterior fossa. On imaging studies, nonacoustic neuromas have the same characteristics as ANs except for their location. Overall ANs represent 95% of intracranial schwannomas, and trigeminal neuromas are the next most common; however, schwannomas have also been reported on the CNs IX, X, XI, and XII. These lesions are distinguished by their different location and by symptoms of dysfunction of the cranial nerve of origin.

Trigeminal neuromas arise both *intradurally,* from the nerve root in the CPA and Meckel's cave, and *extradurally,* from the gasserian ganglion in the middle cranial fossa (McCormick et al, 1988). Typically these lesions enlarge Meckel's cave and produce hypesthesia of the face (Fig. 188-14).

Neuromas of CN IX, X, and XI produce smooth enlargement of the jugular foramen and classically produce hypesthesia and weakness of the palate, vocal cord, and shoulder, respectively (Fig. 188-15). If they arise in the posterior fossa, these tumors may grow to large sizes before producing predominantly acoustic or cerebellar signs. Accurate preoperative differentiation of these tumors from acoustic neuromas is important because residual hearing is more likely to be preserved in lower cranial nerve neuromas.

Hypoglossal neuromas produce motor hemiatrophy of the tongue as well as enlargement of the hypoglossal canal on radiography (Fig. 188-16).

Glomus tumors (paragangliomas)

Paragangliomas are discussed in detail in Chapter 192. Because glomus jugulare tumors and glomus vagale tumors may extend into the posterior fossa, however, they are important lesions in the differential diagnosis of skull base neoplasms.

The first symptom is often pulsatile tinnitus, after which a conductive hearing loss develops. Involvement of the nerves of the jugular foramen and the hypoglossal nerve causes progressive neurologic deficits related to those nerves.

The characteristic appearance on CT scan with bone-review program is irregular destruction of the jugular foramen schwannomas, which produce a smooth enlargement (Fig. 188-17). The vascular pattern of paragangliomas on angiography are characteristic (Fig. 188-18) and biopsy is not indicated in these lesions (Spector et al, 1979). Diagnostic angiography

should be performed concomitantly with preoperative embolization when surgical resection is planned.

MRI produces a unique "salt and pepper" mixture of intensities on both T1 and T2 images (Fig. 188-19). Arborizing flow-voids reveal the prominent tumor vessels of this lesion. Lo (1991) has described two limitations of MRI in evalating paragangliomas. Bone changes and the relation of tumor to bone landmarks are not visualized on MRI. Furthermore, especially on Gd-enhanced T1 images, distinguishing tumor intensity from bone marrow is difficult. Thus MRI may provide complementary information about infralabyrinthine and intracranial tumor extensions, but bone algorithm CT is the cornerstone of imaging evaluation in paragangliomas.

The combination of CT, MRI, and angiography can provide extensive information about involvement of the internal carotid artery. The potential role of magnetic resonance angiography (MRA) is currently being considered to replace angiography (Fig. 188-20). Because angiography is a necessary step for preoperative embolization of glomus tumors, however, it is unlikely that MRA can replace the role of direct intravascular angiography in glomus tumor management. If surgical resection may require manipulation of the artery, preoperative assessment of the adequacy of collateral flow via the circle of Willis is necessary. Temporary balloon occlusion of the carotid artery in combination with radio-isotope imaging techniques or xenon-enhanced CT scan provide accurate quantification of collateral blood flow (Janecka et al, 1990).

Arachnoid cysts

Arachnoid cysts are thin-walled sacs that contain yellow, entrapped CSF. The current theory is that these lesions represent congenital developmental anomalies (Haberkamp et al, 1990).

Symptoms are produced by the mass effect of the cyst on surrounding structures and may be similar to the symptoms of ANs. These patients may develop mild to profound hearing loss with a retrocochlear pattern (Pappas and Brackmann, 1981).

On imaging studies, these lesions have similar CT and MRI characteristics as epidermoids (as discussed earlier in this chapter). Enlargement of the IAC is often noted but is not characteristic of these lesions. The typical appearance is a smooth-surface lesion, which on CT approximates CSF and is nonenhancing, and on MRI exhibits isointensity or hypointensity to brain on T1 and hyperintensity to brain on T2 images (Fig. 188-21). Treatment of these lesions is not total resection. Instead, surgical drainage via retrolabyrinthine exposure is the usual recommended therapy; however, diuretic therapy provides symptomatic relief in a minority of patients.

Hemangiomas

Hemangiomas are hamartomatous neoplasms originating from blood vessels. Although benign, they produce symptoms by compression of adjacent structures. The two types of hemangiomas are capillary and cavernous.

Capillary hemangiomas typically arise in the area of the geniculate ganglion in association with a perigeniculate capillary plexus (Balkany et al, 1991). The lesion is characterized by a progressive facial weakness despite its being much smaller than facial nerve neuromas. The upper basal turn of the cochlea may be exposed by the expanding lesion with the production of pulsatile tinnitus. CT demonstrates a smooth enlargement of the geniculate ganglion and enlargement of the labyrinthine portion of the fallopian canal by a soft tissue mass (Fig. 188-22). Although capillary hemangiomas are enhancing, they produce facial weakness at such an early stage that only subtle findings may be apparent on CT, and a very small enhancement in the area of the labyrinthine segment may be the only finding. Other CT findings include "honeycomb bone", irregular and indistinct bone margins, and intratumoral bone spicules (Fig. 188-23). These findings contrast with those of facial nerve neuromas, which are large, more obvious lesions with sharp bone margins.

Cavernous hemangiomas are the second type of hemangioma. These lesions present in the IAC and produce symptoms typical of an AN. Although they tend to produce symptoms more rapidly than an AN, they are identical with AN on CT. However, on MRI they tend to be slightly more hyperintense than the typical AN (Fig. 188-24).

Petrous Apex Lesions

Petrous apex lesions are an important category of skull base neoplasms that may involve the posterior fossa. Some of the specific lesions have been described in the CPA portion of this chapter. The critical distinction in petrous apex diagnosis is between cholesterol granulomas, which require drainage procedures, and mass lesions, which generally require complete excision.

Cholesterol granulomas

A cholesterol granuloma arises in the pneumatized spaces of the temporal bone as a result of occlusion of the air cell system. Hemorrhage into the air cells results in a foreign body reaction and progressive granuloma formation. An expansile lesion of the temporal bone results, with extension into the CPA and resultant signs and symptoms of CN VIII dysfunction. The CT and MRI findings in this lesion are distinguishable from the other common lesions of the petrous apex (Table 188-3). On CT the lesions result in a punched-out lesion of the temporal bone with an isodense mass of the petrous apex that does not enhance; however, there is rim enhancement with intravenous contrast (Fig. 188-25). On MRI both T1 and T2 images are hyperintense with respect to brain (Fig. 188-26). Primary cholesteatomas are the main lesions from which cholesterol granulomas must be distinguished. Cholesteror granulomas are much more common; in our institution they have occurred 20 times more frequently than epidermoids. Overall, cholesterol granulomas are relatively uncommon in comparison to ANs. Only one cholesterol granuloma is identified for each 35 ANs diagnosed in our institution. On imaging studies, rim enhancement on CT is distinctive; on MRI, epidermoids do not manifest hyperintensity on *both* T1 and T2 images. Total excision of cholesterol granulomas is not necessary; drainage may be achieved by a transmastoid or transcanal infralabyrinthine approach with preservation of cranial nerve function. The transcanal, infracochlear, hypotympanotomy approach is our preferred management as it affords dependent drainage and the possibility of revision, if necessary, via a myringotomy (Giddings et al, 1991).

Table 188-3. Imaging features of petrous apex "cystic" lesions

Features	Cholesterol granuloma	Epidermoid	Mucocele
CT density	Isodense	Hypodense	Hypodense
T1 MRI	Hyperintense	Hypointense	Hypointense
Gadolinium	Nonenhancing	Nonenhancing	Rim enhances Mass is nonenhancing
T2 MRI	Hyperintense	Hyperintense	Hyperintense
Borders	Smooth	Scallopped	Smooth.

Asymmetric petrous apex pneumatization

Although asymmetric pneumatization of the petrous apex does not represent a true neoplasm, this condition can be confusing and must be distinguished from true neoplasms. The fat content of bone marrow in a nonpneumatized petrous apex can produce a hyperintense image on a nonenhanced T1 MRI (Fig. 188-27). This finding is distinguished from a neoplasm by the lack of bone destruction or expansion on CT, the absence of contrast enhancement with Gd, and the hypointensity on T2-weighted images.

Mucocele and mucus retention cysts

Petrous apex air cells may become obstructed, resulting in retained secretions as a mucus retention cyst in the petrous apex (Fig. 188-28) or an expansile mucocele (Fig. 188-29). CT reveals a nonenhancing lesion limited to the petrous apex air cell system. MRI is consistent with a mucus-filled lesion (hypointense on T1 and hyperintense on T2). No specific treatment is required for retained mucus, but a symptomatic mucocele requires drainage.

Petrous carotid artery aneurysms

Although aneurysms of the horizontal carotid artery are rare, they may appear as expansile, well-defined masses (Fig. 188-30). Their preoperative identification is critical for obvious reasons. Carotid aneurysms may be confused with the radiologic appearance of chondrosarcomas.

Giant cell tumors

Giant cell tumors are extremely rare primary neoplasms of the temporal bone. They originate from undifferentiated cells of the supporting connective tissue and consist of multinucleated giant cells in a background of spindle-shaped stromal cells. The one giant cell tumor treated at the House Ear Clinic had retrocochlear signs and symptoms. CT demonstrated a diffuse lesion of the temporal bone compressing the contents of the IAC.

Miscellaneous Cerebellopontine Angle Lesions

Metastatic tumors

Tumors may metastasize to the CPA from other sites. We have treated patients with metastatic lesions from lung, breast, prostate, oropharynx, and cutaneous melanomas. These tumors were distinguished by their rapid progression of symptoms and associated neurologic signs in addition to hearing loss and dizziness. Associated lytic lesions in the petrous apex are another distinguishing feature of metastatic tumors. A rapid impairment of hearing, other cranial neuropathies, and brainstem dysfunction suggest malignant neoplasms of the posterior fossa, especially in a patient with a history of another malignancy.

Chordomas

Chordomas are dysontogenetic neoplasms that arise in remnants of the embryonic notochord. Although more than half arise in the sacrococcygeal region, more than one third of them occur at the skull base in the region of the clivus, or less commonly, the upper cervical vertebrae (Batsakis, 1979). The prominent clinical characteristics are extensive bone destruction and progressive cranial nerve palsies. Extension of clivus chordomas to the petrous apex, sphenoid, or CPA is not unusual. Although frontoorbital headache and vision complaints (such as limitation of visual fields, diplopia, or loss of acuity) are more common, occasionally the initial symptoms reflect extension into the CPA. On CT scans the bone destruction is readily apparent, and the masses are homogeneous with moderate enhancement and a greater density than bone (Fig. 188-31). MRI reveals an isointense T1 image and hyperintense images on T2.

Chondrosarcomas

Chondrosarcomas of the skull base may also appear in the CPA. They are clinically indistinguishable from chordomas except that they are centered more laterally. CT illustrates their characteristic bone destruction and invasiveness (Fig. 188-32). MRI demonstrates this lesion well; it is hyperintense on T2 in the area of bone destruction in the skull base (Fig. 188-33).

Lipomas

Lipomas are hamartomas that are thinly encapsulated and poorly delineated. They appear as soft, multilobular masses of typical adult adipose tissue. We have treated two lipomas within the IAC that produced symptoms typical of an AN. CT scans distinguish these lesions from neuromas in that they are less dense. However, MRI is diagnostic. The lesions are hyperintense on T1, nonenhancing with Gd, and hypointense on T2 (Fig. 188-34).

Dermoid tumors

A dermoid is a skin-lined cystic tumor containing dermal and adnexal structures. The lining of the cyst is mature, stratified squamous epithelium. A dermoid is a slowly expanding lesion that produces symptoms similar to a primary cholesteatoma. It may be differentiated on CT scans as a nonhomogeneous cystic mass that contains calcium but that is otherwise less

dense than brain.

Teratomas

Teratomas arise from multipotential cells that differentiate into a variety of tissue representing more than one germ layer. They contain ectodermal, mesodermal, and endodermal tissues. Carcinomatous or sarcomatous changes occur in 10% to 35% of these tumors. When malignant degeneration occurs, symptoms progress rapidly; otherwise symptoms progress slowly, making the lesions indistinguishable from other benign CPA tumors. CT scans demonstrate a nonhomogeneous lesion of less density than brain without enhancing characteristics.

Intraaxial Tumors

Intraaxial tumors may occasionally be confused with CPA tumors due to extension to the CPA or compression of CPA structures. Intraaxial tumors may arise from the brainstem (gliomas), from the cerebellum (medulloblastomas from the vermis or astrocytomas from the peduncles) or from the fourth ventricle (choroid plexus papillomas and ependymomas). Although such lesions are highly unusual, they cannot be totally ignored. In children, brainstem gliomas are reportedly the most common source of CPA neoplasms. As a group, the intraaxial tumors are usually isointense on T1 MRI images and hyperintense on T2 images (Lo, 1991).

Hemangioblastomas

Hemangioblastomas are tumors of blood vessel origin occurring primarily in the cerebellum. They may also occur in the cerebral hemispheres and in association with a similar retinal tumor and may be multicentric. Although histologically benign, hemangioblastomas may produce major neurologic dysfunction by compression of the brainstem.

Rapidly progressing signs and symptoms of cerebellar dysfunction are characteristic of this tumor. Hearing and balance are likely to remain normal. Cranial CT scans identify a tumor intrinsic to the cerebellum with extension into the CPA (Fig. 188-35).

Medulloblastomas

Medulloblastomas arise from the cells of the external granular layer of the cerebellar folia. They may appear as exophytic masses on the cerebellum with extension into the CPA. Symptoms are produced by destruction of cerebellar tissue and mass effect of the tumor on the CPA's adjacent structures.

Medulloblastomas are characterized by a rapid development of symptoms. In addition to having hearing loss and dizziness, all patients in our series had associated neurologic findings, including facial weakness, dysmetria of speech and hand motion, perverted nystagmus, and abnormal peripheral reflexes. Temporal bone imaging is normal, and CT and MRI demonstrate a lesion intrinsic to the cerebellum.

Brainstem gliomas

Four patients treated at the House Ear Clinic have had brainstem gliomas. Exophytic gliomas may arise on the surface of the pons and grow into the CPA; gliomas may also be intrinsic to the brainstem. Exophytic gliomas may produce signs and symptoms similar to those of ANs, whereas the intrinsic variety produces predominantly long tract signs.

The two patients with exophytic lesions in this institution were thought preoperatively to have ANs. Long tract signs in association with characteristic brainstem distortion on CT made preoperative diagnosis possible in the patients with intrinsic lesions.

Tumors of the fourth ventricle: Malignant choroid plexus papillomas and ependymomas

Choroid plexus papillomas and ependymomas both arise from the fourth ventricle and cause CPA symptoms by growing through the foramen of Luschka. In this location they produce early signs of CN VIII dysfunction. In malignant choroid plexus papillomas, CT scan demonstrates a mass with enhancing characteristics of a schwannoma (Fig. 188-36). It is distinguishable from an acoustic schwannoma by being separate from the IAC. Ependymomas may calcify, and those portions have variable imaging characteristics. Both lesions are isointense with brain on T1 MRI images and mildly hyperintense to brain on T2 images (Fig. 188-37).

Summary

The differential diagnosis of posterior fossa skull base neoplasms begins with a thorough neurootologic history and physical examination. Any suspicious neurotologic finding should be evaluated thoroughly because these lesions produce minimal signs and symptoms until they are far advanced. Although ANs are the most common posterior fossa skull base tumors, the less common lesions can usually be diagnosed accurately with the application of modern audiologic and imaging modalities and a systematic approach to their differing characteristics.