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Introduction

A wide spectrum of diseases involves the temporal bone and skull base. Primary tumors, inflammatory processes, and metastatic disease are a few of the many abnormalities that can exist within the temporal bone and skull base. With the advent of high resolution computed tomography (HRCT) and magnetic resonance imaging (MRI), the diagnosis of many of these abnormalities is more readily apparent. By classifying these processes based on location and identifying the common imaging characteristics, a relatively straightforward differential diagnosis can be formulated.

Lesions of the Middle Ear and Mastoid

Cholesteatoma (Epidermoids)

Cholesteatomas are soft tissue masses caused by aberrant accumulation of keratin debris within a sac of squamous epithelium. These are not true neoplasms, as they do not exhibit cellular growth; rather they are the result of squamous epithelial migration or implantation within the middle ear, mastoid, or petrous apex. Middle ear cholesteatomas are usually distinguished as two types, acquired or congenital. Congenital masses are thought to arise from embryonic rests within the middle ear. Acquired cholesteatomas consist of an abnormal keratinizing squamous epithelium in the middle ear, epitympanum, or mastoid. It has been proposed that primary disease develops behind an intact tympanic membrane as a result of chronic otitis media. Secondary disease results from eustachian tube dysfunction followed by a retraction pocket in the tympanic membrane, which traps epithelium, followed by perforation and accumulation of keratin within the middle ear. Patients typically present with purulent otorrhea and conductive hearing loss. Patients with congenital cholesteatoma, however, may present with an intact tympanic membrane and not present with otorrhea. Diagnosis is typically made during otologic

examination. HRCT may be of value in the preoperative assessment of cholesteatomas but is not always diagnostic of disease. Erosion of the scutum and expansion of the antrum within areas of air cell breakdown and soft tissue density are frequent findings on HRCT. Ossicular destruction, erosion of the otic capsule (lateral canal), and erosion of the facial canal or mastoid tegmen can also be seen. Routine CT is not advocated for cholesteatoma diagnosis but may be important in complicated disease (revision cases, intact TM, history does not correlate well with physical exam). MRI plays a limited role in middle ear and mastoid cholesteatomas. However, on T1-weighted images cholesteatomas are low intensity and on T2-weighted images cholesteatomas are typically high intensity.

Paragangliomas

Paragangliomas of the head and neck are typically benign, slow growing tumors arising from widely distributed paraganglionic tissue thought to originate from the neural crest. Paraganglia in the head and neck region are closely aligned with the distribution of the parasympathetic nervous system and often have a close spatial relationship with neural or vascular structures. Paraganglia have been shown to have chemoreceptor roles with modulation of respiratory and cardiovascular function. Paraganglia are located in many locations in the head and neck including the middle ear, jugular bulb, carotid bulb, ganglion nodosum of the vagus nerve, larynx, and base of the heart. Histologically, paragangliomas are similar in appearance to the normal histology of the paraganglia and includes two cell types. Type I cells (chief) cells are APUD type cells with copious cytoplasm and large round or oval nuclei. Their cytoplasm contains dense core granules that store and release catecholamines. Type II (sustentacular) cells are elongated cells that closely resemble Schwann cells although their function is not entirely clear. The two cell types are arranged into clusters with a core of chief cells surrounded by sustentacular cells embedded in a fibrous stroma. These clusters make up the fundamental histologic structure (termed "Zellballen") and may be somewhat enlarged in paragangliomas. Nuclear pleomorphism and cellular hyperchromatism are common in benign paragangliomas and should not be considered evidence of malignancy. In fact, there are no clear histologic characteristics of malignancy in these lesions. Malignancy is based on the clinical finding of metastasis, not on histologic examination (occurs in only 3-4 % of tumors). The chief cells of paragangliomas have neurosecretory granules that store catecholamines, however only 1-3 % of these tumors actively secrete norepinephrine. Catecholamines are much more likely to be secreted by glomus jugulare tumors than glomus tympanicum tumors. Approximately 5% of patients with paragangliomas eventually present with multiple tumors, however with a familial form of the disease over 50% exhibit multicentricity.

Although described as rare, paragangliomas are the most common true neoplasm of the middle ear and are the most common pathologic entity affecting the jugular foramen. They are second only to schwannomas as the most common neoplasm affecting the temporal bone.

Glomus tympanicum tumors usually originate on the promontory of the cochlea from paraganglionic tissue associated with either Arnold's nerve (CN X) or Jacobsen's nerve (CN IX). Typically, the tumor enlarges to fill the middle ear space and to envelope the ossicles. The tympanic membrane may be displaced laterally but rarely will it perforate through the membrane. The tumor may enlarge into the mastoid, facial recess or retrofacial air cells. If large enough, the tumor may cause bone erosion in the hypotympanum and the jugular fossa or vertical portion of the petrous carotid may be exposed. These large tumors are difficult to differentiate from glomus jugulare tumors.

Glomus jugulare tumors arise in the jugular fossa and are usually large before patients become symptomatic. Compression of neurovascular structures in the jugular fossa and extension along the skull base to the hypoglossal canal can lead to cranial neuropathies. Erosion of the jugular fossa anteriorly and superiorly may expose the petrous internal carotid artery and allows the tumor to invade the middle ear producing pulsatile tinnitus and a conductive hearing loss.

Patients with glomus tympanicum typically present with pulsatile tinnitus and conductive hearing loss and usually present earlier than glomus jugulare tumors. Otoscopic exam usually demonstrates a reddish-blue pulsatile retrotympenic mass. Positive pressure with pneumatic otoscopy may cause blanching known as the Brown sign. The pulsatile nature of the lesion may be diminished with pressure over the ipsilateral carotid artery which is known as the Aquino sign. A bruit may be auscultated over the mastoid in some cases. Jugular foramen syndrome, also termed Vernet syndrome, arises when tumor growth affects cranial nerves IX, X, and XI. Villaret syndrome is a combination of jugular foramen syndrome and Horner's syndrome. Facial nerve weakness may occur and ataxia may develop if the tumor involves the posterior fossa. Higher cranial nerves may become involved especially if the cavernous sinus becomes infiltrated with tumor.

Imaging studies provide essential information for the evaluation of patients with temporal bone paragangliomas. HRCT can help identify the tumor origin, especially if the bony partition between the jugular fossa and hypotympanum is intact. Erosion of the bony spine that separates the jugular bulb from the petrous carotid usually indicates a glomus jugulare tumor. HRCT also helps to exclude other lesions such as an aberrant internal carotid artery or high riding dehiscent jugular bulb. In addition, the CT sections can be expanded to include slices down to the carotid body in order to exclude multicentric lesions. MRI helps identify intracranial extension. It also can reveal diagnostic flow voids within the tumor (salt and pepper pattern). On enhanced T1-weighted images, it delineates further the true deep tissue extent and its relationship with neighboring neurovascular structures. Large tumors are usually evaluated with carotid angiography as well typically with embolization 1-2 days prior to surgical resection.

Vascular Variants in the Middle Ear and Mastoid

Particular variations in vascular anatomy in the temporal bone are a frequent cause of a retrotympenic vascular mass and pulsatile tinnitus. These variants include the jugular bulb anomalies (high riding bulb, dehiscent jugular bulb, and jugular bulb diverticulum), aberrant internal carotid artery (ICA), and the persistent stapedial artery. When these anomalies are present, differentiation from the paragangliomas is crucial.

An asymmetrical jugular bulb is a common vascular variant. HRCT will demonstrate asymmetry in the size of the jugular bulb area with preservation of the jugular spine and all cortical margins. A large, high riding jugular bulb could also simulate a lesion expanding the jugular foramen. However, the bony margins will be well corticated, differentiating this from a pathologic process. If the jugular bulb is dehiscent, a retrotympenic mass may be appreciated. On HRCT, however, the jugular bulb will be contiguous with the middle ear mass confirming its origin. MRI will show the high riding bulb relative to the tympanic cavity.

An aberrant ICA may present with pulsatile tinnitus and a vascular appearing retrotympenic mass that may mimic a paraganglioma clinically. HRCT appearance of such an anomaly is characteristic. The aberrant ICA is seen entering the tympanic cavity through an enlarged tympanic canaliculus, posterior to the normal ICA. It courses anteriorly across the cochlear promontory to join the horizontal carotid canal through a dehiscence in the carotid plate. Angiography shows characteristic features of the aberrant ICA, however this is not necessary for diagnosis. Because MRI does not display bony detail well, the use of MRI even with flow enhancement techniques does not reliably identify aberrant ICA's. HRCT remains the examination of choice.

The persistent stapedia artery is a rare vascular anomaly. It may occur with or without an aberrant ICA. The HRCT findings are characteristic. These include the absence of the foramen spinosum on the affected side and enlargement of the proximal tympanic segment of the facial nerve canal adjacent to the cochleariform process.

Adenomas

Benign adenomas of the middle ear are rare, nonaggressive neoplasms that are most commonly found in young adults. These seem to arise from the glandular elements of the middle ear mucosa. Histology usually reveals benign glandular proliferation. Patients typically present with a middle ear mass and an intact tympanic membrane with a conductive hearing loss. HRCT reveals a soft tissue mass usually arising from the promontory of the cochlea. There is no bony erosion, however the ossicular chain may be involved. The differential diagnosis of such a mass in the middle ear includes a schwannoma, adenoma, cholesteatoma (congenital), aberrant internal carotid artery, high riding jugular bulb, or a glomus tumor.

Endolymphatic Sac Tumors

Endolymphatic sac tumors are a rare, aggressive papillary tumor of the middle ear and mastoid. These are highly vascular, friable, and polypoid masses that on microscopic examination have both papillary and cystic components. The papillary component resembles the rugose portion of the endolymphatic sac while the cystic portion usually contains a proteinaceous material. Such material is similar to thyroid colloid, thus these tumors must be differentiated from metastatic papillary thyroid carcinoma with the use of thyroglobulin staining. These tumors extend along the endolymphatic duct in the direction of the bony labyrinth. Destruction of the labyrinth usually produces a sensorineural hearing loss. From the endolymphatic duct, the tumor may erode into the posterior semicircular canal, the vestibule, or mastoid. From the mastoid, these tumors can envelope the facial nerve and extend into the middle ear.

Screening audiometry in conjunction with a temporal bone scan usually makes diagnosis. Audiograms usually show a SNHL however a CHL may result from middle ear extension of the tumor. HRCT often shows an erosive mass along the posterior petrous face. The tumor may contain areas of calcification and often exhibit an "expansile" growth pattern which helps differentiate these from metastases or other malignant processes such as chondrosarcomas. MRI reveals a pattern of signal intensity that varies with tumor size. In general, tumors less than 3 cm show circumferential rim of high signal intensity whereas tumors over this size can demonstrate a "speckled" pattern. On T1-weighted images with gadolinium, these tumors enhance and may contain flow voids. Angiography is usually used as an adjunct to surgical therapy to help minimize intraoperative blood loss.

There is a strong association with endolymphatic sac tumors and von Hippel-Lindau disease. Von Hippel-Lindau disease is an autosomal dominant disorder that manifests itself as multiple hemangioblastomas of the retina and CNS with associated renal cysts, renal carcinoma, pheochromocytomas, pancreatic cysts, and papillary cystadenomas of the epididymis. Approximately 11% of patients with this disease have endolymphatic sac tumor, however as many as 60% may eventually develop the disease. Thus, early screening and follow-up allows for earlier diagnosis and management of this aggressive disease. MRI with gadolinium enhanced T1-weighted images usually reveal marked enhancement of the heterogenous lesion.

Sarcomas

Sarcomas are the most common temporal bone malignancy in children. Rhabdomyosarcoma is the most common of these neoplasms which accounts for 30% of sarcomatous temporal bone neoplasms and 7% of all temporal bone malignancies. Pleuripotential mesenchymal cells in the middle ear or eustachian tube give rise to this tumor. Most patients present with chronic otorrhea and otalgia that fails to respond with appropriate medications. Facial weakness or paralysis is not uncommon and distant metastases are present in 14% of patients at presentation. Rhabdomyosarcoma is divided into several histologic types with the embryonal form being the most common. This consists of small spindle shaped mesenchymal cells in a loose myxoid matrix. Radiological imaging typically reveals a soft tissue density in the middle ear or mastoid with extensive surrounding bony destruction.

Metastatic Disease

Temporal bone metastases resulting from distant malignancies are an infrequent finding. Primary sites in descending order of frequency include breast, lung, kidney, prostate, and GI tract. Metastases are usually hematogenously spread and are usually associated with diffuse metastatic disease in other locations. Patients typically present with hearing loss and possibly facial weakness and dysequilibrium. The mastoid and petrous apex shows a predilection for metastatic disease compared with other areas in the temporal bone. On both HRCT and MRI scans, the appearance of lesions is highly variable depending on the tissue of origin. Although most metastases destroy bone, a few are osteoblastic. Osseous destruction is usually irregular but may be well marginated.

Langerhans cell histiocytoses (Histiocytosis X)

Langerhans cell histiocytoses are a group of neoplasms that are characterized by idiopathic histiocytic and eosinophilic proliferation. Three major categories are described. These include the infantile form (Letterer-Siwe disease), Hand-Schuller-Christian disease, and eosinophilic granuloma. Letterer-Siwe disease is a diffuse, systemic form of the disease that is uniformly fatal within 1-2 years. Eosinophilic granuloma is a localized form of the disease and usually presents as a unifocal bony lesion usually involving the temporal or frontal bone. The tumor is characterized by a local collection of histiocytes and eosinophils that causes resorption of bone producing a radiolucent lesion. Diagnosis is confirmed with open biopsy. HRCT reveals "punched-out" defects or "moth-eaten" holes. Hand-Schuller-Christian disease is a systemic variant of eosinophilic granuloma that usually presents with chronic otorrhea and hearing loss. There are typically multiple punched-out defects in the temporal bone and is often bilateral. The disease may affect other organs including the abdominal viscera and skin.

Lesions of the Petrous Apex and Clivus

Anatomy (Asymmetric Bone Marrow and Giant Air Cells)

The petrous apex is the part of the temporal bone that lies between the inner ear and the clivus. It is divided into two compartments by the internal auditory canal. Disease processes most frequently involve the much larger anterior portion, which lies anteromedial to the cochlea and internal auditory canal. The smaller, and clinically less significant posterior portion lies between the semicircular canals and the IAC. This region is comprised primarily of solid bone of otic capsule origin but may contain some pneumatization.

Anteriorly and medially, the petrous bone articulates with the basal portion of the occipital bone at the clivus. Laterally, it articulates with the greater wing of the sphenoid. The anterosuperior surface of the apex forms a portion of the floor of the middle cranial fossa. The posteromedial surface extends laterally from the clivus, and defines the anterolateral limit of the posterior fossa. The internal carotid artery traverses its apex along its long axis. The cavernous sinus and trigeminal nerve lie anterior and superior. The abducens nerve and confluence of the cavernous and petrosal sinuses pass anterior and medial. The cochlea and labyrinth are lateral and the IAC is posterior to the apex.

The cells of the petrous apex contain bone marrow. The extent of bone marrow infiltration varies among patients and may be asymmetric in patients due to unequal pneumatization in up to 68% of patients. On HRCT, asymmetric pneumatization and marrow formation is easily recognizable, however on MRI the signal returned from an asymmetric marrow may simulate a lesion in the petrous apex. On T1-weighted scans, bone marrow is hyperintense because of its rich fat content. It is hypointense on T2 weighted images in contrast to pathologic entities such as cholesteatoma, cholesterol granulomas, and most tumors.

A few patients have an extraordinary degree of mastoid pneumatization. Giant air cells may indent the anterior wall of the IAC and may simulate a destructive lesion. The giant cells are usually smooth and contain air which is easily discernable on HRCT.

Cholesterol Granulomas

A cholesterol granuloma, the most common primary lesion of the petrous apex, results from chronic obstruction of pneumatized air cells. The pathway for ventilation of petrous apex cells is circuitous, eventually reaching the middle ear space to communicate with the nasopharynx via the eustachian tube. When edema occurs, negative pressure develops within the lumen of apex cells leading to fluid transudation and hemorrhage. It is believed that the red blood cells break down releasing cholesterol from their cell membranes which produces crystals formation and incites a sterile inflammatory reaction. Granulation tissue forms secondary to repeated hemorrhage, thus creating an expansible lesion. Mass effect typically produces hearing loss, tinnitus, vertigo, or facial twitching.

On HRCT, these lesions appear as smoothly delineated expansile masses located in the anteromedial portion of the apex. The density is similar to that of surrounding brain. There is no enhancement of these lesions with contrast. HRCT provides essential information in

determining the most favorable route for drainage of these lesions either via an infralabyrinthine, infracochlear, or transsphenoid routes.

MRI provides the diagnosis in virtually all cases. These lesions are markedly hyperintense on both T1- and T2-weighted images, likely secondary to the subacute and chronic hemorrhages with associated breakdown products. They do not enhance further after the administration of gadolinium. MRI also plays a key role in assessing for recurrent lesions. Serous fluid, mucous, or scar tissue that fills a drained cholesterol granuloma will not produce the expansile mass effect and will have a much lower T1-weighted signal on MRI. Recurrence of a cholesterol granuloma should be suspected when cyst fluid contents demonstrate renewed hyperintensity on T1-weighted scans.

Primary Cholesteatomas

Primary or congenital cholesteatomas are rare lesions composed of desquamitized keratin surrounded by a capsule of stratified squamous epithelium. These likely arise from aberrant embryonic rests within the petrous apex.

On HRCT, cholesteatoma of the petrous apex appears as a smoothly marginated expansile lesion that does not enhance following administration of contrast. Typically, the mastoid air cells are normal in patients with congenital cholesteatomas in this region. Unfortunately, primary cholesteatomas are generally not distinguishable from cholesterol granulomas on HRCT.

With MRI, petrous apex cholesteatomas appear inhomogeneously hypointense on T1-weighted images and homogeneously hyperintense on T2-weighted sequences, thus differentiating these from cholesterol granulomas. While cholesteatomas are hyperintense on T2 images, they are not usually as hyperintense as is typically seen with cholesterol granulomas.

Effusions in the Petrous Apex

Because of the circuitous nature of the ventilation of the petrous apex, effusions can develop in the petrous apex as a result of eustachian tube dysfunction, upper respiratory tract infections, or barotrauma. A simple effusion is not associated with osseous erosion, but may develop into a cholesterol granuloma or mucocele over time if not resolved. On HRCT, an effusion will appear as a soft tissue density filling an air cell tract without any accompanying signs of bony destruction. As with mastoid effusions, the delicate septae between cells will be preserved.

On MRI, a petrous air cell effusion is similar to a cholesteatoma. It is typically low in signal on T1-weighted images and high intensity on T2-weighted sequences. Thus, MRI alone is not able to distinguish an effusion from a cholesteatoma in most cases.

Petrous Apicitis

Infection of the petrous apex may involve either the mucosal spaces (apicitis) or the bone and marrow component (osteomyelitis). Acute petrous apicitis may occur as a result of abscess formation in a well-pneumatized petrous tip. The symptoms in this setting are usually rapid and may progress to Gradenigo's syndrome. The classic presentation of such case is typically

described as retroorbital pain, abducens nerve palsy, and otorrhea. Longstanding chronic middle ear infections may also lead to the evolution of petrous apex symptoms and may progress to osteomyelitis of the petrous tip bone and marrow spaces.

On HRCT, acute petrous apicitis in a well-pneumatized petrous apex appears as an expansile lesion that may have irregular margins. Chronic infection may lead to bone destruction similar to what is seen in skull base osteomyelitis. Apicitis typically does not enhance, however if the infection has coalesced to form an abscess, enhancement may be marked.

On MRI, acute petrous apicitis with abscess formation demonstrates low intensity on T1-weighted images and high intensity on T2-weighted images. It may also reveal rim enhancement after gadolinium administration.

Skull Base Osteomyelitis

Skull base osteomyelitis (SBO) usually evolves after long standing chronic otitis externa in an immunocompromised or diabetic individual. The petrous apex becomes involved usually as part of a larger process typically involving an extensive region of the skull base.

On HRCT, soft tissue density is usually seen in the ear canal and middle ear as well as in the pneumatized spaces of the temporal bone. Demineralization can occur late in SBO which makes HRCT insensitive in some individuals. In advanced cases, an irregular lytic pattern may be seen.

On MRI, an ill-defined, irregular increase in signal intensity is present, especially on T2-weighted images. It is of little use in determining the extent of the lesion, as bony detail is poor.

Technetium scans can document the presence of early SBO and gallium scans can provide a map of the area involved with inflammation. Since the technetium remains positive long after the infection has cleared, gallium scanning is used to monitor antibiotic efficacy.

Aneurysms

Aneurysms of the petrous portion of the carotid artery are extremely rare. They are, however, an entity that must be differentiated from other lesions affecting the petrous apex. Their etiology is likely a congenital weakness of the artery wall but may also occur as a sequelae to trauma or infections. On HRCT, aneurysms have the appearance of a smoothly marginated bone-eroding lesion. With contrast, the lesion may be homogeneous and dense, or heterogeneous, depending on the amount of flowing blood and thrombus.

On MRI, intrapetrous aneurysms can be complex with flow voids. Relative intensity on T1- and T2-weighted images depends on the amount of thrombus present and the age of the aneurysm. Angiography may be required to confirm the diagnosis and MRA may be a useful adjunct.

Chondrosarcomas

Chondrosarcoma of the skull base is thought to arise from embryonic rests of cartilage that occur near the foramen lacerum and petrous apex. Patients usually present with headaches or symptoms suggesting multiple cranial nerve compromise. Although benign chondromas may also occur, chondrosarcoma appears to be much more prevalent. On HRCT, benign chondroma has been described as a non-enhancing, relatively regular, lytic lesion. In the more common chondrosarcoma, irregular bone destruction is the rule with areas of contrast enhancement. Multiple calcifications may occur and have been said to resemble “popcorn”.

On MRI, chondrosarcoma enhances markedly with gadolinium. Differentiating chondrosarcomas with chordomas may be difficult, however chondrosarcomas tend to arise more laterally (within the petrous apex) as compared to chordomas.

Chordomas

Skull base chordomas are low to intermediate grade malignancies that result from defective embryonic remnants of the notocord. While the notocord is typically a midline structure, lateral projections of notocordal tissue may reside in the medial aspect of the petrous apex. Thus, while most chordomas arise in the clivus and spread into the petrous apex, they may also develop entirely within the petrous apex. Headache, diplopia from abducens palsy, and visual deficits are typical complaints of patients. Diagnosis can be made from radiographic imaging and from cytological or histological evaluation revealing the typical “soap bubble” or physaliphorus cells from these tumors. HRCT images show a destructive lobulated soft tissue mass often containing foci of calcification. There is typical erosion of the clivus and basiocciput that may extend laterally to the petrous apex. Enhancement is typical with intravenous contrast.

On MRI, chordomas are hypointense on T1-weighted images and markedly hyperintense on T2-weighted images. They usually enhance with administration of gadolinium. Given their location and propensity for bony erosion, and imaging characteristics, chordomas may be difficult to differentiate from chondrosarcomas discussed earlier.

Metastases

The petrous apex and clivus are two of the most common sites in the temporal bone for metastatic disease. As discussed earlier, primary sites in descending order of frequency include breast, lung, kidney, prostate, and the GI tract. On both HRCT and MRI scans, the appearance of lesions is highly variable depending on the tissue of origin.

Lesions of the Internal Auditory Canal, Cerebellopontine Angle, and Skull Base

Epidermoids

Epidermoids represent 1% of all intracranial masses and most occur in the region of the cerebellopontine angle (CPA). Ectodermal rests likely are the etiologic factor similar to primary cholesteatomas of the petrous apex. Epidermoids enlarge by expanding to fill empty spaces such as the CPA, IAC, or petrous apex. These masses can reach very large sizes before they begin to compress adjacent structures. Thus, most patients become symptomatic in a gradual manner

with their symptoms being related to the location of the mass. Patients with CPA masses tend to present with SNHL and dysequilibrium, similar to the presentation of an acoustic neuroma. Epidermoids, however, are more likely to present with facial paresis or hemifacial spasm. These tumors are usually diagnosed on imaging studies. HRCT usually shows a well-defined, homogeneous mass that may contain areas of calcification. MRI is diagnostic, showing similar characteristics as congenital cholesteatomas of the petrous apex; the T1-weight images are hypointense and the T2-weighted sequences are hyperintense with no enhancement with gadolinium.

Schwannomas

Schwannomas of the temporal bone and the skull base are benign neoplasms that arise from the sheaths of cranial nerves. Schwannomas in this location can be categorized as vestibular, facial, trigeminal, or jugular foramen tumors, depending on the primary nerve involved. Vestibular schwannomas are by far the most common comprising 7% of all intracranial tumors and 80% of CPA tumors. These tumors usually expand centrally from the internal auditory canal into the CPA and may compress the pontine brainstem and cerebellum. Thus, the most common symptoms are unilateral hearing loss, tinnitus, and dysequilibrium. Large tumors extending anteriorly may compress the trigeminal nerve producing facial hypesthesias or may compress the brainstem producing hydrocephalus.

Facial nerve schwannomas can arise anywhere along the course of the facial nerve, however these most commonly arise in the perigeniculate, tympanic, or mastoid segments. Facial weakness or paralysis that progresses is the most common presentation.

Trigeminal schwannomas usually originate from the gasserian ganglion and may expand into the middle cranial or posterior fossa. Patients usually present with facial neuralgias, paresthesias, or hypesthesias in one or more divisions of the trigeminal nerve.

Jugular foramen schwannomas arise from the jugular fossa from the nerve sheaths of cranial nerves IX, X, and XI. The vagus nerve seems the most common however it is difficult to ascertain the nerve from which the tumor is derived. Patients typically present with dysphagia, hoarseness, and shoulder weakness.

Physical examination can reveal functional deficits, however diagnosis is most often made with radiographic studies. HRCT may show expansion of IAC, fallopian canal, or jugular fossa depending on the etiology of the disease. They can reveal inhomogeneous enhancement with contrast. MRI with the use of gadolinium permits definitive diagnosis in the vast majority of cases. MRI shows a low signal intensity on T1-weighted images that enhances markedly with intravenous gadolinium administration.

Meningiomas

Meningiomas are the second most common mass in the CPA. These tumors arise from the arachnoid layer of the meninges. The most frequent location for meningiomas of the lateral skull base is on the posterior surface of the petrous bone between the inferior and superior petrosal sinuses. The clinical presentation varies depending on the location of the tumor. Cranial nerves V, VII, and VIII, XI, X, and XII are variably affected.

Meningiomas are hyperdense to isodense with surrounding brain on HRCT and they exhibit homogenous enhancement after administration of contrast. Calcification within the tumor and associated hyperostosis supports the diagnosis of meningioma over schwannoma.

MRI reveals a densely enhancing, sessile tumor adjacent to but not usually centered within the IAC. A “tail” of “flare” can often be seen at the margin of the tumor. The most common type of meningioma shows isointensity on T1-images but becomes hyperintense on T2-weighted images. Focal signal voids may indicate calcifications and enlargement of the adjacent signal void from nearby bone may indicate hyperostosis. As both schwannomas and meningiomas brightly enhance with gadolinium on T1-images, features such as a mushroom or half-moon shape lesion eccentric to the IAC supports the diagnosis of meningioma.

Lipomas

Lipomas of the IAC and CPA are rare but potentially problematic tumors. As they can present clinically with hearing loss, tinnitus, dysequilibrium, and headache, they can be misdiagnosed as acoustic schwannomas. However, lipomas do exhibit unique imaging characteristics that can provide a definitive diagnosis. On MRI, lipomas are similar to subcutaneous fat with high signal intensity on T1-weighted images and diminished signal intensity on T2-weighted images. In addition, T1-weighted images do not enhance with gadolinium as the tumor signal is already near complete saturation. MRI with fat suppression may further confirm the diagnosis.

Hemangiomas

Hemangiomas are benign vascular proliferations that arise from capillaries, arterioles, or venules. They may occur in a variety of locations including the external ear, tympanic membrane, middle ear, IAC, and the geniculate ganglion of the facial nerve. Clinical presentation depends on the tumor location. Geniculate hemangiomas are the most common temporal bone hemangioma and consistently arise from the superior aspect of the geniculate. Because of the intimate relationship of the hemangioma with the facial nerve, facial nerve dysfunction (paresis, twitching, or spasm) is common. HRCT may show a mixed density mass with calcifications centered at the geniculate ganglion, thus they are often termed ossifying hemangiomas. Hemangiomas are hyperintense on T1- and T2 weighted images on MRI and may contain flow voids.

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