

TITLE: Temporal Bone Lesions

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RESIDENT PHYSICIAN: Alan Cowan, MD

FACULTY PHYSICIAN: Matthew W. Ryan, MD

SERIES EDITORS: Francis B. Quinn, Jr., MD and Matthew W. Ryan, MD

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This discussion of temporal bone lesions is meant to serve as an overview. No attempt is made to include all possible lesions arising within the temporal bone. This is designed as a logical, anatomically categorized review of common lesions which may be encountered both clinically and radiographically. In general, the emphasis is placed on the diagnosis of the lesions, and in-depth discussion of the treatments of the lesions will be withheld in the interests of time.

External Auditory Canal

Benign Neoplasms

The most common benign lesions of the external auditory canal include exostosis and osteomas. Exostoses are more common than osteomas. They tend to arise in the medial portion of the canal along the tympanomastoid and tympanosquamous suture lines. Exostoses are frequently bilateral. Patients usually have a history of exposure to cold water, and this is thought to play a causative role. Osteomas are less common than exostosis. They tend to be unilateral, and are not associated with cold water exposure. Where exostosis are often multiple with broad bases, osteomas are single with pedunculated shapes. They may also occur in the mastoid or IAC, but the EAC is by far more common.

Malignant Neoplasms

Malignant neoplasms of the EAC are uncommon. Most involve the canal by local extension. Skin neoplasms such as squamous or basal cell more commonly occur on the auricle and then extend into the canal. Salivary gland neoplasms such as mucoepidermoid carcinoma or adenoid cystic carcinoma may also invade the EAC from the parotid. Primary neoplasia of the EAC is extremely rare, but tumors of the cerumen glands such as ceruminous adenoma, ceruminous adenocarcinoma, pleomorphic

adenoma, adenoid cystic carcinoma, and mucoepidermoid carcinoma have been reported. Of note, squamous cell carcinoma of the EAC may mimic necrotizing external otitis due to the findings of pain, otorrhea, and bone destruction on CT. All patients with the diagnosis of malignant otitis externa should therefore undergo a histologic examination of the involved lesion.

Malignant Otitis Externa

The term malignant, when referring to OE, does not imply tumor, but rather the aggressive nature of the disease. Other terms for the disease include necrotizing external otitis and skull base osteomyelitis. This disease arises in immunocompromised individuals, classically in elderly diabetics, but may occur in any immunocompromised patient. The offending organism is most often *Pseudomonas aeruginosa*, but *Staphylococcus epidermidis* has also been identified. As the infection progresses, it may extend through the fissures of Santorini and access the deep neck spaces and infratemporal fossa resulting in life-threatening infections and cranial nerve deficits. Radiologic workup most commonly consists of CT scan to delineate bony destruction. If intracranial complications or extension is suspected, then MRI is preferred. If the diagnosis of skull base osteomyelitis is difficult to make on CT, special radionuclide studies may be of help. Once treatment has begun, monitoring the outcome of therapy is difficult with current techniques. Changes may persist on CT, MR, technetium, and Gallium-67 scans for months after infection. Currently, Indium-111 WBC scans and SPECT scans are most useful to evaluate for treatment response, since these scans return to normal much sooner after resolution of disease.

Cholesteatoma

While very common in the middle ear and mastoid, cholesteatoma occurs only about 0.1 – 0.5% of otologic patients. These lesions tend to occur in the posterior aspect of the EAC, directly lateral to the annulus. Due to this position, CT is often recommended to evaluate for involvement of the facial nerve. These lesions are commonly focal, do not extend around the circumference of the EAC, and often present with persistent otorrhea and pain. On CT the lesions usually appear focal and show erosion of local bony cortex.

Keratinosis Obturans

This condition usually occurs in patients with a history of sinusitis or bronchiectasis. As opposed to patients with cholesteatoma of the EAC, KO patients are usually younger (less than 40) and do not have persistent drainage. Where EAC cholesteatomas are often focal near the posterior aspect EAC directly lateral to the annulus, keratinosis obturans occurs throughout the EAC, often causing total obstruction of the canal. Rather than destroying the bone, KO progressively widens the EAC in a broad, usually circumferential manner. The bony cortex should appear intact on CT.

Middle Ear and Mastoid

Otitis Media

This disease will be encountered frequently in practice due to its high incidence. This lesion is often confused for the more aggressive process of mastoiditis. Clinically, OM is less aggressive than mastoiditis. It may cause local pain and drainage, but pain over the mastoid cortex and swelling over the mastoid cortex would be unusual for the diagnosis of OM. Radiographically, fluid may partially or totally fill the middle ear space and mastoid cavity, which is the source of confusion in the differential. OM may have complete opacification of the mastoid and ME space, but will have no destruction of bony septae or mastoid cortex. If the diagnosis is questionable radiographically due to a heavily sclerotic mastoid or other conditions, the diagnosis is usually rather easy to make on clinical grounds.

Mastoiditis

Where OM will have partial to complete ME and mastoid opacification, mastoiditis commonly will have complete mastoid opacification. The presence of bony septae destruction may allow for the application of the term “coalescent” mastoiditis which indicates the destructive nature of the disease. Care should be taken to examine for areas of cortical bone disruption, as this may lead to spread of the infection. Lateral cortex disruption may result in a sub-periosteal abscess. Inferior cortical disruption (mastoid tip) may result in a Bezold’s abscess. Superior disruption of the tegmen may result in meningitis, venous sinus thrombosis, or several types of intracranial abscesses.

Cholesteatoma

Cholesteatoma may present as a primary or acquired lesion. Congenital lesions are by far less common, accounting for only 2% of cholesteatomas. They may occur anywhere within the temporal bone, and radiologic differentiation may not be possible. Acquired cholesteatomas are thought to arise from aberrant rests of epithelial cells which may be present due to trauma, prior surgery, or more commonly, eustacian tube dysfunction or recurrent otitis media. Both eustacian tube dysfunction and recurrent OM are thought to produce a prolonged negative pressure within the middle ear space. This allows for a slow retractive process to occur along the weak area of the TM at the pars flacida. The result is a pocket of epithelial cells which produce keratin but cannot clear this debris. The result is a cholesteatoma. As a result, most acquired cholesteatomas will present within Prussak’s space which is just deep to the pars flacida. Early cholesteatomas of this region may begin to erode the scutum (superior EAC wall) or the ossicles. As the mass grows, openings in Prussak’s space allow the tumor to spread posteriorly where it then has access to the remainder of the middle ear space and mastoid. When it expands, it causes remodeling or erosion of surrounding bone, which may show as scutum, ossicle, or septae erosion. Large lesions may erode into vital structures, and care should be taken to evaluate the tegmen for possible dura exposure, fallopian canal for dehissences of the facial nerve, and the bony labyrinth for canal fistulas.

Paragangliomas

Paragangliomas or glomus tumors arise from nests of neuroendocrine tissue which are present in the region of Arnold's and Jacobson's nerves. These lesions are noted clinically by a red or pink mass within the middle ear, which usually presents with pulsatile tinnitus or conductive hearing loss. Although several studies may be done, CT is the first study of choice. Fine cut CT allows for differentiation between a glomus tympanicum and a glomus jugulare. If a lesion is isolated to the middle ear space, and does not appear to erode the jugular plate, then the diagnosis of a tympanicum may be made. These lesions arise in the middle ear space, and tend to grow in areas of least resistance, only causing bony destruction late in the course, as opposed to cholesteatomas. Once the jugular plate has been eroded, distinction between the two lesions may be difficult to impossible. Extension from the jugular bulb may proceed in almost any direction. Extension inferiorly along the great vessels is common and presents with neck involvement and possible cranial nerve deficits. Extension medially may invade the CPA and result in brainstem or acoustic symptoms. Involved bone appears friable or "moth-eaten", and smooth cortical lines are not visualized. Angiography may be necessary in large tumors to assess the vascular supply and also provide embolization for larger tumors. The most common supply is via the ascending pharyngeal artery, but the posterior auricular or occipital arteries may be involved. Very large tumors may also be fed anteriorly by the maxillary artery or even from the ipsilateral or contralateral internal carotid.

Labyrinth

Labyrinthitis

This disease has multiple causes; bacterial, viral, autoimmune, or traumatic. Viral is the most common, and symptoms include vertigo, sensory hearing loss, tinnitus, and possibly nausea/vomiting. While radiographic confirmation of this disease is not necessary for diagnosis in most cases, common features may be noted on imaging. MRI is the study of choice. This usually demonstrates enhancement of the membranous labyrinth on T1 after contrast administration. Pre-contrast images should not enhance, and if present may represent labyrinthine hemorrhage due to trauma.

Labyrinthitis Ossificans

This disease is the result of labyrinthine inflammation. Bacterial causes are more often implicated when inflammation leads to ossification. If the patient has a history of hearing loss with a congenital or prior labyrinthine infection, it is necessary to rule this disease process out before considering cochlear implantation. CT may show a non-descript fibrous or bony opacification of the normally fluid filled membranous labyrinth. MRI may show a signal void on T2 images which would normally be bright due to the presence of perilymph.

Otosclerosis

This disease is due to resorption of the endochondral layer of the otic capsule with deposition of new spongy bone. This process is usually limited in nature, involving only the fissula ante fenestram, a region at the anterior aspect of the oval window. Deposition of spongy bone here impinges on the oval window and stapes footplate causing a progressive conductive hearing loss. Clinical evaluation may reveal no abnormalities other than a slight pink hue to this area due to increased vascularity. CT scans are usually not necessary for diagnosis, but when performed may show a small focus of soft tissue density at the anterior aspect of the oval window. This may be small, or may obscure the oval window entirely. Care should be taken to evaluate the position of the facial nerve, since a facial nerve overlying the oval window may preclude surgical intervention. When the sclerosis is present beyond the area of the oval window, it may be referred to as retro-fenestral or cochlear otosclerosis. This almost always has a fenestral component. The hearing loss in this case is often sensory as well as conductive. The otic capsule may show almost total replacement by spongy bone on CT.

Internal Auditory Canal and Cerebellopontine Angle

Acoustic Neuroma

Acoustic neuroma or vestibular schwannoma is the most common mass of the cerebellopontine angle. It typically presents in the middle to late decades of life, and is usually unilateral. Bilateral disease or disease early in life should lead to suspicion of neurofibromatosis, a diagnosis which has implications on the treatment of choice for these lesions. Acoustic neuromas are benign masses. These masses tend to grow slowly over several years, with growth rates commonly from 0.2 – 2 mm per year. However, some tumors have been found to progress at rates up to 10 mm per year. These lesions may also remain stable for multiple years with no signs of growth on long-term follow-up. The masses themselves occur with equal frequency on the superior and inferior vestibular nerves. Schwann cells are more numerous in the region of Scarpa's ganglion, and as a result, more tumors are found near this location. This is also why AN, as opposed to several other CPA masses, almost always has an intracanalicular component. While the vestibular portion of the vestibulocochlear nerve commonly is involved, the cochlear portion is rarely involved with tumor. However, when the schwannoma arises from this nerve, the lesion may extend into the cochlea, due to the glial-schwann junction occurring distally at the modiolus.

The radiologic features of acoustic neuroma have been well documented. The anatomic location of the tumor is usually centered about the porus acousticus. The tumors usually have an intracanalicular component, and may extend into the CPA. Other lesions may arise completely within the CPA, and have no extension into the IAC. These lesions may grow to a larger size before diagnosis due to their lack of compression of the nerves within the IAC. Smaller lesions are becoming more common as new techniques for imaging are being found. The range of size at diagnosis therefore varies from mere millimeters up to 6-8 centimeters. As the tumors grow they generally enlarge the porus of the IAC. Although the diameter of the IAC varies greatly from individual to

individual, the diameter of the contralateral IAC has been found to be nearly equal in normal patients. Therefore, an enlargement of the porus greater than 2 mm in comparison to the contralateral side is a relatively good predictor of AN. Since the lesion commonly arises near the area of the porus and enlarges in a spherical fashion, it is considered a concentric mass. Therefore, as the tumor enlarges, it will have acute angles to the remainder of the petrous portion of the temporal bone. Masses such as meningiomas, which are eccentric and tend to spread along the petrous area will have obtuse angles. Other features that may favor AN in the differential are: lesions rarely extend anteriorly and superiorly, they almost never penetrate into the middle cranial fossa, and they lack prominent vasculature, as well as dural tails.

CT scans of acoustic schwannomas tend to show the above features: a porus-centered mass, acute angles, IAC involvement. They also demonstrate the homogeneous nature of the mass. The majority of lesions, excluding previously treated tumors and very large tumors, will show uniform density on CT. Calcifications and central necrosis are rare, however, central clearing has been noted in some larger lesions. The density of AN on CT is similar to that of nearby brainstem, and more dense than surrounding CSF. If given IV contrast, the tumor will most likely show homogeneous uptake and turn very bright. Again, non-homogeneous uptake may be seen with previously treated lesions and large tumors.

MRI is the study of choice if the diagnosis of AN is in question. The T1 weighted exam with Gadolinium contrast has been shown not only to be 100% sensitive for the diagnosis of AN, but also to have the highest negative predictive value for the lesion as well. On standard T1 images, the tumor should be relatively isointense to pons but more intense than CSF. On T2 images, the lesion should be mildly brighter than pons, but darker than CSF. After Gadolinium, the T1 sequence should show a very intense lesion, brighter than all other surrounding structures.

Meningioma

Meningioma is the second most common diagnosis of a primary CPA lesion. These lesions arise from the cap cells of arachnoid villi which are in high concentration near the foramen of cranial nerves and dural venous sinuses. The CPA contains a large number of cranial nerves (V-XII), and is also bonded near its anterior superior aspect by the superior petrosal sinus. Any of these may give rise to a CPA meningioma. Whereas AN is strictly a primary CPA mass, meningioma can arise within the CPA, extend from the middle fossa to the CPA, or extend from the CPA into the middle fossa. Most tumors of this area though, arise from the posterior portion of the petrous bone, and extend along its medial boundary. These lesions uncommonly enter the IAC, and thus, may be large at the time of diagnosis. As opposed to AN, the meningioma is a vascular tumor, and may show flow voids of the pial blood vessels. Meningiomas are also not homogeneous masses, and may show central clearing. Calcifications can be present in up to 25%, and when present, greatly lean towards the diagnosis of meningioma over AN. Another significant finding is that of a “dural tail”. This tapering of the mass along the dura is present 50-72% of the time, and is considered a major diagnostic feature. Also, as a result of this tapering effect, the angles formed between the tumor and the surface of the

petrous bone are generally obtuse.

On CT scan, meningiomas may appear isodense to surrounding structures. The widening of the porus noted in acoustic schwannomas should be absent. CT scan may also show calcifications within the tumor, which are highly suggestive of the diagnosis. MRI is the study of choice. T1 images will show a lesion near the intensity of pons, however, it may not be homogeneous, and may have a central hypointensity in larger lesions. On T2 images the lesion is between pons and CSF in intensity. After administration of gadolinium, the T1 image should show an intense lesion, although not as intense as AN. This sequence is also of value in evaluation for the dural tails. Flow voids caused by the tumor's vasculature may be noted on any sequence, but is easier to see on T1 post-gad due to the brightness of the tumor after contrast.

Epidermoid

Epidermoids are the third most common masses of the CPA angle, comprising 2-6% of the total lesions. These lesions are identical to the cholesteatomas of the middle ear, and may arise within the CPA or within the petrous portion of the temporal bone (congenital cholesteatomas). They are composed of a stratified squamous epithelium lining which surrounds a mass of keratin debris. Unlike most other CPA tumors, epidermoids are associated with facial weakness, paralysis, and spasm. Radiologically they are found to expand into nearby structures. They can cause erosion of the petrous bone, they may extend into the middle fossa with a "dumbbell" appearance, or they may cross midline and expand in the contralateral cistern.

On CT, epidermoids are close to CSF in density and generally do not enhance. MRI is the study of choice for the diagnosis of epidermoid, because the lesions often are difficult to differentiate from arachnoid cysts. T1 images show a very low intensity mass which approximates CSF. T2 images show a very bright mass which again approximates CSF in intensity. These findings are identical to arachnoid cysts. Special MRI protocols such as constructive interference steady state (CISS) or diffusion weighted imaging are required for positive identification. On CISS the epidermoid appears moderately intense, while the arachnoid cyst is hypointense. The same is true for diffusion weighting. This differentiation is crucial, since arachnoid cysts can often be medically managed with diuretics.

Arachnoid Cyst

The arachnoid cyst is important to remember for two reasons. First, it is difficult to distinguish radiologically from an epidermoid. Both epidermoids and arachnoid cysts appear dark on T1 weighted MRI and are very bright on T2. The best method of differentiation may be through diffusion weighted MRI or CISS sequence MRI, which would show the epidermoid as a solid tissue mass, and the arachnoid cyst as a fluid filled structure. Second, the treatment for epidermoids and cysts are very different. Surgery is the mainstay for epidermoid lesions of the CPA, where symptoms of arachnoid cysts may be controlled without surgery through the use of diuretics.

Neuromas

Although acoustic schwannomas may constitute up to 90% of CPA lesions, schwannomas of other cranial nerves probably only account for 2%. Trigeminal schwannomas more commonly arise in the middle fossa and later extend into the CPA, where they may cause typical vestibular or acoustic symptoms. These lesions may present with facial numbness or trigeminal neuralgia. The lesions are radiographically similar to acoustic neuromas on CT and MRI, except for their location. When presenting with vestibulocochlear symptoms they are commonly found to arise within the region of Meckel's cave and the dumbbell into the CPA. Any neuroma lesion found within the CPA and middle fossa is most likely to originate from the middle fossa and extend to CPA (CN V lesions) rather than progressing the other direction. Facial schwannomas are important to remember because of their similarity in appearance to acoustic schwannomas. Radiographically, they can be indistinguishable from acoustic neuromas when they arise between the CPA and geniculate ganglion. The only time the diagnosis can be made with confidence is when extension beyond the geniculate ganglion into the tympanic or mastoid segments of the facial nerve occurs. Neuromas of cranial nerves IX, X, and XI may present with isolated cranial nerve symptoms, or when involving the foramen, present with a constellation of symptoms known as the "jugular foramen syndrome." This includes dysphagia (CN IX), hoarseness and possible aspiration (CN X), and shoulder weakness (CN XI). Radiographic densities and intensities of neuromas are found on CT and MRI, respectively, as well as widening of the jugular foramen. Hypoglossal neuroma may present with hemiatrophy of the tongue and enlargement of the hypoglossal canal on CT.

Petrous Apex

Cholesterol Granuloma

This is the most common primary lesion of the petrous apex. Patients often have a history of recurrent purulent otitis. Inflammation in the apical air cells leads to deposition of cholesterol debris which leads to a foreign body reaction. This reaction leads to friable vessels and repeat hemorrhage and cholesterol deposition, thus restarting the cycle. These lesions have a characteristic appearance on MRI, with intense signals on both T1 and T2 differentiating it from other lesions.

Cholesteatoma

Cholesteatoma of the petrous apex may be congenital (also known as epidermoid), or may result from extension of mastoid disease. The presentation may be similar to a middle ear cholesteatoma, with recurrent otorrhea. Radiographic findings are similar with soft tissue densities, erosion of surrounding bone. MRI may show low intensity on T1 and high intensity on T2, differentiating this from a cholesterol granuloma. Epidermoids may occur within the petrous bone or CPA.

Petrositis

Acute petrositis is an infection of the air cells of the petrous apex. This typically occurs from extension of a middle ear or mastoid infection through the peri-labyrinthine air cells, but in rare cases may be from hematogenous spread. The classic signs and symptoms were described by Guiseppi Gradenigo in 1904, and consist of otitis with or without otorrhea, retro-orbital pain, and lateral rectus palsy. The pain associated with petrositis is thought to be a result of inflammation of the trigeminal nerve in the region of Meckel's cave. Inflammation in this area may also involve the abducens nerve as it enters Dorello's canal and result in the cranial neuropathy. While "Gradenigo's syndrome" is well known, acute petrositis often occurs without all elements of the classic triad. Pain and otitis are the most consistent findings, and multiple cranial nerves in the area of the cavernous sinus may be affected. CT findings resemble those of mastoiditis, including opacification of the petrous air cells with destruction of bony septae. MR will likely show enhancement of the periphery of the lesion including local meninges and possibly the inflamed portion of the trigeminal nerve. Acute petrositis is severe diagnosis, and complications include brain and epidural abscesses, meningitis, and venous sinus thrombosis.

Ubiquitous Lesions

Dysplasias

There are a large number of dysplastic syndromes which may affect the temporal bone. These syndromes are characterized by the nature and location of the remodeling process. Some diseases may occur as single bone or monostic lesions such as fibrous dysplasia, while others occur as polyostic disease such as McCune-Albright syndrome. The remodeling process may involve only the lamellar bone, and spare the bony cortex, or it may involve the entire bone. Replacement with fibrous or cartilaginous rather than bony elements may also occur. The differences in these disease patterns are often subtle, and diagnosis by CT may be difficult. Other dysplastic diseases in addition to those above include Paget's disease, osteogenesis imperfecta, and hyperparathyroidism.

Sarcoma

These lesions are rare within the temporal bone. Chondrosarcoma may occur near the petroclival region due to the persistence of chondral bone in this area. Rhabdomyosarcoma is a disease of childhood. It frequently presents with recurrent otitis media, possibly with drainage. This tumor is rapidly expansive and may result in cranial neuropathies as it grows. Successful treatments have occurred, however, the disease is usually rapid and fatal.

Metastasis

Metastasis to the temporal bone has been found with almost any type of tumor. The most frequent lesions reported in literature include breast, lung, prostate, and melanoma being the most common. These lesions may arise at almost any location

within the temporal bone. In retrospective studies the most common symptom by far was hearing loss, with asymptomatic lesions also being frequent. Metastasis should be suspected with any lesion of the temporal bone in a person with a known history of cancer, especially those listed above.

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