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Imaging in Pulsatile Tinnitus: Case Based Review

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ABSTRACT

Tinnitus refers to auditory perception of internal origin. It is a relatively common problem and affects men and women equally. Clinically, it may be divided as pulsatile or non-pulsatile and subjective and objective. Although pulsatile tinnitus (PT) is less common, it is more likely to be associated with underlying vascular tumors, lesions or anomalies. Imaging forms the baseline for evaluation of objective tinnitus, primarily in the form of computed tomography or magnetic resonance imaging. We present a review of common causes of PT, along with emphasis on key imaging findings.

Keywords: Tinnitus, Glomus tumor, Pulsatile, Intracranial hypertension, Dural arteriovenous fistula

INTRODUCTION

Tinnitus is defined as an auditory perception of internal origin, usually localized and rarely heard by others.[1] It is relatively common and has maximum prevalence between 40 years and 70 years of age. Both men and women are equally affected.[2,3] Pulsatile tinnitus (PT), when present, is perceived as discrete repetitive sound that is synchronous with the patient's pulse. Continuous (or non-pulsatile) tinnitus, on the other hand, refers to all other rhythms, usually a constant unrelenting noise. Tinnitus may also be classified as subjective versus objective based on whether it can be perceived by the patient alone or both by the patient and the physician. Non-PT is almost always subjective, whereas PT can be subjective or objective.[2] Non-PT is also more common than PT as is subjective tinnitus than objective tinnitus [Table 1].[4,5]

PT may result from an underlying vascular tumor, a vascular malformation or etiology, developmental abnormalities, and in systemic conditions associated with high cardiac output.[4,5] Non-vascular causes of PT include lesions of middle ear, temporal bone, internal auditory canal (IAC), and cranial cavity.[3]

High-resolution computed tomography (HRCT) of temporal bone with contrast is the preferred modality, more so if a retro tympanic mass is seen on the otological examination.[3,6] Aberrant or lateralized internal carotid artery (ICA), persistent stapedial artery (PSA), high-riding jugular bulb, glomus tympanicum, or cholesteatoma are the best demonstrated on computed tomography (CT). For a patient with a normal otologic examination or with no abnormalities on HRCT, an magnetic resonance imaging (MRI) examination and an magnetic resonance angiogram (MRA) are obtained to evaluate for causes related to common carotid artery disease, ICA disease, and benign intracranial hypertension.[3,6] CT angiography/venography or conventional angiography

Table 1: Classification and common causes of pulsatile tinnitus.

Etiology	Common examples	Imaging
Neoplastic/Tumoral	Glomus jugulare Glomus tympanicum Cavernous hemangioma Vestibular schwannomas Hypervascular skull base tumors	MRI Skull base protocol
Arterial	Aberrant ICA Dehiscent ICA wall Persistent stapedial artery Aberrant anterior inferior cerebellar artery loops	CT or MRA
Venous	Jugular bulb diverticulum High-riding jugular bulb Arteriovenous fistulas AV malformations Dural sinus stenosis/IIH	CT or MRA and venogram
Skull base/Temporal bone	Paget's disease Otospongiosis	CT Temporal bones
Miscellaneous	Anemia Hyperthyroidism	Unremarkable

MRI: Magnetic resonance imaging, ICA: Internal carotid artery, CT: Computed tomography, MRA: Magnetic resonance angiogram, AV: Arteriovenous, IIH: Idiopathic intracranial hypertension

is used to demonstrate dural fistula and other rarer venous causes of tinnitus on patients in whom the initial workup is unyielding.[2,3,6] Most patients with isolated non-PT do not show any significant abnormality on imaging. In patients with PT, however, imaging may be positive in up to 57–100% of patients though about 20–30% of these patients have "normal variants" of controversial significance.[3]

TUMORAL CAUSES OF PULSATILE TINNITUS

Glomus tumors/paragangliomas of the temporal bone arise from glomus cells that accompany tympanic branch (Jacobson's nerve) of the glossopharyngeal nerve and the auricular branch (Arnold's nerve) of the vagus nerve.[2,7] These tumors may arise from the middle ear cavity (glomus tympanicum), jugular bulb (glomus jugulare) or may involve both the jugular bulb and middle ear cavity (glomus jugulotympanicum). Both glomus jugulare and glomus tympanicum tumors have a predilection for females (F:M: 3:1) in fifth and sixth decades of life. Besides PT, hearing loss, vertigo, and symptoms related to adjacent cranial nerve involvement (CN IX–XI) may be noted.[7]

Glomus tympanicum arises in the middle ear from the paraganglia along Jacobson's nerve near the cochlear promontory. These tumors are variable in size and usually present as a small soft-tissue nodule classically located at the cochlear promontory, though occasionally, the tumor may be large enough to fill the tympanic cavity and extend posteriorly into the mastoid or anteriorly in to the Eustachian canal and nasopharynx [Figure 1]. Larger tumors may encase the ossicular chain without destruction.[7,8] These tumors enhance brilliantly on the post-contrast MRI images.[3,5] However, most small tumors can be reliably evaluated on thin-section CT alone.[9,10]

Glomus jugulare tumors arise primarily in the jugular fossa while glomus jugulotympanicum involves both jugular bulb and middle ear cavity.[3,11] On CT, irregular moth-eaten cortical erosions and widening of the jugular foramen are typical [Figure 2]. Middle ear invasion with destruction of ossicular chain and adjacent bony landmarks may occur with larger lesions.[10] Intracranial or infratemporal fossa involvement again may occur in advanced tumors.[10,11] Intense homogenous enhancement is seen on the contrast enhanced CT scan. MRI may reveal a characteristic "salt and pepper" appearance on both T2 weighted (T2W) and post-contrast T1W images [Figure 3].[10,12] MRI is superior to CT in showing the location and extent of these tumors and relation to adjacent vascular structures to assess operability.[10] Multi-centricity is seen in 10% of paragangliomas.[13] Familial paragangliomas constitute approximately 10% of cases but have a higher incidence of multicentric tumors, being seen in 35–50% of cases.[12,13]

Hemangiomas are benign neoplasms that may involve the IAC, geniculate ganglion, or the middle ear cavity. Although IAC tumors usually present with sensorineural hearing loss and geniculate lesions with facial weakness, both may cause PT.[2] Geniculate hemangiomas demonstrate stippled

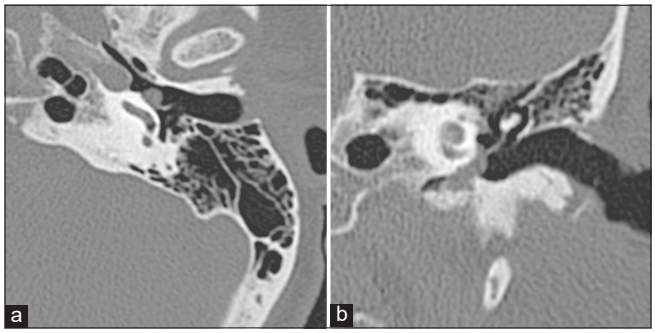

Figure 1: A 39-year-old woman with glomus tympanicum. (a) Axial high-resolution computed tomography and coronal multiplanar reformation image (b) through the middle ear shows presence of a well-defined soft-tissue mass overlying the cochlear promontory (a), and extending into the hypo tympanum.

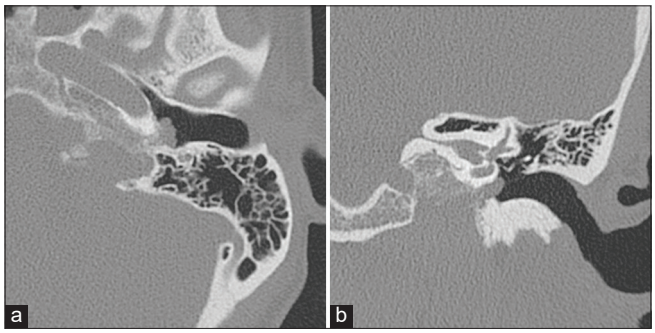

Figure 2: A 37-year-old woman with glomus jugulotympanicum. (a) Axial high-resolution computed tomography and coronal multiplanar reformation image (b) through the middle ear shows presence of permeative bony destruction in the region of the jugular bulb, associated with presence of abnormal soft-tissue in the hypotympanum.

calcifications on CT, which is rather typical [Figure 4].[14] In the IAC and middle ear, imaging appearances are non-specific with most lesions presenting as avidly enhancing soft-tissue masses, often indistinguishable from paragangliomas or neuromas [Figure 5].

PT has been described, albeit rarely, in patients with schwannomas, meningioma, hemangiopericytomas, endolymphatic sac tumors (ELSTs), and vascular metastases from breast lung and thyroid.[6] Vestibular schwannomas cause non-PT more commonly than PT and the exact mechanism for the same is not well understood.[5] On MRI, they appear as well-defined oval lobulated extra axial tumors, heterogeneously hyper intense on T2W image. Post-contrast enhancement is variable and large cystic components have been described [Figure 6].[15,16]

ELSTs are rare, destructive, hyper vascular lesions that arise from the endolymphatic sac situated in the retrolabyrinthine region. These tumors are strongly associated with Von Hippel-Lindau syndrome.[6,17] On CT scan, ESLTs appear as erosive

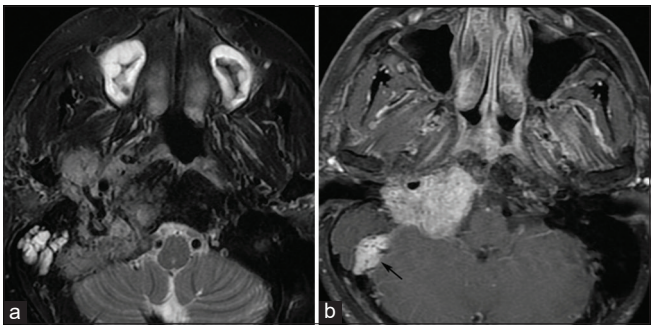

Figure 3: A 41-year-old man with advanced glomus jugulotympanicum. (a) Axial T2W fat-suppressed (T2-FS) and axial post contrast T1-FS image (b) show a large soft tissue mass centered over the right jugular bulb. Note the presence of multiple flow voids on both the T2W image and post contrast image, giving rise to the "salt and pepper" appearance. There is associated extension into the right transverse sinus (arrow).

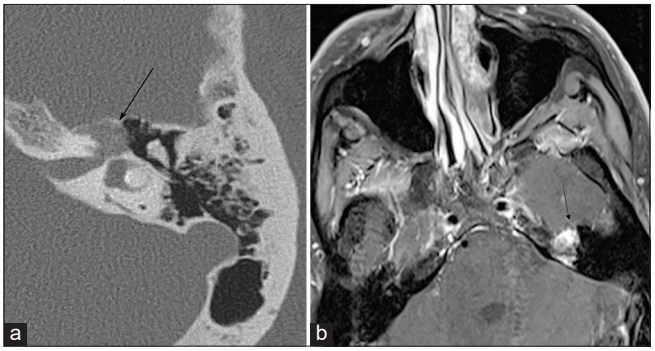

Figure 4: A 50-year-old man with facial nerve hemangioma. (a) Axial high-resolution computed tomography image shows characteristic stippled calcification in the region of the left geniculate ganglion (arrow) (b) post-contrast axial T1 fat-suppressed image reveals focal enhancement in the same region (arrow).

lesions centered on the posteromedial aspect of the petrous bone [Figure 7]. Intratumoral calcifications may be present.[18] On MRI, the ELSTs usually have a heterogeneous appearance with cystic components. Multiple high-signal intensity foci on both T1- and T2-W images may be due to blood products, proteinaceous cysts, or cholesterol clefts. Signal voids may also be seen.[6,17,18] Avid enhancement of the solid portions of the tumor is noted on post-contrast studies.[17,18]

NON-TUMORAL CAUSES OF PULSATILE TINNITUS

Arterial causes

An aberrant carotid artery results from abnormal regression of the cervical segment of the embryonic ICA, necessitating development of a collateral pathway to provide vascular supply to the developing brain.[5,19] This occurs though an anastomosis between the inferior tympanic artery (branch

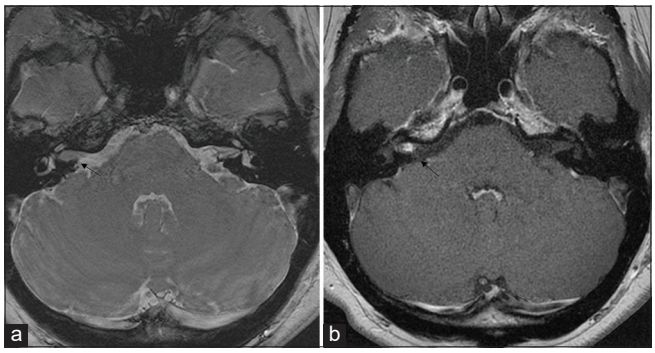

Figure 5: A 53-year-old man with vestibular hemangioma. (a) Axial constructive interference in steady state image reveals presence of a small T2 iso-intense mass in the right internal auditory canal (arrow) (b) axial post-contrast T1 weighted image reveals relatively homogeneous enhancement (arrow). Though imaging findings would be consistent with a vestibular schwannoma, this turned out to be a hemangioma on pathology.

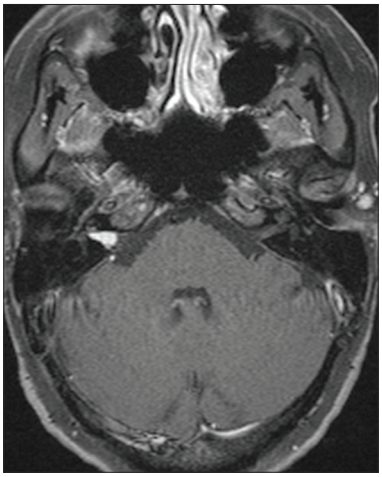

Figure 6: A 31-year-old woman with right vestibular schwannoma. Axial T1 fat-suppressed image reveals presence of an enhancing lesion in the right internal auditory canal, reaching up to the porus acusticus, consistent with a vestibular schwannoma.

of ascending pharyngeal artery) and the caroticotympanic artery (branch of the horizontal segment of ICA).[3,19] An aberrant carotid artery is, thus, a collateral pathway resulting from agenesis of the first embryonic segment of ICA.[20]

When the ICA develops normally, the inferior tympanic artery may be seen as a tiny vessel coursing through the inferior tympanic canal [Jacobson canal] between the carotid and jugular canals in the carotid-jugular spine.[5,20] Imaging in patients with aberrant ICA, however, reveals enlargement of the inferior tympanic canal with the anomalous vessel coursing through the middle ear [Figure 8].[3,5,20] Based on whether the overlying bony plate is preserved or dehiscent, otoscopy may be normal or show a pulsatile retro-tympanic mass.[5]

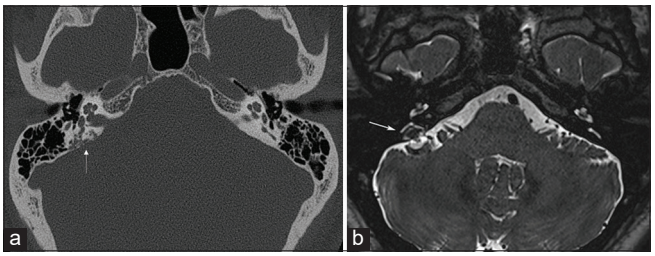

Figure 7: A 40-year-old man with endolymphatic sac tumor. (a) Axial high-resolution computed tomography image reveals focal bony destruction in the region of the right endolymphatic sac (arrow) (b) axial T2-weighted image at the same level shows a heterogeneously hyperintense mass (arrow).

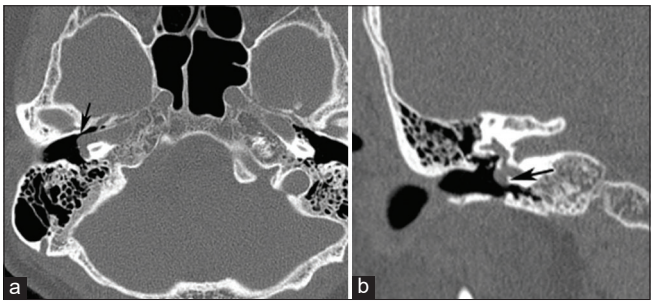

Figure 8: A 29-year-old man with aberrant right internal carotid artery (ICA). (a) Axial high-resolution computed tomography image and coronal multiplanar reformation (b) reveals abnormally aberrant right ICA (arrows), coursing through the middle ear cavity and overlying the cochlear promontory.

An aberrant ICA is often smaller when compared to the normally developed ICA.[3,20] MR and conventional angiography more clearly show the unusually lateral course of the aberrant vessel, which gives rise to the so called reverse seven sign.[3] Although it is usually unilateral, sporadic cases of bilaterally aberrant ICA have been reported.[20]

A PSA results from failure of regression of the embryonic stapedial artery, a branch of the hyoid artery.[21] During the fetal life, PSA pierces through the stapes primordium, forming the obturator foramen. It then divides in to dorsal and ventral divisions which subsequently link with the branches of the external carotid artery to form the meningeal (dorsal division) and maxillary and mandibular arteries (ventral division), respectively. In the normal course, the stem of the stapedial artery regresses completely.

The exact incidence of PSA is unclear and varies from 0.02% to 0.01% in surgical series and is about 0.48% in pathology studies. PSA may be associated with aberrant ICA in about 30% of cases. Clinically, PSA is often asymptomatic and a PSA large enough to cause PT is quite rare.[2,22]

On imaging, PSA is often seen to arise from the petrous ICA and enter the hypo tympanum in a bony canal. It then courses through the obturator foramen, subsequently travelling along

the tympanic segment of the facial nerve in the facial canal inferior to the lateral semicircular canal. It finally exits the facial canal just before the geniculate ganglion to enter the middle cranial fossa.

On imaging, it may be seen in HRCT as subtle soft-tissue thickening along the stapes foot plate and facial nerve within the middle ear. Since it replaces the middle meningeal artery, the foramen spinosum is absent [Figure 9].[5] The latter is, however, not specific for PSA and may be seen in about 3% of all skull base CT studies.

PT may also result from dehiscence of the bony carotid canal near basal cochlear turn, leading to herniation of the carotid artery into the tympanic cavity, referred to as a dehiscent/lateralized carotid artery.[3] In some cases, it may be accompanied by an aneurysm.[22] Although the embryologic development is normal in these cases, they may mimic an aberrant ICA clinically since they often show up as retro tympanic pulsatile masses. On imaging, however, the vertical segment of the carotid artery is present, and the inferior tympanic canal is not dilated.[19]

Atherosclerotic plaques and stenosis are a common cause of PT, accounting for approximately 8–20% of total cases.[6] These are felt to be the most common cause of PT in the elderly. Interestingly, the tinnitus may be ipsilateral secondary to turbulent flow or even contralateral due to compensatory increased flow in the normal vessel.[6,23]

Patients with PT may show severe stenosis (>70%), ICA involvement or even complete occlusion [Figure 10].[6] This may be seen with carotid duplex, CT, or MRA. PT in association with atherosclerosis has been reported with carotid, brachiocephalic, and even subclavian arterial involvement.[22,24]

PT may also occur in association with fibro-muscular dysplasia (FMD), an idiopathic, non-inflammatory, and non-atherosclerotic vasculopathy that usually affects middle aged women.[22,25,26] The renal arteries are most frequently involved (60–70%), followed by the craniocervical vessels (20–30%). Although symptomatic FMD most frequently manifests with cerebral ischemia, PT is the second most frequent manifestation, being seen in about 30% of the patients.[3,6,26]

Interestingly, despite the low prevalence, it is a more common cause of PT than atherosclerosis, possibly due to proximity of stenosis to the petrous bone.[27] Angiography classically shows "string of beads" appearance in up to 85% of carotid FMD [Figure 11].[22] Less commonly, imaging may show unifocal or multifocal tubular stenosis, focal involvement of arterial wall or pseudoaneurysm formation. Bilateral carotid involvement may occur in up to 60% of cases.[12] Cervicocranial FMD may be associated with intracranial aneurysms in 13–57% of cases.[25]

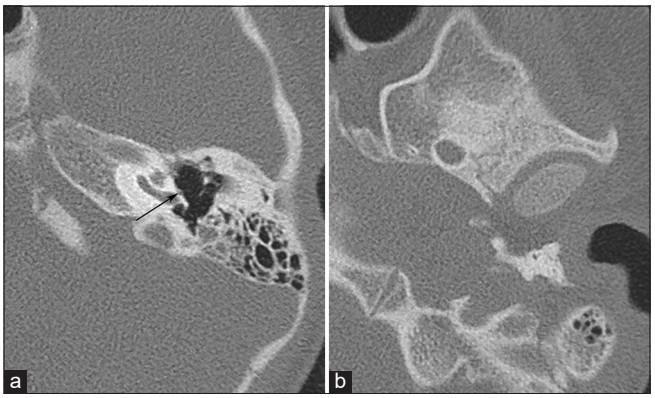

Figure 9: A 27-year-old woman with persistent stapedial artery. (a) Axial high-resolution computed tomography images at the level of middle ear and (b) floor of the middle cranial fossa. There is a tiny soft-tissue focus along the cochlear promontory (arrow). This was seen to course along the horizontal segment of the facial nerve on sequential images (not shown). Image at level of skull base (b) reveals absent foramen spinosum.

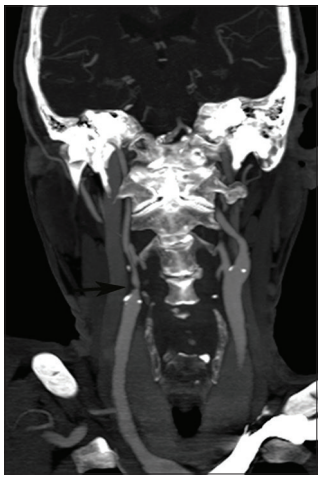

Figure 10: A 61-year-old man with carotid stenosis. Coronal multiplanar reformation computed tomography angiogram image in an elderly patient with pulsatile tinnitus shows moderate to severe narrowing of the right internal carotid artery at its origin (arrow).

Spontaneous carotid dissection often presents clinically with neck pain, cerebrovascular ischemia, or Horner syndrome, but can occasionally result in PT.[5,6,23] The intimal flap may be seen on both CT and MRA with consequent narrowing of the true lumen. Non-contrast, fat-suppressed T1W images may demonstrate the hyper intense crescentic thrombus, referred to as the "fried-egg" sign on axial images [Figure 12].[3,5,6] The dissection is often unilateral, though bilateral ICA dissection may occur [Figure 13]. Conventional angiography is usually not necessary.[5]

Aneurysms involving the petrous carotid artery are most often congenital but also be seen secondary to infection or

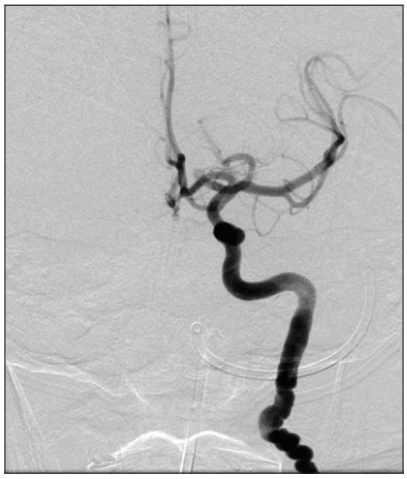

Figure 11: A 57-year-old man with fibromuscular dysplasia (FMD). Conventional angiogram, anteroposterior projection with injection of the left internal carotid artery (ICA). There is marked irregularity of the left cervical ICA which shows a beaded appearance, consistent with FMD.

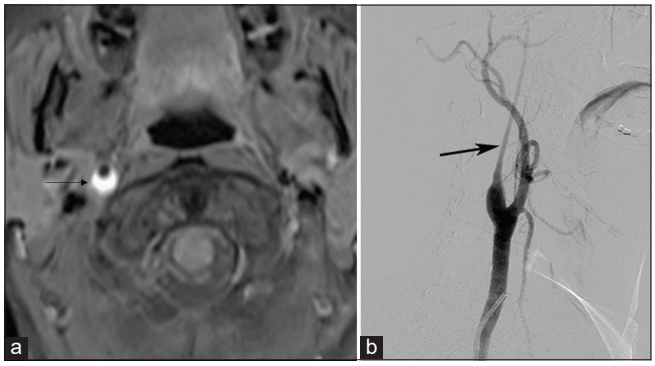

Figure 12: A 30-year-old man with carotid dissection. (a) Axial magnetic resonance T1 fat-saturated image shows T1 hyperintense eccentric mural thrombus in the right internal carotid artery (ICA) (arrow), consistent with dissection (b) conventional angiogram image in another patient with dissection reveals smooth tapering (arrow) of the ICA starting just beyond its origin.

trauma. They may present with hearing loss, PT, headache, or facial pain.[22,28] On CT, they appear as soft-tissue lesions with remodeling of the petrous canal, showing intense post-contrast enhancement [Figure 14]. MRI appearance may be variable, being susceptible to turbulent flow. In about 25% of cases, rupture of the aneurysm may be the initial presentation, manifesting clinically as a triad of otorrhagia, epistaxis, and neurologic deficit.[28]

Some controversy exists regarding PT occurring secondary to aberrant looping of the anterior inferior cerebellar artery (AICA) in the IAC. Some authors regard them as causes of PT and claim resolution of symptoms after microvascular decompression, while others claim no significant correlation between presence of these loops and PT.[6,23,29] Regardless,

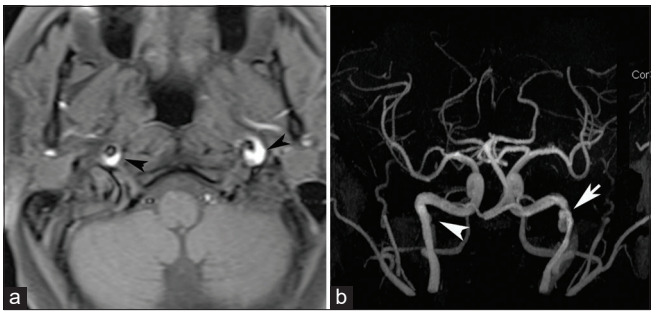

Figure 13: A 28-year-old woman with bilateral internal carotid artery (ICA) dissection. (a) Axial T1 fat-suppressed image reveals eccentric hyperintense mural thrombi involving the cervical ICA bilaterally (arrowheads) (b) multiplanar reformation image of the 3D time of flight magnetic resonance angiogram reveals focal aneurysms in the dissected segments, larger on left side (small arrow), smaller on right side (arrowhead).

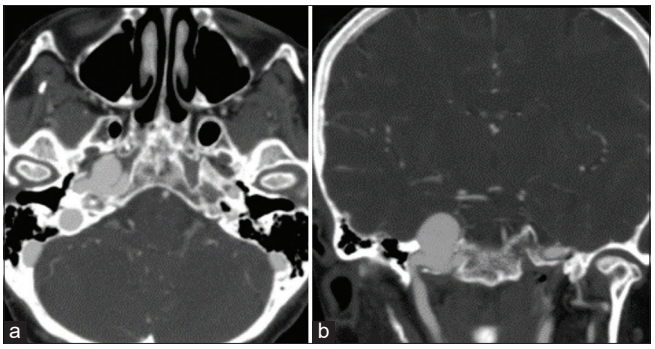

Figure 14: A 67-year-old man with right carotid aneurysm. (a) Axial computed tomography angiogram image with coronal multiplanar reformation (b) shows presence of a large aneurysm involving the petrous segment of right internal carotid artery. There is associated bony remodeling with intracranial extension through the petrous roof.

AICA loops are significantly more common in patients with PT as compared to the general population.[3,29]

Venous causes

Idiopathic intracranial hypertension (IIH), also known as pseudotumor cerebri syndrome, is characterized by increased pressure of the cerebrospinal fluid (CSF) without an identifiable cause.[30] It is more common in obese women in childbearing age group, with mean age of 30 years at the time of diagnosis.[30] IIH is thought to be the commonest etiology for PT, diagnosed clinically based on the modified Dandy criteria.[31] The main diagnostic criteria for this include papilledema with normal neurologic examination, normal CSF composition, and elevated lumbar puncture opening pressure (more than 250 mm CSF in adults). Imaging findings suggesting IIH on MRI include empty sella, distension of perioptic subarachnoid space, flattening of posterior aspect

of globe, and stenosis of the transverse sinus [Figure 15].[31] Studies have shown that flattening of the posterior globe may be highly specific for IIH (specificity 100% and sensitivity 43.5%).[3] Although the literature is limited, narrowing of the Meckel's cave and cavernous sinuses on MRI and dilation of foramen ovale on CT has also been described with IIH.[32,33] In addition, imaging is also useful to exclude secondary causes of transverse sinus stenosis such as thrombosis or compression.[34] The treatment of IIH is guided by visual symptoms since it may be the most debilitating sequel and varies from conservative measures (weight loss, low sodium diet, and/or acetazolamide) to surgical procedures such as optic nerve sheath fenestration or CSF diversion.[1] Stenting of transverse sinus may be helpful in some cases.[34,35]

Anatomical variants of the jugular bulb, a dural venous sinus prominence at the junction between the sigmoid sinus and the internal jugular vein, have been reported in association with PT and include a high-riding bulb, dehiscent jugular bulb, and jugular diverticulum. All these variants are approximately 2–3 times more common on right side.[36] High-riding jugular bulb is most common of these variants and can be seen in approximately 4–20% of normal population, even though PT is seen in only 4.5% of these patients.[37] Jugular bulb is said to be high-riding when it extends over the tympanic annulus and the round window or the basal turn of the cochlea [Figure 16].[38] Sometimes jugular bulb is called "very" high riding if it reaches to the level of IAC.[37]

Absence of the bony plate between the middle ear and the jugular bulb is referred to as a dehiscent jugular bulb [Figure 17].[38] This may manifest clinically as a bluish retro tympanic mass and be seen in association with PT.

Jugular diverticulum is an extra luminal outpouching of the jugular bulb through a bony defect [Figure 18]. It typically occurs between the posterior wall of the IAC and the vestibular aqueduct but it can also occur at the superior surface of the petrous bone, middle ear cavity, and the endolymphatic duct.[37,39] Although clear etiology of jugular bulb diverticulum is unclear, venous hypertension and turbulent venous flow leading to bony defect and herniation are the most accepted hypothesis.[39]

All of the jugular bulb variants can present with PT but since these (especially the high-riding bulb) can also be seen in asymptomatic population, other cause PT must be ruled out before any causal relationship is associated with these. These variants can be diagnosed on CT or MRI but CT is better for delineation of bony outline, excluding underlying bony dehiscence, and to assess relationship with inner ear structures.

Sigmoid sinus anomalies can also cause transmission of venous pulsations to the inner ear leading to PT. If the sigmoid sinus follows an aberrant anteromedial course along the posterior semicircular canal and endolymphatic sac, it can cause PT.[37] Similarly, a laterally placed sigmoid sinus has also been reported to be associated with PT. Less commonly; there may a defect in anterior sigmoid wall (also known as sigmoid sinus diverticulum) leading to extension of sigmoid sinus into the mastoid temporal bone.[40,41] This entity is increasingly recognized as cause of PT and these patients have been effectively treated by sigmoid sinus reconstruction.[41] Other rarely reported causes of PT include enlarged retromandibular vein, dilated mastoid emissary vein, and pre-auricular vascular malformations.[2,6,42]

Arteriovenous (AV) causes of PT

Dural AV malformation (AVM) or AV fistula (AVF) may be congenital or acquired and are characterized by abnormal communication between dural arteries and venous system. While AVM have a tangle of vessels that form the nidus, AVF is characterized by absence of an intervening nidus. The acquired causes of dural AVF (DAVF) include venous thrombosis, head injury, pregnancy, transcranial surgery, or tumors (associated with dural venous outflow obstruction).[43] In patients with normal otoscopic examination, dural AVMs (DAVMs) or AVFs are one of the most common objective cause of PT.[3] PT is typically

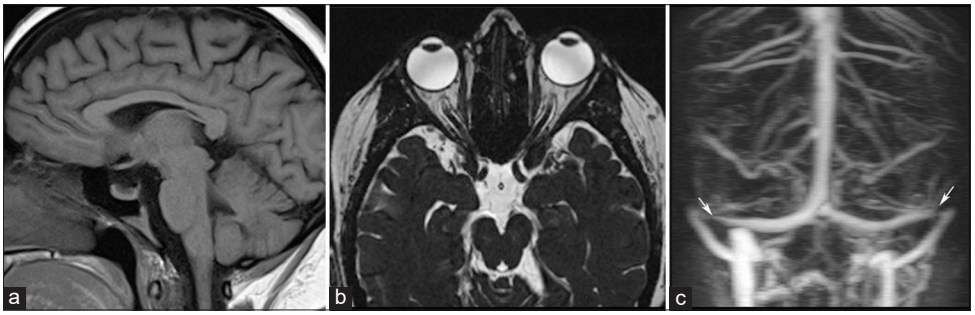

Figure 15: A 31-year-old female with intracranial hypertension. (a) Sagittal T1-weighted image shows empty sella (b) axial constructive interference in steady state image shows distension of bilateral optic nerve sheaths. (c) Coronal maximum intensity projection image of magnetic resonance venogram shows typical stenosis of transverse sinuses bilaterally (arrows).

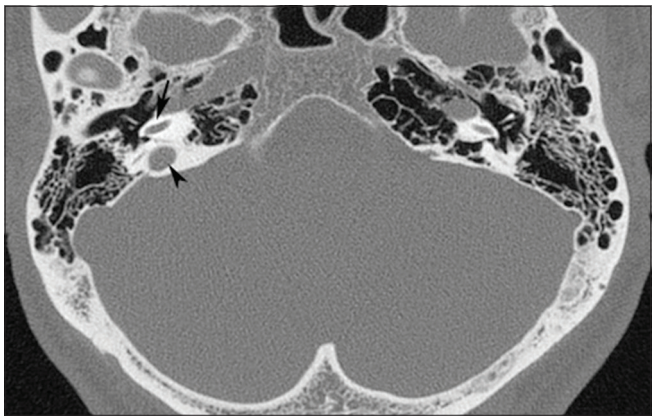

Figure 16: A 24-year-old woman with high riding jugular bulb. Axial high-resolution computed tomography image shows jugular bulb (arrowhead) at the level of the basal cochlear turn (arrow).

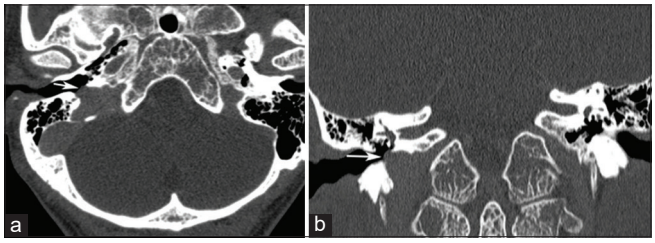

Figure 17: A 29-year-old man with jugular bulb dehiscence. (a) Axial post-contrast high-resolution computed tomography image with coronal multiplanar reformation (b) shows presence of a small bony defect involving the jugular plate with herniation of the jugular bulb in to the middle ear (arrows), consistent with jugular dehiscence.

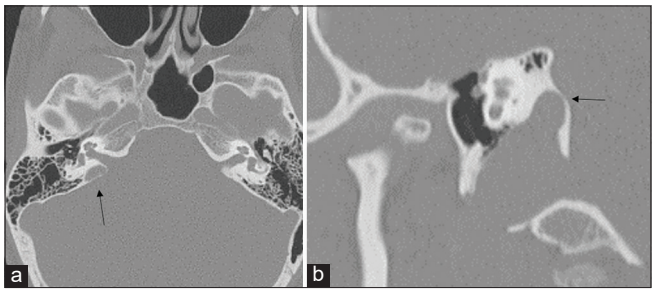

Figure 18: A 33-year-old woman with jugular diverticulum. (a) Axial high-resolution computed tomography image demonstrates a bony defect along the posterior wall of internal auditory canal (arrow) (b) multiplanar reformation image in the Poschl plane demonstrates focal outpouching from the jugular bulb (arrow), consistent with a diverticulum.

associated with venous drainage into the sigmoid or transverse sinus, although it has also been reported with cavernous sinus drainage or with extra-cranial AVF's that usually involve branches of vertebral artery.[6] DAVM or AVF are mostly supplied by branches of external carotid artery. Conventional angiography is preferred for diagnosis and a normal CT or MRI does not rule out a diagnosis of DAVF. If abnormal, non-invasive imaging may show abnormal vascular cluster beneath the skull base, dilated supplying extracranial vessel, engorged leptomeningeal or medullary veins, dilated ophthalmic vein, intracranial hemorrhage, or gliosis of the underlying brain. Sometimes, CT or MRA may show a direct fistula although it is seen only on digital subtraction angiography in most of the cases [Figure 19].[3] If symptomatic, these can be treated by endovascular intervention, radiation therapy, surgery, or combination of therapies.[43]

Miscellaneous causes of PT

Otospongiosis is a bony dysplasia that is characterized by replacement of dense normal enchondral bone of the bony labyrinth by spongy vascular bone.[6] It is more common in females and is bilateral in majority of patients. Depending on the site of involvement, it can be characterized as fenestral or retrofenestral/cochlear. Fenestral subtype is more common and classically involves the region of fissula ante fenestram just anterior to the oval window [Figure 20]. Retrofenestral or cochlear subtype is usually the extension of the disease process to the cochlear capsule, semicircular canals or IAC.[3] Although hearing loss is the most frequent manifestation, up to 65–85% of patients can have tinnitus, which may be pulsatile.[6] PT is attributed to presence of AV micro-fistulae with increased neovascularization in the abnormal spongy vascular bone.[44] In early stage, the abnormal bone seen as single or multiple radiolucent foci in region of fissula ante fenestram (fenestral otospongiosis) or around the cochlea (retro cochlear/cochlear otospongiosis). If severe, there may be a low-attenuation ring around the cochlea, also known as double-ring sign. In healing stage, there is new bone formation that is seen as "heaped-up" appearance along the oval/round window or the cochlea.[3]

MRI in the active phase may reveal high T2 signal in pericochlear and perilabyrinthine areas or post-contrast enhancement of the involved bone. These findings, however, may be absent in inactive or chronic phase of disease.[45] HRCT is, therefore, preferred over MRI for evaluation of otospongiosis.

Paget's disease often affects the middle aged and elderly population. Involvement of the temporal bone can lead to tinnitus.[3] Continuous tinnitus is reported in approximately 60% of cases, but patients can also manifest PT.[6] Similar to otospongiosis, development of AV communications is likely contributory. Imaging findings vary and may include a lytic lesion during the acute phase (osteoporosis circumscripta), diffuse cotton-wool appearance with foci of variable density in the mixed phase or osteosclerosis in the late inactive or blastic phase [Figure 21].

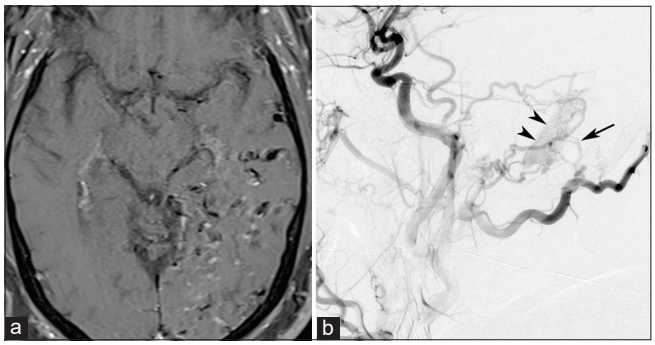

Figure 19: A 43-year-old woman with dural arteriovenous fistulas (DAVFs). (a) Axial post contrast T1 fat-suprressed image demonstrates multiple flow voids in the left temporal and occipital regions secondary to venous congestion (b) conventional angiogram with injection of the conventional coronary angiography demonstrates a prominent occipital artery with fistulous communications to the dural veins (arrow) with early venous filling (arrowheads), consistent with DAVF.

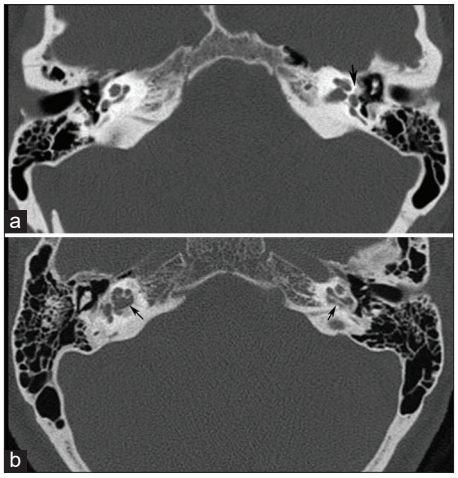

Figure 20: A 66-year-old man with otospongiosis. (a) Axial high-resolution computed tomography (HRCT) image demonstrates a small focus of decreased bone density in the region of the left fissula ante fenestram (arrow), consistent with fenestral otospongiosis (b) axial HRCT image in another patient with retro fenestral otospongiosis demonstrates low attenuation regions around the cochlea bilaterally (arrows) known as the "double ring sign."

Few common metabolic conditions such as anemia and hyperthyroidism can also result in PT. This is primarily due to increased cardiac output associated with these entities and is usually proportional in extent to the severity of the underlying condition. Imaging work in these patients is unrevealing and correction of the underlying metabolic abnormality results in resolution of tinnitus. However, these are diagnosis of exclusion and many of these patients eventually end up getting imaging work-up to rule out other structural causes.[46]

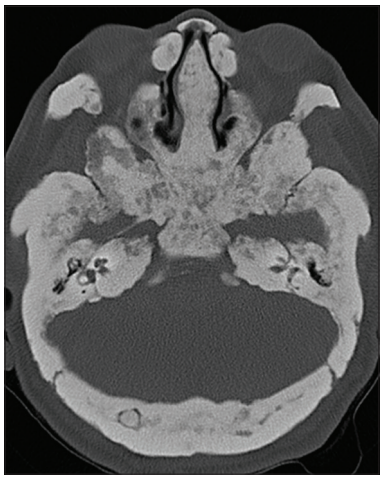

Figure 21: A 61-year-old man with Paget disease. Axial computed tomography image demonstrates extensive bony sclerosis and expansion involving the skull base.

CONCLUSION

PT is relatively common and may be successfully evaluated with imaging. The underlying etiology is variable and may include vascular or tumoral lesions. Although many anatomical variants have been associated with PT, these should be considered only after the more significant etiologies have been excluded through a comprehensive workup.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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