Imaging In Pulsatile Tinnitus: When Should It Ring A Bell?

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Learning objectives

1) To review causes of Pulsatile Tinnitus (PT) and role of different modalities in its evaluation.
2) To review spectrum of imaging findings patients with PT.

Background

Tinnitus is defined as a discrete auditory perception in the absence of any external stimulus (1,2,3). It is relatively common and has maximum prevalence between 40-70 years of age. Both men and women are equally affected (3, 4).

Tinnitus is said to be pulsatile when it is synchronous with patient's pulse. Alternately, it may be non pulsatile and continuous in nature. Tinnitus can also be classified as subjective when only the patient hears it and objective when it is heard both by the patient and physician/ another person (1, 2, 3).

More patients have non-pulsatile tinnitus than PT, and subjective tinnitus is more commonly seen than objective tinnitus (1, 2).

PT is usually a consequence of underlying vascular tumor, a vascular malformation or abnormality that may be congenital or acquired (1, 2). PT may also be seen in systemic conditions that increase cardiac output (anemia, thyroid dysfunction, heart disease) (5). Non-vascular causes of PT include lesions of middle ear, temporal bone, internal auditory canal (IAC) and cranial cavity (4).

Imaging work up of tinnitus in general can be frustrating, both to the patient and the physician (6). Most patients with isolated non-pulsatile tinnitus do not show any significant imaging abnormality. 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 (4).

High-resolution contrast enhanced CT of temporal bone is the preferred initial modality (4). If no abnormality is found, further investigation may be done with CT angiography/ venography, MRI, MR angiography or conventional angiography (3,4, 5).

Imaging findings OR Procedure details
TUMORAL CAUSES OF PT:

- Glomus tumors / paragangliomas arise from paraganglia or glomus cells that accompany cranial nerves (2). 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). Glomus tympanicum is the most common tumor involving the middle ear cavity and usually arises along the course of tympanic nerve (4). These lesions are most commonly seen over the cochlear promontory and can vary in size from a few millimeter to a mass that fills the middle ear (2, 3). There is intense post contrast enhancement, a finding better appreciated on MR as compared to CT scan (2, 4). A glomus jugulotympanicum should be considered when there is associated erosion of floor of middle ear cavity (4) with involvement of both the jugular bulb and the middle ear [Fig 1]. Glomus jugulare arise in the jugular bulb and may secondarily involve the middle ear. These lesions may cause erosions of the anterior and lateral walls of the bony jugular fossa, a finding best appreciated on HR CT scans (2) [Fig-2]. Jugular spine erosion is also common (4). Hypertrophy of the feeding inferior tympanic artery may manifest as enlargement of inferior tympanic canaliculus (2). MR imaging may reveal a characteristic salt and pepper appearance (2, 4). As with glomus tympanicum, there is intense post contrast enhancement. Both glomus jugulare and jugulotympanicum may grow in to the neck with in the lumen of internal jugular vein (3) [Fig-3].

- Hemangiomas are rare benign neoplasms that may also present with PT. These lesions may be seen in the IAC, geniculate ganglion or in the middle ear cavity. Although IAC tumors usually present with SNHL and geniculate lesions with facial weakness, all of these tumors may cause PT (2). Except for geniculate hemangiomas, which may have a characteristic stippled appearance on HRCT, most of these lesions present as non-specific soft tissue lesions that show prominent post contrast enhancement. Infact, lesions confined to the middle ear may be indistinguishable from glomus tympanicum on imaging (2).

- PT may rarely be seen in patients with acoustic neuroma or meningioma, although the exact mechanism is unclear (7). Similarly, lesions involving the jugular fossa may also present with PT, presumably secondary to compression of jugular vein (8).

- PT has also been rarely reported in association with cholesterol granuloma and histiocytosis of the petrous bone (9,10).

VASCULAR CAUSES OF PT
These may be arterial, venous or arteriovenous in etiology. Clinically, venous tinnitus differs from arterial tinnitus. Venous tinnitus is usually right sided, since the right jugular fossa is larger than the left in majority of population. It is heard as a continuous murmur with accentuation during systole and may be abolished by compressing ipsilateral jugular vein (9).

**ARTERIAL CAUSES:**

- **Aberrant carotid artery:** It is a rare congenital anomaly that occurs secondary to abnormal regression of cervicalICA during embryogenesis (4). This is then compensated by hypertrophy of the inferior tympanic artery, which anastomoses with the caroticotympanic artery in the middle ear, thereby restoring flow to the petrous part of carotid artery (2, 4). On imaging, there is enlargement of the inferior tympanic canalculus with presence of a lateralized carotid artery coursing through the middle ear (2) [Fig-4, 5]. The ipsilateral vertical segment of carotid canal is absent (4,9). On MRA, the abnormal lateral course of aberrantICA is termed as the reversed-7 sign (4).

- **Persistent stapedial artery (PSA):** it may arise from an aberrant carotid artery, a laterally displaced or even a normal carotid artery (11). A PSA large enough to cause PT is quite rare (4, 9). The artery, when present, courses through the obturator foramen and enlarges the tympanic facial nerve canal before entering the middle cranial fossa (4). Since it replaces the middle meningeal artery, the foramen spinosum is absent (2).

- **Dehiscent/lateralized carotid artery:** This usually occurs secondary to dehiscence of bony carotid canal near basal cochlear turn with consequent herniation of the carotid artery into the tympanic cavity (4). In some cases, it may be accompanied by an aneurysm (9).

- **Atherosclerosis:** Atherosclerotic plaques may cause PT secondary to turbulent flow [Fig-6]. However, this is an uncommon cause of symptomatic PT, possibly be due to lack of proximity between the stenosis and petrous bone (12). Nonetheless, cases of PT have been reported secondary to atherosclerotic involvement of carotid, brachiocephalic and subclavian arteries (9, 13).

- **Fibro muscular dysplasia (FMD):** This is a non-inflammatory, non-atherosclerotic vasculopathy that usually affects young to middle aged women (6, 9). Although much less common than atherosclerosis, it is otherwise a more common cause of PT than atherosclerosis, possibly due to proximity of stenosis to the petrous bone (12). PT is the second most common presentation after cerebral ischemia. Angiography classically shows "string of beads" appearance in up to 85% of carotid FMD (9).

- **Carotid dissection:** This may rarely manifest as PT (4) [Fig-7]. Non-contrast, fat-suppressed T1W images may demonstrate the hyper intense thrombus (2). The intimal flap may be seen on both CT and MR angiography with
consequent narrowing of the true lumen. Conventional angiography is usually not necessary for diagnosis (2) [Fig-8].

- Aneurysms involving the petrous carotid artery may have PT as the initial presenting complaint (9).
- Aberrant AICA loops: These are of controversial significance. Some authors regard them as causes of PT and claim resolution of symptoms after micro vascular decompression, while others claim no significant correlation between presence of these loops and PT (1, 2, 14).

VENOUS CAUSES

- Idiopathic intracranial hypertension (IIH): This is reported to be the most common diagnosis in patients with PT (2, 4). Although the exact pathogenesis is not clear, there is direct relation with CSF pressure (9). Patients are usually young, obese women. On MR imaging, there may be flattening of posterior globe, distension of optic nerve sheath, tortuous optic nerve or empty sella turcica. Flattening of the posterior globe is the only sign that is considered to be suggestive of IIH (specificity 100%, sensitivity 43.5%) (15). In most cases however, the diagnosis is mostly clinical and can be confirmed by elevated opening pressure and prompt relief in tinnitus on CSF drainage (2, 9).
- Transverse sinus stenosis: Focal stenosis of venous sinus may cause tinnitus, presumably due to turbulent flow. This may be demonstrated on both MR and CT venography. Some patients may respond to dilatation and stent placement (4).
- Jugular bulb variants: A high riding jugular bulb by itself is unlikely to be a cause of PT, given it's high prevalence in normal population (9) [Fig-9]. In some cases, dehiscence of the sigmoid plate may result in extension of jugular bulb in to the middle ear cavity (2, 4) [Fig-10, 11]. Another anatomical variant is jugular bulb diverticulum that is defined as a focal protrusion of the jugular bulb in to the deep temporal bone (4). This may be associated with PT (2). In general, however, in patients with PT and presence of any of the above venous anomalies, another treatable cause should be excluded (2, 4).
- Venous anomalies/ variants: An aberrant sigmoid sinus which runs in close proximity to the posterior semicircular canal and endolymphatic sac may also present with PT (2). Similarly, a laterally placed sigmoid sinus has also been reported to be associated with PT.
- Other reported causes of PT include enlarged retromandibular vein and preauricular vascular malformations (9).

ARTERIOVENOUS CAUSES

- Dural AVF: Most dural AVF are acquired and usually involve the transverse, sigmoid and cavernous sinuses (4, 9). These account for 10-15% of intracranial AV malformations but are the most common cause of objective
PT in patients with a normal otoscopic examination (4, 9, 16). Although CT/ MR imaging may show dilated dural venous sinuses, leptomeningeal vessels, cortical veins or prominent extra cranial vessels, the exact site of fistula is often not demonstrated and conventional angiography remains the most sensitive diagnostic tool (2, 4) [Fig-12].

- Otospongiosis: PT may be seen occasionally in these patients, presumably secondary to arteriovenous micro fistulas and neovascularization (17). On imaging, radiolucent foci may be seen in region of fissula ante fenestram (fenestral otospongiosis) or around the cochlea (cochlear otospongiosis) (2, 4) [Fig-13, 14].
- Paget's disease: It is a rare cause of PT, which is again presumably secondary to intraosseous shunts (2, 18) [Fig-15].
- Cerebral and head and neck arteriovenous malformations: These may rarely present with PT [Fig-16].

Images for this section:
Fig. 1: Axial T2W fat suppressed image shows presence of a heterogeneous intensity mass involving the region of right jugular fossa and extending in to the right middle ear cavity. Multiple flow voids are seen within the mass. Posteriorly, it is seen to extend into the right sigmoid sinus (black arrow). Fluid opacification of right mastoid cells is also seen.
**Fig. 2:** Axial high resolution CT image from a different patient. There is permeative destruction involving the walls of the jugular fossa on the right side with extension in to the petrous bone. The mass was seen to extend in to the middle ear cavity (not shown), in keeping with a jugulotympanicum.
Fig. 3: Post contrast axial fat suppressed image from same patient as Fig-1. The mass is seen to involve the jugular bulb with extension into the cranial cavity posteriorly and right para pharyngeal space anteriorly. The uninvolved left jugular bulb is also seen (white arrow).
**Fig. 4:** Axial, non contrast high resolution CT image from a patient who presented with PT. Aberrant course of the right carotid artery, which is seen to traverse the right middle ear cavity (black arrow).

**Fig. 5:** Coronal image, same patient as Fig-4. The artery is seen to abut the cochlear promontory and lie directly opposite the tympanic membrane, which would also explain characteristic appearance on otoscopy.
Fig. 6: CT angiogram, coronal MIP image in a patient with right sided PT. There is focal high grade stenosis of the right ICA (black arrow).
Fig. 7: Axial T2W image in a patient with carotid dissection and PT. There is circumferential wall thickening of the right ICA (white arrowhead). The left ICA appears normal (white arrow).
**Fig. 8:** Conventional angiogram image from the same patient. There is long segment smooth narrowing of the right ICA, giving a string like appearance (black arrow).

**Fig. 9:** Axial high resolution CT image in a patient with PT. The right jugular bulb is high lying and is seen at the level of basal cochlear turn (black arrow).
Fig. 10: Axial post contrast high resolution CT image in a patient with PT shows dehiscence of jugular plate with herniation of the jugular bulb into the middle ear cavity (white arrow)

Fig. 11: Coronal MPR reformation from the same scan again reveals dehiscence of the jugular bulb, which is seen to project into the floor of the middle ear cavity.
Fig. 12: Conventional catheter angiogram in a patient with PT. Selective contrast injection into the left ECA reveals presence of a dural fistula which is supplied by the ascending pharyngeal (arrowhead) and occipital branches (black arrow) of left ECA.
Fig. 13: Axial high resolution CT image shows a tiny lucent focus involving the left fissula ante fenestram (black arrowhead), in keeping with fenestral otospongiosis.
Fig. 14: Axial high resolution CT from a different patient. Curvilinear lucency along the cochlear turn is seen on left side, suggestive of a ‘third cochlear turn sign’ (black arrow). Similar lucencies are also noted on the right side (black arrowhead).
Fig. 15: Axial CT image in a patient with known Paget's disease shows extensive bony sclerosis and expansion involving the skull base.
Fig. 16: Catheter angiogram in a patient who presented with a skull lump and PT following blunt trauma. Catheter angiogram reveals a complex network of vascular channels involving the scalp with multiple feeding arteries and enlarged draining veins. The patient subsequently underwent partial embolization of the vascular malformation with resolution of PT.
Conclusion

PT is relatively common and may be successfully evaluated with imaging. However, considering the overlap of findings in patients with and without PT, close correlation of findings with the patient’s symptoms remains critical before any intervention.

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References