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Imaging Case of the Month: Autophony in a Patient with Giant Cell Tumor of the Temporal Bone

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Giant cell tumor (GCT) of the temporal bone is a relatively rare but well-documented benign bone tumor. Presenting symptoms often include headache and involvement of cranial nerves(1). Autophony, or the unusually loud or disturbing sound of a patient's own voice, can be a symptom of several disorders. It is often accompanied by hyperacusis to one's body sounds(2). We describe the case of a woman who developed GCT of the temporal bone with autophony, tinnitus, and left-sided conductive hearing loss and symptom persistence following incomplete resection. To our knowledge, this is the first case reported of autophony in a patient with giant cell tumor of the temporal bone and demonstrates how a soft tissue lesion impinging on the ossicles can be a source of autophony.

CASE REPORT

The patient is a 54 year old woman with a previous history of giant cell tumor of the temporal bone who presented to our clinic for management of persistent left-sided autophony and pulsatile tinnitus. She also complained of left ear pain, hearing loss on the left, dry mouth, and decreased taste sensation on the left side of her tongue. Previous work-up at an outside clinic discovered a mass in the external auditory canal which appeared to impinge on the ossicles and extended anteriorly towards the temporomandibular joint and temporal lobe. Pre-operative audiogram a significant sensorineural hearing loss above 4kHz. At lower frequencies the hearing loss was conductive with an air-bone gap of 35, 25, and 15 dB at 0.5, 1, and 2 kHz. Biopsy revealed giant cell tumor and incomplete resection of the tumor was performed approximately 10 months before presenting to our office.

Physical examination revealed an erythematous mass in the anterior superior portion of the external auditory canal abutting the tympanic membrane and malleus that was hard on palpation. Our review of the CT and MRI scans already performed following incomplete resection revealed residual tumor at the anterior border of the petrous ridge (Figure 1). Preoperative pure tone audiometry of the left ear revealed an air conduction (AC) of 40 bone conduction (BC) of 75dB, with a 35dB air-bone gap.

A left preauricular approach to the middle fossa was used through the previous craniotomy site. On initial dissection of the tumor from the temporal bone, the mass was found to extend anteriorly and into the middle ear space where the tegmen was eroded. The tumor was carefully removed from the malleus, which remained intact and mobile, as well as from the posterior aspect of the external auditory canal. A tympanomeatal flap was raised to address the tumor involving the anterior external auditory canal, and a complete resection was achieved. Bone grafts were used to reconstruct the canal wall and repair the tegmen defect.

In follow-up, the patient reported complete resolution of autophony and other symptoms as well as air-bone gaps that were improved by 15 dB, 0 dB, and 10 dB at 0.5, 1, and 2 kHz. At higher frequencies, the hearing loss was predominately sensorineural and air conduction thresholds remained within 5 dB of pre-operative levels.

DISCUSSION

We report a case of autophony in a patient with giant cell tumor (GCT) of the temporal bone. GCTs are generally benign, locally aggressive lesions of the metaphysis of long bones. GCTs in the skull are rare, but most commonly occur in the sphenoid and temporal bones, which develop by endochondral ossification unlike other skull bones(1). GCTs of the temporal bone commonly present with pain on the affected side, hearing loss, swelling in the affected region, facial weakness, headache, and involvement of cranial nerves(3). They have a high recurrence rate and a risk for malignant transformation and potential to metastasize (most often to the lungs)(4). The differential diagnosis for lesions in the middle ear includes more common lesions including cholesteatomas, adenomas, paragangliomas, sarcomas, vascular variants, and Langerhans cell histiocytosis.

GCTs of the temporal bone frequently manifest on CT as mixed-density masses with varying amounts of bone destruction. Some cases demonstrate calcification and cystic change. Tumors are often heavily vascularized and are, thus, usually contrast-enhancing(1,3). GCTs consist of three types of cells: round monocyte-resembling cells, spindle-shaped fibroblast-like stromal cells, and multinucleated giant cells. Grossly, the tumors are gray to yellow-brown and may be soft, or firm and friable. Surgical resection is the primary management of choice(3).

Autophony is a common symptom of several disorders and may be accompanied by hyperacusis to one's body sounds. The differential diagnosis for a patient presenting with autophony is broad. Autophony may occur with canal occlusion (e.g. hearing aids, cerumen) but is classically associated with superior canal dehiscence syndrome (SCDS)(2) and patulous eustachian tube (PET).

In SCDS, the thinning or absence of bone overlying the superior canal acts as a third window into the inner ear, exposing the membranous semicircular canal to the middle cranial fossa. The third window then allows vibration to be transferred from the soft tissue into the cochlea., resulting in autophony. The current case demonstrates a novel mechanism by which soft tissue in the middle fossa can transmit vibration to the inner ear.

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Figure 1.

Top: Non-contrast CT, coronal view, soft tissue windows. Residual tumor in the left anterior border of the petrous ridge (black arrow). Bottom: Same cut, bone windows, tumor location shown with white arrows.

Figure 2. T1-weighted fat-suppressed MRI following gadolinium, coronal view showing $1.9 \times 1.8 \times 0.7$ cm enhancing mass in squamous portion of the left temporal bone (white arrows).