Epidermal Inclusion Cyst of the Styloid Process: A Case Report

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ABSTRACT

We report an unusual case of epidermal inclusion cyst of the styloid process which presented to us with decreased hearing and tinnitus. The patient underwent complete excision via a combined postaural cervical approach. The unusual location of the tumor and the absence of a history of trauma or intervention in that area added confusion to the diagnosis, which was finally made on histopathology.

KEYWORDS: Epidermal inclusion cyst, styloid process, intraosseous cyst, aneurysmal bone cyst

Epidermal or epidermoid inclusion cysts have been infrequently seen in bone. Such cases occur most frequently in the flat intradiploic calvarial bones and terminal phalanges,¹ with occasional mention of cases of the temporal bone, sphenoid,² or maxilla. We present an unusual case of epidermal inclusion cyst of the styloid process mimicking a glomus jugulare tumor. To the best of our knowledge, such an unusual site of presentation has not been mentioned in the literature.

CASE REPORT

A 26-year-old male patient was referred to a specialized cancer center with a history of decreased

hearing in the right ear over the past 3 months. This patient had a history of associated unilateral tinnitus. There was no history of otorrhea, otalgia, prior ear surgery, or any trauma to the head-neck region. Examination of the ear revealed a non-pulsatile bulge in the anteroinferior wall of the right external auditory canal with an intact tympanic membrane, which was hard in consistency. A neurological examination of the patient was normal. Pure tone audiogram revealed a mild conductive hearing loss of 30 dB in the right ear. A computed tomography (CT) scan showed a homogeneously enhancing soft tissue mass in the region of the right jugular foramen and styloid process, abutting the petrous bone and posterior border of the carotid canal, and bulging into the external auditory canal,

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Figure 1 Coronal computed tomography image showing an enhancing soft tissue mass in the region of the right jugular foramen bulging into the external auditory canal.

raising a suspicion of glomus jugulare tumor (Fig. 1). However, due to the clinical suspicion, it was decided to supplement the imaging with magnetic resonance imaging (MRI) scan. The MRI revealed a lobulated soft tissue mass involving the right styloid process with a smooth, well-defined outline with an intermediate signal intensity on T1-weighted, hyperintense with fluid-fluid levels on T2-weighted, and moderate homogenous postcontrast enhancement (Figs. 2–4). The lesion bulged into the inferior portion of the right middle ear cavity; however, there was no evidence of destruction of the middle ear or ossicles. These MRI features gave a probable diagnosis of giant cell tumor or aneurysmal bone cyst. Surgery was offered to the patient.

A postaural incision continuing inferiorly into the neck was taken to approach the styloid process via the cervical route. After identifying the internal jugular vein, external and internal carotid arteries, and preserving the tenth, eleventh, and twelfth cranial nerves, the parotid fascia was elevated to identify the posterior belly of the digastric. The facial nerve was identified at the stylomastoid foramen. Then, the cystic mass involving the styloid process was identified and dissection carried all around. However, during dissection, the

Figure 2 T1-weighted magnetic resonance image showing an intermediate signal intensity involving the styloid process.

cyst wall was accidentally punctured and so, after completely draining the cyst, further medial dissection was continued. The whole styloid process had been thinned out due to pressure erosion and had to be removed piecemeal. The hypotympanum was drilled and the facial nerve was exposed to its extratemporal course. The internal carotid artery was bare due to pressure erosion of the overlying bone; hence all the epithelial remnants were

Figure 3 Axial postcontrast fat-saturated T1-weighted magnetic resonance image of a lesion involving the styloid process bulging into the middle ear cavity.

Figure 4 Coronal short T₁ inversion recovery image with lesion involving the styloid process bulging into the middle ear cavity.

carefully dissected off the internal carotid artery and cranial nerves. Fat was used to obliterate the resultant defect.

On histopathology, sections showed an intraosseous cyst containing keratin debris. The cyst wall was composed of true epidermis formed by stratum basalis, stratum granulosum, and stratum corneum (horny layer), and subepithelial fibrovascular tissue showing mild chronic inflammation, giving a diagnosis of epidermal inclusion cyst of bone (Fig. 5).

Postoperative recovery was uneventful with normal facial function and decrease in symptoms. The patient is under observation and is presently asymptomatic after 3 years of follow-up.

DISCUSSION

Epidermal inclusion cysts are rare tumors that occur from the inclusion of epidermal elements into the dermis after trauma. These cysts have a predilection for the frequently traumatized sites of the body like fingers, soles, and palms. Rarely do they affect bone, with a predilection for the flat intradiploic calvarial bones of which around 250 cases have been described in the world literature.¹ However, a few cases of inclusion cysts occurring at unusual sites like the sphenoid,² infratemporal fossa, orbit, and parapharyngeal space have also been described. We believe that our case of an epidermal inclusion cyst of the styloid process has never been described before.

Epidermal inclusion cysts can be congenital or acquired. Congenital epidermoid cysts of the anterior fontanelle or those that are orogenital in location presumably result from the sequestration or trapping of epidermal rests along embryonic fusion planes during development. The acquired

Figure 5 Intraosseous cyst lined by true epidermis and containing keratin debris. (A) Hematoxylin & eosin, 40 $\times$. (B) Hematoxylin & eosin, 200 $\times$.

cysts arise from inclusion of epidermal structures in the dermis and other deeper tissues²⁻⁴ and are most likely as a result of sharp penetrating trauma, either accidental or iatrogenic (e.g., after surgery). Cases have been reported where epidermal inclusion cysts have been found in the parotid region after temporalis fascia harvest,⁵ in the parapharyngeal space after mastoidectomy,⁶ and after facial nerve decompression.⁷

Occasionally, they may arise following blunt trauma where the epithelial cells are driven down into the deeper tissues as suggested by the epithelium implant theory.⁸ However, in our case there was no history of any surgical intervention or any remembered blunt trauma to the head and neck region, suggesting a possible congenital origin.

The presentation of epidermal inclusion cysts is very varied. In the hands and feet, they generally present as asymptomatic slow-growing masses. However, in the head-neck region, there have been reports of symptoms due to pressure erosion. White and colleagues² described an intradiploic epidermoid cyst of the greater wing of the sphenoid that presented with seizures due to intracranial rupture. Sargent and associates⁹ reported an epidermoid cyst of the greater sphenoid wing that caused unilateral proptosis and loss of visual acuity. Our patient, however, presented only with tinnitus and a conductive hearing loss. The findings of a bulge in the external auditory canal raised our suspicion, leading to the imaging that aided the diagnosis.

With MRI, an epidermoid cyst has a low signal with T1 weighting and a high signal with T2 weighting. In comparison, aneurysmal bone cysts (ABC) reveal well-delineated expansile lesions surrounded by a fibrous capsule with hypointense signal intensity on all sequences with internal septations with hypointense signal intensity, separating the multiple cystic components of the lesion, which show up as fluid-fluid levels.^{10,11} Giant cell tumors, on the other hand, reveal low-signal intensity regions on T2-weighted images and intermediate signal intensity on T1-weighted

images which are related to the presence of hemosiderin from prior hemorrhage.

The ultimate diagnosis of epidermal inclusion cysts is by histopathology. On histopathology, the epidermal inclusion cyst is basically a keratin-filled cavity lined by stratified squamous epithelium usually with an apparent granular layer. Morphologically, it must be distinguished from a dermoid cyst. The dermoid cyst is also lined by keratinizing stratified squamous epithelium; however, it contains one or more skin appendages, such as sebaceous glands, sweat glands, or hair follicles, and the lumen is filled with keratin or sebaceous material.

The treatment of such lesions is complete surgical excision with care to excise the thinned-out bony wall. Failure to do so may result in recurrences.¹ However, care must be taken to avoid damage to vital structures, in which cases complete excision may not be possible. Delaying surgical management in patients with lesions in critical areas may lead to permanent nerve deficits due to pressure erosion.

The follow-up of these patients must be done clinically as well as by radiological means.

CONCLUSION

Epidermal inclusion cysts of bone in the head-neck region are uncommon. Any pressure symptoms associated with swelling must raise a suspicion of such entities especially with a past history of trauma or intervention. However, inclusion cysts can still occur spontaneously, possibly due to the sequestration of epidermal rests. The treatment in these cases is complete excision, with removal of all epidermal components.

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