

Clinical Image

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Atypical presentation of an intradiploic epidermoid cyst

Clinical Presentation

A 31-year-old female patient with a history of right canal wall down mastoidectomy with meatoplasty for extensive primary acquired cholesteatoma at the age of 6 years, now presented with right ear pain, discharge, and trismus lasting for one month. Symptoms did not respond to medical treatment in another center in the form of a full course of both oral and ototopical antibiotics. On examination, there was a supra and preauricular well-demarcated firm swelling pushing the right auricle anteroinferiorly. The skin of the superior and anterior walls of the right external auditory canal was tender and edematous. There was discharge and granulation tissue emanating from the area of the temporomandibular joint (TMJ). We noted a bony defect in the same area as well as keratin flakes with suctioning. The mastoid cavity was lined by healthy skin and the middle ear by dry, healthy mucosa separated from the external auditory canal by a large central perforation in the tympanic membrane. There was no facial nerve weakness or objective sign of malocclusion. Upon these findings, we suspected that the patient's otalgia and otorrhea arose as a result of the mass. Audiologic assessment revealed a right-sided, moderately severe conductive hearing loss with speech reception threshold at 55 dB, speech recognition score of 95%, and normal hearing in the left ear. Computed tomography of the temporal bone showed a soft tissue mass occupying the space between the 2 bony plates of the right squamous bone, with erosive changes of the outer and inner plates. There had been erosion of the middle fossa floor laterally and over the TMJ capsule (**Figures 1A & 1B**). Upon reviewing a CT that was obtained 2 years earlier, we noted an aeriated right squamous temporal bone with hypoplastic condyle and ramus of the mandible as well as a small intra-diploic soft tissue mass without bone destruction (**Figure 1C**).

Magnetic resonance imaging of the brain revealed heterogeneous soft tissue attenuation within the right temporal region that occupied the right squamous bone. It measured 2.8 by 3.7 cm. The lesion was inseparable from the ipsilateral temporalis muscle, sitting on the superior aspect of the mastoid cavity and attic region. It displayed low signal intensity on a T1 weighted image (T1WI), heterogeneous intensity on a T2 weighted image (T2WI) with diffusion restriction, and peripheral faint enhancement (**Figure 1D**). We noted pachymeningeal enhancement of the right temporal region with non-sizable subdural collection and no intracranial extension.

Clinical Image

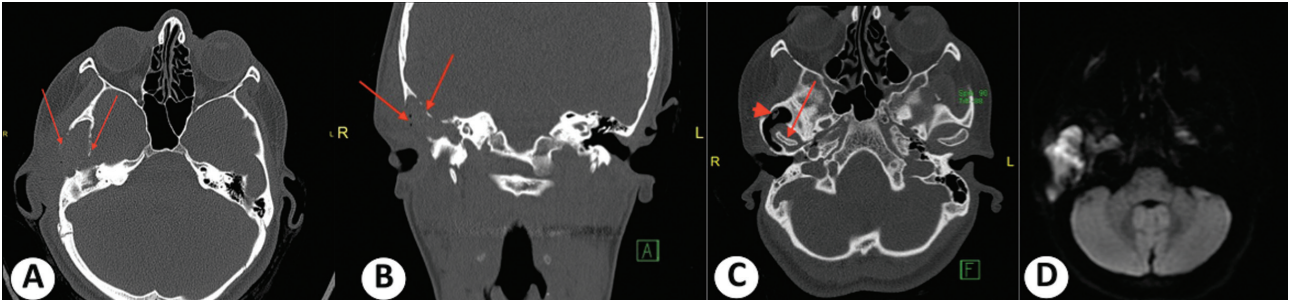


Figure 1 - Computed tomography showing the A & B) temporal bone of a soft tissue mass occupying the intradiploic space of the right squamous bone with erosive changes of the outer and inner bony plates (arrows), C) temporal bone of an abnormally aeriated squamous temporal bone with hypoplastic condyle, ramus of the mandible (arrow), and intact bone with small soft tissue shadow (arrow head). D) Magnetic resonance imaging of the brain showing marked hyperintensity on diffusion-weighted imaging within the right squamous temporal bone.

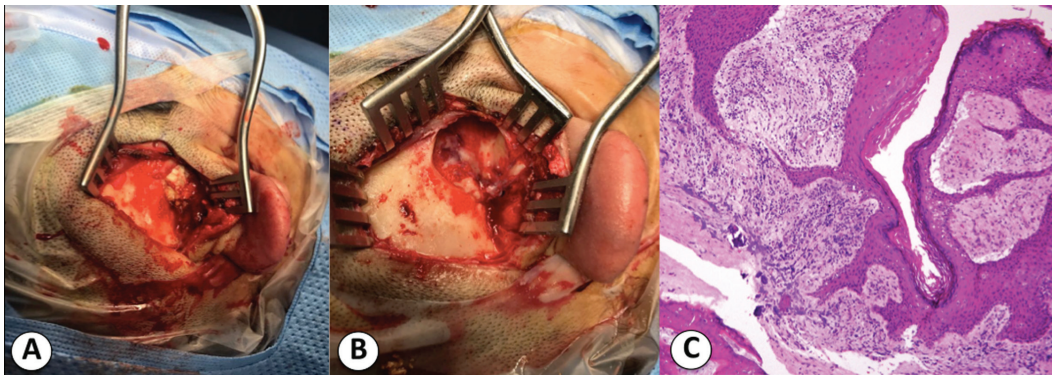


Figure 2 - Intraoperative images showing A) before and B) after complete excision of the epidermoid cyst. C) Histologic photomicrograph of the cyst wall (100x, Hematoxylin and eosin stain).

Questions

1. What is the diagnosis?
2. What is the etiology of this condition?
3. What is the management?

Answers

1. The diagnosis was an epidermoid cyst (EC) of an aerated squamous part of the temporal bone. The distinguishing characteristics in this case, which raised our suspicion of the diagnosis, were a defect in the anterior wall of the external canal to the TMJ from which pus and keratin could be suctioned, clean mastoid and middle ear cavity on examination, the earlier radiologic evidence of an abnormally aerated squamous bone with a small soft tissue shadow inside it, and a hypoplastic condyle and ramus of the mandible. The positive history of cholesteatoma during early childhood in this patient was a misleading factor to the possibility of recurrence of the condition.
2. Epidermoid cysts could either be congenital or acquired. Ectoderm implantation at the time of merging of the epithelial fusion lines can lead to the formation of congenital ECs.¹ Traumatic implantation of epidermal tissue; however, leads to acquired EC development.²
3. The patient underwent surgical excision of the EC through a supra-auricular skin incision. Total removal of the EC was accomplished with closure of the defect between the squamous bone and the mastoid cavity (Figures 2A & 2B). Histopathological study confirmed the diagnosis of EC (keratin type) with reactive bone trabeculae without malignancy (Figure 2C). The patient is on regular follow up with the otology clinic without any reported adverse events following her surgery and no recurrence of her symptoms. At follow up visit 2 years post surgery, she was found to have complete functional recovery.

Discussion

Epidermoid cysts are slow-growing lesions of the skull, which may be congenital or acquired. They are rare lesions of the cranial bones with the intradiploic type being far less common.^{3,4} Intradiploic ECs are mostly located in the parietal and frontal bones rather than the temporal bone.⁴ Patients present with a soft mass within the calvarial bone or with symptoms related to pressure effects. Symptoms of headache, dizziness, tinnitus, change of mental status, and seizures have been reported in the literature.^{4,5}

We discuss the case of an intradiploic epidermoid cyst of the squamous part of the temporal bone, which presented as otitis externa. In our patient, the intradiploic EC was growing slowly in an abnormally aerated squamous bone, and it extended to the available space of the TMJ because of the congenitally hypoplastic condyle and ramus of the mandible. It eventually resulted in destruction of the posterior wall of the TMJ.

Infection of the EC led to ear discharge through communication with the external auditory canal and otalgia, which mimicked otitis externa. This type of presentation is not reported in the literature.

Intracranial pressure effect of the EC could lead to the development of neurological deficits.³ In our patient, the presence of the defect in the posterior wall of the TMJ into the external ear canal from which keratin could be suctioned was a favorable sign for early suspicion of the diagnosis before the occurrence of neurological symptoms.

Radiological features are those of a well-defined, low-density mass and no enhancement post contrast on a CT scan. Destruction of both the inner and outer tables is possible with irregular margins. Magnetic resonance imaging of epidermoids showed invariably hyperintense signal on T2WI and low signal on T1WI. Marked hyperintensity on diffusion-weighted imaging (DWI) is characteristic.¹ The radiological

findings were typical of EC in our patient. Histologically, ECs are lined by stratified squamous keratinized epithelium covering keratinous material and cholesterol crystals.⁴ Despite the usual benign nature of ECs, possible malignant transformation has been reported, and surgical excision is warranted to avoid complications.⁴ Total removal is the aim of surgery, which we conducted in our patient. The follow up CT temporal bone one year after surgery was clear.

In conclusion, we report a case of an intradiploic EC of the squamous part of the temporal bone that presented as otitis externa. While atypical presentations of these cysts have been reported, it is important to note that patient history can be misleading, and that revisiting a patient's clinical data and radiological studies is paramount to making an accurate diagnosis.

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