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GIANT EPIDERMOID CYST OF TEMPORAL BONE: A CASE REPORT AND REVIEW OF THE LITERATURE

ABSTRACT

Epidermoid cyst is a thin walled cyst lined by stratified squamous epithelium containing pearly white, soft cheesy material composed of fatty acid and keratin. Intracranial epidermoid cyst is usually located in the cerebellopontine angle, parasellar regions however, temporal bone epidermoid cyst is very rare. We present a case of giant epidermoid cyst in a 42-year-old male of the right periauricular region, temporal region and infratemporal fossa with lytic destruction of right middle ear and inner ear structures, mastoid and squamous part of temporal bone with intracranial but extradural extension which was completely excised. Histological examination confirmed the diagnosis of epidermoid cyst.

Keywords: Epidermoid cyst, mastoid, temporal bone

INTRODUCTION

Intracranial epidermoid cyst (IEC) is an uncommon benign lesion of the central nervous system (CNS) with the prevalence of 0.2–1.8% of all intracranial tumors.[1]Epidermoid cyst develops from epidermal and connective tissue forming a sac lined by stratified squamous epithelium containing epithelial debris. Epidermoid cyst is common over the skin however it may occur in the scalp, base of the brain, cerebellopontine (CP) junction . IEC contains white cheesy material and is well-defined, homogeneous, lobulated mass, fibrous capsule with cholesterol crystals, keratin debris. IEC is expansile with tendency for progressive growth compressing surrounding middle ear, eroding the bony structures a result of pressure necrosis and enzymatic lysis.[2]It is asymptomatic but in rare cases, it grows intracranially and produces features of brain compression.

Case: A 42-year-old male presented with history of gradually progressive painless right periauricular swelling with loss of hearing in right ear for 5 years. There was a history of middle ear surgery in the same side attempted with suspected extensive cholesteatoma 3 years back at another centre with the surgery abandoned due to unsuccessful attempt for disease removal. The examination revealed a 8x6 cm painless, firm, rubbery swelling located postero-superiorly to right external auditory canal (EAC), over mastoid process and anteriorly up to lateral canthus of right eye

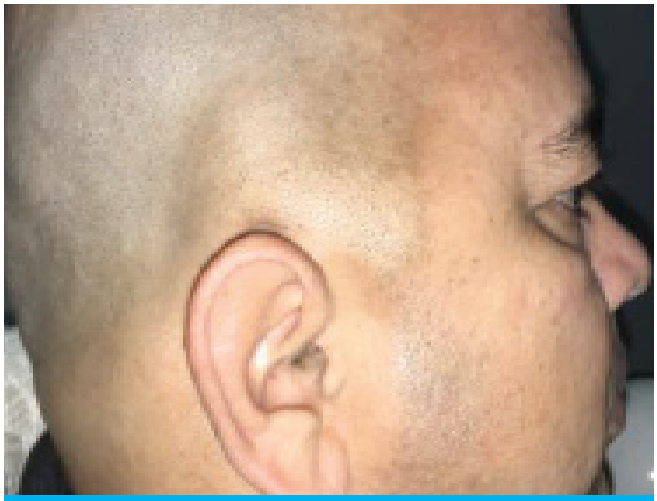

Fig. I: Swelling in right temporal region (Preop)

(Fig. I). The right auricle was normal in size and shape but displaced laterally. On Otoscopic examination, there was totally occluded EAC at cartilaginous part with soft fluctuant swelling arising from postero-superior part of the canal. On the left side auricle, EAC and tympanic membrane were normal. Rinne test showed no response on right ear and positive in left side. Weber test was lateralized to the ear. Facial nerve and other neurologic examination results were normal. On Pure Tone Audiometric evaluation, there was profound hearing loss in right side and the hearing level was normal in the left.

In preoperative HRCT of temporal bone, the lesion was located in temporal region involving

middle ear cleft, the labyrinth and destructing the cortical bone of mastoid and the superior bony portion of EAC (Figure II) The MR

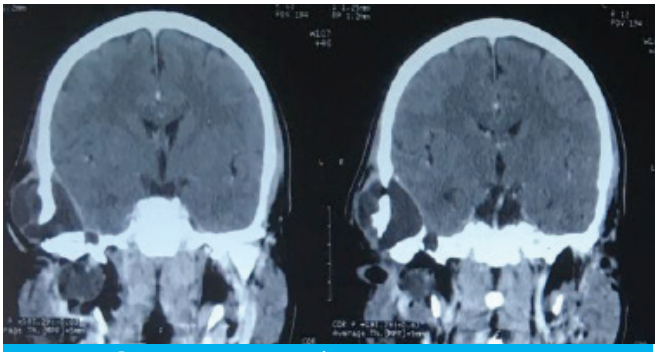

Fig. II: CT scan coronal image

imaging revealed a heterogeneous cystic mass filling the EAC that was destroying the surrounding mastoid air cells extending into middle and posterior cranial fossa with high signal intensity in the peripheral rim and irregular and low signal intensity in the center (Figure III). Mastoid

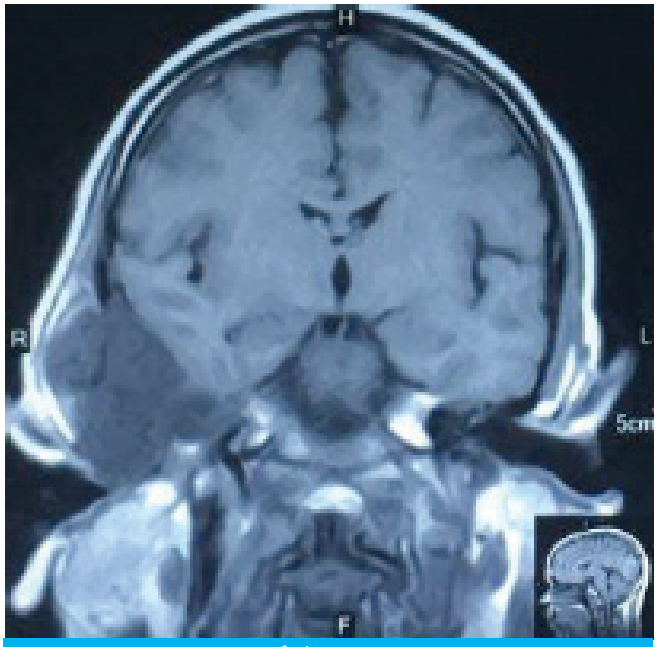

Fig. III: MR image of the mass

exploration was done via post-aural approach which revealed huge light pearly mass beneath the mastoid periosteum along with defect of cortical bone near mastoid tip and squamous part of temporal bone. The dural plate was dehiscent superiorly with the cystic mass adhering to the dura. All structures of bony EAC middle ear and inner ear were eroded however, facial nerve was intact and entrapped within the cyst. The cyst contained approximately 40 cc of cheesy white material (FigureIV). The entire content of the cyst was suctioned out and cyst wall was completely

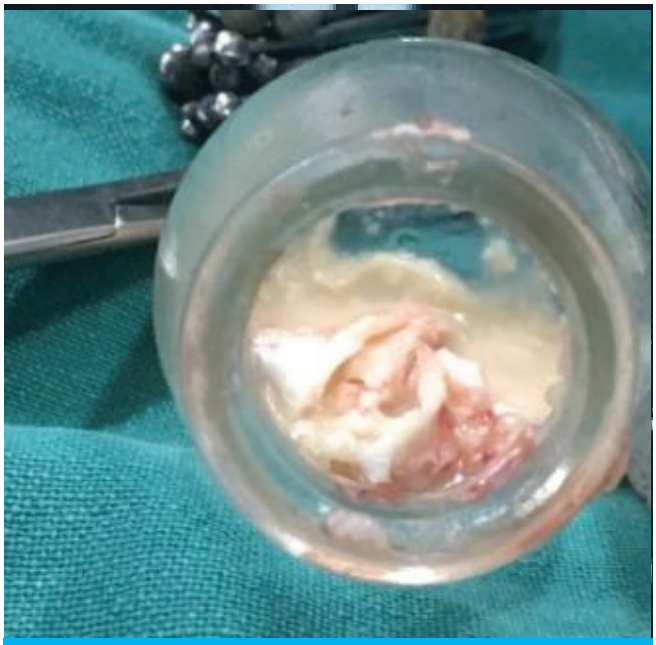

Fig. IV: Cheesy Material of the cyst

dissected. The facial nerve was preserved. There was an intra-operative CSF leak from the area of VIth nerve origin which was identified and controlled with fascia, tissue glue and fat in multiple layers. The cavity was obliterated with thigh fat and Cul-de-sac closure was performed. He had post-operative grade III Facial palsy, however, rest of the post-operative period was uneventful. The post-operative histopathology report revealed keratinizing squamous epithelium with keratinous debris arranged in laminated layers with many anucleate squame consistent with the epidermoid cyst.

DISCUSSION

Epidermoid cyst is a benign tumor that originates from an ectopic ectodermal tissue. It is lined by stratified squamous epithelium and filled with keratineous debris.[3] A pearly appearance is characteristic of the epidermoid cyst. It occurs more frequently in male between the ages of 40 and 60.[4] It is located most commonly over the skin but may occur within the diploe of the skull at the cerebellopontine junction (CP) and parasellar regions. It appear as well-defined, homogeneous, lobulated masses with a fibrous capsule filled with cholesterol crystals, keratin, protein, debris and cerebrospinal fluid (CSF) .[4] The cause for the development can be congenital tumours which result from inclusion of foetal squamous epithelial remnants or acquired tumours developing from ectopic skin elements which are implanted by

trauma, puncture or iatrogenic.[5] Epidermoid cyst at temporal bone was also reported as iatrogenic as a complication of ear surgery in the past.

Intradiploic epidermoid cysts commonly present with a painless lump under the scalp. Arana et al.[6]studied 37 intradiploic epidermoid cyst of temporal bone and found 10.8% asymptomatic.[6] In temporal bone, it may damage labyrinth leading to hearing and balance problems. Although the majority of epidermoid cysts are benign, malignant degeneration has been reported in few. Malignant changes of intradiploic epidermoid cyst should be suspected when there is contrast enhancement in MR imaging or if the tumor is fast-growing.[7]The enormity of the present lesion along with its bony erosions raised the strong clinical suspicion of malignancy.

The diagnosis solely by imaging is difficult. Conventional CT and MRI usually exhibit poor contrast from surrounding CSF making identification and exact extent very difficult.[8-10] They are mostly isodense or hypodense with density of lesion ranging from + 20 to 50 HU.[11] Cholesteatoma usually appears to be a well-defined, homogeneous, and hypodense mass in noncontrast CT scans. The margins of the bone adjacent to the bony labyrinth may be sclerotic and scalloped. No definite relation between signal intensity has been found in MRI. The typical MR image appearance of epidermoid tumors is hypo to slight hyperintense on T1W images, iso hyperintense on T2W images and hyperintense on DWI.However, rarely in T1W images, the epidermoids are hyperintense and in T2W images are of low signal intensity.[9-13]

Both congenital cholesteatoma and epidermoid cyst present as diagnostic dilemma as both show keratinized squamous epithelium including keratin. It is hard to differentially diagnose with MRI because both have a low signal in T1-weighted images and a high signal in T2-weighted images.[12-13] The congenital dermoid cyst containing elements of dermal cell layers can be distinguished from epidermoid cysts composed of epidermoid cell debris rich in cholesterol.

In this case report, it was very difficult to identify the site of origin. It might be tertiary cholesteatoma due to past surgery done at the age of 7 years. It was growing very slowly and might be late presentation of congenital cholesteatoma. However, the facial nerve was intact. Both congenital cholesteatoma and epidermoid cyst have a low signal on T1- and a high signal on T2-weighted images.[12-13] Next possibility is late presentation of epidermoid originated from mastoid bone or cysts originating in the CP angle.

It is not radiosensitive and has a tendency to recur if only partly removed. So, it should be treated by radical surgical resection. It has high chance of recurrence.[2]

CONCLUSIONS

Epidermoid cyst of temporal bone is a rare entity which is diagnosed by clinical presentation, physical examination, radiological, histological and intraoperative findings. The diagnosis should be considered in a case of extensive cholesteatoma with periauricular swellings or complications.

CONFLICTS OF INTEREST

"The authors declare that there is no conflict of interest regarding the publication of this paper."

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