

## Case Report

# Study of intradiploic epidermoid cyst in the right occipital bone

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**Abstract:** Intracranial epidermoid cyst (IEC) is a rare tumor which is often seen in the cerebellopontine angle, saddle area, brain parenchyma, and ventricles, but the diploe of cranial bones is extremely seldom affected. We report an unusual case of intracranial epidermoid cyst that developed from the right occipital. The IEC presented as a round cystic and solid lesion protruding through an occipital bony defect. The lesion was completely excised, and histologic result showed stratified squamous epithelium on the cyst wall and cholesterol crystal in the cyst.

**Keywords:** Cholesteatoma, epidermoid cyst, diploe, treatment

### Introduction

Intracranial cholesteatoma is an uncommon condition that is usually regarded as one of the important diseases involved in neurosurgery. Though considered rare, incidence of intracranial cholesteatoma is on the rise and now it accounts for 0.2 to 1.8% of all tumor lesions. Intracranial cholesteatoma is often seen in the cerebellopontine angle, saddle area, brain parenchyma, and ventricles [1]. Review of current literatures showed that the diploe of cranial bone is seldom affected. Moreover, some reports showed that frontal and parietal bones were the most common locations while occipital was rarer for the cholesteatoma [2, 3]. The following description will report this rare case of diploic cholesteatoma in the occipital bone and illustrate the difficulties in its diagnosis and treatment.

### Clinical report

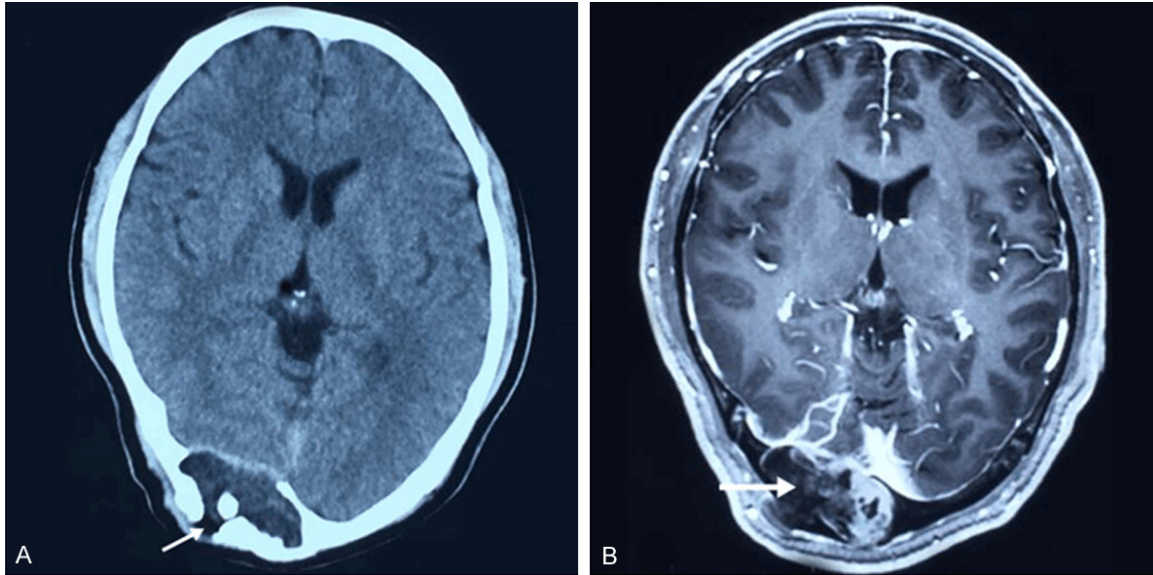
A 46-year-old male patient had complaints of progressive severe headache for one month and an occipital mass under the scalp for 30 years. There was no other positive signs except for an obvious occipital mass which was round, nontender, and without good activity, measuring 3 cm × 5 cm. Computed tomography (CT)

examination showed that there was an expansion of the occipital bony lesion (**Figure 1A**), which was irregular and destructive with well-defined boundary. Magnetic resonance imaging (MRI) disclosed a well-defined, spindle-shaped, mixed T1 and T2 signals on the right-side of occipital, which took on slightly heterogeneous image when enhanced with gadolinium. The right occipital lobe shifted as a result of suffering compression (**Figure 1B**). The patient underwent surgical operation. We observed that the mass originated from the diploe and the skull were destroyed because of tumor erosion, however, local dura mater was unaffected, indicating that the mass arose from the diploe. Then the patient underwent cranioplasty consisted of repairing the bone window with the titanium plate. Pathologic analysis demonstrated that the result was cholesteatoma (**Figure 2**). After 8 days the patient was discharged as cured with no surgery complication discovered. The patient progressed favorably, without recurrence of the tumor or neurological deficits over a 6-month follow-up.

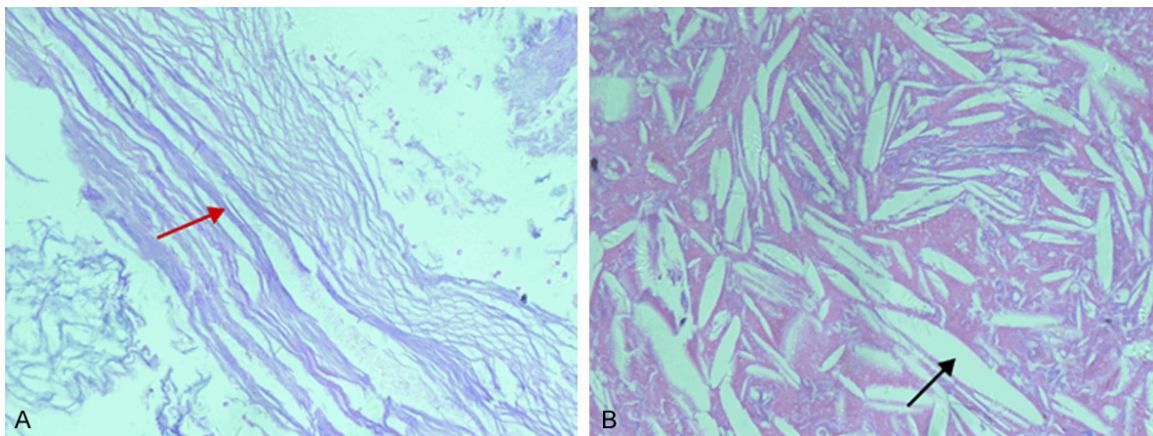
### Discussion

IEC, which is also known as cholesteatoma or pearl tumor, is a rare benign lesion that accounts for less than 1% of all primary intracra-

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**Figure 1.** Images of giant diploic epidermoid cyst of the right occipital bone in the present case. Preoperative CT scans show giant occipital extracerebellar hypodense lesion with extensive bone destruction (arrow) and mass effect in the posterior cranial fossa (A). Preoperative magnetic resonance images (MRI) show giant right occipital extradural diploic tumor (arrow) which is mixed hypointense and hyperintense. It has an enhancement rim of the dura mater and slight compression of the cerebellum and the right occipital lobe, without corresponding encephal-edema (B).



**Figure 2.** Photomicrograph showing massive keratinized matters (red arrow) in the cyst (A), and cholesterol crystals (black arrow) were easy to be seen in the partial cyst (B) (H&E stain,  $\times 200$ ).

nial neoplasms [2]. It can be divided into intracranial cholesteatoma, external auditory canal cholesteatoma and cholesteatoma otitis media depending on its location of distribution. Etiology of epidermoid cyst is still controversial, however, a result of the neural tube closure with the ectoderm composition during the embryonic period about 3 to 5 weeks of gestation may be the most acceptable reason [4]. Michaels L suggested that it may originate from residual cells divided from the ectoderm of the

first branchial groove in embryonic and early in fetal life when neural tube closes [5].

IEC was especially common in the sites of frontal and temporal bone. However, little is known with regard to diploe due to its rarity, which can occur in all age groups with a peak incidence in the third to fourth decades. In general, the round tumor is characterized by slow-growth and being asymptomatic with little periosteum or dura mater structures involved, which be-

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come distinct by the prominence of the soft tissue, therefore, presenting merely less frequently headache [6]. The prevalence is similar in terms of sex, and the patient often sees a doctor in an unintentional physical examination or when a constant cephalgia emerges as a result of the oppression of enlarged tumor.

Clinical history, physical examination, and relative imagings are important diagnostic tools in analyzing the unpopular tumor [3]. The typical x-ray images are the skull lesion and clear hardening band, which arise from caducous epithelial cells of epidermoid cyst, and therefore oppress adjacent bones for swelling growth. MRI is a more accurate modality in diagnosing cholesteatoma, which shows fusiform heterogeneous T1-weighted and T2-weighted signals that reveal inhomogeneous enhancement when strengthened. However, we should highlight differential diagnosis consisted of meningioma, glioma, giant cell tumors, cavernous hemangioma and eosinophilic granuloma due to the deficiency of radiological texts. The defined diagnosis relies on postoperation histopathological examination [7].

The treatment strategy for this tumor is surgical excision, since expansive mass will evolve into a penetrative tumor inside and outside the skull, which could lead to potential complications such as infection, cerebral hemorrhage, abscess formation or malignant transformation [8]. In theory, the total swelling along with the whole cyst wall should be removed for the fear of recurrence [9]. Besides, we must maintain keen vigilance in operation when the endocranium or widespread skull are involved in order to avoid the leakage of cerebrospinal or other complications [10].

### Conclusions

Intradiploic epidermoid cyst is a rare and benign lesion. Compared with other bony lesions, there are no typical representations in symptoms, signs, and images. The defined diagnosis relies on postoperation histopathological examination. Because it is benign, we should try our best to remove en bloc tumor, and then, reconstruct the defect of the skull.

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### Disclosure of conflict of interest

None.

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