Letter to the Editor

A case of intradiploic epidermoid cyst

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Introduction

Epidermoid cysts (EC) are benign neoplastic lesions that can be classified as congenital or acquired (1). Congenital conditions are most common and are caused by the retention of the neuroectodermal epithelium when the neural tube is closed at 3 to 5 weeks of the embryo. Acquired natures are often related to factors such as trauma or surgery (2). EC can occur at any age and is most common between the ages of 20 and 60; the incidence is not gender-specific (2-4). Intradiploic EC account for approximately 0.2–1.8% of primary intracranial tumors, of which approximately 90% are located under the dura mater (cerebellopontine angle area is most common) and 10% are located in the skull plate barrier (5). Intradiploic EC are relatively rare, with slow growth, a long course of disease, and no specific signs and symptoms, and most lesions are painless masses (5). When the mass is large, it can cause infection and secondary bleeding, and malignant transformation can lead to headache and epilepsy (6). Because intradiploic EC is rare, it is prone to misdiagnosis. Here, we report a misdiagnosed case of EC, which is expected to help radiologists and clinicians diagnose EC early and accurately.

Case presentation

All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient's legal guardian for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

Medical history

The patient, female, 11 years old, found frontal mass without obvious inducement 2 years ago. To seek further diagnosis and treatment, the patient's family came to author's hospital. The patient has no relevant family or genetic history.

Imaging assessment

Plain scanning computed tomography (CT) showed invasive and expansive bone destruction of the midline area of the frontal skull, thinning of bone, and hardening of the edges. A nodular with mixed density was observed in the tumor, the cross-section size was approximately 2.0 cm × 1.8 cm, the boundary was clearer, the edge was more regular (*Figure 1*).

Magnetic resonance imaging (MRI) showed uneven low signal in T1 weighted imaging (T1WI) and high signal in T2 weighted imaging (T2WI), fluid-attenuated inversion recovery (FLAIR) and diffusion-weighted imaging (DWI), and the enhancement scans showed slightly enhancement (*Figure 2*).

The patient completed relevant auxiliary examinations and performed resection of skull lesions.

Intraoperative findings

The lesions of the midline area of the frontal skull protruded outward, approximately 2.5 cm × 2.5 cm in size, the corresponding periosteum thickened significantly, and the dura mater adhesion was obvious. The tumor has a poor blood supply, soft texture and clear boundary with surrounding tissue. The pathology report confirmed that

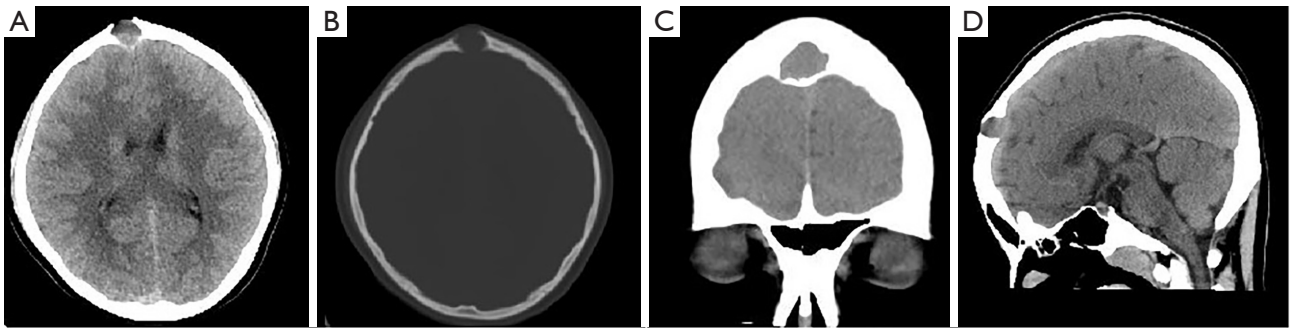

Figure 1 Preoperative cranial CT examination. Axial (A), sagittal (C), and coronal (D) head CT showed a 2.0 cm × 1.8 cm nodular mixed density shadow. The bone window (B) showed erosive and expansive bone destruction. CT, computed tomography.

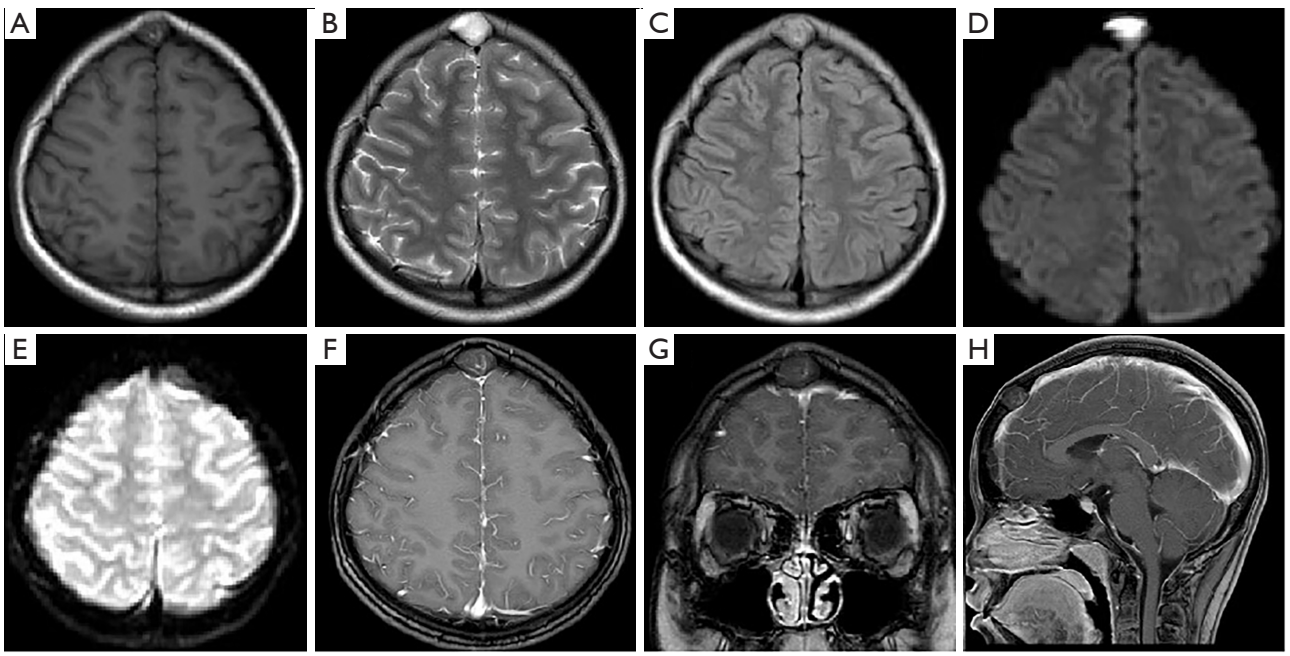

Figure 2 Brain MRI demonstrated a 2.0 cm × 1.6 cm × 1.4 cm mass in the midline area of the frontal skull. The tumor showed overall low signal with patchy high signal shadow in T1WI (A) and mixed high signal shadow in FLAIR (C). T2WI (B) and DWI (D) showed high signal shadow. The ADC (E) showed low signal. Axial (F), sagittal (G), and coronal (H) enhanced T1WI showed no significant enhancement, connecting with the adjacent meninges by a wide base and compressing the adjacent brain parenchyma. MRI, magnetic resonance imaging; T1WI, T1 weighted imaging; FLAIR, fluid-attenuated inversion recovery; T2WI, T2 weighted imaging; DWI, diffusion-weighted imaging; ADC, apparent diffusion coefficient.

the tumor was an intradiploic EC, histologically, an EC has squamous epithelium (hematoxylin & eosin, ×100) (*Figure 3*). The section surface of the mass was gray-white and soft in solid nature, and the surrounding bone tissue was involved. Postoperative CT showed complete resection of the lesion (*Figure 4*).

Discussion

EC are congenital benign neoplastic lesions that tend to occur in the posterior cranial fossa (7). Moreover, such neoplasms are also known as pearl tumors or cholesteatomas because of their lustrous white appearance under the naked eye. EC are well-circumscribed neoplastic

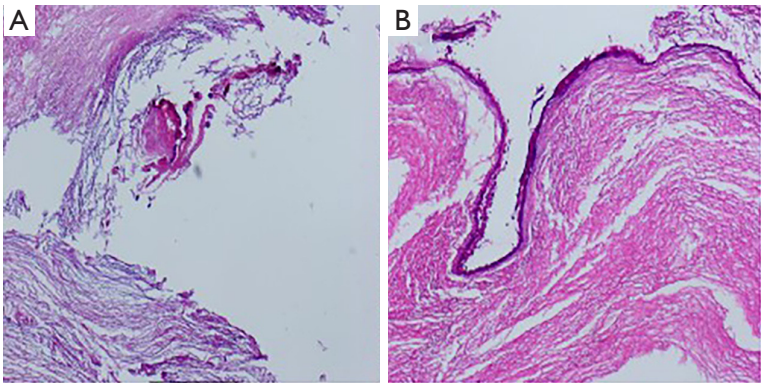

Figure 3 Pathological biopsy. Photomicrograph of pathology (A,B) shows the squamous epithelium image in the mass (hematoxylin & eosin, ×100), the pathology report confirmed that the tumor was an epidermoid cyst of the skull.

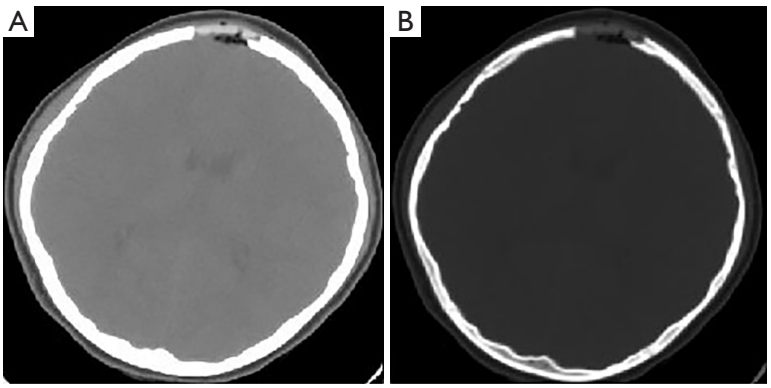

Figure 4 Postoperative re-examination of cranial CT: axial (A) and bone window (B) showed complete removal of the lesion. CT, computed tomography.

lesions, most located in the cerebellopontine horn or sellar area, and a few are located in the parenchymal brain, ventricles, and skull (8,9). Most EC are mainly composed of keratin and cholesterol, do not contain blood vessels, and lack mass effects, so most cysts have no special clinical manifestations. However, with the enlargement of the tumor, different clinical symptoms will appear due to different growth locations: cerebellopontine angle EC often begin with trigeminal or accessory neuralgia resulting in musculoskeletal shoulder disease; sellar EC are often accompanied by vision loss or visual field defects; EC in the brain parenchyma often have seizures or ataxia; ventricle EC can cause intracranial hypertension symptoms; and intradiploic EC are generally not tender (9-11). In this case, the patient found frontal mass without obvious clinical symptoms, and underwent surgery to remove it. EC grow slowly, and patients often go to the hospital when the tumor grows large or clinical symptoms appear, which is difficult to control with drugs, so the clinic is mainly surgery. It is important to note that even asymptomatic or small EC should be surgically removed because invasion of the brain parenchyma can lead to intracerebral hemorrhage. After complete resection of surgery, the long-term prognosis effect is good (3).

Intradiploic EC have typical imaging findings (12). Digital radiography (DR) shows round or oval bone destruction, well-bordered with clear boundaries, while CT shows low-density shadows or mixed density dominated by low density located in the skull plate barrier. The contrast-enhanced scan is generally not significantly enhanced, and calcifications can be seen in some lesions. The CT density depends on the triglycerides, cholesterol and protein content inside the lesions. The inner and outer plates of the skull are thinned or interrupted, showing osteolytic or

expansive bone destruction, and if the outer plate is broken, the lesion may be a "crater"-like bone defect. When the lesion is large, intracranial brain parenchyma compression may occur. MRI is mostly manifested as long T1 and long T2 dominated by uneven mixed signals, FLAIR sequences are often uneven high signals, DWI high signals and ADC low signals, and the enhanced scanning is not significantly enhanced. When the soluble lipid content in the cyst was high or the cyst was complicated by hemorrhage, T1WI and T2WI scans showed high intensity; indeed, when the soluble lipid content is small, cholesterol crystals, calcification and old bleeding are more common, and T1WI and T2WI can show low signal.

Differential diagnosis and typical features

Skull EC should also be distinguished from eosinophilic granuloma (EG), aneurysmal bone cyst (ABC), giant cell tumor of bone (GCBT), fibrous dysplasia of bone (FDB), and other tumors on radiography. EG typically show pathological changes as granulation tissue replacing normal bone tissue, skull destruction with or without soft tissue mass, residual small bone pieces in the lesion, typically "button" type dead bone, and contrast-enhanced scanning shows significant strengthening of soft-tissue masses (13). ABC is a benign neoplastic lesion of unknown etiology that tends to occur in long tubular bones and the spine (13). CT often manifests eccentric dilated osteolytic destruction, multiple cystic low-density areas, and bony separation in the lesion, while MRI showed cystic multilocular lesions, in which an obvious liquid-fluid level was visible, and there was a low signal ring at the edge of the lesion on T1WI and T2WI scans, and the spacing of the cyst wall and lesion was strengthened after enhanced scanning (14). GBT of bone needs to be distinguished from polycystic swelling of EC. Firstly, GBT originating in the skull are rare and often have "soap bubble" changes. Moreover, GBT is a blood-rich tumors, and significant enhancement is often seen on contrasting examination (15). Lastly, FDB, also known as dysplastic bone fibers, is a neoplasia in which bone marrow and reticular bone are replaced by fibrous connective tissue and irregular bone. CT and MRI often show enlargement of the affected bone, increased density or uneven decrease in signal of the bone marrow cavity, or ground-glass changes, the lesion extends in the direction of bone growth. In FDB, there is no clear demarcation from normal bone tissue, there is no periosteal reaction and soft tissue mass, and it is significantly strengthened (16).

Surgery is currently the most commonly used treatment for EC (17,18). Surgery focuses on removing the primary tumor capsule and cyst contents without damaging peripheral nerves and blood vessels. Surgical removal of the lesion can not only relieve the compression of brain tissue but also confirm the pathological diagnosis. EC are mostly benign lesions, and a few have been reported to have malignant changes, so it is necessary to remove the lesion as completely as possible during surgery. However, intracranial EC can sometimes infiltrate the brain parenchyma or adhere to neurovascular structures, resulting in incomplete resection (19). Previous studies have reported that recurrence rates in EC range from 1% to 54%, with reoperation required when the patient clinically reappears (17,19). In this case, the patient was not operated on for tumor recurrence but was treated symptomatically. After the operation, the patient's lesion was completely removed, and the patient recovered well.

Because benign lesions of the skull are relatively rare and some imaging findings are similar, they are easily confused or misdiagnosed during diagnosis. However, the imaging of these diseases has relatively unique characteristics, and radiologists should be familiar with the imaging manifestations and characteristics of these diseases for accurate diagnosis based on clinical history and laboratory tests. This case suggests that it is necessary to analyze and think comprehensively according to the patient's condition in clinical work, while strengthening the learning of relevant professional knowledge, and improving the correct rate of diagnosis and treatment.

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Footnote

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at https://qims.amegroups.com/article/view/10.21037/qims-22-1338/coif). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research

committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient's legal guardian for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

References

1. Hoang VT, Trinh CT, Nguyen CH, Chansomphou V, Chansomphou V, Tran TTT. Overview of epidermoid cyst. Eur J Radiol Open 2019;6:291-301.
2. Jamjoom DZ, Alamer A, Tampieri D. Correlation of radiological features of white epidermoid cysts with histopathological findings. Sci Rep 2022;12:2314.
3. Kuwano A, Arai K, Sakata S, Kawamata T. Intradiploic Epidermoid Cyst Causing Otitis Media with Effusion: Case Report and Review of the Literature. World Neurosurg 2020;144:71-3.
4. Nakajima K, Korekawa A, Nakano H, Sawamura D. Subcutaneous dermoid cysts on the eyebrow and neck. Pediatr Dermatol 2019;36:999-1001.
5. Lee P, Krisht KM, Mukunyadzi P, Krisht AF. Resection of an Isolated Pituitary Stalk Epidermoid Cyst Through a Pretemporal Approach: Case Report and Review of the Literature. World Neurosurg 2021;146:26-30.
6. Telmesani LM, Aldriweesh BA. Atypical presentation of an intradiploic epidermoid cyst. Saudi Med J 2019;40:624-7.
7. Kurihara Y, Kawamura K, Furue M. A Case of Epidermal Cyst with Underlying Lipoma on the Back: A Rare Presentation. Case Rep Dermatol 2021;13:171-5.
8. Ghartimagar D, Shrestha MK, Ghosh A. Recurrence of ruptured intracranial epidermoid cyst - A rare case report and presentation. Int J Surg Case Rep 2020;76:310-4.
9. Mohan A, Unni C, Kannoth P, Parambil RM. A Rare Case of Giant Intradiploeic Epidermal Cyst of the Frontal Bone with Intracranial Extension. Asian J Neurosurg 2020;15:670-3.
10. Brindisino F, Lorusso M, De Carlo L, Mourad F, Marruganti S, Passudetti V, Salomon M. Intracranial Epidermoid Cyst Mimics Musculoskeletal Shoulder Disease: Findings from a Case Report in Physiotherapy Clinical Practice. Int J Environ Res Public Health 2022;19:13590.
11. Yufeng Z, Xiaoqing J, Lulu X, Pei H, Shengwu L, Zhongsheng L. Giant epidermoid cyst penetrating the skull: a case report and literature review. Br J Neurosurg 2021. [Epub ahead of print]. doi: 10.1080/02688697.2021.1950635.
12. Spinato G, Gaudioso P, Falcioni M, Mosto MCD, Cocuzza S, Maniaci A, Fabbris C, Politi D, Volo T, Ferri E, Cristalli G, Mannelli L, Salvatore M, Rizzo PB, Frezza D. Giant Epidermoid Cyst of Posterior Fossa-Our Experience and Literature Review. Dose Response 2021;19:15593258211002061.
13. Davis TA, Mosquea TRJ, Belzarena AC. Multifocal eosinophilic granuloma with femoral epiphyseal lesion mimicking an aneurysmal bone cyst. Radiol Case Rep 2022;17:4176-82.
14. Li M, Gan Y, Shi D, Zhao J. Huge aneurysmal bone cyst secondary to giant cell tumor of the hand phalanx: a case report and related literature. BMC Cancer 2020;20:233.
15. Ravikanth R, Kamalasekar K. Imaging appearances of giant cell tumor of the bone. Med J Armed Forces India 2021;77:108-10.
16. Butt A, Patel K, Agrawal K, Arya A, Singh J. Fibrous dysplasia of the clivus - A case study and literature review. Radiol Case Rep 2020;16:230-6.
17. Choi MK, Chung KJ. Factors affecting complications after treatment of epidermal cyst. Yeungnam Univ J Med 2019;36:136-40.
18. Kim JN, Park HJ, Kim MS, Won SY, Song E, Kim M, Shin H. The reproducibility of shear wave and strain elastography in epidermal cysts. Ultrasonography 2022;41:698-705.
19. Patibandla MR, Yerramneni VK, Mudumba VS, Manisha N, Addagada GC. Brainstem epidermoid cyst: An update. Asian J Neurosurg 2016;11:194-200.

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