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Case Report

Parasellar epidermoid cyst with unique radiological features: A case report and review of the literature [☆]

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ABSTRACT

Intracranial epidermoid cysts (ECs) are encapsulated lesions lined by squamous cell epithelium and the most location is the cerebellopontine angle and appears with cerebrospinal fluid-like irregular mass. Occasionally, ECs present as high-density masses on computed tomography and atypical features in magnetic resonance images in the unusual area, which makes the diagnosis difficult. Here, we report a case of a female subject who complained of episodic left facial convulsions for more than 3 months. Computed tomography plain scan revealed a large hyperdense parasellar mass with atypical magnetic resonance findings. In this report, we analyzed retrospectively the radiological characteristics and histopathology of the parasellar EC, thus increasing awareness about this unusual image features.

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Introduction

Intracranial epidermoid cysts, commonly referred to as cholesteatomas, are rare benign intracranial lesions of the brain that make up 0.2%–1.8% of all intracranial neoplasms, even up to 7% of cerebellopontine angle tumors [1]. Although the vast of epidermoid cysts are intradural, some of them rarely are extradural. They are commonly located close

to the skull base, along cisternal spaces, and attached to adjacent neurovascular structures [2]. Stratified squamous epithelium lines the thin walls of intracranial epidermoid cysts, which are benign and have debris, keratin, cholesterol, and water as their typical contents [3]. Most of them show up as low-density lesions on computed tomography (CT) scans, but occasionally they may appear as high-density lesions, making diagnosis challenging. We report a case of high-density parasellar epidermoid cyst on CT, which

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was also examined by multimodal magnetic resonance imaging (MRI). By merging the cyst's pathology and imaging characteristics, the cyst's contents are discussed. The imaging features were evaluated in conjunction with earlier reports to help radiologists better understand the condition, raise the diagnostic bar, and prevent misdiagnosis.

Case report

A 39-year-old woman with periodic left facial convulsions for more than 3 months with a mild headache, twitching of the left masticatory muscle, difficulty opening her mouth during the attack, and no accompanying dizziness, facial numbness, nausea, or vomiting. She had undergone a partial thyroidectomy 2 years prior for "papillary thyroid cancer with lymph node metastases." The patient was admitted with a fever of 36.5°C, a pulse rate of 90 beats per minute, a respiration rate of 20 breaths per minute, and a blood pressure reading of 123/89 mmHg. She was conscious, in a voluntary position, walking normally, cooperating during the physical examination. The

heart, lungs, abdomen, or neurological system were all confirmed to be normal. Facial electromyography, lateral spread response, and somatosensory evoked potential are approximately normal.

In a precontrast CT scan, there is a circular, well-circumscribed mass next to the left parasellar region with hyperdensity and patchy hypodensity that is not surrounded by brain edema (Figs. 1A-D). The roughly well-defined lesion was performed by multimodal MRI, including conventional MRI plain scan, enhancement, diffusion-weighted imaging (DWI), susceptibility-weighted imaging (SWI), arterial spin labeling (ASL), magnetic resonance spectroscopy (MRS) and magnetic resonance angiography (MRA). In MRI, the signal intensity of the lesion showed heterogeneously high on T1-weighted sequence and heterogeneously low on T2-weighted sequence and Fluid attenuated inversion Recovery (FLAIR). Corresponding to the low-density on CT within the lesion showed hypointense on T1-weighted image (T1WI) and hyperintense on T2-weighted image (T2WI) and Fluid attenuated inversion Recovery (Figs. 2A-C). There was no enhancement on T1WI obtained after addition of contrast material (Fig. 2D). Arterial spin labeling images (Fig. 2E) showed the tumoral

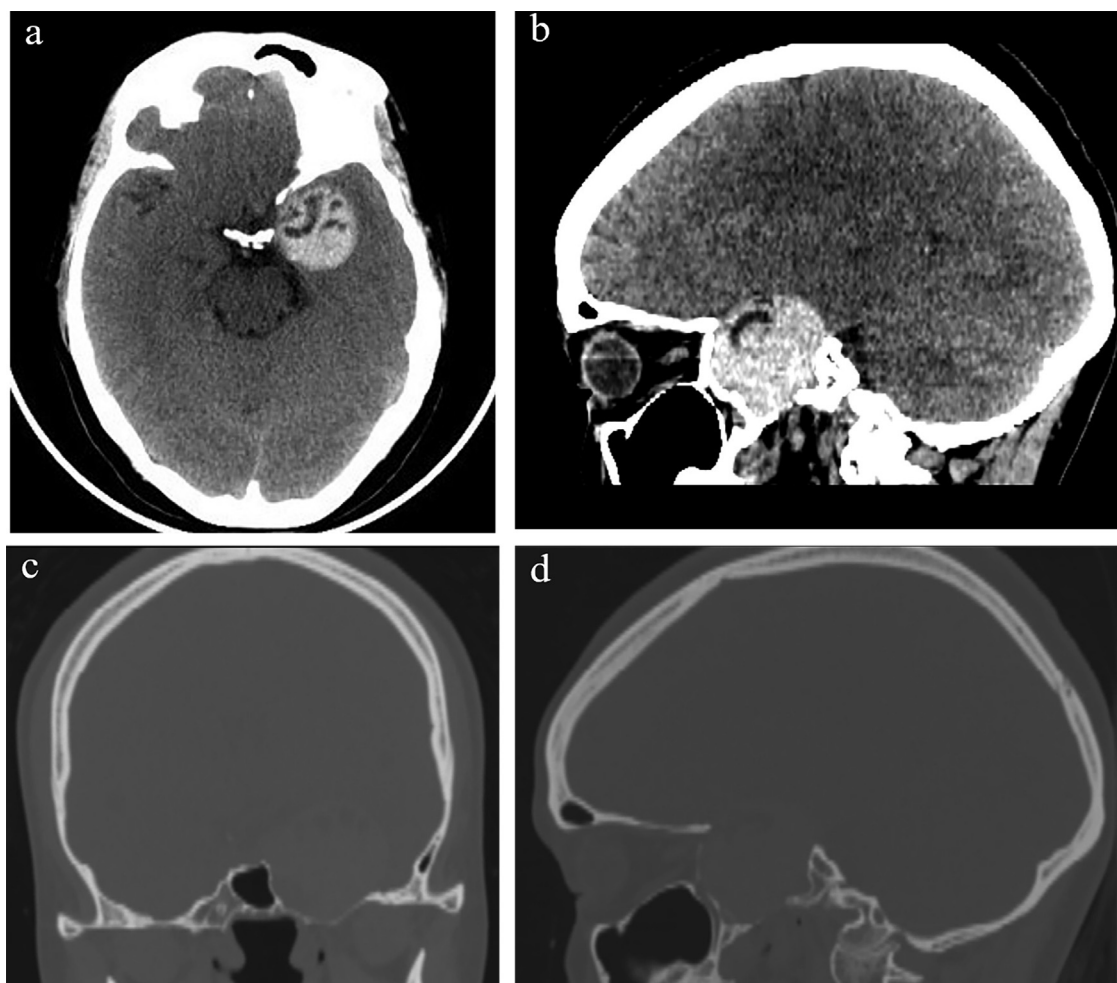


Fig. 1 – The CT scan showed the cysts as density, well-demarcated lesion with low density striated shadows in axial (A) and sagittal (B) view. Bone window showed that the mass causes compression of the surrounding bone but no signs of bone destruction in coronal (C) and sagittal (D) view.

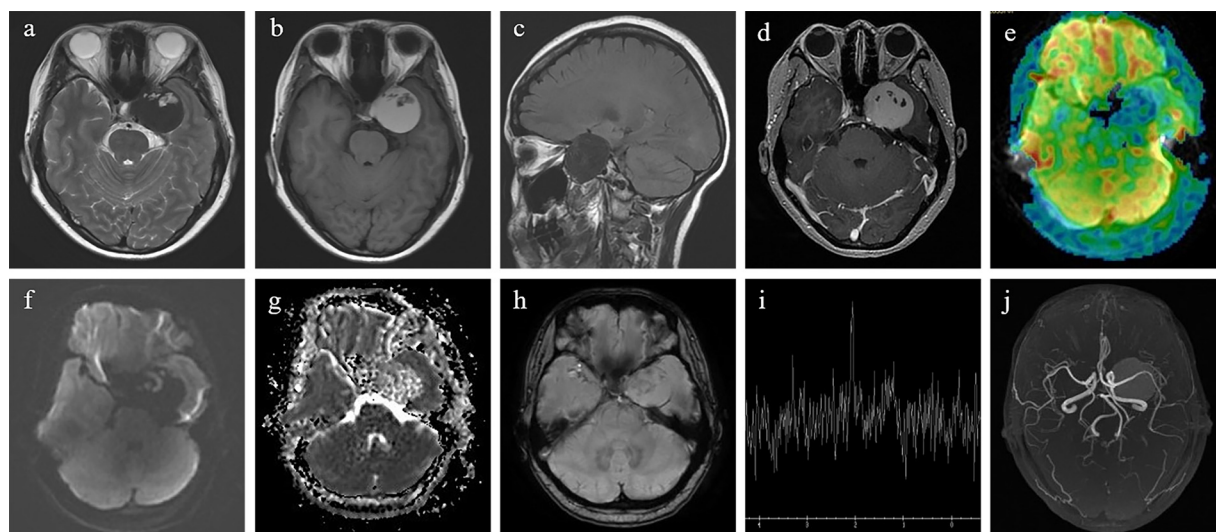


Fig. 2 – The MRI images revealing a lesion that was mostly hyperintense on a T1-weighted (A) and hypointense on a T2-weighted (B) and Fluid attenuated inversion Recovery (C) image. There was no enhancement (D). ASL showed the decreased blood perfusion in the lesion area (E). The majority of tumor showed that the DWI (F) was significantly lower than that of normal brain parenchyma, and demonstrate an elevated ADC (G). No signal reduction area was found in SWI (H). MRS (I) was failed. No obvious abnormality was found in MRA (J). ADC, apparent diffusion coefficient; DWI, diffusion-weighted imaging; SWI, susceptibility-weighted imaging.

blood perfusion was lower than that in the nontumor brain parenchyma, which means the tumor lacks blood supply. The majority of tumor showed that the DWI was significantly lower than that of normal brain parenchyma, and demonstrate an elevated apparent diffusion coefficient (ADC). The small part of tumor had ADC and DWI similar to that of normal brain tissue (Figs. 2F and G). No signal reduction area was found in SWI (Fig. 2H), which would rule out calcification and hemorrhage. Magnetic resonance spectroscopy was failed due to magnetic field in inhomogeneity of the parasellar region (Fig. 2I). The digital subtraction angiography (DSA) and magnetic resonance angiography (Fig. 2J) showed no tumor staining in intracranial mass and no obvious abnormalities in arteries.

The patient underwent “Left frontotemporal craniotomy with left parasellar tumor resection” under general anesthesia. Intraoperative findings showed that the tumor was located in the parasellar region with white capsule and new small blood vessels visible on the surface while its boundary with surrounding brain tissue also appeared clear. Microscopically, the covering of the lesion was the parasellar dura mater, suggesting that the lesion was epidural (Fig. 3A). When incision was made into the capsule, light yellow and brown “oil like” viscous content was seen (Fig. 3B). It was accompanied by yellow cholesterol crystal-like substance, which was lacked blood supply and adhered to the bottom of the cyst (Fig. 3C). Few crystal-like substances were sent to pathology. Photomicrographs of tissue samples obtained for pathological examination revealing the diagnosis of epidermoid cyst. The squamous epithelium (Fig. 4A) and stratified keratin debris (Fig. 4B) were classic pathological features showing of epidermoid cysts.

Discussion

Intracranial epidermoid cysts are always located close to the skull base, and the exact pathogenesis remains obscure. They are extra-axial rather than midline [3]. Previous reported cases of hyperdense intracranial epidermoid cysts (HIECs) mostly occurred in the posterior fossa of females [4]. But subsequent reports have shown that HIECs can also occur in rare places, such as the parasellar area, cerebellum, clival region, brainstem, cerebello-pontine angle and lateral ventricle, etc. [5–9].

Most intracranial epidermoid cysts typically appear as homogeneous, low-density lesions on CT scans and present with low signal intensity on T1-weighted sequences and high signal intensity on T2-weighted sequences on MRI scans [3]. The hypotheses that could explain their imaging features one is the solid cholesterol crystals, another is their content of CSF [10]. However, the reason for unusual CT and MR imaging characteristics of HIECs is not clear. Composition analysis of the cystic fluid concluded that too low concentrations of calcium and iron to explain the high CT density. The combination of high protein content and high viscosity may be responsible for the atypical imaging findings [6–8]. Some cases have reported that hemosiderin deposition in cysts caused by continuous or intermittent bleeding of the lesions, which is the cause of high T1 signal intensity [8,11]. Rare causes, such as the presence of melanin deposits, large numbers of polymorphonuclear leukocytes and infection, were also thought to be causes of atypical imaging findings [6,12]. The latest research suggested that HIECs containing high protein levels and viscosity resulted in low T2 signal, while blood products resulted in high T2 signal [13].

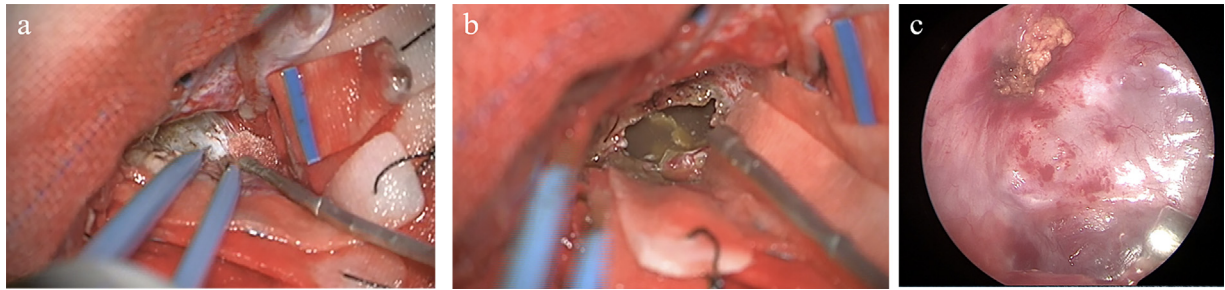


Fig. 3 – Microscopically, the covering of the lesion was the parasellar dura mater (A). Cholesterol crystals and the contents of a yellowish oil-like capsule were seen under fiberscope (B). Close observation under neuroendoscopy showed that a small amount of residual cholesterol crystals adhered to the bottom of the cyst (C).

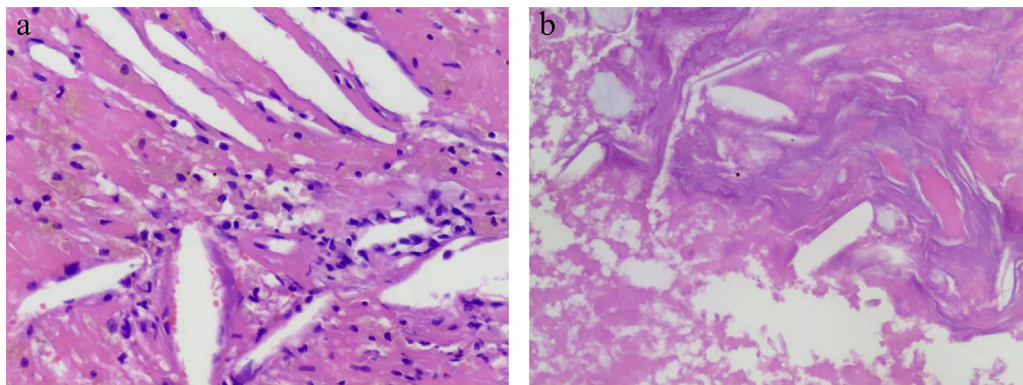


Fig. 4 – Pathological examination revealing cholesterol crystals (A) and stratified keratin debris (B). H & E, original magnification $\times 200$.

In our case, the main body of the tumor showed short T1 and T2 relaxation time. And calcification and bleeding were excluded by no signal reduction area in SWI. Thus, based on intraoperative findings and previous literature, we speculated that the high signal on T1WI is mainly related to the high protein content in the cyst, while the low signal on T2WI may be explained by the high viscosity of the cyst contents. According to the previous literature, intracranial epidermoid cysts demonstrate a markedly hyperintense signal compared to brain tissue and CSF on DWI because of the combination of T2 and diffusion effects, especially the T2 penetration effect rather than diffusion effects when ADC values were significantly higher than other brain area [13,14]. It can be inferred that the low signal on T2WI of the epidermoid cyst in this case resulted in the low signal on DWI.

HIECs located in the sellar region are normally misdiagnosed as other diseases, such as parasellar meningiomas, thrombosed giant aneurysms, cavernous angioma, craniopharyngioma, and teratoma. It is important to diagnose them properly by taking the consideration of their locations and imaging features before the operation.

The epidermoid cyst in this case was similar to the parasellar meningiomas on CT scan. However, meningiomas have significant contrast enhancement and hyperplasia or erosion of the neighboring bones [15].

Although the DSA results were normal in this case, the aneurysm cannot be ruled out in this case according to the location, morphology of the mass and CT density. DSA could not be used for identification because the occluded aneurysms cannot be seen by angiography. In aneurysms with thrombus formation, MRI is better than DSA in displaying the actual tumor size. The signal of thrombus varies greatly due to the time and form of thrombosis: fresh thrombus shows high signal, while old thrombus shows low signal, stratified thrombus shows high and low stratified or vortex signal [16].

The CT images of cavernous hemangiomas in sellar region showed circular, quasi-circular isodensity or mixed density lesion, which could be accompanied by calcification and had clear boundaries. The lesions showed heterogeneous enhancement at the start and then became homogeneous over time, and the vessels were thickened and tortuous. DSA is more useful for demonstrating the feeding arteries and blood supply of the masses [17].

Craniopharyngiomas are difficult to distinguish from epidermoid cysts in sellar region. However, the cyst walls of craniopharyngioma are thicker and uneven, and the cyst walls or solid parts can be significantly enhanced by enhanced scanning [18].

Teratoma is also one of the differential diagnoses, which is more commonly calcified in general.

Conclusion

HIECs are exceedingly rare lesions, especially with atypical MRI images, including reversal of the typical T1 and T2 signal, especially hypointense on T2, as well as a lack of restricted diffusion in the unusual area [19]. Although high signal on DWI is the main basis for distinguishing it from other tumors [14,20], it is not totally applicable. It still needs to be considered in the differential diagnosis for intracranial tumor as a novel entity. Multimodal MRI should be fully taken full advantage of in diagnosis to infer the composition of cyst contents.

Authors' contributions

All authors contributed to the study conception and design. The first draft of the manuscript was written by Sisi Wang and all authors commented on previous versions of the manuscript. All authors read and approved the final manuscript.

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Patient consent

Informed written consent was obtained from the patient for publication of the case report and all imaging studies.

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