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# Suprasellar epidermoid cyst in an adult female, presenting as amnesia and somnolence: A rare case report



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#### ABSTRACT

*INTRODUCTION:* Intracranial epidermoid cysts (ECs) are rare benign congenital lesions and account for approximately 0.3 to 1.8% of all intracranial brain tumors. They frequently occur at the cerebellopontine angles and parasellar regions, insinuating between brain structures. The author reports here a case of pathologically proven suprasellar epidermoid cyst in an adult female, presented with amnesia and somnolence to increase awareness about this unusual presentation.

*CASE REPORT:* A 58-year-old female was presented to our hospital complaining of amnesia for one year, followed by weakness and somnolence for 2 months. Radiological imaging showed the features of the suprasellar epidermoid cyst which resected through the transsphenoidal endoscopic approach.

DISCUSSION: Epidermoid cysts are slow-growing, benign lesions however, they may rarely undergo malignant transformation into a squamous cell carcinoma. The mean age at presentation of these lesions is 40 years. Suprasellar/sellar lesions usually present with non-specific headaches and visual disturbances. However, our case presented with amnesia and somnolence.

CONCLUSION: Surasellare epidermoid cysts (ECs) are rare benign congenital lesions. They often manifest with headache and visual field defects but, they may present with atypical symptoms as amnesia and somnolence. Endoscopic transnasal and trans-sphenoidal approaches can help to remove the lesion in most cases. Attention needs to the possible postoperative complications and longtime imaging follow-up because this lesion may recur after a few years.

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#### 1. Introduction

Intracranial epidermoid cysts (ECs) are rare benign congenital lesions and account for approximately 0.3 to 1.8% of all intracranial brain tumors. They may arise from displaced dorsal midline ectodermal cell rests between the third and fifth weeks of embryogenesis during neural tube closure. However, cerebellopontine angle (CPA) and parasellar location could be explained by the result of the proliferation of multipotential embryogenic cell rests or lateral displacement of ectodermal cells by the developing otic vesicles [1,2]. They frequently occur at the cerebellopontine angles and parasellar regions, insinuating between brain structures [3]. The slow growth of these tumors often results in them remaining asymptomatic until their size is large enough to compress surrounding structures [4]. Symptoms depend on their location and include hearing loss, dizziness, gait disturbance, trigeminal neuralgia, tinnitus, diplopia, visual impairment, apathy, headache, and gait ataxia [2]. The author reports here a case of pathologically

proven suprasellar epidermoid cyst in an adult female, presented with amnesia and somnolence to increase awareness about this unusual presentation.

## 2. Patient information

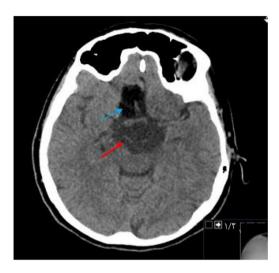
## 2.1. Patient presentation

A 58-year-old female was presented to our hospital complaining of amnesia for one year, followed by weakness and somnolence for 2 months. The patient was a known case of post-thyroidectomy hypothyroidism on medication and regular follow up for the last 10 years. She had a long history of headaches. She was an ordinary smoker but, she did not give any history of food or drug allergy and psychosocial problems. On physical exam, her Glasgow coma scale scores were 15. She had no facial asymmetry, gaze limitation, and no significant focal neurologic deficit. Her muscle's motor function was normal.

Abbreviations: ECs, epidermoid cysts; MRI, Magnetic Resonance Imaging.

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**Fig. 1.** Axial brain CT image shows a hypodense mass in the suprasellar region, extended to basal cisternae and frontal region, and surrounded the vascular structures, chiasm, and stalk.

#### 2.2. Laboratory and radiological findings

Her routine blood exam, thyroid hormones, and electrolyte levels were within normal limits. She was negative for HIV and her HbsAg and anti HCV tests were non-reactive. On brain CT images a hypodense mass measuring approximately  $5.3 \times 4$  cm in size was observed in the suprasellar region, extended to basal cisternae and frontal region, and surrounded the vascular structures and chiasm and stalk (Fig. 1). In contrast-enhanced brain MRI, the same findings were noted (Fig. 2a, b). The lesion was extended to the frontal region as seen on CT, and some fat compatible signals were detected in this region (Fig. 2c and d). There was no post-contrast enhancement in the lesion but in diffusion-weighted MRI images, the restricted diffusion was observed within the lesion (Fig. 3a and b). On Perfusion MRI sequences, the lesion was hypo-perfused in and all metabolite amounts were decreased on MR spectroscopy. The pathologic exam of the resected strictures showed lamellar keratin, consistent with the epidermoid cyst.

#### 2.3. Therapeutic intervention

The patient was taken up for a transsphenoidal endoscopic procedure. The sphenoid sinus was reached by the experienced ENT team with the trans-nasal transsphenoidal approach. Dura was incised and the suprasellar area was reached and the mass was excised as much as possible without damaging the surrounding tissues. Bleeding was controlled with the help of FloSeal and Surgicel. Fat tissue taken from the leg was placed to prevent fistula, and the lumbar drainage was maintained. No intraoperative complications were noted, and the patient was transferred to the neurosurgical intensive care unit (ICU) for further postoperative management. Her consciousness was recovered postoperatively and the lumbar drain was removed, but on the second day of post-operation, she became hypotensive that managed appropriately. The postoperative brain CT and MRI performed at this time showed resection of the suprasellar lesion and postoperative changes. (Fig. 4a, b, and c). By post-operative day nine, the patient developed rhinorrhea and lumbar drainage was placed again, and  $4 \times 30$  cc was started to be drained. On day 14 post-operation she developed a fever and in consultation with infectious disease specialists, her antibiotics were changed. After 23 days post-operation her rhinorrhea stopped and she was referred to the ENT for further evaluation of rhinorrhea but, CSF rhinorrhea was not considered. The lumbar drainage of the patient, who had not had rhinorrhea for a few days and was not considered by the ENT, was removed. By postoperative day 32 the patient, whose postoperative complications were treated was discharged with full recovery.

#### 3. Discussion

Epidermoid cysts popularly known as Tumeur perlees(pearly tumors) were first described by Cruveilhier in 1835. They are slow-growing, benign lesions however, they may rarely undergo malignant transformation into a squamous cell carcinoma. The mean age at presentation of these lesions is 40 years [5]. Typically, they are irregular, insinuating lesions that encase adjacent neurovascular structures. Suprasellar/sellar lesions usually present with non-specific headaches and visual disturbances. In other locations, headache (either non-specific or that of intracranial hypertension), seizures, and motor/sensory deficits have been noted. Hydrocephalus is rare and is usually a late manifestation [6]. Our patent was 58 years old and presented with amnesia and somnolence. However, she had a long history of a headache without any visual disturbance.

On CT, ECs are hypodense due to the low absorptive value of their fat content. On MRI epidermoid cysts appear hypo-, iso-, or hyperintense on T1-weighted MRI imaging, with or without rim enhancement following contrast administration. On T2-weighted MRI imaging, however, ECs typically appear hyper-intense. The most specific MR sequence appears to be diffusion-weighted imaging where ECs show a restricted pattern. This allows differentiating these lesions from other cystic pathologies, including arachnoid cysts, which show isointensity to CSF on all MR sequences without diffusion restriction [7].

Generally, suprasellar epidermoid cysts have been excised by transfrontal craniotomy, but in the last years, a less invasive transnasal endoscopic approach has been adopted. The first craniotomy for a suprasellar epidermoid cyst was reported by Sadeh et al. when a subtotal resection was performed [8]. Epidermoid cysts are rarely completely resected and the degree of resection obtained is limited by adherence to nearby structures. Firm adhesion of the cyst to the underlying pia matter or leptomeninges around cranial nerves are examples of situations in which gross versus subtotal resection would have to be weighed. The clinical decision to reach for total resection is surgeon and patient dependent; the risks and benefits of operating near key vascular structures must be included in the pre-operative discussion, with expectations should complications occur clearly outlined before surgery [4]. Despite better outcome has been reported by C.W. Huo et al. [7] in patients underwent endoscopic intervention, however, due to its rare occurrence and the overall low number of published studies, caution is required when drawing any definitive conclu-

Although epidermoids are benign, patients should be followed closely after the tumor has been removed surgically because these tumors are associated with different clinical symptoms and malignant transformation does occur in rare cases [9].

This work has been reported in line with the SCARE 2018 criteria [10].

#### 4. Conclusion

Suprasellar epidermoid cysts (ECs) are rare benign congenital lesions. They often manifest with headache and visual field defects but, they may present with atypical symptoms as amnesia and somnolence. Endoscopic transnasal and trans-sphenoidal approaches can help to remove the lesion in most cases. Attention needs to

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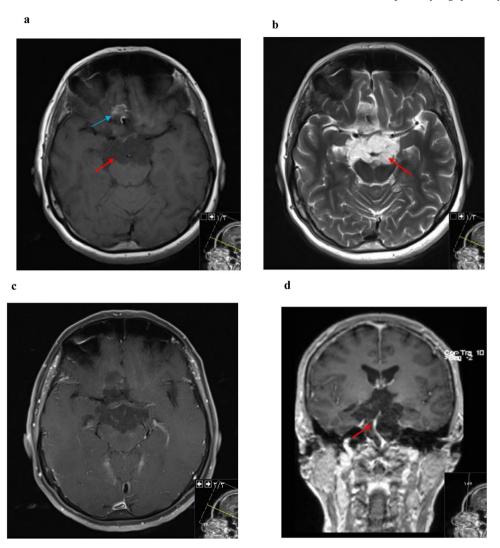


Fig. 2. (a) Axial T1 weighted brain MRI shows a hypo-intense mass in the suprasellar region (red arrow), extended to basal cisternae and frontal region (blue arrow) and surrounded the vascular structures, chiasm, and stalk. (b) Axial T2 weighted brain MRI shows a hyper-intense mass in the suprasellar region (red arrow), extended to basal cisternae and frontal region and surrounded the vascular structures, chiasm, and stalk. (c) Axial T1 weighted fat-suppressed, post-contrast brain MRI shows a hypo-intense mass in the suprasellar region, extended to basal cisternae and frontal region and surrounded the vascular structures, chiasm, and stalk. (d) Coronal T1-weighted fat-suppressed, post-contrast brain MRI shows a hypo-intense mass in the suprasellar region, extended to basal cisternae and displaced the vascular structures, chiasm and stalk.

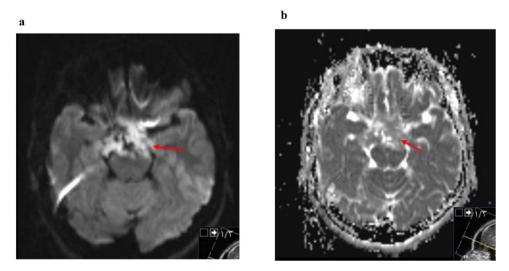
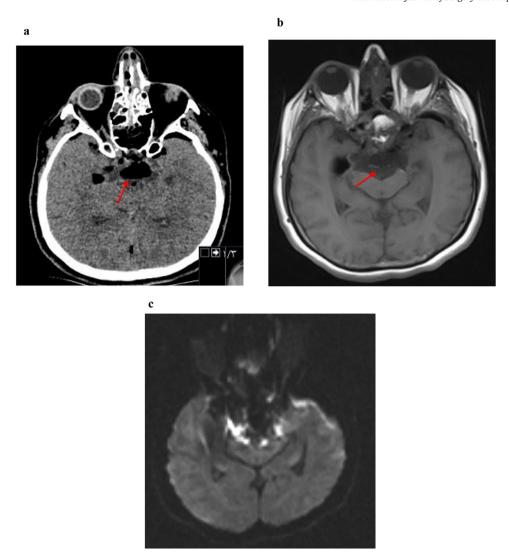


Fig. 3. (a) Axial DWI-MRI shows restricted diffusion in the suprasellar region. (b) Apparent diffusion coefficient maps show actual diffusion in the suprasellar region.

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**Fig. 4.** (a) Postoperative axial CT image shows post-operative changes and pneumocephalus in suprasellar region with no mass lesion. (b) Postoperative axial T1 weighted brain MRI image shows post-operative changes and pneumocephalus in the suprasellar region with no mass residual. (c) Postoperative axial Diffusion-weighted brain MRI image shows no restricted diffusion in the suprasellar region.

the possible postoperative complications and longtime imaging follow-up because this lesion may recur after a few years.

#### **Declaration of Competing Interest**

The author has no potential conflicts of interest to disclose.

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## **Ethical approval**

The manuscript has got an ethical review exemption from the Ethical Review Committee (ERC) of our institution, as case reports are exempted from review according to the institutional ethical review committee's policy.

### Consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the editor of this journal.

#### **Author contribution**

Not applicable.

## Registration of research studies

Not applicable.

#### Guarantor

The corresponding author (Dr. Habib Ahmad Esmat) is the Guarantor for the work and he has the responsibility of access to the data, and controlling the decision to publish.

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