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Epidermoid cyst of the sphenoid sinus with extension into the sella turcica presenting as pituitary apoplexy: case report

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Abstract

Background: Epidermoids of the central nervous system are rare tumors. They are usually found in the fourth decade of life and most commonly off midline in the cerebellopontine angle. We report here a rare case of an epidermoid arising from the sphenoid sinus with extension into the sella and adjacent structures with acute onset of neurological deficit. The significance of the clinical presentation resembling pituitary apoplexy and magnetic resonance imaging (MRI) findings is noted.
Case Description: A 25-year-old man presented with acute severe headache, diplopia, and decreased visual acuity. Examination revealed right-sided ptosis and paresis of the third cranial nerve on the right side. Computed tomography and MRI were suggestive of a slow-growing sphenoid sinus mass with extension into the sella. The sublabial transsphenoidal approach was used to remove the mass under direct visualization. The patient's neurological status improved to baseline both subjectively and objectively after the operation.
Conclusion: Epidermoids, although rare, should be considered as part of the differential diagnosis when evaluating lesions of sphenoid sinus or sellar origin.

Keywords: Epidermoid; Sphenoid sinus; Sella turcica; Skull base

1. Background

Epidermoids typically occur between the ages of 20 and 60 with a peak incidence in the fourth decade. There is no gender predilection [9,10]. They comprise 1% of all intracranial tumors [23] and are usually found off midline in the extraaxial, subdural regions, most commonly the cerebellopontine angle cistern (40%-50%), followed by the parasellar/middle fossa space (10%-15%) [1,3,10]. About 10% are extradural, most commonly the cranial diploic spaces [21] of the frontal, parietal, and occipital bones, with the sphenoid bone less commonly effected [2,4,17,19]. Epidermoids arising from the ethmoid [5] and maxillary sinuses [22] and one case of a sphenoid sinus cyst [14] have been previously reported.

We report here a rare case of an epidermoid tumor originating within the sphenoid sinus with extension into the sella turcica that presented with acute sudden onset of headache and cranial nerve defects suggestive of pituitary apoplexy. The lesion was successfully resected by the transsphenoidal route. The unique magnetic resonance imaging (MRI) characteristics and pathological results will be presented.

2. Case description

A previously healthy 25-year-old man presented with acute severe headache and worsening diplopia. He denied any nausea, vomiting, weakness, numbness, or constitutional symptoms. Examination revealed an afebrile well-nourished young male with right-sided ptosis and paresis of the right third cranial nerve. Visual acuity was 20/60 OD

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and 20/40 OS by handheld acuity testing. No gross visual field defects were found. The rest of the neurological examination findings were normal. Complete blood count, serum electrolytes, and endocrinologic function studies were also normal.

On MRI, T1-weighted images showed a high signal mass with an isointense border in the sphenoid sinus with cephalad extension to the sella turcica. There was a mass effect on the optic chiasm and the cavernous sinus, right greater than left (Fig. 1A). The lesion extended anteriorly to the posterior margin of the ethmoid sinus and posteriorly into the prepontine cistern (Fig. 1B). There was no enhancement of the lesion with administration of gadolinium. The T2-weighted images revealed an isointense mass with a bright signal border as shown in Fig. 1B.

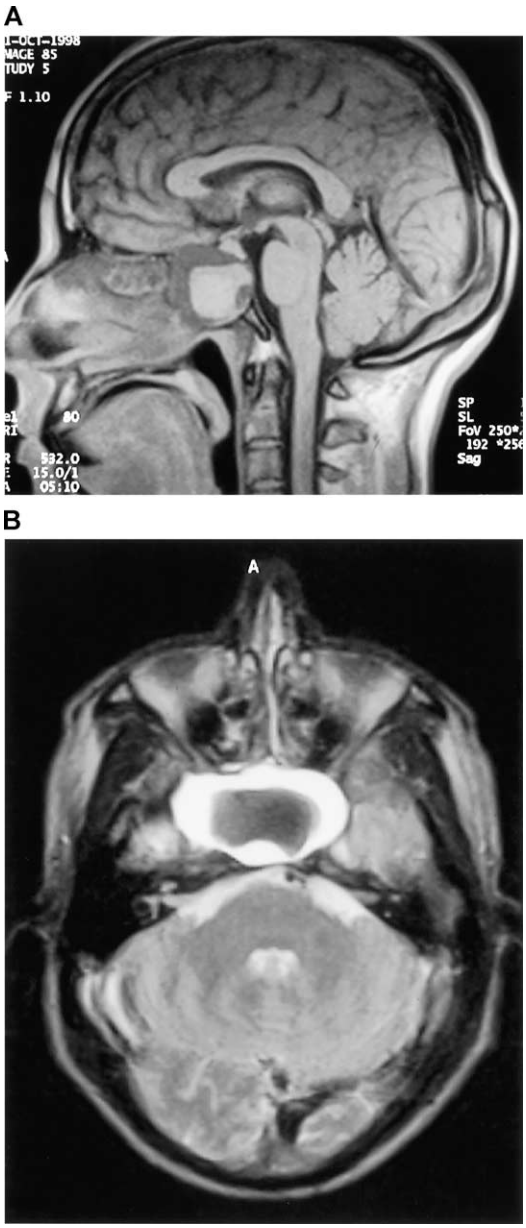

Fig. 1. Magnetic resonance image of brain. A: Sagittal T1-weighted image shows the mass extending superiorly from the sphenoid sinus into the sella. B: Axial T2-weighted image reveals an isointense mass with a bright signal border causing mass effect on the cavernous sinuses.

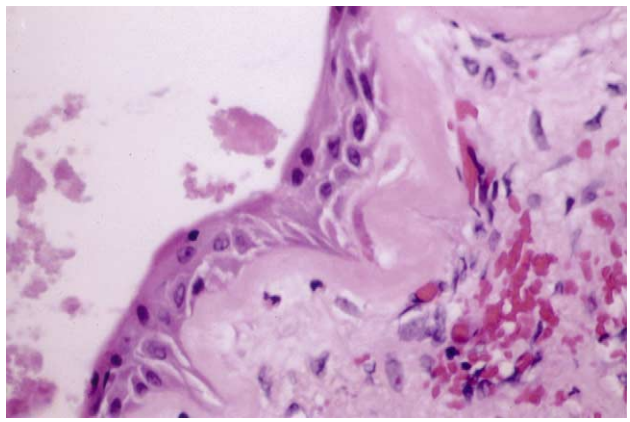

Fig. 2. Hematoxylin and eosin–stained section of mass demonstrating strips of stratified squamous epithelium, and a few superficial non-nucleated squamous cells amidst eosinophilic granular material.

The sublabial transsphenoidal approach was used. The anterior wall of the sphenoid sinus was soft and easily entered with a microcurette. Upon entering the sphenoid sinus, yellow adherent cheesy material was encountered. No hemorrhage or cystic fluid was found. The floor of the sella and clivus was completely eroded and the dura was visible. Under direct fluoroscopy, the anterior, posterior, and superior extent of the lesion was curetted, delivering the lesion from the sella to the clivus.

The specimen consisted of dry, flaky, cheesy material and tiny fragments of soft tissue. Light microscopy revealed a few preserved superficial non-nucleated squamous cells amidst eosinophilic granular material, and strips of stratified squamous epithelium resting on fibrosed chronically inflamed pericyst tissue (Fig. 2). No keratohyaline layer was evident on the superficial epithelial lining. This was consistent with an epidermoid cyst.

The patient's visual acuity improved to 20/40 OD and 20/40 OS by postoperative day 7. The diplopia had been resolved and the third cranial nerve function had returned to baseline by 3 months' follow-up.

3. Discussion

In 1829, Cruveilhier [7] reported the first description of the "tumeurs perlees." Several years later, after recognizing the presence of cholesterol crystals in a diploic epidermoid, Müller [13] introduced the term "cholesteatoma," but Virchow [25] soon recommended the abandonment of the term after realizing cholesterol was neither essential nor constantly seen in these tumors.

The location of this tumor with sellar extension and the acute neurological deterioration caused by such a slow-growing tumor are rare and unique characteristics. This clinical presentation warrants the addition of this lesion to the differential diagnosis of a patient who presents with a sellar mass and symptoms suggestive of pituitary apoplexy.