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Intrasphenoidal Rathke's Cleft Cyst: Case presentation and review of the literature

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Abstract: Rathke's cleft cyst is a benign lesion of embryological origin with sellar-suprasellar localization. It is found in 12-33% of normal pituitary glands in autopsy series. Although it is mostly asymptomatic, it can cause symptoms by compressing surrounding neural and pituitary tissues. The most common symptoms are endocrine problems, visual problems and headache. Uncommonly, the lesion can present with apoplexy. It is rarely reported outside sellar-suprasellar sites, such as the cerebellopontine angle, prepontine cistern, and intrasphenoidal locations. We present an intrasphenoidal Rathke's cleft cyst found during investigation of a headache and operated on. We discussed the case with literature review and two similar reported cases.

Key words: Cyst, intrasphenoidal, rathke cleft, sellar-suprasellar

Introduction

Rathke's cleft cyst (RCC) is a benign, intrasellar, epithelial cystic lesion with mucoid content (6, 9). It is found in 12-33% of normal pituitary glands in autopsy series (9). It is thought to originate from Rathke's pouch remnants. Although these sellar and suprasellar lesions are usually asymptomatic, they can also cause symptoms due to pressure on surrounding neural tissues and the pituitary gland (6). The most common symptoms are endocrine problems, visual problems and headache. Rarely, the lesion can present with apoplexy (6, 10). A few case presentations have reported in places other

than sellar and suprasellar locations. Two cases with pure intrasphenoidal localization were reported in the literature (4, 9). We present our intrasphenoidal case and discuss the case with literature review.

Case presentation

A 28-year-old female patient presented with headache extending to the left eye. Investigations revealed an isointense lesion filling the sphenoid sinus and eroding the planum sphenoidale and with contrast enhancement at the periphery on T1-weighted images. The sella base, the pituitary gland and stalk was intact (Figures 1 and 2). The examination of the patient revealed no

neurological deficit. No abnormality was present on laboratory analyses. The patient was operated through the transnasal transsphenoidal route. Fluid with the viscosity of motor oil discharged from the cystic structure filling the sphenoid sinus. Then the cyst wall was totally excised. Pathology preparation showed cytoke-
 ratin (+), S100 (-),

CD1a (-), and CD68 (+) in the cyst epithelium. The pathology report was Rathke's cleft cyst accompanied by xanthogranulomatous inflammation (Figure 3). The patient's headache resolved in the postoperative period. Postoperative 1st year follow-up MRI and paranasal sinus tomography revealed no contrast enhancing lesion (Figures 4 and 5).

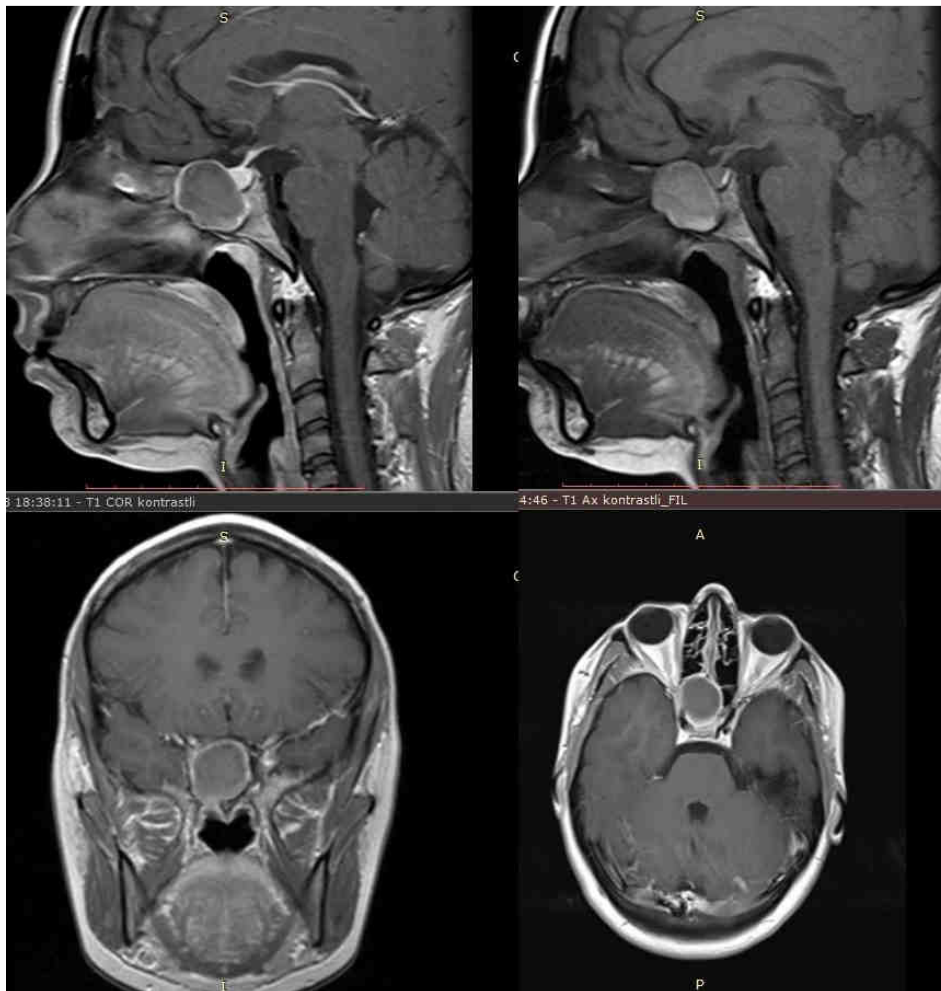


Figure 1 - Preoperative brain MRI images reveal an isointense lesion filling the sphenoid sinus and extending to the ethmoids with contrast enhancement of the periphery on T1-weighted images. The pituitary gland and stalk are seen to be intact

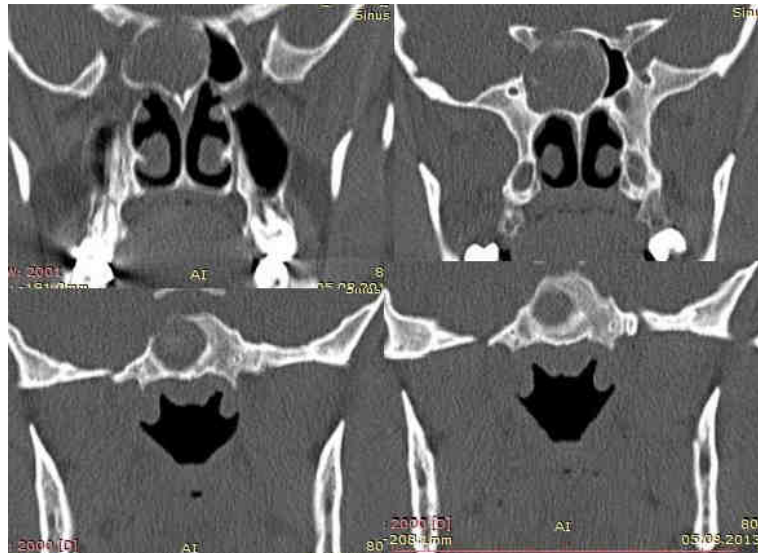


Figure 2 - Preoperative parasagittal sinus tomography reveals a cystic lesion with intrasphenoid sinus, mostly on the right, eroding the planum sphenoidale and extending to the ethmoids

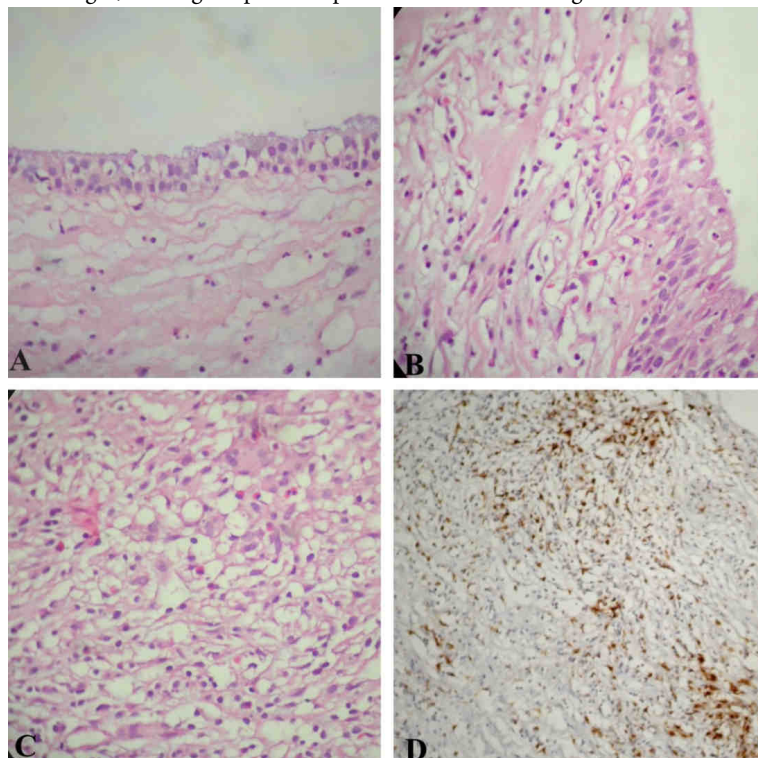


Figure 3A - HE X 20 with mucous and ciliated cyst epithelium. **3B**: Squamous metaplasia of cyst epithelium HE X 20. **3C**: Xanthogranulomatous inflammation at cyst wall HE X 20. **3D**: CD68 (+) histiocytes at the wall DAB X 10

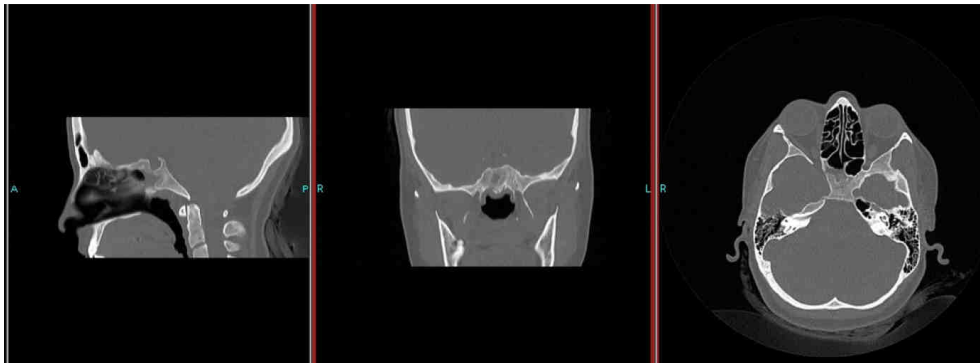


Figure 4 - Postoperative 1st year MRI follow-up reveals no cystic lesion in the sphenoid sinus

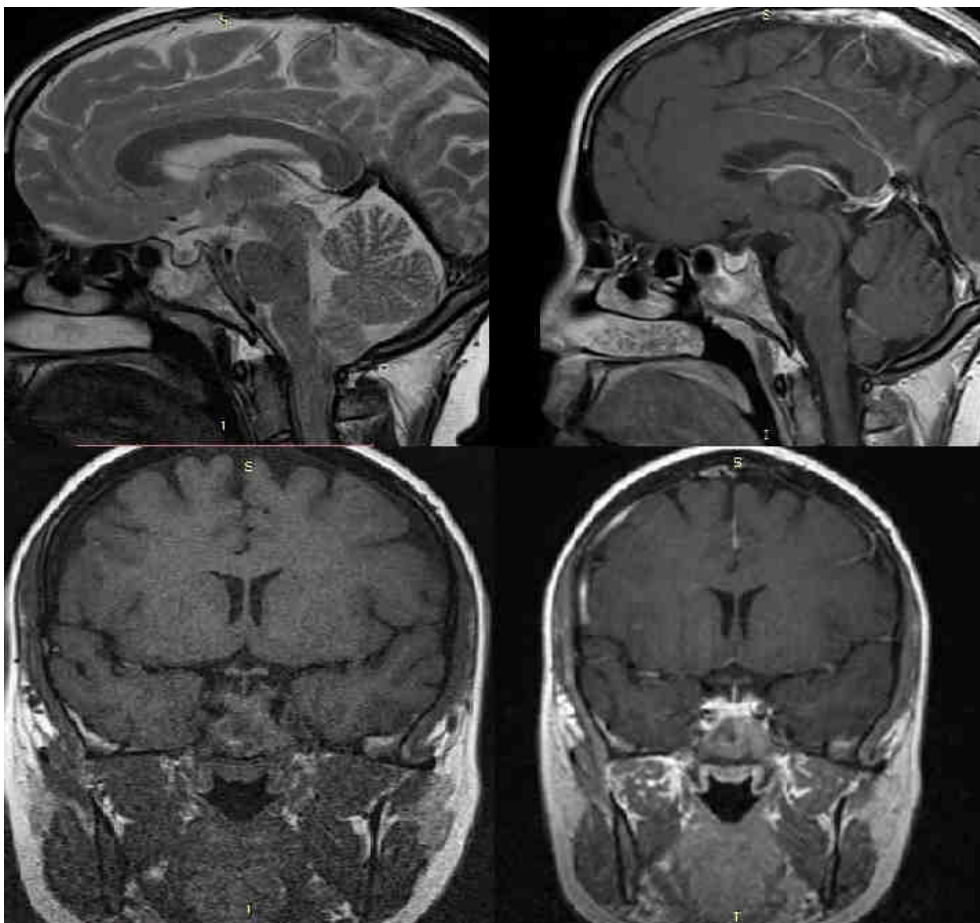


Figure 5 - Postoperative 1st year paranasal sinus tomography reveals no cystic lesion in the sphenoid sinus

Discussion

RCCs usually have an anterior sellar or anterior intrasellar-suprasellar localization. A couple of cases located at the pontocerebellar angle (3, 12) or prepontine cistern (5) have been reported. An intrasphenoidal symptomatic RCC was reported only in 2 cases in the literature (4, 9).

A pure sphenoidal location can be explained with the lesion's embryological origin (9). RCCs originate from Rathke's pouch remnants (9). This pouch develops from the ceiling of the stomodeum coated with epithelial cells of ectodermal origin as a diverticulum towards the diencephalon in the 4th gestational week (9). The infundibulum develops from the neuroepithelium of the diencephalon origin in the same period. It migrates towards the craniopharyngeal canal. While the hypophyseal diverticulum is extending in the 5th week, the part connected to the oral ectoderm narrows and then closes and degenerates in the 6th week. The infundibulum and Rathke's pouch become in contact again at the 5th week. The adenohypophysis and neurohypophysis develop in this way (9). This embryological process indicates that RCC's can be found at any location in the craniopharyngeal canal (9).

RCC lesions are usually small and asymptomatic. The presenting signs may be hormonal problems, apoplexy, hypophysitis, abscess, hypothalamic dysfunction, sphenoid sinusitis, oculomotor involvement, metabolic encephalopathy or visual problems depending on the location (1, 6, 7, 10, 11). Chronic cephalgia can also be seen with RCC. Acute

and severe headache can develop in patients with RCC due to bleeding inside the cyst, chemical meningitis, increased sellar pressure or local inflammation (6, 8).

The first intrasphenoidal RCC in the literature was presented by Megdiche-Bazarbacha et al (9). Headache and diplopia were present in a 41-year-old male patient in this case. The symptoms disappeared after spontaneous rhinorrhea but the headache and left visual loss recurred 4 months later. Radiological analyses revealed an intrasphenoidal lesion compressing the pituitary gland and optic chiasm. The patient was operated with the transrhinoseptal approach and the symptoms resolved postoperatively (9). The second case in the literature was presented by Kalina et al (4). A lesion that could be an intrasphenoidal Rathke's cleft cyst was found during investigations performed for seizures in a 13-year-old male patient in their case presentation. Mucocele, meningocele and craniopharyngioma were considered in the differential diagnosis. It was decided that the lesion was RCC and endoscopic fenestration was performed. The lesion had disappeared on 3rd month follow-up investigations (4).

Radiological RCC diagnosis has become more common with the start of MRI use. An RCC can be seen as a well-delineated lesion that is hyper/iso/hypointense on T1-weighted and hyperintense on T2-weighted MRI images depending on the amount of cholesterol, mucopolysaccharide and protein inside the cyst (2, 9). There is usually no contrast enhancement or circular enhancement may be present. CCT shows hypodense lesions

without contrast enhancement. The pituitary gland is usually around or below the cyst (9). The differential diagnosis mainly includes bleeding craniopharyngioma and adenoma. While RCC is usually not calcified, craniopharyngiomas become calcified and can be detected on CCT. Microscopically, RCC is covered with ciliated columnar or cuboidal epithelium and craniopharyngioma with squamous cells. The cyst content of RCC resembles motor oil. Calcification is usually not seen in RCC cases.

Most RCCs are asymptomatic and radiological preliminary diagnosis and differentiation from craniopharyngioma are therefore important. While the treatment aim for craniopharyngiomas is total excision, cyst drainage through the transsphenoidal route and limited excision of the cyst wall can be enough for RCC patients (9). Preliminary consideration of RCC radiologically will therefore determine how aggressive surgery will be (9).

Written informed consent of the patient was obtained.

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