Intrasphenoidal Rathke Cleft Cyst

SUMMARY: Symptomatic Rathke cleft cysts (RCC) are reported in the sellar and suprasellar regions, but no case of sphenoidal RCC has been reported. We report a case of sphenoidal RCC in a 41-year-old man. The lesion was revealed by headaches and diplopia. Symptoms disappeared transiently after a spontaneous rhinorrhea but relapsed 4 months later. MR imaging showed a cystic sphenoidal lesion, isointense on T1-weighted images (WI) with peripheral gadolinium enhancement and hyperintense on T2 WI. The patient underwent surgery through a transthyrosoptal approach. The wall of the sphenoid sinus was paper-thin. The cyst contained a motor-oil-like fluid and communicated widely with the nasal fossa. Its wall was partially extracted. Symptoms and signs ceased after surgery. MR imaging performed 1 year later showed the disappearance of the sphenoidal cyst. Embryological origin of RCCs is discussed. The hypothesis of a continuum between the different epithelial cystic lesions of the sellar and parasellar region is discussed. Imaging has an important impact on the diagnosis; nevertheless, the specific characterization remains difficult.

Discussion

The sphenoidal, infrasellar origin of this RCC is supported by the absence of a suprasellar extension and by the entirely extradural location of the cyst. Ectopic sphenoidal12–16 or even clival cranioopharyngioma11 have been reported, but no case of RCC has been reported in these locations.

The explanation of this location can be embryologic. Cranioopharyngiomas and RCCs arise from remnants of the Rathke pouch.9,12 This pouch appears during the fourth gestational week and arises as a dorsal diverticulum from the stomodeum lined by epithelial cells of ectodermal origin. Near the same time, the infundibulum forms as a downgrowth of the neuroepithelium from the diencephalon.1 It migrates following the craniopharyngeal canal. By the 5th week, the Rathke pouch comes into contact with the infundibulum; these 2 structures give rise to the adenohypophysis and neurohypophysis, respectively.

According to Erdheim’s theory,13 a cranioopharyngioma can arise from every part of the craniopharyngeal canal. This may also be true for RCC; in this case, the cyst derives from the canal enclosed in the bones of the cranial base, in contact with the walls of the pharynx.

Remnants of the craniopharyngeal canal can originate in a spectrum of cystic lesions ranging from simple RCCs to complex cranioopharyngiomas.2 Harrison et al14 suggested that there is a continuum between epithelial cystic lesions of the sellar and parasellar region (cranioopharyngiomas, RCCs, neuroepithelial cysts, epidermoid and dermoid cysts), which all appear to arise from the ectoderm. In some cases, they cannot be differentiated on the basis of clinical, biologic, and radiologic criteria. Even on histologic examination, there can be overlapping features. Russel and Rubinstein1 reported 2 dumbbell-shaped cysts with intrasellar components lined by cuboidal to columnar ciliated epithelium with an abrupt transition at the diaphragma sella to entirely squamous suprasellar tumors. These lesions demonstrated a histologic continuity of cranioopharyngioma and RCC. In a series of 19 cases of sellar and suprasellar epithelial cystic lesions examined histologically, Harrison et al14 reported that 9 displayed significant overlapping histologic features with a solely intrasellar cranio-
pharyngioma, which appeared to arise from an RCC with extensive squamous metaplasia.

Among the epithelial-lined cystic lesions of the sellar and parasellar region, craniopharyngiomas have the highest recurrence rates, exceeding 90% for partially resected lesions.15 At the opposite end of the spectrum lie RCCs, with a recurrence rate estimated at 5% by Voelker et al16 in their review of the literature.

Preoperative diagnosis of RCC is theoretically important, because the operative management may be limited to excision and biopsy of the cyst wall with drainage of the contents via a transsphenoidal route. CT usually shows a homogenous, low-attenuation, nonenhancing lesion.17,18 This nonspecific description matches with most of the sellar region lesions. MR imaging allows better characterization, showing a cystic intrasellar or intrasellar and suprasellar lesion. The exclusive suprasellar location is very rare.18 The intrasphenoidal location has never been reported, which made the diagnosis difficult in our case. Moreover the maximum size of the cyst usually ranges between 6 and 26 mm in diameter.19,20 In our case, the diameter was 40 mm. Signal intensity varies widely21,22; it seems to depend on the composition of the cyst and the quantity of cholesterol, mucopolysaccharides, and proteins.23 The most constant finding is a high-intensity signal on T2 WI and an isointense or hyperintense signal on T1 WI.23 In the reported case, the cyst showed an isointense signal on T1 and hyperintense signal on T2 WI. After gadolinium-diethylene-triaminepentaacetic acid (Gd-DTPA) administration, the pituitary gland is enhanced but not the cyst, which makes it easy to identify. The position of the pituitary gland is usually around or under the cyst. In the latter situation, a very particular aspect may be realized: the sign of “the egg in a cup” that is very suggestive of RCC.23 Despite all these features, no certain diagnosis can be made, especially when there are uncommon finding as in the present observation. However, imaging gives an exact description of the position of the cyst, which better guides the surgeon and allows preservation of the normal pituitary tissue.

Even in the hands of experienced neurosurgeons, the diagnosis of RCC still remains doubtful, because of the variability of the cyst content; the most difficult diagnosis remains between RCC and craniopharyngioma.2

In a review of 155 cases of RCC, Voelker et al. concluded that the trans-sphenoidal approach is the procedure of choice for sellar and suprasellar RCC and that partial excision and drainage of the cyst is the recommended treatment for these lesions.16 It proved to be a safe and effective procedure.

On the basis of a series of 28 cases of sellar and suprasellar RCC, El Mahdy and Powell2 recommend letting the cyst wall open in the sphenoid sinus to avoid recollection and recurrent cyst formation. He used fascia lata and fat grafts only in cases in which the arachnoid membrane was breached and an important cerebrospinal fluid leak was detected.

Conclusion
The diagnosis of RCC should be suggested in case of cystic lesions of the parasellar region even inside the sphenoid bone. Clinical and radiologic features are not specific. The main differential diagnosis is craniopharyngioma. Both have the same embryologic origin, but RCCs are easier to treat and have a much better prognosis.
References