



Huge intrasphenoidal Rathke's cleft cyst: a case description and analysis of the literature

Da In Lee, Kyung Mi Lee, Eui Jong Kim

Department of Radiology, Kyung Hee University Hospital, Kyung Hee University College of Medicine, Seoul, Korea

Correspondence to: Kyung Mi Lee, MD, PhD. Department of Radiology, Kyung Hee University Hospital, Kyung Hee University College of Medicine, #23 Kyunghee-daero, Dongdaemun-gu, Seoul 02447, Korea. Email: lolabunny0111@hanmail.net.

Submitted May 31, 2023. Accepted for publication Sep 06, 2023. Published online Nov 07, 2023.

doi: 10.21037/qims-23-780

View this article at: <https://dx.doi.org/10.21037/qims-23-780>

Introduction

Rathke's cleft cysts (RCCs) are non-neoplastic cysts derived from the remnant of the Rathke's cleft, which are rarely symptomatic and often incidentally found in 4–33% of autopsies (1). If the cyst is of substantial size, symptoms such as headache, visual impairment, and endocrine disturbance may develop, leading to its discovery. Symptomatic RCCs typically present between the fourth and fifth decades of life, with an average reported size of 10–20 mm (2). RCC greater than 4 cm in size are rarely reported. Here, we report a case of huge RCC with a diameter greater than 7 cm, occurring in an adolescent patient.

Case presentation

All procedures performed in this study were in accordance with the ethical standards of the institutional research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

An 18-year-old male presented with 3-week history of persistent left hemifacial numbness and weakness. The patient soon developed diplopia and hemianopsia in the left eye. Although the patient had a history of orbital blowout fracture, he had no lasting complications or known genetic disorders or additional traumatic events.

The extraocular muscle function test revealed diplopia with a right-down gaze abnormality. The patient promptly underwent brain MR, without conclusive results due to

artifacts of dental braces, which mandated a subsequent nonenhanced CT examination.

CT revealed a large lobulated cystic mass with a small solid portion occupying the parasellar and suprasellar areas (approximately 7.2 cm × 7.0 cm). It caused expansion of the bony sella and both nasal and paranasal cavities, causing a mass effect (*Figure 1*). No calcific lesions were observed in the cyst.

A repeated brain MRI was acquired after removing his dental braces, revealing 7.6 cm sized bone destructing mass occupying both the sphenoid sinuses, right maxillary sinus, posterior aspect of the nasal cavity, sellar, parasellar, and suprasellar areas. The mass was predominantly hyperintense on T2-weighted images (T2WI) and hypointense on T1-weighted images (T1WI) and fluid attenuated inversion recovery (FLAIR) images, suggestive of a cystic mass. After administration of a gadolinium contrast agent, the mass showed rim enhancement with another thin septa-like structure within the lesion. There were small nodular foci of T1-weighted hyperintensity and T2-weighted hypointensity with corresponding dark signal foci on susceptibility-weighted imaging (SWI), indicating small hemorrhages. No definite diffusion restrictions were observed. The mass expanded both orbital cavities and both cavernous sinuses, with a mass effect on both the temporal and posterior fossa, compressing the prepontine cistern. The pituitary gland was visualized at the superior aspect of the mass. There was no evidence of hydrocephalus. The differential diagnoses considered were cystic schwannoma, lymphatic malformation, chordoma, and aneurysmal bone cyst. Craniopharyngioma and encephalocele were also included

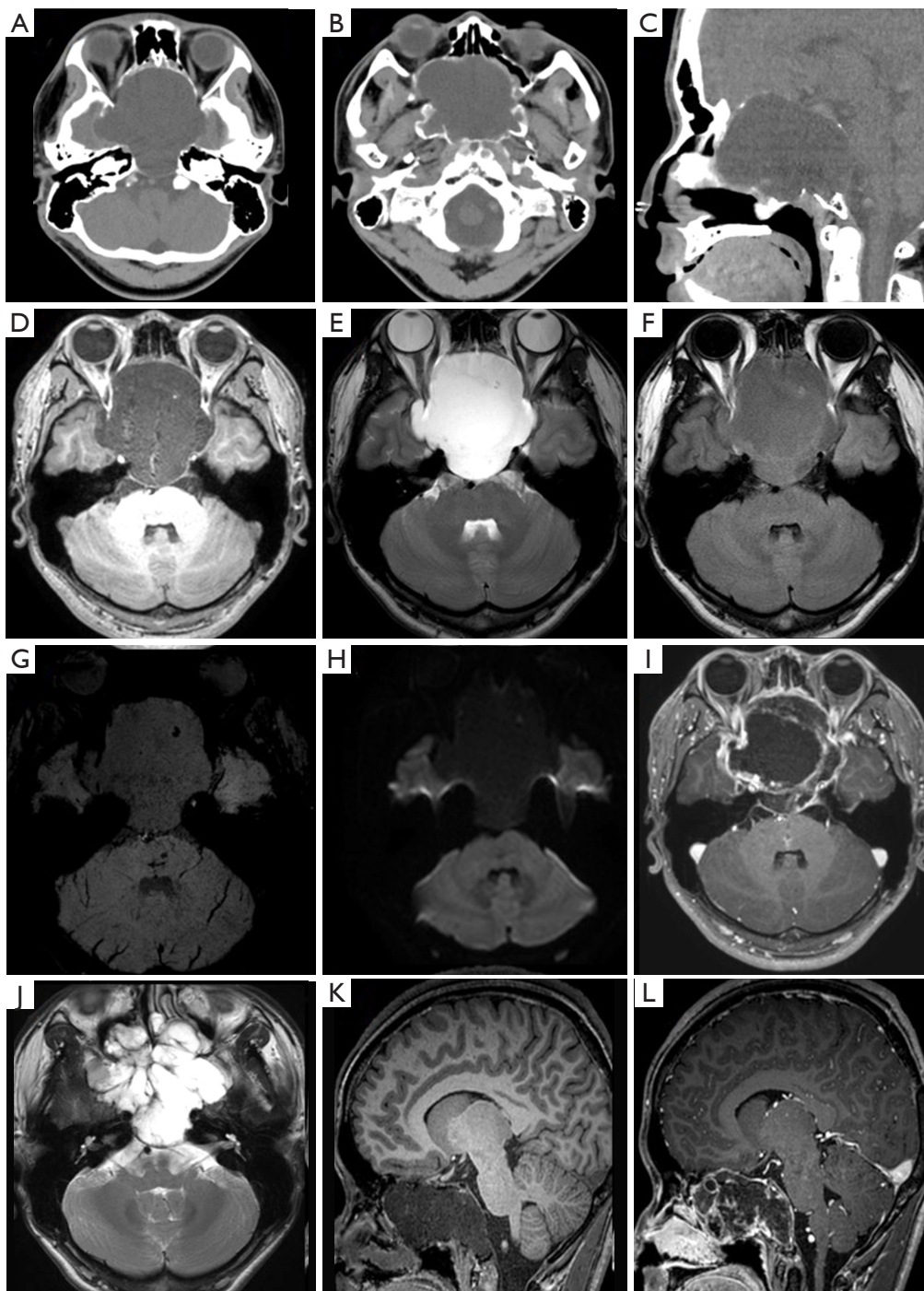


Figure 1 Axial and sagittal non-contrast CT (A-C), T1WI (D), T2WI (E) FLAIR (F), SWI (G), DWI (H) and gadolinium enhanced T1WI (I) showing an intrasphenoidal cystic lesion. Notice small T1-weighted hyperintense and T2-weighted hypointense nodule with blooming in SWI. Contrast enhanced T1WI shows rim enhancement with internal enhancing septa-like structure. Axial T2WI (J), sagittal T1WI (K) and contrast enhanced T1WI (L) showing recurred mass at 11-month follow-up. CT, computed tomography; T1WI, T1-weighted imaging; T2WI, T2-weighted imaging; FLAIR, fluid attenuated inversion recovery; SWI, susceptibility weighted imaging; DWI, diffusion weighted imaging.

Table 1 Case reports of giant Rathke's cleft cyst

Case No.	Reference	Age, years	Sex	Size, cm	Localization	Symptoms	Operation	Recurrence
1	Sivaraju <i>et al.</i> (4)	43	Male	5.6×4.3×3.3	Suprasellar	Headache, seizures	Mass excision with perianal craniotomy	×
2	Meyer <i>et al.</i> (5)	45	Female	>4	Clivus-sphenoid sinus-orbital roofs-prepontine cistern	Neck pain	Mass excision with bifrontal craniotomy with bilateral supraorbital osteotomy	×
3	Chuang <i>et al.</i> (6)	17	Male	4.0×2.9×1.9	Retroclival	Syncope	Cyst content removal with wall marsupialization via left retromastoid approach	×
4	Choudhry <i>et al.</i> (7)	35	Male	6.5 cm in greatest diameter	Suprasellar	Headache, gait disturbances, blindness in left eye	Two-stage operation: initial stereotactic cyst decompression → definitive mass removal	×
5	Liao <i>et al.</i> (8)	39	Female	5.7×3.7×4.8	Suprasellar	Depression, urinary incontinence, cognitive impairment, anisocoria, hyper-reflexia, positive Babinski sign, gait disturbance	Mass removal via left-sided pretemporal approach	×
6	Agarwal <i>et al.</i> (9)	8	Female	8×9×6.5	Right temporo-fronto-parietal	Right visual disturbance, nausea, vomiting	Drainage and marsupialization via right fronto-temporo-parietal craniotomy	×

in the differential diagnosis, but with a lower possibility. Patient underwent removal of mass via the transsphenoidal approach (TSA).

Histopathological examination revealed a cyst with ciliated pseudostratified columnar epithelium and goblet cells. Immunohistochemistry revealed positive SMA staining and negative staining for ALK1, cytokeratin, and STAT6. The features and anatomical location indicated the presence of a RCC.

At the two-year follow-up, the patient did not show any symptomatic recurrence. On follow-up MRI, however, there was an even larger (8 cm × 7 cm) cystic mass occupying both sphenoid sinuses, right maxillary sinus, posterior nasal cavity, clivus, sella, parasellar, and suprasellar areas. The mass showed a similar signal on T2WI and T1WI with rim enhancement, suggestive of recurrence of the previous lesion.

Discussion

RCC is a benign cyst originating from embryonic remnants of Rathke's pouch. Several authors reported that an RCC usually grow about 10–20 mm in diameter and mean cyst size is reported to be 16.3 mm (2). The cysts reported rarely exceed 4 cm in diameter (2,3). In a review of the literature using PubMed, EMBASE, and CENTRAL with keywords “giant”, “rathke”, “cyst”, and “case report” (singly or in combination), we screened 8 articles and identified 6 cases with diameter greater than 4 cm, and only one case had diameter exceeding 7 cm (*Table 1*) (4–9). To our knowledge, this is the first case with recurrent symptomatic RCC exceeding 7 cm in diameter with predominantly infrasellar growth.

Although the radiological characteristics of RCCs have been extensively investigated, differentiating RCCs from other cystic sellar/parasellar lesions remains

challenging. RCCs are often misdiagnosed as other cystic sellar and parasellar lesions such as craniopharyngioma, pituitary adenoma, chordoma, and epidermoid cysts. Often, MR imaging shows variable signal intensities on T1WI and T2WI, with varying locations. Usually, RCC is hypointense on T1WI and mostly hyperintense on T2WI, with varying heterogeneity. These MR signal intensities depend on the composition of the cyst such as proteins, mucopolysaccharides, and cholesterol. Contrast-enhanced MRI shows rim enhancement, sometimes due to the surrounding adenohypophysis mimicking wall enhancement. In addition, RCCs may also show patchy enhancement attributed to squamous metaplasia in the cyst wall or inflammatory changes (10,11). The presence of an intracystic nodule, with an incidence of 37.5–45%, has been suggested as a characteristic MR feature of RCC compared to cystic pituitary adenoma. However, in rare cases, pituitary adenomas may have intracystic nodules (67.9% *vs.* 16.7%), and these nodules may often be confused with hemorrhages (12).

In the present case, the location of the cyst was unusual. RCC originates from remnants of the Rathke pouch that appear at the 4th gestational week, arising as a dorsal diverticulum from the stomodeum lined by epithelial cells of ectodermal origin. At the 5th week of gestation, the Rathke's pouch contacts the infundibulum growing downward from the neuroepithelium of the diencephalon along the craniopharyngeal canal (13). Remnant of the craniopharyngeal canal can give rise to a spectrum of cystic lesions, such as simple RCCs, complex craniopharyngiomas, neuroepithelial cysts, epidermoid cysts, and dermoid cysts, all of which arise from the ectoderm (14).

Intrasphenoidal RCCs are rare and rarely present with symptoms such as visual disturbance or endocrinopathy. To the best of our knowledge, only eight cases have been documented in the literature presenting with diplopia, owing to tumor extension into the middle cranial fossa or orbit (15). Histopathological analysis is the gold standard for diagnosing RCC in patients with sellar/parasellar cystic lesions. On microscopy, RCC demonstrates a simple columnar or cuboidal epithelium with ciliated goblet cells or pseudostratified columnar cells. Stratified squamous epithelium and chronic inflammation favor craniopharyngiomas. Stratified squamous epithelium from metaplasia of the epithelial lining has been described, which may be related to recurrence and its more aggressive nature (4).

The primary treatment for symptomatic cases is wall

excision and cyst decompression, usually using the TSA. Common complications following surgery for RCC include recurrence, CSF rhinorrhea, and endocrine dysfunction. The reported recurrence rate for RCC varies between 0–30%. In a meta-analysis, the recurrence rate was found to be 12.8% owing to factors such as squamous metaplasia in the cyst wall, enhancement of the wall on MRI, and surgical methods.

Compared to other cases in the literature, our case shows combination of uncommon features in RCC where the age of the patient, size, location, and recurrence after surgical treatment was atypical. Large intrasphenoidal RCC is extremely rare. Although histopathology plays a key role in diagnosis in the absence of specific clinical and radiological features, being aware of such a presentation of RCC may be valuable in clinical management and consideration of periodic follow-ups, even without recurring symptoms.

Acknowledgments

Funding: None.

Footnote

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at <https://qims.amegroups.com/article/view/10.21037/qims-23-780/coif>). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study were in accordance with the ethical standards of the institutional research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

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Cite this article as: Lee DI, Lee KM, Kim EJ. Huge intrasphenoidal Rathke's cleft cyst: a case description and analysis of the literature. *Quant Imaging Med Surg* 2023;13(12):8864-8868. doi: 10.21037/qims-23-780