

# Symptomatic Rathke's Cleft Cyst with Rapid Enlargement Masquerading as Rathke's Cleft Cyst Apoplexy

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To the Editor: A 35-year-old Chinese woman was admitted to our hospital complaining of a 1-year history of bilateral temporal chronic intermittent moderate headache which used to be alleviated by ibuprofen tablets or rest and had increased in intensity in the past 1 month. The patient also presented with the slow development of visual deficit. In January 2012, the patient presented with polyuria and polydipsia suggestive of central diabetes insipidus. Endocrine studies showed no clinically significant results. The patient also underwent a brain magnetic resonance imaging (MRI) which demonstrated an ovoid sellar lesion measuring 6 mm × 6 mm, with hyperintense, with no apparent enhancement after administration of gadolinium-diethylenetriaminepentaacetic acid (Gd-DTPA) on T1-weighted images and iso- to hypo-intense on T2-weighted images [Figure 1a]. The patient was initially diagnosed with Rathke's cleft cyst (RCC) and refused to undergo the operation. By the end of 2013, she once again presented with polyuria and polydipsia that were in spontaneous remission in the following 6 months with no medical intervention. A radiographic follow-up was administered in July 2014 demonstrating an ovoid sellar lesion measuring 9 mm × 11 mm × 14 mm indicative of enlargement of the former cyst, with hyperintense, with no obvious enhancement after administration of Gd-DTPA on T1-weighted images and iso- to hyper-intense on T2-weighted images [Figure 1b], and apparently, the signal characteristics of the MRI were almost the same as the initial one suggesting likely hemorrhage. From July 2014 to May 2015, she sought for medical services in the Department of Traditional Chinese Medicine and regularly took medicinal herbs aiming to promote circulation and remove stasis. In May 2015, she complained of a slightly blurred vision of her right eye. In November 2015, she was attacked by a severe unbearable headache, a vision's sharp decline of the right eye, and mild right temporal hemianopsia. The pain was diffuse, dull, and persistent for a week and could not be alleviated by tablets or rest as before. Her medical history was otherwise unremarkable. The general physical examination was uneventful. The neurological examination showed an awake, alert, and completely oriented patient with no focal sign. The muscular strength and tension of her four limbs were completely normal. Various physiological reflexes could be elicited in response to external stimulus. Pathological signs were absent. The results of ophthalmologic tests were in accordance with her self-reported symptoms indicating remarkable changes of visual acuity (VA) and visual field. Preoperatively, VA

of the right eye was 0.5+, and VA of the left eye was 0.8. Partial temporal hemianopia was observed with no suspect [Figure 1c]. The results of hematologic and biochemical tests were uneventful. Endocrine evaluations were within the normal ranges. MRI studies demonstrated a sellar and suprasellar lesion with hyperintense, with no apparent enhancement after administration of Gd-DTPA on T1-weighted images and hyperintense on T2-weighted images, measuring 16.8 mm × 16.0 mm × 14.6 mm [Figure 1d]. Located in the inferoposterior position of RCC, there was an irregular-shaped lesion showing hypointense on T1-weighted images and hypointense on T2-weighted images suggesting internal apoplexy. The mixed signal intensity was thought to be consistent with the symptom of apoplexy indicating intracystic old hemorrhage of RCC. Repeated MRI findings revealed the rapid enlargement of RCC within <4 years which was quite uncommon and likely to be described in published cases for the first time. The patient underwent an endoscopic endonasal transsphenoidal resection for RCC. In the procedure, the intracystic contents, which were seemingly yellowish cholesterol crystals instead of expected stale hemorrhage, were completely drained off, and the wall of RCC was resected as much as possible in a safe manner. Intracystic hemorrhage of RCC apoplexy was denied by surgical exploration. Intra- and post-operatively, leakage of cerebrospinal fluid and intracranial infection did not happen. The results of histopathological examinations proved the cystic lesion to be RCC. The patient recovered from the chronic headache; however, the visual deficits failed to improve significantly.

According to a very limited quantity of published reports on RCC apoplexy, the prevalence of RCC apoplexy in women is higher than that in men. Clinical manifestations of the patients with RCC apoplexy are similar to those with pituitary tumor apoplexy. Chronic headache is the most dominant presentation. The headache of our patient was severe, with sudden onset which was highly suggestive

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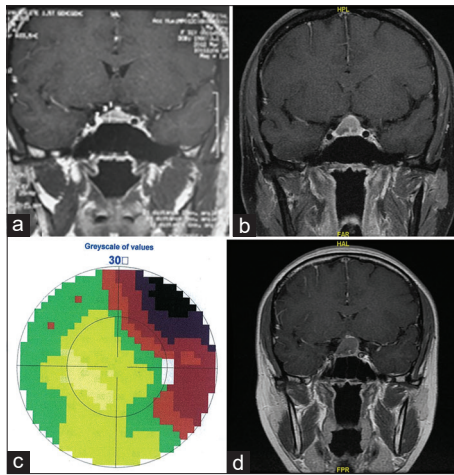
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**Figure 1:** T1-coronal MRI with Gd-DTPA showed enlargement of RCC during the follow-up period in 2012 (a), 2014 (b), and 2015 (d). The figure of visual field showed partial temporal hemianopia of the right side (c). MRI: Magnetic resonance imaging; Gd-DTPA: Gadolinium-diethylenetriaminepentaacetic acid; RCC: Rathke's cleft cyst.

of apoplexy and was believed to result from the sudden expansion of the mass in the pituitary fossa, and in the meantime, there was strong radiographic evidence of hemorrhage on MRI. The patient also complained of the deterioration of her VA of her right eye and the ophthalmological tests revealed temporal hemianopia of the right side. MRI indicated the radiographic mass effect showing a large cystic lesion which was also proven by intraoperative findings. Hemorrhage into RCC has been reported, but to our knowledge, RCC with rapid enlargement behaving like RCC apoplexy is first reported in our case.

Up to now, no typical and consistent MRI features have been identified for RCC. It is suggested that the presence of a posterior ledge of the diaphragma sellae or an intracystic nodule should be pathognomonic of RCC leading to early diagnosis. The signal intensity widely varies from lesion to lesion directly depending on the biochemical nature of each intracystic material. Considering the highly variable appearances of RCC on MRI, differential

radiologic diagnosis is regularly required. The patient in our case exhibited a cystic lesion in the intrasellar region with mixed signals. According to the MRI features of the patient, we regarded the cystic lesion very likely to be RCC apoplexy. However, intraoperative visualization and/or pathological confirmation of hemorrhage on the background of RCC remain the gold standard in diagnosing RCC apoplexy. The preoperative diagnosis of RCC apoplexy of our patient mainly according to MRI features was not in consistency with the gold standard.

RCC, a nonneoplasm cystic mass, usually remains steadily small and asymptomatic throughout the life in most patients.<sup>[1]</sup> However, some appear clinically significant for the growth and the subsequent mass effect on the surrounding structures in the sellar region. Although RCC apoplexy is rarely reported in published data, vigilance should be raised in the differential diagnosis of cystic mass in the sellar region.<sup>[2]</sup> Surgical treatment is the optimal approach for those patients with symptomatic RCC because histopathological results and intraoperative findings of the cystic lesion remain the gold standard for diagnosing RCC or RCC apoplexy, and surgery can alleviate mass effect-related symptoms and even lead to the recovery of endocrine dysfunction.<sup>[3]</sup> Through the case report, we hope that our experience can help enrich the clinical and radiographic differential diagnosis of RCC apoplexy and the further understanding of RCC.

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### Conflicts of interest

There are no conflicts of interest.

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