Ruptured Rathke Cleft Cyst Mimicking Pituitary Apoplexy

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Abstract

Keywords

- Rathke cleft cyst
- pituitary apoplexy
- ► sellar mass
- ► RCC rupture
- nonhemorrhagic RCC apoplexy

Rathke cleft cysts (RCCs) are benign cystic lesions of the sellar and suprasellar region that are asymptomatic in most cases. Occasionally, compression of the optic pathway and hypothalamo-pituitary structures may cause clinical symptoms, such as headaches, visual deficits and endocrinopathies. Acute presentation caused by hemorrhage into an RCC have been described in the literature, and the term "Rathke cleft cyst apoplexy" has been coined. We present the case of a 32-year-old man with acute onset of meningitistype symptoms and imaging findings resembling hemorrhagic pituitary tumor apoplexy. In retrospect, clinical symptoms, intraoperative appearance, and histologic examination were compatible with the diagnosis of nonhemorrhagic rupture of an RCC. Thus, the clinical presentation of "Rathke cleft cyst apoplexy" is not necessarily caused by hemorrhage.

Introduction

Rathke cleft cysts (RCCs) are benign cystic lesions of the sellar or suprasellar region developing from poor obliteration of the craniopharyngeal duct.^{1,2} RCCs consist of a single layer of epithelium composed of ciliated columnar or cuboidal cells.³ Common incidental findings on autopsy,⁴ RCCs are asymptomatic in most cases. Clinical symptoms are mainly caused by compression of perisellar structures such as the optic chiasm or the hypothalamo-pituitary region.⁵ There have been a few reports on RCCs presenting with a syndrome resembling pituitary tumor apoplexy.⁶ Recently, a new clinical entity called "Rathke cleft cyst apoplexy" has been suggested by Chaiban et al, strictly referring to intracystic hemorrhage into an RCC.⁶ We present a case of nonhemorrhagic rupture of an RCC presenting with clinical symptoms

received October 24, 2012 accepted after revision February 11, 2013 published online May 21, 2013 and imaging findings mimicking pituitary apoplexy (i.e., bleeding into a pituitary adenoma).

Case Report

A 32-year-old man with no relevant past medical history presented to our emergency department with a 5-day history of bifrontal headaches accompanied by nausea, vomiting, and photophobia resistant to analgetic and antiemetic drugs. Out of his usual state of health, the patient initially experienced a sudden onset of tingling paresthesias in the fingertips of his right hand as well as fevers and chills. About 1 hour later, he noticed bifrontal headaches. The next day (day 1), he was seen as an emergency at another hospital; the initial physical examination findings were normal. In the wake of elevated temperatures (38.1°C), headaches, and photophobia, the

© 2013 Georg Thieme Verlag KG Stuttgart · New York DOI http://dx.doi.org/ 10.1055/s-0033-1343985. ISSN 2193-6315. workup mainly consisted of a lumbar puncture (2 cells/µl) to rule out meningitis and contrast-enhanced magnetic resonance imaging (MRI) of the brain, which was inadvertently read as normal. The patient was discharged on an analgetic regimen. Whereas fevers, chills and paresthesias were only present on the first day, the bifrontal headaches, nausea, vomiting, and photophobia failed to improve, and the patient subsequently presented to our hospital (day 5). We repeated the lumbar puncture as well as MRI (day 6). The cerebrospinal

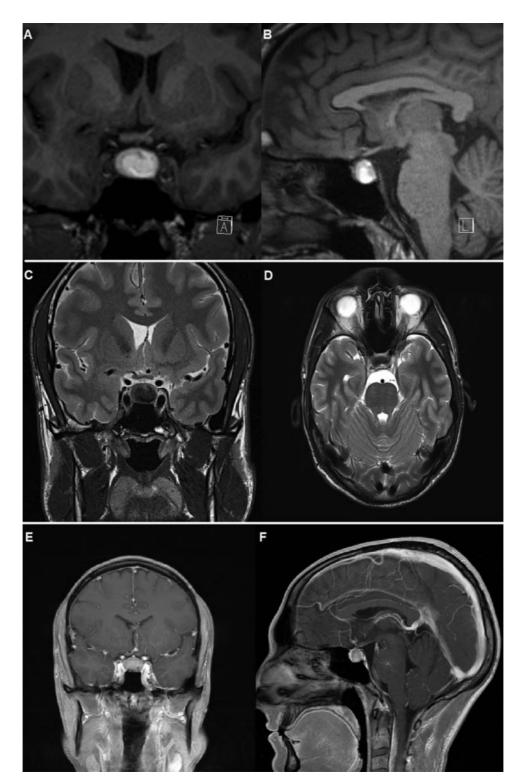


Fig. 1 Magnetic resonance imaging of the brain (day 6) showed the lesion to be hyperintense in nonenhanced T1-weighted coronal (A) and sagittal (B) images and hypointense on T2-weighted coronal (C) and sagittal (D) views. After gadolinium administration, slight intralesional contrast enhancement was seen (E, F), most likely representing the pituitary gland. Sphenoid sinus mucosal thickening is seen focally at the sellar floor (F).

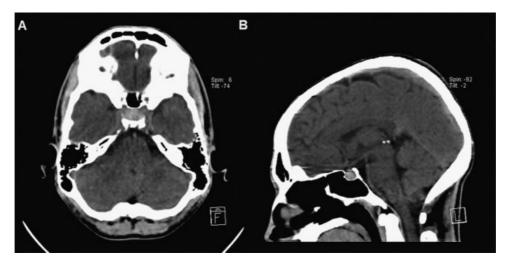


Fig. 2 Nonenhanced computed tomography scans showed the lesion to be hyperdense in axial (A) and sagittal (B) views.

fluid (CSF) contained 7 white blood cells/µl, a slightly decreased lactate level (1.4 mmol/L; reference range, 1.5–2.1 mmol/L), and a minimally elevated protein level (0.497 g/L; reference range, 0.2–0.4 g/L). Routine blood chemistry laboratories (C-reactive protein [CRP], 1.0 mg/L; sodium, 143 mmol/L; glucose, 4.9 mmol/L) and a complete blood count (hemoglobin, 132 g/L; Leukocyte count 4.97 X 10^9 / L, or 4.97 G/L) were within normal limits. On neurologic examination, visual function was normal, and no cranial nerve palsies or focal neurologic deficits were present. Classical nuchal rigidity was absent because full range of motion was present with flexion of the head, but the headaches were exacerbated by this maneuver. MRI of the brain revealed ballooning of the

pituitary gland with T1-hyperintense and T2-hypointense signal changes consistent with either an RCC or hemorrhagic pituitary apoplexy (**>Fig. 1**). A subsequent computed tomography (CT) scan then showed the lesion to be hyperdense consistent with hemorrhagic pituitary apoplexy (**>Fig. 2**). Endocrinologic workup revealed neither hormone excess nor hormone deficiency of the anterior pituitary except for a slightly elevated prolactin (26.4 µg/L; reference range, 4.0–15.2 µg/L).

The patient subsequently underwent transnasal-transsphenoidal surgical exploration (day 7). Intraoperatively, the pituitary gland appeared to be displaced and showed inflammatory changes. Colloidal material was seen within the

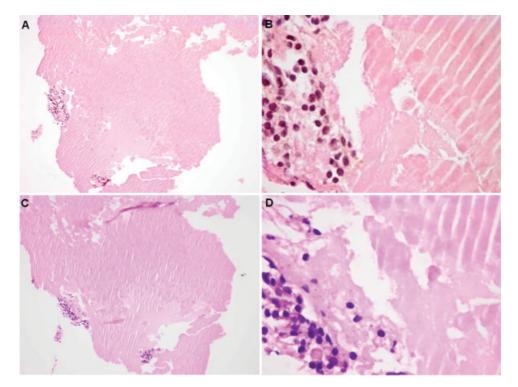


Fig. 3 Elastica van Gieson staining (A, 100x) demonstrates the amorphous character of the main material (B, 600x). Hematoxylin and eosin staining (C, 100x) shows the small fraction of cuboidal cells with no evidence for ciliae in high magnification (D, 600x).

sella; numerous gelatinous pieces of tissue were sent for histologic examination. Evidence of hemorrhage was not seen throughout the entire procedure.

Histologic examination showed mainly amorphous material and some cuboidal cells, which did not show characteristic ciliae (**Fig. 3**). These findings in the presence of compatible radiologic features are diagnostic for RCC. Postoperatively, the patient recovered quickly, and 3 days later he was discharged without focal neurologic deficits, free of headaches, and in his usual state of health.

Two weeks postoperatively, the patient presented again to our emergency department complaining of polyuria and polydipsia. Although he was drinking 5 L of water daily, sodium and osmolality in serum and urine were within the normal range. The diagnosis of diabetes insipidus (DI) could not be made. However, because the history of the patient was very suggestive for DI, we started a low-dose desmopressin therapy for symptomatic relief. Possibly these symptoms will resolve in the near future.

Discussion

RCCs are rarely symptomatic, and clinical symptoms may include-apart from headaches-endocrinopathies and visual dysfunction caused by mass effect of the lesion.⁵ Chaiban and colleagues presented a series of 11 cases and reviewed another 10 literature cases of patients undergoing surgery for hemorrhage into an RCC.⁶ They coined the term "Rathke cleft cyst apoplexy" describing this clinical condition. Based on clinical findings and neuroradiologic features, a distinction between pituitary tumor apoplexy and hemorrhage into an RCC cannot be made without surgical intervention and histologic confirmation. An earlier report by Binning et al also describes cases of patients with no signs of hemorrhage but with clinical and radiologic findings resembling pituitary tumor apoplexy or hemorrhagic RCC apoplexy.⁷ They argue that nonhemorrhagic RCC apoplexy might mimic imaging features of hemorrhage caused by intracystic nodules with similar MRI characteristics. Nevertheless, Chaiban and colleagues doubt that the cases reported by Binning et al represent a true variation of the "Rathke cleft cyst apoplexy" syndrome but believe that in the nonhemorrhagic cases patients underwent surgery a couple of months after initial onset of symptoms, allowing the blood components to be absorbed after the initial apoplectic event. However, the patient we present had never been symptomatic before the initial and sudden onset of symptoms, and he underwent imaging on days 1 and 6 and surgery on day 7 after initial symptoms. If this apoplexy-like event would have been hemorrhagic, intraoperative and histologic findings would clearly have indicated this. Rather, our assessment of the presented case is a rupture of an RCC, causing some colloidal fluid to leak out and to produce a mild form of chemical meningitis (headache, photophobia, fevers, chills, nausea, vomiting, and slightly abnormal CSF findings). The elevated prolactin level could be attributable to stalk compression or use of metoclopramide, which our patient repeatedly received as an antiemetic; its antidopaminergic effects readily explain mild hyperprolactinemia.⁸

Liu and Couldwell have suggested a grading scale for pituitary apoplexy in which grade I patients present with headache, endocrinopathy, or both; grade II describes patients with additional cranial nerve deficits; and grade III is reserved for patients with a decreased level of consciousness.⁹ Using this grading scale, if applicable, the clinical symptomatology of our patient would be best described as grade I.

Regarding the histopathology in this case, only a few cuboidal cells and mainly amorphous material were seen. In the presence of presurgical neuroradiologic findings consistent with RCC, these histologic features can be considered diagnostic.¹⁰

Conclusion

Clinical and radiologic features mimicking pituitary apoplexy can be consistent with a nonhemorrhagic RCC. Rupture of the cyst and leakage of its colloidal material is a potential pathomechanism in this case. Therefore, potential suddenonset clinical complications in patients with RCCs are not restricted to hemorrhagic cases alone as suggested by the term "Rathke cleft cyst apoplexy," and the event of a rupture of the cyst should be included in the differential diagnosis of pituitary apoplexy, which is most commonly caused by bleeding into a pituitary adenoma.

Conflict of Interest

References

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