Rathke cleft cyst presenting with hyponatremia: an unusual presentation

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The authors report a case of Rathke cleft cyst presenting with severe hyponatremia. A 33-year-old man suffered sudden severe headaches, visual changes, dizziness, nausea, vomiting, and a metallic taste in his mouth. Initial laboratory values demonstrated severe hyponatremia. Magnetic resonance imaging revealed a cystic lesion with questionable intracystic hemorrhage, concerning for pituitary apoplexy. Transphenoidal decompression and drainage of the cyst confirmed the diagnosis of Rathke cleft cyst and resolved the symptoms. Postoperative follow-up studies at 6 months demonstrated normal endocrine function and no evidence of a cyst. (DOI: 10.3171/2011.4.FOCUS1180)

KEY WORDS • Rathke cleft cyst • hyponatremia • transphenoidal surgery • pituitary • endocrine dysfunction • intrasellar cyst • pituitary apoplexy

Rathke cleft cysts are benign cystic remnants of the craniopharyngeal duct. They are commonly located in the sellar and suprasellar region. First described by Luschka in 1860 as “an epithelial area in the capsule of the human hypophysis resembling oral mucosa,” these remnants of the Rathke pouch have been estimated, based on autopsy examinations, to be present in 22% of the population. Despite this high prevalence, they remain largely incidental findings, with an increase in recent diagnoses likely resulting from improvements in imaging of the sellar and suprasellar region. Occasionally, however, they may become large enough to cause symptoms, and these cysts now comprise 5%–10% of all resected sellar lesions. Presenting symptoms typically include headache, visual disturbances, and pituitary dysfunction. Endocrinopathies associated with pituitary dysfunction include growth hormone deficiency, hypogonadism, hypothyroidism, and hypocortisolemia.

Rarely, RCCs can present in apoplexy, with acute-onset headache, vision changes or loss, nausea, vomiting, meningismus, and even hypothalamic dysfunction. The variation in clinical and imaging appearance and the fact that RCCs can present with nonhemorrhagic apoplexy make diagnosis of this presentation difficult. We report a case of an RCC in which the patient had profound hyponatremia and for whom we had preoperative concern for pituitary apoplexy, an unusual presentation.

Abbreviation used in this paper: RCC = Rathke cleft cyst.

Case Report

History and Presentation. This 33-year-old man without a significant medical history was initially seen at an outside hospital after a motor vehicle accident. At that time, he was thought to have a possible cervical spine injury, for which he was started on a hydrocortisone taper. In addition, he was scheduled for MR imaging the following week. Although he initially had improvement in his symptoms, 5 days later he experienced an acute-onset headache, which progressively worsened over the next 2 days, along with dizziness, nausea, vomiting, and a metallic taste, and he returned to the outside hospital for evaluation. Physical examination showed bitemporal hemianopsia, and laboratory results at that time revealed severe hyponatremia (Na 112 mmol/L). Treatment with 3% hypertonic saline was initiated. Magnetic resonance imaging revealed a 1.3-cm cystic, enhancing intrasellar lesion compressing the optic chiasm superiorly (Fig. 1). Given the acute onset of symptoms and presence of hyponatremia, there was concern for pituitary apoplexy, and the patient was transferred to our facility.

Examination and Operation. On admission to our facility, the patient underwent full endocrinological examination. The levels of follicle-stimulating hormone, luteinizing hormone, and growth hormone were normal, but adrenocorticotropic hormone, cortisol, thyroid-stimulating hormone, and T4 levels were low (Table 1). As the
patient appeared to be euvolemic and had normal kidney function (blood urea nitrogen level 14 mg/dl and creatinine level 0.75 mg/dl), he was not thought to have cerebral salt-wasting syndrome. Because sick euthyroid syndrome was a possibility, the patient was given levothyroxine, and hydrocortisone treatment was restarted. After his endocrinological levels were optimized, the patient was taken to the operating room for decompression and drainage of the cystic mass. A portion of the cyst wall was taken for histological confirmation, and the pathological analysis was consistent with RCC (Fig. 2).

Postoperative Course. The patient’s symptoms resolved, and his sodium levels normalized. He was discharged home on postoperative Day 4. At his 6-month follow-up examination, he remained neurologically intact, and follow-up MR imaging revealed complete resolution of the lesion (Fig. 3). In addition, given his normalized endocrine function, the levothyroxine treatment was discontinued, and the hydrocortisone was tapered off as well.

Discussion

Despite the relatively high prevalence of RCC seen on postmortem analysis, these lesions are largely asymptomatic. Patients with symptomatic RCCs commonly present with headaches and symptoms related to the compression of the optic chiasm, hypothalamus, pituitary gland, and structures within the cavernous sinus.4,10,12,17 Ophthalmological symptoms include vision loss and various visual field defects. Concerning endocrinopathies, deficiencies in growth hormone, hypogonadism, hypothyroidism, and hypocortisolism have been noted (Table 2).1,5,9 Hyponatremia is a rarely documented presentation of RCC.2,6,13 Our case is similar to the recently reported presentation of nonhemorrhagic RCC apoplexy by Binning et al.3 unique to the patient in the current case, however, is that he presented with severe hyponatremia. Previous reports of RCCs presenting with apoplexy all described intracystic hemorrhage seen at the time of surgery.11,12 In the case series by Binning et al., preoperative MR imaging also demonstrated mixed signal intensities suggestive of hemorrhagic pituitary tumor, which intraoperatoratively was found to be the intracystic nodule of the RCC. Because intracystic hemorrhages have also been noted within RCCs, the diagnosis becomes even more difficult. Our patient also presented with acute clinical symptoms of apoplexy with preoperative MR imaging findings suggestive of a possible hemorrhagic pituitary tumor. Intraoperatoratively, we discovered a nonhemorrhagic, cystic lesion consistent with an

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<th>Hormone</th>
<th>Patient’s Values</th>
<th>Normal Range</th>
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<tr>
<td>Na (mmol/L)</td>
<td>121</td>
<td>136–144</td>
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<tr>
<td>adrenocorticotropin hormone (pg/ml)</td>
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<td>cortisol (μg/dl)</td>
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<td>0–9</td>
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<td>growth hormone (ng/ml)</td>
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<td>0.01–1</td>
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<td>luteinizing hormone (IU/L)</td>
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<td>1.7–8.6</td>
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<tr>
<td>prolactin (mg/ml)</td>
<td>6.6</td>
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<tr>
<td>thyroxine (μg/dl)</td>
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Fig. 1. Initial MR images. Contrast-enhanced sagittal (A) and coronal (B) T1-weighted MR images showing an intrasellar lesion. Note the rim enhancement and superior displacement of optic chiasm. The lesion has both hypointense (arrowhead) and isointense (intracystic nodule) components. Axial T2-weighted MR image (C) demonstrating hyperintensity (arrow) with isointense component and a questionable fluid level.

Fig. 2. Photomicrograph of tissue sample illustrating benign ciliated epithelium (red arrow) and reactive foamy macrophages (white arrow). H & E, original magnification × 400.
Rathke cleft cyst with hyponatremia

The RCC provides evidence that an RCC may present with symptoms shortly after decompression and drainage of the RCC. The cause of the hyponatremia presents an interesting clinical quandary. There have been 10 reported cases of hyponatremia as the presenting symptom in RCC. In a retrospective analysis, Ogawa et al. described the largest series. They described 8 patients with an RCC who presented with symptomatic hyponatremia, which was postulated to be due to hypocortisolemia caused by inflammatory damage from the RCC within the anterior pituitary gland. Because our patient was on corticosteroid therapy prior to admission to our hospital, it is difficult to say with certainty whether the RCC caused a transient hypocortisolemia from the RCC within the anterior pituitary gland. The resolution of our patient’s hyponatremia, hypothyroidism, and clinical symptoms shortly after decompression and drainage of the RCC provides evidence that an RCC may present with apoplectic symptoms and acute related endocrinopathies including hyponatremia.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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References