CASE REPORT

A CASE OF SYMPTOMATIC RATHKE'S CLEFT CYST IN AN ADOLESCENT GIRL

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Abstract

Rathke's cleft cysts (RCCs) are benign, nonneoplastic sellar and suprasellar lesions believed to arise from remnants of Rathke's pouch. RCCs tend to remain asymptomatic throughout an individual's lifetime and are often discovered incidentally. Rarely, a patient may present with frontal headaches, visual difficulties, or pituitary dysfunction. Most of these cases have been described in middle-aged adults, whereas symptomatic RCCs in children and adolescents are far less frequent. We describe a case of a 17-year-old girl presenting with generalized weakness, bifrontal headaches, and amenorrhea. Her signs and symptoms, attributed to a Rathke's cleft cyst after careful histopathological evaluation, were almost completely resolved although she required three operative procedures within 1 year.

Keywords

Rathke's cleft cyst, headaches, pituitary, amenorrhea, endocrine dysfunction

Introduction

Rathke's cleft cysts are benign epithelial lesions of the sellar and suprasellar region that arise from remnants of Rathke's pouch, (1) which forms at 3 to 4 weeks' gestation. (2,3) Although most remain clinically silent throughout an individual's lifetime, these cysts occasionally become symptomatic by enlarging and compressing surrounding neurovascular structures. Most symptomatic patients experience bifrontal headaches, visual disturbances, and endocrinopathies. (4) Only a handful of cases described in the English language literature report similar manifestations in adolescents. (2,4-6) We report a case of symptomatic Rathke's cleft cyst in an adolescent girl and shed light upon the clinical and radiologic features unique to this case. Of particular interest is the uncharacteristic rapid reaccumulation of cyst fluid in this patient and the necessity for three operative procedures within 1 year before her symptoms were relieved.

Case Report

An obese 17-year-old girl presented with reports of excessive fatigue, amenorrhea, and bifrontal headaches occurring over the past several months. Her developmental and family history was otherwise unremarkable. Magnetic resonance imaging (MRI) of the brain revealed a large, thin-walled cyst expanding the sella and abutting, but not displacing, the optic chiasm (Figure 1). An endoscopic transnasal approach with image guidance was used in order to access and drain the lesion. Pathologic analysis revealed a cyst wall lined by benign columnar ciliated cells compatible with a Rathke's cleft cyst, and cytologic analysis of the cyst fluid indicated areas of previous hemorrhage. After the initial surgery, the patient's headaches improved. However, she still reported feeling fatigued. A repeat MRI 3 months postoperatively demonstrated a persistent cyst. A repeat endoscopic transnasal operative procedure was performed including sphenoidotomy and drainage of the cyst with image guidance, but it was not possible to fully marsupialize the cyst. After the second drainage, the patient reported compressive ophthalmologic symptoms, particularly episodes of blurry vision predominantly in the left eye. A decision was made to perform a definitive drainage procedure with the aid of neurosurgery (Figure 2). We approached the lesion via a transphenoidal endocopic, four-handed surgeon technique. The posterior nasal septum was removed with a 2mm diamond bur, and the mucosa on the rostrum of the sphenoid sinus was elevated until each sphenoid ostium was visible. Then, the rostrum itself was drilled with the same diamond bur until the mass was fully exposed and bilateral access was obtained. The surgery was uncomplicated and her postoperative recovery period was uneventful. She never developed diabetes insipidus nor had any signs suggesting a cerebrospinal fluid leak. After this third drainage, the patient reported an improvement in symptoms. At her follow-up visits as recently as 30 months after her latest operative procedure, she reported intermittent headaches, but her vision remained stable as per her ophthalmologist. In addition, there has been no radiologic evidence of recurrence of the cyst.

Figure 1: Preoperative coronal T2-weighted MRI showing hyperintense sellar mass in an adolescent girl with a Rathke's cleft cyst.

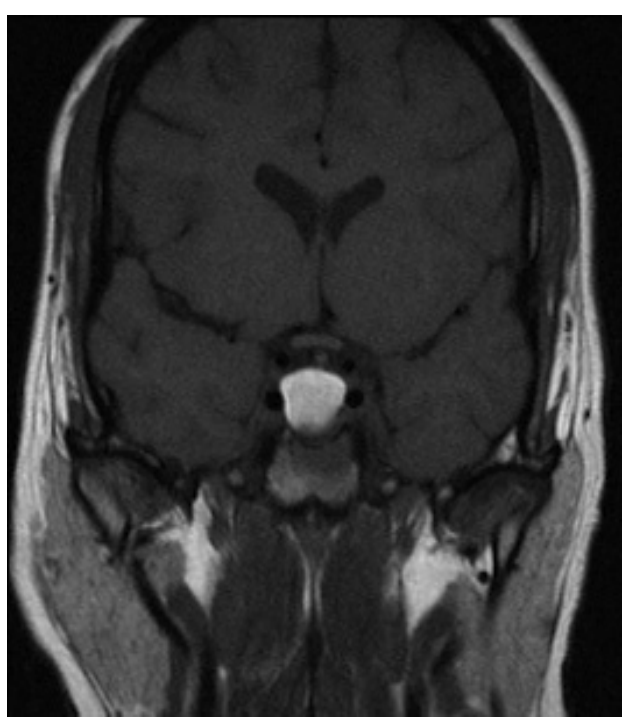
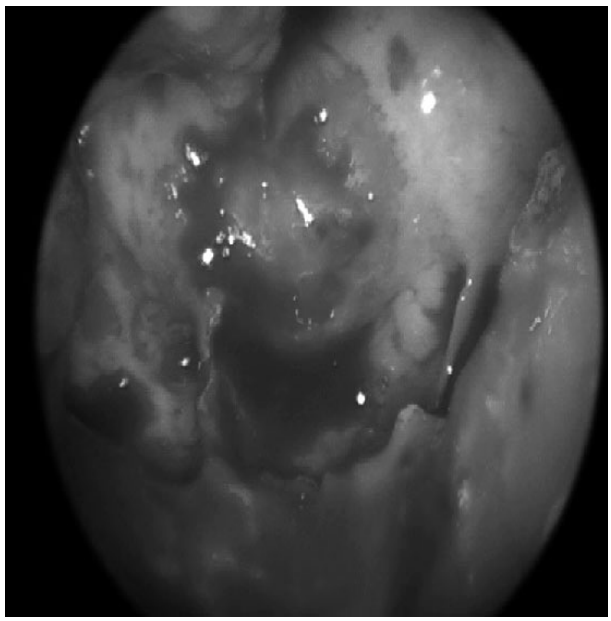


Figure 2: Endoscopic transsphenoidal view of marsupialized sellar mass in an adolescent girl with a Rathke's cleft cyst



Discussion

Rathke's cleft cysts are believed to arise from remnants of Rathke's pouch, a dorsal invagination of the stomodeal ectoderm. (7) With development, the pouch proliferates to form the pituitary gland, pars tuberalis, and pars intermedia. The lumen of the pituitary is normally obliterated by epithelial infoldings, but retention of this lumen forms a Rathke's cleft cyst. (2,3) Commonly discovered as an incidental finding, RCCs have been reported as occurring in up to 22% of normal autopsies. (1) Most, however, tend to remain asymptomatic during an individual's lifetime. (8)

Rarely, symptomatic Rathke's clefts cysts occur, and these typically manifest with episodic headaches, visual deficits, and endocrine dysfunction. (1,7-9) The majority of these symptomatic cysts occur in middle-aged adults whereas adolescent cases are much rarer. Since the initial description of RCC by Goldzieber in 1913, only a small percentage of subsequent reports have described cases in adolescents. (5) The clinical presentation of symptomatic cysts differs between adults and adolescents, with diabetes insipidus occurring more frequently and visual disturbances less frequently in the younger population. (6) Our patient, in contrast, developed no signs or symptoms suggesting diabetes insipidus, and her initial predominant symptom was weakness.

Surgical intervention is crucial for symptomatic RCCs to alleviate compression of and prevent further damage to surrounding neurovascular structures. (3,10) Replacing traditional craniotomies, the transsphenoidal approach has now become the method of choice for these lesions because it is less invasive and has lower rates of postoperative complications. (11,12) However, intractable cases and reaccumulation have been reported. (3,8) Ogawa et al (3) reported that eight of their 155 patients required reoperation owing to fluid reaccumulation and neurologic deficit, with none requiring a third operative intervention. Raper and Besser (8) retrospectively reviewed 12 patients with RCC at their institution. Only one patient required a total of three operations for a particularly aggressive lesion; however, this was over a 7-year span. In contrast, our patient needed three operative procedures within 1 year before her symptoms stabilized.

Recurrent RCCs were once thought to be rare entities, with a recurrence rate estimated to be 5%. (8) Mukherjee et al (9) and Kim et al (4) found that recurrences may be more common than previously thought. Recurrences may be identified more often when the case is followed up aggressively with routine MRI, or in some instances recurrences may reflect cyst fluid reaccumulation caused by incomplete surgical removal.

Conclusion

Due to their indolent nature, histopathologically confirmed Rathke's cleft cysts are rarely symptomatic in the pediatric population, and therefore are rarely discovered. For those lesions which become symptomatic, early surgical intervention is key. We emphasize the importance of a thorough and complete cyst evacuation and cyst wall excision to prevent reaccumulation, and stress routine, post-operative MRI to monitor for reoccurrence in this population.

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Conflict of Interest : None

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