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A Rare Cause of Headache: Rathke Cleft Cyst

CASE REPORT

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ABSTRACT

Headache is the most common symptom encountered in clinical practice. Although the causes of primary headaches are common, intracranial space-occupying lesions also cause headaches, albeit more rarely. Rathke cleft cyst (RCC) is a rare, usually asymptomatic cystic lesion of the sellar/suprasellar region. Despite being benign in nature, these should be immediately treated in case they display compressive signs. We report a case of a 36-year-old man with big-sized RCC diagnosed after presenting with a 3-month history of headache in the light of the existing literature.

Keywords: Rathke cleft cyst, headache

INTRODUCTION

Headache is the most common symptom encountered in clinical practice. Although causes of most primary headaches are common, intracranial space-occupying lesions can also cause headaches, albeit more rarely. Rathke cleft cyst (RCC) is a rare, usually asymptomatic cystic lesion of the sellar/suprasellar region originating from the embryonic remnants of the Rathke cleft. Most of these lesions are incidentally detected upon imaging or during autopsy series (1). Most of the symptomatic lesions have a diameter of 5–15 mm and rarely reach a larger size. Despite being benign, these lesions should be immediately treated in the case of compressive symptoms. In this report, a case of RCC diagnosed in a 36-year-old man with a 3-month history of headache is discussed in light of existing literature.

CASE REPORT

A 36-year-old man presented to our hospital with a 3-month history of moderate-to-severe headache. The patient occasionally felt pressure around the head and sometimes a boring pain as well. Although his headache was usually progressive, the patient reported that the headache decreased without any treatment for 3 days in the second month. His headache was occasionally aggravated by Valsalva-like maneuvers and was not treated with analgesic medical therapy. The patient had no additional neurological symptoms or a history of any systemic disease.

Following a confrontation visual field testing, he was diagnosed with bitemporal visual field loss. Neurological examination revealed that motor, sensory, and cerebellar functions were normal. There was no pathological reflex. He had normal blood biochemistry and hemogram parameters. The magnetic resonance imaging (MRI) of the brain revealed a 31×25×22 mm cystic mass lesion with contrast enhancement that filled the sella turcica and appeared isointense with cerebrospinal fluid in the T1A and T2A sequences and hyperintense in fluid attenuation inversion recovery (FLAIR) sequences and showed contrast enhancement in postcontrast images (Figures 1). In addition, a pituitary MRI showed a dilated sella and a cystic mass with dense content and suprasellar extension that obliterated the cistern.

A detailed patient medical history revealed reduced libido and impotency but no galactorrhea, acromegaly, or polyuria-polydipsia. Hormonal tests showed a prolactin level of 300 ng/mL (normal range: 2.5–17 ng/mL). He was referred for a surgery; the cyst was drained via a trans-sphenoidal approach, and a yellowish mass that was difficult to aspirate was removed. A pathological examination confirmed RCC. The patient experienced no postoperative complication. His headache, visual impairment, and impotency regressed after 1 month. Written informed consent obtained from the patient.

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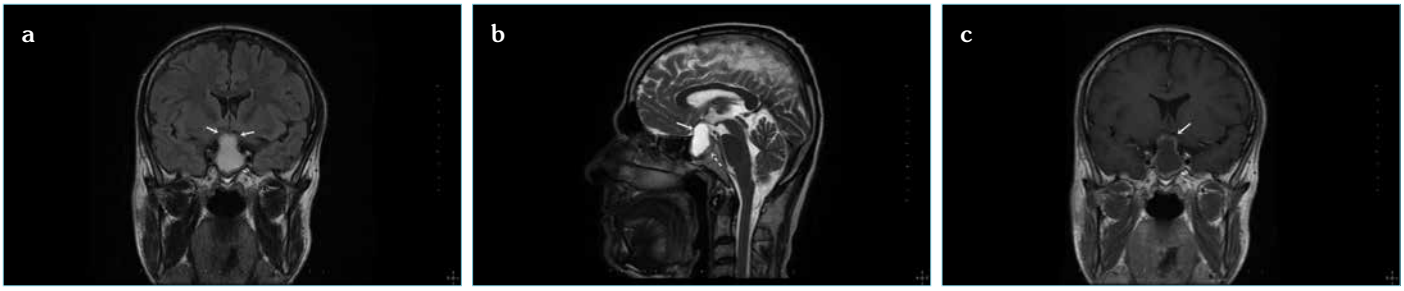

Figure 1. a-c. (a) Coronal in fluid attenuation inversion recovery (FLAIR) sequences weighted MR images showed hyperintense lesion that filled the sella turcica (arrows). (b) Saggital T2-weighted MR images show a 31×25×22 mm cystic mass lesion which appeared isointense with cerebrospinal fluid in the T2A sequences (arrow) and pressed pituitary gland (striped arrow). (c) Coronal T1-weighted MRI with contrast enhancement that appeared isointense with cerebrospinal fluid and showed contrast enhancement in postcontrast images (arrow).

DISCUSSION

Rathke cleft cysts are benign cysts seen in the sellar and suprasellar regions. They originate from the remnants of the Rathke cleft and are covered by epithelial cells. The cells in the anterior wall of the Rathke cleft proliferate and form the distal part of the pituitary gland, whereas the cells in the posterior wall form the intermediate part. According to a theory, they are formed due to an injury of the Rathke cleft that is secondary to rapid cell proliferation and increased secretion (2). Although they are mostly small mass lesions, Siveraju et al reported huge and multicompartmental lesions (3). In our patient, the cyst size was 31×25×22 mm.

These lesions usually appear in the fourth and fifth decades of lifetime and are more common in women (4, 5). They are usually asymptomatic. The symptomatic lesions frequently cause headaches, visual field defects, and endocrinological disorders. Abscess, asymptomatic meningitis, empty sella, and sphenoid sinusitis may also be encountered. (1, 6, 7). Headache is the most common symptom and is a reflection of increased intracranial pressure and dural tension; it was the most common symptom seen in eight patients of a 14-patient study (8). In another study, headache was present in three of nine patients (9). Data suggest a symptom duration of 2 months–10 years before diagnosis (9).

The patient presented to our hospital with headaches for 3 months. The headache was usually progressive, and a Valsalva-like maneuver occasionally aggravated it. According to The International Classification of Headache Disorders, the patient's headache was attributed to an intracranial tumor, although some features were fluctuant.

Visual loss occurs due to optic chiasm compression represents one of the most common symptoms. Kim et al. (8) studied 14 patients and found no visual loss, whereas the rate of visual loss was 22% in another study (9). Our patient had bitemporal visual loss.

About one-half of patients experience hormonal disturbances. Elevated prolactin and reduced growth hormone levels are the most common hormonal disturbances, followed by gonadotropin deficiency, panhypopituitarism, hypothyroidism, and hypocortisolism (5). One study found endocrinological symptoms in 44% of the patients and biochemical/hormonal disturbance in 77% (9). Our patient had hyperprolactinemia and associated libido loss.

Rathke cleft cyst (RCC) display varying positions and density signals in MRI. Depending on the serous, mucinous, or mixed cyst content, they appear hyper, hypo, or isointense, respectively. They appear hyperintense in FLAIR sequences, but they show no internal contrast enhancement in enhanced T1A sequences. Similarly, in our patient, the lesion appeared isointense on T1 and T2 sequences, and only the rim of the cyst was enhanced in the T1 images following contrast uptake.

The differential diagnosis of these lesions includes craniopharyngioma, cystic pituitary adenoma, colloid cyst, arachnoid cyst, and epidermoid cyst. They show similarities to craniopharyngiomas with respect to the origin and localization. The distinction between the two is made by observing ciliated epithelium in RCC by a pathological examination. Cystic pituitary adenomas appear heterogeneous in intensity in MRI. Colloid cysts are seen in older patients and present hydrocephalus associated with an obstructed foramen of Monro.

These lesions are usually classified as benign cysts. However, they may also lead to anterior pituitary functional disorders, extrasellar enlargement, and visual loss (10). They must be immediately treated when they cause the aforementioned lesions. In a recent study, spontaneous involution was detected in patients with large masses that had optic nerve contact but no visual disturbances (8).

As the patient's lesion caused visual disturbance, we referred him for a surgery. The surgical treatment of RCC includes radical surgery or trans-sphenoidal cyst drainage, which has been reported adequate with low rates of postoperative endocrinological and neurological disorders (9). Our patient underwent trans-sphenoidal cyst drainage, and his symptoms regressed following the operation. The rates for postoperative regression and recurrence were reported to be 78% and 19%–28%, respectively (10).

In our patient, RCC was the underlying etiology of headache, which is common in neurology practice. It is important to diagnose this lesion at an early stage and refer an appropriate treatment to reduce morbidity and mortality.

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