A rare case of symptomatic Rathke's cleft cyst in a 26-year-old female is reported. The clinical and radiological features of this lesion are described with emphasis on the differential diagnosis between Rathke's cleft cyst and craniopharyngioma.

Rathke's cleft cysts present a common incidental intrasellar autopsy finding occurring in 13% to 22% of randomly examined pituitary glands [1]. These lesions rarely, however, enlarge sufficiently enough to cause compression of adjacent neural structures. Although to date, approximately 155 cases of symptomatic Rathke's cleft cysts have been reported in the pertinent literature [2]; the condition continues to be largely unknown to clinicians.

The purpose of this case report is to draw attention to this pathological entity which should be considered in the differential diagnosis of cystic lesions in the sellar area. To the best of our knowledge, this is the first case of symptomatic Rathke's cleft cyst to be reported from Saudi Arabia.

Case Report

This 26-year-old Filipino female dietitian presented to the Neurosurgical Division of the King Khalid University Hospital with a two month history of constant headaches which were associated with nausea and non-projectile vomiting. There were no visual disturbances and apart from oligomenorrhea since puberty, her past medical history was unremarkable.

Examination

The patient was short in stature (height 140 cm and weighed 37.5 kg). Skin and hair distribution were normal. Neurological status of the patient was normal except for a bi-temporal hemianopia.

The plain x-ray of the skull revealed erosion of the dorsum sellae with no sellar abnormality. Cranial computerized tomographic (CT) scan showed a large low density lesion measuring 30×39 mm with faint ring enhancement filling the entire suprasellar space with compression of the third ventricle (Figure 1). The magnetic resonance imaging (MRI) demonstrated the lesion extending into the sella and elevating the floor of the third ventricle. The lesion was hyperintense on both T1 and T2 weighted images except for a small nidus which could have represented a solid component of the lesion. The vertebral angiography disclosed marked dorsal displacement of the distal end of the basilar artery.

The basal secretion of the following hormones was normal: T3, T4, TSH, cortisol, prolactin, LH, FSH, and growth hormone. The pituitary dynamic tests after injection of insulin, TRH, and GnRH revealed inadequate GH and FSH responses.

Operation

The patient underwent a right frontotemporal craniotomy. A thin-walled, brown-greenish cyst was encountered. The right optic nerve was stretched over the cyst wall and the right carotid artery was displaced laterally. The cyst was punctured and 35 cc of milky greenish fluid was aspirated. The inferior part of the cyst wall was excised leaving the superior part, which was firmly attached to the floor of the third ventricle, unre moved.

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Postoperative Course

The patient recovered well but developed transient diabetes insipidus which was controlled with vasopressin. Two years following surgery, the patient remains asymptomatic except for irregular menstruations. The basal secretion of the pituitary hormones is normal and her visual fields have normalized. The cranial CT scan was normal (Figure 2).

Pathological Examination

The histopathological examination of the excised cyst wall showed loosely arranged fibrous connective tissue covered on one side by a layer of ciliated cuboidal to columnar epithelium in some areas and flattened cells in others (Figure 3). A few single goblet cells that showed strong staining by alcian blue were scattered among the cuboidal epithelium. No areas of hemorrhage or hemosiderin deposits were observed.

Discussion

The exact origin of Rathke's cleft cyst is unknown. The most popular hypothesis is that it derives from remnants of the Rathke's pouch, a superior diverticulum of the stomodeum [3-5]. Alternatively, it has been suggested that Rathke's cleft cyst is a neuroepithelial derivative [6] or develops by reverse metaplasia from anterior pituitary cells [7]. Based on the demonstration of amyloid stroma in a Rathke's cleft cyst, a neural crest origin was also proposed [8]. On the other hand, electron microscopic evidence of a coating material surrounding cells with microvilli raised the possibility that Rathke's cleft cyst could present endodermal inclusions from the upper respiratory tract [5,9].

The majority of Rathke's cleft cysts remain asymptomatic throughout life. The reason why only a small number of cysts progressively enlarge until they become symptomatic remains obscure. Mucous secretion and hemorrhage into the cyst and inflammatory reactions (septic or aseptic) in the cyst wall may contribute to cyst enlargement [5,10,11].

Most of the symptomatic Rathke's cleft cysts present between the third and sixth decades of life. Females are doubly more affected than males [2]. The clinical symptoms produced by Rathke's cleft cyst are almost uniformly a result of compression of the pituitary and/or suprasellar structures. Decreased visual acuity, visual field defects, hypopituitarism, diabetes insipidus, and headaches are the main manifestations [2,3,5].

Occasionally, Rathke's cleft cyst may present...
similar to a craniopharyngioma with aseptic meningitis with pyrexia and impaired consciousness [12,13]. Obstructive hydrocephalus and bacterial abscess formation in a Rathke’s cleft cyst have also been reported in a few cases [14,15].

The diagnosis of Rathke’s cleft cyst may be suspected in cases with a CT scan showing low density intra- and suprasellar lesions with no calcification and either no enhancement or a ring-like enhancement following injection of contrast material [10,16]. A Rathke’s cleft cyst rarely presents with higher or mixed attenuation values with or without calcification [2]. The MRI features of Rathke’s cleft cysts are also unspecific and depend on the content of the cyst [11,16]. Cysts containing CSF-like fluid appear hypointense on the T1-weighted image (T1-WI) and hyperintense on the T2-weighted image (T2-WI). Hyperintensity on both T1-WI and T2-WI, as in our patient, has been considered suggestive of the presence of blood [11]. Nevertheless, the histopathological examination of the cyst wall in our case did not reveal any evidence of old or recent bleeding into the cyst.

Rathke’s cleft cyst is typically lined by ciliated cuboidal or columnar epithelium and, therefore, is quite distinct from craniopharyngioma which consists of squamous epithelium [3-6,17]. However, cysts with mixed histological features of Rathke’s cleft cysts and craniopharyngioma have been described [5,9,12,18]. Such observations suggest that Rathke’s cleft cysts and craniopharyngioma may be closely related and share a common cellular origin. However, some authors attributed the presence of squamous epithelium in some Rathke’s cleft cysts to metaplasia from the columnar epithelium [19]. Rathke’s cleft cyst is considered to be a more benign lesion than craniopharyngioma. While complete removal or partial resection followed by radiotherapy presents standard treatment of craniopharyngioma, Rathke’s cleft cysts may be managed adequately by simple aspiration of the fluid [4,20,21]. However, to avoid recurrence [5,22] evacuation of the cyst and liberal resection of the cyst wall are recommended [23]. Intrarassellar cysts with or without suprasellar extension are best approached transphenoidally while cysts which are mainly suprasellar such as our case usually require a transcranial approach [2]. Although postoperative radiotherapy is generally considered to be unnecessary, it has been employed in a few cases [12].

References