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Rathke's Cleft Cyst and Panhypopituitarism: Two Cases Report

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Abstract: Rathke's cleft cysts are benign, usually asymptomatic cystic epithelial congenital lesions. The symptomatic forms are exceptional with endocrine manifestations due to hypophyseal compression by the cyst. We present two cases of Rathke's cleft cyst revealed by panhypopituitarism. The prognosis for the final height and fertility of these patients remains the major concern. The early management of these cystic lesions of benign appearance remains crucial.
Keywords: Rathke's cleft cysts, Adenohypophyseal deficiency, Late diagnosis, Prognosis.

INTRODUCTION

The Rathke's cleft cysts sellar lesions are frequently discovered accidentally. These are benign epithelial cystic lesions, most often asymptomatic [1]. However, the symptomatic forms exist, although rare and are related to bulky cysts, causing endocrine syndromeandan intracranial hypertension syndrome. These multiple pituitary deficits are defined by insufficient secretion of at least two anterior pituitary hormones, most often linked to a somatotropic deficit with another gonadotropic or thyrotropic but all associations are possible [2]. We report the cases of two young patients with large Rathke's cleft cysts of insidious evolution withpanhypopituitarism which was diagnosed late.

CASE REPORT

The first patient is 28 years old, complaining of intense frontal headache with visual disturbances, it was worsening slowly and progressively, during 9 years, and was treated as migraines. A cerebral MRI performed in 2013 revealed suprasellar cystic lesion measuring 20 mm in diameter, suggesting Rathke's cleft cysts with stable appearance of the lesion over the past three years. The clinical and hormonal preoperative exploration found signs of thyrotropic and corticotropic insufficiency with asthenia, hypotension and hypoglycemia;on clinical examination, patient had a small size (1, 50m); webbed neck; bilateral adipomastia, external genital organs corresponding to stage G3P4of Tanner. In front of the persistence and intensity of headaches surgery is indicated. The hormonal assessment revealed: a corticotropic deficit with a plasma cortisol of 8h at 6.6µg / dl (N: 62-194µg / L); thyrotropic insufficiency with normal TSH at 0.3mIU / L (N: 0.27-4.20pmol / L) and T4L low at 5pmol / l (N: 12-22pmol / L); as well as gonadotropic insufficiency with testosterone at 0.025 ng / ml and FSH at 0.59 IU / l. The morphological assessment including a pituitary MRI showed an intrasellar and suprasellar cyst of 20 mm in diameter, fills the chiasmatic cistern, comes into contact with the floor of the 3rd ventricle and internal carotids, squeeze the pituitary gland, brainstem and basilartrunk. The visual field was slightly altered in right and left eye with early appearance of chiasmatic lesions. Practical management consisted of the substitution of corticotropic and thyrotropic axes. The patient was operated on surgically with as postoperative complications a polyuro-plydipsic syndrome treated with ADH. The histological examination showed a Rathke's cleft cysts. Postoperative evolution noted the persistence of corticotropic and thyrotropic deficits linked to poor therapeutic compliance. Subsequent management of the gonadotropic deficiency was encouraged.

The second case is that of an 18-year-old, referred by neurosurgery for preoperative hormonal evaluation of a sellar cyst revealed by severe stunting and delayed puberty. History take noted a correct birth weight and psychomotor development, but a decline in the growth curve since the age of 12. The patient didn't mention any tumoral syndrome, in particular no headaches or visual disturbances, no signs in favor of anterior pituitary hyposecretion. On medical examination, a face without dysmorphic syndrome, a height of 1.47m (- 4DS) and a weight of 38kg at (- 3SD) for age, mild bilateral adipomastia, stage TANNER G1P1, without other clinical features. The hormonal assessment revealed thyrotropic deficit with a T4L at 10 pmol / L (N: 12-22) and a normal TSH at 3.12 mIU / L, gonadotropic deficiency with testosterone at: 0.025 ng / ml (2,49-8,36), FSH: 1.2 IU / L (N: 10.9-13.9) and LH: 1.2 IU / L (N: 6.9-10.3) as well as a somatotropic

deficit with GH <20 IU / L after stimulation; in addition, an 8h cortisolemia of 11.59µg / Dl (62-194 µg / L) and a prolactinemia of 12.15ng / ml. Metabolically, the lipid and phospho-calcium level was correct. Pituitary MRI revealed compatible image with a choristoma or cyst 18x15mm remodeled atRathkeclift. His bone age corresponding to 12 years for a chronological age at 18 years and 8 months. Goldman's visual field was slightly altered in right and left eye; with widening of the 2 blind spots, peripheral localized deficit in right and left eye (supero- temporal and super-nasal of the right eye in the supero-temporal and infero-nasal of the left eye), thus a relative chiasmatic lesion. Medical management consisted initially ofthyrotropic deficit correction. Afterward, it has been encouraged to correctsomatotropic and gonadotropic deficits. The patient was operated transsphenoidally with postoperative complications ofpolyuro-polydipsic syndrome and treated with ADH. Histological examination revealed an aspect of Rathke's cleft cysts a neuroepithelial type.

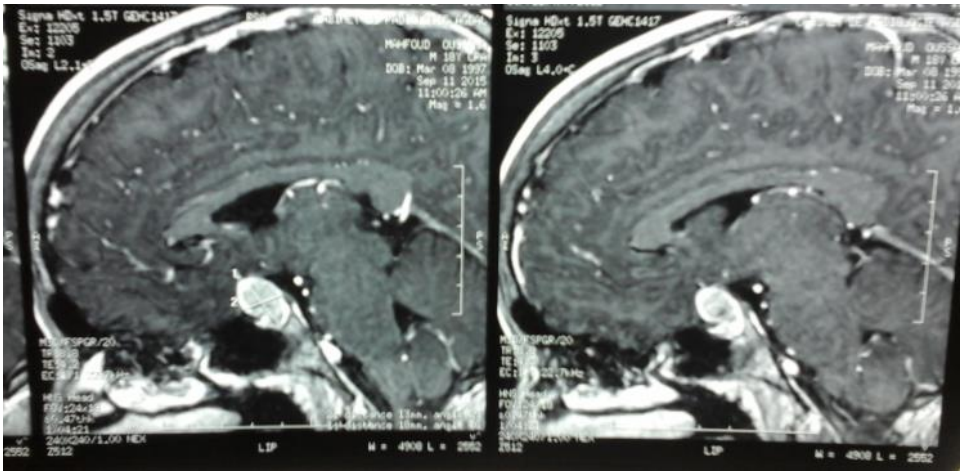

Fig-1: Pituitary image of a Rathke's cleft cysts

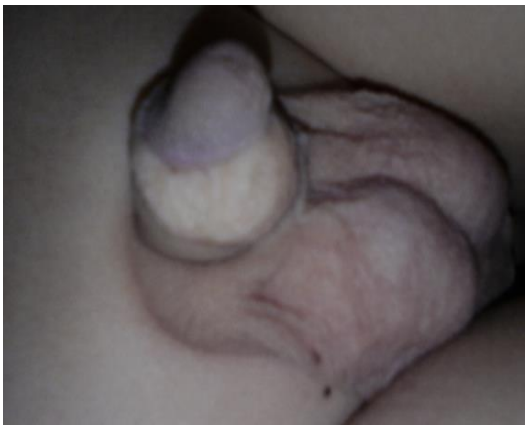

Fig-2: EGO image illustrating pubertal delay in an 18-year-old patient

DISCUSSION

The Rathke's cleft cysts are lesions developed from the epithelial fragments of Rathke's pouch. This is a pouch formed from an outgrowth of the stromodeum during the third or fourth week of embryonic life. The anterior wall of the pouch is the origin of the anterior lobe of the pituitary gland and the posterior wall forms the intermediate portion [3-5]. The residual opening between these two parts closes most often, but can sometimes persist and give a cleft lined with stratified epithelium that can dilate by accumulation of secretions, which will lead to the formation of a cystic lesion from which the cyst name of Rathke's pouch [1,3]. It is basicallya benign lesion, usually asymptomatic therefore its discovery is often accidental. On the other hand, bulky cysts, averaging between 10 and 20 mm, are most oftensymptomatic, intra and supra-sellarapplying mass effect on neighboring structures [4,5]. Among the clinical manifestations of Rathke's cleft cysts, endocrine disorders account for 50% [1,2,4]. A panhypopituitarism found in one of our patients and multiple combined anterior pituitary deficiencies observed in the other, including a somatotropic, gonadotropic and thyrotropic deficit. The common point in these two patients is the diagnostic delay, at the respective ages of 28 years and 18 years. Most patients improve symptoms such as headache and visual disturbances, but hormonal disorders have rarely

returned to normal in patients with severe preoperative hormone deficiency [2,4]. Pituitary MRI remains the test of choice for the Rathke's cleft cysts discovery. The symptomatic forms most often correspond to lesions with suprasellar extension, the differential diagnosis with a craniopharyngiomacystbased on the study of contrast enhancement of the cystic wall after injection of gadolinium. Contrast enhancement is generally not observed because of the unicellular character of the wall of Rathke's cleft cysts[5-7]. Surgical treatment is indicated in symptomatic forms, in spite of possible spontaneous decrease of volume of these cystic lesions. The endonasaltranssphenoidal endoscopic approach remains the usual approach in the surgical management of pituitary and parasellar tumors [1,8-10], except in the case of large cyst with suprasellar extension, frontotemporal craniotomy is preferred, as was the case of our patient. However, postoperative cyst recurrence is not uncommonly reported in 10 to 33% of cases and occurs on average 2 years after surgery [1,11, 12]. Endocrine dysfunction remains one of the major concerns regarding the prognosis of final body size and fertility, but also the quality of life. Monitoring and adaptation of replacement therapy remains problematic in these patients.

CONCLUSION

The Rathke's cleft cystsare frequent benign lesions of the sellar region. However, symptomatic forms may appear as endocrine disorders including panhypopituitarism. These pituitary deficits have consequences on the quality of life and the prognosis of the final body size and fertility. Reactive inflammation of the normal pituitary gland may have a role in the pathogenesis of hormonal deficiency, in addition to compressing effect. The early management of these cystic lesions of benign appearance remains crucial.

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