PANHYPOPITUITARISM APPARENTLY CAUSED BY HYPOPHYSISITIS MASKING A RAPID DEVELOPMENT OF A CRANIOPHARYNGIOMA.
A CASE REPORT.

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INTRODUCTION

We report here the unusual medical history of a male subject presenting with panhypopituitarism presumably due to hypophysitis as suggested by MRI, but who a few months later developed a visual field defect due to a large cystic lesion then suggestive of Rathke's cleft cyst on MRI. The final pathological diagnosis was craniopharyngioma complicated by a Propionibacterium acnes infection.

CASE REPORT

A 39-year-old male subject was referred with complaints of polydipsia and polyuria for three months. He also complained of headache, weight gain, loss of libido and tiredness. Of interest was the personal medical history of Graves' hyperthyroidism. Endocrine assessment confirmed the diagnosis of diabetes insipidus. Hypopituitarism was present with low-normal to decreased serum levels of free T3 (3.6 pmol/L; normal 3.1-6.8), free T4 (6.8 pmol/L; normal 12-22), TSH (1.1 mU/L; normal 0.3-4.2), testosterone (0.6 nmol/L; normal 10-28), LH (<0.1 U/L; normal 2-10) FSH (0.9 U/L; normal 2-10), IGF-I (130 μg/L; normal 200-250) and an inadequate response to a insulin tolerance test for cortisol (peak value of 125 nmol/L, normal >180 nmol/l) and for GH (peak value of 0.3 μg/L, normal > 3 μg/l). Coronal and sagittal T1-weighted MRI showed a focal thickening of the pituitary infundibulum as well as infiltration of the anterior pituitary lobe with
of squamous, non-dysplastic epithelium, with a limited amount of chronic inflammatory cells. Keratinisation was focally present. No distinctive architectural pattern could be discerned and no pituitary tissue was identified.

Two weeks later, the subject again developed headaches and visual disturbances. MRI confirmed the presence of a similar sellar mass with unchanged characteristics. Because of bacterial surinfection, antibiotics were given for one month. The headaches diminished but shrinkage of the pituitary mass and correction of the visual field defects were not observed. A second neurosurgical exploration through a subfrontal approach was successful in decompressing the optic chiasm. Pathological examination of the resection specimen confirmed the diagnosis of craniopharyngioma.

DISCUSSION

In this case of craniopharyngioma the straightforward diagnosis and adequate therapy have been obscured by the initial aspecific radiological images. The original presentation of hypopituitarism and diabetes insipidus was explained by the findings on MRI, showing focal thickening of the pituitary infundibulum as well as infiltration of the anterior lobe of the pituitary. A lesion leading to both anterior and posterior deficiency in association with an autoimmune thyroid disease made the diagnosis of lymphocytic panhypophysitis very likely (1). Secondary forms of hypophysitis also have been described, usually due to systemic diseases such as sarcoidosis or tuberculosis. Local lesions such as Rathke’s cleft cyst or craniopharyngiomas have rarely been associated with secondary forms of hypophysitis and in these cases the lymphocytic or granulomatous infiltrate focuses around the lesion rather than to be diffuse. A pituitary abscess usually presents on MRI as a cystic lesion with peripheral ring enhancement after gadolinium administration, but a thickening of the pituitary stalk, as in this case, may also occur and suggests an infiltrative rather than an infectious process (2). The most common pathogenic organism is a coagulase-negative staphylococcus, while Aspergillus fumigatus and Escherichia coli are less frequent. Propionibacterium acnes has been recognized as a cause of post-neurosurgical infection, but until now it has never been reported outside a surgical procedure (3). Its presence as a causative agent for the radiological image of hypophysitis remains therefore doubtful, also since no adjacent infection could be observed.
The subsequent presentation of the subject suggested an acute development of a large Rathke's cleft cyst, eventually initiated due to bacterial contamination despite the absence of an infectious syndrome. Rathke's cleft cyst is a common finding at routine autopsy, but rarely evolve into a symptomatic presentation with hypopituitarism and visual field defects. The diverse clinical expression and the lack of radiological distinctiveness make therefore the diagnosis of Rathke's cleft cyst not always evident. An accurate preoperative diagnosis has however surgical importance. The usual management of symptomatic Rathke's cleft cyst is simple surgical drainage with partial excision of the cyst wall. This surgical attitude, also taken by the surgeon in this case, is advocated since recurrence of a Rathke's cleft cyst is rare.

The final pathological diagnosis in this subject rather surprisingly was a papillary craniopharyngioma. While the classical mucinous Rathke's cleft cyst is lined by cuboidal or columnar ciliated epithelium, the identifying mark of a craniopharyngioma is the keratinisation of individual cells or the presence of nodules of keratinized cells (4). The clinical presentation and the hormonal deficit in craniopharyngiomas do not allow to separate this entity from other sellar and parasellar lesions. The MRI appearance of craniopharyngiomas depends on the proportion of solid and cystic components and of its content, but, although the radiological image maybe very suggestive, specific characteristics do not exist. In contrast to Rathke's cleft cyst, the present surgical option is to remove the whole craniopharyngioma cyst in order to avoid recurrence, but without destroying neighbourhood structures.

In conclusion, the main interest of this case is its unusual radiological presentation. We can assume that the radiological image of hypophysitis was the first expression of the development of a craniopharyngioma, inducing local irritation and leading to panhypopituitarism. It is therefore important to perform a control MRI after a reasonable time in case of a hypophysitis treated conservatively. Another point of interest is the extremely rapid development of a cystic craniopharyngioma, leading to visual field defects within eight months. A similar phenomenon of accelerated growth has been reported during pregnancies, and in a 55 year old woman with a normal MRI two years before (5).

REFERENCES