LYMPHOCYTIC HYPOPHYSITIS, VISUAL FIELD DEFECTS AND HYPOPITUITARISM

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SUMMARY A 45-year-old man presented with anterior pituitary failure, requiring thyroxine, hydrocortisone and androgen replacement. An MRI scan revealed a large cystic pituitary mass and thickening of the pituitary stalk. Over three years, diabetes insipidus and bitemporal hemianopia developed and the cystic mass had enlarged on MR scanning. Transphenoidal resection was performed with normalisation of the visual fields. Histology revealed lymphocytic hypophysitis, which is rare in men. The presentation with cystic enlargement is unique. (Int J Clin Pract 1999; 53(8): 643-644)

Lymphocytic hypophysitis (LH) is rare, usually involving the anterior pituitary gland. It is characterised by massive infiltration of lymphocytes and plasma cells followed by necrosis, macrophage infiltration and surrounding parenchymal fibrosis. Most reported cases are in women during pregnancy and the immediate postpartum period, with few in postmenopausal women and in men. Patients usually present with varying degrees of hypopituitarism.

CASE REPORT
A 45-year-old caucasian male presented with a six-month history of weight loss, weakness, lethargy, reduced libido and cold intolerance. There was no family history of autoimmune or endocrine disorders. He had reduced pubic and axillary hair and dry skin. Visual fields were full. Investigations showed a normal full blood count and renal function. Basal serum endocrine tests revealed free thyroxine 6 pm/l (normal >9 pm/l); TSH <0.5 mU/l; cortisol 50 nmol/l (low); testosterone 5 nmol/l (normal >8 nmol/l); LH <1 IU/l; FSH <1 IU/l; growth hormone <1 mU/l. Serum prolactin was mildly raised at 700 mU/l.

MR scanning of the sellar region revealed a large mass arising from the pituitary fossa, thickening of the pituitary stalk, peripheral ring enhancement and a cystic appearance. The optic chiasm was not compressed. The diagnosis of panhypopituitarism probably secondary to a non-secreting adenoma of the pituitary gland was made. A conservative management plan was agreed with the patient. However, pituitary exploration was to be considered if the mass enlarged on MRI scan. Thyroxine, hydrocortisone and androgen replacement was started, with marked clinical improvement. Two years later, the patient reported polyuria and polydipsia. A water deprivation test confirmed diabetes insipidus and treatment with desmopressin controlled these symptoms.

During the next year, blurring of vision was reported and the patient developed a partial bitemporal hemianopia. MRI of the sellar region showed further enlargement of the cystic sellar mass with upward displacement of the optic chiasm (Figure 1). At transphenoidal exploration of the pituitary, soft white tissue and a cystic cavity filled with creamy fluid were found. The wall of the cyst and macroscopically abnormal surrounding tissue were removed. No normal anterior pituitary gland was identified at surgery. Paraffin sections revealed anterior pituitary, featuring a dimorphic pattern.

Figure 1. MRI scan showing a cystic mass arising from the pituitary fossa and causing chiasmal compression

Figure 2. Heavy lymphocyte infiltrate in between anterior pituitary glandular structures (arrows)

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643
beside a rim of gland with normal trabecular architecture and viable cells. There was necrotic anterior pituitary tissue with heavy macrophage infiltration, foci of infarction and collaginous necrosis and cavitation. Reticulin staining highlighted areas of preserved glandular architecture but there was collapse of the reticulin network and increased collagen in the areas bordering the necrosis. A heavy mixed T and B lymphocytic infiltrate were present among the surviving glandular elements at the periphery (Figure 2). There was no evidence of a granulomatous or caseating process, neoaplasm, Rathke's pouch remnants, crianiopharyngioma or bacteria. The appearances were those of lymphocytic hypophysitis with foci of necrosis, cavitation and surrounding interstitial reactive fibrosis.

Postoperatively, serum was tested for autoantibodies to the following: nuclear factor, mitochondria, adrenal, gastric parietal, intrinsic factor, thyroid (microsomal and colloid), parathyroid and pituitary. All were negative. The patient's visual fields immediately returned to normal and endocrine tests showed persistence of panhypopituitarism, including diabetes insipidus.

DISCUSSION

The presentation of LH can vary from prolactin hypersecretion, presumed secondary to stalk compression, to complete anterior pituitary hormone failure mimicking Sheehan's syndrome.1 Diabetes insipidus has been reported but is unusual and is termed infundibulo-hypophysitis.2 In rare cases the swollen pituitary gland may extend into the suprasellar region or even the cavernous sinus.3 Clinical and radiological distinction between non-functioning pituitary adenoma and LH is practically impossible, although atypical MRI features such as peripheral enhancement and thickening of the pituitary stalk may suggest LH.4 Our patient did have a thickened pituitary stalk at presentation. Most patients recover spontaneously and a trial of steroid therapy to aid resolution has been advocated.5 Some patients, however, continue with a degree of residual hypopituitarism even though the pituitary has reduced in size. Co-existing autoimmune disorders are described but the exact incidence is not known.6,7 Our patient is of interest because it is the first case of LH described with cystic appearance and homogeneous peripheral thickening on MRI. Also, LH in a male is unusual.

CONCLUSION

LH should be included in the differential diagnosis of a cystic pituitary mass and the diagnosis confirmed by surgical exploration.

REFERENCES