A case of granulomatous hypophysitis with hypopituitarism and minimal pituitary enlargement

PATRICE HASSOUN, ELYAS ANAYSSI, IBRAHIM SALTII

From the Departments of Pathology and Internal Medicine, Faculty of Medicine, American University of Beirut, Beirut, Lebanon

SUMMARY A case of hypopituitarism and minimal sellar enlargement was found at hypophysectomy to have a giant cell granuloma of the pituitary. The clinical and histopathological features of this rare entity are reviewed. It is proposed that hypopituitarism which is out of proportion to minimal sellar enlargement may be a suggestive clue to the preoperative diagnosis of giant cell granulous which normally simulates a pituitary tumour.

Granulomatous diseases of the pituitary are uncommon disorders which are rarely diagnosed in patients presenting for hypophysectomy. The majority of reported cases come from neuropys material and include infectious and systemic diseases such as tuberculosis, sarcoïdosis, syphilis and fungal infections. However, when such specific aetiologies are excluded, there remains a small group of patients in whom the granulomatous disorder is of unknown origin and in whom there is no systemic involvement with the granuloma apparently restricted to the pituitary gland.

In this report we describe a patient who presented with hypopituitarism together with radiological findings of a small intrasellar tumour. The patient was subjected to a trans-sphenoidal hypophysectomy and proved to have a pituitary giant cell granuloma.

Case report

A 65-year-old woman presented with a two months history of anorexia, nausea, vomiting and weight loss of about 10 kg. She had no headaches, visual complaints or abdominal symptoms. Physical examination revealed a sick elderly woman with a dry skin and slowing of speech and mentation. The visual fields, fundoscopic and neurological examinations were normal. The blood pressure was 90/70 mm Hg and heart rate 76/minute. The following tests gave normal results: fasting serum glucose, urea nitrogen, creatinine, albumin, globulin, cholesterol, calcium, phosphate, sodium, potassium and chloride. Urine analysis was normal with a specific gravity of 1028. Haemoglobin was 11 g/dl and haematocrit 34%. The leucocyte count, leucocyte differential count and platelet count were normal. The erythrocyte sedimentation rate was 50 mm/hour. Serum VDRL was nonreactive.

In view of the persistent nausea and vomiting, computed tomography was performed and revealed rarefaction of the dorsum sellae and the lamina dura of the sella turcica. There was a double floor of the sella but the total sellar volume was not increased. There were no abnormalities in the suprasellar region.

Baseline endocrine tests were performed and are summarised in the table. Intravenous administration of 200 μg of thyrotropin releasing hormone (TRH), resulted in no rise in serum prolactin or thyroid stimulating hormone. In view of the clinical state of the patient, an insulin-induced hypoglycaemia test was not done. Serum was negative for antithyroglobulin, antimicrosomal, anti-islet cell and antilactotrope antibodies. An intradermal tuberculin test and chest radiography were negative. Serum was also negative for Brucella antibodies.

Treatment with intravenous hydrocortisone resulted in prompt clinical improvement and disappearance of the vomiting. A transphenoidal hypophysectomy was well tolerated by the patient. Thereafter, the patient was placed on maintenance doses of l-thyroxin and hydrocortisone. On regular bimonthly follow-up for the subsequent two years she was asymptomatic.

Histopathology

The excised pituitary tissue weighed 1.8 g and was firm in
consistency. Microscopic disruption of the adenohypophysis, cells of the hypophysitis, with the sellar abnormalities that were observed on CT scanning, was suggestive of a pituitary tumour. A review of the previously reported cases of giant cell granuloma of the pituitary revealed a similar presentation simulating a pituitary tumour with hypopituitarism and radiological evidence of minimal sellar enlargement. However, a common feature amongst these patients (including the present case) is that unlike pituitary neoplasms, the degree of pituitary failure is out of proportion to the minimal degree of sellar enlargement. A pituitary tumour producing little or no sellar enlargement is not expected to cause hypopituitarism unless it has major supra sellar extension.

Another feature of giant cell granuloma is that the course of the disease is more rapid than in the usual case of pituitary tumour (refs and present case).

The pathogenesis of pituitary giant cell granuloma remains unknown. Although its clinical presentation bears many similarities to the recently described entity of autoimmune lymphocytic hypophysitis, the two disorders are histologically distinguishable. Moreover, in lymphocytic hypophysitis which occurs mostly in postpartum women, there is an association with other autoimmune endocrine and non-endocrine disorders and the patients' sera are sometimes positive for antibodies against pituitary cells or other endocrine cells or antigens. These antibodies were not detected in the serum of our patients.

Despite the obvious histological and immunological differences between granulomatous and lymphocytic hypophysitis, a recent study by Mckee suggest that there are common ultrastructural features between the two entities. The author suggests that the two disorders may simply be different stages of a single disease. This proposition needs further study.

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P Hassoun, E Anayssi and I Salti

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