Case Report

Autoimmune panhypophysitis presenting as pituitary mass

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Abstract

Inflammatory involvement of pituitary has been described as of autoimmune, granulomatous, xanthomatous or secondary forms. Correct histological diagnosis is required for adequate treatment. We herein report a case of lymphocytic panhypophysitis in a middle aged male patient, a known case of seizures with multiple calcified lesions in brain who presented with polyuria & polydipsia. The patient was evaluated and was diagnosed as lymphocytic hypophysitis based on imageological, immunological, histopathological studies.

Key words: lymphocytic hypophysitis, autoimmune, granulomatous, infundibulohypophysitis.

Abbreviations: lymphoadenohypophysitis (LAH), lymphocytic infundibuloneurohypophysitis (LINH), lymphocytic panhypophysitis (LPH), Anti Diuretic Hormone (ADH).

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Case report

This 42-year-old male patient, a known case of primary generalised seizures for past 30 years on carbamazepine, presented with complaints of polyuria, polydipsia, decreased visual acuity, lethargy, headache and erectile dysfunction of 2 months duration. Patient’s visual acuity was reduced to finger movements at one meter distance. Visual fields showed bitemporal hemianopia, and primary optic atrophy. There were no motor or sensory deficits. No signs of meningeal irritation.

Investigations and Radiology

Laboratory investigations revealed low serum TSH, low serum cortisol, low FSH, LH, Testosterone levels, moderately elevated serum prolactin level, normal erythrocyte sedimentation rate, and normal serum electrolytes. Urine specific gravity and osmolality were within normal limits. CSF analysis showed no evidence of meningitis.
Figure 1: FLAIR image, axial sections showing a hyperintense mass lesion in the sella, extending into the suprasellar space, reaching up to the chiasma and also involving the hypothalamus.

Figure 2: Sagittal T2W image showing well defined lesion in the sella distinct from the pituitary (which is located inferiorly), and extending to the suprasellar region.
Management

Patient was treated with hydrocortisone and replacement of thyroid hormones. Pterional craniotomy and subfrontal approach to the suprasellar region showed bulge of the diaphragm sella which on opening showed grayish granulation tissue. Stalk was thickened. Subtotal resection of the sellar mass and decompression of chiasm performed. Histopathological examination of the specimen showed destruction of normal pituitary parenchyma with areas of fibrosis with dense lympho-mononuclear infiltrate. Definite lymphoid follicles not identified. There was no evidence of granulomas (Figure 4).

Patient was started on prednisolone 1mg/kg/day along with desmopressin(20 micrograms twice daily), and replacement of thyroxine (100microgram/day). Patient had significant improvement in vision, urine output and thirst. Serum prolactin showed significant decrease with elevation of serum cortisol and thyroxin levels. Evaluation for secondary causes of hypophysitis like c-ANCA, serum calcium, VDRL, TPHA, Sputum for AFB, Mantoux were negative.

The patient is under follow-up for past three years and is doing well on hormone replacement.

Figure 3: Contrast enhanced T1W images in sagittal plane showing uniformly enhancing lesion in the sella, suprasellar space and extending along the optic tracts and into the hypothalamus

Figure 4: Histopathological examination of the specimen shows destruction of normal pituitary parenchyma with areas of fibrosis and dense lympho-mononuclear infiltrate with no evidence of lymphoid follicles or granulomas.
Discussion

Granulomatous hypophysitis is a rare entity was first described in 1917 by Simmonds (1). The first antemortem case was reported in 1980 (2). Pathologically hypophysitis is broadly classified as primary or secondary. Primary is further sub classified as autoimmune (lymphocytic), granulomatous, and xanthomatous (3). The term “secondary hypophysitis” indicates a pituitary inflammation that originates from neighboring lesions or is part of systemic disease and focuses around the lesion rather than diffusing to the entire gland.

Histological classification of lymphocytic hypophysitis is described as lymphoadenohypophysitis (LAH), lymphocytic infundibuloneurohypophysitis (LINH), lymphocytic panhypophysitis (LPH) based on involvement of pituitary gland. Absence of granulomas and presence infiltration of pituitary tissue by the lymphocytes is labeled as lymphocytic hypophysitis. Our patient presented with features of posterior pituitary deficiency and on evaluation had panhypopituitarism. Visual deficits were due to the inflammatory mass compressing the chiasm which improved with steroids. Persistent polyuria and polydipsia were due to deficient post pituitary hormones. Calcified lesions could well be healed tubercular granulomas which might have caused pituitary insufficiency and been undetected in histology due to limited sampling. However the patient improved in symptomatology with steroids and desmopressin and there was no evidence of lesion on repeat contrast enhancement.

Two cases of granulomatous involvement of pituitary have been described in literature from India. Bhardwaj etal (4) reported a case of granulomatous hypophysitis which was treated later with antitubercular drugs. Mehdiratta etal (5) described a case presenting as pituitary mass with non caseating granulomas treated with decompression and antitubercular drugs. Our patient is currently on maintenance dose of wysolone and desmopressin after six months of treatment.

The treatment of AH is, at the moment, only symptomatic. It includes reducing the size of the pituitary mass and/or replacing the defective endocrine function. Mass reduction can be achieved by pituitary surgery, lympholytic drugs (glucocorticoids, azathioprine, or methotrexate), or radiotherapy. Surgery has been the most common form of treatment and aims to reduce the pituitary mass and the associated compressive effects on the surrounding structures, without introducing new endocrine or neurological deficits.

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References