Lymphocytic Hypophysitis Mimicking Pituitary Macroadenoma

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Abstract
Lymphocytic hypophysitis is an inflammatory/autoimmune disorder that primarily involves the pituitary gland and the pituitary stalk. The common clinical presentations include headache, nausea, vomiting, fatigue, features of hypopituitarism and diabetes insipidus as well as diplopia, orbital pain and bitemporal hemianopia. We report a case of lymphocytic hypophysitis which presented as hemichorea. Neuroimaging showed a mass in the sella turcica region which, on histopathological examination was suggestive of lymphocytic hypophysitis. After excision of this mass, patient showed marked improvement in his symptoms albeit he developed panhypopituitarism. The patient was treated with pituitary hormonal replacement therapy and is currently asymptomatic.

Introduction
Lymphocytic hypophysitis is a rare disease and represents an inflammatory/autoimmune disorder that primarily involves the pituitary gland and in many cases the pituitary stalk.¹,² The clinical presentation of this inflammatory condition may mimic a pituitary adenoma,³ the spectrum of which includes headache, nausea, vomiting, fatigue, features of hypopituitarism, diabetes insipidus, diplopia, orbital pain and bitemporal hemianopia.⁴ Lymphocytic hypophysitis is most commonly diagnosed in women during pregnancy or in the post-partum period and can be associated with other types of autoimmune diseases like autoimmune thyroiditis, orbital pseudotumour, pernicious anemia, type I diabetes mellitus and primary biliary cirrhosis.¹,³,⁵-¹⁰ Although most commonly occurring in women during the child bearing years, lymphocytic hypophysitis has now been recognized to affect men and women of any age.¹¹

Case Report
A 76 years old male presented to us with history of fever, headache, and right sided abnormal limb movements for seven days. There was no history of head injury, loss of consciousness, vomiting, convulsion, bladder or bowel disturbances, or visual difficulty. There was no history of similar complaints in the past or any other medical disease like hypertension or diabetes. On examination, patient had choreoathetoid movements in right upper limb and lower limb. Rest of neurological and other systemic examinations were normal. Routine blood investigations were normal. Patient underwent an MRI brain which revealed a well-defined enhancing soft tissue intensity mass lesion in the sellar region including the infundibulum, extending into suprasellar region and inferiorly into the right sphenoid sinus associated with right sphenoid sinusitis. Cerebrospinal fluid (CSF) examination, thyroid function test, peripheral blood film for acanthocytes and serum angiotensin converting enzyme (ACE) levels of this patient were normal. Patient was treated with sodium valproate, haloperidol and clobazam with no respite from symptoms. Transnasal endoscopy and excision of pituitary macroadenoma was performed by a team comprising of experts from otorhinolaryngology and neurosurgery. Histopathological examination of the tissue fragment in the right posterior ethmoidal sinus and sphenoid sinus revealed respiratory mucosa lined by pseudo stratified columnar epithelium with focal areas of necrosis. The underlying stroma exhibited moderate to heavy mixed inflammatory infiltrate of lymphoplasmacytic cells and eosinophils along with Periodic acid Schiff (PAS) positive branching septate mycelia filaments, morphological resembling Aspergillus species (Figures 1 and 2).

Fig. 1: Periodic acid schiff (PAS) staining shows PAS positive branching septae mycelia filaments suggestive of Aspergillus

Fig. 2: H & E stain of biopsy shows inflammatory infiltrate of lymphoplasmacytic cells and eosinophils suggestive of lymphohypophysitis

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Vascularity was of moderate nature, hence mimicking a pituitary adenoma.

After surgery, patient’s choreoathetoid movements subsided completely. Patient also developed pan hypopituitarism in the form of apathy, hypotension, and features of hypothyroidism like constipation, dry skin and sluggish reflexes. Further investigations revealed henceforth: thyroid stimulating hormone (TSH) =0.04µIU/mL, tri-iodothyronine (T3) =1.38µg/dL, thyroxine (T4)=7.4µg/dL, serum cortisol =1.80µg/dL, serum testosterone= 0.16nmol/L and serum prolactin =19.98ng/mL . Antineutrophil cytoplasmic antibody (ANCA) and antinuclear antibody (ANA) were negative.

Patient was treated with anterior and posterior pituitary hormone replacement therapy in the form of hydrocortisone, levothyroxine and voriconazole for two months. Currently the patient is asymptomatic.

Discussion

Hemichorea refers to choreaform movements in one half of body. It sometimes occurs after lesions that selectively involve the caudate nucleus, putamen, and globus pallidus. Some common causes being acute stroke, non-ketotic hyperglycaemia, and systemic lupus erythematosus. Lymphocytic hypophysitis is an autoimmune condition in which the pituitary gland becomes infiltrated by lymphocytes, resulting in pituitary enlargement and impaired function. It demonstrates a wide spectrum of presentations, imaging results and treatment strategies.

This case presented to us with the symptom of hemichorea which can be attributed to the autoimmune process found in association with lymphocytic hypophysitis. As per our knowledge and reviewing of literature this is the first case of lymphocytic hypophysitis which is presenting as hemichorea. There is also a variation in the response to therapy, in particular corticosteroids. Those patients who have partial responses and relapses continue to represent therapeutic challenges. Longitudinal follow up and diligent anterior and posterior pituitary hormonal replacement therapy are essential for successful management of this condition.

Conclusion

Hemichorea could be a rare clinical manifestation of lymphocytic hypophysitis. Autoimmunity can thought to be the common denominator in both these conditions. We urge researcher to report more such associations to confirm our hypothesis.

References