CASE REPORT

LYMPHOCYTIC HYPOPHYSITIS; A RARE AUTOIMMUNE DISORDER THAT PRESENT AS LARGE SELLAR SUPRA SELLAR MASS WITH COMPLETE RESOLUTION WITH STEROIDS

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Significant proportion of sellar masses is seen in clinical practice. They range from most common pituitary adenomas to rare inflammatory lesions. Presentation can vary and depends if it secretes any hormone or imparts a pressure effect upon the surrounding vital structures. Radiological imaging coupled with histopathology is important tools of diagnosis. Management options depend upon type of disease.

Keywords: Lymphocytic hypophisitis, autoimmune, pituitary, sellar mass, steroids

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INTRODUCTION

Lymphocytic infundibulo-neurohypophysitis (LYH) is an unusual and rare autoimmune disorder affecting pituitary gland.[1] It is characterized by lymphocytic and plasma cell infiltration of the posterior lobe of the pituitary and the pituitary stalk.[2] It most frequently occurs in women of child-bearing age.[3] It, along with other Inflammatory lesions of the hypophysis, account for 0.5% of all symptomatic diseases of the pituitary.[4] Its course is unpredictable at some time.[5] Patient can present with headache, diplopia, galactorrhea, diabetes insipidus, hypopituitarism and psychosis.[2,3,6] The initial pituitary enlargement, secondary to infiltration and oedema, can evolve to remission, for spontaneous or pharmacological resolution of the inflammation, or evolve to progressive diffuse destruction with gland atrophy for fibrotic replacement, thus leading to various degrees of pituitary dysfunction. The autoimmune process against the pituitary gland is made evident by the appearance of circulating autoantibodies (APA), mainly detected by indirect immunofluorescence on cryostatic sections of human or primate pituitary. Among the target autoantigens recognized by APA are alpha-enolase, gamma-enolase, the pituitary gland specific factors (PGSF) 1 and 2 and corticotroph-specific transcription factor (TPIT).[7] In addition, pituitary microscopic deposits can be found in the liver, pancreas, and submandibular gland.[8] Magnetic resonance imaging (MRI) of the brain is a reliable investigation that can reveal an enhancing mass involving the sella and suprasellar region.[1] MRI findings of LYH and pituitary adenomas are similar. However the parasellar T2 dark sign can be a specific finding used to distinguish pituitary adenoma from LYH.[9] Surgical biopsy can be taken and can confirm histologic diagnosis.[10] Transnasal transsphenoidal (TNTS) is used for this purpose.[7,11] Microscopic examination shows a marked infiltration of lymphocytes and plasma cells in the posterior pituitary gland.[9] Course of the disease as well as its treatment is still controversial.[5] Steroid therapy with glucocorticoides[1] and hydrocortisone can cause complete resolution of mass.[5] Gamma knife surgery is reserved for persistent disease.[3]

CASE REPORT

A 36 years old lady presented with headache visual deterioration and vomiting. She had these symptoms for the last three months. She visited local clinicians and had all relevant necessary investigations at hand. She had normal vitals, her vision was 6/12 in both eyes. She had bitemporal hemianopia and had bilateral papilloedema. Her pituitary hormonal profile was normal. She had an MRI brain with contrast (Figures-1 to 4) showing large sellar lesion with right parasellar and sub-temporal extension. She had trans-nasal trans-sphenoidal biopsy which confirmed non-secreting pituitary adenoma. She was put on steroids and was put on next available list for staged procedure. Initially it was planned to decompress sella and optic chiasma as patient was losing vision. After trans-cranial sub-frontal approach, it was a great surprise not to have lesion at all. Patient was returned to ward and after recovery a computerized scan (CT) brain with contrast (Figures-5 and 6) was done. There was no lesion at all. There were early postoperative changes. Right skull base was eroded but there was no evidence of lesion. A thorough investigation was made including confirming patient's data, MRI scan scrutiny and tracing and taking opinions on histopathology report. Patient's data was correct. Biochemical profile and MRI scans were of the same patients. We reviewed histopathology slides by three centres and discussed the case in detail. Predominance of lymphocytes on slides (Figures-7 and 8) coupled with clinical presentation and radiological data led us to the conclusion: diagnosis of pituitary lymphocytic hypophysitis. It was one of its type in which there was a huge lesion with extension in right sub-temporal region causing erosion of the base of skull and vanished with steroid completely.

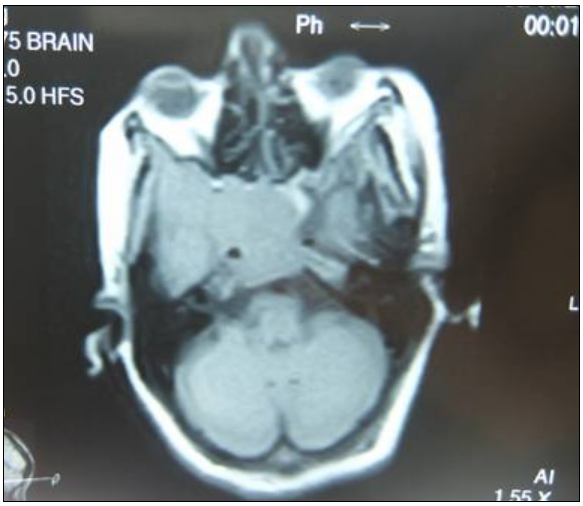

Figure-1: MRI Brain with contrast

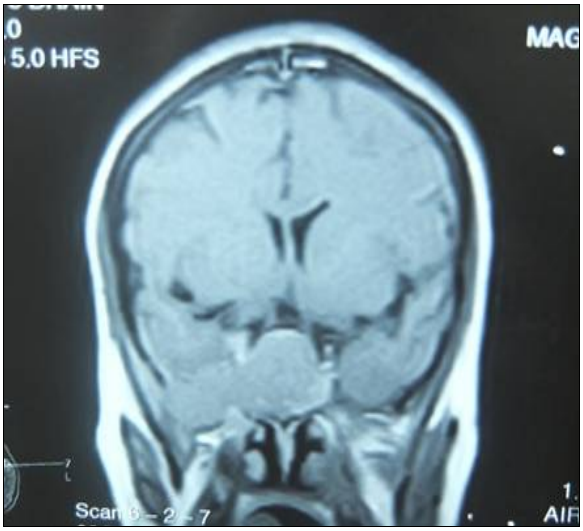

Figure-2: MRI Brain with contrast

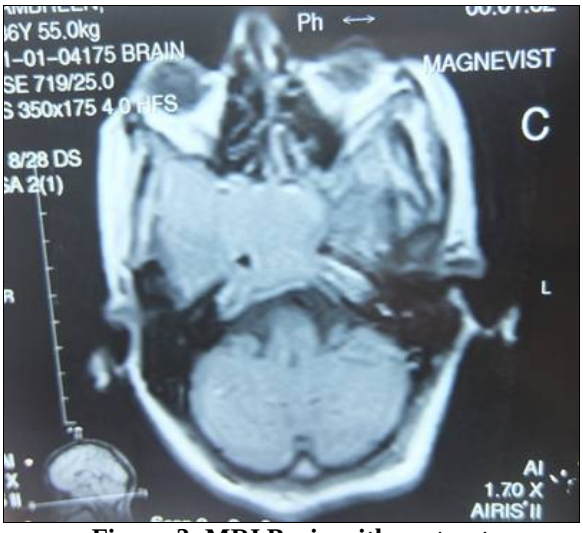

Figure-3: MRI Brain with contrast

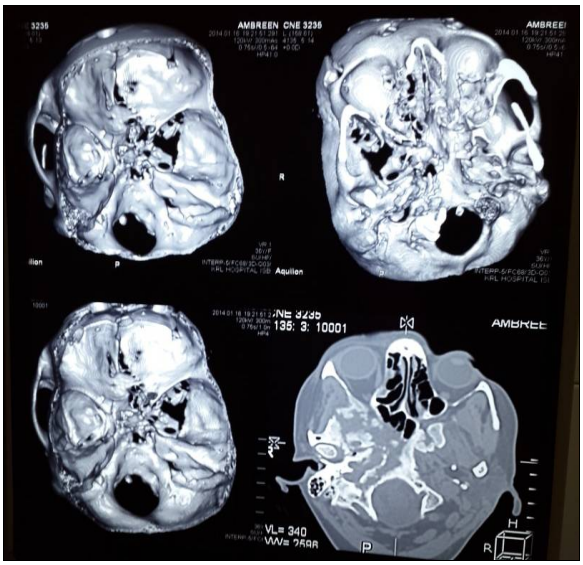

Figure-4: MRI Brain with contrast

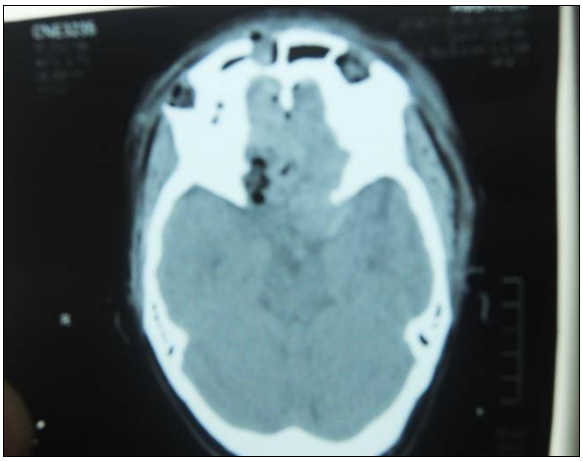

Figure-5: CT Scan with contrast

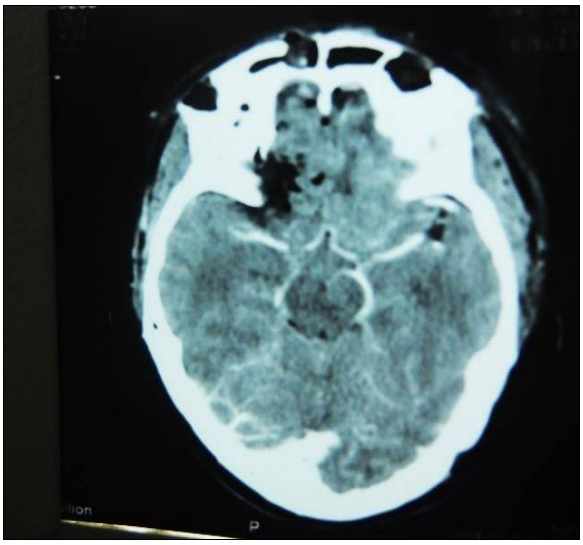

Figure-6: CT Scan with contrast

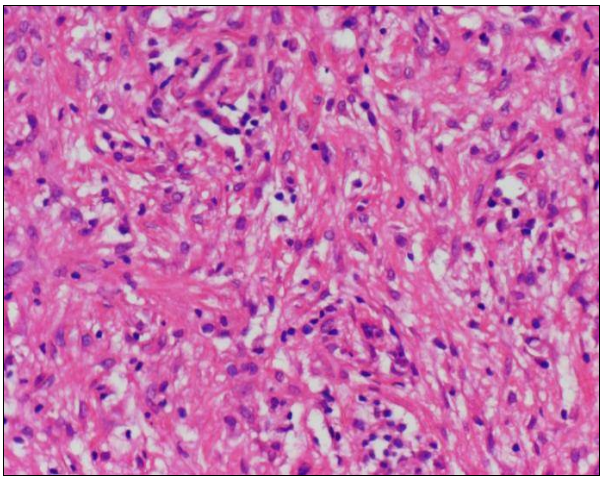

Figure-7: Predominance of lymphocytes

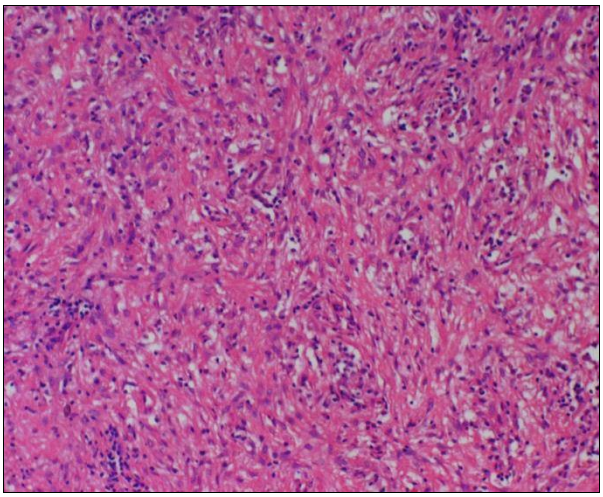

Figure-8: Predominance of lymphocytes

DISCUSSION

Sellar and supra sellar masses are unique and have interesting pattern of presentation. Clinical scenario is straight forward in most of the cases as presentation can be because of hormonal effect or mass of lesion causing significant pressure upon nearby vital structures to produce various symptoms and signs. Modern radiological and biochemical tests help clinician not only in diagnosis but decision making regarding optimum treatment of the disease. However inflammatory lesion of the sellar region that accounts only 0.5% of all symptomatic sellar lesions may mislead not only in diagnosis but in treatment sometimes.[4] The same happened in our case. Although we had biochemical, radiological and histopathological evidence of pituitary adenoma yet we were surprised by not finding lesion per operatively. This was because no one thought of having an inflammatory lesion. There was no suspicion in mind. So a suspicion in mind of having inflammatory lesion can avoid misdiagnosis as well as aggressive surgical approach.[8] Various diagnostic tools, both radiological and biochemical, aid in diagnosis.

MRI is considered one of the reliable diagnostic radiological investigations but even with this the diagnosis of inflammatory sellar lesions is very difficult. It is because of the similar MR findings of both disorders. Thickened stalk, pituitary symmetry, homogeneous enhancement, and parasellar dark signal intensity on T2-weighted images can occur in both of them. However parasellar T2 dark sign on MR imaging in patients with lymphocytic hypophysitis can be considered reliable.[10] Theoretically it seems very easy but practically overlapping clinical and biochemical scenario can mislead the diagnosis as was in our case. We had hormonal profile and we MRI brain with contrast showing large mass lesion. We had histo-pathological specimen taken from trans-sphenoidal route confirming lesion as non-secreting pituitary adenoma.

Steroids play as important role in the treatment. Adequate steroid doses not only improve symptoms but also regress mass. This was the course of the disease in our case also. There was complete regression of mass with steroids. However in rare instances it can recur upon lowering of steroid doses.[12] Fortunately this did not happen in our case. She was on three months' follow up and it is now over a year the lesion has not recurred.

Role of histopathology also comes in consideration. We had histopathology report which was confirming pituitary non-hormone secreting adenoma. There was no doubt in the mind of the surgeon as well as pathologist so it led not only in misdiagnosis but also patient had to go through a major surgical procedure.

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