LYMPHOCYTIC INFUNDIBULO-NEURO HYPOPHYSITIS: A RARE CAUSE OF DIABETES INSIPIDUS

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A 69 years old female with polydipsia, polyuria and intermittent headache for 6 months was admitted to hospital due to intractable symptoms for last 3 months. On Admission, physical examination, blood glucose level, pituitary and thyroid hormones level were within normal limits. Urine Specific gravity was slightly decreased. MRI showed slight enlargement of sella turcica, with uniform flattening of floor, pituitary showed uniform, mild enlargement (1.1cmX1.5cmX1.2cm), isointense to white mater in T1WI with loss of normal “bright spots” of posterior pituitary, heterogeneous signal with low intensity posteriorly in T2WI and intense homogeneous enhancement with “dural tail” in T1WI with Gadolinium. Pituitary stalk was slightly thickened but undisplaced. The lesion was diagnosed as pituitary adenoma and underwent transsphenoidal excision of pituitary.

Intraoperatively, pituitary gland was soft, dark red, moderately vascular with clear margin measuring 1.2cmX1.5cmX1.3cm, with flattened sellar floor. Light microscopy of the pituitary showed normal pituitary tissue with a large number of lymphocytes, few plasma cells and little fibrous tissue and pathologically proved to be lymphocytic hypophysitis. (Fig. 1A-D).

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**Figure 1A**: T1WI shows uniformly mildly enlarged pituitary isointense to white mater and loss of normal "bright spots" of posterior pituitary (1B) T2WI shows heterogeneous signal with hypo intense signal posteriorly (1C) Gd enhancement shows intense homogeneous enhancement of the lesion with "dural tail" sign (arrow) (1D) Light microscopy showing normal pituitary tissue with large number of lymphocytes, little plasma cells and fibrous tissue (HE X 400)
Discussion

Lymphocytic hypophysitis (LyH), an extremely rare autoimmune disease first reported by Guodie and Pinkerton in 1961,\(^1\) characterized by lymphocytic infiltration of the pituitary with female predominance (M: F = 1.5:1.8) mainly in late pregnancy or postpartum. But, can also occur in non-pregnant and elderly female and very rarely in males.\(^2,3,4\) LyH can be classified into (a) Lymphocytic adenohypophysitis (b) Lymphocytic infundibulo neuro hypophysitis and (c) Lymphocytic infundibulo panhypophysitis. Clinical presentation like headache, vomiting, visual field impairment can be due to mass effect whereas anterior hypopituitarism and diabetes insipidus are due to pituitary dysfunction.\(^2\)

In MRI, LyH shows diffuse pituitary enlargement, with or without symmetrical sellar floor flattening or symmetrical suprasellar expansion. If pituitary stalk is involved, thickening of pituitary stalk without obvious deviation and loss of normal “bright spots” of posterior lobe in T1WI is present. In Gadolinium enhancement mostly lesion has homogeneous enhancement.\(^2,3,4\) “Dural tail” sign can be seen in advanced disease with dural involvement.\(^2\)

Our patient showed mild but diffusely enlarged pituitary and sella tursica with symmetrical flattening of the sellar floor. In T1WI pituitary was isointense to white matter with loss of normal “bright spots” of posterior lobe and in T2WI pituitary was heterogeneous with low signal posteriorly corresponding to posterior lobe. In Gadolinium enhancement, there was intense and homogenous enhancement of whole pituitary with “dural tail” sign. Through combined clinical and imaging characteristics Lymphocytic infundibulo-neuro hypophysitis with features of Diabetes Insipidus was of high probability. Pituitary adenoma sharing identical imaging pictures shows asymmetric pituitary enlargement with asymmetric flattening of sellar floor, asymmetric suprasellar extension and contralateral deviation of normally thickened pituitary stalk, with more clear margin, slightly delayed inhomogeneous enhancement and persistence of “bright spots” of neurohypophysis in T1WI and lack of “dural tail sign”.\(^2,3,4\)

Pathological diagnosis is still the gold standard for LyH. Glucocorticoids have been used for the treatment of this disease, but the dose and treatment are controversial with inconsistent efficacy. Transsphenoidal partial resection of pituitary is indicated in LyH leading to intractable headache, visual field defects, or non-responsive to glucocorticoid therapy. Immunosuppressive or radiation therapy is considered if Medical and surgical treatments are in effective.\(^5\)

References