

中文題目：IgG4 相關性腦垂腺炎：案例報告

英文題目：IgG4-related Hypophysitis: Case Report

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Case presentation:

IgG4-related hypophysitis is a rare disease characterized by easily mimic pituitary tumor and receive unnecessary surgical biopsy/resection but actually good response to steroid treatment. It usually presents with excessive polyuria, polydipsia, visual field defect and headache. Its presentation is usually chronic. In this case report, we present a 39-year-old male patient with thirsty and polyuria initially. He was found to have a urine osmolality of 69 mOsm/kg. Diabetic insipidus was diagnosed by water deprivation test. Sella magnetic resonance imaging showed no pituitary lesion and lack of normal T1W1 hypertense spot of posterior pituitary lobe, which compatible with diabetic insipidus. However, he had symptom of easily fatigue after 9 months later. There was an elevation of prolactin and decrement of LH, FSH, and testosterone. Follow-up sella MRI revealed one enhancing nodule about 0.7cm in pituitary stalk. Further evaluation showed elevation of IgG4(147.2mg/dL). A medium dose of prednisolone was prescribed. IgG4 levels of the serum decreased and follow-up MRI revealed mild thickening and enhancement of pituitary stalk in regression obviously following glucocorticoid therapy. IgG4-related hypophysitis was confirmed by fulfilling Leporati criteria 2,4, and 5.

Conclusion:

IgG4-related hypophysitis has good response to steroid therapy. However, patient presented with diabetic insipidus alone usually did not receive immediate steroid therapy. After introducing of new diagnosis criteria by Leporati in 2011, the diagnosis of IgG4-related hypophysitis became easier and faster. The prevalence of IgG4-related hypophysitis may be more common than previously thought. Given that the presentation of IgG4 hypophysitis maybe diabetic insipidus alone and the MRI finding was also negative initially in our reported case, it is important to consider this entity in the differential in order to prevent delaying treatment.

Abbreviations: IgG4 = immunoglobulin G4, IgG4-RD = IgG4-related disease, MRI = magnetic resonance imaging

Keywords: glucocorticoid, hypophysitis, IgG4-related disease, pituitary hypofunction, diabetic insipidus