自體免疫性下視丘炎

Autoimmune hypophysitis

吳婉禎

台大醫院 內科部

Wan-Chen Wu

Division of Endocrinology and Metabolism, Department of Internal Medicine, National Taiwan University Hospital, Taipei, Taiwan

Hypophysitis is characterized by inflammation and cellular infiltration of the pituitary gland, and is generally accepted to be an autoimmune cause. The true incidence and prevalence are unknown. The disease is rare with an estimated incidence of 1 in 9 million, and reported prevalence among other pituitary masses on the basis of surgical series consistently under 1%. The disease can be classified by etiology, anatomy, and histopathology. Etiology distinguishes hypophysitis into primary (or idiopathic) and secondary to systemic diseases, medications, sellar diseases, or adjacent parasellar masses. Depnding on the anatomic location of the infiltrate, lymphocytic hypophysitis (LYH) is classified into 3 subtypes: adenohypophysitis (LAH) – involving mainly anterior pituitary lobe, infundibuloneurohypophysitis (LINH) – involving mainly the stalk and posterior pituitary lobe, and panhypophysitis (LPH) – involving both pituitary lobes. Based on the histolopathologic features, there are two main forms: lymphocytic and granulomatous, and three rare variants: necrotizing, xanthomatous, and IgG4 plasmacytic.

Hypophysitis generally occurs in the fourth decade of life and is rare in children and the elderly. LAH is 3.4 to 6 times more common in women, especially during late pregnancy and in the postpartum period. LINH affects both genders equally. LPH is more frequent in men.

Clinical manifestations vary, but usually the result of an expanding sellar mass, hypopituitarism, central central insipidus, and change of prolactin secretion. Sellar mass effect induces headaches, visual disturbances, and diplopia. The sequential order of trophic hormone deficiency specific to hypophysitis is as follow: ACTH > TSH > LH/FSH > PRL > GH.

Autoimmune hypophysitis (AH) should be suspected in women presenting

pituitary enlargement in the peri-partum, or with pituitary dysfunction and underlying autoimmunity and in all patients with a rapidly growing pituitary mass. Although the definite diagnosis can be achieved only by histology on the trans-sphenoidal biopsy specimen, non-invasive diagnosis can be putatively assessed by MRI and endocrinological deficit pattern. The clinical applicability of pituitary autoantibodies in the routine diagnosis and management of patients with AH is limited at present by a low diagnostic specificity and sensitivity and by conflicting results generated by different methods.

The aim of acute management is to decompress the pituitary mass and prevent adrenal crisis. Mass reduction can be achieved by pharmacotherapy, surgery, and radiotherapy. Glucocorticoid can effectively reduce pituitary mass volume. Surgery is recommended for patients who required diagnostic confirmation or decompression of the optic chiasm. The purpose of chronic management is to restore adequate hormone levels. Long-term follow-up studies have yet to be performed for patients with hypophysitis.