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ABSTRACT

IgG4-Related Hypophysitis Presented with Diabetes Insipidus, a Diagnostic Problem Case Report

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A seventeen years old female came to the hospital with complaints of three-years history of poliuria and amenorrhea. She had signs of diabetes insipidus and secondary amenorrhea. There were increasing in prolactin hormon, but decreased spontaneously to normal level. MRI showed pituitary stalk enlargement.

Patient was consulted to Toranamon Hospital, Tokyo, and anterior pituitary provocative test revealed severely impaired response of gonadotropin (FSH and LH) to GnRH and GH respons to GHRP-2 (growth hormone-releasing peptide-2), blunted response of TSH to TRH. ACTH and cortisol respons to CRH were within normal limit. Her serum estradiol (<11 pg/ml) and IGF1 (71 ng/ml) were low, indicating that patient had hypogonadotropic hypogonadism and severe growth hormone deficiency. Lumbal bone mineral density test showed osteoporosis sign (Z score -3,7).

The differential diagnosis of her pituitary stalk enlargement include germinoma, lymphocytic infundibulo-panhypophysitis, granulomatous hypophysitis secondary to sistemic illness (e.g sarcoidosis, collagen deseases, and tuberculosis) and IgG4 related hypophysitis. Analysis of PLAP (germinoma marker), serum lysozyme and angiotensin-converting enzyme (for predicting sarcoidosis), anti-nuclear antibody, rheumatic factor, PR3-ANCA, MPO-ANCA (for predicting collagen deseases), T-Spot (test for tuberculosis) and others including sIL-2 receptor, Beta HCG, AFP were all within normal limit. However her serum IgG and IgG4 were elevated (1780 mg/dl and 144 mg/dl). The patient was suggested as having IgG-4 related infundibulo-hypophysitis.

IgG4-related disease involves in many variable organs. Patient was then examined with thyroid ultrasound and the result compatible with chronic thyroiditis. Thyroid autoantibodies TPOAb and TGAb were negative. Abdominal ultrasound showed no abnormality in pancreas, liver and retroperitoneal region. Although the definitive diagnosis of IgG4-related infundibulo-hypophysitis is biopsy, but because of the risk for panhypopituitarydism, the test was not done.