Lymphocytic Hypophysitis in Association with Relapsing Polychondritis – A Case Report.

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Inflammatory lesions of the pituitary gland, known as hypophysitis, are uncommon disorders which mimic tumors of the sellar region causing signs of mass effect. Hypophysitis may be primary or secondary. Lymphocytic hypophysitis, the most common form of primary pituitary inflammation, is initially characterized by lymphocytic infiltration and enlargement of the pituitary gland. It classically present during late pregnancy or early postpartum, and it is thought to be an autoimmune phenomenon. Bacterial, viral, and fungal infections are rare causes of secondary hypophysitis. Systemic inflammatory diseases can also involve the pituitary; reported diseases include sarcoidosis, Wegener’s granulomatosis, Takayasu’s arteritis, Crohn’s disease, and Langerhans cell histiocytosis. In this report, we describe a patient who presented with clinical and radiological features of hypophysitis with panhypopituitarism, as well as with the diagnostic criteria of relapsing polychondritis, ie nasal chondritis, signs of ocular inflammation, cochlear and vestibular dysfunction in addition to CNS involvement. Serologic studies in our patient were also compatible with relapsing polychondritis and included positive ANA, anti-cardiolipine antibodies and VDRL. Positive c-ANCA in the absence of diagnostic criteria for Wegener’s granulomatosis suggest vasculitis which can be associated with relapsing polychondritis. Histologic studies of biopsy specimens from affected sites were also compatible with the diagnosis and a pituitary biopsy confirmed lymphocytic hypophysitis. This is to our knowledge the first reported case of hypophysitis in association with relapsing polychondritis. Also worthy of note is the rapid and dramatic response of the patient to medical treatment with corticosteroids, the favorable outcome persisting after an 18-month follow-up period.

Notes take at the poster.

A 40 years old woman with recurrent headache, loss of vision, amenorrhea, and galactorrhea. Physical exam noted signs of hypothyroidism (pallor, lethargy, swollen face, slow speech), and of her previously diagnosed (10 years ago) polychondritis (saddle nose post-inflammatory necrosis of her nose cartilage). MRI showed pituitary enlargement with suprasellar extension and infiltration of the stalk. Hormone tests showed pan-hypopituitarism and increased prolactin. Pituitary biopsy (October 2002) showed lymphocytic hypophysitis. Patient began replacement with thyroxine and prednisone.

In April 2004 the patient developed headache, blurred vision, lethargy, amenorrhea, hearing loss, conjunctivitis, and dizziness. Visual field exam revealed blindness in the right eye and central scotoma in the left eye. Retinal angiography showed arteritis. CSF analysis showed lymphocytosis and CSF cultures were negative. MRI revealed the same pituitary findings as 2 years ago, establishing the diagnosis of recurrent lymphocytic hypophysitis. Patient received oral pulse therapy with steroids with marked clinical improvement.