CASE REPORT

Visual disturbance by lymphocytic hypophysitis in a non-pregnant woman with systemic lupus erythematosus

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The authors present the case of a patient with systemic lupus erythematosus who developed visual disturbance and amenorrhea. Though the clinical and radiological findings resembled those of pituitary adenoma, the patient was finally diagnosed as having lymphocytic hypophysitis after the operation. We briefly describe this relatively rare entity in relation to its autoimmune pathogenesis.

Keywords: lymphocytic hypophysitis; systemic lupus erythematosus (SLE); antipituitary antibody; pregnancy; autoimmunity

Case history

A 26-year-old non-pregnant woman with a two-year history of systemic lupus erythematosus (SLE) was referred to us for decreased visual acuity in both eyes, bitemporal hemianopsia and amenorrhea. Neurological findings were unremarkable but she had a nephrotic syndrome caused by lupus nephritis. Though the activity of lupus was generally sedated by the administration of prednisolone, the dose of which was reduced to 5 mg/d at the time of presentation, pancytopenia was detected by blood analysis and anti-DNA antibodies and antinuclear antibodies were positive.

Endocrinological examination revealed panhypopituitarism, which included prolactin <1.0 ng/ml (normal, 2–5), luteinizing hormone 1.0 mIU/ml (normal, 5–30), follicle-stimulating hormone <0.1 IU/ml (normal, 5–40), adrenocorticotropic hormone 3.0 pg/ml (normal, 5–35) and cortisol <1.0 μg/dl (normal, 3–15). Provocative testing for thyrotropin-releasing hormone, luteinizing-hormone-releasing hormone and insulin failed to show any significant elevation of these hormones.

Head computed tomography disclosed an isodense sellar mass, extending to the suprasellar region, which was homogeneously enhanced by contrast medium. Magnetic resonance imaging demonstrated the mass to be iso intensity on both T1 and T2 weighted images and homogeneously augmented by gadolinium-diethylene-triaminopentaacetic acid (Figure 1). Compression of the optic chiasm could be depicted. Since the subject had already been prescribed prednisolone (5–50 mg/d) as the treatment for SLE and we could not preclude the possibility of a non-functioning pituitary adenoma, we therefore decided to establish a histological diagnosis by surgical intervention. An elastic hard and whitish-gray mass was observed. The specimen sampled for the intraoperative frozen sections was composed of infiltrating lymphocytes and proliferating vessels with no evidence of neoplasia. Therefore, partial resection of the mass for decompression of the optic chiasm was performed. Hematoxylin and eosin staining of the pathological specimen revealed diffuse infiltration of lymphocytes with stromal fibrosis and destruction of parenchymal cells (Figure 2). Most infiltrating lymphocytes were positive for UCHL-1 (T-cells) (Figure 3), while L26-positive cells (B-cells) were sparse. Staining for κ and λ light chains showed almost equal distributions of both. All these pathological findings suggested a diagnosis of lymphocytic hypophysitis.

Circulating antipituitary antibodies (anti-AIT-20 cell antibodies and antibodies against rat anterior lobe cytosol) were proven to be present postoperatively.
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Figure 1 A sagittal view of enhanced T1-weighted magnetic resonance imaging showing compression of the optic chiasm by the homogeneously augmented mass.

Figure 2 Hematoxylin and eosin staining of a pathological specimen demonstrating diffuse infiltration of lymphocytes with stromal fibrosis and destruction of parenchymal cells. (× 100).

Figure 3 Most infiltrating lymphocytes are positive for UCHL-1. (× 400)

Lymphocytic hypophysitis is a rare entity which tends to occur predominantly in women during the peripartum period. Due to multiple complicated immunologic changes at this time, it has been believed that autoimmunity may contribute to the pathogenesis of the disease. While there have been several reports of lymphocytic hypophysitis in men or non-pregnant women, as in our case, further support for the idea of autoimmune basis of the disease is the concurrence of conditions such as Hashimoto’s thyroiditis and adrenalitis in approximately 20% of the patients. However, we are not aware of any histologically proven description of lymphocytic hypophysitis with SLE. Why has an association with such a common autoimmune disease not been reported so far? The answer is not obvious, but a clue might be given by the fact that all the autoimmune associates with lymphocytic hypophysitis previously documented as well as lymphocytic hypophysitis itself are encompassed in a disease group in which organ-specific antibodies such as anti-thyroid antibodies and antipituitary antibodies are produced, while SLE is classified as a non-organ specific or systemic autoimmune disease like rheumatoid arthritis and dermatomyositis. The present case, however, provides concrete evidence that the pathogenesis of lymphocytic hypophysitis is strongly ascribable to autoimmunity showing that it can be associated even with...
systemic autoimmune disease. Therefore, visual disturbances in patients with collagen diseases of the autoimmune mechanism such as SLE should alert clinicians to the possibility of pituitary disease including not only adenoma but the very uncommon lymphocytic hypophysitis.

References