Case Report

Postpartum Pituitary Hypophysitis

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The case of a young woman who developed lymphocytic hypophysitis 2 weeks after delivery of a healthy baby is reported. The patient presented with clinical features suggestive of a pituitary mass lesion, but surgery was avoided when other clinical and radiologic features were considered. The patient recovered with steroid treatment only. We review the literature on this increasingly recognized condition and argue that medical management may be more suitable than previously thought. Key Words: Lymphocytic hypophysitis—Pregnancy—Steroid treatment.

Inflammatory growths in the area of the pituitary gland should be considered in the preoperative differential diagnosis of every pituitary tumor that is not associated with important hormonal dysfunction, particularly in the peripartum period. It may also present in the early stages of pregnancy, with symptoms mimicking hyperemesis gravidarum or meningeal irritation (1). Hypophysitis has been reported in men and in association with a number of autoimmune diseases, including Sjogren syndrome, Graves disease, diabetes, and rheumatoid arthritis.

The precise etiology remains unclear, but animal studies have shown that rubella virus glycoproteins can induce autoimmune lymphocytic hypophysitis in Syrian hamsters and that neonatal thymectomy prevents the disease (2). A definitive diagnosis can be made by endoscopic endonasal biopsy (3), but we suggest that histologic examination may not always be necessary to diagnose hypophysitis when characteristic clinical features are found, as illustrated by the present case.

CASE REPORT

A twenty-year-old right-handed woman presented with a week-long history of progressive bilateral visual deterioration in early January 2000. During the previous week, she had given birth to a baby girl by Caesarean section. Initially, she noticed visual blurring, particularly in the temporal fields of vision, when she moved her head quickly. She also began to complain of vague frontotemporal headaches without diurnal variation or change with posture. There was no nausea, vomiting, diplopia, or ocular pain. She had no other symptoms. At presentation, she had not begun to lactate. The pregnancy had been uneventful, and before conception, her menstrual cycle had been regular. She had a miscarriage 3 years previously. There was no other relevant past medical history and no family history of relevance—in particular, no past or family history of autoimmune disease.

On examination, her visual acuity was 6/12 OD and 6/9 OS. There was a bitemporal hemianopia on field testing with normal fundus examination. There were no other abnormal neurologic or endocrinologic signs. A computed tomograph (CT) scan was reported as showing a pituitary macroadenoma extending into the suprasellar region.

Routine hematology tests revealed a hemoglobin level of 10.7 g/dL, an increased platelet count of 436, and a normal biochemical profile. Her prolactin level was normal (149 nmol/L), but her morning cortisol level was low at 27 U/L (normal, 170–720 U/L); the low cortisol level may have been influenced by the steroids being used in her treatment. Her thyroxine level was low (< 6.4), but her thyroid-stimulating hormone level and bone profile were normal.

Before and after contrast, T1-weighted magnetic resonance images (MRI) showed a homogeneously enhancing pituitary mass with suprasellar extension compressing the optic chiasm and the hypothalamus (Fig. 1). The mass was intimately related to, but not surrounding, the cavernous and supraclinoid internal carotid arteries. No intracavernous extension or evidence of hemorrhage was noted. She was given 4 mg of dexamethasone four times daily for a presumed diagnosis of lymphocytic hypophysitis. During the first 3 days of treatment, her headaches improved, and her visual acuity improved to 6/4 bilaterally. There was mild red desaturation in the temporal field OU, indicating marked improvement in visual fields. Cortisol and thyroxine replacement was begun.

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A second MRI scan was performed 4 days after the first imaging study (Fig. 2). The homogenous enhancement around the pituitary gland had reduced considerably and was now clear of the optic chiasm. She was discharged on a reducing dose of dexamethasone and replacement thyroxine and hydrocortisone therapy. On the day of discharge, her visual function was normal, including normal fields, visual acuity, and color vision.

DISCUSSION

Our patient represents a classic example of this unusual condition, the etiology of which remains unknown.

FIG. 1. A: Precontrast T1-weighted sagittal image of the pituitary gland. The gland is enlarged and the suprasellar extension compresses the optic chiasm. B: The enlarged pituitary enhances homogenously after administration of gadolinium diethylene triamine penta-acetic acid.

FIG. 2. A: Coronal and B: sagittal postcontrast T1-weighted images. After 4 days of intravenous dexamethasone, the pituitary has reduced in size and no longer compresses the optic chiasm. The pituitary stalk remains thickened.
She displayed features of a suprasellar lesion, and the clinical context and imaging findings suggested lymphocytic hypophysitis. These clinical features include a rapid onset of visual impairment, often in the peripartum period (4). CT and MRI imaging show a diffuse pituitary mass and enlarged pituitary stalk with homogenous contrast enhancement. Diabetes insipidus and persistent amenorrhea may occur; later, panhypopituitarism may also occur. Dynamic MRI has been used to demonstrate an abnormality of the hypophysial vasculature, even if the pituitary disease is seen to regress on conventional MRI studies (5). Gallium-67 scintigraphy may show abnormal uptake in the lesion (6). Autoantibodies to human pituitary cytosol proteins have been detected in as many as 70% of patients with biopsy-proven lymphocytic hypophysitis (7) and may represent markers for an immunologic process affecting the pituitary gland. As many as 25% of patients may have autoimmune thyroiditis (8).

Histologic examination of the pituitary shows a lymphoplasmacytic infiltrate with occasional neutrophils, eosinophils, and macrophages. There may be focal or diffuse adenohypophysial destruction of variable severity with associated fibrosis.

Most reported cases have been managed surgically, but steroid therapy has been advocated (9). One prospective study of nine patients with lymphocytic hypophysitis treated with high-dose methylprednisolone pulse therapy demonstrated improvement of adenohypophysial function in four patients. There was also a cessation of diabetes insipidus in all four patients (10). The presumptive noninvasive diagnosis of lymphocytic hypophysitis seems possible in a high proportion of patients, and high-dose steroids may result in improvement of clinical, endocrinologic, and MRI findings. In the absence of a visual field defect, however, it has been recommended that surgery and steroid therapy be withheld with periodic reassessment (11). In the absence of a controlled clinical trial, it is possible only to speculate whether there is a higher incidence of hypopituitarism after surgery for this condition. The current report illustrates that the response to steroids may be so rapid that very little time is lost if the diagnosis is incorrect. The major differential diagnosis includes other pituitary adenomas common in pregnancy: meningiomas (which may be distinguished from this condition by imaging characteristics) and neurosarcoidosis (the initial management of which is the same as the management for lymphocytic hypophysitis).

The natural history of lymphocytic hypophysitis begins with enlargement of the pituitary secondary to inflammatory infiltration and progresses to atrophy of the gland with destruction of pituitary tissue and replacement with fibrosis. The prognosis of the condition remains unclear, and there are only a few reports of pregnant women who have had an episode of lymphocytic hypophysitis (11). Recurrence after 2 years has also been reported (12).

REFERENCES