

A Rare Case of Male Lymphocytic Hypophysitis

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Lymphocytic hypophysitis (LH) is characterized by lymphocytic pituitary infiltration, which occurs mostly during or after pregnancy. Its involvement in male is very rare. The authors report herein a LH mimicking pituitary macroadenoma clinically and radiologically in male patient who presented with visual disturbance and hypogonadism.

KEY WORDS : Pituitary · Lymphocytic hypophysitis · Pituitary adenoma · Hypopituitarism · Transsphenoidal surgery.

Introduction

Lymphocytic hypophysitis (LH) is a rare inflammatory disorder involving pituitary gland and pituitary stalk. This is associated with autoimmune disease and most LH produces an intrasellar mass in women during late pregnancy or in the postpartum period^{2,4}. On literature review^{3,9}, only a few cases of LH have been reported in men, and even more rare for Korean man. In this report, we describe a rare male LH case, preoperatively diagnosed as pituitary macroadenoma.

Case Report

This 34-year-old man presented with progressive visual blurring and headache. He also complained of decreased libido. On admission, ophthalmology consultation demonstrated the reduced visual acuity and bitemporal hemianopsia. Baseline hormonal assay revealed hypofunction mainly in pituitary-gonad axis. Serum testosterone level was 0.14ng/dL (2.4-10.0ng/dL). An early morning cortisol level was 7.2ug/dL (9.4-26.2ug/dL), but he did not show any symptoms of hypocortisolism. His past medical history was unremarkable and there was no evidence of autoimmune disease. Preoperative sellar magnetic resonance (MR) images showed a strong homogeneously enhanced, 13×20mm-sized, pituitary mass with suprasellar extension which pushed up the optic chiasm. The normal pituitary gland was displaced anteriorly and superiorly, and pituitary stalk was not thickened (Fig. 1).

Under presumptive diagnosis of nonsecreting pituitary macroadenoma, transsphenoidal operation to the sellar lesion was performed. Intraoperatively, white-yellowish granular, friable tissue was noted and subsequently removed. The frozen section examination was consistent with an adenoma.

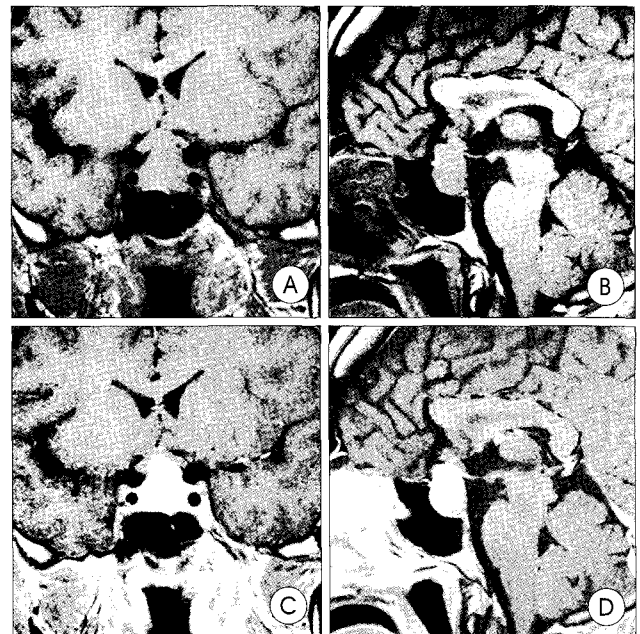


Fig. 1. Magnetic resonance images of the sella in a male presenting with sexual dysfunction and visual loss. Note a large pituitary mass compressing optic chiasm by suprasellar extension (A, B). A uniform and intense contrast enhancement in the sellar lesion is demonstrated, but the pituitary stalk appears normal (C, D).

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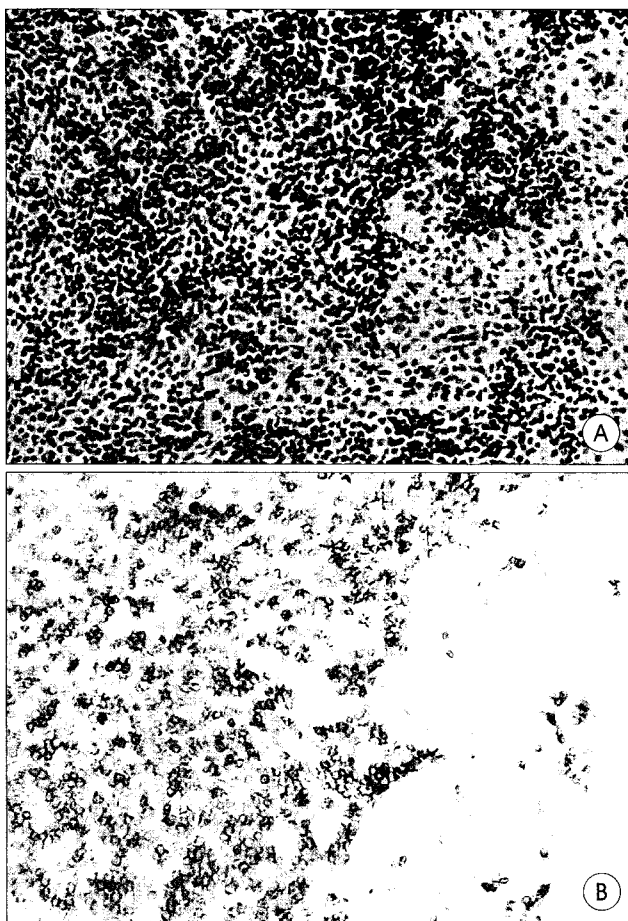


Fig. 2. Photomicrographs of lymphocytic hypophysitis. A : Lymphoid cells are diffusely infiltrated into the atrophic pituitary acini (H&E, original magnification x 200). B : Immunohistochemistry demonstrates the inflammatory cells consisting of mainly CD3-positive T-lymphocytes (original magnification x 200).

After the successful removal, his visual disturbances and headaches resolved. Postoperatively, the patient's cortisol (25.2ug/dL) and testosterone (2.4ng/dL) levels were normalized. One year later, he continues to have normal pituitary function without hormone replacement, including testosterone levels and sexual potency. A postoperative MRI scan was normal. Further immunological studies were performed, and antinuclear, antipituitary, antithyroid microsomal, and antiparietal cell antibodies were all negative. Histologically, the pituitary was overrun by an infiltrates of lymphocytes and plasma cells that deform the structural integrity of the anterior lobe. Immunophenotyping of the infiltrating lymphoid cells consisted of CD3+T-lymphocytes and CD20+B-lymphocytes (Fig. 2).

Discussion

LH is one of the non-neoplastic inflammatory mass lesions of the sella¹⁰. With its rarity in male, young females,

usually late in pregnancy or within the first year postpartum, are mostly affected⁵. By autoimmune proliferations and inflammatory process, the pituitary gland is diffusely involved and infiltrated by lymphocytes, sufficient to produce sellar enlargement, and with disease progression, the pituitary cells are replaced with fibrous tissue⁸. As noted in the present case, and in some instances, the gland may be so massively enlarged that it protrudes into the suprasellar space and cavernous sinus.

The clinical presentation of LH is variable and not distinguishable from a nonfunctional pituitary adenoma or other sellar mass lesions. Visual disturbance, low-grade hyperprolactinemia, headache, diabetes insipidus, and hypopituitarism of variable degrees have been reported as its presenting features^{4,6,10}. The symptoms and signs are similar between both men and women of LH, however, as noticed in this patient, impotence associated with decreased libido is also quite prevalent⁹. Unlike the first male case of LH in Korea⁹, who had a definite eosinophilia considered as indirect clue of autoimmune basis, our case showed no evidence of systemic autoimmune disorder on clinical and laboratory tests. High index of suspicion on LH is required preoperatively because it can be treated medically with corticosteroids. However, as our experience in this patient, many cases of LH are confirmed only after surgery for suspected pituitary adenoma has been undertaken. Histopathologic diagnosis of LH seems to be feasible, but reactive changes secondary to the tumoral hemorrhage, necrosis, or infarction may cause difficulty in differentiating LH from complicated pituitary adenoma^{1,5}. Histologic examination of this patient did not reveal fibrotic change within the lesion indicating early inflammatory in nature.

Radiologically, the differential diagnosis may include pituitary adenoma, pituitary abscess, granulomatous pituitary lesions including sarcoidosis, histiocytosis X, and tuberculosis, and other parasellar tumors and cysts^{5,7}. On radiographic diagnosis of LH, it is occasionally regarded as a variant form of pituitary adenoma, but MR findings such as, symmetrical and diffuse enlargement of the hypophysis, intense pituitary enhancement extending to the stalk and basal hypothalamus, and parasellar involvement and para-meningeal enhancement within the diaphragm sella, sphenoid sinus or cavernous sinus may be significant features of LH⁷. Our case did not show hypertrophy and enhancement of pituitary stalk that are also suitable image findings of LH.

Although its natural history and optimal management remains uncertain, use of corticosteroid in cases of typical clinical presentation should be considered. But, as shown in our patient, transsphenoidal surgery should be undertaken in LH cases having remarkable compressive feature, and in whom radiographical or neurological deterioration is observed during

corticosteroids trial^{1,4}). Furthermore, pituitary surgery is also the first line treatment when it is impossible to establish the diagnosis of inflammatory hypophysitis. Consequently, transsphenoidal surgery is both diagnostic and therapeutic option in the management of LH patients. In general, pituitary dysfunctions in LH are the result of cell destruction and rarely improves with treatment¹⁰). Nevertheless, our case exceptionally illustrate that transsphenoidal decompression can offer the chance of a cure with restoration of pituitary function. Even though, it is difficult to determine whether the extent of surgical resection affects outcome of LH series, a radical surgery on inflammatory pituitary lesion is not usually required or desirable. For this case, a sample submitted for frozen section was thought to be consistent with an adenoma, and then all visible tumor was explored and removed during the surgery. However, hypopituitarism may occur or be worsened following extensive surgery in several reports^{4,5}), highlighting the importance of intraoperative confirmation with frozen biopsy and conservative surgical strategy to avoid further resection of potentially viable pituitary tissue. Unlike advanced LH cases who showed complete hypopituitarism and pituitary fibrosis, this patient revealed visual symptom, only pituitary-gonad axis defect, and large pituitary mass. Therefore, clinically, radiologically, and pathologically, this male case may represent acute glandular enlargement and less destructive stage of LH.

Conclusion

The authors report a rare case of LH in young male patient who presented with visual loss and hypogonadism.

This intra- and suprasellar mass masqueraded as a pituitary adenomas and its definite diagnosis was made postoperatively.

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