Therapy for Lymphocytic Adenohypophysitis

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Lymphocytic adenohypophysitis is characterized by inflammation, swelling and destruction of the anterior pituitary. It has been reported as an autoimmune disorder, with other autoimmune diseases, such as, lymphocytic adrenalitis, chronic thyroiditis, subacute thyroiditis, pernicious anemia and diabetes insipidus (1–3). However, if diabetes insipidus is present in this disease, it should be considered that diabetes insipidus represents the neurohypophysitis.

The important factors to determine therapy in lymphocytic adenohypophysitis are accurate diagnosis of the disease, visual disturbance and an increase in intracranial pressure. MRI, some endocrine tests and findings of pituitary antibody are employed to determine the diagnosis.

The endocrine tests are not specific and showed no sign of adenohypophysial deficiency in some cases with stimulation tests, although it was reported that ACTH and TSH secretion were often impaired, and hyperprolactinemia occurred in many cases and moreover, GH response to GRH was low or rarely markedly high, and ACTH response to CRH was low. CRP, WBC are not confirmed as a marker of inflammation and anti-pituitary antibody has also been reported as an unclear marker, although many investigators have reported that specific human antibody is useful, these are not perfect (2, 3). Pituitary biopsy is a clear method, however, it requires a special technique and informed consent of the patients, and the results are not always clear. Only MRI has revealed the change of size and quantities of mass in pituitary. For instance, the homogenous enhancement by gadolinium during the early phase (4) or ring-enhancement (5) on magnetic resonance imaging is highly suggestive of autoimmune hypophysitis. Furthermore, MRI is advantageous to follow the disease course because it can be repeatedly performed.

The treatment has to be selected for each case, among the various approaches. Two therapies are generally considered: operation for decompression of intracranial high pressure by pituitary mass and the steroid pulse therapy (4). Mass with inflammation is not necessarily indicative of surgery. But in most cases with the provisional diagnosis of pituitary adenoma, previously the approach of transsphenoidal surgery was performed. Recently, diagnosis is considered carefully at first, and then conservative therapy is selected, because lymphocytic adenohypophysitis has been well known as a benign disease in some cases with a pituitary mass.

Glucocorticoid pulse therapy and low dose glucocorticoid therapy are presented as conservative treatments. Glucocorticoids can improve the patient’s symptoms. Whether as a low dose or high dose, glucocorticoid therapy is reported to be of benefit. In some cases, therapy with daily doses of 15 to 40 mg glucocorticoid (methylprednisolone equivalent of hydrocortisone and prednisolone) for 2 weeks to 3 months, has been successful in decreasing mass size and in recovering hormonal abnormality by relieving pituitary gland inflammation (6). In other cases improvement occurred within 6 weeks or 12 months after high dose methylprednisolone pulse therapy for 2 weeks (7). Therefore, conservative therapy is considered as a first choice, if the case has no intracranial brain compression and no visual disturbance, because there are numerous complications of surgery, such as hypopituitarism, incomplete surgical resection, or postoperative recurrence.

However even in cases of suprasellar mass and disturbance of visual field, it is reported for lymphocytic adenohypophysitis in remission steroid pulse therapy was successful in decreasing lesion size and in promoting hormonal recovery by reducing lymphocytic infiltration of the pituitary, because clinical features disappear, although the marker of improvement is unknown (8).

The long prognosis in lymphocytic adenohypophysitis is unclear. But spontaneous recovery from hypopituitarism with lymphocytic adenohypophysitis was reported (9, 10). Avoidance of hypopituitarism should be considered for therapy, especially, in young women who hope to give birth. Conservative therapy can prevent the risk of hypopituitarism from pituitary surgery. Surgery must be performed after progression of lymphocytic hypophysitis despite steroid treatment or after the discontinuation of steroid treatment.

In conclusion, in lymphocytic adenohypophysitis in cases with full informed consent, at first, conservative therapy must be tried to decrease the inflammation and pituitary mass, despite intracranial high pressure due to compression of the large pituitary mass.

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See also p 168.
References


