**INFUNDIBULONEUROHYPOPHYSISITIS PRESENTING A LARGE SELlar-JUXTASELLAR MASS: CASE REPORT**

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**BACKGROUND**

Infundibuloneurohypophysitis (INH) is reported to be a self-limiting inflammatory disease involving neurohypophysis. The authors experienced a case of INH presenting a large mass compressing the brain stem.

**CASE PRESENTATION**

The patient exhibited polyuria followed by left hemiparesis and dysarthria lasting a year. Magnetic resonance imaging showed a large sellar mass extending into the right cavernous sinus and prepontine cistern and compressing pons. Endocrinologically, diabetes insipidus was diagnosed and anterior pituitary function was almost normal. Microscopic examination of the surgical specimen obtained by a transsphenoidal route demonstrated diffuse infiltration of lymphoid cells with predominance of B cells over T cells and the granulation tissue. The patient underwent 40 Gy local radiation because of initial misinterpretation of histologic findings as malignant lymphoma and short-term corticosteroid administration.

**CONCLUSION**

The mass gradually shrank and the patient has become neurologically intact in 6 months. At this moment, 67 months after the onset, the patient is free from disease and has no other lesion. INH seems to be a clinical entity possessing a wide spectrum from infundibular tumorlet to an aggressive sellar mass trespassing on surrounding structures. © 2004 Elsevier Inc. All rights reserved.

**KEY WORDS**

Infundibuloneurohypophysitis, diabetes insipidus, MRI.

Infundibuloneurohypophysitis (INH) is an inflammatory disorder usually confined to the posterior pituitary lobe, stalk, and hypothalamus [7]. There are clear-cut differences between symptoms of INH and that of classic lymphocytic adenohypophysitis [2]. INH does not usually cause hypopituitarism but can cause diabetes insipidus, whereas permanent diabetes insipidus has rarely been described as part of the clinical presentation of lymphocytic adenohypophysitis. The inflammatory process in INH is confined to the neurohypophyseal system, forming a mass lesion in the neurohypophysis and infundibulum, and adenohypophysitis was spared on magnetic resonance imaging (MRI).

The authors experienced a case of INH with unusual radiologic feature and unique clinical course, suggesting wide spectrum of this disease. INH appears to be a concept that calls for further consideration.

**Case Reports**

A 46-year-old otherwise healthy female who had two normal full-term deliveries more than 20 years earlier developed polydipsia and polyuria toward the spring of 1995. She suffered from severe headache 1 year after onset. Admission to Hiroshima University Hospital on March 27, 1996, was followed by onset of left hemiparesis, restlessness, and dysarthria. MRI revealed a sellar mass eroding clivus, extending into the right cavernous sinus and suprasellar cistern, and compressing the brain stem (Figure 1). The mucosa of the sphenoid sinus was considerably thickened, which disappeared in a week. A high intensity adjacent to the lesion indicating edema was demonstrated in the brain stem on T2-weighted image (T2WI). Hyperintense signal of the neurohypophysis was absent on T1-weighted sagittal image (T1WI). The right internal carotid angiogram (CAG) demonstrated about 90% stenosis of internal carotid artery (ICA).

Examination of cerebrospinal fluid revealed moderate leukocytosis (cell count: polymorphonuclear granulocyte 104, monocyte 120, unclassified 236).
Basal and reserve function of adenohypophysial hormones, except ACTH secretion, was preserved (ACTH level was 7 pg/ml).

Newly developed symptoms rapidly improved owing to injection of steroids and osmotic diuretics for several days. Biopsy of the lesion via the transphenoidal approach was then performed. The lesion that eroded sellar floor was found fibrous and yellowish. Microscopic examination of the surgical specimen demonstrated the formation of granulation tissue and diffuse infiltration of slightly atypical lymphoid cells. Immunohistochemical staining confirmed the predominance of B cells over T cells. B cells with a little atypia were found with scattered plasmacytes and histiocytes. Necrosis and fibrosis were shown surrounded by inflammatory cells. Angiocentric infiltration or lymphoid cells with a variable appearance, commonly seen in the section of malignant lymphoma, was not observed (Figure 2).

Postoperatively, steroids were given in short-term interval. The lesion, which pathologists initially suggested as being malignant lymphoma, was irradiated with total dose of 40Gy. The patient then underwent superficial right temporal artery (STA)-middle cerebral artery double anastomoses for the right ICA stenosis because the single photon-computed tomography (CT) showed severely compromised cerebral blood flow of the affected hemisphere. The right CAG after the operation confirmed a good patency of STA to angular and posterior temporal artery. Adenohypophysial functions remained normal. Diabetes insipidus gradually improved. Now, 67 months after the treatment, the patient is an active housewife and working part-time. MRI has demonstrated nearly normalization of the size of hypophysis. No other lesion is demonstrated by body CT.

**DISCUSSION**

INH, which was first described by Kojima in 1989, is a distinct clinicopathological entity with involvement of neurohypophysis [10]. Patients with INH present with diabetes insipidus and occasionally hypopituitarism. The etiology of INH is unclear, although autoimmune pathogenesis was suggested in lymphocytic adenohypophysitis [4]. Pathologic findings in our case were consistent with those of previously reported INH and incompatible with malignant lymphoma. The patient is in good clinical condition 67 months after onset. Local recurrence
and other lesions have not been detected. Local 40 Gy irradiation and short-term steroid administration do not usually control malignant lymphoma. The patient’s clinical course also refuted the initial suggestion by a pathologist of malignant lymphoma.

To our knowledge, histologically proven INH has been reported in 17 cases [2,3,5,6,7,9,10,15,18,24,27]. MRI showed an enlargement of pituitary stalk and neurohypophysis, which was generally self-limiting. Even tiny nodular thickening of the stalk with normal hypophysis was reported [7,12], as is seen in our experience (Figure 3). With the advent of high-resolution MRI, INH has been perceived to account for a cause of central diabetes insipidus [21]. Some INH cases with isolated prominent thickening of the pituitary stalk were reported to reveal regression [8]. When diabetes insipidus is the only presenting symptom with characteristic appearance of a thickened enlarged pituitary stalk, observation with follow-up MRI and replacement of antidiuretic hormone may be a better choice.

Hemiparesis and dysarthria were accompanied with diabetes insipidus in our patient, lasting one year. MRI demonstrated the lesion extending into suprasellar cistern, cavernous sinus, and the brain stem. No INH has been reported to be large enough to compress the brain stem or constrict the ICA. But patients with INH were occasionally reported to present with hypopituitarism, suggesting progression of inflammation to the adenohypophysis [2,3,5,15]. Spreading of inflammation to the neurohypophysis is occasionally seen in patients with lymphocytic adenohypophysitis [1,11,13,14,16,17,19,20,23,25,26]. In addition, chronic inflammation of the neurohypophysis and infundibulum that was thought to have been spread from the cavernous sinus has been reported [5]. Even optic neuritis associated with INH has been mentioned [22]. Therefore, sellar and parasellar inflammatory diseases including INH are considered to have potential to spread inflammatory process beyond the physioanatomic border, as is seen in our case. Consequently, these inflammatory diseases may have a wide range spectrum of clinical presentation.

As for treatment, in 10 out of the 16 reported cases which underwent surgical intervention, replacement doses of desmopression acetate was administered after surgery. Steroid therapy following surgery did not improve diabetes insipidus in 6 reported cases. Improvement of severe headache and reduction of mass effect were achieved by steroid treatment in two reported cases [2,27]. In our case, considerable benefit (marked size reduction) with steroids was also obtained. When the inflammation forms a mass large enough to cause neurologic symptoms like in our case, surgical intervention may be a choice for mass reduction. The external irradiation, which was misapplied because of initial diagnosis as malignant lymphoma, might have accelerated the reduction of lesion size. Thus, low-dose irradiation may be a last choice for INH with mass effect that resists steroid therapy and surgical treatment. Inflammatory diseases forming a large sellar and parasellar mass should be ruled out from other pathology including meningioma, malignant lymphoma, chordoma, and malignant neoplasms. Because there are no pathognomonic appearances on MRI or symptomatic characteris-
tics of these conditions, transsphenoidal biopsy is recommended to establish a diagnosis and pertinent treatment plan.

REFERENCES


COMMENTARY

The authors report an unusual case of large pituitary mass diagnosed as infundibuloneurohypophysitis (INH). When these rare lesions are small and a diagnosis of INH is suspected by the clinical presentation and MRI characteristics, medical management is the treatment of choice, with careful follow-up. In those even rarer cases when the mass lesion results in parasellar symptoms, transsphenoidal surgery is necessary to decompress the pathology and establish a definitive diagnosis.

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Response:
First of all, I thank Drs. Ausman and VanGilder very much for accepting my report and for the commentary.

The patient in my report lived a hopeless life because the histological diagnosis was believed to be malignant lymphoma. I had a foreboding of her