Tolosa-Hunt Syndrome with Atypical Intrasellar and Juxtasellar Lesions
— Two Case Reports —

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Summary: Two patients with Tolosa-Hunt syndrome (THS) who had atypical lesions in the intrasellar and juxtasellar regions are reported. They manifested with painful ophthalmoplegia. Magnetic resonance imaging (MRI) commonly showed an increase in the volume of the cavernous sinus occupied by homogeneously well-enhanced lesions in both cases. These lesions extended to the intrasellar and juxtasellar regions with meningeal enhancement. Follow-up MRI after steroid treatment demonstrated normalized or decreased size of the cavernous sinus. These findings suggested nonspecific inflammatory granulomatosis with atypical extension.

Key words: Tolosa-Hunt syndrome, MRI, cavernous sinus, nonspecific inflammatory granulomatosis, meningeal enhancement

Introduction

In 1954, Tolosa reported a patient with ophthalmoplegia caused by nonspecific inflammation in the cavernous sinus. Hunt also reported 6 patients with the same symptoms and proposed the clinical definition of this syndrome in 1961. The characteristic pathological findings were proliferation of the fibroblasts, lymphocytes, and plasma cells within the adventitia of the intracavernous carotid artery. Steroid therapy is generally used for these lesions, but the effect is variable among patients.

The advent of magnetic resonance imaging (MRI) has allowed clear demonstration of the fine details of the cavernous sinus lesions in patients with Tolosa-Hunt syndrome (THS) (Kwan et al. 1987; Kojima et al. 1991) In this report 2 patients with THS who had nonspecific inflammatory granulomatosis involving the pituitary gland and its stalk were described.

Case Report

Case 1

A 46-year-old woman presented with continuous retro-orbital pain and double
vision in August 1994. Neurological examination on admission revealed IIIrd, IVth, and VIth nerve palsies on the right side associated with low grade fever. Hematological examination showed leukocytosis with a white blood cell count of 10600/mm³. The erythrocyte sedimentation rate was 98 mm in 1 h and 124 mm in 2 hs. C-reactive protein was 15.7 mg/100 ml. Lumbar puncture revealed an opening pressure of 160 mm H₂O, with a total protein level of 50 mg/dl and a glucose level of 42 mg/dl. The cerebrospinal fluid (CSF) contained 129 nucleated cells and 80% of them were lymphocytes. Bacterial and fungal cultures were negative. No abnormal findings were noted in hormonal examination.

Precontrast and post contrast routine computed tomography (CT) failed to reveal any organic lesion. However, MRI demonstrated a mass in the right cavernous sinus, which was hypointense to isointense on T1 and T2-weighted

**Fig. 1.** Case 1. T1-weighted MR image (upper) and T2-weighted MR image (lower) on admission, showing enlargement of the bilateral cavernous sinuses and narrowing of the right carotid artery. (arrow head) The signal intensity of the mass lesion in the cavernous sinus is isointense to gray matter.

**Fig. 2.** Case 1. Follow-up T1-weighted MR image with Gd-DTPA enhancement.

a. The initial coronal images demonstrated heterogeneous enhancement in the bilateral enlarged cavernous sinuses, meningeal enhancement (arrow head), and extension to the pituitary gland.

b. At 1 month after starting steroid therapy, a coronal T1-weighted image shows a marked reduction in the size of the mass in the cavernous sinus.

c. At 4 months after starting treatment, the bilateral cavernous sinuses and the pituitary gland have returned to normal.
images (WI). Stenosis of the cavernous portion of the right internal carotid artery was also demonstrated (Fig. 1) and was confirmed by cerebral angiography. MRI with gadolinium diethylene triamine penta-acetic acid (Gd-DTPA) demonstrated heterogeneous enhancement not only the enlarged cavernous sinus but also the pituitary gland. Meningeal enhancement was also shown the cerebral convexity areas (Fig. 2a).

She was treated by 60 mg of prednisone daily for 3 weeks, and then was gradually tapered over 4 weeks. The steroid therapy dramatically improved all her signs and symptoms and they resolved completely after 1 month.

Follow-up MRI after steroid treatment demonstrated a significant decrease in the volume of the mass lesion and improvement of the narrowing of the right internal carotid artery (Figs 2b, c).

Case 2

A 54-year-old woman presented with headache and diplopia in October 1994 and admitted to our hospital 2 weeks later. Neurological examination revealed neck stiffness, complete paralysis of the left eye movement in all directions, and hypoesthesia in the area of the first branch of the Vth nerve. Marked pupillary dilatation and absence of the direct light reflex were noted on the left side. The erythrocyte sedimentation rate was 16 mm in 1 h and 32 mm in 2 hs. C-reactive protein was 3.2 mg/100 ml. Hormonal studies showed low levels of TSH (0.10 μU/ml), F.T3 (2.2 pg/ml), F.T4 (1.0 ng/ml), and ACTH (5 pg/ml). Lumbar puncture revealed an opening pressure of 120 mm H2O, with a total protein of 103 mg/100 ml and a glucose of 12 mg/100 ml. The CSF contained 1214 nucleated cells. Bacterial and fungal cultures were all negative. Systemic investigations failed to show any evidence of collagen diseases, vasculitis, or malignancy.

MRI showed a mass lesion not only

Fig. 3. Case 2. Follow-up T1-weighted MR image.
a. Axial images obtained after gadolinium injection on admission showing enlargement of the cavernous sinus and enhancement of the cerebellar tentorium. (arrow head).
b. Coronal images with Gd-DTPA enhancement obtained on admission demonstrate a nonhomogeneously enhancing lesion which extends to the pituitary gland and enlargement of the pituitary stalk.
c. Coronal images obtained 1 month after treatment. The size of the mass lesion has decreased. The pituitary gland can be seen.
in the left cavernous sinus lesion with low to isointensity on T1WI but also in the pituitary gland and stalk. The pituitary gland was not detected, but the size of the stalk was enlarged. MRI with Gd-DTPA demonstrated nonhomogeneous enhancement of the cavernous sinus extending to the intrasellar region along the edge of the cerebellar tentorium posteriorly (Figs 3a, b).

Steroid therapy, hormone replacement therapy and antibiotics were then started (60 mg of predonine, 100 μg of T4 and 4 g of PIPC daily). The steroid was tapered over 4 weeks. The right VIth nerve palsy and hypopituitarism slightly improved, but the left IIIrd and Vth nerve palsies remained.

Follow-up MRI one month later showed a decrease in the size of the sellar to juxtasellar lesion and enhancement of the meninges had disappeared (Fig. 3c).

Discussion

MRI is quite useful for evaluating lesions around the cavernous sinus. The MRI features of THS appear to be similar to those of sarcoidosis, meningioma, lymphoma, lymphocytic hypophysitis, and Wegener’s granulomatosis (Hayes et al. 1987; Chang et al. 1990; Sherman and Stern, 1990; Nussbaum et al. 1991). Several MR studies of THS have been reported (Goto et al. 1990; Yousem et al. 1990; Desai et al. 1991; Oda et al. 1991; Zournas et al. 1995). But in some patients with this syndrome, the clinical symptoms are atypical and the mass lesions are not always localized to the cavernous sinus. They can extends into the posterior ethmoid sinus, the orbital apex, and/or the parasellar region (Kojima et al. 1991; Imai et al. 1995). Nishihara et al. (1994) reported of the MRI findings of 6 patients with a nonspecific inflammatory process in the parasellar region. They described isointense lesions in the cavernous sinus on T1WI with variable intensity on T2WI. These lesions are usually enhanced by Gd-DTPA and extend to the orbital apex, the infratemporal fossa, or the cerebellar tentorium in some cases. In our two patients, the inflammatory granulation detected by MRI were not only located in the intrasellar and juxtasellar regions but also in the surrounding dura mater. The original inflammation in the cavernous sinus therefore might have extended into the surrounding dura mater in the cranial base.

Nonspecific inflammation also involve other than the cavernous sinus (Flanders et al. 1989; Atkin et al. 1990). Orbital pseudotumor is also known to extend to the cavernous sinus and cerebellar tentorium (Bourruat et al. 1995). Therefore, they may have a similar inflammatory processes in the mode of involvement at the skull base.

According to previous reports on nonspecific parasellar inflammation including THS, steroid therapy is effective for improving symptoms and signs. In our Case 1, steroid therapy markedly improved the symptoms, but in Case 2 the effect was limited and the mass lesion remained on follow-up MRI. Thus, the effect of steroid treatment is variable among patients with THS. Review of follow-up MRI findings in 20 cases indicates that the mass lesion in the cavernous sinus persisted in 17 cases.
TABLE 1.
Follow-up MRI findings and recovery of symptoms in patients with nonspecific inflammatory processes involving the cavernous sinus region

<table>
<thead>
<tr>
<th>Author &amp; Year</th>
<th>No. of Cases</th>
<th>Follow-up period</th>
<th>Size of cavernous sinus</th>
<th>Complete recovery</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td>no change</td>
<td>decreased</td>
</tr>
<tr>
<td>Yousem 1989</td>
<td>3</td>
<td>not described</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td>Goto 1990</td>
<td>3</td>
<td>not described</td>
<td>0</td>
<td>3</td>
</tr>
<tr>
<td>Oda 1991</td>
<td>1</td>
<td>19 months</td>
<td>–</td>
<td>1</td>
</tr>
<tr>
<td>Kojima 1991</td>
<td>3</td>
<td>15 days - 1 month</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td>Desai 1991</td>
<td>1</td>
<td>15 weeks</td>
<td>–</td>
<td>1</td>
</tr>
<tr>
<td>Nishihara 1994</td>
<td>6</td>
<td>1 month - 2 years</td>
<td>2</td>
<td>4</td>
</tr>
<tr>
<td>Imai 1995</td>
<td>2</td>
<td>1 year</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Zournas 1995</td>
<td>1</td>
<td>10 mouths</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>Takahashi 1995</td>
<td>2</td>
<td>1 month - 4 months</td>
<td>0</td>
<td>1</td>
</tr>
</tbody>
</table>

The limited effect of steroids may be influenced the degree of the inflammation in the cavernous sinus. Cyclophosphamide was reported to be effective in a patient with an orbital pseudotumor who was unresponsive to steroid (Bourruat et al. 1995). Cyclophosphamide may be a treatment of choice for steroid-resistant disease.

Neurological recovery is not always correlated with the improvement of radiological findings. Follow-up study by MRI is needed to detect the recurrence of nonspecific inflammation in the skull base if the size of the lesion was temporarily decreased by the treatment.

References


Flanders AE, Mafee MF, Rao VM, and Choi KH. CT characteristics of orbital pseudotumor and other orbital inflammatory process. J


