
Gadopentetate Dimeglumine-Enhanced MR in the Diagnosis of the Tolosa-Hunt Syndrome

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Summary: A 54-year-old man first was admitted with a right oculomotor nerve palsy that ameliorated spontaneously. Two months later, he was readmitted with right proptosis, ophthalmoplegia, and optic nerve involvement. MR showed an enlarged right cavernous sinus. There was dramatic improvement after high doses of steroids. MR findings 10 months later were normal. Thus, the diagnosis of the Tolosa-Hunt syndrome was established.

Index terms: Eyes, inflammation; Cavernous sinus, magnetic resonance

Syndromes of the cavernous sinus and the orbital apex, although very well specified by clinical findings, are often difficult to differentiate etiologically. This is attributable to the great variety of diseases that affect this region, the position of which makes histologic confirmation both difficult and dangerous. One of these syndromes, characterized by painful acute or subacute ophthalmoplegia, with or without involvement of the optic nerve and the ophthalmic division of the trigeminal nerve, which are promptly responsive to steroids, is the Tolosa-Hunt syndrome. According to Tolosa this is attributable to a nonspecific inflammation in the cavernous sinus area, sometimes extending to adjacent structures (orbit) (1, 2). Before computed tomography (CT) it often was a diagnosis of exclusion. Modern high-resolution CT and magnetic resonance (MR) images have been able to demonstrate soft-tissue abnormalities in some of these cases (3–6). A case of Tolosa-Hunt syndrome diagnosed with only MR imaging with gadopentetate dimeglumine enhancement is presented.

Case Report

A 54-year-old man first was admitted to our clinic in February 1991 with a 4-week history of right-sided frontal

and retroorbital boring pain, followed by ptosis and diplopia. The only finding on physical examination was a right oculomotor nerve palsy.

The laboratory data, including complete blood count, erythrocyte sedimentation rate, blood urea and sugar, electrolytes, liver enzymes, collagen disease tests, coagulation profile, triiodothyronine, thyroxine, thyrotropin, and serologic tests for syphilis, all were within normal limits. The cerebrospinal fluid routine examination and cerebrospinal fluid culture did not reveal any pathologic findings. Tests for myasthenia also were normal. The electrocardiogram, electroencephalogram, skull x-rays, brain CT scan, MR imaging, and digital arteriogram revealed no abnormalities. The only pathologic finding was a slightly abnormal glucose tolerance test.

Vitamins and gangliosides were recommended as short-term therapy, because the patient was considered to have peripheral neuritis with spontaneous amelioration of his symptoms.

Two months later, the patient was readmitted with right proptosis and rapidly deteriorating visual acuity. The clinical examination revealed a right exophthalmos with affection of ocular motility to all directions. No direct light reflex was elicited, and a serious diminution of visual acuity was found ipsilaterally. There were no other pathologic findings. Routine laboratory and x-ray tests remained normal. The new brain MR showed enlargement of the right cavernous sinus. The lesion was isointense with muscle on short-repetition-time/short-echo-time images (Fig 1A). After the intravenous injection of paramagnetic agent (20 mL gadopentetate dimeglumine), there was enhancement of the enlarged cavernous sinus (Fig 1B). There was extension into the orbital apex (Fig 1C).

The patient was treated for 2 weeks with high-dose prednisolone (100 mg/d). There was dramatic improvement within 24 hours. In the first week, ocular motility returned to normal, no exophthalmos was detected, and visual acuity from 1/10 was found to be 6/10. Steroid therapy was continued for 2 months with gradual reduction. The patient has been asymptomatic since. In February 1992, new brain MR findings with gadopentetate dimeglumine were normal (Fig 1D).

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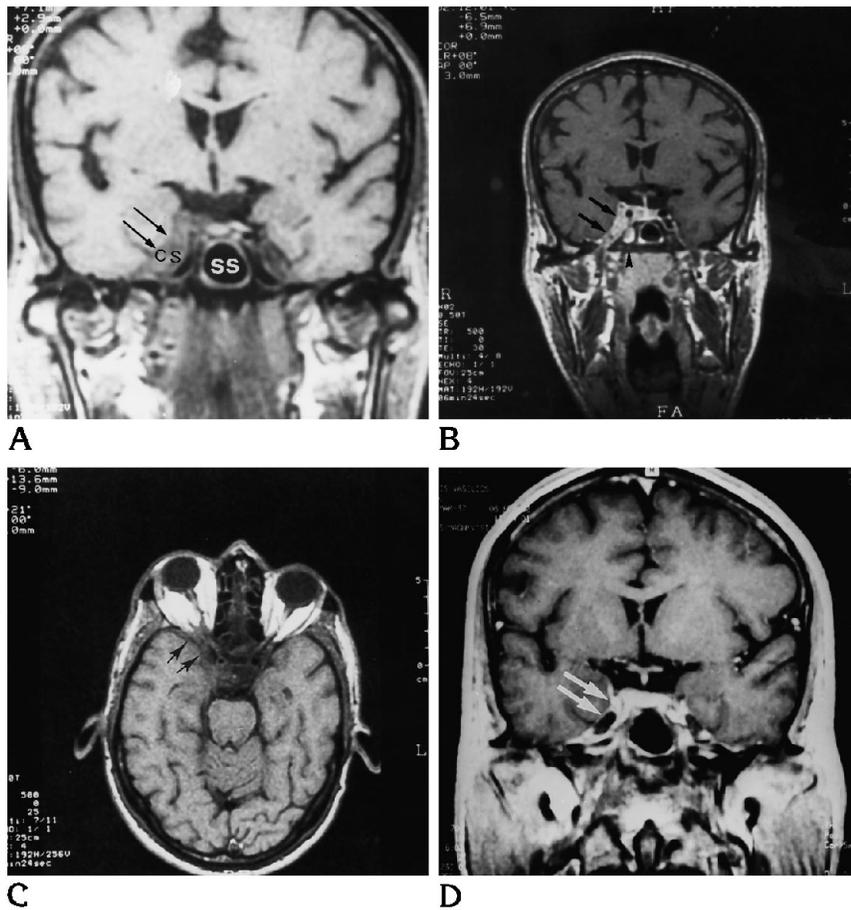


Fig 1. Coronal MR images (500/30/4 [repetition time/echo time/excitations]) before (A) and after (B) intravenous administration of the gadopentetate dimeglumine show widening of the cavernous sinus (arrows). Note mucosal thickening in the sphenoid sinus. There also is opacification of the air cell in the right side of the sphenoid sinus, contiguous with the abnormal cavernous sinus (arrowhead). CS indicates cavernous sinus; SS, sphenoid sinus.

C, Axial T1-weighted image (500/25/4) before contrast enhancement shows extension of the inflammatory abnormality into the orbital apex (arrows).

D, Coronal MR images (500/10/2) after intravenous administration of gadopentetate dimeglumine show normal appearance of the right parasellar region, after treatment (arrow).

Discussion

Our patient fulfills the criteria of Tolosa-Hunt syndrome, namely, recurrent painful ophthalmoplegia, optic nerve involvement, and dramatic response to high-dose steroid therapy with rapid clearing of pain and improvement of vision. It is important to recognize that many of the features of the syndrome develop with other types of pathologic processes occurring in the same region. The clinical improvement after steroid therapy is not absolute proof because lymphoma, meningioma, and giant-cell tumors also respond symptomatically to steroids, although with some delay, but signs do not resolve (7). Other differential diagnoses in this case include diabetic ophthalmoplegia caused by ischemic optic nerve disease, which is relatively unresponsive to steroids. This diagnosis has no radiologic evidence of inflammation and requires history of diabetes (8). Aneurysm of the cavernous portion of the internal carotid

artery can be excluded by MR, CT, and standard angiography. Tumors of the nasopharynx are excluded by CT and MR images and physical examination.

In our patient, opacification of the air cell in the lateral sphenoid sinus contiguous with the abnormal cavernous sinus on the initial image should be explained as secondary reaction to the nonspecific inflammatory syndrome. That this improved on steroids rather than on antibiotics does go along with the diagnosis of Tolosa-Hunt syndrome.

The lesion suspected clinically was eventually demonstrated on gadolinium-enhanced MR imaging and resulted not only in the resolution of clinical signs, but also in resolution of the MR finding. Because the prognosis of this condition is benign, the importance of differentiating this from other lesions in the area, especially with noninvasive and harmless procedures, is obvious for the patient and his family.

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