Tolosa-Hunt Syndrome
Preceded by Facial Palsy

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The Tolosa-Hunt syndrome consists of a painful ophthalmoplegia related to a granulomatous inflammatory process in the cavernous sinus, which may be documented by cerebral magnetic resonance imaging with gadolinium enhancement.

Two cases of Tolosa-Hunt syndrome preceded by facial palsy observed in 1998 at the Department of Neurosurgery of the Second University of Naples are presented here. Both patients developed Tolosa-Hunt syndrome following an ipsilateral facial palsy that resolved in about 15 days with medical treatment. Cerebral magnetic resonance imaging with gadolinium enhancement showed, in both cases, inflammatory tissue in the cavernous sinus. The patients underwent corticosteroid therapy (prednisolone, 80 mg per day, intravenously) with pain regression. In the first case, the patient experienced recurrence of the syndrome that was definitively resolved with further corticosteroid treatment.

The rare reports of facial palsy in patients with Tolosa-Hunt syndrome suggest the inclusion of this disease in the so-called multiple cranial nerve palsy syndrome. It is probable that Tolosa-Hunt syndrome has an inflammatory pathogenesis.

Key words: Tolosa-Hunt syndrome, ophthalmoplegia, cavernous sinus, magnetic resonance imaging, corticosteroid treatment, headache

Abbreviations: THS Tolosa-Hunt syndrome (Headache 2000;40:393-396)
rosurgery of the Second University of Naples in 1998 are presented here.

CASE HISTORIES

Patient 1.– A 25-year-old man presented with a left, peripheral, facial nerve palsy. Cranial computed tomography (CT) with contrast enhancement did not show any intracranial abnormality. Treatment with corticosteroids and neurotrophics was followed by regression of the palsy in about 15 days. After 2 weeks, the patient was again admitted to our department complaining of a severe and continuous, left, frontotemporal headache that was refractory to common analgesic therapies. The findings of the neurological examination were normal. Approximately 7 days after the onset of the pain, the patient developed a partial, left, oculomotor nerve palsy with diplopia, divergent strabismus, and ptosis of the left eye; he also had proptosis and conjunctival chemosis. Cerebral MRI with gadolinium enhancement showed an inflammatory tissue mass in the left cavernous sinus, isointense to gray matter on T1-weighted images and isointense on T2-weighted images without contrast enhancement (Figures 1 and 2). Magnetic resonance angiography excluded vascular malformations. Therefore, corticosteroid treatment (prednisolone, 70 mg per day intravenously) was initiated with pain regression in 48 hours and resolution of the cranial nerve palsies in approximately 2 weeks. The patient was discharged on the 20th day, with no evidence of recurrence 1 year after the onset of the disease.

Patient 2.– A 58-year-old woman was admitted to our department because of the abrupt onset of pulsating, left, fronto-orbital headache followed by diplopia. Neurological examination showed left peripheral-type facial nerve palsy and partial ipsilateral abducens nerve palsy. The findings of cranial enhanced CT were negative. Cerebral MRI with gadolinium revealed the presence of enhancing tissue in the left cavernous sinus, isointense to gray matter on T1-weighted images and isointense on T2-weighted images. No systemic signs of granulomatous disease were found. Magnetic resonance angiography excluded vascular malformations. Therefore, corticosteroid treatment (prednisolone, 70 mg per day intravenously) was initiated with pain regression in 48 hours and resolution of the cranial nerve palsies in approximately 2 weeks. The patient was discharged on the 20th day, with no evidence of recurrence 1 year after the onset of the disease.

COMMENTS

Tolosa-Hunt syndrome is a rare cause of painful ophthalmoplegia. Since 1954, when Tolosa first described this syndrome,12 about 270 cases have been

![Fig 1.—Cerebral MRI. T1-weighted axial image demonstrating soft tissue isointense to gray matter in the left cavernous sinus, with narrowing of the intracavernous portion of the internal carotid artery.](image-url)
Clinical criteria for the diagnosis of THS were suggested by Hunt et al in 1961 and reviewed by Hannerz in 1992. This syndrome has an age of onset ranging from 3 to 75 years and occurs equally in both sexes. It is bilateral in 5% of cases, probably due to the extension of a granulomatous process to the contralateral cavernous sinus. The third cranial nerve is involved in 85% of cases, the sixth cranial nerve in 70% of cases, and the fourth cranial nerve in 29% of cases; the first (ophthalmic) trigeminal branch is involved in 30% of cases, with hypoesthesia and absence of the corneal reflex. Involvement of pericarotid sympathetic fibers causes proptosis by means of contraction of Müller’s orbital muscle.

Tolosa-Hunt syndrome should be differentiated from other more frequent causes of painful ophthalmoplegia, such as ophthalmoplegic migraine, diabetic ophthalmoplegia, giant cell temporal arteritis, meningiomas, lymphomas or parasellar tumors, or vascular malformations of the posterior communicating artery or intracavernous carotid artery. Systemic signs of granulomatous-specific diseases (syphilis, tuberculosis, Wegener granulomatosis) should be sought.

Some authors have reported positive serum antineutrophil cytoplastmic antibodies in patients with THS, suggesting that the condition could be an atypical form of Wegener granulomatosis exclusively located in the cavernous sinus.

Cerebral MRI with gadolinium is the examination of choice in THS, because it shows inflammatory tissue in the cavernous sinus, excluding other cavernous sinus lesions. Alvarez de Arcaya et al have recently stated the MRI criteria for the diagnosis of THS. Magnetic resonance imaging should show enlargement of the affected cavernous sinus, with the outer dural margin convex and bulging laterally; abnormal soft tissue surrounding the internal carotid flow void should be identified, with focal narrowing of the intracavernous portion of the internal carotid artery. It is important to note that a small number of patients might have a normal MRI examination. Magnetic resonance imaging can also exclude acute Bell palsy, in which the facial nerve enhanced after gadolinium administration; neither of our cases showed facial nerve enhancement. Furthermore, the clinical resolution of symptoms often precedes by weeks or even several months the normalization of MRI studies. Cranial CT is normal in almost all patients with THS, though high-resolution CT might show the presence of inflammatory tissue in the cavernous sinus or superior orbital fissure. Diagnosis of THS can be confirmed by performing an orbital phlebography. It shows narrowing or occlusion of the third segment of the superior ophthalmic vein and partial occlusion of the cavernous sinus in 68% of patients, possibly due to venous vasculitis. Indeed, a biopsy from an eye muscle of a patient with THS showed venous vasculitis, probably indicating the basic pathology behind the phlebographic changes in patients with THS.

CONCLUSIONS

The Tolosa-Hunt syndrome is very interesting because of its rarity and because it features in the differential diagnosis of many neurosurgical and neurological diseases. Furthermore, to our knowledge, only 11 cases of THS accompanied by facial palsy have been reported. Among these cases, only 1 patient had THS documented by MRI. Therefore, our cases rep-
resent the second and the third examples of documented THS associated with facial palsy.

Cerebral MRI with gadolinium enhancement is the most valuable imaging technique for demonstrating lesions in the cavernous sinus that are directly responsible for the symptoms of THS.

REFERENCES


