Tolosa-Hunt syndrome

Ben Barnard, *MB ChB*
Delme Hurter, MB ChB, MMed (D Rad)
Department of Radiology, Kimberley Hospital Complex, Kimberley

Francois Roux, *MB ChB, DA (SA)*
Tygerberg Academic Hospital, University of Stellenbosch, Tygerberg

Shaheer Aboobaker, *MB ChB, Dip Opth (SA)*
Department of Ophthalmology, Kimberley Hospital Complex, Kimberley

Corresponding author: *B Barnard (benwbarnard@gmail.com)*

Introduction

Tolosa-Hunt syndrome (THS) is a rare disorder indicated by recurrent painful ophthalmoplegia caused by non-specific inflammation of the cavernous sinus or superior orbital fissure (SOF). The disease shares histopathological features with idiopathic orbital pseudotumour; however, owing to its anatomical location, it produces characteristic clinical manifestations.[1] Recurrent retro-orbital pain, with palsies of the third, fourth or sixth cranial nerves as well as the first and second divisions of the trigeminal nerve, are typical. Clinically, immediate response to steroid therapy is a hallmark of the condition.

The clinical presentation of THS has a wide differential diagnosis, and timely and appropriate imaging – as an adjunct to pertinent laboratory investigations – can greatly assist clinicians with early accurate diagnosis and management.

Case report

Our patient was a 17-year-old girl who presented with a 2-week history of a sharp peri-ocular headache and drooping of the left eyelid. She also complained of a decrease in eye movement as well as visual acuity. She indicated that she had had similar episodes of headache preceded by blurring of vision in the same eye for more than a year. She was previously healthy.

On clinical examination, her vitals were normal. She had a left-sided ptosis and impairment of adduction and elevation of the left eye, consistent with an oculomotor (IIIrd) nerve palsy. The left pupil was mildly dilated but responsive to light. No facial sensory or motor loss in the distribution of the trigeminal (Vth) nerve was detected. Fundoscopy was normal.

Bloodwork and lumbar puncture (LP) were non-specific. Of note, the white cell count (WCC) was mildly elevated and the erythrocyte sedimentation rate (ESR) significantly increased. She tested retroviral negative.

The patient was referred for an urgent brain CT; this showed a diffusely thickened left optic nerve with associated dilatation of the left superior ophthalmic vein (SOV). In addition, a soft-tissue mass was noted in the SOF, extending into the left cavernous sinus. An MRI of the brain and orbits was then performed on a 1.5T Toshiba Vantage machine. Whole-brain FLAIR and T2W imaging was done, followed by thin-slice (2.5 mm slice thickness) coronal, axial and sagittal CE (Magnevist) FS T1W sequences through the cavernous sinus and orbits. This demonstrated an avidly enhancing mass lesion in the orbital apex extending into the left SOF, causing compression on the neurovascular elements. The lesion extended into the cavernous sinus. The diffusely thickened optic nerve was secondary to oedema but was not infiltrated. There was no luminal narrowing of the intracavernous segment of the internal carotid artery (ICA), and no evidence of cavernous sinus or SOV thrombosis. The rest of the study was non-contributory.

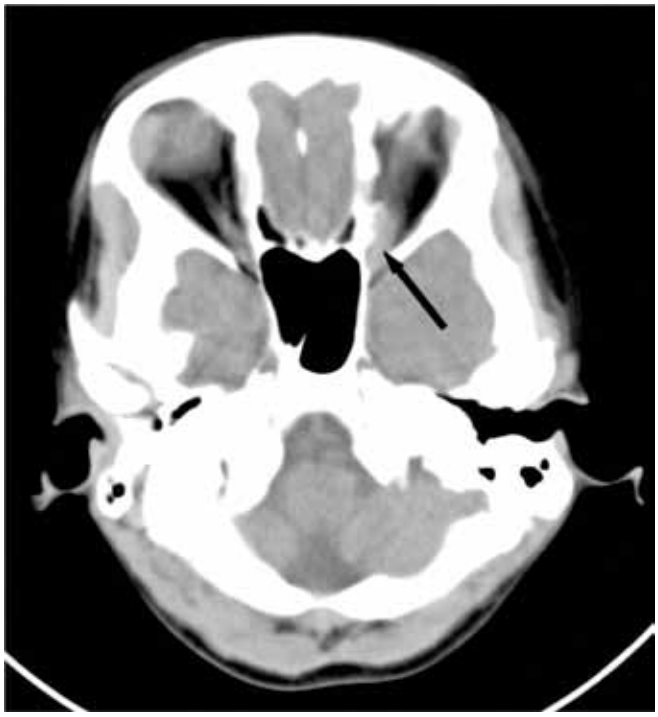

Fig. 1. Axial CT brain demonstrates a soft-tissue mass in the left superior orbital fissure. Note also the thickened optic nerve.

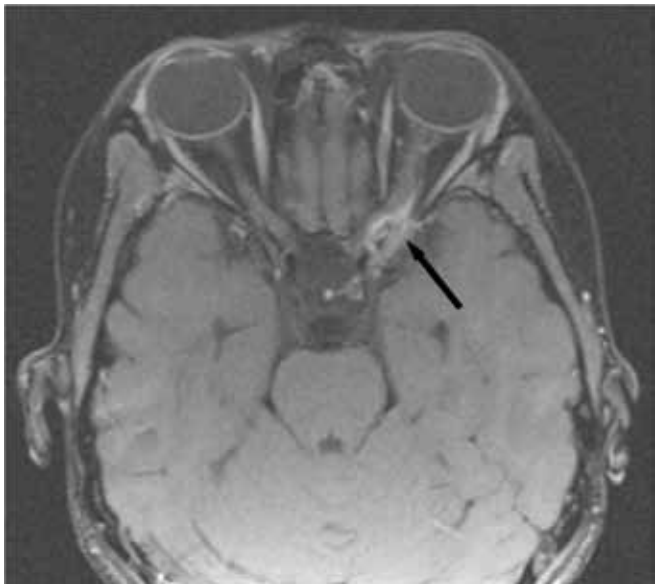

Fig. 2. Axial CE FS T1W image reveals intense enhancement of the soft-tissue mass within the left SOF. The lesion extends into the cavernous sinus.

The patient was treated with oral steroids and showed significant relief of symptoms over the following 48 hours. She had complete resolution of the ptosis as well as the oculomotor nerve palsy within one month. Further follow-up showed no recurrence of symptoms.

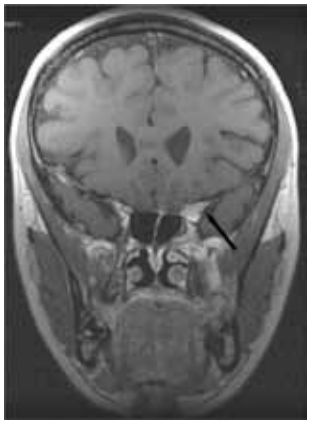

Fig. 3. Coronal CE FS T1W image again shows the intensely enhancing soft-tissue mass.

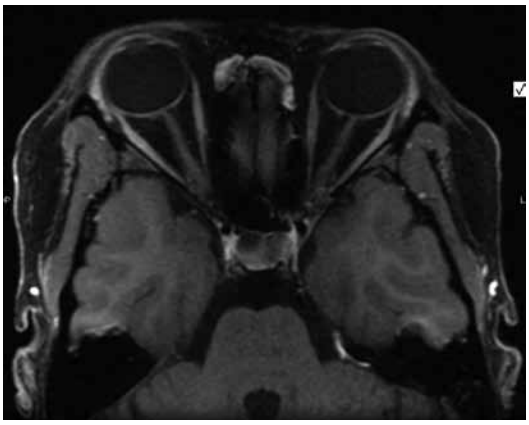

Fig. 4. Axial CE FS T1W follow-up image 4 months later demonstrates complete resolution of the enhancing soft-tissue mass.

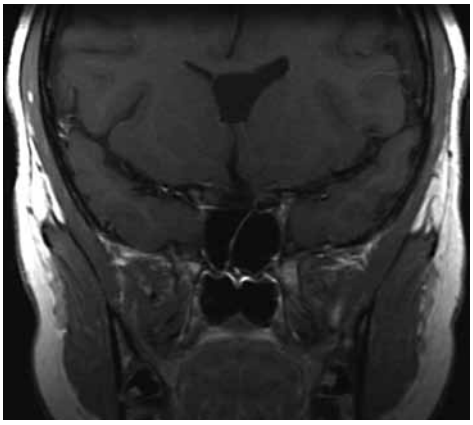

Fig. 5. Coronal CE FS T1W follow-up image again shows the resolution of the mass.

Discussion

Tolosa first described the condition in 1954, in a patient with unilateral recurrent painful ophthalmoplegia involving cranial nerves III, IV, VI and V_1. The patient was imaged using carotid angiography, and segmental narrowing of the carotid siphon was seen.[1]

Hunt *et al.* described 6 patients with similar clinical findings in 1961, and proposed a low-grade non-specific inflammation of the cavernous sinus and its walls as the cause of the syndrome. Pathologically, infiltration of lymphocytes and plasma cells as well as thickening of the dura mater was seen.[1] The condition was termed Tolosa-Hunt syndrome by Smith and Taxdal in 1966.[2] The latter authors stressed the importance of the dramatic rapid response to steroid therapy.

In 1988, THS criteria were provided by the International Headache Society (IHS), and further revised in 2004 (Table I).[3,4]

Table I. THS diagnostic criteria

A	One or more episodes of unilateral orbital pain persisting for weeks if untreated
B	Paresis of one or more of the third, fourth and/or sixth cranial nerves and/or demonstration of granulomas by MRI or biopsy
C	Paresis coincides with the onset of pain or follows it within 2 weeks
D	Pain and paresis resolve within 72 hours when treated adequately with corticosteroids
E	Other causes have been excluded by appropriate investigations

Neuro-imaging – in particular MRI – is an essential part of the workup of any patient presenting with features of THS, as these features are non-specific and have a wide differential diagnosis, including meningioma, sarcoidosis, pituitary tumours, tuberculous meningitis (TBM) and lymphoma.[2]

MRI findings classically demonstrate a soft-tissue mass lesion involving the SOF or cavernous sinus. Signal characteristics are typically hypointense to fat and isointense to muscle on short TR/TE sequences and isointense to fat on long TR/TE sequences.[5] Significant enhancement of the mass lesion is demonstrated on CE sequences. Of particular value is the post-contrast fat-saturated thin-slice coronal images through the orbital apex and cavernous sinus.

THS essentially remains a diagnosis of exclusion. The role of the radiologist is to exclude other conditions causing similar clinical features. Distinctive MRI findings and rapid resolution of clinical symptoms with steroid therapy are characteristic.

1. Som PD, Curtin HD. Head and Neck Imaging. 4th ed. St Louis: Mosby, 2003:587-591.
2. Sathyanathan BP, Rajasundaram R, Sankaravadivelu ST, Nadhamuni K. A case of Tolosa-Hunt Syndrome – MR imaging appearance. Ind J Radiol Imag 2006;16(1):97-98.
3. Mendez JA, Arias CR, Sanchez D, et al. Painful ophthalomoplegia of the left eye in a 19-year-old female, with an emphasis on Tolosa-Hunt syndrome: a case report. Cases Journal 2009;2:8271.
4. Mora-de-Onate J, Pascual-Perez-Alfaro R, Izquierdo-Vazquez C, Gonzalez-Ruiz M, Aguirrebena-Olmos A, Diez-Villalba R. Painful ophthalmoplegia (pseudotumour of the orbit and Tolosa-Hunt Syndrome). Arch Soc Esp Oftalmol 2007;82:509-512.
5. Yousem DM, Atlas SW, Grossman RI, Sergott RC, Savino PJ, Bosley TM. MR imaging of Tolosa-Hunt Syndrome. AJNR 1990;154:167-170.
6. Dornam TL, Espir MLE, Gale EAM, Tattersall RB, Worthington BS. Remittent painful ophthalmoplegia: the Tolosa-Hunt syndrome? J Neurol Neurosurg Psychiatry 1979;42:270-275.