

## A Rare Case of Tolosa Hunt Syndrome

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### ABSTRACT

Tolosa Hunt syndrome (THS) is a rare cause of painful ophthalmoplegia. It is caused by a nonspecific inflammatory process, of unknown etiology, involving the cavernous sinus, the superior orbital fissure and/or the orbital apex. It is classically described as an episodic orbital pain associated with paralysis of one or more of the 3rd, 4th and 6th cranial nerves, resolving spontaneously or with the commencement of steroids. We report a female patient presented with complain of severe headache from one month, nausea and retro-orbital pain.

### Introduction :

Tolosa Hunt syndrome (THS) is a rare cause of painful ophthalmoplegia. It has an estimated annual

incidence 1 case per million per year. It is characterized by nonspecific granulomatous inflammation of the cavernous sinus, superior orbital fissure and / or the apex of the orbit. Clinically patient presents with Painful ophthalmoplegia characterized by multiple cranial nerve palsies. Various etiologies have been reported; these include infections, inflammations, sphenoid sinus mucocele, tumors, dural arteriovenous

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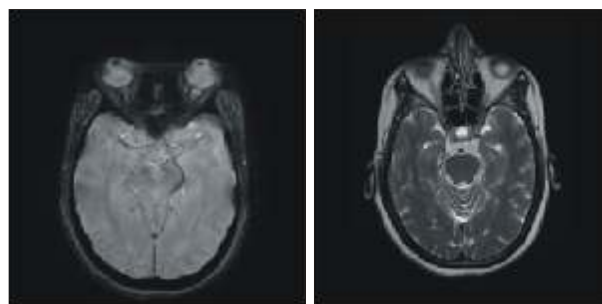
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malformation, trauma, and diabetes mellitus. In 2004, the International Headache Society (IHS) redefined the diagnostic criteria of THS, specifying that granuloma, demonstrated by MRI scan or biopsy, is required for diagnosis.<sup>1,2</sup> Following criteria were proposed

- A. One or more episodes of unilateral orbital pain persisted 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 MRI scan or biopsy is required for diagnosis.

#### Case Report :

A previously healthy, 42 year old female presented with left sided retro-orbital pain and frontal headache since 2 months, increased since 5 days associated with nausea and vomiting. It was followed by ptosis of left eye, diplopia and numbness of upper part of left side of face. The patient denied history of similar complaints in the past, facial or head trauma and drug intake. On examination, there was ptosis of left eye and loss of adduction and abduction, slight upward movement of left eye globe with exophthalmos and loss of sensation in the area of ophthalmic branch of trigeminal nerve. Pupils were mid-dilated but reacting to light. The visual acuity and fundoscopy were normal in both eyes. All other neurologic and general examinations were normal. In lab parameters her Erythrocyte sedimentation rate (ESR) was slightly elevated (25 milliliter / 1st hour). Thyroid function test, fasting blood sugar, kidney function test and complete blood count were normal. CSF examination was normal. MRI Brain revealed an altered intensity lesion in left cavernous sinus that approaches left cavernous carotid artery, extending till superior orbital fissure, appearing isointense on T1W1 and hyperintense on T2W2 & FLAIR. Lateral border of affected cavernous sinus was convex.



**Figure 1 : MRI picture in our patient**

#### Diagnosis :

On the basis of clinical presentation, imaging and laboratory results she was diagnosed to have Tolosa hunt syndrome.

Patient was started on high dose prednisolone (1mg/kg/day). On 3rd day of therapy the patient's headache disappeared completely with partial improvement of ptosis and ophthalmoplegia. but left eye ptosis and ophthalmoplegia required 3 weeks to improve.

#### Before treatment



#### After treatment



#### Discussion :

Tolosa-Hunt syndrome (THS), is characterized by nonspecific granulomatous inflammation of the cavernous sinus, superior orbital fissure and/or the apex of the orbit. THS usually presents with recurrent periorbital pain, headaches on the same periorbital side and diplopia. Clinical signs include slight exophthalmos, extra ocular palsies involving the third, fourth, and sixth cranial nerves and numbness in the area of V1 and V2 branch of the trigeminal nerve<sup>2</sup>.

THS is usually reported as unilateral, while in few it can have bilateral affection .Uniformly, patients

complain of pain, which is a defining symptom. The pain lasts an average of 8 weeks if untreated. Ocular motor cranial nerve palsies may coincide with the onset of pain or follow it within a period of up to 2 weeks. The Pupillary reactions may be normal. The 3rd nerves is involved in 85% of cases, the 6th nerve in 70%, the ophthalmic branch of 5th nerve in 30% and the 4th nerve in 29%. Sympathetic innervation of the pupil is occasionally affected. Facial palsy is not uncommon and systemic symptoms, as back pain, arthralgia, chronic fatigue and gastrointestinal upset, are also noted<sup>3</sup>. Clinically, painful ophthalmoplegia and immediate response to steroid therapy are a hallmark of the condition. The aetiology is unknown, although it shares histopathological features with idiopathic orbital pseudo-tumour. However, owing to its anatomical location, it produces characteristic clinical manifestations. Over the years, there have been cases reporting the involvement of cranial nerves outside the cavernous sinus and some authors raised the possibility that the syndrome is in fact part of a larger spectrum of idiopathic recurrent cranial neuropathy. The condition can be classified, according to neuroimaging, into benign (when no abnormal neuroimaging can be found), inflammatory (when inflammatory findings are shown on MRI or biopsy) and symptomatic (when neuroimaging reveals specific lesion). Treatment should be with high dose steroids (1 mg/kg/d) tapered slowly over 3 to 4 months. It can affect people of age group of 1st to the 8th decades of life, with no sex predilection<sup>4</sup>.

A review of the literature by La Mantia L et al<sup>5</sup> on THS from 1988 to 2002, analysed individual cases in relation to the new IHS criteria. They identified 124 cases. It appeared that clinical presentation was similar in all, but 44 (35%) were reported to have

inflammation on MRI or biopsy evidence of granuloma, 41/124 (33%) had normal neuroimaging findings and 39 (31%) had a specific lesion, so the THS was secondary. This confirms that clinical criteria for THS are not unique and their application alone does not assure a correct diagnosis. The requirement for inflammation on MRI will result in better classification of painful ophthalmoplegias<sup>5</sup>. Other features specifically highlighted by the Society are: If left untreated, the painful episodes typically last eight weeks. Treated, the pain responds to steroids within 48 hours. Cranial nerve involvement coincides with the pain or occurs no more than two weeks after the onset of pain. Other conditions should be excluded by neuroimaging ( $\pm$  angiography)<sup>2</sup>.

#### Conclusion :

THS is a rare disorder painful ophthalmoplegia. It is extremely important to have high index of suspicion. MRI is necessary for diagnosis of THS. If treated early in the course of disease with high dose corticosteroids, patient responds dramatically.

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