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# Tolosa-Hunt Syndrome in a Kenyan Patient: Case Report and Review of the Literature

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**Abstract:** Tolosa-Hunt syndrome is a rare cause of painful ophthalmoplegia accompanied by ipsilateral ocular motor palsies, oculosympathetic paralysis and peri-orbital pain around the distribution of the ophthalmic division of the trigeminal nerve. Though the cause is unknown, idiopathic inflammation has been implicated. The pathology can be localized in the cavernous sinus, superior orbital fissure or the apex of the orbital cavity. Localization of the pathology requires a combination of careful history, physical examination and imaging, preferably MRI. We present a 40 year old Kenyan female patient with Tolosa-Hunt syndrome. She presented with left-sided periorbital headache and ptosis. Further examination revealed left-sided trochlear and abducens palsy, and partial left-sided oculomotor palsy. She had allodynia and hyperesthesia on the distribution of the left ophthalmic division of the trigeminal nerve. After an initial normal head CT scan, a magnetic resonance imaging revealed a soft-tissue hyperintensity extending from the left cavernous sinus, superior orbital fissure and apex of the orbit. She responded positively to steroid therapy, with cessation of headaches, correction of ptosis and ophthalmoplegia and an improved quality of life. There was minimal improvement in vision, with persisting pallor of the disc. This case shines the spotlight on a relatively rare disorder; whose diagnosis requires careful history, examination and interpretation of MRI findings.

**Keywords:** Tolosa-Hunt, Ophthalmoplegia, Ptosi, MRI

## 1. Introduction

Tolosa-Hunt Syndrome (THS) is a rare disorder, estimated to occur in one case per million per year [1]. It is characterized by painful unilateral ophthalmoplegia, thought to occur due to idiopathic inflammation of the cavernous sinus and surrounding structures in the superior orbital fissure [2]. It has a similar incidence between men and women and can occur in any age group between the first and eighth decades of life.

Though mostly unilateral, the symptoms may be bilateral. The most common complaint is pain, which is described as intense, severe, boring or stabbing. The orbital pain extends to periorbital, frontal and temporal regions [3]. Ocular motor nerve palsies may start with the onset of pain, or follow within 2 weeks. In addition to involvement of extraocular movements, the pathological process may involve the orbital apex, attended by a swollen optic disc, or a pale disc. This might be followed by visual decline and may lead to progressive decline and eventual permanent visual loss. Rarely, other cranial nerves outside the cavernous sinus and superior orbital fissure may be affected; these include the maxillary and mandibular branches of the trigeminal nerve and the facial nerve [4, 5].

A thorough history and clinical examination involving localization of the lesion, a high index of suspicion coupled with imaging and response to steroid therapy remains the route to diagnosis of THS.

## 2. Case Report

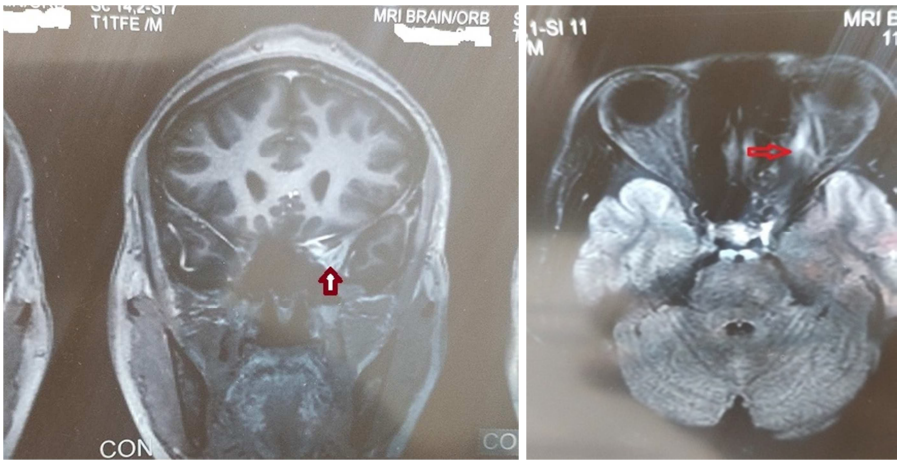
A 40 year old patient presented with 1 month history of left-sided drooping of the eyelid, severe orbital, periorbital and fronto-temporal headache, and gradual decline in vision. The pain was so severe that she experienced episodes of nausea and vomiting. She did not have left or right-sided

hemiparesis or hemiplegia.

On examination, her vital signs, including blood pressure were within normal range. Cranial nerve examination revealed internal ophthalmoplegia of left eye, with reduced adduction and intorsion of the eye ball, but normal lateral movement. The left pupil was slightly dilated and sluggishly reactive to light. She had hyperesthesia and tenderness to light palpation around the left orbit. Fundoscopy revealed a pale optic disc. The right eye exam was normal. The rest of the cranial nerves were intact.

Her full haemogram was within normal limits, and her erythrocyte sedimentation rate was slightly raised at 25mm/hour. Her VDRL was normal, as were her urea, electrolytes, creatinine and liver function tests. Additionally, we performed an anti-nuclear factor assay (ANA) and anti-dsDNA which were negative.

Magnetic Resonance Imaging of the brain and orbit revealed an extensive biconvex intra-orbital and extra-conal left orbit hyperintense lesion, insinuating through the optic canal to the left cavernous sinus and causing pressure at the left orbital apex. The lesion partially encased the left internal carotid. The lesion enhanced diffusely with gadolinium administration.




(a) (b)

***Figure 1.*** *a: Magnetic Resonance Imaging showing a hyperintense lesion on the medial aspect of the cavernous sinus. b: Magnetic Resonance Imaging showing a hyperintensity in the superior orbital fissure.*

The patient was started on prednisone 1mg/kg for an initial 2 weeks, and showed remarkable resolution in pain. However, the ptosis and ophthalmoplegia only slight improved and the vision did not improve. A review within 1 month showed slight improvement in ptosis, but the vision did not improve. By 2 months, the ptosis had almost resolved.

## 3. Discussion

THS was reported in 1954, first by Tolosa, who described a patient with ophthalmoplegia, visual loss and left orbital pain, and was found at post-mortem to have granulomatous inflammation affecting the carotid artery and the region of the cavernous sinus [6]. A case series by Hunt seven years later reported six patients with painful ophthalmoplegia caused by inflammation around the cavernous sinus [7].

Pathologically, the granulomatous inflammation is characterized by infiltration of lymphocytes and plasma cells in the cavernous sinus, with variable extension into and beyond the superior orbital fissure and orbital apex [8]. Almost seven decades after the first cases were described, the etiology is yet to be elucidated, although it is thought that traumatic injuries, tumours or aneurysms could be potential triggers. It is almost always unilateral, but in around 5% of cases may be bilateral [2].

The THS diagnostic criteria as recommended by the International Headache Society includes: a) Unilateral headache, b) Granulomatous inflammation of the cavernous sinus, superior orbital fissure or orbit confirmed by biopsy or magnetic resonance imaging, c) weakness of one or more of the ipsilateral III, IV or VI cranial nerves and d) headache must precede cranial nerve palsies by <2 weeks and there should be no other alternative diagnosis to account for the symptoms [3, 9].

A contrast-enhanced MRI is crucial to the diagnostic workup of a patient with painful ophthalmoplegia, chiefly to exclude other causes [10]. In patients with THS, MRI features can include enlargement of the cavernous sinus with abnormal tissue that is strongly enhanced with gadolinium, abnormal convexity of the wall of the cavernous sinus, and focal narrowing of the intra-cavernous internal carotid artery. Sometimes, a period of time is required before lesions become visible on imaging, suggesting that a normal MRI may not completely exclude the diagnosis of THS. Indeed, previous studies have shown that nearly 50% of THS patients have negative MRI results [11].

The main differential diagnoses of THS that should be considered and ruled out especially before glucocorticoid administration include other causes of painful ophthalmoplegia such as tumors, vasculitis, meningitis, sarcoidosis, diabetic ophthalmoplegia, and pseudotumor [12, 13].

Glucocorticoid administration has diagnostic and therapeutic values. Rapid pain resolution (within 24 to 72 hours) confirms suspected THS [14], and mprovement of cranial nerve palsies with regression of MRI abnormalities over the next two to eight weeks provides added confirmation of the diagnosis [15]. THS generally carries a good prognosis, but recurrences occur in about 21–50 % of the cases over an interval of months to years [11, 16].

## 4. Conclusion

In conclusion, THS is a very rare clinical entity, and its diagnosis depends on proper history and thorough neurological examination, proper interpretation of MRI and dramatic response to steroid therapy within a few days.

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