



### **Imaging of Tolosa Hunt Syndrome: Pre and Post Corticosteroid therapy**

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#### **Abstract**

Tolosa–Hunt syndrome (THS) is a rare painful ophthalmoplegia due to an idiopathic granulomatous inflammation in the cavernous sinus, superior orbital fissure or the orbital apex. The estimated incidence of Tolosa-Hunt syndrome is 1 per 1,000,000 person-years with an average age of onset at 41 years. We present case series of Tolosa Hunt Syndrome and its prognosis to corticosteroid therapy, which was confirmed using 3.0 TESLA MRI scanner.

**Keywords:** Tolosa hunt syndrome, Painful ophthalmoplegia, Orbital apex syndrome;

#### **Introduction**

Tolosa–Hunt syndrome (THS) is rare a painful ophthalmoplegia due to an idiopathic granulomatous inflammation in the cavernous sinus, superior orbital fissure or the orbital apex. The estimated incidence of Tolosa-Hunt syndrome is 1 per 1,000,000 person-years with an average age of onset at 41 years [1]. Tolosa Hunt

syndrome is one of the rare disorders recognized by the National Organisation for Rare Disorders (NORD) [2]. Tolosa first described as an inflammatory process of the cavernous sinus in a patient whose cerebral angiography revealed a suggestive narrowing of the carotid siphon just distal to the cavernous sinus in 1954 [3]. Hunt et al. set forth the clinical criteria by which the painful ophthalmoplegia syndrome can be recognized in 1961 [4]. Hence the name Tolosa-Hunt Syndrome [5]. Tolosa Hunt Syndrome also finds a place in the IHS Classification ICHD-3 Beta, in part three, under Painful cranial neuropathies and other facial pains. The IHS lays down diagnostic criteria for THS [6]: Unilateral headache with both of the following: 1. Presence of granulomatous inflammation of the cavernous sinus, superior orbital fissure or orbit, as seen on MRI or biopsy. 2. Palsies of one or more of the cranial nerves III, IV, and/or VI on the same side. And corroboration of the cause as evidenced by both of the following: 1. Palsies of cranial nerves III,

IV, and/or VI have followed headache in two weeks or less, or have developed simultaneously with a headache. 2. Localization of a headache around the eye on the same side [7]. The most sensitive neuroradiological modality for the diagnosis of THS is widely accepted as MRI, and the use of MRI helps in demonstrating the lesions that are directly responsible for the symptoms of THS. Symptomatic improvement following corticosteroid therapy in a patient with painful ophthalmoplegia is an essential clinical feature of THS. According to literature, corticosteroid therapy dramatically resolves both the clinical and radiological findings of THS. However, caution should be taken when confirming the diagnosis of Tolosa Hunt syndrome with steroid response as improvement clinically as well as radiologically is known to occur with other disease processes such as malignancies, infection, or vasculitis.

### Case Report

Our patient is a 70 year old male presented with difficulty in opening left eye (Ptosis) since 5 days, diplopia and blurring of vision since 3 days associated with headache in the left forehead region, paraesthesia of left forehead. With complete blood count, serum biochemistry (LFT, RFT, Glucose levels), erythrocyte sedimentation rate, thyroid function tests, antinuclear antibody, HIV, ELISA was also performed, which turned out to be normal. MRI Brain plain using 3.0 TESLA MRI scanner following a contrast study was performed in the above patient. Cavernous sinuses, orbital apices, and orbits were evaluated. The parameters used for the patients were as follows: Turbo-spin echo T1 and T2 weighted sequences on the axial plane, turbo spin echo fat-saturated T2 weighted sequence on the coronal plane, turbo spin echo T2 weighted sequence on the sagittal plane, spin echo fat-saturated T1 sequences repeated on the axial and coronal planes followed by intravenous

administration of gadolinium. In all sequences the slice thickness was 3 mm. MRI Brain plain following a contrast study was performed and showed a relatively well defined T1, T2 hypointense homogenously enhancing lesion measuring ~ 0.6 x 0.7 x 1.4 cm (CC X TAS X AP) noted involving the left superior orbital fissure lateral to optic nerve, with extension into anterior aspect of left cavernous sinus, lesion did not show diffusion restriction. Left optic nerve and extraocular muscles appeared normal. A diagnosis of left superior orbital fissure syndrome with mild extension into the cavernous sinus, more likely tolosa hunt syndrome and less likely inflammatory pseudo tumour was considered. As patient requested for discharge, he was discharged on Injection Dexamethasone- initial high dose therapy and gradual tapering regime was followed. When patient came for a follow up after a week, his symptoms were reduced, ptosis and pain had significantly reduced. A follow-up MRI in patient after three months showed relative significant reduction in signal intensity and enhancement of the left superior orbital fissure lateral to optic nerve and left cavernous sinus. Thus, confirming the diagnosis of tolosa hunt syndrome.

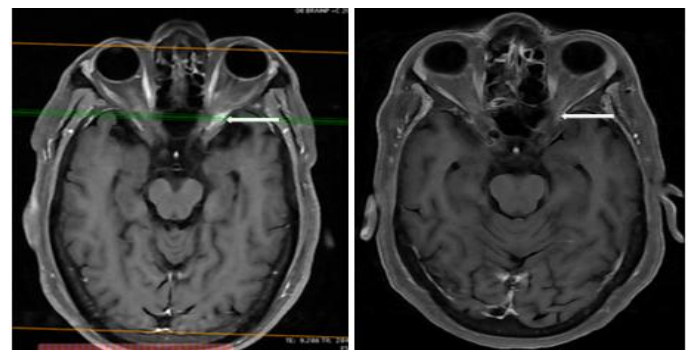


Figure 1: Pre-corticosteroid therapy post contrast T1-FS contrast image (left image) shows a well-defined homogenously enhancing lesion (straight white arrow) measuring ~ 0.6 x 0.7 x 1.4 cm (CC X TAS X AP) with extension into anterior aspect of left cavernous sinus.

Pos-corticosteroid therapy T1-FS contrast image (right image) shows near complete resolution of the lesion.



Figure 2: Above figure represents improvement of ptosis Pre-corticosteroid therapy (white arrow) and Pos-corticosteroid therapy (black arrow) within one week of treatment.

### Discussion

Tolosa hunt syndrome (THS) is not a diagnosis, but a cluster of symptoms, which may result from a number of pathological conditions. While the word aids easy recall of the elements of the syndrome, the fact is that THS is rare and constitutes only 2.9%–3.4% of all the cases of painful ophthalmoplegia. Idiopathic Pseudotumor of the Orbit (IPO) and Tolosa hunt syndrome (THS) are two nearly related syndromes treated as distinct entities until now. Both are unspecific granulomatous infiltrations of unknown local or systemic origin. They share the same clinical features, with periorbital pain, cranial nerve palsies and often dramatic response to corticosteroid therapy. MRI and CT findings show identical signal intensity with different localisation, IPO preferentially intra-orbital and THS is retroorbital in location. Therefore, our patient was considered as a case of tolosa hunt syndrome. Magnetic resonance imaging (MRI) is the most valuable imaging modality to differentiate tolosa hunt syndrome from other differential diagnoses. It offers

accurate assessment and facilitates further plans of management and also aids in follow up scans to confirm clinical status of the patient.

### Conclusion

Difficulty in opening left eye (Ptosis), diplopia and blurring of vision associated with headache in the left forehead region, paraesthesia of left forehead, puts the patient in lot of uneasy and uncomfortable painful condition. So, appropriate diagnosis of the condition using Magnetic Resonance Imaging helps the clinicians to start the patient on corticosteroid therapy as early as possible which can reduce the symptoms in 48-72 hours. Thus, pre and post corticosteroid treatment confirms the diagnosis as Tolosa Hunt Syndrome and regular follow up of the patient can completely resolve the patient's condition. Thus, confirming its good prognosis.

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