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Tolosa–Hunt Syndrome with Favorable Response to Corticosteroid Therapy: A Case Report

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Abstract | **Case Report**

Tolosa–Hunt syndrome (THS) is a rare, idiopathic granulomatous inflammation of the cavernous sinus, superior orbital fissure, or orbital apex, characterized clinically by painful ophthalmoplegia. We report the case of a 53-year-old woman with a five-month history of right periorbital headaches. MRI revealed enlargement of the right cavernous sinus with convex dural margins, iso- to slightly hyperintense T2 signal, and marked homogeneous enhancement after gadolinium administration. There was possible focal narrowing of the cavernous segment of the ipsilateral internal carotid artery, without bone erosion or mass effect. Follow-up MRI after four months of corticosteroid therapy demonstrated significant regression. This case highlights the role of MRI in diagnosing Tolosa–Hunt syndrome and monitoring therapeutic response.

Keywords: Tolosa–Hunt syndrome, cavernous sinus, painful ophthalmoplegia, MRI, corticosteroid therapy.

INTRODUCTION

Tolosa–Hunt syndrome (THS) is a rare inflammatory disorder first described by Tolosa in 1954 and further characterized by Hunt *et al*. in 1961. It is defined by unilateral orbital or periorbital pain associated with paresis of one or more cranial nerves (III, IV, VI), caused by nonspecific granulomatous inflammation of the cavernous sinus or superior orbital fissure. The annual incidence is estimated at one case per million. Although the diagnosis is primarily clinical, MRI plays a crucial role in detecting characteristic findings and excluding other etiologies such as tumors, aneurysms, and infections. Corticosteroid therapy typically results in rapid symptom relief and radiologic improvement. We present a case of THS in a middle-aged woman, with emphasis on MRI findings and follow-up.

OBSERVATION

We report the case of a 53-year-old female, referred to the Radiology Department of ibn hospital – Mohammed VI University Hospital, for evaluation of persistent right periorbital headaches lasting five months. There was no history of trauma, fever, or systemic inflammatory disease. Neurological examination was unremarkable except for tenderness in the right periorbital region.

MRI Findings:

- Cavernous sinus: Enlargement of the right cavernous sinus with convex dural margins.
- Signal: Iso-signal on T1-weighted images; iso- to slightly hyperintense on T2-weighted images.
- Post-contrast enhancement: Marked and homogeneous enhancement after gadolinium administration.
- Vascular: Possible focal narrowing of the cavernous segment of the right internal carotid artery.
- Extensions: Potential spread toward the ipsilateral orbital apex, sphenoidal sinus, and middle cranial fossa.
- Other: No bone lysis, erosion, or mass effect.

The diagnosis of Tolosa–Hunt syndrome was suggested based on the imaging and clinical presentation. The patient received systemic corticosteroid therapy. Follow-up MRI after four months showed marked reduction in the cavernous sinus thickening and normalization of the vascular caliber, correlating with complete resolution of symptoms.

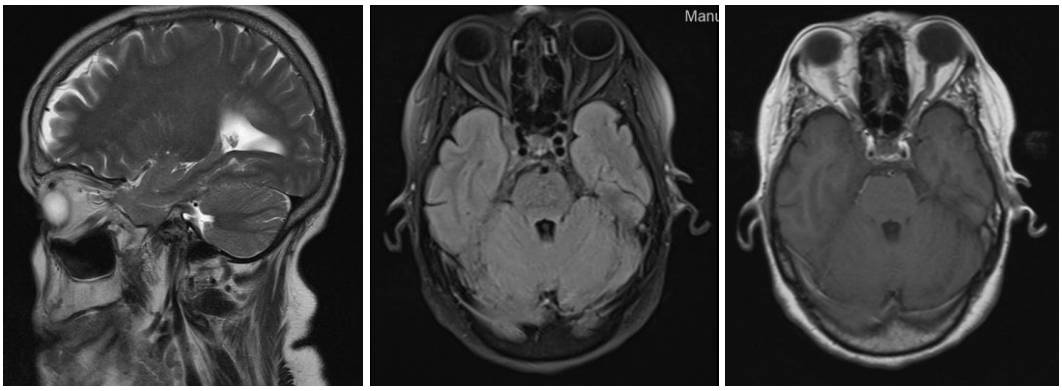

Figure 1: Axial T1-weighted MRI showing enlargement of the right cavernous sinus with intense homogeneous enhancement and convex dural margins

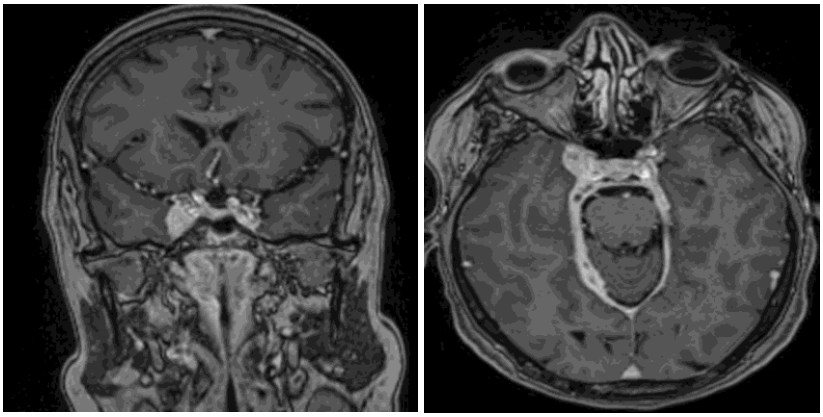

Figure 2: Follow-up coronal axial post-contrast T1-weighted MRI after four months of corticosteroid therapy, demonstrating regression of the lesion

DISCUSSION

THS is caused by a nonspecific granulomatous inflammation involving the cavernous sinus, superior orbital fissure, or orbital apex. The pathophysiology remains unclear, and diagnosis is based on clinical and radiological criteria as defined by the International Headache Society.

MRI features include:

- Enlargement of the cavernous sinus.
- Iso- to hypointense T1 signal, iso- to slightly hyperintense T2 signal.
- Intense, homogeneous enhancement after contrast.
- Possible narrowing of the cavernous segment of the ICA without complete occlusion.
- Absence of bone erosion, helping to differentiate from meningiomas and invasive neoplasms.

Differential diagnoses include meningioma, lymphoma, metastatic disease, sarcoidosis, fungal infections, and carotid–cavernous fistulas. In our case, the absence of mass effect or bony changes, along with rapid steroid response, favored THS.

Corticosteroid therapy is the mainstay of treatment, leading to pain relief within 72 hours in most cases and radiological improvement within weeks. Relapses can occur, necessitating follow-up imaging.

CONCLUSION

THS should be considered in the differential diagnosis of unilateral periorbital pain with MRI evidence of cavernous sinus involvement. MRI is indispensable for diagnosis, exclusion of mimics, and assessment of treatment response. Early corticosteroid therapy is highly effective in symptom control and lesion regression.

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