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Tolosa-Hunt syndrome revealed by MRI: A radiological perspective on painful ophthalmoplegia

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Abstract

Tolosa-Hunt syndrome (THS) is a rare and idiopathic granulomatous inflammation of the cavernous sinus and adjacent structures, classically presenting as painful ophthalmoplegia. We report a case of a 53-year-old woman who presented with right periorbital headache and an incomplete cavernous sinus syndrome affecting cranial nerves III, IV, V1, and V2. MRI demonstrated a strongly enhancing lesion in the right cavernous sinus without evidence of thrombosis or orbital invasion. The patient showed rapid clinical improvement following corticosteroid therapy. This case highlights the critical role of MRI in the diagnosis, exclusion of mimickers, and monitoring of treatment response in THS.

Keywords: Tolosa-Hunt syndrome; Cavernous sinus syndrome; Painful ophthalmoplegia; MRI; Neuroimaging

1. Introduction

Tolosa-Hunt syndrome (THS) is a rare neurological disorder characterized by unilateral orbital pain associated with paresis of one or more of the cranial nerves passing through the cavernous sinus. The pathogenesis is believed to be a non-specific granulomatous inflammation, although the exact cause remains unknown. Imaging, particularly magnetic resonance imaging (MRI), plays a key role in confirming the presence of inflammatory lesions and excluding other serious conditions such as neoplasms or infections. According to the International Classification of Headache Disorders (ICHD-3), THS remains a diagnosis of exclusion (1).

2. Case Presentation

A 53-year-old woman presented with subacute onset of right-sided periorbital headache and progressive diplopia. Neurological examination revealed an incomplete right cavernous sinus syndrome with involvement of cranial nerves III, IV, V1, and V2. There were no systemic symptoms, and ophthalmological examination showed no signs of visual acuity loss or papilledema.

Brain and orbital MRI was performed using T1-weighted pre- and post-contrast sequences with fat suppression, T2-weighted sequences, FLAIR, and DWI. The post-contrast T1-weighted images revealed a homogeneously enhancing lesion in the right cavernous sinus. The lesion appeared slightly hyperintense on T2-weighted images and showed no restricted diffusion. The internal carotid artery was surrounded but not narrowed or displaced. No orbital apex involvement or optic nerve compression was seen.

Extensive laboratory testing was performed to rule out alternative diagnoses. Complete blood count, C-reactive protein, erythrocyte sedimentation rate, ANA, ANCA, serum ACE, and infectious serologies (HIV, syphilis, tuberculosis) were all

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within normal limits or negative. Chest X-ray and CT scans excluded systemic inflammatory or neoplastic conditions. Cerebrospinal fluid analysis was not suggestive of infection or malignancy. Based on clinical presentation, imaging, and negative systemic work-up, a diagnosis of Tolosa-Hunt syndrome was retained.

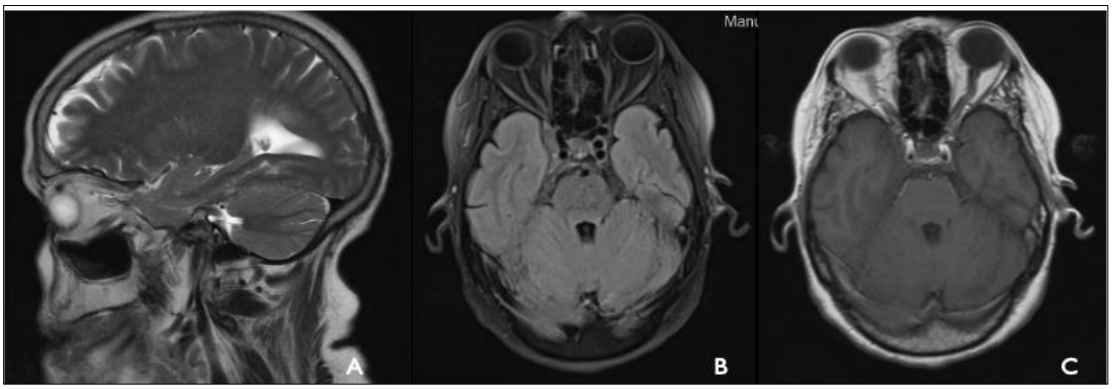

Figure 1 Sagittal and axial magnetic resonance imaging (MRI) without contrast revealed right cavernous sinus enlargement slightly hyperintense on T2 and FLAIR and isointense on T1 weighted images. (A :T2 weighted sagittal images, B: FLAIR axial images, C: T1 weighted axial images)

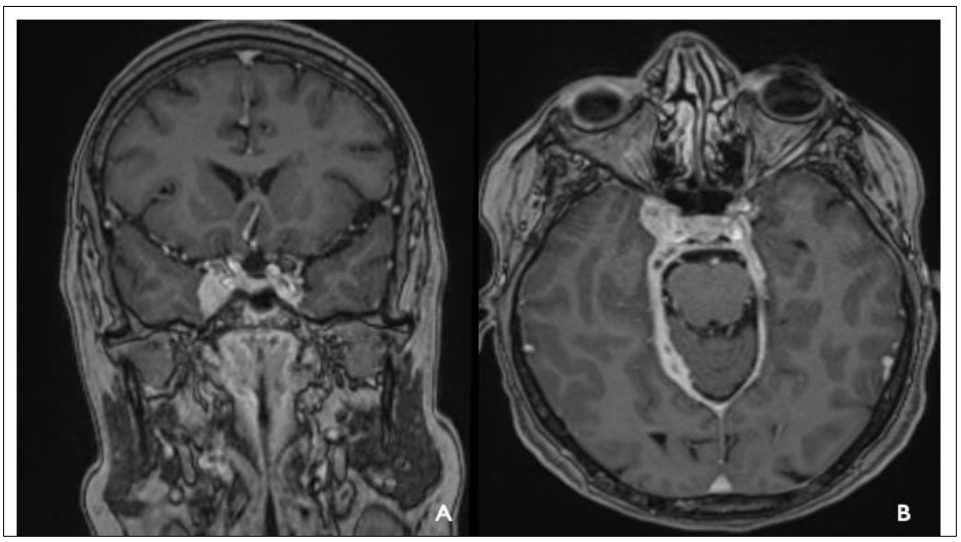

Figure 2 Coronal (A)and axial (B) magnetic resonance imaging (MRI) T1 weighted images with contrast revealed a homogeneously enhancing lesion in the right cavernous sinus surrounding the internal carotid artery without stenosis or displacement

3. Discussion

Tolosa-Hunt syndrome (THS) is a rare disorder characterized by unilateral orbital or periorbital pain associated with cranial nerve palsies, most frequently involving the oculomotor (III), trochlear (IV), and abducens (VI) nerves. In some cases, the ophthalmic (V1) and maxillary (V2) branches of the trigeminal nerve may also be affected, reflecting the anatomical proximity of these structures within the cavernous sinus (2,3).

The underlying pathophysiology of THS involves a granulomatous inflammatory process centered on the cavernous sinus, with potential extension into adjacent regions such as the superior orbital fissure and the orbital apex. This localized inflammation results in the compression and dysfunction of the cranial nerves traversing these compartments. Magnetic resonance imaging (MRI) plays a crucial role in the evaluation of suspected THS. Typical imaging findings include a soft tissue lesion with isointense signal relative to muscle on T1-weighted sequences, slight hyperintensity on T2-weighted images, and avid enhancement following gadolinium administration (1,6). Nevertheless, while MRI findings can be highly suggestive, they are not pathognomonic and must be interpreted within the appropriate clinical context.

The differential diagnosis for lesions involving the cavernous sinus is broad and encompasses neoplastic, infectious, vascular, and inflammatory conditions. Neoplastic etiologies include lymphoma, metastases, and meningioma, while infectious causes may involve fungal or bacterial pathogens. Vascular considerations primarily include cavernous sinus thrombosis, and inflammatory mimics encompass sarcoidosis and granulomatosis with polyangiitis (3,4,5). Careful clinical assessment, laboratory workup, and imaging are therefore essential to exclude these alternative diagnoses.

A hallmark feature of THS is its dramatic response to corticosteroid therapy, with significant improvement typically observed within 48 to 72 hours after initiation (7). This therapeutic response serves both as a key diagnostic criterion and as confirmation of the inflammatory nature of the lesion. In our case, the clinical presentation fulfilled the International Classification of Headache Disorders (ICHD-3) diagnostic criteria for THS, which include characteristic orbital pain, evidence of granulomatous inflammation on imaging, exclusion of alternative diagnoses, and a positive response to corticosteroids (1).

Furthermore, follow-up MRI in our patient demonstrated regression of the previously observed lesion, thereby reinforcing the diagnosis of THS. Despite the generally favorable response to therapy, it is important to note that recurrence occurs in approximately 30–40% of cases, often necessitating prolonged clinical and radiological surveillance (8). Early recognition of relapse is critical to prompt management and to prevent potential long-term neurological sequelae

4. Conclusion

Tolosa-Hunt syndrome is a rare but treatable cause of painful ophthalmoplegia. MRI plays an essential role in diagnosis and follow-up, allowing exclusion of more serious etiologies. Radiologists are central to patient management by identifying typical imaging findings, guiding diagnosis, and monitoring therapeutic response.

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