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Ptosis Revealing A Tolosa Hunt Syndrome: A Case Report

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ABSTRACT

Tolosa-Hunt Syndrome is a rare condition with an undetermined etiology that manifests clinically as unilateral orbital pain and ophthalmoplegia. We report the case of a 48-year-old female who developed acute eight upper lid ptosis within 2 days, which was preceded by retro-orbital pain without other neurological deficits. Unilateral 3d, 4th and 6th cranial nerves palsy were observed. An emergent brain magnetic resonance image (MRI) revealed a cavernous sinus asymmetry, with an oblong, well-defined thickening of the right cavernous sinus. The MRI revealed no signs of cavernous thrombosis nor intracranial lesion. THS was diagnosed, and corticosteroid treatment was initiated, with complete improvement in symptoms. A non-specific inflammation of the cavernous sinus causes the Tolosa-Hunt syndrome. Diplopia and ptosis can occur as a result of nerve paralysis. THS management is challenging and should be multidisciplinary, involving ophthalmologists, neurologists, and neuroradiologists. It is characterized by rapid response to treatment with steroids.

Keywords: Ptosis, retro-orbital pain, Tolosa-Hunt syndrome.

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I. Introduction

Tolosa hunt syndrome is defined as a rare inflammatory process of the cavernous sinus or superior orbital fissure. It results in painful ophthalmoplegia and unilateral periorbital headaches. We report a case of Tolosa Hunt syndrome in a woman and we will discuss its clinical features, treatment and evolution.

II. Case Report

We report the case of a 48-year-old woman, who has a medical history of kidney failure and is going through hemodialysis. She presented brutal and severe unilateral retro-orbital pain that started one week before her examination and right-sided ptosis two days before her examination.

The clinical examination reported a patient in good general condition, conscious, cooperative, and orientated; with 7/10 P2 visual acuity in both eyes. The local examination revealed a right upper lid ptosis associated with limitation of ocular movement-III and IV palsy and limitation of abduction of the right eye (Fig. 1 - 2).

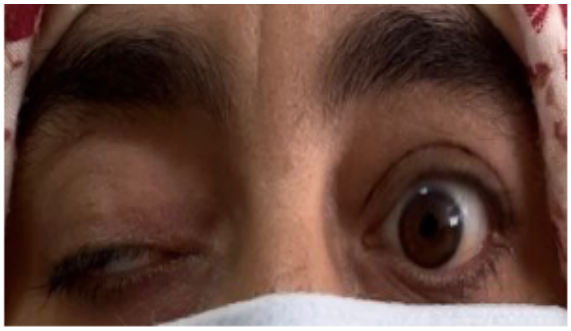

Fig. 1. Right eyelid ptosis

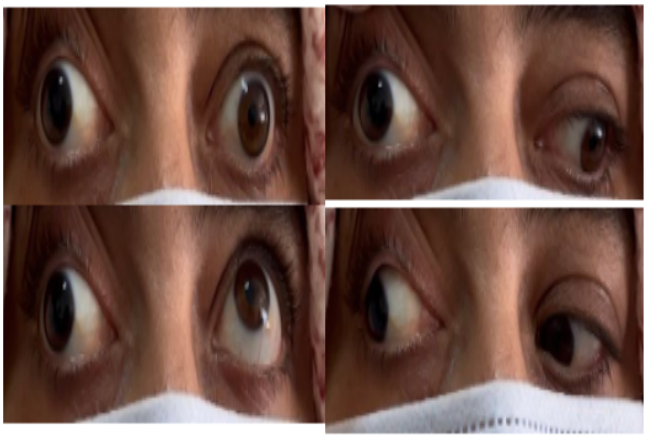

Fig. 2. Limitation of eye movements when eye forced-open characterized by a defect in adduction, upward gaze and downward gaze.

The anterior segment and the fundus examination were normal in both eyes. Optic discs presented large physiological cups.

Cranial magnetic resonance imaging revealed a cavernous sinus asymmetry, with oblong, well-defined thickening of the right cavernous sinus measuring 31.9 mm x 11 mm. This thickening extends anteriorly to the right orbital apex through the right superior orbital fissure. Behind this thickening extends towards the pre-pontic cistern (Fig. 3).

The patient received a 3-day bolus of corticosteroids followed by oral corticosteroids which helped cause rapid resolution of the symptoms. 9 days after we noticed an improvement in ocular movement and total regression of ocular limitations at 3 weeks.

Orthoptic rehabilitation was also prescribed.

The follow-up was up to 10 months, with no signs of recurrence of ptosis or ocular nerve palsy.

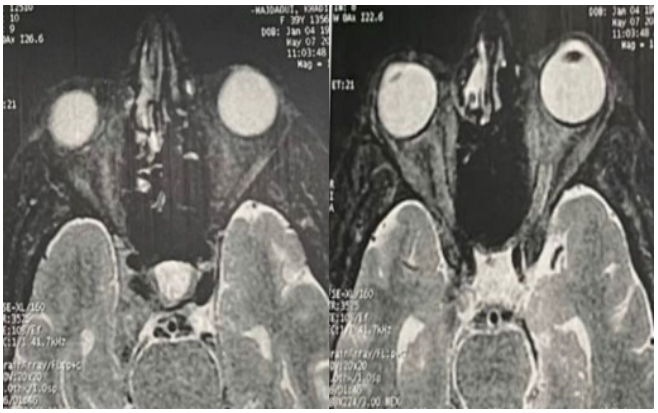

Fig. 3. Cranial magnetic resonance imaging revealed a cavernous sinus asymmetry, with oblong, well-defined thickening of the right cavernous sinus.

III. Discussion

It is yet undetermined what causes the inflammatory response and process that leads to the Tolosa-Hunt syndrome. Tolosa initially described a non-specific, chronic inflammation with fibroblast proliferation and lymphocyte and plasma cell infiltration of the cavernous sinus septa and wall; Hunt later added that these inflammatory changes, in a tight connective tissue, may put pressure on the penetrating nerves. These nerves can be cranial nerves III, IV, and VI in addition to the superior division of the fifth cranial nerve[1]. Clinically, it manifests by painful ophthalmoplegia.

The International Classification of Headache Disorders (ICHD) establishes the diagnosis criteria for Tolosa Hunt syndrome THS as seen in Table I. [2].

TABLE I: ICHD-3 Beta Diagnostic Criteria for 13.8 Tolosa-Hunt Syndrome

A-	Unilateral orbital or periorbital headache fulfilling criterion C
B-	Both of the following:
○	granulomatous inflammation of the cavernous sinus, superior orbital fissure or orbit, demonstrated by MRI or biopsy
○	paresis of one or more of the ipsilateral IIIrd, IVth and/or VIth cranial nerves
C-	Evidence of causation demonstrated by both of the following:
○	headache is ipsilateral to the granulomatous inflammation
○	headache has preceded paresis of the IIIrd, IVth and/or VIth nerves by ≤2 weeks, or developed with it
D-	Not better accounted for by another ICHD-3 diagnosis.

It is a diagnosis of elimination, which implies that all other diagnoses that could be more dangerous such as metabolic, vascular, tumoral, infectious and inflammatory etiologies must be ruled out [3].

Although it has low specificity, the use of an MRI brain with contrast is critical for excluding other disease processes [4]. MRI demonstrates cavernous sinus thickening due to the presence of abnormal soft tissue which is isointense on T1, iso or hypointense on T2, and enhances with contrast. MRI can also demonstrate the extension into the orbital apex and the convexity of the lateral wall of the cavernous sinus [5].

Bolus and oral corticosteroid therapy is the treatment of choice and good response to treatment is disease-specific [1]. There is insufficient evidence to support the optimal dose, administration route and treatment duration. It has been shown that the retro-orbital pain fully resolves within 72 hours of the onset of steroid treatment, but the time required for normalization of the cranial nerve palsies has been reported to be extended, with an average of 26 days [6].

IV. Conclusion

The THS is a rare entity, with poorly known etiopathogenic. THS diagnosis is a diagnosis of elimination that needs an accurate initial assessment, an appropriate treatment and meticulous follow-up. The use of MRI and the good response to steroids remain important to the diagnosis.

Conflict of Interest

The authors declare that they do not have any conflict of interest.

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