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Tolosa Hunt syndrome: A Diagnosis to Remember

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Abstract:- Tolosa-Hunt syndrome (THS) is characterized by painful ophthalmoplegia in relation to a nonspecific inflammatory process of the cavernous sinus, it can have a variable clinical presentation, depending on the location and intensity of the compressive inflammatory process, Corticosteroids have a dramatic effect with clinical improvement within a few days, We report the case of a 65-year-old female patient diagnosed with this syndrome.

Keywords:- *Tolosa-Hunt syndrome, Corticosteroids.*

I. INTRODUCTION

Tolosa-Hunt syndrome (THS) is characterized by painful ophthalmoplegia in relation to a nonspecific inflammatory process of the cavernous sinus. This syndrome is as rare as it is unrecognized in the ophthalmologic setting. It remains a diagnosis of elimination. (1)

II. MATERIALS AND METHODS

We report the case of a 65-year-old female patient, hypertensive on treatment, who consulted the ophthalmology emergency room for decreased visual acuity progressing for 20 days.

- The interrogation reported a notion of hemi-cranial headaches and binocular diplopia.
- The ophthalmological examination found a corrected visual acuity of 3/10 in the right eye and a positive light perception in the left eye, a ptosis and a left abduction paralysis (figure 1). The photomotor reflex is also lazy in the affected eye.
- Biomicroscopic examination of the anterior segment is without abnormalities. While fundus examination OG finds a pale papilla.
- A neurological examination is requested, not showing any associated neurological damage.

III. RESULTS

➢ *A brain MRI is performed, and finds:*

- Thickening of the walls of both cavernous sinuses, convex and in T2 hyposignal, intensely enhanced after injection of PC.
- Infiltrated aspect of the extracervical fats, clearly predominant on the left, with intense contrast in the fats and muscles. This infiltration fills both the upper and lower left orbital fissures, with moderate extension to the left infra temporal fossa.
- Sheathed appearance of the optic nerves, especially on the left at the level of the apices and optic holes, with T2 hypersignal of the left NO
- A complete general, immunological and infectious workup was performed, which came back negative.
- The diagnosis of Tolosa Hunt syndrome was retained in view of the negativity of all the complementary examinations performed.
- The patient was put on a general corticosteroid treatment, which resulted in a spectacular regression of the oculomotor disorders in 3-4 days. The visual acuity persisted with positive light perception.

IV. DISCUSSION

Tolosa-Hunt syndrome can have a variable clinical presentation, depending on the location and intensity of the compressive inflammatory process, and on the time to management. MRI has a central place in the diagnosis, allowing a fine analysis of the cavernous sinus and orbital apex, eliminating differential diagnoses. (2)

The latter are represented by traumatic, vascular, neoplastic, and above all infectious causes. They represent the largest proportion of painful ophthalmoplegia, Tolosa-Hunt syndrome remaining a rare cause (one case in one million inhabitants per year). (3)

Corticosteroids have a dramatic effect with clinical improvement within a few days. However, there are no precise recommendations on dosage, duration of treatment or even route of administration. As far as evolution is concerned, recurrences are classic, in which case immunosuppressants

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(azathioprine, methotrexate) and even radiotherapy can be used as a second line treatment, most often for corticosteroid sparing.

V. CONCLUSION

Tolosa-Hunt syndrome is a rare condition that requires extensive and accurate initial evaluation before a diagnosis can be made, which remains one of elimination. MRI is a valuable examination because of its better sensitivity for the detection of cavernous sinus lesions and its reproducibility. In the presence of painful ophthalmoplegia, the diagnosis of Tolosa-Hunt syndrome must be evoked. Recurrences, corticosteroid dependence and its side effects make it a disabling disease.

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FIGURES

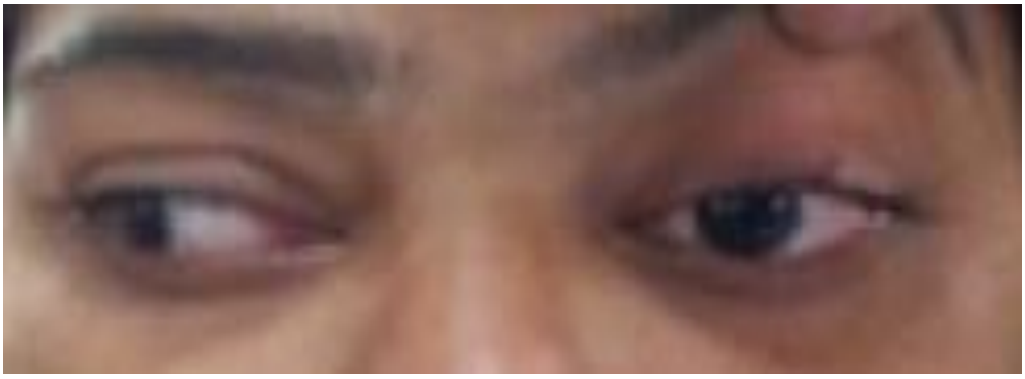

Fig 1 : ptosis and abduction paralysis on the left

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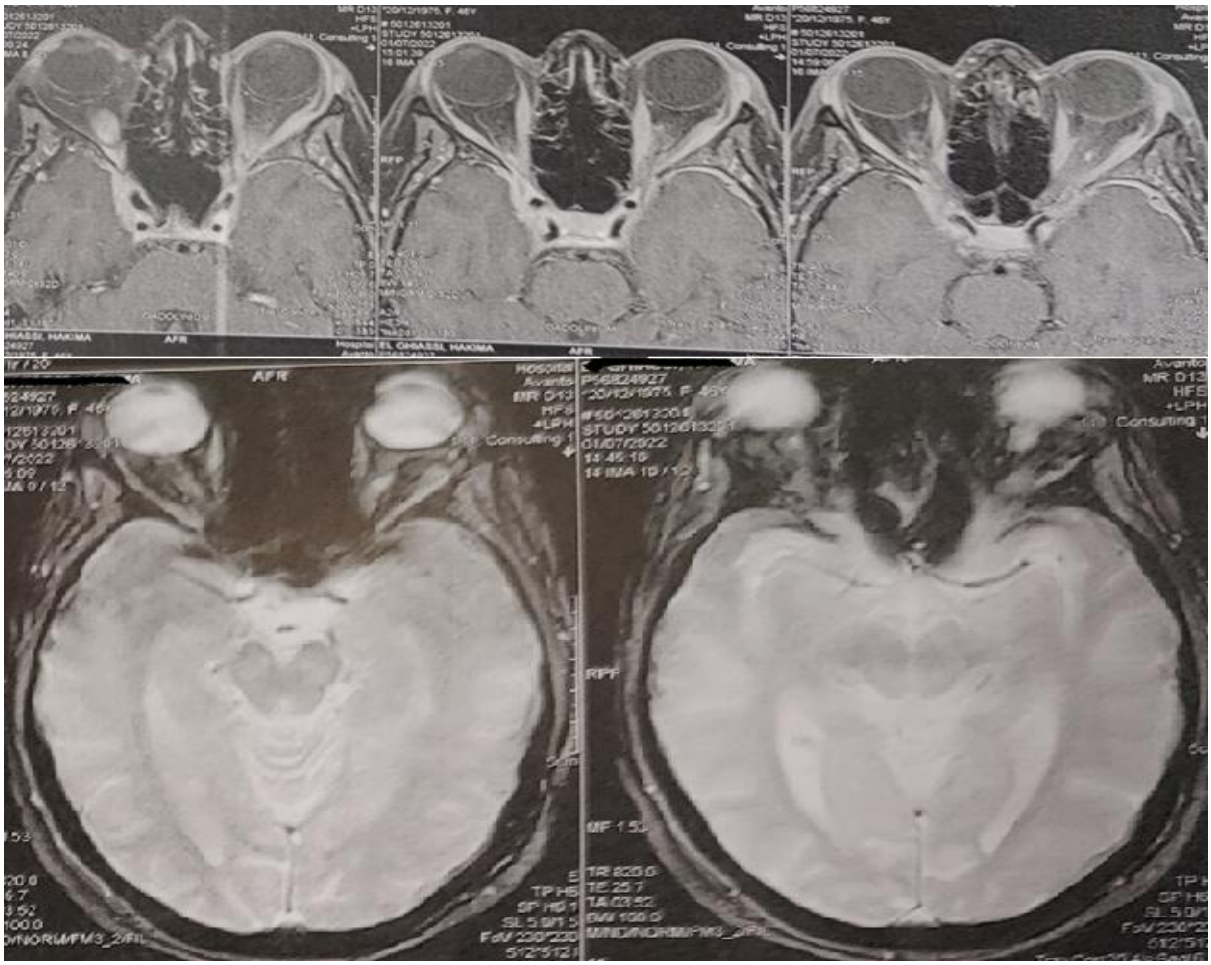

Fig 2: Brain MRI

- Thickening of the walls of both cavernous sinuses, convex and T2 hyposignal, intensely enhanced after PC injection.
- Infiltrated aspect of the extra cervical fats, clearly predominant on the left, with intense contrast in the fats and muscles. This infiltration fills both the upper and lower left orbital fissures, with moderate extension to the left infra temporal fossa.
- Sheathed appearance of the optic nerves, especially on the left at the level of the apices and optic holes, with T2 hypersignal of the left optic nerve.