A CASE REPORT ON TOLOSA HUNT SYNDROME

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Abstract:

Toloso-Hunt syndrome (THS) is an extremely rare disorder characterised by painful unilateral ophthalmoplegia triggered by idiopathic inflammation of the cavernous sinus affecting the 3,4,5 cranial nerve. corticosteroid therapy effectively improves THS symptoms. We report the case of 54-year-old male patient who presented with Blurred vision and double vision (for a week), one sided Headache (left side) in periorbital area with severe pain from 10 days, Patient gone for ocular examination in that it was found that left eye ptosis, pupil -left sluggishly reacting to light, left eye examination showed visual activity -6/36, CNS examination of facial sensation decrease in ophthalmic division on left side, acute onset of left eye vision distribution, double vision III, IV, VI- CN involvement in the left eye. MRI Brain impression found that thickening of left lateral -rectus with abnormal enhancement of left cavernous sinus extending up to orbital apex through above investigations it was diagnosed as THS. The patients' symptoms improved dramatically upon on starting the oral corticosteroid therapy.

Key words:

auto immune, inflammation, cavernous sinus, ptosis, rectus muscle, periorbital muscles

Introduction:

TOLOSA HUNT SYNDROME is a rare neuroimmunology disorder characterised by severe periorbital headache, ophthalmoplegia, unilateral pain in periorbital region (sharp, shooting, stabbing, severe and intense [(1)]. It is associated with inflammation of specific areas behind the eyes (cavernous sinus and superior orbital fissure). There will be more involvement of maxillary and mandibular division of 5th cranial nerve (trigeminal), optic nerve, facial nerve [(2)]. The effected individual may exhibit signs of paralysis of certain cranial nerves such as drooping of upper eye lip(ptosis), double vision, large pupil, facial numbness. The exact cause of the disorder is unknown but causes were explained by few theories, one theory states that due to abnormal autoimmune response linked with inflammation in a specific area behind the eye (cavernous sinus and superior orbital fissure) , and some cases had shown that inflammation may be due to clumping of certain type of cells(granulomatous inflammation), others described that the disorder include generalised inflammation and constricted or inflamed cranial blood vessels near the eye[(3).] The more onset of age is 41 years, mostly cases were reported among 30-45 age group individuals, rare cases were found in children (<10 years). Both men and women get equally effected [(2)]. TOLOSA HUNT SYNDROME- codified by the International Headache Society which is the standard diagnostic method for the identification of the disorder. MRI, CT- Scan are another diagnostic method [(2).]

Case report:

A male patient of 54 years was admitted in the Neurology ward with chief complaints of Blurred vision and double vision (for a week), one sided Headache (left side) in periorbital area with severe pain from 10 days and also difficulty in looking too far objects. He had past history of hypertension on Amlong-10mg-OD. Patient gone for ocular examination in that it was found that left eye ptosis, pupil -left sluggishly reacting to light, left eye examination showed visual activity -6/36. Neurological examination revealed that left oculomotor, trochlear, and abducens nerve paresis, with hypoesthesia over area supplied by left ophthalmic division of trigeminal nerve. Lab values – Hb -11.1g/dl, TC – 8800 cum.mm, sr.cr-1.89mg/dl, CSF Proteins- 18mg/dl which are found to be normal. MRI Brain impression found that thickening of left lateral -rectus with abnormal enhancement of left cavernous sinus extending up to orbital apex, multiple micro haemorrhage in between parenchyma basal ganglia. CT Brain found that thickening of lateral rectus muscle with enlargement of left cavernous sinus with orbital apex as shown in the Fig:1 and left ICA. CNS examination of facial sensation decrease

in ophthalmic division on left side, acute onset of left eye vision distribution, double vision III, IV, VI- CN involvement in the left eye.

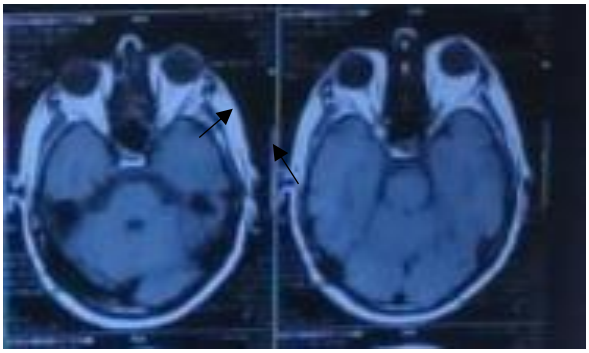

Figure 1: left lateral rectus with inflamed cavernous sinus

The patient was treated with high dose intravenous methylprednisolone 1 gram/day for 3 days along with gastroprotective therapy. There was a significant reduction in left periorbital pain and ptosis after 72 hours with a moderate improvement of left eye ocular movement. The patient was discharged on day 7 with oral prednisolone 50mg daily for 13 days and should follow-up in Neurology.

Case discussion:

Tolosa first described this disease condition in 1954, in a patient with the unilateral recurrent painful ophthalmoplegia involving cranial nerves III, IV, and V1. Similar cases were reported by Hunt et al. in 1961. Smith and Taxdal called it as Tolosa-Hunt Syndrome for the first time in 1966[(2)]. The later authors discussed the importance of the rapid response towards steroid therapy. Tolosa-Hunt syndrome is a rare neuroimmunology nonspecific granulomatous inflammation characterized by infiltration of lymphocytes and plasma cells primarily in and around the cavernous sinus, with variable extension into and beyond the superior orbital apex. Etiology is still unknown however, traumatic injury, tumours, or an aneurysm could be the potential triggers [(2).] It does not have any age or sex predilection [(5)]. It is almost always unilateral (except in 4-5% of cases) Both men and women get equally effected [(6).] It is considered a very benign illness, but exclusion of more malignant diseases bears utmost importance whenever any patient presents with such clinical features.

The International Classification of Headache Disorders (ICHD) criteria for THS include the following [(7)]:

A. Unilateral headache fulfilling criterion C

B. Both of the following:

1. granulomatous inflammation of the cavernous sinus, superior orbital fissure or orbit, demonstrated by MRI or biopsy.

2. paresis of one or more of the ipsilateral III, IV, and/or VI cranial nerves.

C. Evidence of causation demonstrated by both of the following:

1. Headache has preceded paresis of the III, IV, and/or VI nerves by ≤2 weeks, or developed with it.

2. Headache is localized around the ipsilateral brow and eye.

D. Not better accounted for by another ICHD-3 diagnosis.

In a study of 22 cases reported with THS, pain and diplopia were found in 100% and 91% cases and the pain was followed by paresis. The pain was relieved in 91% within 72 hours of treatment and no patient had reported regarding complete relief from paresis [8]. These findings were consistent in this case.

MRI brain with contrast, especially the coronal view, is one of the crucial diagnostic study and helps to rule out other disease processes, but it has low specificity [2]. Yousem et al. examined 11 patients and described that pathological MRI findings in the cavernous sinus were found in nine, among them six had affected cavernous sinus was enlarged; in five of nine, had a convex lateral wall and extension into the orbital apex was seen in eight patients [9].

High-dose glucocorticoids are the first-line treatment for Tolosa-Hunt syndrome due to its inflammatory pathology. It causes rapid resolution of orbital pain within 1-3 days, which is considered as a diagnostic confirmation [10]. Our patient responded similarly with a consequential reduction of pain within 72 hours of glucocorticoid therapy. In one study, 40% of patients were achieved pain relief within 72hours and 78% within seven days. After an initial high dose of corticosteroid (loading dose), an oral taper dose over the course of several weeks is recommended, along with frequent follow-up with subsequent MRI monitoring is recommended to document about the resolution of the disease. Immunosuppressive drugs are the other therapeutic choice [10]. Despite treatment, recurrences are very common and the overall quality of life of a patient is poor [1].

Tolosa-Hunt Syndrome is a rare entity with unknown etiology, presenting clinically with unilateral orbital pain and ophthalmoplegia, is a diagnosis of exclusion, which will resolve spontaneously even though it can recur and have a substantial response to systemic steroids. The representative clinical presentation of this case with dramatic response to steroids

and exclusion of other conditions from investigation and imaging techniques were crucial for concluding the diagnosis [3].

CONCLUSION:

Tolosa-Hunt Syndrome is a rare inflammatory condition, should be carefully diagnosed by excluding the possible differential diagnosis like peri orbital syndrome, infections and inflammations. Corticosteroid treatment is the standard treatment for THS. Recurrent MRI imaging should be considered in patients with repeated episodes. In this case MRI and positive response to treatment of corticosteroids (methyl prednisolone) were relevant for making diagnosis.

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