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Research Article

AN INTERESTING CASE OF TOLOSA-HUNT SYNDROME (A RARE CASE REPORT)

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A B S T R A C T

Background: Tolosa- Hunt Syndrome Is A Painful Ophthalmoplegia Caused By The Inflammatory Process of Various Etiologies Involving The Cavernous Sinus, Presents With Ocular Motor Weakness And Retro-Orbital Pain. Facial Sensation And Visual Acuity May Be Diminished.

Case Study: A 55-Year-Old Female Presented To Our Medical Opd With Complaints of Severe Headache And Facial Pain And Pain In The Right Eye And Diplopia For The Past 10 Days. On General Physical Examination, We Found That Her Vitals Were Stable, And Central Nervous System Examination Showed That The Patient Is Unable To Abduct Her Right Eyeball, Rest All Other Extraocular Movements Were Normal On The Right Eye. On The Left Eye, All Extraocular Movements Were Absolutely Normal. Her Routine Blood Investigations Were Normal And In View of 6th Cranial Nerve Involvement We Ordered Mri Brain And It Showed Minimal Asymmetrical Enhancing Soft Tissue Thickening Along The Posterolateral Aspect of The Right Cavernous Sinus With Mild Small Vessel Ischemic Changes. On The Basis of These Findings, A Diagnosis of Tolosa- Hunt Syndrome Was Made And The Patient Was Started On Corticosteroid Therapy. Dramatic Improvement In Diplopia And Pain Noted Within 48 Hours Of Steroid Therapy.

Conclusion: Although The Pathogenetic Basis of Tolosa- Hunt Syndrome Remains Unknown, From A Practical Clinical Standpoint It Can Be Regarded As A Distinct Entity That May Be Stimulated By Various Other Disorders. Cortico Steroids Form The Mainstay of Treatment For This Syndrome.

INTRODUCTION

Tolosa Hunt syndrome (THS) is described as severe and unilateral periorbital headache associated with painful and restricted eye movements. Synonyms for Tolosa Hunt syndrome include painful ophthalmoplegia, recurrent ophthalmoplegia, ophthalmoplegia syndrome. Tolosa Hunt syndrome is one of the rare disorders recognized by the National Organisation for Rare Disorders (NORD). It is also included as one of the painful cranial neuropathies by the International Headache Society (IHS) in its headache classification. Tolosa Hunt syndrome was first described in the year 1954 by Dr. Eduardo Tolosa, a Spanish neurosurgeon. Tolosa- hunt syndrome is a painful ophthalmoplegia caused by inflammatory process of various aetiologies involving the cavernous sinus, presents with ocular motor weakness and retro orbital pain. Facial sensation and visual acuity may be diminished.

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CASE REPORT

A 55-year-old female presented to our medical OPD with complaints of severe headache and facial pain and pain in the right eye and diplopia for past 10 days. She is also a known case of diabetes mellitus on irregular medication for past 10 years and she had history of similar type of headache in the past few months ago and took treatment for the same for 1 week in a primary care centre in her native suspecting it to be a sinusitis. On examination her vitals parameters were stable and central nervous system examination showed that the patient is unable to abduct her right eyeball(Figure 2), rest all other extra ocular movements were normal on right eye. On the left eye all extra ocular movements were absolutely normal. Her routine blood investigations were normal and in view of 6th cranial nerve involvement we ordered MRI brain to rule out intracranial pathology and it showed (Figure 1) minimal asymmetrical enhancing soft tissue thickening along posterolateral aspect of right cavernous sinus with mild small vessel ischemic changes. Calcium level was 8.8 mg/dL, ANA was negative, rheumatoid factor was negative, and erythrocyte sedimentation rate was not elevated. Chest X-ray did not show hilar adenopathy and was unremarkable otherwise. A complete infectious aetiology was ruled out, which included the

following: Lyme disease, herpes simplex virus, *Aspergillus*, *Coccidioides*, West Nile, syphilis, *Cryptococcus*, *Histoplasma/Blastomyces*. Complete vital panel was negative. A lumbar puncture was done, and cerebrospinal fluid (CSF) studies revealed normal glucose, normal protein, and one nucleated cell. CSF angiotensin-converting enzyme level was normal. However, a serum angiotensin-converting enzyme level was not checked. CSF cytology was negative; CSF IgG index was normal. Serum and urine protein electrophoresis were normal. The patient was immediately started on IV methylprednisolone 500 mg BID for 3 days. The patient's pain and ocular movements began to marginally improve within 12 hours of the first dose. The patient continued IV steroids, and on day 3, he reported that his pain had completely resolved with significantly improved adduction of the right eye (Figure 3). The patient was released home on day 4 with almost complete normalization of her right eye adduction. The patient was discharged home on prednisone 60 mg daily for 7 days with follow-up with Neurology.

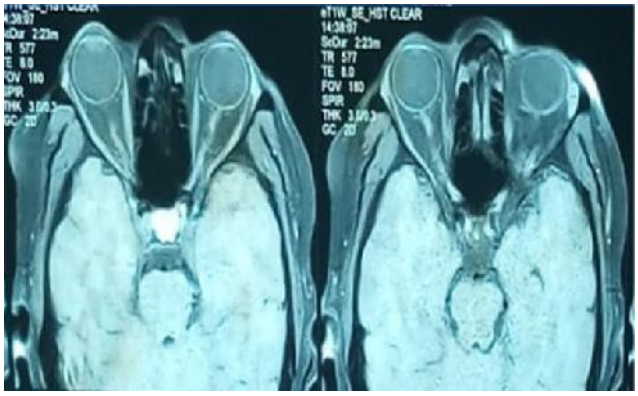

Figure 1 MRI Brain

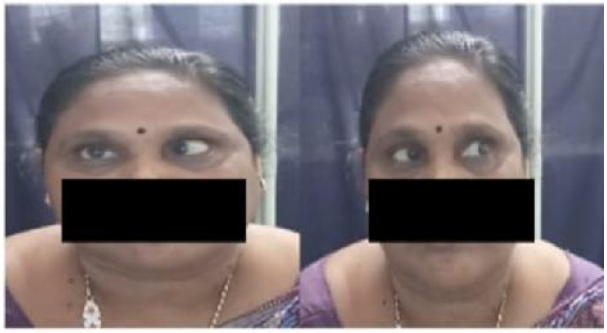

Figure 2 Right Eye Lateral Rectus Palsy (Before Treatment)

Figure 3 Right Eye Lateral Rectus Palsy Recovered (After Treatment)

Differential Diagnosis

- Anisocoria
- Benign Skull Tumours
- Brain Mets
- Cavernous Sinus Syndromes
- Cerebral Aneurysms
- Cerebral Venous Thrombosis
- CNS Whipple Disease
- Epidural Hematoma
- Lyme Disease
- Meningioma
- Migraine Headache
- Neurosarcoidosis
- Paediatric Craniopharyngioma
- Polyarteritis Nodosa
- Primary CNS Lymphoma
- Systemic Lupus Erythematosus
- Tuberculous Meningitis
- Varicella Zoster

DISCUSSION AND CONCLUSION

The International Headache Society guidelines for diagnosis of THS include the following: retro-orbital pain with an oculomotor palsy, granulomatous inflammation within the cavernous sinus, superior orbital fissure or orbit confirmed by MRI or tissue biopsy, the onset of the oculomotor palsy must be at the same time or within 2 weeks of the onset of orbital pain, and the pain must be localized around the ipsilateral brow and eye. THS follows a variable course that can last from days to weeks to months. Recurrences are common and can either be unilateral or bilateral. Other causes must be excluded by appropriate investigations. The retro-orbital pain has been shown to completely resolve within 72 hours of onset of steroid treatment, but the time needed for normalization of the cranial nerve palsies has been broad with an average of 26 days. Our patient met the aforementioned criteria and all infectious and autoimmune aetiologies were ruled out making a diagnosis of THS more likely.

We believe this case demonstrates the importance of including THS as a differential diagnosis after all other diagnostic studies rule out more common aetiologies for retro-orbital pain with oculomotor nerve palsy. Our patient visited 2 different emergency departments over a 2-week period and received separate diagnosis of sinusitis and cluster headaches, respectively, and only was sent to our emergency department after an ophthalmologist evaluation. Since THS responds well to steroid treatment, early detection is beneficial. Per the literature review, no standard dose for steroid treatment has been established. Our patient responded well to IV methylprednisolone 500 mg, and on day 3, he had almost complete resolutions of his pain and oculomotor nerve palsy. This response was faster than reported in the majority of cases.Studies have shown that younger patients are more likely to have a faster improvement in their oculomotor palsy, which could also explain our patient's almost complete resolution of symptoms within 3 days of steroid treatment.

Declaration of Conflicting Interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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