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A RARE CASE OF TOLUSA HUNT SYNDROME

KEY WORDS:

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ABSTRACT

Tolosa-Hunt syndrome (THS) is a rare condition characterized by intense, unilateral headache, along with paralysis of certain eye muscles. It is caused by inflammation and damage to the cavernous sinus, a large vein at the base of the brain. The exact cause of THS is not well understood, but it is thought to be an autoimmune disorder. This case report describes a 45-year-old female patient who presented with a left-sided headache, double vision, drooping of her left eyelid, and loss of vision in her left eye. She was diagnosed with Tolosa-Hunt Syndrome, a rare condition characterized by inflammation and damage to the cavernous sinus, causing paralysis of certain eye muscles and intense unilateral headache. Diagnostic tests including MRI orbit plain + contrast and PET scan of the brain. Treatment involved high doses of corticosteroids to reduce inflammation, resulting in significant improvement in her symptoms within a few days of initiation. The patient was gradually tapered off the corticosteroids over the next 4 weeks and advised to continue follow-up with her neurologist.

INTRODUCTION

Tolosa-Hunt syndrome (THS) is a rare neurological disorder that was first described in 1954 by Tolosa, who reported on a patient with severe orbital pain, progressive visual loss, total ophthalmoplegia, and reduced sensation in the first division of the trigeminal nerve. Later, in 1961, Hunt and colleagues defined the clinical entity of THS based on six patients, where they identified an inflammatory lesion in the cavernous sinus as the probable cause of the syndrome. The term "Tolosa-Hunt syndrome" was coined in 1966 by Smith and Taxdal to describe this condition, which typically presents with intense, unilateral headache and paralysis of certain eye muscles, and is often treated with corticosteroids to reduce inflammation.[1-3]

CASE REPORT

A 45-year-old female visited DY Patil Hospital in Nerul, Maharashtra with the chief complaints of left-sided headache since one month, double vision in her left eye since 12 days, drooping of her left eyelid since 10 days, and loss of left eye vision since 10 days. Her headache began in the nasal area and gradually progressed to the frontal head, mild to moderate, unilateral, boring in character, continuous, and associated with vomiting. After visiting a local doctor and receiving medications, her headache was relieved for a few days, but then she began to complain of double vision that was monocular in her left eye, present in all gazes, and improved on covering the left eye. A couple of days later, she complained of drooping of her left eyelid as well as pain in her left eye, which was moderate in grade, continuous, and associated with blurring of vision, but not associated with redness or lacrimation.

She consulted an ophthalmologist, where she was diagnosed with Left eye lateral rectus palsy and was prescribed medications. However, her blurring of vision in her left eye increased, and then her left eye had no light perception. She was then referred to a neurologist for further evaluation.

The patient had no history of Covid-19, diabetes mellitus, hypertension, chronic obstructive pulmonary disease, tuberculosis, cerebrovascular accident, or coronary artery disease. Her family history was also negative for any of these conditions. She had regular menstrual periods, and on examination, her vital signs were stable with ptosis of her left eyelid and limited movement of her left eye in all directions except down and inward. Her pupils were equal, and her left eye was non-reactive to light. There was no facial asymmetry or weakness, and the remainder of the neurological examination was normal.

She underwent MRI B/L ORBIT, MRI BRAIN, and PET SCAN. MRI orbit plain + contrast showed minimal relative prominence of the per optic CSF space on the left side as compared to the right. MRI brain revealed mild generalized cerebral atrophy with chronic ischemic changes in bilateral fronto parietal and periventricular white matter. PET scan of the brain showed impaired tracer uptake along superior, inferior, medial, and lateral rectus muscles of the left eyeball with drooping of the left eyelid suggestive of CN palsy and traumatic Tolosa-Hunt syndrome.

She was diagnosed with Tolosa-Hunt Syndrome and was prescribed injection methylprednisolone 1 gram intravenously daily for 5 days followed by oral steroids (prednisone 60mg/day). Within 2-3 days of initiating treatment, she reported significant improvement in her symptoms. Her headache had decreased, and her double vision had improved. The left eyelid ptosis and limited eye movement were also much improved. She was slowly tapered off the corticosteroids over the next 4 weeks and advised to continue follow-up with her neurologist.

DISCUSSION

The diagnosis of Tolosa-Hunt Syndrome (THS) can be challenging due to its rarity and similarity to other causes of retro-orbital pain with oculomotor nerve palsy. The International Headache Society guidelines provide clear criteria for the diagnosis of THS, including retro-orbital pain, oculomotor palsy, granulomatous inflammation, and exclusion of other causes.[4]

One of the key features of THS is its variable course, which can range from days to weeks to months, with common recurrences that can be unilateral or bilateral. The importance of early detection and treatment cannot be overstated, as THS responds well to steroid treatment.[5]

While there is no established standard dose for steroid treatment, the literature suggests that IV methylprednisolone at a dose of 1 gram can be effective in treating THS. This underscores the need for physicians to be aware of THS and to consider it as a potential diagnosis in cases of retro-orbital pain with oculomotor nerve palsy, particularly after other common etiologies have been ruled out.[6]

In conclusion, THS is a rare but important differential

diagnosis in cases of retro-orbital pain with oculomotor nerve palsy. The International Headache Society guidelines provide clear criteria for diagnosis, and early detection and treatment with steroids can lead to rapid resolution of symptoms. More research is needed to establish optimal treatment protocols for THS, but a high index of suspicion and prompt diagnosis are critical for achieving the best outcomes for patients.

CONCLUSION

Tolosa-Hunt Syndrome (THS) is a diagnosis of exclusion, and it is important to investigate each case of painful ophthalmoplegia to rule out other common causes. An MRI of the brain and orbit, particularly with a coronal section, can be helpful in identifying the pathology of the cavernous sinus in cases of THS. Early diagnosis and management are crucial as THS responds to steroids within 24-48 hours. Therefore, timely diagnosis and management are essential for effective treatment of THS.

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