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TOLOSA HUNT SYNDROME: A RARE CASE REPORT

Ophthalmology

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KEYWORDS

Tolosa hunt syndrome, a neuro-ophthalmic entity, painful ophthalmoplegia, recurrent ophthalmoplegia, ophthalmoplegia syndrome.

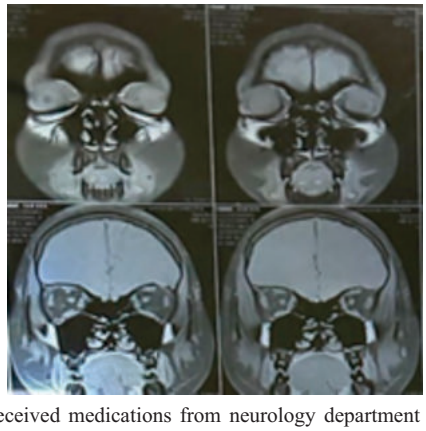
INTRODUCTION

Tolosa Hunt Syndrome is a rare steroid responsive etiology for painful ophthalmoplegia caused by nonspecific inflammation of the cavernous sinus or superior orbital fissure [1]. It is one of the rare disorders recognised by the National Organization for rare disorders and is also included as one of the painful cranial neuropathies by the International Headache Society (IHS) in its headache classification[2]. It was first described by Tolosa in 1954 and explored further by Hunt. The estimated annual incidence is one case per million per year. It is characterized by unilateral orbital pain associated with paresis of one or more of third, fourth or sixth cranial nerve, commonly leading to diplopia. It is caused by idiopathic granulomatous inflammation.

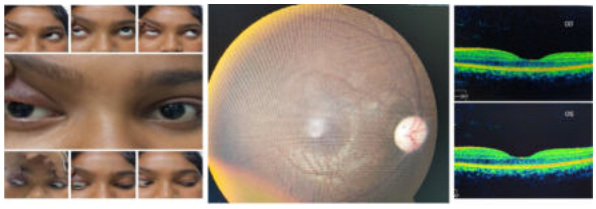
CASE PRESENTATION

Tolosa Hunt Syndrome is a rare disease characterized by painful ophthalmoplegia affecting third, fourth and/or sixth cranial nerve caused by non-specific inflammation in the cavernous sinus or superior orbital fissure of unknown etiology. We present a 14-year-old girl who presented to OPD in December 2022 with sudden loss of vision and drooping of right eyelid for 2 weeks. On examination, perception of light was positive in right eye with moderate ptosis, RAPD and restricted EOM in all directions. The left eye examination was within normal limits with visual acuity: 6/6. The intraocular pressure was normal.

Routine investigations revealed increased WBC (13,500), RBS was normal (65mg/dl). Chest X-ray came out to be normal, viral markers negative, CRP high (14.70 mg/dl). In CSF, Mycobacterium Tuberculosis and Cryptococcal antigen were not detected. VDRL was non-reactive. MRI brain with orbit shows heterogeneously enhancing soft tissue anterior aspect of right cavernous sinus and right superior orbital fissure suggestive of tolosa hunt syndrome.



Patient received medications from neurology department including pulse therapy of IV methylprednisolone followed by oral steroids (prednisolone) with multivitamins after which the symptoms improved, extraocular movements and drooping of lids improved. Patient's vision was same as perception of light in right eye with Fundus showing pale disc. OCT came out to be normal.



extraocular movements after treatment

Fundus of right eye showing pale disc

OCT of both eyes: WNL

DISCUSSION

Etiology is still unknown, however traumatic injury, tumours or aneurysm could be potential triggers [2]. It does not have any age or sex predilection and can affect anyone in the first to eighth decade of life [3]. It is almost always unilateral and if bilateral shifts the balance in support of other differential diagnoses [4]. It is considered a very benign illness, but exclusion of more malignant diseases bears utmost importance [1]. MRI brain with contrast especially the coronal view, is a crucial diagnostic study and helps to exclude other disease processes [2]. High dose glucocorticoids are the are the first line treatment for Tolosa Hunt considering its inflammatory pathology. It causes rapid resolution of orbital pain within 1 to 3 days, which also serves as diagnostic confirmation. Our patient responded similarly with a significant reduction of pain.

After an initial high dose of corticosteroid, an oral taper over the course of several weeks is recommended, along with regular follow up with subsequent MRI studies to document resolution of disease. Despite treatment, recurrences are common and overall quality of life is poor.

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