Asian Journal of Case Reports in Surgery

9(4): 1-5, 2021; Article no.AJCRS.69142

Sudden Painful Loss of Vision – Exquisite Case of Tolosa Hunt Syndrome

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Authors' contributions

This work was carried out in collaboration among all authors. Authors SD and Saranya prepared rough manuscript. Author SS has review the manuscript. Author SAP has prepared the final manuscript. Author LK has collected the data. All authors read and approved the final manuscript.

Article Information

Editor(s):
(1) Prof. José Francisco de Sales Chagas, Sao Leopoldo Mandic Medical School, Brazil.
Reviewers:
(1) Zunaina Embong, Universiti Sains Malaysia, Malaysia.
(2) Bárbara Alexandra Rubio Lastra, Chile.
Complete Peer review History: http://www.sdiarticle4.com/review-history/69142

Case Study

Received 25 March 2021
Accepted 02 June 2021
Published 09 June 2021

ABSTRACT

Tolosa Hunt Syndrome is a painful ophthalmoplegia with periorbital or hemi cranial pain plus ipsilateral oculomotor nerve palsy with or without oculo-sympathetic paralysis. Many causes can lead to painful ophthalmoplegia like inflammatory, infective, neoplastic, vascular, etc. Various investigations need to be done to rule out the organic causes of painful ophthalmoplegia. Here we would like to report a case of Tolosa Hunt Syndrome in a 45-year-old female who was presented to our hospital with history of retro orbital pain followed by sudden loss of vision. Computed Tomography and Magnetic Resonance Imaging showed features suggestive of Tolosa Hunt Syndrome and she was treated with high dose steroids. The patient responded well to high dose steroids showing significant improvement in both vision and retro orbital pain.

Keywords: Tolosa hunt syndrome; opthalmoplegia; hemi cranial, oculosympathetic paralysis.

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1. INTRODUCTION

Tolosa Hunt syndrome (THS) is a rare steroid-responsive painful ophthalmoplegia with the palsy of third, fourth and sixth cranial nerves [1,2]. This condition receives its name due to its discovery by Tolosa in 1954 and further research by Hunt who treated six similar patients with steroids [3]. It is characterised by severe unilateral headaches with ophthalmoplegia. The incidence of Tolosa Hunt syndrome is found to be 1 in a million [4]. It has no sex predilection and bilateral involvement is seen only in 5% of the patients. Diagnosis of THS is a tedious process of ruling out all other causes but a fruitful one since it can be cured using steroids.

2. CASE REPORT

A 45-year-old female with no relevant past medical history came to our hospital with complaints of left-sided severe retro-orbital pain extending to the left hemi cranium for 10 days. The boring pain though kept progressing and after 5 days of its onset, the patient developed left eye double vision and ptosis with the development of an apparent squint. On the tenth day, the patient developed sudden loss of vision on the left eye followed by a right eye decrease in vision. There were no complaints of neck stiffness, nausea, vomiting, recent upper respiratory symptoms, numbness, tingling, weakness in any part of the body, no past or family history of headaches, migraines or neurological disease. The patient's vitals were normal. On ophthalmology consultation, there was ptosis and 15% exotropia with no light perception in all quadrants of the left eye with restriction of movements on all sides to 25% and grade 1 Relative Afferent Pupillary Defect (RAPD). The vision in the right eye was the perception of light in all quadrants with a full range of extra ocular movements. Bilateral corneal, trigeminal nerve sensations and fundus were normal. Bilateral superficial temporal and carotid artery pulsations were present and were equal. The rest of the neurological examination was normal. On investigation, her complete blood counts were normal, except for mild leucocytosis (White Blood Cells of 13400/mL) and slight higher range Erythrocyte Sedimentation Rate (20 mm/hr). The chest X-ray was negative for lesions. Cerebrospinal fluid parameters; including mycobacterial DNA and CSF IgG index; were within normal limits. ANCA, ANA and dsDNA were negative. Other bacterial, viral and fungal causes for encephalitis were ruled out. Incidentally, she was diagnosed as diabetic with HbA1c of 12.1% and was started on injectable insulin after consultation with an endocrinologist. Contrast Enhanced Computed Tomography (CECT) scan of Brain was unremarkable except for mild asymmetric thickening of the left optic nerve sheath complex and mucosal thickening of the bilateral sphenoid sinus, left more than the right (Figs. 1a and b).

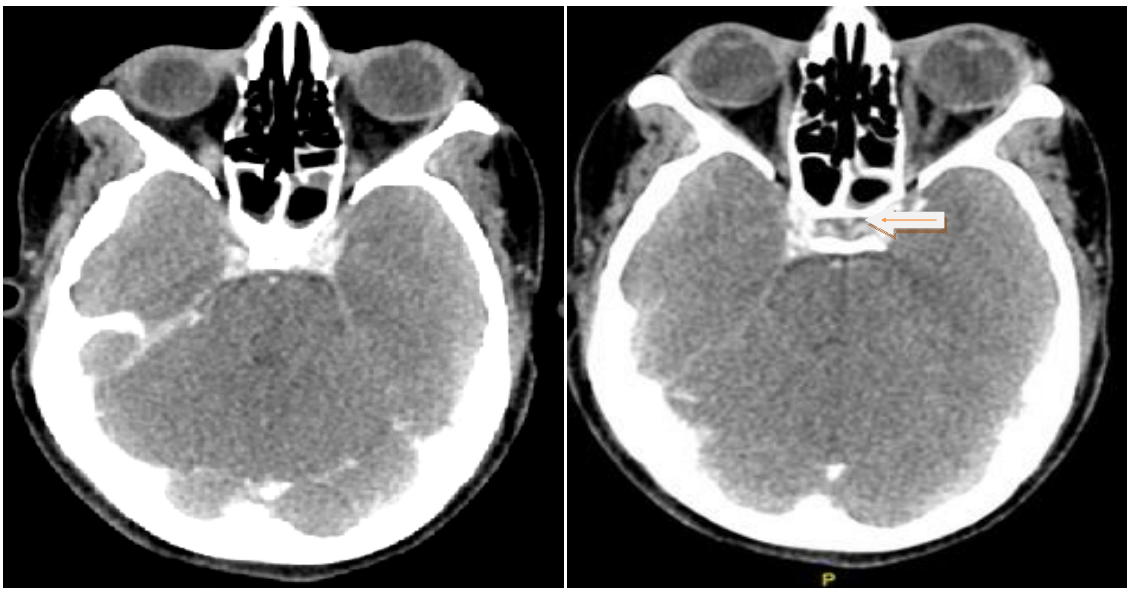

Fig. 1a,b. Left optic nerve sheath complex and mucosal thickening of the bilateral sphenoid sinus

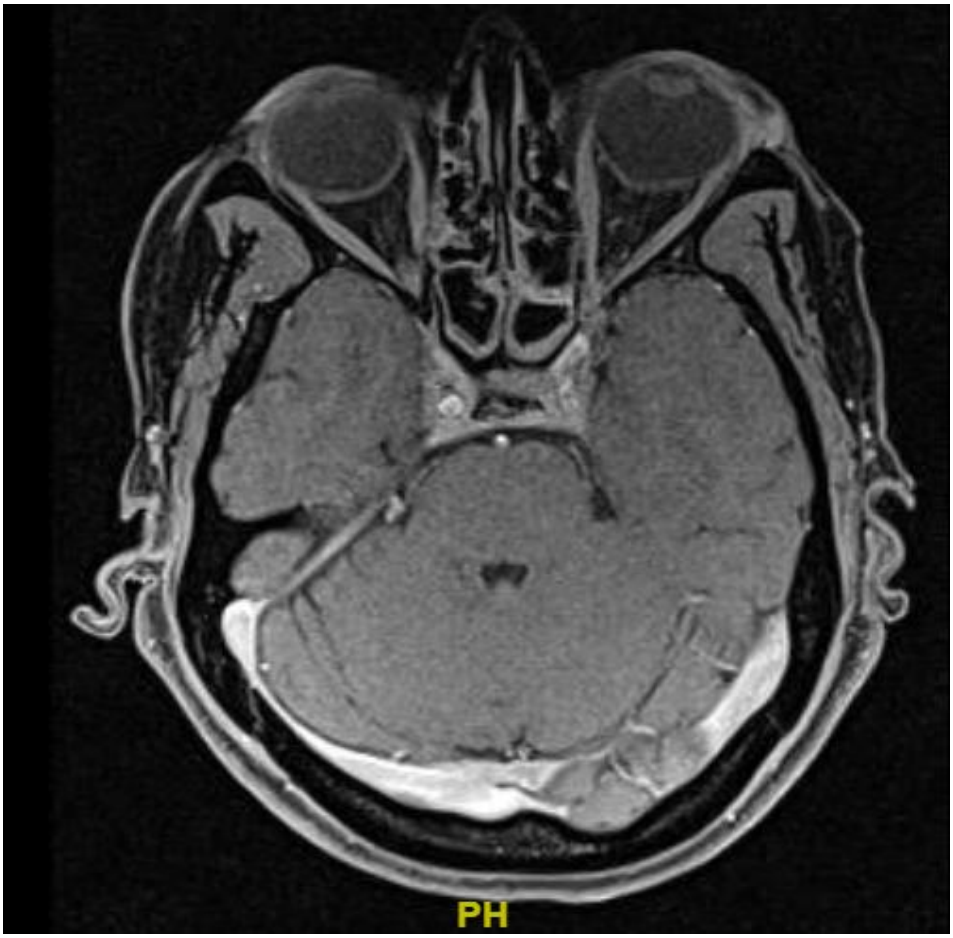

Fig. 2. Magnetic Resonance Venography (MRV) of brain and orbits

Subtle soft tissue density was present around the left superior orbital fissure. Magnetic Resonance Venography (MRV) of Brain and orbits (Fig. 2) showed subtle enhancing soft tissue thickening in the left orbital apex, extending into the left superior rectus.

The left cavernous sinus appeared to be more homogenously enhanced compared to the right. We finally concluded the diagnosis of THS after discussion with Radiologist and Neurologist, by ruling out all other causes of painful ophthalmoplegia.

The patient was started on Intravenous Methyl Prednisolone 1 g Q24 hourly for 5 days. The patient reported that her retro-orbital pain had completely come down by 2 days and had a drastic improvement in the vision with recovery to the perception of fingers in the left eye within 2 days. After 5 days the patient was changed to tablet prednisolone at the dose of 1 mg/ kg and advised to have regular follow up with the neurology OPD and progressive tapering of the drug and strict sugar control and monitoring. At follow up after 3 months the patients had no ophthalmoplegia in her left eye and her vision was 6/12 and 6/ 24 in the right and the left eye respectively. On examination she had a normal direct and consensual light reflex. The patient had no headaches and was under regular follow up with neurology.

3. DISCUSSION

The International Headache Society has made guidelines for the diagnosis of THS which were fulfilled by our patient (International Classification of Headache Disorder {ICHD 3 beta} [5] – unilateral headache, paresis of third, fourth and/or sixth cranial nerves. THS follows a variable course that can last from days to weeks to months.

The etiology of THS is still unknown. A nonspecific inflammation in the region of the cavernous sinus, superior orbital fissure, or orbit provokes THS. In rare cases, the inflammation may spread intracranially [6]. The pain frequently extends to the retro-orbital area as in our patient. It can also present in frontal, and temporal regions with cranial nerve palsies at the onset of pain or within 2 weeks. Pupillary reactions may be normal or there may be sympathetic or parasympathetic involvement. In 30% of cases, there is involvement of the ophthalmic division of the trigeminal nerve. Optic nerve dysfunction has been reported, indicating the orbital apex extension. Infrequently, other cranial nerves may be affected: maxillary and mandibular branches of the trigeminal, facial, or acoustic nerves [6,7]. Our patient also presented with features of sudden onset ipsilateral brow pain and headache with decreased sensations over the forehead, temporal, and frontal regions indicating the involvement of the ophthalmic division of the trigeminal nerve.

THS might have normal MRI findings, but it mostly shows an isointense mass on T1-weighted images and isointense or mildly hypointense signal on T2-weighted images, with enhancement after injection of a contrast medium [6,8]. Usually, the lesion extends to contiguous areas, mainly the sub temporal fossa and orbital apex [9]. On starting steroid, the retro-orbital pain resolves completely within 72 hours [5]. We also found almost same response in our patient. The pathological area also generally decreases in size after corticosteroid therapy [6]. However, some cases do not respond to corticosteroid therapy [8]. Sometimes the therapy relieves the cranial nerve dysfunctions within 6–8 weeks with corticosteroids and residual nerve palsies rarely persist [6,7]. The disappearance of symptoms following the systemic corticosteroid treatment may precede the normalization of neuroradiological studies by weeks or even several months [4]. Recurrences are common and can either be unilateral or bilateral [8]. Our patient also showed improvement of symptoms and vision within two days of steroid therapy.

4. CONCLUSION

Headache with ophthalmoplegia is an unusual emergency which is not routinely encountered. THS has to be always kept as a differential diagnosis for such conditions, after ruling out more common aetiologies for retroorbital pain with multiple nerves palsy. Multidisciplinary approach is warranted to diagnose such entity in early time. Once diagnosed early, we can reverse its course by giving high dose of steroids. Regular follow up is required to check for the recurrence.

CONSENT AND ETHICAL APPROVAL

As per international standard or university standard guideline patients consent and ethical approval has been collected and preserved by the authors.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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Peer-review history:
The peer review history for this paper can be accessed here:
http://www.sdiarticle4.com/review-history/69142