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AN INTRIGUING CASE OF PAINFUL EXTERNAL OPTHALMOPLEGIA IN A WOMAN- BILATERAL TOLOSA-HUNT SYNDROME

Neurology

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ABSTRACT

Tolosa-Hunt syndrome is a rare, cause of reversible and painful ophthalmoplegia characterized by recurrent unilateral orbital pain, ipsilateral oculomotor paralysis and prompt response to steroids. We describe a patient who presented with painful ophthalmoplegia and was ultimately diagnosed via contrast enhanced magnetic imaging resonance studies and was successfully treated for Tolosa-Hunt syndrome. High index of suspicion of the disease allows for immediate intervention and thus decrease the length and severity of symptoms .We describe the case presentation and keys for diagnosis emergency medicine that physicians should know for this potentially devastating condition.

KEYWORDS

Tolosa-Hunt Syndrome, Ophthalmoplegia.

INTRODUCTION

Tolosa-Hunt syndrome (THS) is a diagnosis of exclusion and a rare but important steroid-responsive etiology for painful ophthalmoplegia. Tolosa described this entity in 1954 and Hunt later on explored further where he published 6 cases of remittent unilateral retro-orbital pain with extraocular nerve palsies which were steroid responsive. The estimated annual incidence is one case per million per year. (1) It presents as unilateral orbital pain associated with palsy of either or more of the oculomotor, trochlear and abdomens cranial nerve, leading to external ophthalmoplegia which can cause diplopia. The basic pathology described being underlying granulomatous inflammation of anterior portion of cavernous sinus.(2) Being a diagnosis of exclusion one must consider and rule out more sinister disease processes including malignancy, infection or vascular occlusion. Rapid recognition of the condition through neuroimaging studies allows for therapeutic intervention thereby decreasing the morbidity of the illness. There is no specific presentation other than ophthalmoplegia and may afflict patients of any age and gender. (3)

CASE REPORT

A 30-year-old woman, with no existing co-morbidity presented to us with right periorbital eye pain with visual disturbances in the form of drooping of right eye lid with binocular diplopia on looking medially and upward. The pain was right peri-orbital which was throbbing, continuous, moderate to severe intensity without any aura with episodes of extremely sharp pain worsened with even gentle palpation of the eye since 15 days, although there was no proptosis, diminution of vision. The patient was seen at a tertiary care centre where Magnetic Resonance Angiogram Brain was done to rule out any PComm aneurysm, Magnetic Resonance Venogram didn't reveal any cavernous sinus thrombosis of any sinus or bleeding. She denied prior history of head injury or other cranial nerve involvement, fever, illness, foreign travel, nausea, vomiting, vertigo, or any constitutional symptoms. Examination at admission revealed stable haemodynamic parameters. Nervous system examination revealed right ptosis with right complete external ophthalmoplegia with right pupil being semi-dilated, sluggishly reactive to light and indirect light reflex was present (Figure 1, 2,3). There was mild ptosis on left side also with normal extraocular movements in all gaze. Left pupil was 2 mm, reactive to light and indirect reflex was present. Intraocular pressure were normal. Bilateral fundi were normal. Rest other neurological evaluation was normal. MRI Brain with contrast revealed contrast enhancement in right cavernous sinus with no evidence of thrombosis or bleed or infarct or sinusitis or orbital fat enhancement. Serum ACE levels were normal. ANA, Anti-Ds DNA were negative. Repetitive nerve stimulation did not reveal any decremental response. ESR was 88. Cerebro Spinal Fluid analysis revealed normal study with gene X pert for Tuberculosis was negative. Visual evoked potentials were within normal limits. A diagnosis of Tolosa-Hunt syndrome was made and patient was managed with steroids. Our patient responded well to IV methylprednisolone 1gm, and on day 3, she had almost complete resolutions of her pain with improvement in ptosis and ocular movements on further follow up in Out Patient Department.

Figure 1. Bilateral ptosis (R>L).

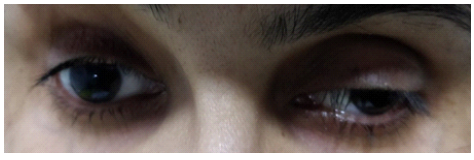

Figure 2. Right medial rectus palsy is evident on left horizontal gaze whereas left eye extraocular movements are normal.

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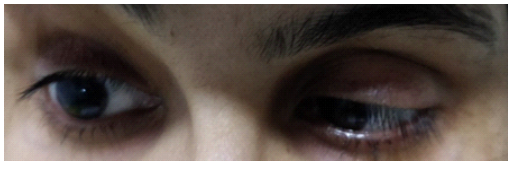

Figure 3. Mild Right lateral rectus palsy is evident on right horizontal gaze whereas left eye extraocular movements are normal.

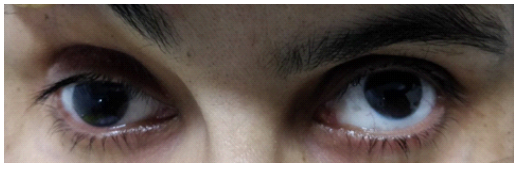

Figure 4. Right superior rectus palsy is evident on left horizontal gaze whereas left eye extraocular movements are normal.

DISCUSSION

Patients with Tolosa-Hunt Syndrome often present with severe peri-orbital pain with opthalmoplegia which can involve the cranial nerves III,IV,V and VI. Its underlying pathology is an idiopathic granulomatous inflammation of the cavernous sinus. It is a diagnosis of exclusion, however neuroimaging can be used to detect inflammatory changes in the cavernous sinus or orbital fissure and a biopsy can be

used to confirm the diagnosis. Biopsy is rarely required as the clinical setting and the response after steroid administration is quite characteristic. The International Headache Society laid guidelines for diagnosis of Tolosa-Hunt syndrome which included retro-orbital pain, localized around the ipsilateral brow and eye 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. (7) Its course is variable and can last from days to weeks to months. Recurrences are not uncommon and can manifest as unilateral or bilateral manifestations. (8) Other causes need to be excluded by appropriate investigations. The retro-orbital pain shows complete resolution usually within 72 hours of onset of steroid treatment, but time needed for clinical improvement in opthalmoplegia varies considerably, may take up to 4-6 weeks.(9) Possible common differentials to cause both orbital and facial pain include infections, orbital cellulitis, cavernous-sinus thrombosis and various neoplastic diseases. Magnetic resonance with contrast enhancement and fat suppression techniques are the best imaging tools for a patient with painful ophthalmoplegia. It often detects inflammatory changes and mass-like lesions in the anterior cavernous sinus with or without extension of the changes into the superior orbital fissure and orbital apex. We believe that this case signifies the importance of considering Tolosa-Hunt syndrome as a well-recognized and easily treatable etiology for retro-orbital pain with oculomotor nerve palsy after all other diagnostic modalities rule out more common etiologies. Since Tolosa-Hunt syndrome responds well to steroid treatment, early detection is beneficial. No standard steroid dosage has been established. Younger patients have faster improvement in their neurological deficit. (10) The clinical course of Tolosa-Hunt syndrome in our patient confirms the possibility of bilateral infiltration of the supraorbital fissures and cavernous sinuses. Therefore, although criteria of the syndrome classify unilateral lesions as typical, we believe that the differential diagnosis in patients with bilateral, painful ophthalmoplegia should include Tolosa-Hunt syndrome. (11)

CONCLUSION

Tolosa-Hunt syndrome is a reversible and painful ophthalmoplegia characterized by recurrent unilateral or bilateral orbital pain, ipsilateral oculomotor paralysis and prompt response to steroids. While the differential diagnosis of both sudden vision loss and eye pain is broad, there are few specific causes of unilateral painful vision loss. Prompt recognition of the disease through appropriate diagnostic imaging and laboratory studies can allow for immediate intervention and thus decrease the length and severity of symptoms including vision loss. Patients should see an ophthalmologist as soon as possible and be started on high dose corticosteroids immediately.

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