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CASE REPORT OF TOLOSA HUNT SYNDROME IN ELDERLY FEMALE

General Medicine

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ABSTRACT

We describe a patient who presented with unilateral orbital pain, unilateral headache and partial ptosis of left eye who was successfully treated for Tolosa-Hunt syndrome(THS). Early recognition of THS can allow for immediate treatment and thus reduce the severity of symptoms.
Symptoms may not spontaneously resolve without treatment and leads to unnecessary suffering. Corticosteroids should be started as soon as THS is diagnosed.

KEYWORDS

Tolosa Hunt Syndrome, Unilateral Orbital Pain, Corticosteroids

Introduction

THS is very rare and reversible cause of recurrent unilateral orbital pain and ipsilateral oculomotor palsy. THS responds very well to steroids within 72 hours. Other features include unilateral headache, ptosis and diplopia.[1]

The clinical features are varied in cases of THS. It can affect patients of any age and sex. There are no specific diagnostic tests for THS and it remains as a diagnosis of exclusion. [2]

Magnetic Resonance Imaging(MRI) and Computed Tomography(CT) Scans of the brain and orbits can be done to look for any inflammatory changes present in cavernous sinus or superior orbital fissure. Quick diagnosis of the disease with appropriate imaging techniques and laboratory investigation is helpful in starting the treatment. Some symptoms may resolve without any intervention, while delaying treatment can cause unnecessary suffering to the patient. [3]

Case Presentation

A 65 years old female patient came to Medicine Department with chief complaints of unilateral headache, orbital eye pain, partial ptosis on left side. Patient also had diplopia since three weeks.

Headache was throbbing and sharp shooting and was localized in left frontal, parietal and temporal regions. (Below photograph).

Patient did not give any history of nausea, vomiting, difficulty in swallowing, respiratory problems or fever. Patient had no history of diabetes, hypertension, thyroid disorder and tuberculosis. Patient was not on any medications except, for pain relief.

On general examination, patient was vitally stable. Third cranial nerve paresis was noted on neurological examination.

Hemoglobin was 11 g/dl, total count 9000/cu. mm, platelets 2.4 lakh/cu. mm, ESR was 11 mm/hour and CRP was 4 mg/litre.

Magnetic Resonance Imaging(MRI) along with Magnetic Resonance Angiography (MRA) of brain and orbits was done, which was within normal limits. However, mild thickening of left cavernous sinus was seen. Cerebrospinal fluid(CSF) did not show any significant finding.

So, THS was considered and patient was given Prednisolone 40 mg per day for seven days and called the patient for follow up after 7 days. On follow up, all her symptoms subsided. Pain was relived and there was no ptosis or diplopia.

Discussion

THS is a rare disorder characterized by hemicranial or periorbital pain associated with ophthalmoplegia and has dramatic response to corticosteroid therapy. [4] It was described by Tolosa and Hunt in 1954 and 1961 as an idiopathic granulomatous inflammatory process. [5,6]

In 1988, Tolosa Hunt Syndrome diagnostic criteria were provided by the International Headache Society (IHS), and again changed in 2004.They are:

1. One or more episodes of unilateral orbit pain which continues for weeks if untreated.
2. Paresis of one or more of the third, fourth and/or sixth cranial nerves and/or presence of granuloma of cavernous sinus by MRI or biopsy.
3. Paresis coincides with the onset of pain or within 2 weeks of onset of pain.
4. Pain and paresis resolve in 72 hours when treated with steroids.
5. Other causes have been ruled out by proper investigations. [7]

MRI is an important part of the workup of any patient presenting with features of Tolosa Hunt Syndrome, as these features are non-specific and have many differential diagnosis, including meningioma, ophthalmoplegic migraine, herpes zoster, tuberculous meningitis (TBM) and lymphoma. [8]

Tolosa Hunt Syndrome remains a diagnosis of exclusion. The patient described above did not show the classical MRI findings and was diagnosed on the basis of exclusion and response to steroid therapy. The role of MRI also, is to exclude other conditions having similar clinical features.

Conclusion

Tolosa-Hunt syndrome is a reversible disease characterized by recurrent unilateral orbital pain, ipsilateral oculomotor paralysis and fast response to steroids. Early recognition of the disease by appropriate diagnostic imaging and laboratory studies will allow immediate treatment and reduce the severity of symptoms including vision loss. Patients should be started on corticosteroids immediately, once diagnosed as having THS.

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