ORIGINAL RESEARCH PAPER

Medicine

UNUSUAL PRESENTATION OF AN UNUSUAL DISEASE-TOLOSA HUNT SYNDROME

KEY WORDS: Tolosa-Hunt syndrome, unusual presentation

Dr Sanjay P Khare*

MD (internal medicine; Director- Bariatric Medicine; Chief Consultant in Internal Medicine; Coordinator-Academic and Clinical training-Apollo Hospital Navi Mumbai.* Corresponding Author.

Dr Nazia Khan

Physician assistant-Apollo Hospital Navi Mumbai.

ABSTRACT

The Tolosa-Hunt syndrome is a rare syndrome with an estimated annual incidence of one case per million per year [1]. It is recognised as a rare disorder by the National Organisation for Rare Disorders (NORD). It presents as a severe periorbital headache along with restricted eye movements. The presentation is mostly unilateral, with either side being affected equally. Very rarely (in 5% cases), there's a bilateral involvement. We present a case of Tolosa Hunt which had some irregularities, making it a rare presentation of a rare case.

INTRODUCTION

Tolosa Hunt syndrome is believed to occur following a non-specific inflammation in the region of the cavernous sinus and/or superior orbital fissure. Most of the times cause is labelled as idiopathic. Other possible aetiologies/triggering factors might be trauma, tumour (2), aneurysm, connective tissue disorders etc. It was first described by Dr. Eduardo Tolosa in 1954, in Spain. (3) He described it as "inflammation within cavernous sinus with the proliferation of fibroblasts and infiltration with lymphocytes and plasma cells."

In 1962, Hunt et al. endorsed this syndrome and also noted that "inflammatory changes within a tight compartment exerted pressure upon the cranial nerves III, IV, VI, as well as the superior division of the fifth cranial nerve. Attention was drawn towards granulomatous deposits along with epithelioid cells and giant cells. (4)

From 1966 onwards, this came to be known as the Tolosa-Hunt syndrome. (5)

Classical presentation is severe unilateral retro orbital pain with extra ocular nerve palsies. Facial sensation and visual acuity may be diminished. Usually there is high ESR. Neuro imaging shows typical features. A sharp response to steroid is characteristic.

CASE REPORT:-

Mr H.M, M/76 Years was a known case of
1) Type II Diabetes on Tablet Vildagliptin+ metformin 50/500 twice daily,
2) Hypertension on Tablet Metoprolol XL 25 mg 1 OD.
3) Ischaemic Heart Disease (PTCA done in 2018) on tablet Aspirin 75 mg 1 OD+ Rosuvastatin 10 mgs.
4) Bronchial asthma On Budesal Nebulisation 0.5 mcg thrice daily.

He had had Bell's palsy in 2019, from which he had recovered.

Mr HM presented with complaints of
(a)Drooping of right eyelid since last 3 days. (b) Reduced visual acuity and (c) Swelling of right eyelid. (d) Tingling and numbness-right forehead. On direct questioning, he confessed to have diplopia.

Despite recurrent questioning, there was NO orbital pain/ discomfort.

On **Examination:** There was ptosis of the right eyelid. Vision was restricted to finger counting. There was a loss of corneal reflex on affected side. The right pupil rested in right lower and outer hemisphere with patient unable to adduct it. Pupillary light reaction was compromised.

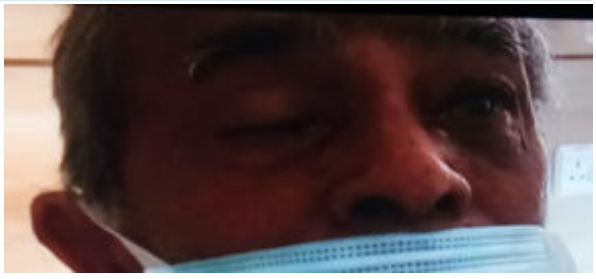
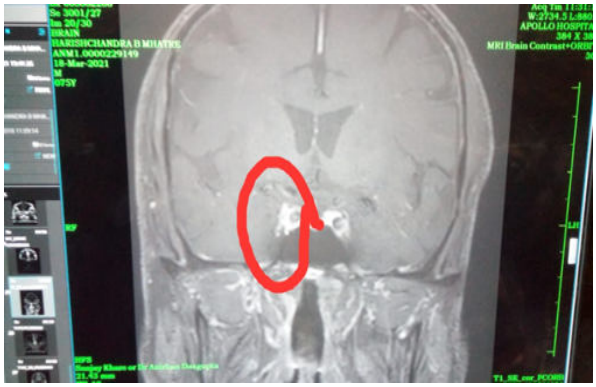
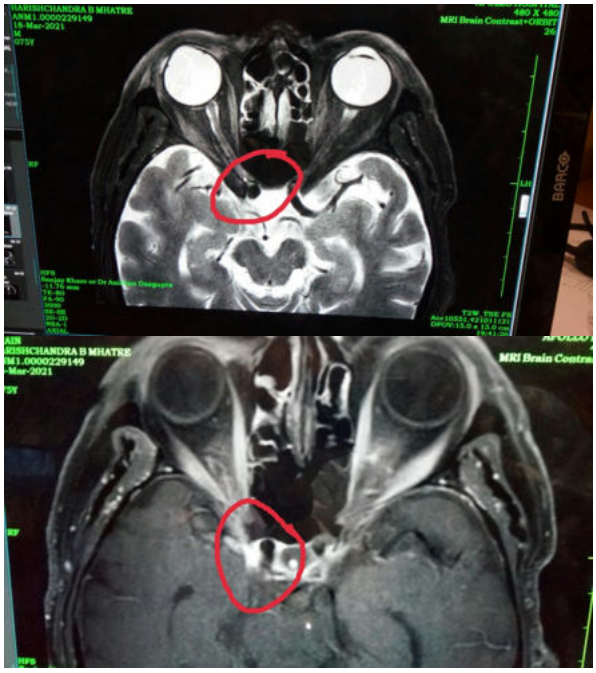


Table Of Investigations:

Creatinine	0.8
Random blood sugar	171
SGOT	22
SGPT	20
ESR	13
CB-NAAT FOR COVID-19	Negative
A N A BY IFA	Negative
CSF REPORT	Red blood cells= 37 White blood cells =01 No xanthochromosia Glucose =75 Protein =52 Gene xpert for mycobacterium tuberculosis not detected
NT PRO BNP	1527
2D ECHO	Suggestive of status post PTCA ischemic disease changes. Paradoxical motion +. Left ventricular ejection fraction 50% Grade I diastolic dysfunction.

Radiological Findings :- MRI Brain With Orbital Cuts With Contrast





MRI Brain Showed:
Mild soft tissue fullness and enhancement in the lateral aspect of the right cavernous sinus, representing inflammatory aetiology. Signal alteration in the posterior right optic nerve and surrounding fat stranding suspicion for optic neuritis. Mild fat is also seen around the left optic nerve. No acute infarct. No mass effect or abnormal enhancement noted in the neuroparenchyma. Age related mild chronic small vessel ischemic changes.

Diagnosis :-
Tolosa Hunt Syndrome. Most peculiarly, there was no pain at any stage of his illness.

Treatment Plan:
His usual medications were continued.

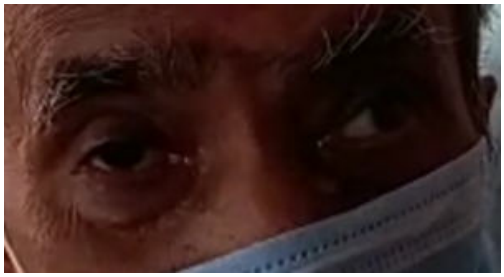
Steroids are the mainstay of management. In fact, steroids are looked upon as diagnostic AND therapeutic in Tolosa-Hunt syndrome. (6,7,8). Resolution of clinical features along with imaging abnormalities following a course of systemic corticosteroids is looked upon as "diagnostic" of Tolosa-Hunt syndrome.

Once his CSF was documented to be clear without any infections etc., Injection Solumedrol (500 Mg in 100 Ml Ns over 45 Minutes BD) was started.

Predictably, the sugars rose following steroids, which were countered with appropriate insulin doses as per sugar monitoring.

Pt was discharged after 3 days of IV solumedrol and on day 6 of admission.

Course And OPD Follow Up:-



He was reassessed on day 10 of illness, in the OPD. There was a significant resolution of symptoms and signs. The Ptosis had resolved. There was a slight improvement in the adduction of right eye pupil. The right eyelid swelling had abated to a huge extent.

CONCLUSION:-
Tolosa-Hunt syndrome is in itself a rare condition. In this case, there was further peculiarity in that there was NO pain at any stage of his illness. To the best of our knowledge, there are hardly any recorded cases like these, in the medical literature.

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