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“Hunting” a case of External Ophthalmoplegia: TOLOSA-HUNT SYNDROME

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Clinical History

A 35-year-old female, who is a housewife, hailing from Mysuru district came with complaints of pain in the left eye, sudden in onset, retro-orbital, dull aching, continuous, radiating to the left hemicranium since 15 days. One week later, she developed diminution of vision in the left eye which was insidious in onset and progressive.

The patient also complained of diplopia since 15 days.

The patient complained of numbness on the left side of the forehead since a week.

The patient had no h/o photo-phobia, phonophobia, nausea, or vomiting.

No h/o excessive watering of eyes, drooping of eyelids, painful eye movements.

No h/o similar complaints in the past.

Examination

A middle-aged woman, moderately built, and nourished is conscious, co-operative, well oriented to time, place, and person.

Vitals-

PR: 84bpm, BP: 120/80mm Hg, RR: 20cpm, Temperature: Afebrile

No pallor/icterus/cyanosis/clubbing/lymphadenopathy/edema

Central Nervous System-

Higher Mental functions: Conscious and oriented.

Cranial Nerves:

Optic nerve- Visual acuity of 6/18 in the left eye and 6/12 in the right eye.

Relative Afferent Pupillary Defect + in LE

Color vision and field of vision are normal.

Oculomotor, Trigeminal, and Abducent nerves- The patient has restricted movement in the Left eye in abduction (Lateral Rectus), Depression, and Intorsion (Superior Oblique).

Ocular position-Esotropia of the left eye.


Image showing Esotropia of the left Eye

Aperture, Pupil size, and accommodation are unremarkable.

No nystagmus or Ptosis.

Trigeminal nerve- Loss of sensation over the V1 branch.

Other cranial nerves- Intact.

Motor system: Normal bulk, tone normal in both upper and lower limbs, Power 5/5 in both upper and lower limbs, plantars-flexor bilaterally.

Sensory system: unremarkable.

Cerebellar signs and meningeal signs absent.

Peripheral nerve examination-Normal.

Fundoscopy: Normal fundus. No papilloedema.

Cardiovascular System: S1 S2 +

Respiratory System:B/L Normal vesicular breath sounds

Per Abdomen-Soft, non-tender, no organomegaly

Investigations

Hemoglobin-12.8 g/dl

Total leucocyte Count-8900 cells/cumm

Platelet count-3.86 lakh/cumm

T3-1.12 ng/ml

T4-9.0 mcg/ml

TSH-1.12 mcIU/ml

PT-14.5 sec

INR-1.13

Blood urea-23 mg/dl

Serum creatinine-0.8 mg/dl

CSF Fluid analysis-

:cells- 10(lymphocytes)

: Glucose-71 mg/dL

: Protein-122.6 mg/dL

: Chloride-116 mmol/L

:Gram stain-Nil

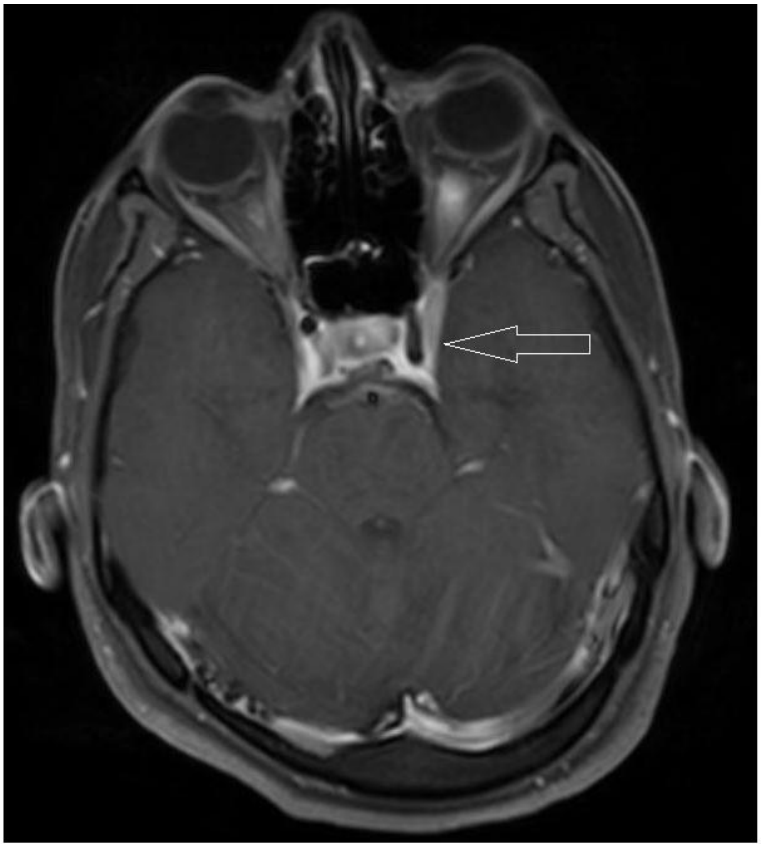
:KOH-No fungal elements

:Acid Fast Bacilli not seen.

MRI BRAIN(plain and contrast): Enhancing soft tissue lesion in left cavernous sinus with extension and mild compression and medial displacement of the optic nerve at the orbital apex.

Mildly hyperintense left lateral rectus muscle.

Bulky left lacrimal gland- Tolosa Hunt Syndrome to be considered.



MRI Brain – Arrow shows enhancing soft tissue lesion in Left Cavernous Sinus

Diagnosis

TOLOSA-HUNT SYNDROME –ORBITAL APEX SYNDROME

Treatment

The patient was started on IV Dexamethasone and switched over to oral corticosteroids which is the mainstay of treatment.

Treatment given:

INPATIENT: INJ DEXAMETHASONE 4mg IV 1-1-1 for 5 days f/b 1-0-1 for 2 days

OUTPATIENT: TAB DEXAMETHASONE(1mg/kg body wt) 40mg 1-0-0 for 1 week f/b

30mg 1-0-0 for 1 week f/b

20mg 1-0-0 f/b

10mg 1-0-0 for 1 week then stopped.

Usually, oral steroids are tapered over 4-6 weeks according to the steroid response of the particular patient.

Discussion

DEFINITION: It is a syndrome of painful ophthalmoplegia which is characterized by periorbital and/or hemicranial pain, palsy of the ocular motor nerves (III, IV, and VI) on the same side, loss of sensation along with the ophthalmic division of the V cranial nerve, occasionally the maxillary division due to chronic non-specific inflammation.
Superior orbital fissure syndrome refers to symptom complex produced by the involvement of the III, IV, VI, and V1 cranial nerves.

Orbital apex syndrome refers to the symptom complex produced by the involvement of structures passing through superior orbital fissure and optic canal, characterized by involvement of II, III, IV, VI, and V1 cranial nerves[1].

The incidence of Tolosa-Hunt syndrome is equal among both the sexes with a prevalence of 1-2 cases per million.[2]

CLINICAL FEATURES:[1]

1. Pain-unilateral, periorbital, retro-orbital and/or hemi-cranial.
2. Ophthalmoplegia, i.e, the involvement of the III, IV, VI cranial nerves with or without the involvement of the II cranial nerve on the same side.

3. Vision may or may not be affected.
4. Ptosis may occur due to the involvement of III cranial nerve on the same side.
5. Loss of sensation over the forehead and face may occur due to the involvement of the ophthalmic division, occasionally maxillary division of the V cranial nerve, on the same side.
6. Uncommonly, patients may also have systemic features like nausea and vomiting, due to severe pain.

DIAGNOSTIC CRITERIA(IHS CLASSIFICATION ICHD-3 BETA)[3]

1. Unilateral headache
2. Including both of the following criteria:
3. MRI or Biopsy showing the presence of granulomatous inflammation of the cavernous sinus, superior orbital fissure, or orbit.
4. Ipsilateral palsy of one or more Occulomotor nerves(III, IV, VI)

3. Headache which is not explained by any other etiology.

Although according to the diagnostic criteria Tolosa Hunt syndrome is unilateral ophthalmoplegia, there have been few reported cases of bilateral ophthalmoplegia, evidenced by the fact there was a response to steroids and the symptoms not explained by any other conditions[4].

DIFFERENTIAL DIAGNOSIS FOR PAINFUL OPHTHALMOPLEGIA:[1]

1. Inflammatory causes: Rhinocerebral mucormycosis, bacterial sinusitis, orbital cellulitis, Herpes Zoster, Mycobacterial tuberculosis, Sarcoidosis, Tolosa-Hunt Syndrome, Wegener’s Granulomatosis.
2. Vascular causes: Carotid-cavernous fistula, Carotid-cavernous thrombosis, carotid artery aneurysm.

3. Neoplastic causes: Meningioma, Pituitary adenoma, Nasopharyngeal carcinoma, Craniopharyngioma, Neurofibroma.
4. Trauma

Rhinocerebral mucormycosis is the most frequent cause for cavernous sinus syndrome, which has to be ruled out in patients with Diabetes, transplantation, prolonged neutropenia, or malignancy, with complete ENT examination and MRI brain[5].

INVESTIGATIONS:[1,5]

1. Hematology: Complete hemogram, Liver and Renal function tests, Blood glucose, Erythrocyte Sedimentation Rate, C-Reactive Protein, Hemoglobin A1c, Treponemal tests, Antinuclear Antibody Profile, protein electrophoresis, Angiotensin-Converting Enzyme levels.
2. Cerebrospinal fluid: pressure, Cell count, Differential count, Microscopy, Glucose, Protein, Chloride Culture, Cytology, Angiotensin-converting enzyme, ADA.
3. Neurological studies: Magnetic Resonance Imaging, Computed Tomography, Cerebral Angiography

When MRI is being used to detect the presence of granulomatous tissue in the orbital region, the T1 sequence can be used with slice thickness being as low as 2mm, as the granulomas are not larger than 1 or 2mm.[4]

4. Biopsy: Cavernous Sinus

TREATMENT: The mainstay of treatment is glucocorticoids. Oral prednisone (60mg daily) is given for 2 weeks and gradually tapered over a month or longer if the pain is still persistent[5].

There is a dramatic response to the steroids in terms of the symptoms, the pain is relieved within 24-72 hours, cranial nerve palsies improve gradually over 2-8 weeks.

Some patients may also require immunosuppressive drugs such as Azathioprine, Methotrexate, Mycophenolate mofetil, Cyclosporine, Infliximab for either suppression of the disease process or to avoid the long term effects of steroid therapy.

In patients with contraindications to Steroid therapy, Radiotherapy can be used as the first line of management after a biopsy-proven diagnosis of Tolosa-Hunt syndrome.

Relapses can occur in 40-50% of the patients and are more common in younger patients. The role of steroids in relapse is unclear[3].

References

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