AN UNUSUAL CASE OF TOLOSA HUNT SYNDROME

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ABSTRACT

Tolosa-Hunt syndrome is a rare disorder characterized by severe periorbital headaches, along with decreased and painful eye movements (ophthalmoplegia).

Symptoms usually affect only one eye (unilateral). In most cases, affected individuals experience intense sharp pain and decreased eye movements [(3)].

Key words –Tolosa hunt syndrome - extensive involvement

CASE

His family physician referred him to our centre.

On examination it was found that his left pupil was non reacting and he was completely blind in his left eye (no PL).

A 62-year-old diabetic patient, weighing approximately 55 kg presented with acute onset ptosis. This was after about 2 weeks of his 2nd dose of his Covid vaccination. Progressively he developed painful complete ophthalmoplegia in 48 hours.

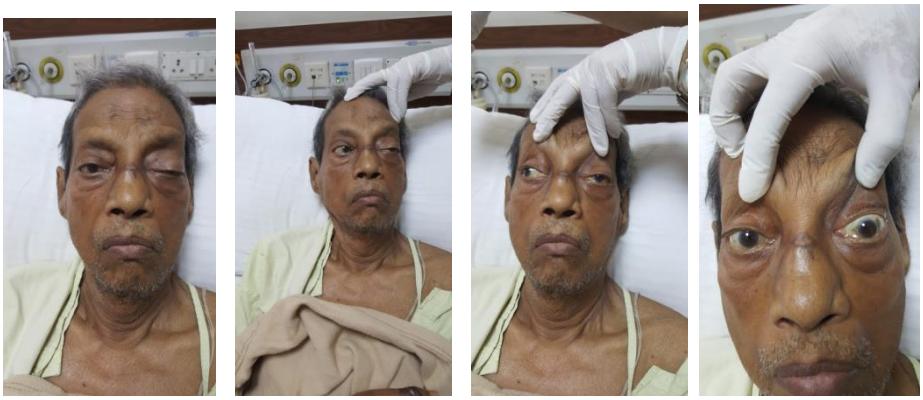

Figure -1

SHOWING THE PTOSIS AND OHTHALMOPLEGIA.

Also surprisingly on examination it was found that his left pupil was not reacting and he was completely blind in his affected side, that his left eye.

The ophthalmoplegia is shown in the figures below.

Routine investigations did not reveal any abnormality except a small rise in CRP and leucocyte counts. All his immune and paraneoplastic markers were negative.

An MRI [(2)] was done (pictures given below) and it was seen that he was having Superior orbital syndrome (Tolosa Hunt) and the extent was up to Cavernous sinus, explaining the highly compromised optic nerve as well, which is not a common finding.

CSF study was not done.

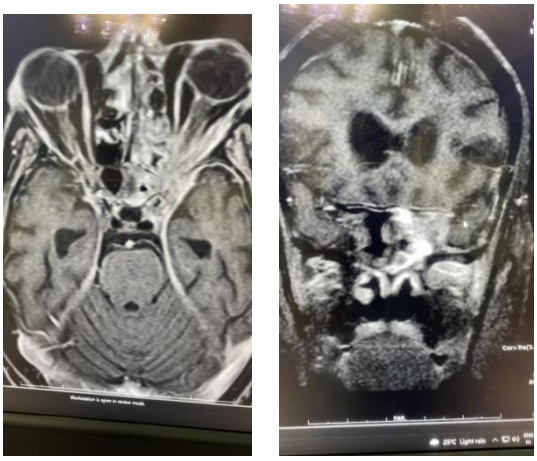

Figure -2

MRI SHOWING THE LESION AND EXTENSION OF THE EXUDATES TO CAVERNOUS SINUS.

COURSE

He was immediately started on 1gm of Methyl-prednisolone for 5 days, keeping his diabetes and electrolytes monitored.

He was then shifted to oral gluco-corticoids at 1mg/kg and planned to continue for 2 weeks and then progressively taper off by 5 mg each week.

Progressively in 5 days he could visualise light and faintly see movement of hand, although could not count fingers. However, at discharge, on 10th day, he could move slightly the left eye (only down and up, not laterally or medially) and could faintly count fingers. He developed a mild degree of probably steroid psychosis for which quetiapine was added and was stable.

Repeat MRI showed slight reduction in Exudates (Peri superior orbital fissure, and no increase in muscle oedema. The compression over Optic nerve was reduced).

Repeat VEP did not show any change. (No signal detected in left eye).

DISCUSSION

Anatomical explanation of Tolosa Hunt syndrome.

The oculomotor nerve enters the orbit through the superior orbital fissure. It then divides into two branches between the lateral rectus: superior and inferior rami. The oculomotor nerve is inferior to the trochlear nerve and the nasociliary nerve runs in between the two rami.

It is divided into 3 parts from lateral to medial:

The space transmits: superior ophthalmic vein, lacrimal nerve, frontal nerve, trochlear nerve (CN IV), recurrent meningeal branch of lacrimal artery (anastomotic branch of lacrimal artery with the middle meningeal artery)

Definition:

Tolosa–Hunt syndrome (THS) is a **rare disorder** characterized by severe and unilateral headaches with orbital pain, along with weakness and paralysis (ophthalmoplegia) of certain eye muscles (extraocular palsies).

Symptoms:

Affected individuals may exhibit signs of paralysis (palsy) of certain cranial nerves such as drooping of the upper eyelid (ptosis), double vision (diplopia), large pupil, and facial numbness. The affected eye often abnormally protrudes (proptosis). The exact cause of Tolosa-Hunt syndrome is not known, but the disorder is thought to be associated with inflammation of specific areas behind the eye (cavernous sinus and superior orbital fissure).

Diagnosis:

Tolosa Hunt syndrome is diagnosed through the clinical presentation, neuroimaging studies.

Laboratory tests and cerebrospinal fluid (CSF) studies are supportive tests but help in ruling out other causes of ophthalmoplegia

Treatment:

He hallmark is steroids, glucocorticoids [(1)], more specifically.

Cause:

It's not very clear but supposedly an autoimmune disease. However common immune markers may not be detected.

DIFFERENTIAL DIAGNOSIS:

Orbital apex syndrome is characterized by vision loss from optic neuropathy and ophthalmoplegia due to the involvement of ocular motor nerves in the anatomical region of the orbital apex

Prognosis:

Left untreated, symptoms may resolve spontaneously after an average of about eight weeks.

UNUSUAL ELEMENT IN THIS CASE:

Normal vision is not affected in Tolosa Hunt. But n this case it was likely due to that the left sided blindness was caused by the extension of the inflammatory process to the left optic nerve at its intracranial, intra-canalicular or proximal orbital segment.

SUMMARY

Tolosa Hunt is hence an uncommon autoimmune disorder causing unilateral painful Ophthalmoplegia.

However, in this case there was associated blindness which is unusual. Hence if such a patient complains of vision urgent imaging and glucocorticoids should not be delayed. [4]

Although we initially tried to link this to his post Covid vaccination, but usually post vaccination reactions are rare after 5-7 days of vaccination.

But the carry home message is to identify the condition early and start steroids as the extension of infiltrates, as in this case can extend to Cavernous sinus and cause visual loss.

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