Case Report

A case report of Tolosa — Hunt syndrome

M Senthilvelan[1], Jayaram Kosalram[2], D Kanagaraj[3]

Tolosa Hunt syndrome is a rare clinical entitycharacterized by sudden onset of painful ophthalmoplegia and prompt response to steroid therapy[1]. Generally it involves third, fourth and sixth cranial nerves due to the presence of non specific granulomatous inflammatory process in the region of cavernous sinus and /or superior orbital fissure[2,3]. Here we present a case report on a 17 year old female who presented with Unilateral headache, drooping of left eyelid and diplopia. MRI brain of the patient showed Left cavernous sinus enlargement and T1 & T2 isointense lesion causing narrowing of Left ICA extending anteriorly to the orbital apex. The diagnosis of Tolosa Hunt syndrome was made after ruling out other differentials on the basis of history, examination and investigations.

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***Key* words : Ophthalmoplegia, ptosis, diplopia, tolosa hunt syndrome, cavernous sinus.**

Tolosa Hunt syndrome is a rare neurological entity which can affect people of any age with no sex predilection[4]. It is a steroid responsive painful ophthalmoplegia which was first described by Tolosa in 1954[5]. In 1961, Hunt *et al* reported six cases of painful ophthalmoplegia which rapidly improved with steroids and in 1966, Smith and Taxdal gave theeponym "Tolosa Hunt syndrome" to this entity[4]. The criteria for diagnosis of Tolosa Hunt syndrome is framed by the International Headache society and modified in 2013[6]. Although it is considered as a benign condition with prompt response to steroid therapy, sometimes patients may require prolonged immunosuppressive therapy[7].

Case Report

A 17 year old female presented with left sided Headache with double vision, Fever and drooping of left eyelid for 10 days. Headache was constant, dull aching nature in the left sided periorbital region extending to the left frontal region. There was no history of visual loss and sensory disturbances over face. Fever was low grade, intermittent, relieved by antipyretics.

The patient was conscious, oriented with Pulse rate of 98/mt, Blood Pressure of 100/70 mmHg, Respiratory rate of 15/mt, Temperature of 100°F.

Cranial nerve examination showed partial ptosis, restricted ocular movements and dilated non reacting (loss of both direct and indirect light reflex) pupil in the left eye suggestive of third, fourth and sixth cranial nerve paresis. Other cranial nerves were normal. Higher mental function, motor and sensory system were intact. No signs of meningeal irritation. Other system examination were normal (Fig 1).

Blood investigation showed Hb-11.6 gm%, WBC count-10,200 cells/µl, ESR- 22mm/hr, RBS- 94 mg/dl, T3- 122 ng/dl , T4- 9.9 mcg/dl, TSH- 0.82 mIU/ml, HIV serology, ANA and ANCA were negative, CSF analysis was normal.

MRI Brain with MRA and MRV with contrast showed enlarged left cavernous sinus and T1 & T2 isointense lesion causing narrowing of left Internalcarotid artery extending anteriorly till the orbital apex. Post contrast image showed diffuse enhancement of the same lesion. Brain Parenchyma was normal, No aneurysm or vascular malformations, No cavernous sinus thrombosis, Dural venous sinuses were normal, Extraocular muscles appeared normal. Features were suggestive of Tolosa Hunt Syndrome (Fig 2).

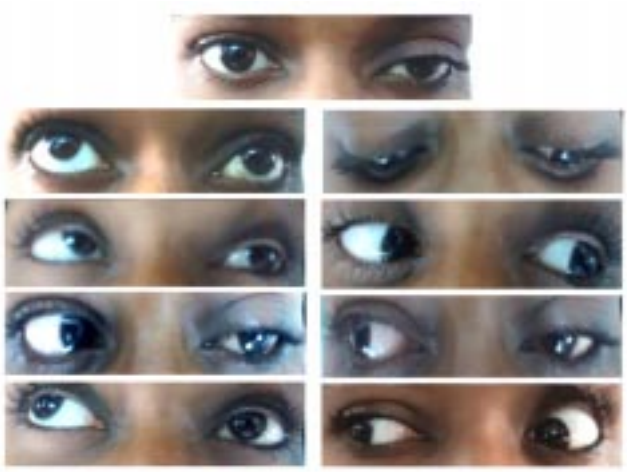

Fig 1 — Showing cranial nerve examination

Patient was treated with Tablet Predisolone 15mg thrice a day for 3 days and tapered slowly. There was significant improvement in ptosis and ocular movements after 2 days of corticosteroid therapy and recovered fully with out residual weakness at 7 day.

Discussion

Tolosa Hunt syndrome is caused by a non specific granulomatous inflammatory process in the cavernous sinus and / or superior orbital fissure[2,3]. The inflammatory process is characterized by non caseating, giant cell granuloma, fibroblast, lymphocyte and plasma cell proliferation within the cavernous sinus and / or superior orbital fissure[4]. The infiltration of the cavernous sinus with a non specific inflammatory tissue leads to compressive neuropathy of the cranial nerves third, fourth, sixth and first and second segments of the trigeminal nerve. The inflammatory process may also involve optic nerve[3].

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Department of General Medicine, Rajah Muthiah Medical College & Hospital, Annamalai University, Chidambaram 608002
[1]MD (Gen Med), Professor,
[2]MD (Gen Med), Postgraduate student and Corresponding author
[3]MD (Gen Med), Postgraduate student

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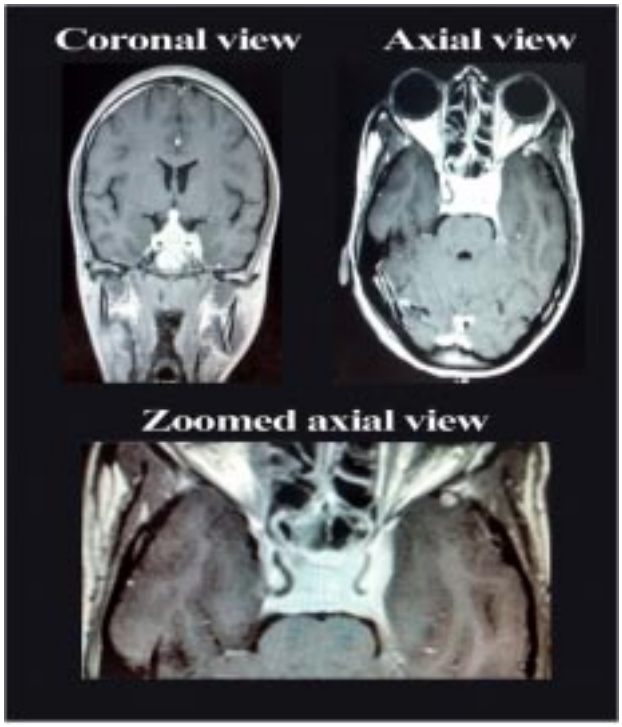

Fig 2 — MRI brain with MRA and MRV with contrast

Criteria for Tolosa Hunt Syndrome by International Headache Society 2013 :

(A) Unilateral headache fulfilling criterion C

(B) Both of the following :

(1) Granulomatous inflammation of the cavernous sinus, superiororbital fissure or orbit demonstrated by MRI or biopsy.

(2) Paresis of one or more of the ipsilateral third, fourth, sixth cranial nerves.

(C) Evidence of causation demonstrated by both of the following :

(1) Headache has preceded paresis of the third, fourth and/or sixth nerves by less than or equal to 2 weeks or developed with it.

(2) Headache is localized around the ipsilateral brow and eye.

(D) Not better accounted for by another ICHD-3 diagnosis[6].

Our patient fulfilled all the criteria for Tolosa Hunt Syndrome. The patient presented with unilateral headache with retro orbital pain and involvement of oculomotor, Trochlear and Abducent nerve. We excluded Neoplastic, Infectious, Vascular causes and thyroid dysfunction by appropriate investigations. Our patient improved with High dose corticosteroids within 2 days. Although the pathologic process and symptoms are self limiting, a short course of corticosteroids is helpful in prompt relief and to avoid long term complications.

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