

An Observational Study of Use of Azathioprine As Immunosuppressant For The Treatment Of Central Serous Retinopathy Associated With Tolosa Hunt Syndrome

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Abstract: Tolosa Hunt syndrome is a rare disorder characterized by painful ophthalmoplegia by nonspecific inflammation of cavernous sinus or superior orbital fissure. In this disorder there is unilateral retroorbital pain and palsy of cranial nerve three, four, six and ophthalmic and maxillary subdivisions of trigeminal nerve. The disorder is often recurrent but usually responding well with glucocorticoid administration. The condition if present with central serous retinopathy is far rarer. Treatment of Tolosa Hunt syndrome with high dose steroid can precipitate central serous retinopathy but coexistence of the two condition is very much rare and if present together, there is therapeutic challenge where to use oral steroid or not. As steroid is contraindicated in this patient with good response. This result signifies azathioprine can be used as alternative therapy in conditions of Tolosa Hunt syndrome where steroid is contraindicated.

Key Word: Azathioprine, Tolosa Hunt syndrome, Central serous retinopathy

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I. Introduction

Tolosa Hunt syndrome was first described by Tolosa in 1954 as recurrent painful ophthalmoplegia of unilateral eye with palsy of other cranial nerves like three, six and ophthalmic division of trigeminal nerve(1). Tolosa Hunt syndrome is low grade nonspecific inflammation of cavernous sinus or its walls(2). Pathologically, there is infiltration of lymphocytes and plasma cells. The unique feature which distinguishes it from other similar conditions like cavernous sinus thrombosis, craniopharyngioma, meningioma etc that it responds well with glucocorticoid administration and even there are chances of recurrence, there is complete recovery with steroid therapy(3). Tolosa Hunt syndrome has got many differentials like tuberculous meningitis, sarcoidosis, pituitary tumors, meningioma, lymphoma. Only striking difference with other condition is rapid response with steroid. It has been said that if the diagnosis is Tolosa Hunt syndrome, it should respond within 72 hours with oral steroid therapy(4).

In our patient, there were two pathology. Central serous retinopathy was probably due to hypertension of the patient. In our case, we were unable to use glucocorticoid as steroid is a precipitating factor for central serous retinopathy. Instead we used azathioprine and the patient responded. So it can be concluded that in presence of central serous retinopathy, we can treat Tolosa Hunt syndrome successfully with other immunosuppressants like azathioprine.

II. Materials And Methods

Our patient is a thirty eight years old hypertensive diagnosed as Tolosa Hunt syndrome four years back and on irregular medication. She presented with chief complaint of left sided retro-orbital pain and left hemicranial headache for two months. She also complained of seeing double images for the same duration which was associated with impairment in her daily living. These events were of insidious in onset and gradually

progressive. She also developed inability to move her left eye in all directions for eight days. In this period she also developed drooping of her left upper eyelid.

When we enquired she also revealed that she has difficulty in distant vision for this two months and the diminution of vision was more on left side. She also stated that she was feeling decreased sensation in her left forehead.

There was no associated fever or other systemic manifestations like prostration. There was no associated redness or bulging of left eye. There was no history of weaknesses of any part of the body.

We asked her about her past illnesses. But she could not remember any major illnesses including any infections like cellulites or boils in the region of dangerous area of face.

The patient was a middle aged woman with one son sixteen years old delivered normally and having her menstruation normally. She had no addiction.

On examination we found no significant abnormality in general examination except hypertension. She was of average built and nutrition

On neurological examination, the patient's higher functions were normal. There were no weakness of any limb. There was ptosis of left eye. There was no movement at all in left eyeball. Movements of right eye was within normal limit. Pupil in the left side was dilated and not reacting to light but right sided pupil was normal in shape and reacting to light normally. Consensual light reflex in left eye was also absent but present in right eye. There was sensory loss in the distribution of V1 division of trigeminal nerve. Visual acuity of both eye were reduced.

We had sent basic blood investigations like complete blood count, urea and creatinine which came out to be normal. CT scan of both orbits was within normal limit (Figure 1). MRI of brain with special attention to cavernous sinus revealed contrast enhancement on left sided cavernous sinus (Figure 2). We consulted with department of Radiology. According to them, the enhancement was most probably due to any inflammatory lesion. Their opinion was more in favor of any granulomatous lesion. So in this way we reached the diagnosis to be Tolosa Hunt syndrome. Fundoscopy and ocular coherence tomography revealed that she had acute central serous retinopathy of left eye and resolving central serous retinopathy of right eye.



Figure 1--CT scan of both orbits was within normal limit

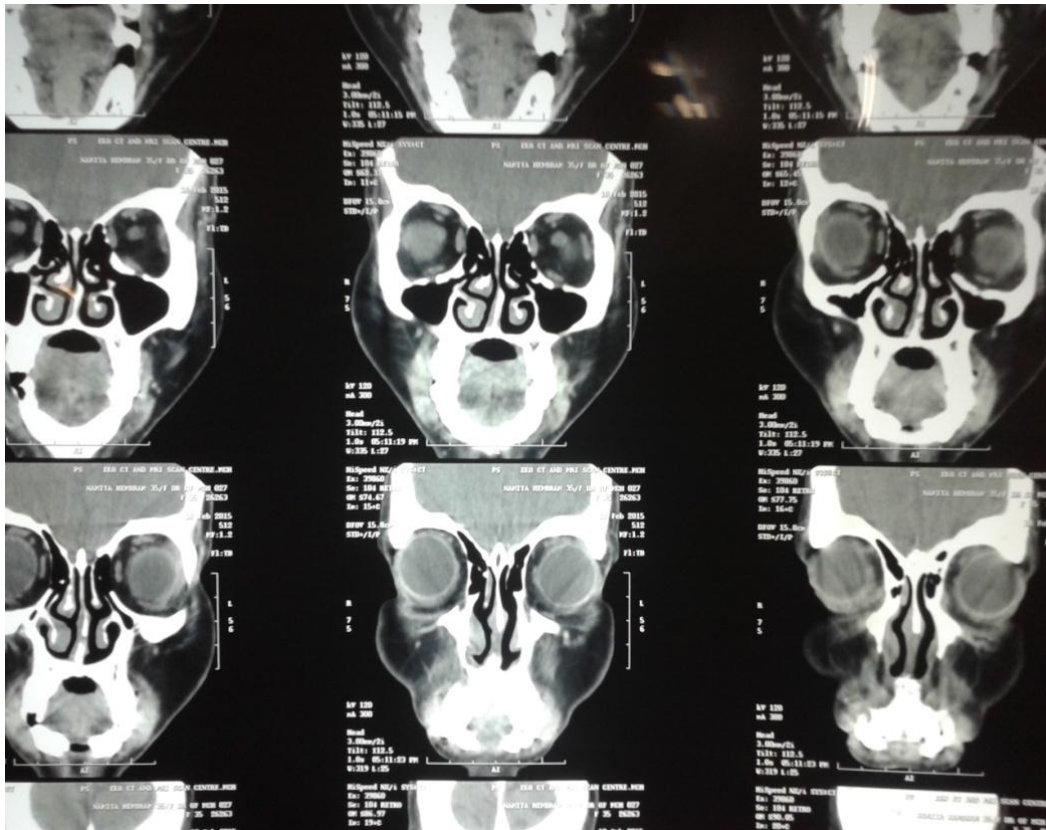


Figure2- MRI of brain showing contrast enhancement on left sided cavernous sinus.

We treated the patient with oral azathioprine 50mg two times daily. The patient showed improvement in movement of left eye five days after starting of therapy. Light reflex on left sided pupil returned. Size of the left pupil also was returning to near normal in comparison to right pupil. Ptosis of the left eye ball improved.

III. Discussion

Tolosa-Hunt syndrome usually presents with unilateral headache with severe painful ophthalmoplegia and sudden in onset (5). There are ups and downs of the symptoms though it usually resolves spontaneously. Granulomatous inflammation of cavernous sinus, unilateral in nature is the etiology (6). Diagnosis is basically clinical and by exclusion though magnetic resonance imaging or computed tomography is used to detect inflammatory changes in the cavernous sinus or orbital fissure. Other investigations like biopsy of the affected tissue can be done though difficult to perform. Oral corticosteroid in high doses is very effective to relieve the symptoms(7). Acute and worsening painful vision loss is a cause for concern in the emergency department of ophthalmology and it is very essential to identify the precipitating cause for further intervention in a timely manner.

In the other hand Central serous retinopathy (CSR), also known as central serous chorioretinopathy (CSC or CSCR), is a disorder of eye presents with sudden loss of vision which is temporary in nature (8). Persons with Central serous retinopathy have higher levels of corticosteroid level(9). In contrary to Tolosa-Hunt syndrome, corticosteroid is the precipitating cause in this situation (10). In our patient Tolosa-Hunt Syndrome was associated with central serous retinopathy. Steroid is indicated for one which may lead causation to the other. For that reason azathioprine, an immunosuppressant was issued with good result.

IV. Conclusion

Tolosa-Hunt syndrome is a rare, reversible and painful ophthalmoplegia .Central serous retinopathy is associated with this syndrome. As steroids cannot be used in this situation, azathioprine is used with good result instead of steroid. This observation may show a good revelation as azathioprine has less side effects in many situations Further exploration is needed in this new modality of treatment.

CONFLICT OF INTEREST: No Such

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