



Case Report

Azathioprine in orbital apex syndrome

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ABSTRACT

Orbital Apex Syndrome (OAS) is commonly caused by specific inflammations, infections, neoplasms, iatrogenic, trauma, vascular and non-specific orbital inflammation (NSOI). OAS secondary to NSOI responds rapidly to corticosteroids. There are very few case reports on azathioprine as additional treatment modality in OAS secondary to NSOI. We are presenting a 42 years old female who had recurrent episodes of painful ophthalmoplegia, treated with steroid therapy. Patient was steroid dependant. Therefore, she was started on azathioprine 2-3mg/kg body weight for 36 weeks. She had complete remission of the painful ophthalmoplegia, with no further recurrence on 6 months follow-up.

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1. Introduction

Orbital apex syndrome (OAS) is described as a syndrome involving damage to oculomotor, trochlear, abducens and ophthalmic branch of the trigeminal nerves (V1) in association with optic nerve dysfunction. Based on anatomical location, two other syndromes that can have overlapping features are the superior orbital fissure syndrome/ Rochon-Duvigneaud syndrome and Tolosa Hunt Syndrome (THS).

The common causes of OAS are specific inflammations, infections, neoplasms, iatrogenic, trauma, vascular and non-specific orbital inflammation (NSOI).

We hereby present azathioprine as a treatment option in OAS secondary to NSOI.

2. Case Report

A 42 years old female presented with complaints of binocular diplopia of 2 days duration, with no history of any systemic illness. Examination revealed 15^o left exotropia and hypertropia with head tilt towards right side. Best corrected visual acuity (BCVA) was 6/6 in both the eyes and colour vision was normal.

Over a period of next 3 days, visual acuity reduced to 6/9 with ptosis in the left eye and painful limitation of all extraocular movements (EOM), corneal sensations were normal. There was no relative afferent pupillary defect (RAPD) and fundus was normal in both eyes. Patient was diagnosed with left side OAS secondary to NSOI. MRI was within normal limits with clear para nasal sinuses and normal orbital apex (Figures 1 and 2). Patient was given intravenous dexamethasone 8mg and 60 mg/day of oral prednisolone for a week. She got relieved of pain immediately after the steroid infusion.

Over the next 2 days, in spite of oral steroids, visual acuity reduced to 6/12p with limitation of all EOM associated with pain, there was increase in ptosis and no RAPD with normal fundus examination. Patient was started on intravenous methylprednisolone 500mg for 3 consecutive days with good response to the medication. She developed mild pain 2 weeks later. Intravenous methylprednisolone 500mg was repeated again along with oral prednisolone at 1mg/kg tapered over 9 weeks.

Patient reported pain in the left eye as the dosage of oral steroids reached 20 mg per day. She was started on oral azathioprine 50 mg twice a day along with 30mg oral prednisolone tapered over 3 weeks. Oral azathioprine was increased to 50mg three times a day with monthly complete

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Fig. 1:



Fig. 2:

blood counts, liver and renal function tests and serum C-reactive protein assay. Patient showed good response to medication with no diplopia or pain.

Oral Azathioprine was continued at a dose of 50mg thrice daily for a period of 5 weeks. When the patient was found to be tolerant to Azathioprine she was maintained for the next 31 weeks and then subsequently stopped. Patient had slight nausea and vomiting sensation on medication which was tolerable. On 6 months follow up, patient is asymptomatic with no pain or diplopia and maintains visual acuity of 6/6 in the left eye.

3. Discussion

NSOI is a benign non-infectious, inflammatory process of the orbit, characterised by a polymorphous lymphoid infiltrate with varying degrees of fibrosis, without a known local or systemic cause.

The etiopathogenesis of NSOI is unknown. Suggested theory for NSOI are immune mediated or molecular mimicry when a foreign antigen has structural similarities with self-antigen and it may happen after an acute infection.

Treatment options for OAS secondary to NSOI include corticosteroids and immunosuppressive therapy. Response to corticosteroids is rapid with dramatic improvement of all symptoms and signs.

The treatment of THS and inflammatory OAS is often long with frequent relapses and remissions. Patients tend to have intolerable side effects due to the long-term steroid therapy and may benefit with immunosuppressive agents.

In some cases of THS, immunosuppressants like methotrexate¹ and azathioprine²⁻⁴ and radiation⁵ have been used.

Smith JR, Rosenbaum JT¹ have used methotrexate as treatment modality in a case of THS, it was used at a dose of 25mg per week for a period of 47 months. The patient was asymptomatic for 19 months, she then experienced recurrent inflammation which required reinstitution of the drug with a tapering course of prednisone.

Mazzotta G et al³ used azathioprine, 2mg/kg of body weight in a patient with recurrent THS. She showed complete remission of the painful ophthalmoplegia within a few days. Azathioprine was continued for another 6 months, with no further recurrence of the ophthalmoplegia.

Bhattacharya R et al⁴ reported a case of THS with central serous retinopathy treated with azathioprine as first modality of treatment. The patient was given azathioprine 50mg BD. The patient showed improvement in the condition in 5 days.

Jan Hannerz² studied azathioprine as treatment modality in 4 patients with THS. They found satisfactory results.

Our case was comparable to the one reported by Mazzotta G et al.³ They reported THS whereas our case had clinical features of orbital apex syndrome with normal MRI findings. We had used 1.5 T MRI which might have missed the early features of cavernous sinus involvement.

The dose of azathioprine used in our case was comparable to Mazzotta G et al.³ Our patient showed complete remission after 3 weeks. Azathioprine was stopped after 9 months in our case. We didn't find any recurrence which was comparable to Mazzotta et al.³

4. Conclusion

There are multiple causes of OAS. Thorough history, examination and imaging is required for etiological diagnosis. NSOI responds well to steroids therapy, azathioprine can be considered as an option of treatment in refractory cases and patients who are intolerant or dependant to steroid therapy.

5. Source of Funding

None.

6. Conflict of Interest

None.

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