



Orbital Apex Syndrome: A Rare Case Report

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Abstract

Orbital apex syndrome also known as Jacod syndrome is a rare disorder. This could be inflammatory, infectious, neoplastic, vascular or traumatic in origin. We present a 40-year-old woman with drooping of right eye eyelid since 15 days associated with headache. Patient was investigated and managed conservatively with systemic steroids. The patient was followed-up regularly in OPD after discharge. Mainly the treatment is by addressing the underlying cause and requires multidisciplinary approach.

Keywords: *Orbital apex syndrome, Jacod syndrome.*

Introduction

Orbital apex syndrome (OAS) involves cranial neuropathies in association with optic nerve dysfunction. Orbital apex syndrome is symptomatically related to superior orbital fissure syndrome and cavernous sinus syndrome with similar etiologies. The distinction is the precise anatomic involvement of the disease process.¹ Orbital apex syndrome is a syndrome characterised by dysfunction of oculomotor nerve (CN III), trochlear nerve (CN IV), ophthalmic branch of the trigeminal nerve, (CN V1), abducens nerve (CN VI) along with optic nerve damage. This could be inflammatory, infectious, neoplastic, vascular or traumatic in origin. It was Kjoer, in 1945, who firstly created the term orbital apex syndrome (OAS).² We present a patient of the OAS in this case report

Case Report

A 40-year-old woman presented with complaint of drooping of RE eyelid associated with headache since 15 days patient was a known case of hypertension since 2 years. Ocular examination of right eye revealed visual acuity - 6/12 with 15 degree exotropia, complete ptosis, restricted ocular movements in all direction. Left eye – Visual acuity -6/9. Anterior segment was normal. Fundus Examination both eyes was within normal limits. No abnormality found in Blood investigations . MRI brain with orbit showed mild edema in the region of the orbital apex suggestive of Inflammatory pathology. Patient was admitted and started on intravenous Methyl Prednisolone 1g x 3 days Followed by oral prednisolone started at 50mg/day. Patient was discharged on 7th day. Patient had full recovery on follow up visits.



Fig. 1 At the time of Presentation showing RE eyelid Ptosis

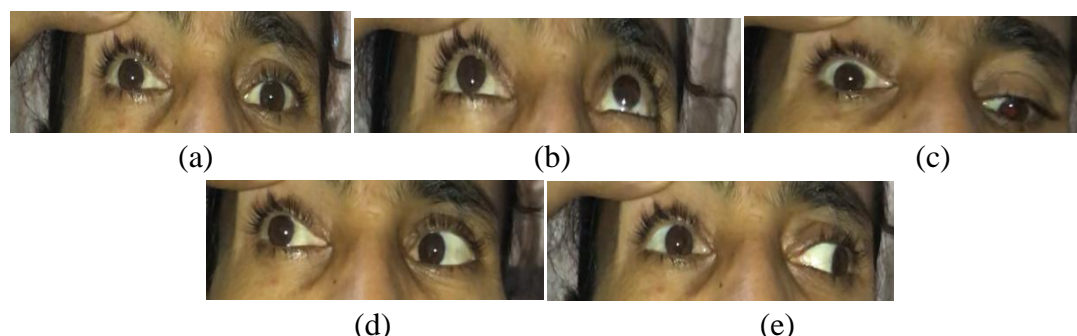


Fig. 2. The ocular movements revealed right complete ophthalmoplegia. (a) resting (b) upward gaze (c) downward gaze (d)look to right (e) look to left.

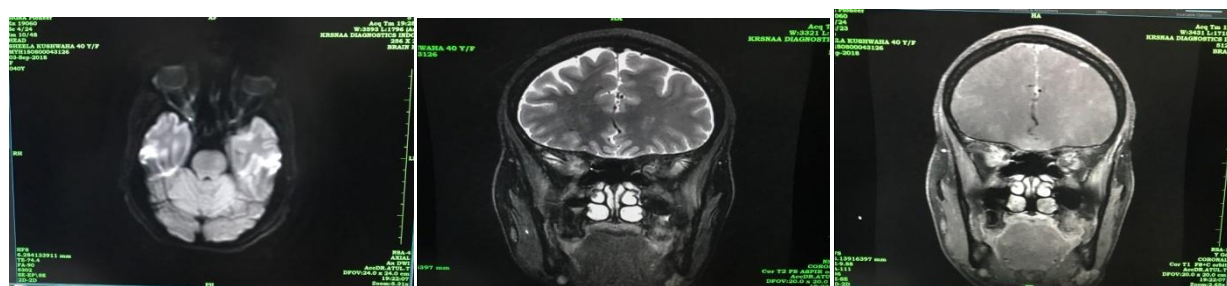


Fig. 3 MRI Brain with orbit showed focal edema s/o inflammatory etiology.



Fig. 4 After Treatment with Steroids

Discussion

The OAS is characterized by visual impairment, ophthalmoplegia, ptosis, proptosis, dilated pupil and light reflex abnormality as well as hypoesthesia of forehead. The ophthalmoplegia is secondary to CN III, CN IV or CN VI dysfunction; ptosis occurs due to weakness of both the superior tarsal muscle, which is innervated by nasociliary nerve, and the levator palpebral muscle, innervated by CN III. Exophthalmos may be attributed to the decreased tension of the extraocular muscles. The forehead and eyelid region anesthesia comes from CN VI damage. Moreover, the dilated pupil is often found related to the parasympathetic fibers

dysfunction in CN III. Disturbance of vision was attributed to optic neuropathy from CN II. All of above symptoms were termed OAS. The symptoms should be differentiated between nerve compression by either superior orbital fissure, orbital apex, or cavernous sinus. OAS may be caused by inflammatory, infectious, neoplastic, iatrogenic/traumatic, or vascular processes.¹ The management of OAS remains uncertain. Many inflammatory etiologies require treatment of the primary process and reduction of inflammation using systemic immunomodulatory agents which include corticosteroids and steroid-sparing agents.³ Decompressive surgery to provide an anatomic expansion of the orbit and radiation

therapy to alleviate inflammation causing compression of optic nerve in thyroid orbitopathy can be pursued.⁴ In infectious etiology treatment usually entails administration of the appropriate broad-spectrum anti-microbial therapy. Surgical intervention via a direct or endoscopic approach may be necessary in cases of orbital abscess or subperiosteal abscess.⁴ In Orbital apex syndrome with no known local or systemic cause, Idiopathic orbital inflammation should be suspected as a diagnosis of exclusion⁵. Steroid therapy classically results in rapid improvement.³

Conclusion

Orbital apex syndrome is an important differential diagnosis for any patient presenting with multiple cranial nerve palsy. Though OAS is not commonly seen, it can be properly diagnosed by careful physical and ophthalmologic examinations, together with radiological findings. Detailed history, a complete neuro ophthalmological examination with systemic review helps in narrowing the differentials. Prompt neuroimaging is mandatory. Treatment is by addressing the underlying cause and requires multidisciplinary approach.

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