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A Case of Sinonasal Carcinoma presenting as Orbital Apex Syndrome

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Abstract : Neoplasia of the orbital apex though rare form a very important differential diagnosis in any patient presenting with multiple cranial nerve palsy Here we present a case of sinonasal carcinoma with orbital apex syndrome ,as the initial presenting feature. Mutidisciplinary approach is required to treat these patients and the prognosis is generally poor.
Keyword : Orbital apex tumors , Sinonasal carcinoma

CASE REPORT:

A 60 year old lady presented to us with complaints of Right eye defective vision , fever with rhinitis and nasal blockade for two months . On examination best corrected visual acuity in her right eye was perception of light with projection of rays accurate and left eye was 6/9. Her right eye showed eccentric proptosis with a relative afferent pupillary defect and complete ophthalmoplegia (fig 1) Corneal sensations and other cranial nerves were intact . Fundus showed temporal pallor . Anterior and posterior segment examination of the left eye was within normal limits. Anterior rhinoscopy revealed a friable fleshy mass in her right nasal cavity(fig 2).There were no palpable lymph nodes in the head and neck region. Since the patient presented with multiple cranial nerve palsy involving her right second, third, fourth and sixth nerves , immediate neuroimaging was warranted .CT Brain and orbits showed a ill defined soft tissue lesion with hyperdense areas involving bilateral maxillary , sphenoid , ethmoid, right frontal sinus extending to the right orbital apex and cavernous sinus region and causing extensive destruction of medial orbital wall and displacement of intraorbital structure (fig 3 & 4) . Patient was referred to an ENT surgeon and endoscopic biopsy of the mass in the nasal cavity showed sinonasal carcinoma of neuroendocrine differentiation. A complete metastatic workup was then done and CT chest was normal (fig 5) and USG abdomen revealed liver metastasis . An oncologist consultation was taken and patient was put on four cycles of chemotherapy (etoposide + cisplatin) followed by radiotherapy On her last visit in October 2013 her condition has remained status quo.

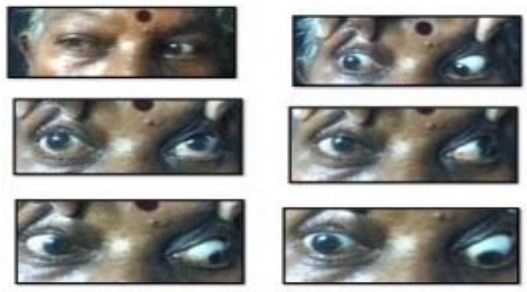
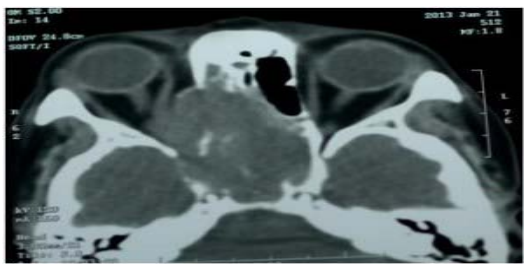


Fig 1- Showing complete ophthalmoplegia of right eye.

Fig 2- Showing friable fleshy mass in right nasal cavity



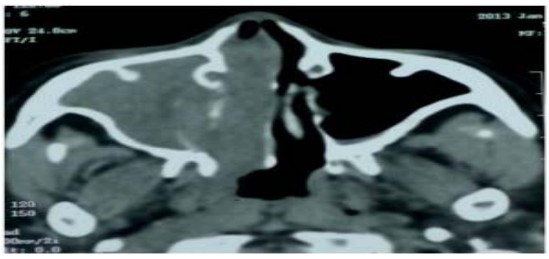


Fig 3 & 4 -CT BRAIN & ORBIT showing ill defined soft tissue lesion with hyperdense areas involving bilateral maxillary , sphenoid, ethmoid , right frontal sinus extending to the right orbital apex and cavernous sinus region into th eorbita after causing extensive medial orbital wall destruction.

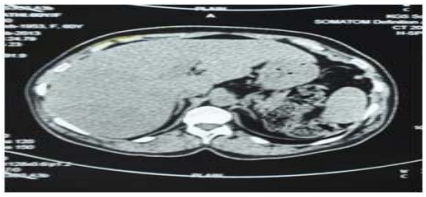


Fig 5- CT chest of the same patient being normal .

DISCUSSION :

Orbital apex syndrome involves damage to the occulomotor , trochlear , abducens nerves,and ophthalmic branch of the trigeminal nerve (V1) in association with optic nerve dysfunction. The cavernous sinus syndrome includes features of orbital apex syndrome with added involvement of the maxillary branch of the trigeminal nerve (V2) and oculosympathetic fibers .Superior orbital fissure syndrome involves lesions , immediately anterior to the orbital apex, external to the annulus of zinn, where the optic nerve is spared .1 The etiologies of these syndromes can be varied ranging from inflammatiory , infectious , neoplastic ,traumatic/iatrogenic to vascular.1 Neoplasia involving the orbital apex should always be kept as a differential in a patient presenting with multiple cranial nerve palsies .In a study conducted by Keane 2et al and Ananthkumar 3et al neoplasia was found to be the commonest cause for orbital apex syndrome . Primary ocular or orbital tumors , CNS tumors and those of the PNS may invade the apex . Neural tumors of the apex are generally benign tumors like meningiomas , schwannomas.1 They have a prolonged course and may only involve a single cranial nerve for a long period of time .2 Most common neoplasia to cause a cavernous sinus syndrome include nasopharyngeal carcinoma, pituitary adenoma ,lymphoma, meningioma and metastasis .2 Godfred et al described a series of 240 malignant nasopharyngeal tumors with neuro ophthalmic sym toms of which 20 % developed cavernous sinus syndrome.2 These tumors tend to involve the abducens and mandibular division of the fifth , while a lateral extension of a pituitary adenoma will involve the occulomotor and ophthalmic division of fifth. In a study conducted by mayo clinic2 metastatic disease was mainly responsible for the syndrome and it includes metastasis from breast, lung, kidney and malignant melanoma.

Orbital apex syndrome has been reported to be the presenting feature of various PNS tumors such as adenoid cystic carcinoma 4 , mucoepidermoid carcinoma of the maxillary sinus 5 ,squamous cell carcinoma of the maxillary sinus .6 Carcinomas of the maxillary sinus are uncommon . They comprise of 0.2-0.5% of all systemic malignancies, 3% of all cases of head and neck carcinoma, and 80% of all cases of paranasal sinus carcinomas.6 Johnson7 et al reviewed 79 patients with sinus and paranasal tumours and found that 47 patients (59%) demonstrated orbital involvement. The maxillary sinus was the most common site of origin of the paranasal tumours and the sphenoid sinus the least common. Squamous cell carcinoma was the most common paranasal tumour to invade the orbit. In their series the most common ophthalmic presenting sign was proptosis and none presented with orbital apex syndrome. Srinivasan et al reported a case of squamous cell carcinoma of the maxillary sinus presenting as orbital apex syndrome . The patient did not have any ENT symptoms prior to onset and the tumor had grown to inoperable dimensions in a short time period before presentation to the ophthalmologist. In our case the patient did have ENT symptoms but presented to the ophthalmologist first with complaints of defective vision and prominence of eyes and the tumor had grown to inoperable dimensions within a short span of time. The neuroimaging suggested a possibility of invasive sinusitis and it was biopsy which revealed the diagnosis. Like most malignant tumors involving the orbital apex , the prognosis was poor in our patient as well.

CONCLUSION : When any patient presents with multiple cranial nerve palsy involving II,III,IV, VI ,V1,V2 in various permutations and combinations, a lesion in the Orbital apex/ cavernous sinus should be ruled out . A detailed history, complete neuro ophthalmological evaluation and systemic review is important in narrowing the differentials . Prompt neuroimaging is mandatory . Neoplasia involving the orbital apex though rare , form an important differential diagnosis. A complete metastatic workup needs to be done , once the diagnosis of malignancy has been clinched . Multidisciplinary approach is required for appropriate management .

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