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An unusual case of third cranial nerve palsy

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Abstract : 62 year old man presented with complaints of drooping of right upper eyelid for 15 days. It was associated with headache , eye pain , diplopia on retracting the eyelid. No history of any systemic diseases or trauma. On ocular examination , best corrected visual acuity in both eyes was 6 on 6 with N6 near vision and normal intraocular pressure . The anterior segment examination of right eye showed complete ptosis , mid dilated pupil with gross restriction of extraocular movements except for abduction.Corneal sensation was slightly reduced in right eye. Anterior segment examination of left eye was normal.Investigations revealed right third cranial nerve palsy . Considering the clinical red flag signs , MRI Brain was done which revealed extra axial lesion involving infra temporal and pterygo palatine fossa and spreading perineurally throught 5th cranial nerve into right cavernous sinus affecting 3rd cranial nerve. A radiological diagnosis of Adenoid cystic carcinoma of minor salivary glands was made.

Keyword : Ptosis, Diplopia , Cavernous sinus An unusual case of third cranial nerve palsy

INTRODUCTION

Third cranial nerve (oculomotor) palsies are quite commonly encountered in neuro-ophthalmology as well as in general ophthalmic practice. Though many adult cases have an ischemic origin due to underlying systemic diseases like diabetes mellitus and hypertension and have a relatively benign and mostly self-limiting course, it is important to find clinical “red flag” signs which may indicate a more sinister etiology and warrant neuroimaging and further investigations. The following case report is of such a case with an unusual etiology.

CASE REPORT

A 62 year old gentleman presented to us with complaints of Drooping of Right upper eyelid since 15 days , associated with headache and eye-pain along with double vision on retracting the right upper eyelid. The eyelid drooping was sudden in onset, complete in extent, stationary during the course, and with no diurnal variation. It was associated with dull-aching pain, behind the right eye, associated with headache. Double vision was appreciated by the patient only On retracting the closed lids, with a horizontal separation of images, more in some gazes than others. The patient was non-diabetic and non-hypertensive, with no other systemic illnesses and no history of any similar episode in the past. There was no history of any head trauma or history suggestive of cerebrovascular disease or other cranial nerve palsies. The patient’s personal history revealed that he was a beedi-smoker for the past 25 years, and consumes alcohol occasionally.

Fig 1. a) showing complete right eye ptosis b) 30 degree exotropia of right eye on fixing light with unaffected left eye and c) more than 45 degree exotropia of left eye on fixing light with affected right eye. (fig b. and c. show that secondary deviation is more than primary deviation)

On general examination the patient was conscious, co-operative, well-oriented to time, place and person, moderately built and nourished and with no clinical evidence of pallor, Icterus, Cyanosis, Clubbing, Lymphadenopathy or Pedal edema. His vital signs were stable and all systems were within normal limits clinically. On Ocular examination, best corrected vision was 6/6(20/20) and N6 near vision and intraocular pressure was normal in both eyes. The anterior segment was normal in left eye with lens changes whereas right eye showed complete ptosis, pupil was mid-dilated and not reacting to both direct and consensual light. Extraocular movements showed gross restriction of movements in all gazes except abduction in right eye and were normal in left eye. The corneal sensation in right eye was slightly reduced. Fundus examination in both eyes was normal. Cranial nerve examination was normal in both eyes except for right third nerve and probable right fifth nerve involvement (decreased corneal sensation). Hess Charting and Diplopia Charting were suggestive of third cranial nerve palsy in right eye. Routine blood investigations were within normal limits.

The patient had already undergone a CT scan brain before coming to us, which was reported as normal study. Considering the clinical red flag signs of painful palsy, pupillary involvement and probable trigeminal nerve involvement an MRI imaging with and without contrast was adviced for the patient. The MRI Brain (Fig 2 A to E), showed a heterogenously enhancing extra-axial lesion, involving Infra-temporal and pterygo palatine fossa, parapharyngeal and masticator spaces, with compression of masticator muscles and spreading perineurally through mandibular division of 5th cranial nerve (V3) into right cavernous sinus region and paracavernous area causing tethering of nerves in precavernous and cavernous area (the probable cause of 3rd cranial nerve palsy). A Radiological diagnosis of probable Adenoid Cystic carcinoma of minor salivary glands, spreading perineurally around mandibular nerve (V3) reaching cavernous sinus was given. The patient was referred to a Neurosurgeon but was lost to follow-up with us.

DISCUSSION

The salivary glands include Parotid, Submandibular, sub-lingual and minor salivary glands. Minor salivary glands are sub-mucosal and are found distributed over the lips, palate, buccal mucosa, tongue, cheeks, retro-molar area, floor of the mouth and upper aero-digestive tract and contribute to 10% of total salivary volume. Tumours of the salivary glands contributes to 3% of all body tumours, majority occurring in parotid gland (85%), only a minority (4 – 5 %) occurring in the minor salivary glands. But, minor salivary gland tumours gain importance because of the fact that 80% of these tumours are malignant. Adenoid Cystic Carcinoma is the second-most common malignant tumor of the salivary glands and is the most common malignant tumor in all salivary glands except parotid gland. Its common characteristics include widespread local infiltration, perineural spread, a propensity for local recurrence and late distant metastases. Histologically, its three subtypes include Tubular, Cribriform and Solid, with the last having worst prognosis and more tendency for perineural invasion.

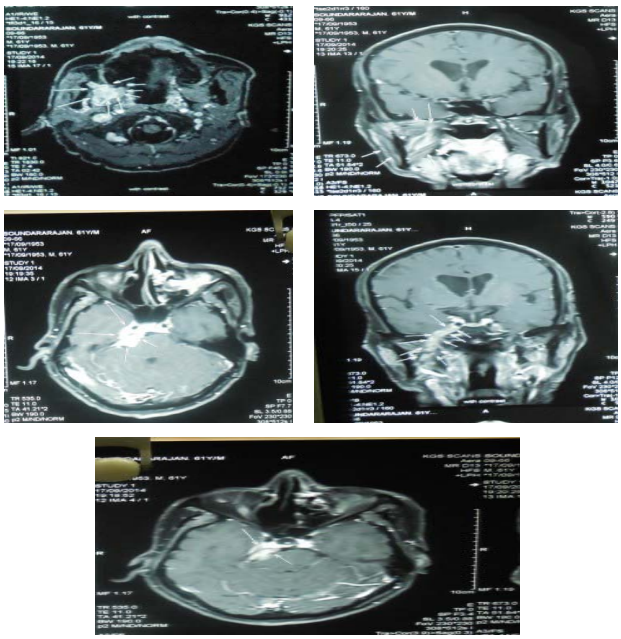

Fig 2.MRI Brain.
A) Axial image-cisterna magna level, arrows showing right pharapharyngeal space tumour
B) Coronal image showing masticator muscles involvement
C) Axial T2 image, inner two arrows showing compression and displacement of Pons
D) T1 weighted with contrast coronary section image showing perineural spread along mandibular division of 5 th cranial nerve
E) Axial image-cavernous sinus involvement

Clinically, ages above 50 years are affected. It may present as a asymptomatic, firm, domeshaped, swelling on lateral to midline of the palate. Evidence of perineural spread may be present before the primary tumour is detected. Literature shows involvement of cavernous sinus occurring in 15 % of patients either by contiguous or perineural spread. Literature is consistent that the time between onset of neurological signs and symptoms, and the time of diagnosis range between few months to 3 years. The treatment of choice consists of total tumor resection-which may involve large scale tissue removal like medial maxillectomy or subtotal maxillectomy, cranio-facial resection etc. depending on site. A combined modality of radical surgery followed by postoperative radiation has been suggested to be the most effective. Chemotherapy is reserved as a palliation therapy for incurable tumours.

CONCLUSION

Third nerve palsy is a common clinical entity, and many of its causes may be benign and selflimiting, but a good clinical acumen regarding important so-called “red-flag” signs is very important to pick out the cases which require further investigations for more sinister etiologies. A normal study in the CT scan Brain for our case also highlights the inadequacy of CT scans as screening tools in soft tissue lesions and the requirement of MRI in these cases. A wide window period between the onset of neurological signs and symptoms and establishing diagnosis found consistently in literature shows the need for high degree of clinical suspicion, which may help in early diagnosis at a potentially curable stage which can be life-saving in these patients.

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