Isolated Sixth Nerve Palsy in Pseudo Gradenigo's Syndrome

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ABSTRACT
Nasopharyngeal carcinoma is a relatively uncommon malignancy in South India. Exact incidence in Indian population is not known. It usually presents with either epistaxis, tumors in the neck, or cranial nerve(s) involvement. This is an interesting case because isolated sixth nerve involvement on presentation is rare.

KEYWORDS:
Nasopharyngeal carcinoma, Isolated sixth nerve palsy, Clinical symptoms, Neuro-ophtalmologic presentation

Case Report:
A 71 yrs old man presented in our hospital with a swelling in the medial canthus of the right eye(RE) past 3 months in duration. The swelling was slowly and progressively increasing in size[Figure 1(i)]. It developed induration and ulceration past 1 month. He also complained of right sided periorbital pain. He suffered from serious discharge from his right ear few months back. There was no history of weakness of any part of body, ataxia, or loss of sensation over body. Bladder and bowel habits were normal. There was no history of fever, epistaxis, swellings in neck, haemoptysis, or weight loss. On examination, patient was conscious, and vitals were stable. Examination of chest, cardiovascular system, abdomen and central nervous system was normal. There were no other systemic or neurologic symptoms. There was no history of smoking or drug/alcohol abuse. His best corrected visual acuity in RE was 20/70 and in left eye(LE) was 20/30. Ocular motility test showed RE abduction deficit[Figure 1(ii)] while movements were preserved on the left side. Thus diplopia was present in right lateral gaze. Pupils were bilaterally equal and normally reacting to light. Clinically it was chronic otitis media presenting with Gradenigo's syndrome. The patient was further sent for radiological investigations to confirm the diagnosis.

Computed tomography(CT)[Figure 2] findings were suggestive of ill defined soft tissue mass(6.9 x 6.8 x 5.6)cm in size in the right nasopharynx with extensions up to the angle of mandible of right side, above up to the base of skull, right cavernous sinus and bilateral sphenoid sinuses. There was lytic destruction with bone rarefaction involving clivus, squamous part of right temporal bone, walls of both sphenoid sinuses and soft tissue extension into both sinuses, right middle ear cavity, right temporo-mandibular joint. The lesion crosses the midline to the left side causing lytic destruction of condylar process in the base of skull with possible mass effect at cervico-medullary junction and right temporo-parietal region. CT with contrast was done to confirm these findings. Therefore, the final diagnosis was nasopharyngeal carcinoma leading to 6th nerve palsy of right side. The triad of Gradenigo's syndrome when presents with nasopharyngeal carcinoma is called Pseudo Gradenigo's syndrome. Audiometry test showed sensorineural hearing loss of both ears.

He was treated with combination chemotherapy with 5-fluorouracil and cisplatinum along with external beam radiotherapy. The patient was given one cycle of chemotherapy and radiotherapy, and is yet to report back for follow-up.

Discussion:
In 1904 Guiseppe Gradenigo described an infection of the apex of the petrous part of the temporal bone from acute otitis media with the clinical symptoms of unilateral pain around the eye, diplopia due to sixth nerve paralysis and persistant otorrhea. Contact with the tip of the petrous pyramid makes the portion of the sixth nerve within Dorelli's canal susceptible to pathologic processes. This is especially important in cases secondary to localized inflammations or extra-dural abscesses due to complicated otitis media. Nasopharyngeal carcinoma may also mimic Gradenigo's syndrome, since it can obstruct the Eustachian tube, resulting in serious otitis media. Additionally, the carcinoma may subsequently invade the cavernous sinus, causing sixth nerve paresis. Therefore, its called as Pseudo-Gradenigo syndrome.

Nasopharyngeal carcinoma is the most dreaded etiology of sixth nerve palsy and most frequently occurs between the ages of 40 and 70 years. It may present with various symptoms including nasal obstruction, rhinorrhea, epistaxis and serous otitis media secondary to obstruction of the Eustachian tube. This tumour may also involve the cranial nerves in many ways. Firstly, the newly may extend superiorly through foramen lacerum, which is an unpaired pathway near the fossa of Rosenmuller into the cranium[4,5], thus involving the cranial nerves in the middle cranial fossa and cavernous sinus. In our patient, the extension of the tumour into cavernous sinus lead to sixth nerve palsy without involvement of other cranial nerves. Sixth nerve is the second most common nerve to be involved in nasopharyngeal carcinoma. Isolated sixth nerve palsy, a rare neuro-ophtalmologic presentation was noted in two cases of nasopharyngeal carcinoma.

References: