

Ophthalmoplegia associated with head and neck malignancies: a case series

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Abstract

Ophthalmoplegia is the paralysis or weakness of the eye muscles. It can affect one or more of the six muscles that hold the eye in place and control its movement. Total ophthalmoplegia refers to the paralysis of all the muscles in the eye. People affected by ophthalmoplegia with orbital apex involvement may have double or blurred vision. They may also experience an inability to position the eyes in sync. Some may also have a hard time moving both eyes in every direction, and many will have drooping of their eyelids. Causes of ophthalmoplegia along with orbital apex involvement can be multiple sclerosis, trauma infarction, brain injury, stroke, thyroid disease, infection, associated with sinonasal malignancy, nasopharyngeal carcinoma, invasive fungal infections needing surgical removal and brain tumours.

Keywords: Ophthalmoplegia, nasopharyngeal carcinoma, Maxillary carcinoma, Olfactory neuroblastoma

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Introduction

Ophthalmoplegia is the paralysis or weakness of the eye muscles[1]. It can affect one or more of the six muscles that hold the eye in place and control its movement. Total ophthalmoplegia refers to the paralysis of all the muscles in the eye, which in turn results in ptosis[2], immobility of the eye, dilated non reacting pupil and loss of accommodation. People affected by ophthalmoplegia with orbital apex involvement may have double or blurred vision[3]. They may also experience an inability to position the eyes in sync. Some may also have a hard time moving both eyes in every direction, and many

will have drooping of their eyelids. Causes of ophthalmoplegia along with orbital apex involvement can be multiple sclerosis, trauma infarction, brain injury, stroke, thyroid disease, infection[4-6]. Among head and neck causes ophthalmoplegia can be associated with sinonasal malignancy, nasopharyngeal carcinoma [7] invasive fungal infections needing surgical removal and brain tumours[8]. Nasopharyngeal carcinoma can manifest with ophthalmoplegia and neurologic features like Horner's Syndrome. Squamous cell carcinoma of nasopharynx is the commonest variety[9] can spread anteriorly through nasal cavity to Pterygopalatine fossa and involve orbital apex. Also by lamina papyraea nasopharyngeal carcinoma can involve the orbit and extra ocular muscles causing ophthalmoplegia. Maxillary carcinoma can also involve the orbital apex, cavernous sinus, optic nerve, middle cranial fossa and presents as a case of ophthalmoplegia[10]. Also Olfactory neuroblastoma,

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known as esthesioneuroblastoma is a specific type of malignant neuroectodermal tumor thought to arise from neuroepithelial elements in the olfactory mucosa, can be associated with ophthalmoplegia.

Diagnosis: The patient's medical history, family history and detailed examination usually help to differentiate the various causes associated with ophthalmoplegia. In addition Computed tomography scan (CT scan) or magnetic resonance imaging (MRI) scan of the brain, orbit and sinuses are needed to rule out underlying pathology in sinuses, nasopharynx, extent of orbital involvement, intracranial tumour extension, extra-ocular muscle involvements and orbital apex involvement.

CASE 1 (Olfactory Neuroblastoma With Nasopharyngeal Angiofibroma With Total Ophthalmoplegia): A female of 9 years was brought to the Department of Eye and Ear Nose Throat in a Tertiary Care Centre in Kolkata; with chief complaints of persistent headache, neck pain, conductive hearing loss, an unilateral left sided nasal obstruction, continuous dribbling of mucus with profuse and recurrent bleeding from nose, diplopia ; pain in both the eyes, abnormal behavior. On examination a polypoidal mass protruding from the left nostril. Associated with proptosis, adenoid facies, slurred speech, high arched palate, increased inter-canthal distance, puffy oedematous face, rhinorrhoea, poor IQ level and

mental retardation. CT scan of PNS revealed large heterogeneous space occupying lesion filling both nasal cavities, nasopharynx, both ethmoid & sphenoid sinuses extending to pterygoid region, orbits, sellar & suprasellar region; associated with gross erosion. Mass is a soft tissue attenuation with relatively homogenous enhancement. There is presence of focal calcification. Biopsy revealed a lesion which is unencapsulated and contains two different elements. One part shows proliferative thin and thick walled blood vessels separated by fibrous septa. The other part shows a tumor composed of small round cells having scanty cytoplasm and prominent round to oval nuclei. These cells show high N:C ratio, anisonucleosis and hyperchromatic nuclei with prominent nucleoli. The cells are arranged in nests with attempts at pseudorosette formation. Stroma shows desmoplastic reaction and sparse chronic inflammatory cell infiltrate. Patient was planned for radiotherapy. The radiation target volume included the nasopharynx and also the paranasopharyngeal space, oropharynx, base of the skull, sphenoid sinus and also the posterior ethmoidal sinus. High megavoltage radiation was planned with a dose of 65-70Gy to the primary tumor & 65 -70Gy to the neck nodes. Furthermore chemotherapy with Cisplatin-based regimens with 5-FU Cisplatin was planned with radiotherapy as an adjuvant therapy. Unfortunately this patient was lost to follow up.



Fig 1: Clinical presentation of the patient with a polypoidal mass protruding from the left nostril. Associated with proptosis, adenoid facies, slurred speech, high arched palate, increased inter-canthal distance, puffy oedematous face, rhinorrhea.

Fig 2: CT scan of PNS revealed large heterogeneous space occupying lesion filling both nasal cavities, nasopharynx, both ethmoid & sphenoid sinuses extending to pterygoid region, orbits, sellar & suprasellar region, associated with gross erosion.

CASE 2 (Total Ophthalmoplegia With Carcinoma Maxilla): A 58 years male patient who is a farmer by occupation appeared in a tertiary care centre of Kolkata with complaints of right nasal discharge, bilateral nasal obstruction, right cheek swelling, protrusion of right eye, inability to move right eye and diminished vision in right eye since 4 months chronologically. There was also history of hyponasal speech, mouth breathing and snoring, Right sided headache, right ear pain and double vision. No history of frank nasal bleeding, postnasal discharge, cheek numbness, difficulty in swallowing, voice change was present. On clinical examination, facial asymmetry was present, fullness in right medial canthus, increased inter canthal distance



Fig 3: Examination shows facial asymmetry, fullness in right medial canthus and increased inter canthal distance

was there. Mucoïd discharge was present in right vestibular region and external nares. On anterior rhinoscopy, a single pink exophytic mass with irregular surface covered with discharge and slough seen filling entire nasal cavity. On probing it was insensitive, firm in consistency, scanty bleeding, not friable, able to pass probe all around except laterally. On ocular examination conjunctiva was congested and chemosed in right eye lower corneal opacity present with absent sensation. fundus examination was normal in both eyes. No eye movement present on right side. The patient was treated Total Maxillectomy with Orbital Exenteration with bilateral neck dissection. There was no recurrence at followup after six months.



Fig 4: CT Scan of the patient showing ca maxilla right side with orbital involvement

CASE 3 (Nasopharyngeal Carcinoma With Total Ophthalmoplegia): A 57 year old male presented with abrupt-onset ocular pain, blurred vision and difficulty in right eye movement which he had first noticed one month prior to presentation. On observation visual loss to no light perception on the right was found with exophthalmos and ophthalmoplegia. The patient's medical history was otherwise unremarkable. Nasal endoscopy revealed a mild contour protrusion without mucosal ulceration or necrosis in the nasopharynx. Neck CT revealed a soft tissue mass in the extraconal and intraconal space of the right orbit. Moreover, asymmetry of the left nasopharynx and metastatic

lymph nodes were present in the left level II, III, IV, and retropharyngeal areas. On PET/CT scan, the main mass in the right orbit was of a hypermetabolic nature with a maximum standardized uptake value (maxSUV) of 3.5. Abnormal FDG uptake was present in multiple left neck lymph nodes, identical to CT findings. Transnasal endoscopic biopsy, of the right orbital mass and left nasopharyngeal mucosa, was performed under general anesthesia. Histopathologic examination of both specimens revealed undifferentiated carcinoma. The patient received two cycles of chemotherapy with Docetaxel and Cisplatin. However, the patient died six months after diagnosis.



Fig 5: Computed tomography (CT) images of case 3. Axial (A, C, D) and coronal (B) images reveal a soft tissue mass (★) in the extraconal and intraconal space of the right orbit. Moreover, asymmetry of the left nasopharynx and metastatic lymph nodes is noted in the level II (black arrow) and retropharyngeal areas (white arrow) of the left neck.

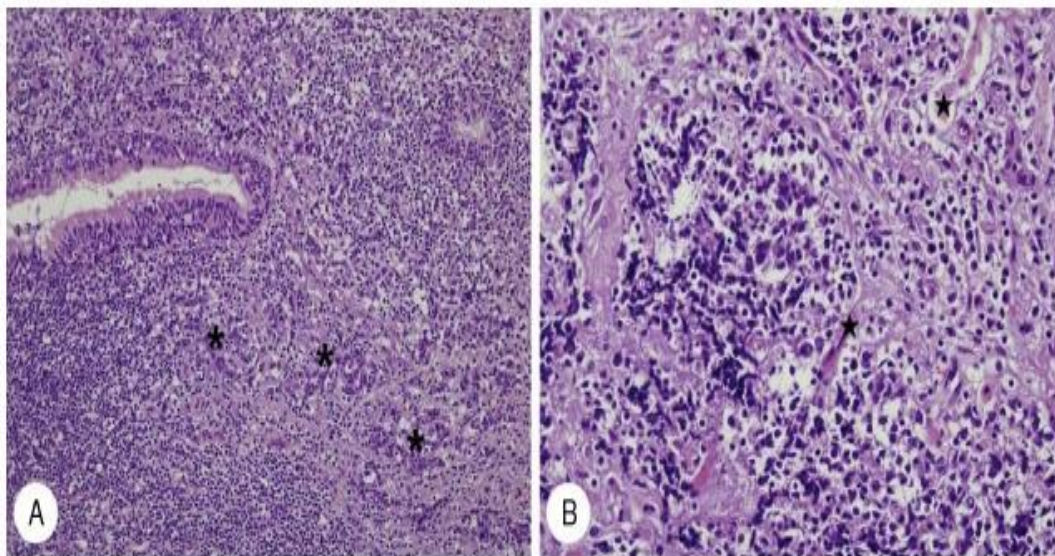


Fig 6: Histopathological Examination shows: (A) Infiltrating undifferentiated non-keratinizing carcinoma (*) of the nasopharynx is noted beneath normal ciliated columnar pseudostratified epithelium.(B) Nasopharyngeal undifferentiated carcinoma infiltrates orbital soft tissue and destroys extraocular striated muscles

Outlook on head and neck cancer with ophthalmoplegia:

Head and Neck cancer is the sixth most common cancer worldwide[11]. It primarily affects men between the sixth and the seventh decades of life. Apparently, the incidence of the association between Head and neck cancers and Ophthalmoplegia is currently unknown and when judging the cases presented herein, one could suggest that this association might be unrecognized and might not be as rare as once believed. The diagnosis of Ophthalmoplegia is clinical; however, a detailed history and physical examination should be performed. Imaging is important in discerning the causative etiology. Most of the patients diagnosed with distant metastasis of HNC are considered to be incurable and are often treated by palliative care protocols, such as chemotherapy or radiotherapy. Overall, up to 80% of all Head and neck cancer patients are diagnosed with advanced stages of the disease, presenting poor prognosis and requiring aggressive multimodality treatment in association with multidisciplinary supportive care where dental oncologists[12] may contribute to the early diagnosis of the disease relapse or relevant complications of tumor progression like ophthalmoplegia.

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