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Case Report

Rochen-Duvigneaud syndrome with nasopharyngeal carcinoma

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ABSTRACT

Nasopharyngeal carcinoma is an uncommon head and neck malignancy which can rarely present with cranial nerve palsies. Here we report a case of nasopharyngeal mass which presented initially with ophthalmic complaints due to compression of cranial nerves in superior orbital fissure resulting in Rochen-Duvigneaud syndrome. It was histologically proven as non keratinising differentiated type of Nasopharyngeal carcinoma

Keywords: Rochen-Duvigneaud syndrome, Nasopharyngeal carcinoma

INTRODUCTION

Nasopharyngeal carcinoma is an uncommon head and neck neoplasm with incidence less than 1 per 100000 except in endemic areas.[1] The most common presentation of NPC is the presence of neck node, followed by nasal and otologic symptoms.[2] Cranial nerve palsy occurs in 8 to 12.4% cases of NPC.[3]

Here we report a rare case of superior orbital fissure syndrome also known as Rochen Duvigneaud syndrome due to the compression of cranial nerves III, IV, V1, VI in superior orbital fissure by nasopharyngeal tumour which presented initially with ophthalmic complaints and later proved on biopsy as nonkeratinizing differentiated type of nasopharyngeal carcinoma.[4]

CASE REPORT

A 54 year old male patient presented to an ophthalmologist with pain right side of the face and drooping of eyelid right side. There was no history of recent dimness of vision. On ophthalmologic examination his visual acuity was 6/9 on right side. His field of vision was reduced on right. He also complained of paraesthesia and numbness on right side of forehead. His CT brain was done which showed a soft tissue density lesion in the posterosuperior nasopharynx. He presented to the ENT department at our hospital with ptosis and proptosis of about 1 month duration, decreased hearing and tinnitus right ear. On examination, there was gross deviation of nasal septum which had occurred due to trauma earlier. He had diminished light reflex on right. Extraocular muscle movements were restricted in all directions. He had decreased sensation on right side of forehead and absent corneal reflex on right side. His sensations were intact over the region of right cheek and jaw. On enquiry, he reported decreased lacrimation from his right eye. MRI brain showed ill-defined hyperintense mass lesion in posterior nasopharynx more towards right side extending superiorly to right superior orbital fissure and adjacent right cavernous sinus. Mass lesion measured 3.2×2 cm in nasopharyngeal region and 2×1.2 cm in right SOF/ cavernous sinus regions. Lesion caused mass effect over right ICA and adjacent cranial nerves in right superior orbital fissure. Bilateral optic nerves appeared mildly bulky with hyperintense signals. MRI cerebral angiogram showed right ICA mildly compressed at the cavernous sinus regions. Nasal endoscopy was done which showed a proliferative slough covered mass in the nasopharynx more to right side. His tympanic membrane was retracted on right side. On pure tone audiometry, he had mild conductive hearing loss, 36 dB and a B type tympanogram was reported. Specimen was taken from the nasopharyngeal mass under endoscopic guidance and

sent for biopsy. Histopathological examination report came out as nasopharyngeal carcinoma nonkeratinizing differentiated type. He did not have any lymph-node involvement or distant metastasis. He was advised chemoradiation as it is a stage IV tumour T4N0M0.

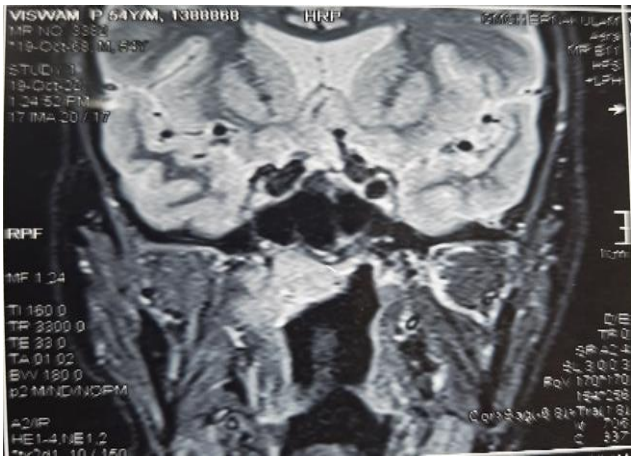

Figure 1: The nasopharyngeal mass.

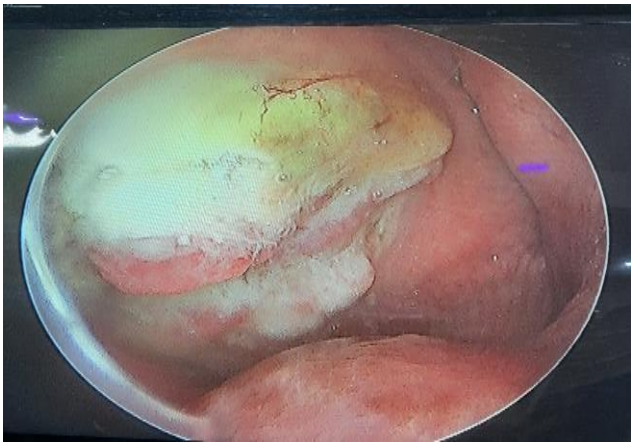

Figure 2: Nasal endoscopy of slough covered mass in nasopharynx.

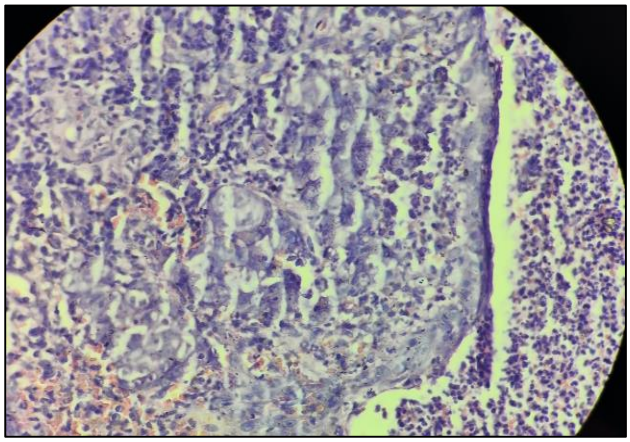

Figure 3: Histopathology of non keratinising differentiated type of nasopharyngeal carcinoma.

DISCUSSION

The nasopharynx is part of the upper airway system connecting the nasal cavities with the larynx and trachea, through the oropharynx. Its superior anatomical limit is the skull base and inferior limit is the soft palate. Important anatomical elements in this area include eustachian tube orifice, torus tubaris and fossa of rosenmuller. The most frequent malignant tumour found at this region is nasopharyngeal carcinoma which arises from the fossa of Rosenmüller.[5]

Nasopharyngeal carcinoma (NPC) is an aggressive malignant tumor that arises from the nasopharyngeal epithelial lining. It is relatively uncommon. The estimated prevalence was said to be 0.7% of all cancers worldwide, according to the world health organization (WHO).[6] NPC can occur at any age, most commonly between the ages of 40 and 60, and has a 3:1 male to female predominance.[7] Nasopharyngeal carcinoma was first described as a separate entity by Regauda and Schmincke 1921.[8]

The common presentation of patients with NPC is with neck swelling, symptoms of nasal obstruction, or serous otitis media (due to obstruction of the Eustachian tube). Less common presentations include cranial nerve palsy, headache, and tinnitus.[7] As per previous studies the most common cranial nerves affected are 5th and 6th.[9]

Our patient presented initially to the ophthalmologist with ptosis indicating early involvement of 3rd cranial nerve. On examination, all extraocular muscle movements were affected which showed paralysis of III, IV, VI cranial nerves. He had severe pain and paraesthesia of right side of forehead and retroorbital pain on right side. He had absent corneal reflex. But sensations over face in maxillary and mandibular regions were intact. This showed the involvement of ophthalmic division of trigeminal nerve (V1). MRI showed mass lesion in the nasopharynx extending to the superior orbital fissure and cavernous sinus on the right side causing mass effect on internal carotid artery and cranial nerves in superior orbital fissure.

The superior orbital fissure lies at the orbital apex and at the border between the orbital roof and the lateral orbital wall and serves as a pathway between the orbit and the middle cranial fossa. It is almost pear shaped with its broader part at the nasal side medially and apex laterally with the long axis extending upward at a 45 degree angle. The size of the fissure in an adult is around 22 mm in length, 2 to 3 mm width at the apex and about 7-8 mm at the base. The tendon of the lateral rectus muscle divides the tissue into two parts: the superior and inferior part. The superior part contains the trochlear nerve (IV), frontal and lacrimal branches of the ophthalmic division of the trigeminal nerve and the superior branch of the ophthalmic vein whereas the inferior part contains the superior and inferior branches of the oculomotor nerve

(III), abducens nerve (VI), nasociliary nerve (V) and the inferior branch of the ophthalmic vein.[10]

Superior orbital fissure syndrome (also known as Rochen-Duvigneaud syndrome) is a collection of symptoms caused by compression of structures just anterior to the orbital apex. This was first described by Hirschfeld in 1858 and officially named by Rochen-Duvigneaud in 1896.[4] The main causes of this syndrome includes traumatic injury, neoplasm, inflammation and vascular.[11] The clinical features of this syndrome are proptosis, ptosis, external ophthalmoplegia and paraesthesia along with lacrimal hyposecretion and retroorbital pain.[4,10]

The important differential diagnosis of superior orbital fissure syndrome are orbital apex syndrome and cavernous sinus syndrome.[4] In orbital apex syndrome cranial nerve II or optic nerve is also affected causing loss of vision. But in our case vision was preserved in right eye. Cavernous sinus syndrome usually involve the whole of trigeminal nerve and may be associated with horners syndrome. But in our case, sensations were intact over maxillary, mandibular divisions of trigeminal nerve and there was no miosis or anhidrosis.

A study by Alrashed et al shows 6 cases of nasopharyngeal carcinoma with ophthalmic presentations and there are 3 other case series in literature earlier with ophthalmic complaints as the initial presentation of NPC.[6] A case of 3rd cranial nerve palsy as the presenting neurophthalmic feature of NPC has been reported by Beckmann et al.[9] A study by Padungkiatsagul et al has reported isolated Horner's syndrome as the initial presentation of NPC.[12] But our patient did not have any miosis or anhidrosis. Studies by Agarwal et al Popovska et al have reported cranial nerve palsies due to cavernous sinus involvement in nasopharyngeal malignancies.[13,14] A case of orbital apex syndrome secondary to nasopharyngeal carcinoma has been reported by Sandhya et al.[15] To the best of our knowledge this is the first case report of nasopharyngeal carcinoma presenting as Rochen Duvigneaud syndrome.

NPC is broadly divided by the WHO into keratinizing (upto 25%) and nonkeratinizing subtypes. Nonkeratinizing NPC, is further subclassified into differentiated (almost 15%) and undifferentiated (upto 65%).[16] On histopathologic examination, our case was nonkeratinizing differentiated type, the least common among the three.

Radiotherapy is the standard treatment for NPC. It is also chemo sensitive. Surgery is reserved for recurrent and resistant cases.[6] Our patient was advised chemoradiation.

CONCLUSION

Our case of nasopharyngeal carcinoma is significant because of its rare presentation with ophthalmologic complaints, cranial nerve palsy due to spread superior orbital fissure and also because of its less common histopathology. To the best of our knowledge this is the first case report of nasopharyngeal carcinoma presenting as Rochen Duvigneaud syndrome.

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