Correspondence:**Ectopic olfactory neuroblastoma**

Nasopharynx, defined as nasal part of the pharynx extending from the base of the skull to the upper surface of the soft palate and in front communicates through choanae with nasal sinuses is a rare site for the occurrence of neuroblastomas. Usually most of the lesions involving the nasal cavity are epidermoid carcinomas but neuroblastomas are rare. Olfactory neuroblastoma arises from sensory epithelium in upper nasal fossa at the level of cribriform plate. It constitutes about 5 % of malignant tumours of nasal cavity. It is highly prevalent in Chinese population with a reported incidence of 1 in 200,000 population.¹ The incidence of carcinoma of nasopharynx in United States is 0.8/100,000 for males and 0.03/100,000 for females. It is most common in the second and fifth decades of life but can be seen at any age and occurs equally in males and females. Pathologically 90% of malignant lesions arising from the nasopharynx are epidermoid or undifferentiated carcinomas, the other 10% are mainly lymphomas, plasmacytomas, melanomas, and rhabdomyosarcoma and chordomas. The literature search revealed involvement of nasopharynx by esthesioneuroblastoma is very rare when compared to other tumours.

A 22-year-old male presented with complaints of blockage of left nasal cavity and anosmia of 2 months duration and epistaxis (10 episodes per day) not associated with foul smelling discharge. He also complained of headache which was holocranial and intermittent, that was aggravated on exertion and relieved spontaneously. This was not associated with either vomiting or blurring of vision. Giddiness on changing the posture from standing to sitting was evident. There was deviation of left eye ball laterally and difficulty in movements of left eye present. There was no history suggestive of diplopia, blurring of vision, loss of conscious-

ness, seizures, earache or discharge from ear, deafness, gait disturbances or weakness, sensory deficits, urine or bowel habit disturbances, fever or trauma to nose. The patient was moderately built and nourished and was anaemic. Examination of the nasal cavity revealed the presence of a friable mass extending into left nasal cavity without the presence of either deviated nasal septum or hypertrophied turbinates. Pre-operative computed tomography showed the presence of a mass extending into left nasal cavity and involving bilateral maxillary sinuses, left frontal, left sphenoid and left ethmoid sinuses. Mass was also extending into extraconal compartment of left orbit involving left superior rectus and eroding the cribriform plate (Figure 1). The patient underwent excision of mass under general anaesthesia by lateral rhinotomy through Moore's approach. A mass was found to be arising from the nasopharynx and extending onto nasal cavity and bilateral nasal sinuses. Wide local excision was done and the excised material was subjected for histopathological examination which revealed the tumour cells seen predominantly arranged in large lobules, separated by fibrovascular stroma with few areas of solid nests and cribriform patterns (Figure 2). Occasional tumour lobules showed Homer - Wright rosettes (Figure 3). The cells were small to intermediate sized having vesicular nucleus with single nucleolus and scanty acidophilic cytoplasm (Figure 3). Atypical mitotic figures (2-3/10 high power fields) was noted. No neurofibrillary background was seen. The stroma showed extensive areas of haemorrhage with mild lymphocytic infiltration and the case was diagnosed as esthesioneuroblastoma. Post-operative period was uneventful. The patient was discharged and referred for radiotherapy.

Figure 1: Coronal CT showing mass extending from left nasal cavity and involving bilateral maxillary sinuses, left frontal, left sphenoid and left ethmoid sinus

Esthesioneuroblastoma is an uncommon malignant neoplasm, representing 2%-3% of sinonasal tract tumors with an incidence of approximately 6%.² These tumours arise from the olfactory neuroepithelium, which extends from the roof of the nose to the area of the superior turbinate and a portion of the nasal septum. The development of olfactory neuroblastoma outside the region where olfactory neuroepithelium does not normally exist is extremely rare. It has

been rarely reported in the literature from the regions such as the maxillary and sphenoidal sinuses, the petrous apex, nasopharynx and the pituitary gland.³

To conclude, ectopic olfactory neuroblastoma is a rare entity and with the invention of new modalities of treatment like intensity-modulated radiation therapy and Image-guide radiation therapy a better prognosis can be expected. The Radiation Therapy Oncology Group reported that combined radio and chemotherapy

Figure 2: Photomicrograph showing the tumour cells seen predominantly arranged in lobules, separated by fibrovascular stroma with few areas of solid nests and cribriform patterns (Haematoxylin and eosin, $\times 400$)

Figure 3: Photomicrograph showing tumour lobules with Homer - Wright rosettes. The cells were small to intermediate sized having vesicular nucleus with single nucleolus and scanty acidophilic cytoplasm (Haematoxylin and eosin, $\times 400$)

will give long term survival rates. In recent systematic review of published series spanning 40 years, a steady improvement of overall survival of patients was noted.⁴

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