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Case Report

Ectopic primary olfactory neuroesthesioblastoma originating from the anterior nasal septum

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ABSTRACT

Olfactory neuroestesioblastoma (ONB) is a rare malignant neoplasm of the paranasal sinus. Ectopic growth of ONB outside the region of neuroepithelial cells are exceedingly rare and so far, it has only been reported to arise from the maxillary sinus, sphenoid sinus, posterior septum and the turbinates. None has been reported to originate from the anterior cartilaginous part of the septum. A 31-year-old lady, who was premorbidly well, presented with 2 episodes of spontaneous epistaxis from the left nostril in 1 year. The epistaxis worsened over the one week prior to her visit. Nasoendoscopy showed a vascularised round mass at the anterior part of the left septum. Initial biopsy was reported as possible ectopic squamous cell carcinoma of the salivary gland. A repeat biopsy then confirmed the diagnosis of ONB. Computed tomography scan of the paranasal sinus showed an isolated mass (18×18 mm) arising from the anterior part of the septum. Endoscopic resection of the tumour was done and histopathology report revealed as ONB. Post-operative radiotherapy was given with a total dose of 66. The rarity of ONB poses a challenge during diagnosis. The histological identification is difficult as these tumours may show little or no differentiation. A simple biopsy from the mass is often insufficient and may produce inaccurate results. Isolated growth without involvement of the upper nasal vault is extremely rare. A possible explanation would be the presence of ectopic neuroepithelial cells at the cartilaginous anterior septal area that has failed to degenerate during fetal development.

Keywords: Olfactory neuroesthesioblastoma, Ectopic growth, Nasal septum

INTRODUCTION

Olfactory neuroesthesioblastoma (ONB) is a rare malignant neoplasm of the paranasal sinus. It represents 2% to 3% of all malignant sinonasal tumours. This slow-growing tumour, first described by Berger and Luc in 1924, originates from the neuroepithelial cells of the olfactory membrane. Commonly, ONB arises from the upper third of nasal septum, cribiform plate and medial surface of the superior turbinate.[1] Ectopic growth outside the region of the neuroepithelial cells are exceedingly rare. So far, it has only been reported to arise from the pituitary gland, maxillary sinus, sphenoid sinus, inferior turbinate and the posterior septum.[2] None was reported to have originated from the anterior cartilaginous part of the septum as it is in our case.

CASE REPORT

A 31-year-old lady, premorbidly well, presented with recurrent episodes of left sided epistaxis for one year. There was no associated history of anosmia, nasal blockage or headaches. Nasoendoscopic examination revealed a firm, vascularised mass at the left anterior septum adjacent to the Kiesselbach's plexus (Figure 1). Paranasal sinus CECT scan reported a moderately enhancing, round, lobulated mass measuring 18×18 mm arising from left nasal septum (Figure 2). There was no

bony erosion or intracranial extension. Two biopsy samples were taken. The initial sample was reported as low grade mucoepidermal carcinoma with the differential diagnosis of pleomorphic adenoma. Immuno-histochemical staining was positive for pancytokeratin, p63 and cytokeratin 5/6/7. Interestingly, the second sample was reported as ONB based on the microscopic findings of pseudorosette formations and fibrillary stroma. A transnasal endoscopic resection of the mass was performed with clear margin and was then followed by intensity-modulated radiotherapy with a total dose of 66Gy over a 6-week period. Histologically, the resected mass appeared to have Homer-Wright and Flexner-Wintersteiner rosettes, as well as perivascular pseudorosettes. The final diagnosis was ONB.

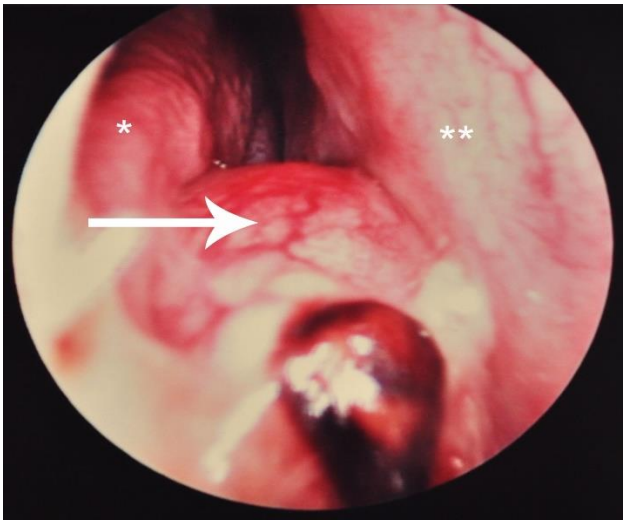

Figure 1: Endosopic finding of left nasal cavity showing a vascularised round mass arising from the anterior part of the nasal septum (arrow); nasal septum, inferior turbinate.

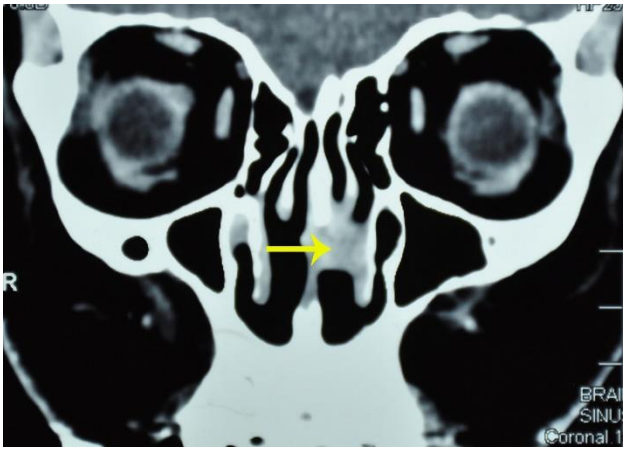

Figure 2: CECT of the paranasal sinus showing a moderately enhancing, well lobulated mass in the anterior nasal septum (arrow).

DISCUSSION

The rarity of ONB presents a diagnostic challenge to the surgeons and pathologists. Ectopic growth of this tumour have been reported in literatures. In our case, this lady had an isolated growth at the anterior cartilaginous part of the nasal septum. To our knowledge, this is the first case that has ever been reported to date. As the specialised sensory neuroepithelial olfactory cells are usually found in the upper third of nasal cavity, a possible explanation for this unusual presentation is that the neuroepithelium has failed to degenerate during fetal development leaving the presence of ectopic cells beyond the neuroepithelial region.[2]

Given that this tumour has a broad histological spectrum, it can easily be confused with other small round cell tumours and undifferentiated carcinomas.[2] This is clearly demonstrated in our patient where there were two different histological interpretations of the biopsy sample taken from the same site. Generally, lower grades ONB (Hyams grade I and II) can be easily identified and diagnosed under light microscopy owing to the presence of rosettes and neurofibrillary matrix. Immuno-histochemical staining will only serve as an adjunct to diagnosis. Higher grades tumour (Hyams grade III and IV) may cause a diagnostic dilemma as the rosettes and neurofibrillary matrix may be absent or scanty. In our case, the biopsy samples represent both ends. The immunohistochemical stain which can reliably diagnose olfactory neuroblastoma is the nonspecific stain neuron-specific enolase which was not done in this case.[3] While cytokeratins and p63 may be used, the results are variable.[2,4]

It has been well established that the primary treatment for ONB consists of open surgery and radiotherapy; open surgery mainly done to obtain the tumour en bloc and protect the optic nerve. However, the advancement in endoscopic technology has made endoscopic approach more feasible and they have been reported to have equal or better outcomes than open surgery.[5] Our patient had endoscopic resection and radiotherapy. She has shown good progression upon treatment completion.

Isolated ONB is rare and the common presenting symptoms are non-specific. Based on our experience, even in the absence of a mass in the upper nasal cavity, the possibility of an ONB should be considered whenever a suspicious mass is seen in the anterior nasal cavity. As histological diagnosis can be difficult, a simple biopsy is often not sufficient for diagnostic purpose as highlighted in our case.

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