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A rare presentation of olfactory neuroblastoma- A case report

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Abstract

Olfactory neuroblastoma is a rare tumor arising from the olfactory epithelium with a varied clinical and radiological presentations. This is a case of a 28-year-old female who presented to our emergency department with unilateral spontaneousuncontrolled nasal bleeding. Contrast enhanced Computed Tomography was done, which showed features of a hyper enhancing polypoidal mass in the upper part of right nasal cavity with no erosion of cribriform plate or lamina papyracea. Patient underwent endoscopic excision biopsy of the mass under general anesthesia.

HistoPathological examination revealed it as Olfactory neuroblastoma and the patient was subjected to Radiotherapy.

The purpose of this study is to report the unusual presentation of a bleeding mass along with the absence of any bony erosion on CECT in this rare Sino nasal tumour.

Keywords: Esthesio neuro blastoma, Rare presentation of Sino nasal tumour

Introduction

Olfactory neuroblastoma is an uncommon neuroectodermal tumour arising from the Olfactory neuroepithelium in the nasal cavity. The various appellations for this tumour include Esthesioneuroblastoma, Esthesioneuroepithelioma, Olfactory placode tumour, Esthesioneurocytoma.

The incidence is relatively low worldwide, about 0.4 cases per million, which accounts for about 2- 3 % of all Sino nasal tumours. First described by Berger and Luc in 1924, it has a bomiodal age distribution at 2nd and 6th decade.

Nasal obstruction is the most common symptom (>90%), while Epistaxis (<50%), headache, facial pain, anosmia,

visual disturbances, excessive lacrimation and rhinorrhea are relatively rare.

The classical picture on Computerized Tomography is that of a" dumbbell-shaped mass", with its upper portion in intracranial fossa, lower portion in the nasal cavity and its waist at the cribriform plate showing surrounding bony erosion of lamina papyracea and the cribriform plate. On CECT it shows moderate to high post contrast enhancement.

Homer wright pseudo rosettes are seen in about 30% of cases and Flexner-Winter Steiner true rosettes are seen in about 5% of patients.

Case report

A 28-year-old female presented to our Emergency Department with right sided spontaneous uncontrolled nasal bleeding, with no history of any trauma, nasal obstruction, nasal discharge or anosmia.

On examination there was profuse nasal bleeding from the right nasal cavity while the left nasal cavity was normal with no post nasal bleeding. Immediately bilateral anterior nasal packing was done and the bleeding was arrested.

Patient's bleeding and clotting profiles were within normal limits.After 48 hours when the nasal pack was removed the bleeding had subsided.

On Diagnostic Nasal Endoscopy, a reddish, polyp was seen arising from upper part of the nasal cavity between the middle turbinate & septum. The exact site of attachment could not be visualized.

Probing was not done as the mass appeared vascular and prone for bleeding. Rest of the right nasal cavity, left nasal cavity & Nasopharynx were normal.

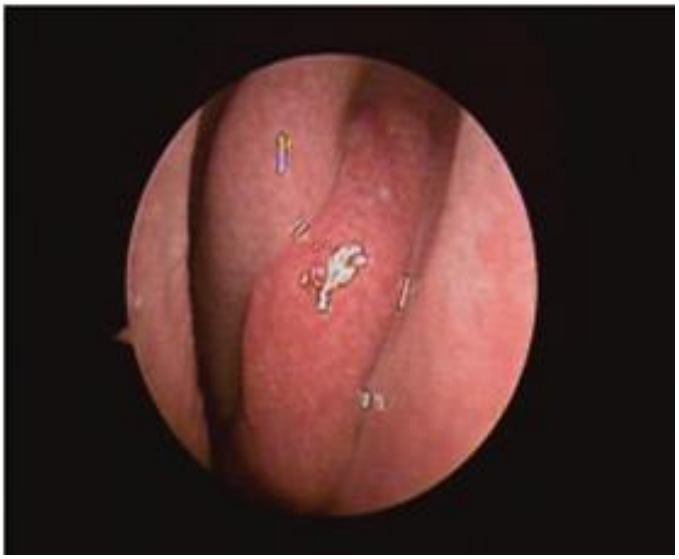

Figure 1: Diagnostic Nasal endoscopy (right) showing reddish polypoidal mass between the middle turbinate and septum

Contrast Enhanced CT PNS [coronal, axial & sagittal] showed a small well defined hyper enhancing polypoidal tissue in the right nasal cavity abutting nasal septum & right middle turbinate, measuring approximately 35× 4× 24 mm (AP×TR×CC) with no adjacent bone erosion. Rest of the nasal cavity and the sinuses were normal. With this history, examination and investigation, a provisional diagnosis of hemangioma of nasal septum was made.

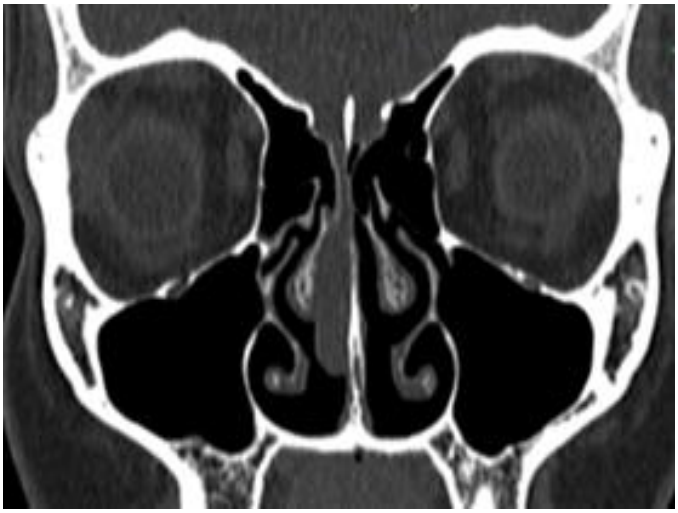

Figure 2: CT PNS (before contrast)

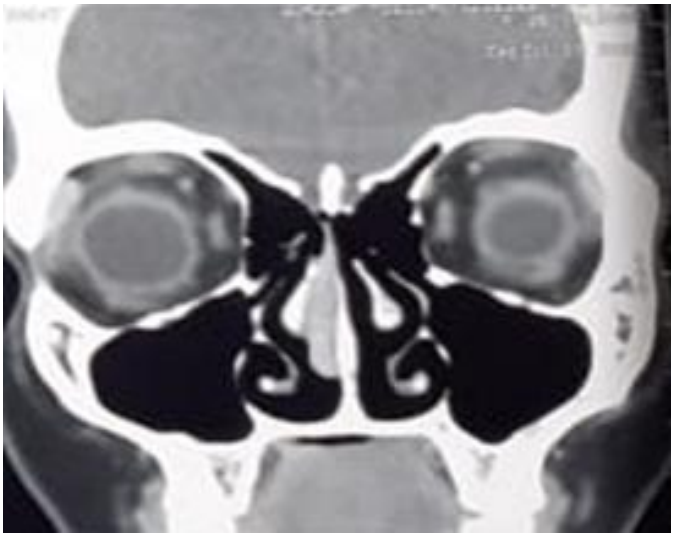

Figure 3: CT PNS (Post- contrast)

The patient was taken for an endoscopic excision biopsy of the mass. Intraoperatively, a single pedunculated mass was seen arising from the upper nasal cavity which bled on touch.The lower part of the mass was excised for HPE using bipolar cautery and the rest of the mass was removed completely using aCoblator. The stalk attachment was found to be in the upper part of nasal septum near the roof.

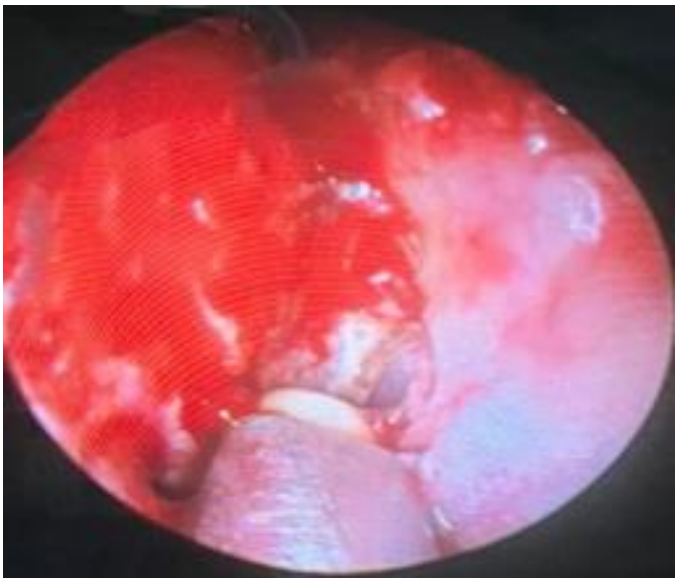

Figure 4: Surgical removal using Coblator

Histopathology report showed uniform round cells in lobules separated by highly vascular interlobular stroma. Nuclei show anisocytosis, stippling of chromatin and indistinct nucleoli. Neurofibrillary stroma was less prominent. No areas of necrosis.

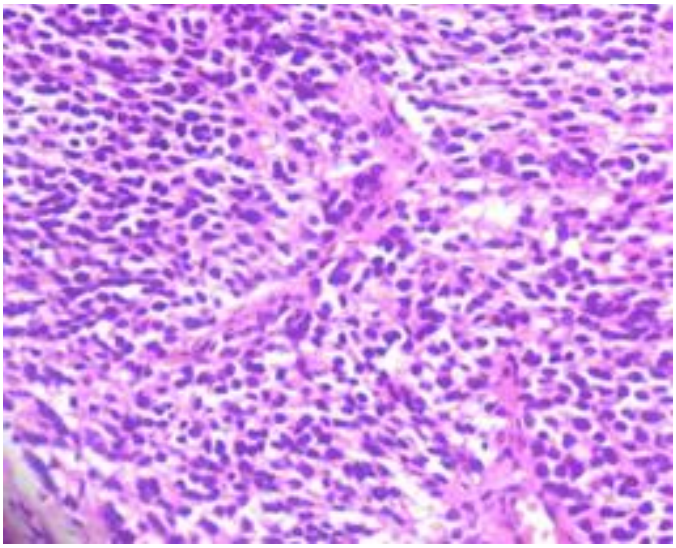

Figure 5: HPE showing uniform round cells in lobules, separated by vascular stroma

Immunohistochemistry showed, the cells were diffusely strongly positive for chromogranin. S100 showed positivity in sustentacular cells. CD 20 and CK (AEI/ AE3) were negative. Based on the histopathological a final diagnosis was made as Olfactory neuroblastoma (Hyam's grade II).

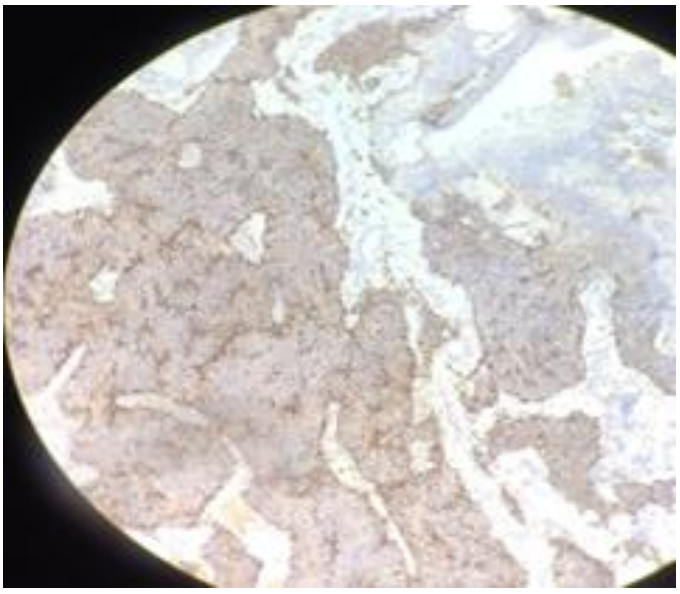

Figure 6: IHC showing Chromogranin positivity

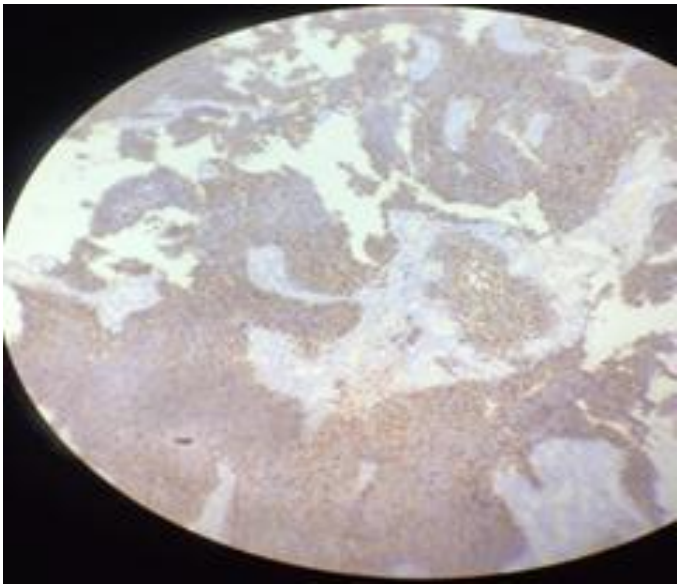

Figure 7: IHC showing S100 positivity

The immediate post op period was uneventful. After 3 weeks, a postop- DNE was done which showed no residual lesion.

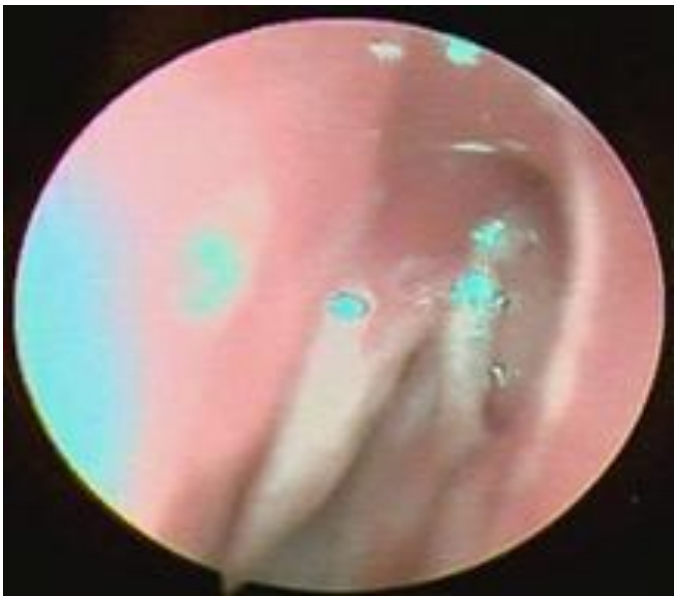

Figure 8: Postoperative DNE at 3 weeks showing no residual lesion

A whole-body PET-CT was done to rule out any residual tumour but no metabolically active lesions were found. Patient was advised radiotherapy based on previous studies suggesting a high recurrence rate.

Discussion

Olfactory neuroblastoma is a relatively rare tumour of the Sino nasal tract(1). It is difficult to diagnose when it doesn't present with distinctive clinical or radiographic features. Clinically it can mimic many other tumours of the Sino nasal tract making the diagnosis much more complex. Epistaxis was the only feature in our patient and is rare as an isolated presentation for olfactory neuroblastomas. Moreover, the classical radiological appearance of a "dumb-bell" shaped mass in the roof of nasal cavity, with erosion of cribriform plate and lamina papyracea was also absent which masked the actual diagnosis. Microscopy shows small round blue cells, with small nuclei and "salt and pepper" nuclear chromatin distribution. Immunohistochemistry helps in a definite diagnosis. IHC markers like CD56, Synaptophysin, Chromogeranin, neuron specific enolase and S-100 are positive, whereas ONBs are negative for myogenic, desmin, CD45RB, CD99 etc.(2).

Chromogeranin, neuron specific enolase and S-100 are positive, whereas ONBs are negative for myogenic, desmin, CD45RB, CD99 etc. (2).

Traditionally olfactory neuroblastoma at the time of presentation usually presents with features of invasion into the surrounding tissues and require a cranio-facial resection followed by radiotherapy to prevent recurrence(3).

However rare cases like ours might present as a solitary polyp without any invasion and in such cases the need for an extensive resection is debatable(4).

The recurrence rates in such solitary lesions are unknown and even though the patient has been advised radiotherapy long term follow up is needed for tumour recurrence(5,6).

Conclusion

• Olfactory neuroblastoma though rare, are an important differential diagnosis of tumours in the nasal cavity especially those arising close to the roof.

• Complete excision followed by radiotherapy is the treatment. Patients require long term follow up for recurrence.

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