



Case Report:

Olfactory Neuroblastoma: Diagnostic Difficulty

Vidya MN, Registrar, Department of Pathology, Apollo Hospitals, Bannerghatta Road, Bangalore 76

Swarna Shivakumar, Consultant Pathologist, Apollo Hospitals, Bannerghatta Road, Bangalore 76

Siddhartha Biswas, Consultant Pathologist, Apollo Hospitals, Bannerghatta Road, Bangalore 76

Vijay Shankar S, Associate professor, Department of Pathology, Adichunchangiri Institute of Medical Sciences, BG Nagara, Nagamangala taluk, Mandya district, Karnataka

Address For Correspondence:

Dr. Vijay Shankar S,

No 45, Ananda shylam,

3rd cross, 1st main ,

KN extension, Yeswanthpur,

Bangalore - 560022,

Karnataka, India.

E-mail: vijayshankarpatho@gmail.com

Citation: Vidya MN, Shivakumar S, Biswas S, Vijay Shankar S. Olfactory Neuroblastoma: Diagnostic Difficulty. *Online J Health Allied Scs.* 2010;9(4):18

URL: <http://www.ojhas.org/issue36/2010-4-18.htm>

Open Access Archives: <http://cogprints.org/view/subjects/OJHAS.html> and <http://openmed.nic.in/view/subjects/ojhas.html>

Submitted: Dec 6, 2010; Accepted: Dec 28, 2010; Published: Jan 20, 2011

Abstract:

Olfactory neuroblastoma is an uncommon malignant tumor of sinonasal tract arising from the olfactory neuro epithelium. The olfactory neuroblastomas presenting with divergent histomorphologies like, epithelial appearance of cells, lacking a neuro fibrillary background and absence of rosettes are difficult to diagnose. Such cases require immunohistochemistry to establish the diagnosis. We describe the clinical features, pathological and immunohistochemical findings of grade IV Olfactory neuroblastoma in a 57 year old man

Key Words: Olfactory neuroblastoma; Nasal cavity; Immunohistochemistry

Introduction:

Olfactory neuroblastoma is an uncommon malignant neoplasm, representing 2-3% of sinonasal tract malignancies. It is a malignant neuro ectodermal neoplasm arising from the olfactory neuro epithelium found in the upper 1/3 to 1/2 of the nasal septum, the cribriform plate and the superior medial surface of the superior turbinate.¹ This tumor has a broad histological spectrum and olfactory neuroblastoma can be confused with other small round cell tumors and undifferentiated carcinomas. The diagnosis becomes difficult when the cells have an epithelial appearance with scant or absent neurofibrillary background. Immunohistochemistry is of great value in differentiating ONB from other sinonasal tract malignancies.

Case Report:

Left nasal polyp biopsy of a 57 year old male patient with 3 year history of left sided nasal obstruction was received in the department of pathology. The patient presented with anosmia, headache and loss vision in the left eye with worsening of symptoms over the last 3 months. CT scan revealed a left paranasal sinus lesion occupying the left maxillary, ethmoidal region with intraorbital and intracranial extensions. MRI revealed a left nasal mass extending and eroding the olfactory

groove at the anterior base with erosion of the left orbital wall. Biopsy was done and sent for histopathological examination.

Gross Features: Multiple irregular grey white soft tissue bits were received with a large polypoidal bit measuring 3x1.5x1 cm. Cut section was grey white with hemorrhagic areas.

Microscopy: Microscopic examination showed an intact respiratory mucosa with a tumor in the submucosa. The tumor cells were seen predominantly arranged in large lobules, separated by fibrovascular stroma with few areas of solid nests, cribriform pattern and comedo necrosis.(Fig.1) Occasional tumor lobules showed Homer – Wright rosettes (Fig 2)

The cells were small to intermediate sized having vesicular nucleus with single nucleolus and scanty to moderate amount of acidophilic cytoplasm (Fig 3). Atypical mitosis of 2-3/10 HPF was noted. No neuro fibrillary background was seen. The stroma showed extensive areas of haemorrhage with mild lymphocytic infiltration.

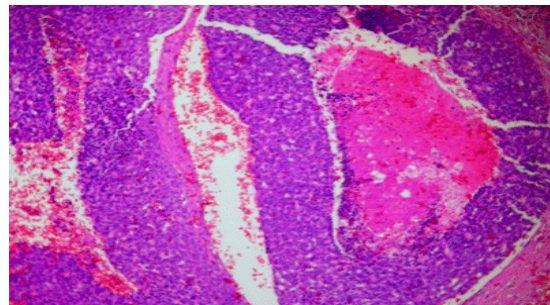


Figure 1: Photomicrograph showing tumor cells in lobules with comedo necrosis (H&E X 100)

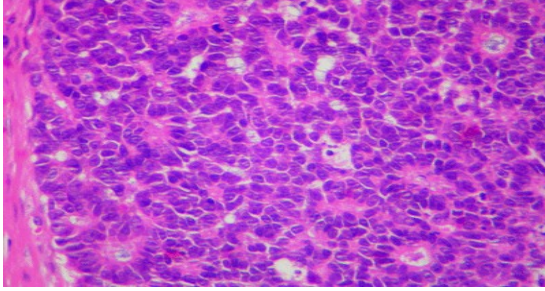


Figure 2: Photomicrograph showing Homer-wright rosettes (H&E X 400)

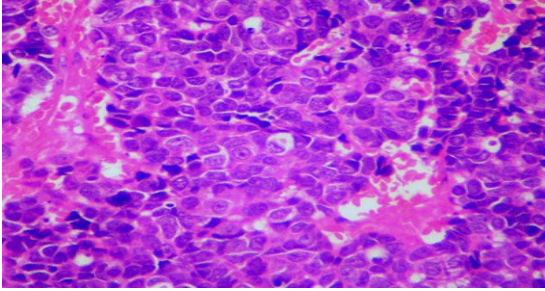


Figure 3: Photomicrograph showing tumor cells with vesicular nucleus with nucleoli & moderate amount of cytoplasm (H&E X 400)

IHC was strongly positive for NSE, CK cocktail and S-100. S-100 showed positivity of sustentacular cells along the periphery of the tumor lobules with few cells within the tumor Lobules (Fig 4 & 5).

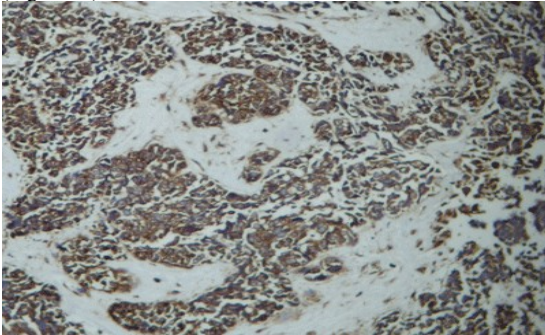


Figure 4: Photomicrograph of immunohistochemical staining showing S-100 positivity of the sustentacular cells in the periphery as well as within the lobules (X 400)

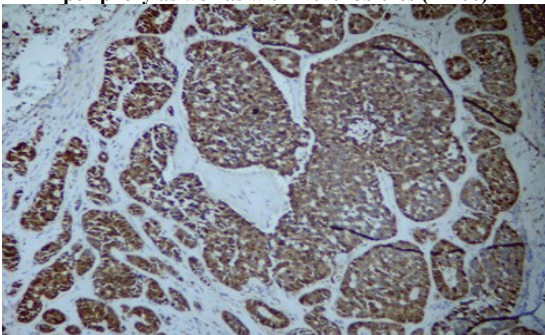


Figure 5: Photomicrograph of immunohistochemical staining of the tumor cells showing Non Specific Esterase positivity (X 400)

The final diagnosis of olfactory neuroblastoma grade IV was made.

Debulking surgery was done. The patient had an uneventful post operative period and was subjected for radiotherapy.

Discussion:

Olfactory neuroblastoma is an uncommon malignant neoplasm, representing 2-3% of sinonasal tract tumors with an incidence of approximately 6%.^{1,2} These tumors arise from the olfactory neuro epithelium, which extends from the roof of the nose to the area of the superior turbinate and a portion of the nasal septum.²

There is no gender predilection;¹ some authors report a male to female ratio of 2:1.³ It occurs in all age groups with a bimodal peak in the 2nd and 6th decade.⁴

The usual clinical symptoms are unilateral nasal obstruction, epistaxis, anosmia, headache, pain and ocular disturbances.¹ ONB has a tendency to spread sub mucosally to involve the paranasal sinuses, nasal cavity and other structures like oral cavity, orbits and the brain.³

Olfactory neuroblastoma of grade I & II (Hyams grade) can be easily recognized as they resemble other neuroblastomas with small round cells arranged in rosettes and an abundant neuro fibrillary background.¹ Grade III & IV (Hyams grade) have little or no neuro fibrillary background with few or no rosette formations making morphological diagnosis difficult. In our case, some of the cells had an epithelial appearance with vesicular nucleus and comedo necrosis closely resembling undifferentiated sinonasal carcinoma. The other differential diagnosis of small cell neuroendocrine carcinoma was considered as the tumor cells showed rosette arrangement with absent neuro fibrillary background. Some areas showed cribriform pattern which is known to occur in ONB and neuroendocrine carcinoma favoring differentiation rather than existence of two separate neoplasms.⁴ With confounding histomorphology, the diagnosis will be difficult which requires immunohistochemistry for confirmation.

ONB shows variable positivity for Cyto keratin, synaptophysin and chromogranin. One of the studies showed cytokeratin (4/11), synaptophysin (7/11) and chromogranin A (1/11).⁵

The main mode of treatment is complete craniofacial resection of the tumor followed by radiotherapy.⁶ The prognosis depends on the grade and stage of the tumor. The 5 year survival Rate is 40% for high grade tumors with a high local recurrence (15-70%).¹

Conclusion:

Olfactory neuroblastoma is an uncommon neoplasm of the sinonasal tract. Histologically, the tumor can have varied cytomorphological appearance with absence of neurofibrillary matrix and occasional areas with rosette formation. IHC will be of immense help in differentiating it from the undifferentiated sinonasal carcinoma.

References:

1. Christopher DM Fletcher. Tumors of the upper respiratory tract. In Christopher DM Fletcher. Diagnostic histopathology of tumors. 3rd ed. Elsevier, 2007. pp 112-118.
2. Ghaffar, Shehzad, Salahuddin, et al. Olfactory neuroblastoma: a case report and review of the literature. *Ear, Nose and Throat Journal*. 2005 March;84(3):150-52.
3. Arafat Ayoub, Mohammed Ali Hiari. Olfactory neuroblastoma: A Case Report. *Journal of Research in Medical Sciences*. Dec 2005;12(2):48-50.
4. Roasi J. Respiratory tract. In Rosai J, ed Ackerman's Surgical Pathology. 9th ed. Elsevier 2004;Vol 1. pp 316-17.
5. Frierson HF Jr, Ross GW, Mills SE, et al. Olfactory neuroblastoma. Additional immunohistochemical characterization. *Am J Clin pathol*. 1991 Jun;95(6):903.
6. Mills, Stacey EMD, Frierson, et al. Olfactory neuroblastoma: A clinicopathologic study of 21 cases. *The Am J of Surg Pathol* 1995;9:317-327.