



ORIGINAL RESEARCH PAPER		Radio-Diagnosis
OLFACTORY NEUROBLASTOMA(ESTHESIONEUROBLASTOMA) – A CASE REPORT		KEY WORDS:

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ABSTRACT	Esthesioneuroblastoma (ENB) is a rare malignant neoplasm of the olfactory epithelium. It can directly invade central nervous system through cribriform plate. However, non-contiguous intracranial involvement without recurrence at the primary site is extremely uncommon. ¹ The optimal therapy is the multimodality treatment. The treatment of ENB correlates with the extent of the lesion, with surgery being the mainstay of therapy followed by postoperative irradiation. ² Herein, we report 37 years old male patient.
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INTRODUCTION Esthesioneuroblastoma is an uncommon cancerous tumor emerging from the olfactory specialized neuroepithelium located in the superior aspect of the nasal vault, commonly invading not only the cranial vault and the floor of the cranial cavity, but also the eye socket. ² Esthesioneuroblastoma is also known as esthesioneurocytoma or neuroepithelioma, or more commonly, olfactory neuroblastoma. The diagnosis is based on histological and immunohistochemical analysis, with surgery and radiotherapy being the main therapeutic options. Olfactory neuroblastoma is characterized by a slow progression with a high recurrence rate even after a long period of remission. It represents less than 5% of all cancers of the nasal cavity. ³	CECT Brain and PNS Coronal view CECT Brain and PNS revealed approx. 55x29x60 mm sized heterogeneously enhancing soft tissue density lesion involving left nasal cavity. Lesion causes erosion of nasal septum and extends into right nasal cavity. Lesion extends into left ethmoid sinus. Lesion causes erosion of left maxillary sinus and extends into it. Lesion causes erosion of all turbinates on left side. Lesion causes erosion of both sphenoid sinuses and extends into it. Lesion causes erosion of cribriform plate and extends intracranially into left frontal lobe. Lesion causes erosion of left orbit and extends into it. Lesion abuts and displaces medial rectus muscle on left side. There is associated mucosal thickening is noted in b/l frontal sinus, b/l ethmoid sinus and maxillary sinus.
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Case Report A 37 years old male patient initially presented in ENT department of medical college in other locality with complaint of nasal blockage on left side and decreased smell sensation. Biopsy sample from nostril made primary diagnosis of malignant round cell tumor. Patient referred to GCRI (Gujarat cancer research institute, Ahmedabad) from there. Oncosurgeon of Head and Neck Unit of GCRI advised CECT scan and MRI of Brain and PNS.	
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Imaging Findings 	
CECT Brain and PNS Sagittal view (Bone window) Showing erosion of bone at roof of nasal cavity 	T1W Post contrast fat suppressed , MRI Brain 
	T2W fat suppressed Transverse View, MRI Brain MRI images revealed same findings and helped delineating tumor accurately. Imaging data in correlation with biopsy findings suggests differential diagnosis of (1) Olfactory Neuroblastoma (2) Rhabdomyosarcoma (3) Ewing's sarcoma. Surgery performed with Endoscopic Trans Nasal Trans Sphenoidal (TNTS) approach and tumor resected and tissue sent for intraoperative frozen section for histopathological examination. The morphological and immunohistochemical data were consistent with Olfactory Neuroblastoma. After around 3 to 4 months of surgery similar complaint re-

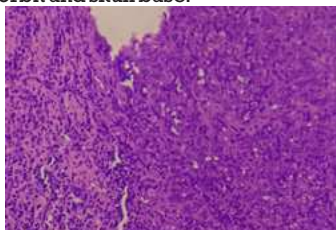
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emerged.

Imaging data revealed recurrence of tumor. Another Surgery performed with Endoscopic Trans Nasal Trans Sphenoidal (TNTS) approach and tumor resected and tissue sent for intraoperative frozen section for histopathological examination, which revealed recurrence of Olfactory neuroblastoma with divergent differentiation. Adjuvant radiotherapy given to patient.

DISCUSSION

The Esthesioneuroblastoma (ENB) is a rare malignant tumor of neuroectodermal origin which arises from olfactory receptors in the nasal mucosa or the cribriform plate of the ethmoid bone. This tumor is most commonly present in the upper part of the nasal cavity which includes the superior nasal concha, roof of the nose, cribriform plate of the ethmoid, and upper part of the nasal septum. It affects both genders equally, with a bimodal age distribution occurring in ages 11-20 and 51- 60 years.⁴ Common symptoms include nasal blockage, epistaxis, pain, loss of smell sensation, headache. Most of the ENBs are non-functioning tumors. ENB often extends into orbit and skull base.



Histology of olfactory neuroblastoma shows neoplastic cells with prominent nuclei

ENB arises as a polypoidal mass in the roof of the nasal cavity. There is high tendency of bleeding on biopsy. Microscopically ENB has a distinct lobular architecture composed of sheets and nests of small cells with scant cytoplasm, uniform round to oval hyperchromatic nucleus in a neurofibrillary matrix. Homer-Wright pseudorosettes are seen in lower-grade tumors and Flexner-Wintersteiner rosettes are seen in higher-grade tumors along with foci of necrosis and increased mitoses.

Surgery is the mainstay of treatment in all resectable cases. Surgery alone (open or endoscopic) with an adequate tumor free margin appears to be enough for small, early-stage tumors with close follow-up and adjuvant radiation kept in reserve in case of recurrence. MRI and CT scan imaging gives information about size and extent of tumor. Radiological imaging data with correlation of biopsy finding makes diagnosis of ENB. Radiological data identifies whether tumor is resectable or non resectable. To achieve tumor free margin surgery is planned according to radiological imaging data. Follow up CT scan/ MRI is done to rule out residual lesion or tumor recurrence.

CONCLUSION

ENB is rare tumor with high rate of recurrence. Establishing careful diagnosis with histopathological findings and radiological imaging and planning of treatment with multidisciplinary approach is of paramount importance. Surgical resection of tumor is mainstay of treatment. Radiation therapy is reserved for aggressive, non resectable or partially resectable tumor and in case of recurrence. Radiological evaluation with MRI and CT scan has important role in treatment planning and follow up.

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