ORIGINAL RESEARCH PAPER
Volume - 13 | Issue - 10 | October - 2024 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

EXTENSION OF NEWLY DIAGNOSED OLFACTORY NEUROBLASTOMA: A RARE TUMOUR

Nuclear Medicine

Dr Patel Meet Vinodbhai Postgraduate Resident, Dept Of Nuclear Medicine, Mahatma Gandhi Medical College & Hospital, Jaipur.

Dr Gurudas Singh Postgraduate Resident, Dept Of Nuclear Medicine, Mahatma Gandhi Medical College & Hospital, Jaipur.

Dr Ram Singh Meena Assistant Professor, Dept Of Nuclear Medicine, Mahatma Gandhi Medical College & Hospital, Jaipur.

Dr Karan Singh Peepre Prof And Head, Dept Of Nuclear Medicine, Mahatma Gandhi Medical College & Hospital, Jaipur.

Dr Yash Mittal* Postgraduate Resident, Department Of Gen. Medicine, Ananta Institute Of Medical Sciences And Research Centre. *Corresponding Author

ABSTRACT

Esthesioneuroblastoma, or olfactory neuroblastoma, is a rare malignant neoplasm of the sinonasal tract originating from the olfactory neuroepithelium with neuroblastic differentiation. It most often presents in the superior nasal cavity. Since its initial description in 1924 by Berger and Luc, there have been reports of more than 1000 cases of esthesioneuroblastoma worldwide. Esthesioneuroblastoma is a locally aggressive neoplasm and metastasizes by both hematogenous and lymphatic routes.

KEYWORDS

olfactory neuroblastoma , rare tumour, PET CT

INTRODUCTION

Olfactory neuroblastoma, or esthesioneuroblastoma, is a tumor that grows in the nasal cavity. The nasal cavity has nerves and other tissue that are responsible for the sense of smell. This kind of tumor begins in the nasal cavity and can grow into the nearby eyes and brain. Olfactory neuroblastoma can also spread to other parts of the body, such as the neck, lungs, and bones.

BACKGROUND

Olfactory neuroblastoma (ONB) is an uncommon malignant neuroectodermal nasal tumor. It comprises about 2% of all sinonasal tract tumors with an incidence of approximately 0.4 per million population. Previously called esthesioneuroblastoma, olfactory placode tumor, esthesioneurocytoma, esthesioneuroepithelioma, and esthesioneuroma, these terms highlight the sensory (olfactory) and primitive neuroectodermal origins, although the use of these older terms is discouraged. ONB are thought to arise from the specialized sensory neuroepithelial (neuroectodermal) olfactory cells that are normally found in the upper part of the nasal cavity, including the superior nasal concha, the upper part of septum, the roof of nose, and the cribriform plate of ethmoid. Specifically, Jacobson's vomero-nasal organ, sphenopalatine ganglion, ectodermal olfactory placode, ganglion of Loci (nervus terminalis), autonomic ganglia of the nasal mucosa, and the olfactory neuroepithelium (cribriform plate and superomedial surface of the superior turbinate) are all sites of origination for this malignant neural crest derived neoplasm. It is also of note that these specialized olfactory neurons are probably the progenitors of neuroendocrine carcinomas of the sinonasal tract and so called "olfactory carcinoma." The olfactory epithelium contains three cell types, which can be histologically identified in the tumor: basal cells, olfactory neurosensory cells, and supporting sustentacular cells. The basal cells are the stem cell compartment, continuously replacing the neurosensory cells throughout adult life, both physiologically and as a response to injury.

CLINICAL FEATURES

ONB may occur at any age (2–94 years), but a bimodal age distribution in the 2nd and 6th decades of life are most common without a gender predilection. The tumors most commonly cause unilateral nasal obstruction (70%), and epistaxis (50%), while less common signs and symptoms include headaches, pain, excessive lacrimation, rhinorrhea, anosmia, and visual disturbances. Even though the tumor arises from the olfactory neuroepithelium, anosmia is not a common complaint (5%). Due to the non-specific nature of the initial presentation and slow growth of the tumors, patients often have a long history before diagnosis. There are isolated cases reports of ONB secreting vasopressin with resultant hypertension and hyponatremia.

CASE STUDY

A 47 year male patient presented with chief complaint of left sided nasal swelling, for nasal swelling , diagnostic nasal endoscpy (dne) biopsy was performed

After hpe reports, a series of other investigation has been done and are as follows

HPE- focal ulceration of mucosal epithelium with submucosal infiltrating broad interconnecting bands and nets. Neoplastic round cell having hyperchromatic nuclei, brisk mitosis seen, foci of necrosis seen s/o- **malignant round cell neoplasm**

IHC – MARKERS WERE POSITIVE S/O- **OLFACTORY NEUROBLASTOMA**

After this PT was operated

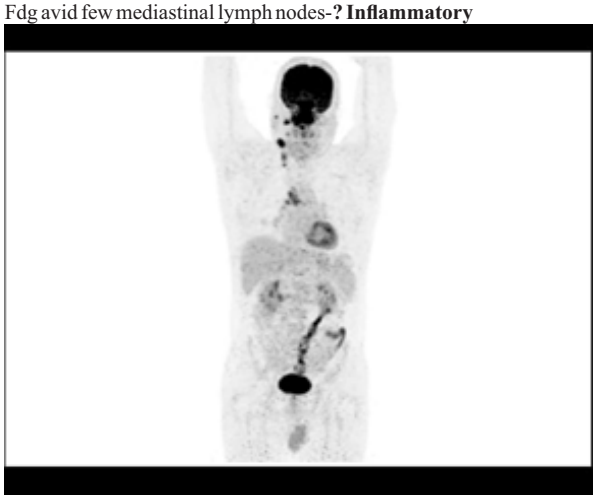
MRI brain-lobulated altered signal intensity lesion is seen along the walls of nasal cavity, b/l omu, medial aspect of maxillary sinus, b/l fsdp, frontal sinus, along the medial wall of b/l orbit with focal bulging with fat stranding on left side. Posteriorly the lesion is abutting the b/l ser with extension of soft tissue into the right sphenoid sinus. Superiorly the lesion is causing erosion of cribriform plate and floor of anterior cranial fossa and causing abutment of b/l basifrontal region with minimal perilesional edema. It is appearing heterogeneous isointense and measuring approx 47 * 38 * 61 mm in size s/o residual lesion with extension.

PET CT was done for disease staging
Suggestive of fdg avid heterogeneously enhancing soft tissue density lobulated mass lesion in the nasal cavity with lytic destrctive changes of nasal bone and maxillary bones. Superiorly, the lesion is infiltrating the b/l ethmoid sinuses, cribriform plate and extending into the anterior cranial fossa with involvement of b/l frontal lobes with perilesional edema. Anteriorly the lesion is involving the b/l frontal sinuses with erosion of adjacent frontal bone.laterally, the lesion is causing lytic destructive changes in the medial wall of the b/l orbits and extending into the b/l maxillary sinuses (right > left) note is made of mild intraorbital extraconal extension of the lesion bilaterally. The lesion is also infiltrating the overlying subcutaneous tissue.

Fdg avid multiple cervical, right intraparotid and right supraclavicular

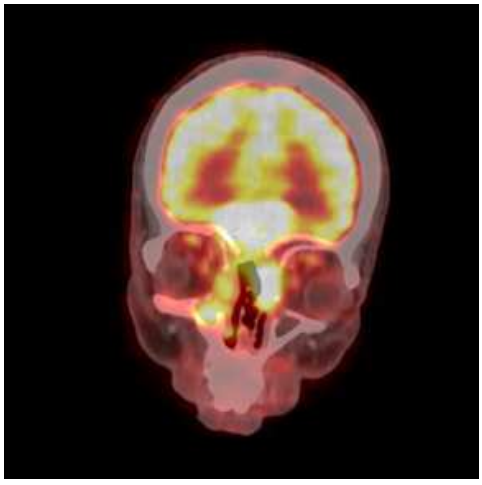
lymphnodes- **likely metastatic lymph nodes**

Fdg avid few mediastinal lymph nodes-**? Inflammatory**



MIP image shows physiological intense FDG uptake in the brain & heart and urinary activity in urinary bladder.

Sagittal image fused PET CT section show FDG avid (SUVmax-18.8) heterogeneously enhancing soft tissue density lobulated mass lesion is noted in the nasal cavity with lytic destructive changes of nasal bone. Superiorly, the lesion is infiltrating the cribriform plate and extending into the anterior cranial fossa with involvement of frontal lobe



Fused PET CT coronal image show FDG uptake (SUVmax- 18.8) heterogeneously enhancing soft tissue density lobulated mass lesion is noted in the nasal cavity with lytic destructive changes of nasal bone and maxillary bones. Superiorly, the lesion is infiltrating the bilateral ethmoid sinuses, cribriform plate and extending into the anterior cranial fossa with involvement of bilateral frontal lobes.

CONCLUSIONS

Extension of rare case report of newly diagnosed olfactory neuroblastoma s/o – extension as described earlier.

REFERENCES:

[1] https://www.cancer.gov/pediatric-adult-rare-tumor/rare-tumors/other-rare-tumors/olfactory-neuroblastoma.
[2] https://www.ncbi.nlm.nih.gov/books/NBK539694/.
[3] https://www.hopkinsmedicine.org/health/conditions-and-diseases/olfactory-neuroblastoma
[4] https://www.ncbi.nlm.nih.gov/pmc/articles/PMC2811627/