



“CASE REPORT OF OLFACTORY NEUROBLASTOMA.”
(Esthesioneuroblastoma)

Dr. Saketh Rao

Resident Doctor, Radiology Department, B.J. Medical college, Civil Hospital Ahmedabad, Gujarat, India

Dr. Zalak Panchal

Resident Doctor, Radiology Department, B.J. Medical college, Civil Hospital Ahmedabad, Gujarat, India

ABSTRACT

Esthesioneuroblastoma (ENB) is a rare tumor arising from the olfactory epithelium of the nasal vault which frequently invades the cranial base, cranial vault and orbit. ENB has a bimodal age distribution between 11 and 20 years and between 51 and 60 years. ENB accounts for approximately 1 to 5% of intranasal cancers. The diagnosis may be delayed for several months due to its slow growing nature. It may be misdiagnosed with other small round cell tumors. Here we describe a case of olfactory neuroblastoma in a 50-year-old female presented with left sided nasal blockage and episodes of epistaxis.

KEYWORDS : Esthesioneuroblastoma, Olfactory neuroblastoma, Malignant tumour of nasal cavity

INTRODUCTION:

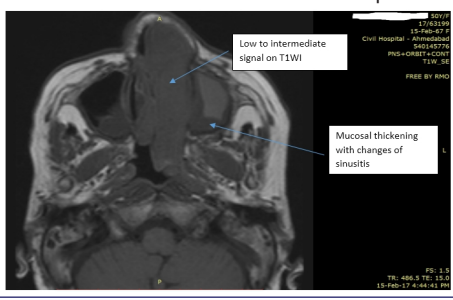
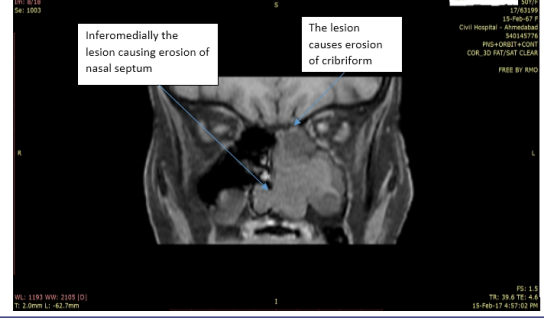
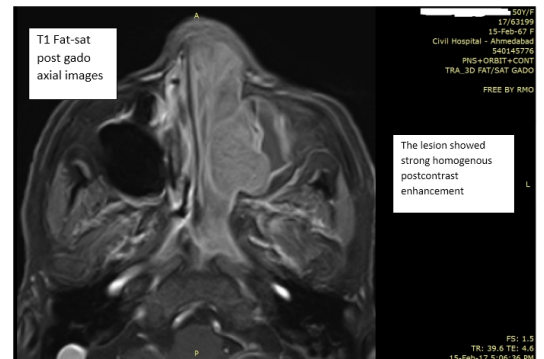
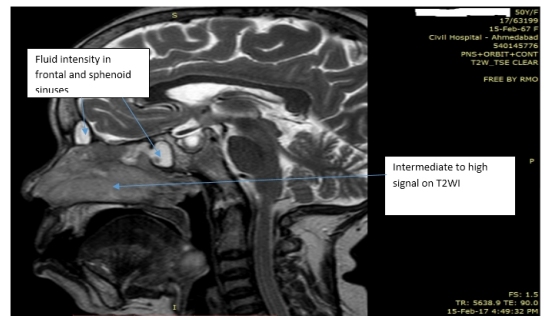
Esthesioneuroblastoma (olfactory neuroblastoma) (ENB) is a rare neuroepithelial tumor that arises from the olfactory epithelium in the cribriform plate or nasal cavity. First described in 1924 by Berger it has a histological pattern similar to that of sympathetic ganglia, retina, and adrenal medulla and only recently became recognized as a distinct pathologic entity probably as a result of immunohistochemistry and by means of electron microscopy techniques. ENB account for 1 to 5% of malignant neoplasm of the nasal cavity. The symptoms are related to sites and invasion of the tumor. The staging system based on tumor extension that was presented by Kadish et al.⁶ in 1976 has been widely accepted. The treatment of choice is a multidisciplinary craniofacial surgical resection that has improved considerably the prognosis.

CASE REPORT

A 50-year-old female presented to the Otorhinolaryngology department with a mass in the left nasal cavity. Her chief complaint was persistent left sided nasal blockage and intermittent epistaxis over the past 1 year associated with headache. A nasal exam was performed using rigid endoscopy, which revealed a large mass, medial to the middle turbinate in the Left nasal cavity. The mass extended through the choana, extending into the nasopharynx clinically. It was attached superiorly with no medial or lateral attachment identified. The lesion was pink and lobulated with a rubbery, non-friable texture.

IMAGING FINDINGS:

MR evaluation of nasal cavity, PNS and orbits revealed a mass lesion almost completely filling the left nasal cavity. The lesion was hypo-isointense on T1WI and hyper-isointense on T2WI compared to gray matter with strong homogenous post contrast enhancement. The lesion extends anteriorly upto the external nares. Posteriorly, the lesion bulges into nasopharynx with effacement of torus tubaris. The lesion causes focal erosion of anteroinferior part of nasal septum. Laterally, the lesion extends into left maxillary antrum with blockage and expansion of osteomeatal complex. The lesion causes focal erosion of left lamina papyracea with extension into extraconal compartment of left orbit. Superiorly, lesion causes erosion of left cribriform plate. However, there was no evidence of intracranial extension or adjacent dural enhancement. Posterosuperiorly, the lesion causes widening of left sphenothmoidal recess with extension into left sphenoid sinus.



DISCUSSION:

Olfactory neuroblastoma (ONB) is an uncommon malignant neuroectodermal sino-nasal tumour.^{1,2,4} ONBs are tumors of neural crest origin that arise in the olfactory mucosa.^{1,2,4} These tumors arise almost exclusively from the highly specialized sensory olfactory neuroepithelium normally encountered within the superior nasal vault, including the superior nasal concha, superior septum, roof of nose, and the cribriform plate of ethmoid.^{1,2,4} ONBs have a wide age range with a bimodal peak in the second and sixth decades of life. ONB is usually not a diagnostic consideration in children.^{1,2,4} The most common symptoms are nasal obstruction and epistaxis. ONBs with intracranial extension may present with headache, proptosis, and cranial neuropathies.^{1,2,4}

IMAGING: The typical finding in ENB is a superior nasal cavity mass at the cribriform plate. A “dumbbell” shape-the upper portion in the anterior cranial fossa and the lower portion in the nose with the narrowest aspect at the cribriform plate - is seen with large masses.¹

CT finding: Bone CT shows bone remodelling mixed with bone destruction, especially of the cribriform plate. Speckled intratumoral calcification is unusual. A homogeneously enhancing mass on CECT is typical.^{1,2,4,5}

MR findings: ENB is hypo- to isointense compared with brain on T1WI and iso- to hyperintense on T2WI. Areas of cystic degeneration and intratumoral hemorrhage are common. Some large ENBs that extend intracranially have benign nonneoplastic tumor-associated cysts around their superior and lateral margins at the tumor-brain interface. ENBs generally enhance strongly and relatively uniformly.^{1,2,4,5}

ENB spread to cervical lymph nodes is common, typically spreading first to level II nodes, with frequent involvement of level I, level III, and retropharyngeal nodal groups at later stages. Local regional and distant metastasis occurs in upto 38% of patients. Late recurrences or metastatic disease can occur upto two decades after initial presentation.

The Kadish classification^{1,2,4,5} is used for staging and recognizes three stages:

Stage A, tumours that are localized to the nasal cavity;
stage B, nasal cavity and paranasal sinuses; and
stage C, extension beyond the sinonasal cavities, including intracranial and involvement.

Management: Craniofacial resection should be performed as the initial procedure in all patients.^{1,3} Adjuvant chemotherapy is recommended for patients with high grade tumors or positive surgical resection margins.^{3,5} Others recommend adjuvant radiotherapy for all patients because combined surgery and irradiation resulted in 92% of disease free status.^{3,5}

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