

Esthesioneuroblastoma – A Rare Case Report

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Abstract: Esthesioneuroblastoma also called as Olfactory Neuroblastoma (ONB) arises from the olfactory placode in the olfactory area of the nasal cavity. The age incidence ranges from 3years to 79years. It usually presents with nasal symptoms like obstructed nasal breathing and epistaxis, The diagnosis may be delayed for several months due to its slow growing nature. It may be misdiagnosed with other small round cell tumors. Its biological activity ranges from indolent growth to local recurrence and rapid widespread metastasis.

Keywords: Esthesioneuroblastoma, olfactory neuroblastoma, nasal cavity, unilateral nasal mass.

INTRODUCTION

Olfactory neuroblastoma is a rare malignant tumor believed to originate in olfactory cells most commonly presented as unilateral polypoid nasal masses often causing nasal obstruction and epistaxis. The olfactory cells, located in the upper rear of the inside of the nose, are responsible for the sense of smell. It has a predilection for invading surrounding regions, such as the paranasal sinuses, orbits skull base and can cause loss of smell, taste, and vision, as well as facial disfigurement in advanced cases [1]. It is a rare disease that was first described in 1924 in the French medical literature and since that time, this tumor continues to be uncommon, incidence being less than 5% of all sinonasal malignancies. Olfactory neuroblastoma is a neuroendocrine tumour capable of causing paraneoplastic syndrome by secreting peptides [2]. ONB often responds well to radiation therapy, but the tumor has a high tendency to recur after excision. Olfactory neuroblastoma is considered distinct from other neuroblastomas, because it does not originate in the sympathetic nervous system [3]. Here we present a case of ONB in a 72 years male patient presented with left side nasal obstruction .

CASE REPORT

A 72 years male patient presented with a 15 days history of left nasal obstruction due to painful swelling in left nostril, obstruction was persistent, non progressive and not relieved with medication, later on the patient developed epistaxis gradually. On palpation, the mass felt as 2x2cm sized single, soft, tender swelling with local rise of temperature, pushing the left ala laterally. He had no history of headache, diplopia, or sensory changes. Physical examination revealed a mass in the left nasal cavity arising from anterolateral aspect of left nostril. Examination of the right nasal cavity was normal, as was the remainder of the head and neck examination. Computed tomography (CT) with contrast showed a heterogeneously enhancing oval shaped mass within the left nasal cavity arising from anterolateral and nasolabial aspect. There is evidence of erosion or destruction of adjacent bone due to mass (Figure 1a & b). Radiologically it was diagnosed as maxillary carcinoma or nasopharyngeal carcinoma. Patient was admitted in E.N.T. department and biopsy was done under all aseptic precautions. Grossly we received a single grayish white soft tissue bit measuring 1 cm x 1cm, which was all embedded and it was diagnosed as Grade 3 ONB microscopically. Patient was referred to higher centre for further management.

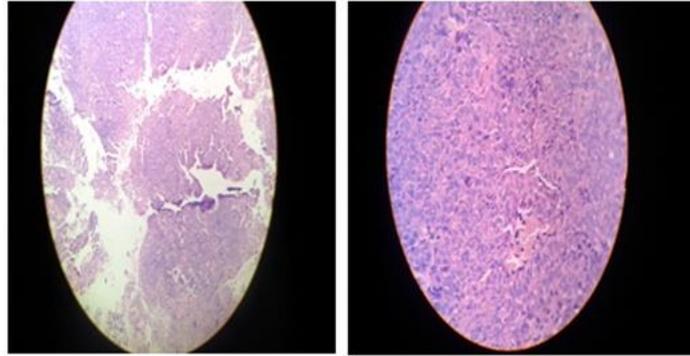


Fig-1: Section showing tumour tissue arranged in rosettes with pleomorphism with no mitotic activity. Grade 3 ONB. [Fig a, x4 H&E. Fig b, x40 H&E].

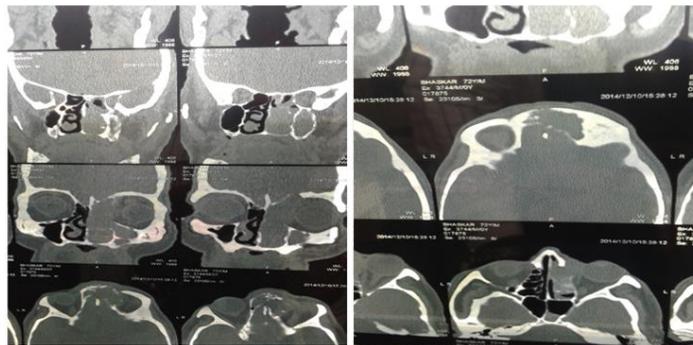


Fig-2: CT scan showing tumour mass eroding the frontal and sphenoidal sinuses and extending intracranially

DISCUSSION

ONB's are solid nasal cavity masses that can erode into nearby osseous structures such as the orbital plate of the ethmoid bone, cribriform plate, and fovea ethmoidalis. The most common finding on physical examination is a unilateral, broad-based, polypoid, friable, and gray-to-red nasal mass. The two most common symptoms presented were unilateral nasal obstruction and epistaxis [1,4,5]. The disease has a bimodal age distribution; it peaks first between 10 and 20 years and then between 60 and 70 years [6]. The distribution is not affected by sex race. The symptoms of ONB can be classified into nasal, neurologic, oral, facial, cervical, and ophthalmologic and are as follows:

- Nasal - Obstruction (70%), epistaxis (46%), discharge, unilateral polyp, anosmia
- Neurologic - Headache, nausea
- Oral (rare) - Mobile tooth, nonhealing tooth-extraction site, ill-fitting dental prostheses, ulceration
- Facial (rare) - Swelling, pain, anesthesia, trismus
- Cervical - Mass
- Ophthalmologic - proptosis, extraocular movement paralysis, and blindness,

The average delay between the appearance of the initial symptom and the diagnosis is 6 months, but diagnosis is delayed for years in some cases.

Nasal examination, particularly if aided by endoscopy, reveals a reddish-gray tumor arising in the upper nasal fossa, which bleeds easily with instrumentation. Although this aspect is strictly

different from the white, glistening appearance of benign nasal polyps, little differentiates ONB from other nasal malignancies. Late findings may include signs related to extensive disease such as orbital, cranial, and cervical involvement. No clear etiologic agent or exposure has been documented in humans; however, a single case of occupational exposure has been reported in a woodworker [7]. Esthesioneuroblastoma can be consistently induced by nitrosamine compounds in rodents.

Olfactory neuroblastoma is an uncommon 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. This tumor has a broad histological spectrum and olfactory neuroblastoma can be confused with other small round cell tumors and undifferentiated carcinomas.

On microscopy the tumor is composed of small round to oval cells arranged in irregular circumscribed nests of cells separated by by stroma. The cells have high N/C ratio, indistinct cell borders, round vesicular nuclei with clumped chromatin and scanty cytoplasm. Pseudorosettes (Homer Wright Rosette) characterized by grouping of cells in circumscribed fashion but without defining basement membrane are seen (Figure 2a & b). We diagnosed it as Grade 3 ONB .Patient was referred to higher centre for further management.

There is no gender predilection, 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.

The usual clinical symptoms are unilateral nasal obstruction, epistaxis, anosmia, headache, pain and ocular disturbances. 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.

ONB 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. Grade III & IV have little or no neuro fibrillary background with few or no rosette formations making morphological diagnosis difficult. [Table 1]

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. With confounding histomorphology, the diagnosis will be difficult which requires immunohistochemistry for confirmation. ONB shows variable positivity for Cyto keratin, synaptophysin and chromogranin.

The main mode of treatment is complete craniofacial resection of the tumor followed by radiotherapy. 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%). [Table 2]

To Conclude ONB 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.

Table-1: Showing Hyams' histologic grading system for ONB.

Histologic features	Grade 1	Grade 2	Grade 3	Grade 4
Architecture	Lobular	Lobular	+/- Lobular	+/- Lobular
Pleomorphism	Absent to slight	present	prominent	marked
NF matrix	Prominent	present	May be present	absent
Rosettes	present	Present	May be present	May be present
Mitoses	Absent	Present	Prominent	marked
Necrosis	Absent	Absent	Present	Prominent
Calcification	Variable	Variable	Absent	absent
Glands	May be present	May be present	May be present	May be present

NF: Neurofibrillary

Table-2 : showing clinical staging for ONB:

Stage	Extent of tumour	5-year survival (%)
A	Tumour confined to nasal cavity	75- 91
B	Tumour involves the nasal cavity plus one or more paranasal sinuses	68 - 71
C	Extension of tumour beyond the sinonasal cavities.	41 - 47

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