World Journal Of ENT & Head-Neck Surgery
(WJEHNS)

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ESTHESIONEUROBLASTOMA: A RARE PRESENTATION

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Abstract:

Introduction- Esthesioneuroblstoma, also known as Olfactory Neuroblastoma, arise from the basal cells within the olfactory neuroepithelium. It accounts for <5% of the sinonasal malignancies. The slow growing nature of the tumor, often delays it's diagnosis. The nature of invasion into surrounding structures including Dura and middle cranial fossa, increases it's morbidity.

Clinical Presentation- A 26 years lady presented with a gradually progressive swelling in the left cheek, which was initially painless but later it became painful and was associated with loosening of teeth of upper jaw. On examination, there was a hard ,non tender and non pulsatile swelling, extending from left nasolabial fold to the zygomatic bone and from the orbit till the angle of mouth. It was fixed to the overlying skin, and the margins were not palpable on any side. Radiological studies (CT and MRI) showed a heterogenous SOL, involving the left side of the cheek, with extensive spread to the surrounding structures including the adjacent maxillary antrum and left side of the nasal canal, with erosion of the adjacent bony alveolus and floor of left maxillary antrum. FNAC from the swelling was suggestive of poorly differentiated malignant neoplasm. Biopsy taken from the nasal mass was diagnostic of neuroblastoma.

Surgical Management- The patient underwent left sided extended maxillectomy by Weber Ferguson's approach with a Dieffenbach's extension under general anaesthesia. Later the patient was sent for chemoradiation. The patient was followed up and after 6 months, an artificial denture was given to the patient after completion of her radiotherapy.

Discussion- Esthesioneuroblastoma is a very rare malignancy of the olfactory neuroepithelium. Certain histological heatures helps diagnosing this tumor. Confirmatory diagnosis requires examination under electron microscope. The staging system proposed by Kadish et al. modified by Morita is now widely accepted. This staging system is predictive of disease-related mortality.

Introduction-

Esthesioneuroblstoma, known as Olfactory Neuroblastoma, arise from the basal cells within the olfactory neuroepithelium. It consists of <5% of the sinonasal malignancies, extremely rare . It was originally described in 1924 by Berger et al[1] in the french literature 'esthesioneuroepitheliome olfactif' with the highest series of cases being reported by Jackson et al in 1984 while highest number of cases was reviewed by Skolnic et al[3] in 1966. Olfactory neuroblastoma exhibits a wide range of morphological diversity ranging an indolent growth to highly aggressive neoplasm with propensity of rapid metastasis[4] It has got a bimodal age distribution of 20 and 50 years and more common in females than males.Though it presents with nasal symptoms like obstruction and intermittent epistaxis,the diagnosis is often delayed due to its slow growing nature. The disease extends its morbidity via direct extension and invasion into the surrounding structure (25% of the cases invades dura and anterior cranialfossa). Radiographic features show a contrast enhancing lesion with expansile and destructive growth with expansile and destructive growth patterns. Genetic studies have shown involvement Hedgehog pathway, MYC and KDR genes implicated for it.

CLINICAL PRESENTATION:

A 26 years female presented with a swelling in the left cheek , which was progressively increasing in size. After a month, she started developing pain over the region and loosening of tooth in the left upper jaw.

It was also associated with a nasal blockage. Her symptoms have been increasing ever since. She also subsequently developed difficulty in chewing and a loss of appetite.

On examination, she had developed trismus. There was a hard ,non tender and non pulsatile swelling measuring about 8 x 9 cm in antero posterior and and craniocaudal directions. It extended from left nasolabial fold to the zygomatic bone and from the orbit till the angle of mouth. It was fixed to the overlying skin, and the margins were not palpable on any side
There were no palpable lymph nodes.

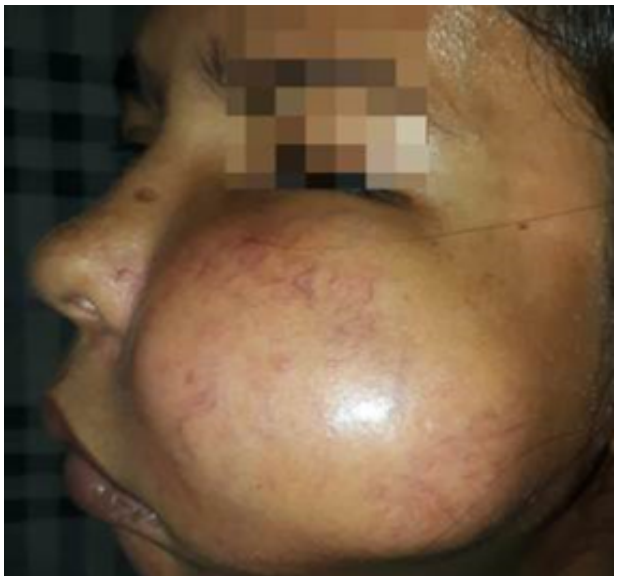

Figure 1

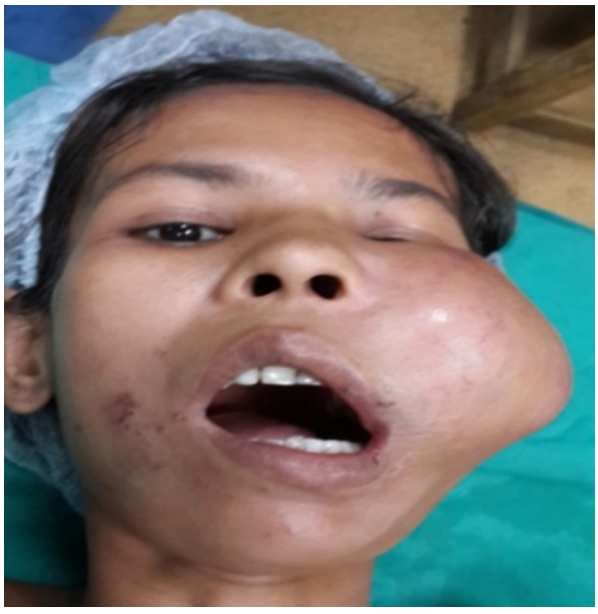

Figure 2

Figure 1 and Figure 2 showing clinical presentation of the patient.

Radiology: CT scan reveals opacity in the sinuses, more marked on the left side and a left upper alveolobuccal mass with erosion of the adjacent bony alveolus and floor of left maxillary antrum.

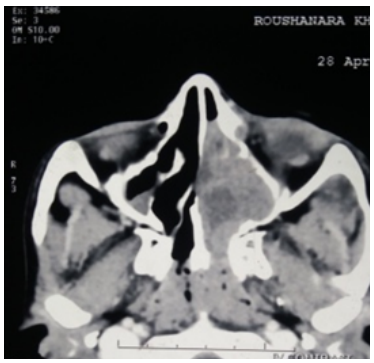

Figure 3

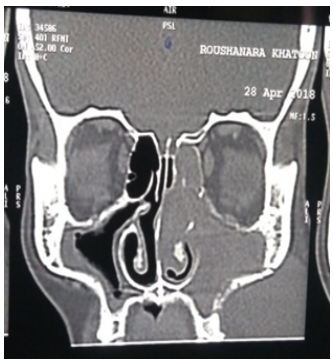

Figure 4

Figure 3 and 4 showing CT scan findings.

MRI (Plain and contrast) are suggestive of large, heterogenous, contrast enhancing soft tissue mass seen involving the left side of the cheek, with extensive spread to the surrounding structures including the adjacent maxillary antrum and left side of the nasal canal.

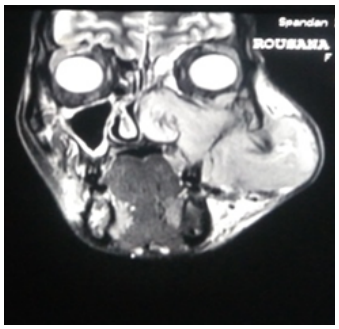

Figure 5

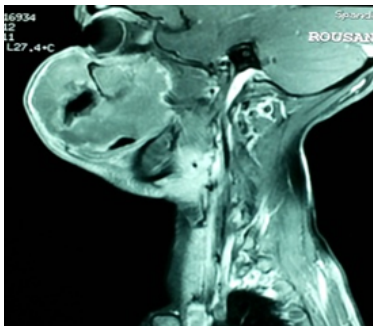

Figure 6

Figure 5 and 6 showing MRI Findings.

Histopathology- FNAC from the swelling was suggestive of poorly differentiated malignant neoplasm. Biopsy taken from the nasal mass was diagnostic of neuroblastoma. The histopathology was confirmed from two different centres and the report was similar from both

SURGICAL PLAN:

The patient was planned for a total left sided extended maxillectomy under general anaesthesia.
Weber Ferguson's incision with a Dieffenbach's extension was given and cheek flap elevated.
Inferior orbital nerve divided. Frontal process of maxilla and the lacrimal bone,floor of the orbit, zygomatic process and the palatae were divided and osteotomy to separate maxillary tuberosity from the pterygoid plates

Involved parts of pterygoid and masseter were excised. A part of the skin with subcutaneous involvement was also excised.

Bleeding from the internal maxillary artery was controlled by ligation and oozing from the pterygoid plexus by packing.

Dacrocystorhinostomy was performed, where the sac was identified and marsupialised.

The skin flap was closed in layers after putting an occluder in the guttaparcha.

Following extended maxillectomy, the patient was referred to the department of Radiotherapy for further treatment.

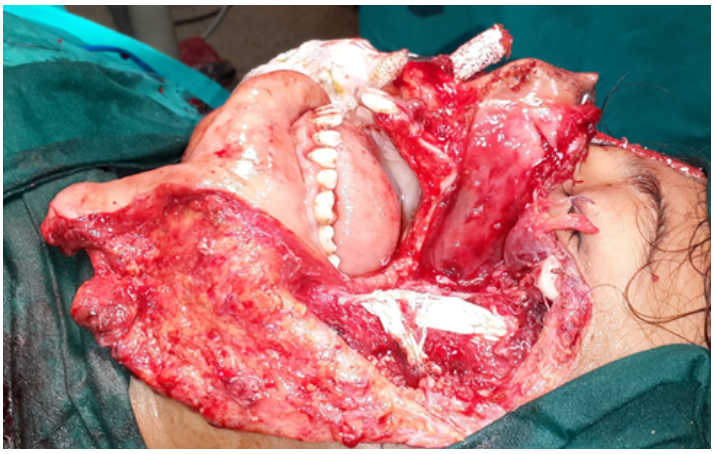

Figure 7 :Intraoperative picture of the patient after removal of the mass.

Figure 8

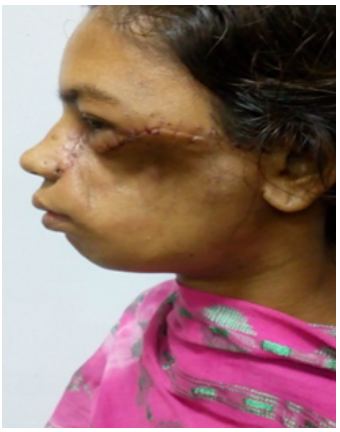

Figure 9

Figure 8 and Figure 9 showing Post operative outcome.

DISCUSSION:

Esthesioneuroblastoma is a very rare malignancy of the neuroepithelium. Embryologically, the olfactory nerves develop from the olfactory placode present in the fetal olfactory mucosa[5,6]. Histologically, there are a number of criterias that help in its diagnosis: neuroepithelial cells arranged in the classic pseudorosette pattern; fibrillar intracellular background; marked microvascularity; and round or fusiform cells approaching the size of lymphocytes with poorly defined, almost nonexistent cytoplasm[7,8,9]

Correct diagnosis often requires confirmatory examination with electron microscopy for the detection of neurosecretory granules. More recently immunohistochemical methods for detection of neuronspecific enolase (NSE) and S-100 protein with negative epithelial, muscle, and lymphoid antigens allowed further confirmation of ENB7, [10].

The staging system presented by Kadish et al.[11] in 1976 based on tumor extension, and modified by Morita[12] has been widely accepted. This staging system is predictive of disease-related mortality. , as our own patient.

It is a slow growing tumor, and patient generally presents late due to non specific symptoms and very rarely presents with a facial swelling.

Our present presented with the facial swelling as her chief complain with other non specific complains of frequent epistaxis and nasal obstruction.

Involvement of the maxilla before involvement of the orbits or the anterior cranial fossa is rare.

An extended maxillectomy was done for the patient. A skin grafting was avoided due to uncertainity of spread. The patient was followed up and after 6 months, an artificial denture was given to the patient after completion of her radiotherapy.

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