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A CASE REPORT OF OLFACTORY NEUROBLASTOMA

Otorhinolaryngology

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ABSTRACT

Olfactory neuroblastoma is a rare tumor of the olfactory neuroepithelium. That is characterized by slow growth and local recurrence. We are presenting a case report of a 52 year old male patient with left side nasal mass which was diagnosed to have olfactory neuroblastoma. This patient was treated with total endoscopic excision of the mass. And radiotherapy was given. Combination of surgery and radiotherapy with or without chemotherapy is considered to be the standard treatment.

KEYWORDS

INTRODUCTION:

Olfactory neuroblastoma is an uncommon malignant neuroectodermal tumour. It is also know as esthesioneuroblastoma. It arises from specialized sensory neuroepithelial (Neuroectodermal) olfactory cells that are normally found in upper part of nasal cavity including superior turbinate, upper part of septum, cribriform plate of ethmoid. It represents 5 to 10% of all sinonasal malignancy(1). It has Bimodal age distribution ,peak incidence in the second and sixth decade. It has no sex predilection. Diagnosis confirmed on histological examination. Recurrence is common in high grade tumours.

CASE – REPORT:

A 52 year old patient presented to ENT department of our hospital with chief complains of unilateral(left) nasal blockage and unilateral(left) nasal discharge with occasional nasal bleed since 1.5 years. He had no complains of visual disturbance, facial swelling/ facial pain.

On Clinical Examination:

- Anterior rhinoscopy revealed mass in left nasal cavity.
- On endoscopic examination, a large greyish white polypoidal mass seen medial to middle turbinate in left nasal cavity, that doesn't bleed on touch. Right side nasal cavity was normal
- No visible/palpable neck nodes

Radiological Investigation:

- CT Scan of paranasal sinuses (plain+contrast) showed Hyperdense soft tissue density lesion involving left nasal cavity & all sinuses on left side and erosion of adjacent bony wall of anterior ethmoid sinus. .
- The lesion within left nasal cavity and ethmoid sinus shows intense post contrast enhancement. Bilateral cribriform plate appear normal. No evidence of intracranial or intraorbital extension. All routine blood investigation were carried out & they were within normal limits.

Surgical Management:

- Under Endoscopic vision: greyish white mass was visualized in left side nasal cavity. Mass was soft to firm in consistency , completely removed from nasal cavity and maxillary osteum widened and sinus cleared of disease Similarly ethmoid , frontal and sphenoid sinuses made free of disease. After removal of mass, circumferential area was analyzed, no active leak from cribriform area was present. Hemostasis was achieved.

And mass sent for further histopathological examination.

- On **Histopathological Examination** of the specimen showed abundant vascularised fibrous stroma. The small round tumour cells with small uniform hyperchromatic nuclei, salt and paper nuclear chromatin suggestive of olfactory neuroblastoma grade ll according to hyam's grading system.

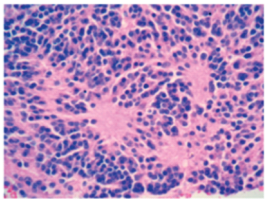

Figure : 1 Microscopic appearance of grade-ll olfactory neuroblastoma

- Patient was referred to radiotherapy department for further management.
- On follow up after 6 months, patient was having no complains. and Operated site was healthy, without any evidence of recurrence on nasal endoscopy.

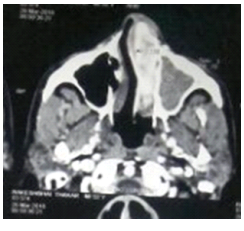

Figure 2: axial section showing hyperdense Mass in left nasal cavity and maxillary sinus

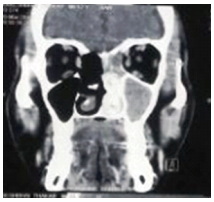

Figure 3: coronal section showing nasal mass in left nasal cavity , frontal and maxillary sinuses

DISCUSSION:

- Olfactory Neuroblastoma is an uncommon Malignant neuroectodermal sinonasal tumour, first described by berger in 1924(2)
- It arise from highly specialized sensory olfactory neuroepithelium normally present within superior nasal concha, nasal septum, roof of nose, cribriform plate of ethmoid.
- Primary clinical features include nasal obstruction (93%), bleeding (53%), rhinorrhoea, anosmia, visual disturbances, facial pain(1)
- Clinically it presents as unilateral soft, polypoidal highly vascular submucosal mass in superior nasal cavity extending to adjacent paranasal sinuses, orbit is often involved. Most of the patients present at a locally advanced stage.
- Differential diagnosis includes small round cell tumours like lymphoma , rhabdomyosarcoma, undifferentiated carcinoma andmalignant melanoma.
- CT Scan assess local invasion in to adjacent bony structures like cribriform plate, erosion of lamina papyracea, fovea ethmoidalis and MR Scan assess extent of soft tissue invasion like anterior cranial cavity and orbit. The typical radiographic findings will show 'dumbbell-shaped' occupying the superior nasal cavity and ethmoid sinus , extending through the cribriform plate and into the anterior cranial fossa with the 'waist' at the level of cribriform plate.

Diagnosis is done on histopathological examination: the hallmark is formation of rosettes, pseudorosettes separated by a fibrovascular stroma.

HYAMS HISTOLOGICAL GRADING SYSTEM (1)

Histological features	Grade I	Gradell	Grade lll	Grade lV
Architecture	Lobular	Lobular	May be lobular	May be lobular
Pleomorphism	Absent or slight	Present	Prominent	Marked
Neurofibrillary	Prominent	Present	May be present	Absent
Rosettes	Homer–Wright (pseudo)rosette	Homer–Wright (pseudo)rosette	Flexner–Wintersteiner (true) rosettes	Flexner–Wintersteiner (true) rosettes
Mitoses	Absent	Present	Prominent	Marked
Necrosis	Absent	Absent	Present	Prominent
Glands	May be present	May be present	May be present	May be present
Calcification	Variable	Variable	Absent	Absent

Complete surgical eradication often followed by Radiotherapy is modality of treatment. Palliative chemotherapy may be reserved for advanced , surgically inoperable tumors or for disseminated disease.
A craniofacial resection has been suggested for all patients with frontal cranial base involvement.

The study by Benfari et al indicated that Radiotherapy should be applied to all patients, with exception of cases with tumor limited to the cribriform plate without bony destruction.(3)

In a study by Feng et al, 24 patients with olfactory neuroblastoma were treated endoscopically. In that study, 19 patients presented with newly diagnosed Olfactory neuroblastoma and 5 with recurrence. Twenty patients (83.3%) received postoperative radiotherapy and 7 patients (29.2%) also received chemotherapy. One patient developed a CSF leak. Four (16.6%) patients, all with Kadish stage C disease, died of tumor recurrence(4).

In a recent systematic review and meta-analysis, Fu et al. evaluated 36 studies containing 609 patients comparing outcomes for open versus endoscopic resection of Olfactory neuroblastoma. In this analysis, endoscopic surgery was defined as a purely endoscopic approach; open surgery included transcranial, transfacial, and endoscope-assisted procedures.In this study, patients undergoing an endoscopic resection had significantly lower rates of recurrence , cause-specific mortality and overall mortality compared to open surgery. (5)

Prognosis is largely dependent upon the inter relationship of tumor stage and grade. The presence of cervical lymphnode metastasis and distant metastasis suggest poor prognosis.

Kadish stages of Olfactory Neuroblastoma, modified by Moritais (1)

Stage	Features
A	Disease confined to the nasal cavity
B	Disease confined to th e nasal cavity and paranasal sinuses
C	Disease beyond the nasal cavity and paranasal sinuses, including involvement of the cribriform plate, base of skull, orbit or intracranial cavity
D	With metastasis to cervical lymph nodes or distant metastases

In our case, by histo pathological and radiological examination, patient had Stage B and grade ll Olfactory neuro blastoma.

CONCLUSION:

- Complete surgical eradiation of tumor followed by full course of radiotherapy is considered the treatment modality of choice for most olfactory neuro blastoma.
- Early Diagnosis and prompt treatment is required for better prognosis of disease.

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