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OLFACTORY NEUROBLASTOMA. "RECENT TRENDS AND MANAGEMENT"

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ABSTRACT

OBJECTIVE- To study the clinical and radiological spectrum of Esthesioneuroblastoma and evaluate the results of surgical treatment.
STUDY DESIGN- Retrospective study of 10 patients.
SETTING- Tertiary centre
METHOD- Epidemiological parameters, clinical presentation, staging and treatment modalities for 10 patients who were diagnosed as esthesioneuroblastoma were studied and analyzed over a period of 2014 to 2017 at Department of ENT and Head & Neck surgery, B.J. Medical college, Ahmedabad
RESULT- Out of 10 patients, 7 (70%) were female being more common in comparison to 3 (30%) male, 9 (90%) patients came with complaint of nasal mass and nasal obstruction and all 10 patients underwent endoscopic resection with or with out chemoradiotherapy.
CONCLUSION- The result of endoscopic resection followed by radiation have been reported to be comparable to those of craniofacial resection.

KEYWORDS

olfactory neuroblastoma, craniofacial resection surgery, radiotherapy.

INTRODUCTION:

Olfactory neuroblastoma (ONB) is an uncommon malignant neuroectodermal sinonasal tumour. It accounts for approximately 6% of cancer cases in the nasal cavity and in the paranasal sinuses. (1) At present, two terms are well-established in the literature: esthesioneuroblastoma and olfactory neuroblastoma. (1) ONBs are tumors of neural crest origin that arise in the olfactory mucosa. 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. 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)]

OBJECTIVE:

Objective is to study the clinical and radiological spectrum of Esthesioneuroblastoma and to evaluate the result of surgical treatment and to review the literature in management of Esthesioneuroblastoma

METHOD:

The collection and use of human materials for the present study were approved by the Ethics Committee on Human Subjects of each institute, and informed consent was obtained from parents of all patients.

A retrospective study of 10 patients who were diagnosed as esthesioneuroblastoma were studied and analyzed for epidemiological parameters, clinical presentation, staging and treatment modalities over a period of 2013 to 2017 at who were referred to Department of ENT and Head & Neck surgery, B.J. Medical college, Ahmedabad.

Treatment modalities adopted were surgery alone or surgery supplemented with radiation. Patients having cervical metastasis were not included in the study.

Staging was done according to Kadish classification.

STAGES	
A	Tumours that are localized to the nasal cavity
B	Nasal cavity and paranasal sinuses
C	Extension beyond the sinonasal cavities, including intracranial involvement
D	Metastasis to cervical node or distant sites

RESULT:

10 patients were taken in the study out of which 3 were males and 7 were females. Age at the time of the diagnosis ranged from 45 to 72 years. 9/10 patients had complaint of nasal mass and nasal obstruction, being the most common symptoms. 5/10 patient had epixtasis, 2/10 headache and diminish vision. According to kadish classification, 7 patients presented at stage B, while 2 patient presented at stage C and 1 patient with stage A. All 10 patients underwent endoscopic resection of tumor with / without radiotherapy. Post operatively, 3 patients had no complaints, while 5 patients had complaint of sinusitis and 2 patient had on and off episode of epistaxis.

DISCUSSION AND REVIEW OF LITERATURE:

Olfactory neuroblastoma (ONB) is an uncommon malignant neuroectodermal sino nasal tumour. It accounts for approximately 6% of cancer cases in the nasal cavity and in the paranasal sinuses. 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.

The most common symptoms are nasal obstruction (70%) and epistaxis(50%). ONBs with intracranial extension may present with headache, proptosis, and cranial neuropathies. ONB 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.

Differential diagnosis: Nasal and paranasal squamous cell carcinoma, Sinonasal polyposis, Chaonal polp, Juvenile angiofibroma, Neuroendocrine carcinoma, Embryonal rhabdomysarcoma, Undifferentiated sinonasal carcinoma, Ewing's sarcoma.

HRCT and MRI can be used as complimentary investigation to precisely delineate the involvement of the cribriform plate, anterior cranial fossa and retro maxillary spaces.

The typical appearance of an olfactory neuroblastoma on microscopic examination includes the presence of characteristic cells separated into nests or compartments by fibrovasclar septae, neurofibrillary intracellular matrices and rosette formation. Immunohistochemistry can lead to a definitive diagnosis. This tumor is positive for neuroendocrine markers, chromogranin, synaptophysin, NSE.

Radical surgical resection with disease-free margin is a strong predictor of tumor freedom and prevention of significant delay of both local and distal recurrence.[(11)] The use of craniofacial approaches led to over a two-fold improvement in disease-free survival.[(12)] It became the gold standard in the treatment of esthesioneuroblastoma [(10) (11)]. It has increased progression-free-survival from 37.5% to 82% in comparison to extracranial resection.[(12)] Biller-et-al recommended craniofacial

resection of all tumours, regardless of whether they invade the anterior cranial fossa or are confined to nasal roof. [7] This is a combined transcranial and transfacial approach, typically including a frontal craniotomy, and lateral rhinotomy or midfacial degloving & resection of the dura over the cribriform plate, olfactory bulb, the entire ethmoid labyrinth, medial maxillae, upper septum and the anterior and posterior walls of the frontal sinuses.

Craniofacial resection, combined with radiotherapy has resulted in a significant improved 5 year survival rate.[8] Several authors still defend en bloc craniofacial resection as the treatment of choice for tumors in general.[3,4]

It provides safer free margins, consequently fewer recurrences, more reliable reconstruction of the skull base.

Limitations are it is surgically traumatic, healthy structures are resected to provide access to the surgical field, the duration of surgery is longer, intensive postoperative care is required, Cerebrospinal fluid leaks, frontal abscesses, pneumocephalus, hydrocephalus, intracranial hemorrhage, subdural hematoma and hygroma, mucoceles, diabetes insipidus, and amaurosis due to thromboembolic disease [5,6]

Yuen, et al. first described the use of an endoscopic approach in the treatment of esthesioneuroblastoma in 1997 [3].Since then, multiple studies have not only reproduced its feasibility but, in some cases, demonstrated its superiority [2]. Endoscopic approaches can either be classified as purely endoscopic or endoscopic-assisted craniofacial resections.

Several series, including our own, show excellent results with endoscopic-assisted approaches, including clear surgical margins and long-term disease-free and overall survival with minimal adverse events [4]. In some cases, endoscopic-only approaches did not compromise the extent of resection in even Kadish C disease [7], though many studies did not produce the same results [8].This finding was comparable to our results, where our patients undergoing endoscopic-only approaches had fewer complications.

It avoids retraction of frontal lobes, it avoids esthetic and functional loss because of transfacial approaches, it allows difficult areas to be visualized, it reduces the recovery period and hospital stay of patients.
Stereotaxic radiotherapy is indicated for patients where surgical resection was incomplete or residual disease is present.[8] Its use alone is not recommended; survival is increased when combined therapy is used.[10]

CONCLUSION:

Esthesioneuroblastoma requires aggressive surgical resection and radiation therapy. Complete surgical resection remains the expectation of the surgical procedure chosen and if not possible with endoscopic techniques should be considered a contraindication to using this approach .Endoscopic techniques seem to be safe approaches for the treatment of ENBs followed by post RT/CT. However, they are intended for selected patients and require extensive experience in base of the skull surgery. The result of endoscopic resection followed by radiation have been reported to be comparable to those of craniofacial resection.

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