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Case Study

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CASE REPORT: ESTHESIONEUROBLASTOMA

Dr. Tarun Nanda[1], Dr. Prithak Madan[2]* and Dr. Ankur Sharma[3]

[1]Post Graduate Resident, Department of Radiation Oncology, Ravindra Nath Tagore Medical College, Udaipur.
[2]Post Graduate Resident, Department of Radiation Oncology, Mahatma Gandhi Memorial Medical College, Indore.
[3]Radiation Oncologist, Department of Radiation Oncology, Ravindra Nath Tagore Medical College, Udaipur.

***Corresponding Author: Dr. Prithak Madan**

Post Graduate Resident, Department of Radiation Oncology, Mahatma Gandhi Memorial Medical College, Indore.

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SUMMARY/ABSTRACT

This report presents a case of esthesioneuroblastoma (ENB) in a 12-year-old male, initially misdiagnosed as necrotizing aspergillosis, highlighting the complexity of diagnosing rare nasal cavity tumors. Despite initial treatment with ayurvedic medicine and later surgical intervention, the correct diagnosis was confirmed through immunohistochemistry. The patient underwent chemotherapy as part of multimodal treatment, underscoring the necessity of an integrated approach for managing ENB, including surgery, radiotherapy, and chemotherapy.

BACKGROUND

Esthesioneuroblastomas are rare neuroectodermal tumors originating from the olfactory epithelium, characterized by their occurrence across all ages, with peaks in the second and sixth decades of life. These tumors present significant diagnostic and treatment challenges, often requiring a multidisciplinary approach.

INTRODUCTION

Esthesioneuroblastomas (ENBs), first described by Berger and Luc, are rare tumours thought to arise in the olfactory receptors in the nasal mucosa or the cribriform plate of the ethmoid bone and superior turbinate, extending to base of the skull and to the intracranial space. Due to uncertain histological origin, it has been described by various names in the literature, but most common terms used are ENBs and olfactory neuroblastoma. It manifests across all ages, but peak occurs at second and sixth decade of life. Most of these patients presents in locally advanced stages and require multimodality treatment in form of surgery, chemotherapy and radiotherapy. Multimodality approach with a risk-adapted strategy is required to achieve good control rates while minimizing treatment related toxicity.

CASE PRESENTATION

A 12-year-old male presented with four months of intermittent, progressive epistaxis, nasal congestion, and difficulty breathing, which worsened to include proptosis of the left eye and a protruding nasal mass. Initial treatment with ayurvedic remedies was ineffective.

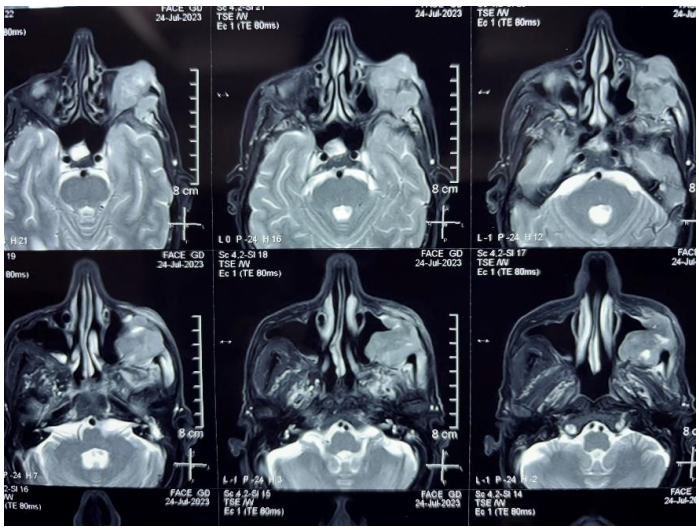


FIGURE 1.

CASE REPORT

This case report accounts a case of 12-year-old male child who had initial complaints of bleeding from left nostril in the form of clots, intermittent, initially scanty in amount, progressive in nature, associated with congestion of nostrils and difficulty in breathing from last four months. Patient had been taking ayurvedic treatment for the same for one and a half months. The symptoms worsened along with the development of proptosis of left eye and protrusion of mass from left nasal cavity. On CECT PNS (from a private laboratory) revealed a soft tissue density mass lesion filling entire left nasal cavity and ethmoid sinus with resorption of ethmoidal septas and superior, middle and inferior nasal turbinates, laterally causing resorption of lamina papyracea and medial wall of maxillary sinus, with posterior extension into nasopharynx with obliteration of nasopharyngeal airway, superiorly extending into the anterior cranial fossa through the left cribriform plate, poster superiorly extending into the sphenoid sinus. Possibility of 1) invasive fungal sinusitis 2) neoplastic aetiology.

On performing Histopathology of nasal cavity mass at a private centre, Necrotising aspergillosis was diagnosed. Patient then presented to our hospital to the ENT department. Here he underwent Left medial maxillectomy under GA. On performing immunohistochemistry, tumour cells are diffusely positive for Synaptophysin, Chromogranin A, NSE, negative for CD99, Desmin and NK 2.2. Diagnosis of olfactory neuroblastoma was confirmed.

Patient was then referred to our department for further management. On repeat MRI FACE mass lesion was detected involving left nasal cavity, left maxillary sinus, nasopharynx, left pterygopalatine fissure, left parapharyngeal space, pterygoid space, superiorly extending into the anterior floor of middle cranial fossa and lateral extension into the left orbit. Multiple enlarged cervical lymph nodes were seen.

Patient has been started on Inj. Cisplatin 25mg/m^2 and Etoposide 100mg/m^2.

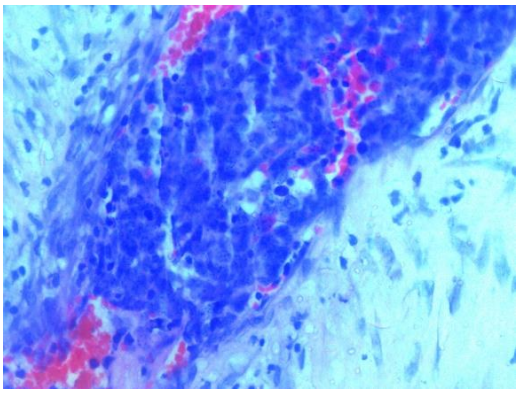
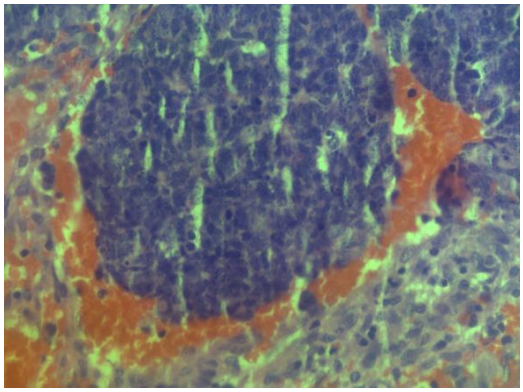


FIGURE 2.



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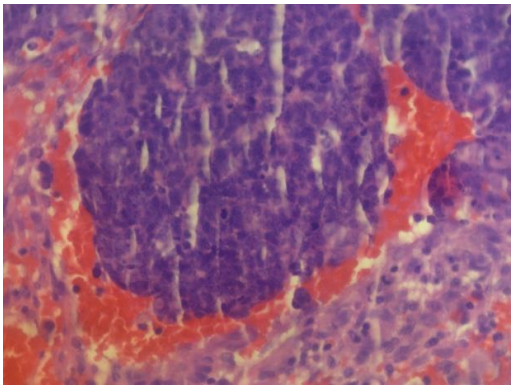

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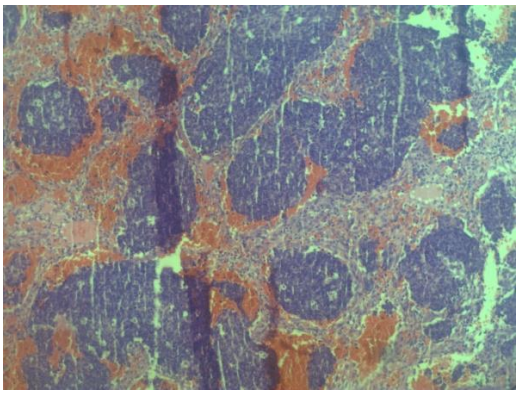

FIGURE 5.

INVESTIGATIONS

CECT PNS identified a mass filling the left nasal cavity and extending into surrounding areas, with possible diagnoses including invasive fungal sinusitis or a neoplastic etiology. Histopathology from a private center suggested necrotizing aspergillosis, but further testing at our hospital confirmed olfactory neuroblastoma.

DIFFERENTIAL DIAGNOSIS

The initial differential diagnosis included invasive fungal sinusitis and other neoplastic causes. The correct diagnosis was established through immunohistochemistry, differentiating it from other tumors.

TREATMENT

Treatment commenced with left medial maxillectomy, followed by chemotherapy with Cisplatin and Etoposide, reflecting the multimodal approach recommended for ENB management.

OUTCOME AND FOLLOW-UP

The case underscores the importance of considering ENB in differential diagnoses for nasal cavity masses and highlights the role of comprehensive treatment strategies. Follow-up will be crucial for monitoring recurrence and managing long-term outcomes.

DISCUSSION

ENBs are rare tumours constituting about 3% of all endonasal neoplasms. They are of neurocrest origin, arising from olfactory sensory cells in the olfactory epithelium.[1] The olfactory epithelium consists of olfactory sensory cells, sustentacular cells and basal cells.[1] There appears to be slight male predominance with a bimodal distribution for age incidence. The peaks are at 11-20 years and 40-60 years, the highest incidence at 51-60 years. These tumours appear to be neuroectodermal in origin in the olfactory epithelium occurring high in the nasal cavity or in the lateral wall adjacent to the ethmoid. Local invasion also occurs, affecting surrounding structures like the paranasal sinuses, the orbit, and the intracranial cavity. Lymphatic spread to the cervical lymph nodes can occur. The incidence for distant metastasis can be as high as 50%. In comparison to other tumours in this region, it has the best prognosis with overall 5-year survival rate of 60-80%.[1]

These tumours are friable and bleed easily on touch. Patient commonly present with epistaxis and nasal blockage. Patients may also have local pain or headache, visual disturbances, rhinorrhoea, tearing, proptosis, or swelling in the cheek.

CT provides the best information about the tumour and its local invasion into surrounding bone structures. MRI allows an estimate of tumour spread into surrounding soft tissue areas, such as the anterior cranial fossa and the retro maxillary space. Bone scintigraphy scan is useful in detecting distant metastases.

Esthesioneuroblastomas consists of lobular sheets with neurofibrillary fibres and rosettes.[2] Hyam's classifications are an important way of determining prognosis.[3] Grade I tumours having an excellent prognosis and grade IV tumours are considered uniformly fatal.

It is characterized by neurofibrillary stroma and neurosecretory granules that are not seen concurrently by any other pathologies in the region.[1] Histological tests such as keratin, CK5/6, S-100 protein or NSE can be run to further differentiate Esthesioneuroblastomas from other tumours.[1]

Kadish et al. were the first to propose a staging classification for ENB. In 1992, Dulguerov et al.[6] proposed TNM staging system based on the TNM system, determined on pre-treatment CT and MRI findings.[4] Nowadays Modified Kadish staging is widely used to stage the sinonasal tumours. A: Tumor confined to nasal cavity. B: Nasal cavity and paranasal sinuses. C: Tumor extends beyond nasal cavity and paranasal sinuses, including skull base, orbit or cribriform plate. D: Tumor metastasizes to cervical lymph nodes and beyond.

Multimodal treatment approach is recommended for increased survival and better quality of life. An ethmoidomaxillary resection with or without orbital sparing is usually necessary. Craniofacial resection allows en bloc resection of the tumour with better assessment of intracranial extension and protection of the brain and optic nerves. The procedure can be combined with pre/postoperative irradiation.

Due to locally infiltrative nature of the disease, surgically clear margins are difficult to achieve. Thus, there is a role of adjuvant RT to minimize the risk of local recurrence.[5,6] In Kadish stage A, surgery alone can suffice but in stage B and C post operative RT is indicated . RT is delivered to the tumour bed and local extension with nodal irradiation reserved for involved nodes. Elective nodal irradiation is not practiced routinely. The dose can vary between 50-60 Gy.

The role of chemotherapy is not very clear in adjuvant settings in early tumours, but in locally advanced and metastatic tumours it has a definitive role. By acting on the systemic micro metastases, it decreases the chances of systemic failure. The common drugs used are Cisplatin, Etoposide, Adriamycin, Vincristine and Cyclophosphamide.

Learning Points/Take-Home Messages

- Esthesioneuroblastoma diagnosis requires careful differential diagnosis and confirmation through specific immunohistochemical markers.
- A multidisciplinary approach is crucial for effective management, combining surgery, radiotherapy, and chemotherapy.
- Early and accurate diagnosis is key to improving outcomes in ENB patients.

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