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Esthesioneuroblastoma- A rare case and its management at Tertiary care hospital in Assam

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Abstract

Aim: To highlight the fact that combined endoscopic and Lateral Rhinotomy approach gives a better exposure of the sino-nasal tumours in cases of extensive loco regional spread.

Background: It was first described by Berger and Luc in 1924, and since then approximately 400 cases have been reported .It accounts for only 3% of all intranasal tumours. It is usually found in young men ,with a secondary peak at the age of 50-60 years.

Case description: 60 year old, male, belonging to poor socioeconomic status, from a small village Keot, Jorhat came to department of Otorhinolaryngology in a Tertiary care centre Assam in January 2019. Patient presented with complaint of severe recurrent epistaxis since 6 months.

Conclusion: Our case was graded according to the Hyam's pathological grading as Grade III and staged as Stage C as per Kadish staging system and T3N0M0 in TNM classification. Epistaxis is not always benign symptom, the underlying cause should be thoroughly investigated.

Clinical significance: Esthesioneuroblastoma (also called olfactoryneuroblastoma) is a rare, slow growing, malignant tumour which is often misdiagnosed at peripheral hospitals, timely diagnosis and management can help decrease significant amount of morbidity in patients.

Keywords: endoscopic, esthesioneuroblastoma, fascia lata dural repair, lateral rhinotomy

Introduction

Esthesioneuroblastoma (also called olfactoryneuroblastoma) is a rare, slow growing malignant tumour that arises from neuroepithelial cells of the olfactory membrane high in the nasal vault and frequently invades the skull base, cranial vault, and orbit. It was first described by Berger and Luc in 1924, and since then approximately 400 cases have been reported.It accounts for only 3% of all intranasal tumours. It is usually found in young men, with a secondary peak at the age of 50-60 years.

Case Report

A 60 year old male, belonging to poor socioeconomic status, from a small village Keot, Jorhat came to

department of Otorhinolaryngology in a Tertiary care centre of Assam in January 2019. Patient presented with complaint of severe recurrent epistaxis since last 6 months for which he was treated at a nearby hospital. He gave history of left side nasal obstruction, swelling on the root of nose which gradually increased in size, anosmia, severe frontal headache unresponsive to analgesic and increased lacrimation of left eye since last 1 month.

Clinical Examination

On examination, a single 4*3 cm square size, firm, nontender, nonmobile swelling was seen involving the root of nose with involvement of skin(Figure 1). On endoscopic examination there was a huge mass in left nasal cavity with local destruction of all anatomical landmarks. It was polypoid in shape, brown red colour, soft fragile and bled on touch. Examination of the right nasal cavity, mouth, ears, neck and cranial nerves were within normal limit.

Investigations

CT scan showed features suggestive of malignant mass in left nasal cavity with extension and bony destructions. There was minimal epidural intracranial extension of the lesion into anterior cranial fossa with bony destruction of Cribriform bone anteriorly.(Figure 2). Endoscopic guided biopsy was taken under local anaesthesia and histopathology came out as Olfactory Neuroblastoma (Esthesioneuroblastoma).

MRI scan of paranasal sinuses showed heterogeneously enhanced polypoidal mass lesion of size 10cm *7.9cm*4cm in left nasal cavity, frontal sinus and bilateral ethmoid complex, exerting mass effects over adjacent structures causing erosion of cribriform plate with extra-axial component in the anterior cranial fossa without brain parenchymal or cavernous sinus invasion.(Figure3)

Staging

Several staging systems, including Hymans, Kadish, and TNM systems, have been proposed as a guide to choosing treatment modalities. Tumour staging is an important guide for prognosis and therapy.

In our case, patient was in stage C regarding Kadish staging system and T3N0M0 in TNM classification.

Operative Procedure

All preoperative investigations were done. Under general endotracheal anesthesia the patient was kept in supine position with perfect alignment of the head and body for suitable placement of the incision and adequate anatomical exposure. Fascia lata graft was harvested from left lateral aspect of the left thigh (Figure 4). Lateral rhinotomy (with supraciliary extension) incision was given on left side and lateral wall of nose was raised to expose the tumour. The tumour was resected and hemostasis was achieved with cauterization of anterior, posterior ethmoidal vessel and septal branch of sphenopalatine artery. Thereafter under endoscopic guidance anterior wall and intersinus septum of frontal sinus was resected and cleared off the tumor. Bilateral sphenoid sinus was excavated and rendered disease free. Posterior bony septum was removed. The Dura near crista galli was found leaking CSF and was repaired with double layered fascia lata graft and tissue glue (Figure 5). Nasal packing was done. The wound was closed in layers with Vicryl 3-0 and Ethilon 3-0.

Post-Operation Histopathology

The resected tumor tissue was sent for histopathological examination (HPE). It showed sheets and lobules of small neoplastic cells separated by vascular and fibrous stroma. The cells showed uniform, sparse cytoplasm, round-oval nuclei, punctate salt and pepper chromatin and inconspicious nucleoli. Some

neoplastic cells are arranged in palisading pattern around central fibrillar neural matrix forming pseudo-rosettes(Homer-Wright rosettes), consistent with the diagnosis of Olfactory Neuroblastoma(Hyams Grading - Grade III)

Post-operative radiotherapy, was given (5500 cGy dose; 30 fraction over 6 weeks).

Followup: Endoscopic examination was done monthly and it did not show any recurrence or complications. On follow-up, repeat CT scan was done after completion of radiotherapy which showed no residual tumor.

Discussion

The low incidence of this disease makes a development of standardised clinical and histological classification difficult. Up to now this tumour is considered to be slow progressive but strained by a high rate of local recurrences[1].

Hyams et al in their study mentioned that the presenting symptoms and signs were nasal obstruction (9/10), epistaxis (3/10), nasal discharge (2/10), while in one case there were no symptoms at all. No patient had regional or distant metastases[2].

In our case report patient presented with complaint of severe recurrent epistaxis since 6 months and gave history of aggravated left side nasal obstruction, swelling on the root of nose which is gradually increasing in size, anosmia, severe frontal headache unresponsive to pain killers and increased lacrimation of left eye since 1 month.

Kadish Set al proposed a system of pretherapy staging in which for Group A, the tumor is limited to the nasal cavity; in group B, the tumor is localized to the nasal acvity and paranasal sinuses; and in group C, the tumor extends beyond the nasal cavity and paranasal sinuses[3].

Pathologic grading of ONB is by Hyams criteria, which groups tumors on a scale of I to IV based on histologic features that roughly represent a spectrum of benign to malignant behavior. Briefly, Hyams grade I tumors display preserved lobular architecture, zero mitotic index, no nuclear polymorphisms, prominent fibrillary matrix, no evidence of necrosis, and cells loosely organized around a central fibrillar eosinophilic material (Homer-Wright pseudorosettes). Hyams grade II tumors have similar findings to grade I but have evidence of low levels of mitoses and nuclear polymorphisms. Hyams grade III tumors begin to have reduced lobular architecture, a moderate mitotic index with moderate levels of nuclear polymorphisms, and a reduction in fibrillary matrix. Flexner-Wintersteiner rosettes, which are true rosettes with cells arranged around an empty space, may be present in Hyams grade III tumors. Hyams grade IV tumors show a high mitotic index and nuclear polymorphism, no fibrillary matrix and rosettes, and frequent necrosis.[4]

In our case, patient was in stage C regarding Kadish staging system and T3N0M0 in TNM classification and HPE showed Hyams Grading - Grade III.

Shah et al in their study mentioned that most patients presented with locally advanced disease. However, regional and distant metastases at the time of initial diagnosis are uncommon. Local recurrence at the primary site was very common, and this reflects either the conservative initial surgical treatment employed or the multicentric nature of the tumor[5].

Stammberger H et al in their study mentioned that the limitation of endoscopic resection result s when the anatomical spread of the tumor has extensive infiltration of orbit, dura/brain and other vital structures exist. However, in experienced hands, endoscopic surgery in this region can be rather radical, bone and

even dura of the anterior skull base can be resected as can the periorbit, and all structures reconstructed in the same session[6].

In our case report, combined Lateral rhinotomy and endoscopic resection was performed.Patient was very carefully studied preoperatively to assess the feasibility and suitability of the procedure.We achieved complete resection of the tumour with excellent local control. There were no postsurgical complications and no long-term cosmetic deformities. The endoscopic skull base reconstruction was done with dura repair with fascia lata graft.

Foote R et al mentioned that the local control was improved in patients who received postoperative adjuvant radiation(5-year rate of local control was 85.9% + 9.3%, compared with 72.7% + 9.5% for those who had operation alone, p = 0.26)[7].

In our case patient recieved 30 fraction of external megavoltage beam with 5500 cGy dose over 6 weeks.

De Vos et al data suggested that metastatic esthesioneuroblastoma is sensitive to platinum-based chemotherapy. They concluded that this tumour is very sensitive to platinum-based chemotherapy and that durable complete response can be achieved, even in a metastatic ENB[8].

Shah J Pet al mentioned that amongst the anterior sull base malignancy, the highest survival rate was observed in patients with esthesioneuroblastoma and lowest in those with mucosal melanoma. Survival was significantly better for those whose tumors could be excised with a limited resection in comparison with those requiring an extended procedure[9].

Federica Sberze et al in their study mentioned that the actuarial 5-year survival was 60%, and the actuarial 5-year disease-free survival was 33%, with a median follow-up of 4.3 years. Recurrences occurred at a median time of 11 months after diagnosis (2.5 mo-18 y). The first site of failure was locoregional alone in 10 of 12 patients who progressed, and in 6 patients involved the brain or the meninges[10].

In our case, in 12 months of followup patient did not show any recurrence.General condition of patient improved over time and he was able to resume his daily activities and job after 6 months of treatment.

Conclusion

Our case was graded according to the Hyam's pathological grading as Grade III and staged as Stage C as per Kadish staging system and T3N0M0 in TNM classification.Epistaxis is not always benign symptom, the underlying cause should be thoroughly investigated. Combined endoscopic and Lateral rhinotomy approach gives a good exposure of the tumour in such extensive loco-regional involvement and allows excellent disease clearance and dura repair can be done in same setting and adjuvant radiotherapy helps achieve a better survival rate.In the present scenario of endoscopic resection of nasal mass, one should not hesitate to use combined approach in such cases for better diseases clearance.

Ethical Clearance: Before commencing the study, necessary permission and approval from ethics committee was obtained from the Institutional Ethics Committee (Human), Assam Medical College and Hospital. Informed written consents was obtained from the patients according to the protocol approved by the Ethics Committee and after explaining the procedure to him in his own understandable language.

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Legend Figures

Figure 1: Preoperative image of the patient

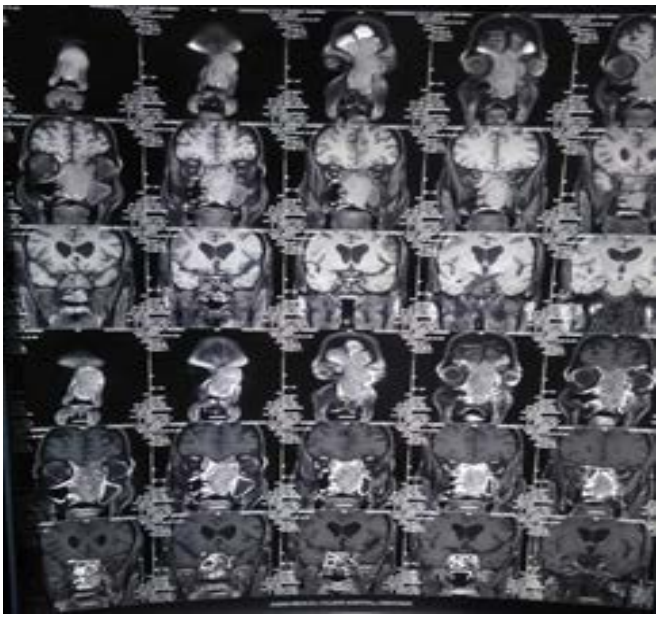

Figure 2: Preoperative CT scan nose and PNS of the patient

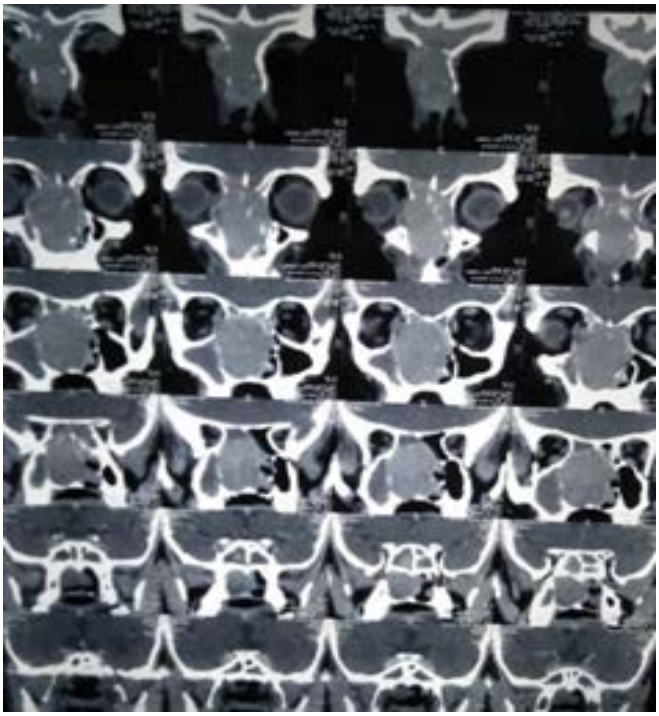

Figure 3: Preoperative MRI Scan Nose and PNS of the patient

Figure 4: Fascia lata graft harvested from lateral aspect of left thigh

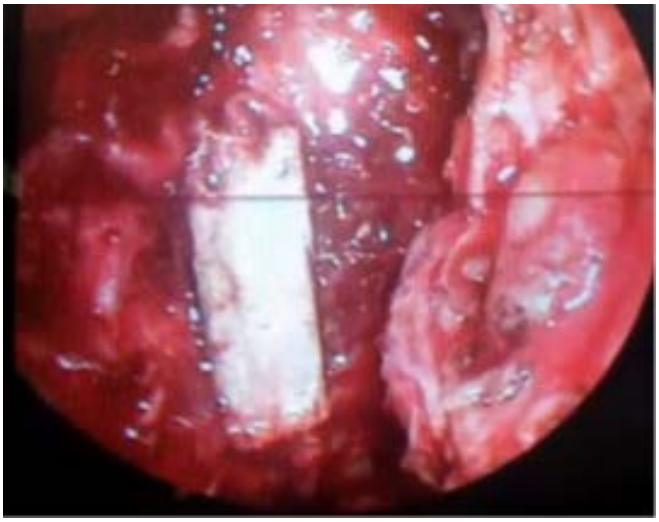

Figure 5: Dura repair by Fascia lata graft