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Ophthalmology

A case of Esthesioneuroblastoma as proptosis.

Dr. Smita Harne Lecturer Department of ophthalmology B.J.Government Medical College Pune

Dr.Apurva Puranik Junior Resident Department of ophthalmology B.J.Government Medical College Pune

ABSTRACT Esthesioneuroblastoma, also known as olfactory neuroblastoma, is an uncommon malignant neoplasm arising from the olfactory epithelium in the roof of the nasal cavity. There are very few case reports published worldwide. The common presenting symptoms of Esthesioneuroblastoma are unilateral nasal obstruction (70%), epistaxis (50%), anosmia, rhinorrhoea, facial pain, headache, excessive lacrimation and rarely proptosis and visual disturbance. Apart from being locally aggressive, it metastasizes by haematogenous and lymphatic routes. We report an extremely rare case of esthesioneuroblastoma in a 18-year-old female with orbital involvement presenting as proptosis. This rare tumour should be considered in the differential diagnosis for young patients presenting to ophthalmic outpatient department with proptosis.

KEYWORDS :

Introduction:

Esthesioneuroblastoma (olfactory neuroblastoma) is a rare neuroectodermal malignant neoplasm that originates from the olfactory sensory epithelium [1-3]. It accounts for upto 5.0% of malignant tumors of the nasal cavity. Berger et al. in 1924 was first to describe this disease. The incidence curve for this disease has a bimodal shape with the first peak in the 2nd decade and 2nd peak in 6th decade with roughly equal sex distribution [2]. Patients present with nonspecific symptoms of nasal obstruction, epistaxis, headache, pain, visual disturbances, anosmia. Owing to the nonspecific nature of the presenting symptoms, patients often have a long history prior to diagnosis [3]. Tumors involving the orbital area generally present with epiphora, decreased visual acuity and proptosis [4]. We report a case of esthesioneuroblastoma in 18 years old female who presented with proptosis and nasal obstruction, diagnosed on computed tomography and histopathology.

Case report:

A 18 years old female presented to opd with complaints of nasal obstruction, nasal bleeding and bulging of left eye with decreased vision for 3 month. She also noticed a rapidly increasing swelling over nasal bridge for 1 month. There was insignificant past history and family history. On systemic examination, there was loss of smell sensation. Local examination revealed a diffuse, firm, nontender swelling with ill-defined margins over the nasal bridge, glabella and over left maxillary region. There was no rise in local temperature. . There was proptosis of left eye (3cm* 2.5 cm)and mild lateral deviation with limited eyeball movement; best corected visual acuity was 6/6 in right eye and 6/24 in left eye with .Extraocular movement were equal and painless in all direction and gazes in right eye. Extraocular movement in left eye were restricted in elevation ,dextroversion dextroelevation and dextrodepresion. There was no evidence of lymphadenopathy. Blood profile demonstrates normal limits of complete blood count, renal function test and liver function test.

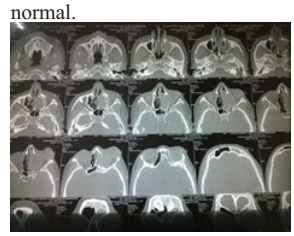
Left eye paraxial proptosis

Pre op Restricted dextroelevation dextroversion and sursumversion in left eye

Magnetic resonance Imaging with contrast of obit revealed large well defined , altered signal intensity area in left nasal cavity, left ethmoidal air cells, left osteomeatal complex ,left maxillary sinus, left sphenoid sinus, left frontal sinus. Break in medial wall of left orbit with extension of lesion in left extra ocular space at medial aspect which was causing compression of left orbital globe, extra ocular muscles , left optic nerve. On post contrast , it shows heterogeneous contrast enhancement likely to represent fungal sinusitis / neoplastic.

Considering fungal etiology intravenous amphotericin b was started for 5 days .However the proptosis did not reduce.

Computed tomography (CT) scan of paranasal sinuses including orbit was performed in axial section with coronal and sagittal reconstructions. CT revealed large, ill-defined, heterogeneously enhancing soft tissue density mass in left nasal cavities and ethmoid sinuses with destroyed nasal septum. There was destruction of medial wall of left maxillary sinus and left lateral nasal wall with extension into left maxillary antrum. Laterally, the mass extended into the left infratemporal region with destruction of the postero-lateral wall of left maxillary sinus .There was also destruction of medial wall of left orbit with intraorbital extension causing antero-latero-inferior displacement of the eyeball resulting in proptosis (The mass extended superiorly involving frontal sinuses; however cribriform plate was normal.



Funtional endoscopic sinus surgey was undertaken that revealed a mass in maxillary sinus that progressed into ethmoidal sinus and it was encroaching on the left globe after breaking the medial wall of left orbit. The mass was removed in toto by FESS.

Post operatively the patient's proptosis reduced (2cm *1 cm).Best corrected visual acuity was 6/6 in right eye and 6/24 in left eye with no improvement on pin hole examination. The patient developed intraop third nerve palsy.

Histo patho examination report showed columnar ciliated epithelium with subepithelial tissues showing tumour composed of sheets of uniform round cells with high N/C ration ,round to oval nuclei and scanty cytoplasm on fibrillary background .Few pseudorossetts seen. No necrosis no mitosis.

Impression malignant small round cell tumour suggestive of olfactory neuroblastoma.

Discussion:

Olfactory neuroblastoma (also known as esthesioneuroblastoma) is a very rare cancer that develops in the upper part of the nasal cavity.Olfactory neuroblastomas generally grow slowly, but in some cases may progress rapidly and aggressively. The faster growing tumors are capable of widespread metastasis.Esthesioneuroblastoma is 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.[3] Esthesioneuroblastoma is made up of lobular sheets with neurofibrullar fibers and rosettes.[1]Diagnosis of ENB requires a high index of suspicion and is ultimately based on biopsy. The most common complaint of ENB patients is unilateral nasal obstruction. Symptoms are typically present for months, and sometimes for years before presentation. Patients may have had multiple procedures for removal of "polyps." Another common symptom is recurrent epistaxis. Other symptoms include anosmia, facial pain, headache, proptosis, diplopia, syncope, and lethargy. The most common physical sign is a high nasal mass.[1] , The mass is usually gray to pink or red, firm, and polypoid. It bleeds easily. Less common findings include neck mass, extraocular paralysis, or nasopharyngeal mass. If clinical suspicion is high, imaging studies, usually CT scan, may be ordered prior to biopsy in order to assess extent of tumor involvement. Biopsy can be performed in the clinic or the OR, but brisk bleeding is to be expected.There are several staging systems described for olfactory neuroblastoma; Kadish system [7] has been the frequently used staging system with good prognostic correlation. It has divided the tumors in three stages: Stage A- tumors restricted to the nasal cavity; Stage B- tumors involving the nasal cavity and paranasal sinuses and Stage C- tumors extending to beyond the paranasal sinuses (orbit, skull base or metastasis).

Esthesioneuroblastoma can resemble small blue cell tumors like squamous cell carcinoma, sinonasal undifferentiated carcinoma, extranodal NK/T cell lymphoma, nasal type, rhabdomyosarcoma, Ewing/PNET, mucosal malignant melanoma and neuroendocrine carcinomas (NEC) that occur in the intranasal tract.Open craniofacial resection or endonasal endoscopic resection is done for stage A and B tumors. Chemotherapy is usually implemented in patients with locally advanced, metastatic or recurrent disease [7-12]. The commonly used chemotherapy combinations are cyclophosphamide plus vincristine and cisplatin-based regimens [12]. The combination of surgery and radiotherapy is the most frequent treatment approach with highest cure rates, however definitive radiotherapy as a conservative management is also used. Despite definitive local therapy, local recurrence and distant metastases have been reported with the metastasis being 25.0-50.0% of cases. In patients with recurrent or metastatic olfactory neuroblastoma, chemotherapy is therefore often implemented. Radiotherapy is usually adopted as an adjuvant following surgery, especially for cases with neck metastasis or as a neoadjuvant before surgery [10,11]. Compared with other sinonasal malignancies, the prognosis of esthesioneuroblastoma is much better, with a disease-free survival at 5 years of more than 80.0% [12].

Conclusion:

In conclusion, clinicians should be aware of this malignant disease and esthesioneuroblastoma. should be considered as one of the differential diagnosis of proptosis.

Consent:

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Competing interest:

Authors declare no conflict of interest.

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