CASE REPORT

OLFACTORY NEUROBLASTOMA PRESENTING AS BLEEDING POLYP

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Malignant tumours of the nasal cavity are rare. Olfactory neuroblastomas (esthesioneuroblastomas) account for only 6% of these neoplasms. Fewer than 1,000 cases have been reported in the literature since this tumour was first described more than 75 years ago. A 13 year old girl presented with bleeding nasal polyp to ENT Department. She was operated as there were no signs of tumour the histopathology report revealed it to be olfactory neuroblastoma. As there was no intracranial extension she was given postoperative radiotherapy.

Keywords: olfactory neuroblastoma, bleeding polyp, nasal, epistaxis

INTRODUCTION

Nasal polypi olfactory neuroblastomas arise from olfactory neuroepithelium, which extends from the roof of the nose to the area of the superior turbinates and a portion of the nasal septum. From there, they can readily extend into the cribriform plate of the ethmoid sinus. Most of the cases described in the literature involved adults, but one case has been reported in a child as young as 2 years of age.[1] Another child aged 8 years is reported in Africa who died later.[2] Fewer than 1,000 cases have been reported in the literature since this tumour was first described more than 75 years ago. A case of olfactory neuroblastoma is being reported because of its rarity and unique presentation.

CASE HISTORY

A girl aged 13 years reported to ENT OPD complaining of recurrent episodes of epistaxis for the last four years. The blood was fresh and clotting, she also had episodic headaches in the frontal bone. She had nasal obstruction on the right side, which was persistent in nature. On examination there was a reddish pollypoidal mass, soaked in blood in the right nostril, it bled on touch. It was arising from the area of middle turbinate which was not visible. Inferior turbinate and septum were intact. Eye movements were normal and no diplopia was noticed. Visual aquity and colour vision were normal. Considering it to be a bleeding polyp surgery with intranasal removal under general anaesthesia was planned. Preoperative investigations were normal. She was operated upon and polyp was removed and under suction and packing the nasal cavity was cleaned. During surgery it was noticed that the Mass was arising from the roof and destroyed the middle meatus, as we could enter the Maxillary sinus easily. The nose was packed with BIPP which was removed on the 5th postoperative day. Tissue removed was sent for histopathology which had blue cell suggestive of olfactory neuroblastoma. The CT scan PNS was done with and without contrast (fig-2) which showed enhancing mass in the right nasal cavity extending from the cribriform plate obliterating the medial orbital wall and middle meatus. No intracranial extension was noticed. Keeping in mind the facilities available in our hospital, she was referred to specific centre for craniofacial resection and postoperative radiotherapy.

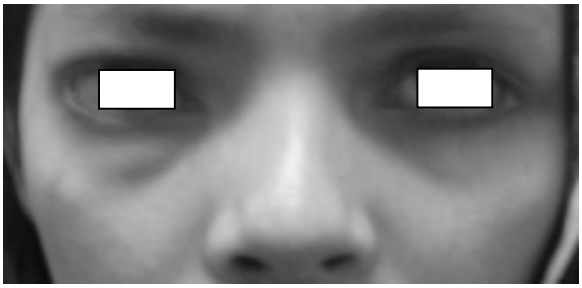

Figure-1: Normal-looking nose of the patient

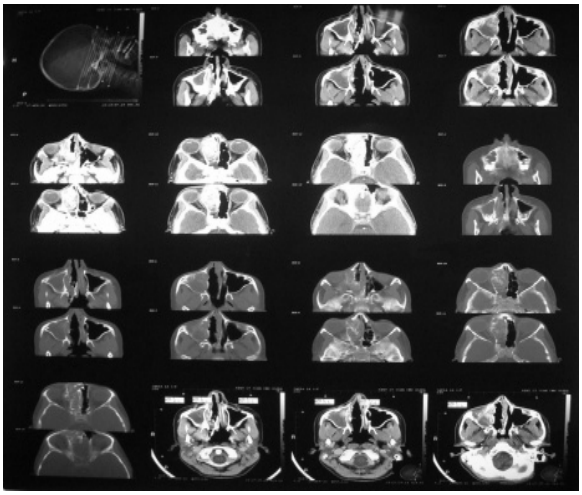

Figure-2: CT Scan paranasal sinuses showing the Mass at different levels

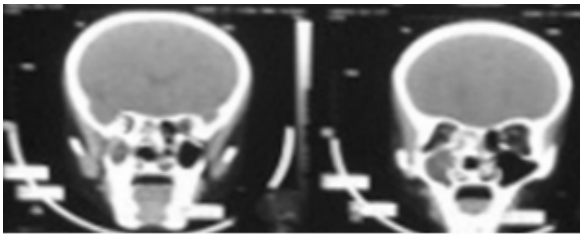

Figure-3: CT scan showing the mass in right side of nose. Right maxillary sinus is opaque

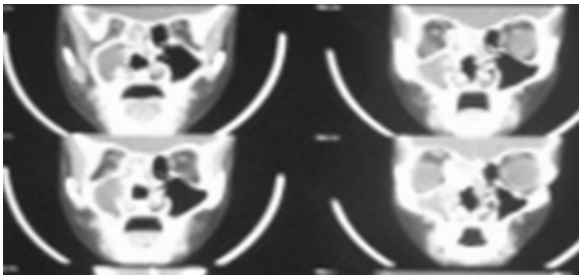

Figure-4: CT scan showing the mass and opaque right maxillary sinus

DISCUSSION

Olfactory Esthesioneuroblastoma is an uncommon neuroectodermal tumor that originates from the olfactory sensory epithelium in the upper nasal fosse at the level of the cribriform plate.[3] It represents up to 5% of malignant tumours of the nasal cavity. First described by Berger et al[4] in 1924, almost 1000 cases have been published. The probable origins reported for this tumour include the sphenopalatine ganglion, the vomeronasal organ of Jacobson, the neuroepithelial cells of the olfactory membrane, the ectopic olfactory epithelium in the nasal mucosa, and the amine precursor uptake and decarboxylation cells. These tumours affect both sexes equally. A bimodal age distribution (the 2nd and 6th decades of life) has been documented, although patients of all ages can be affected. Patients present with non-specific symptoms of nasal obstruction (70% of cases) and epistaxis (50%); and headache patients with extensive tumours may have orbital symptoms such as proptosis, excessive lacrimation and anomie (<5%).[5]

The Hyams classification (grades I through IV) is based on histological differentiation; the grade IV designation is used to describe undifferentiated sino-nasal carcinomas.[6] The grade correlates with the prognosis. More well accepted is the Kadish classification system (stages A through C), which is based on the clinical spread of the tumour: stage A tumours are confined to the nasal cavity, stage B lesions involve the sinuses, and stage C masses involve the middle fossa.[7]

Owing to the 'small, round, blue-cell' nature of the neoplasm, the differential diagnosis is quite broad; it includes melanoma, rhabdomyosarcoma, sinonasal undifferentiated carcinoma, lymphoma, Ewing's sarcoma, pituitary adenoma, plasmacytoma, paraganglioma, and primitive neuroectodermal tumour.

CONCLUSION

Olfactory neuroblastoma is a rare entity; still we should have high indices of suspicion. Although it has a bimodal age distribution no age should be considered safe. Treating physician should be aware of the capabilities of self and the centre in which he/she is working and should not feel shy in referring the patients to specified centers.

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