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RESEARCH ARTICLE

A CASE SERIES ON OLFACTORY NEUROBLASTOMA

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Abstract

Olfactory neuroblastoma (ON), also called Esthesioneuroblastoma (ENB) is a rare malignant nasal tumour that arises from the olfactory neuroepithelium in the olfactory rim of nasal cavity. Here, we present a case series of two patients who presented with nasal obstruction, epistaxis and anosmia. Clinical examination revealed reddish mass within the nasal cavity and extending into the nasopharynx. An endoscopic craniofacial resection was done and biopsy confirmed the masses to be olfactory neuroblastoma.

Introduction:-

Esthesioneuroblastoma (ENB), also called olfactory neuroblastoma (ON), is a rare malignant nasal tumor. It accounts for about 1–5% of all malignant paranasal sinus tumours. The incidence of Olfactory neuroblastoma is around 0.4 per million. It arises from the olfactory neuroepithelium in the olfactory rim of the nasal cavity. Berger and Luc, termed the tumour "esthesioneuroepithelioma", when they first described it in 1924. ENB presents with peaks in the second and sixth decades of life. Clinical features associated with ENB are unilateral nasal obstruction, epistaxis, headache, rhinorrhea, anosmia and visual changes. The gold standard of diagnosis is Biopsy of the mass. Other modalities of evaluation include contrast enhanced computed tomography (CECT) and Magnetic resonance imaging (MRI). Treatment options consist of surgery or RT only, surgery and RT, surgery and chemotherapy combined with RT, or only chemotherapy and palliative care.

The present study reports the case of a patient with a mass in the nasal cavity who was treated by endoscopic craniofacial resection.

Case Report

Case 1:

A 23 years old male patient presenting with complaints of bilateral nasal obstruction for 6 months. He also gave a history of headache in both frontal region, epistaxis and anosmia. There was no history of visual abnormalities or diplopia. Anterior rhinoscopy showed reddish mass in both nasal cavities which bleeds on touch. Posterior rhinoscopy showed a reddish mass occupying the whole of nasopharynx.

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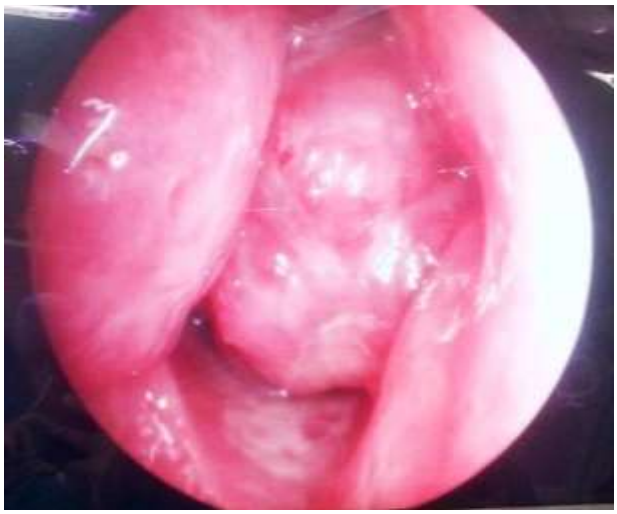

Figure 1: Endoscopic picture of the Right sinonasal mass

Figure 2: CT Paranasal sinuses showing Right sinonasal mass

MRI paranasal sinuses showed a large multilobulated mass of 70 X 46 X 40 mm occupying the nasal cavity and nasopharynx extending into bilateral frontal sinuses, floor of anterior cranial fossa, floor of nasal cavity, ethmoid sinuses and maxillary sinus. Anteriorly it was found to be extending into the nasal cavity and posteriorly into the nasopharynx in the lower part and in sphenoid sinuses in the upper part. The mass was centered at the superior olfactory recesses bilaterally and involves ethmoid air cells. Surrounding bones were found to be destroyed. The walls of anterior cranial fossa and bilateral orbits were intact. Biopsy of the mass showed features suggestive of olfactory neuroblastoma which as strongly positive for Neuron-Specific Enolase and negative for Chromogranin. An endoscopic craniofacial resection was done to remove the mass. Patient underwent radiotherapy postoperatively. Postoperative MRI scan showed no residual mass.

Case 2:

47 years old male patient presented with complaints of bilateral nasal obstruction, epistaxis and anosmia for 4 months. There was no history of visual abnormalities or diplopia. Anterior rhinoscopy showed reddish mass in the right nasal cavity which bleeds on touch. Posterior rhinoscopy showed a reddish mass occupying the whole of

nasopharynx. MRI paranasal sinuses revealed fairly defined lobulated T2 hyperintense and T1 hypointense lesion in right nasal cavity and ethmoid air cells extending from anterior to posterior choana. The lesion was seen displacing the nasal septum to left. An endoscopic resection was done to remove the mass. Biopsy of the mass confirmed diagnosis of olfactory neuroblastoma. Postoperative MRI scan showed no residual mass.

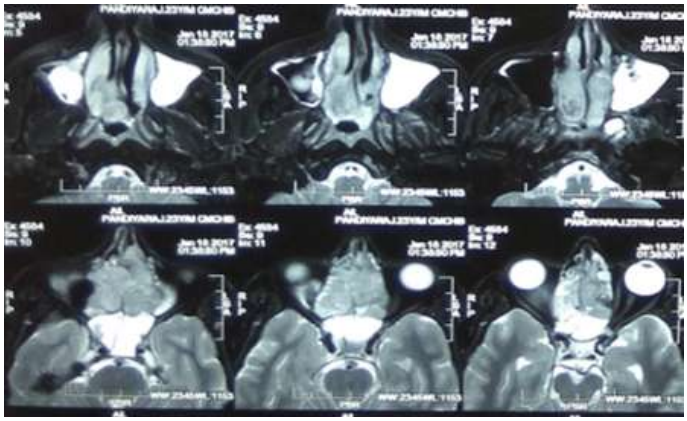

Figure 3: MRI paranasal sinuses showing Right sinonasal mass

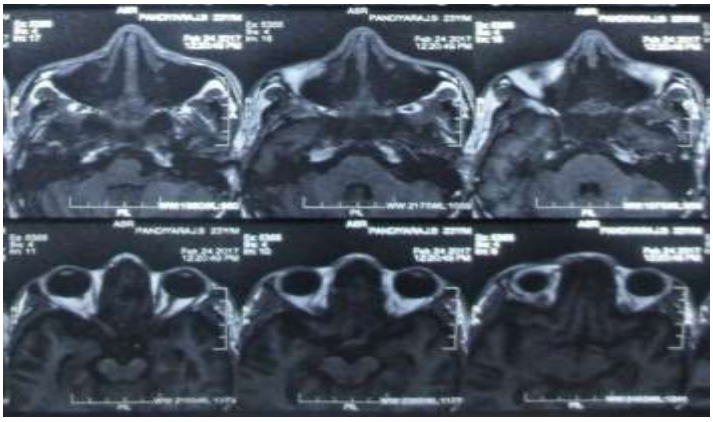

Figure 4: Postoperative MRI showed no residual mass

Discussion:-

Olfactory Neusroblastoma (ON) is an uncommon malignant nasal tumour. They make up less than 5% of paranasal sinus malignancies. It occurs more commonly in the 2nd and 6th decades of life. They are of neuroectodermal origin derived from olfactory epithelium. The most common symptoms are one-sided nose obstruction and epistaxis, while rhinorrhea and anosmia may also occasionally accompany these symptoms. Extensive lesions may cause frontal headaches and diplopia. Clinically, ON presents most commonly as a large, unilateral, polypoid, glistening, reddish-grey mass in the superior nasal cavity. Extension into the adjacent paranasal sinuses, orbits and cranial vault may also be seen.

Radiography shows the presence of a characteristic dumbbell-shaped occupying the superior nasal cavity and ethmoid sinus, extending through the cribriform plate into the anterior cranial fossa. CT scan shows homogenous enhancement, focal calcifications, remodeled or resorbed bone margins. ON can spread quickly and easily into the intracranial structures via the cribriform plate. Cribriform plate and orbit involvement in ON are important prognostic factors. Blood borne metastasis occurs in 20% of cases, mostly to lungs and bones and lymphatic metastasis in less than 10% of cases.

Histopathologically, the tumor consists of small medium sized round cells with uniform nuclei, scanty cytoplasm and few mitoses having intercellular fibrillary stroma. On immunohistochemical study, ON is usually diffusely positive forsynaptophysin, chromogranin, CD56, and NSE. S-100 protein positive sustentacular cells highlight the periphery of the tumor. ON is negative for desmin, myogenin, actin, CD99, EBER, TTF1, and CD45RB.

The Kadish staging system is the most widely accepted classification system for olfactory neuroblastoma. According to this system, group A tumors are limited to the nasal cavity, group B tumors extend only to the paranasal sinus, and group C tumors extend beyond the nasal cavity and sinuses. A modified Kadish system includes group D tumors, which have cervical lymphadenopathy or distant metastasis. This system continues to serve as a reliable prognostic predictor of outcome and long-term patient survivability in ON. There is decrease in survival as the stage increases: 75-91% for Stage A, 68–71% for Stage B and 41–47% for Stage C. The overall 5-year survival rate is 60–80%. Hyams et al. developed a four-tiered grading system, which has also shown good overall correlation with outcome.

Complete surgical elimination of the tumor requires a bicranial-facial approach, which removes the cribriform plate. This followed by a course of radiotherapy is considered as the treatment of choice and found to achieve the best long term outcome. Endoscopic resection can also achieve similar results for limited tumor.

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