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Olfactory Neuroblastoma Presenting as Fibro-Osseous Lesion of Maxillary Bone.

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ABSTRACT

A seven year female child reported to department of oral and maxillofacial surgery with complaint of swelling at left upper jaw since 5months. Provisional diagnosis of fibro-osseus lesion was made and curettage was done. After 2 months, patient again appeared with complain of swelling of left maxilla and neck. Computed tomography (CT) scan of face and neck was done which was suggestive of osteodestructive, osteoexpansile mildly homogenously enhancing soft tissue lesion involving left maxilla with features and extension as described-neoplastic etiology. (? Rhabdomyosarcoma ?? lymphomatous infiltration) while histomorphology and immunohistochemistry report revealed olfactory neuroblastoma (ONB)with positivity of synaptophysin, CD-56 and FLI-1. Ki prolification index was 60-70%. Patient was given chemotherapy but did not respond.

Key words: Fibro-osseus lesion–CT scan–Rhabdomyosarcoma–lymphomatous infiltration–olfactory neuroblastoma

1 INTRODUCTION:

Olfactory epithelium in the nasal cavity and the paranasal sinuses gives rise to rare malignant tumor ONB which was first explained by Berger et al. in 1924 [1]. Early diagnosis of ONB is very difficult, because of its nonspecific symptoms and signs. The most common clinical presentation of ONB, whose exact aetiology is unknown till now, is nasal obstruction, craniofacial pain, olfactory and ophthalmic disturbances and epistaxis. Local invasion of ONB occurs in paranasal sinuses and its side structures. Slowly arising this tumour is also known by the name esthesioneuroblastoma [2]. Among all nasal carcinomas existence of ONB is about 3% to 5% whose incidence is expected up to 1 per 2.5 million [3]. It causes increased morbidity and mortality along with high recurrence rate. Secondary to ONB, presentation of Cushing syndrome, ectopic adrenocorticotropic hormone syndrome, paraneoplastic syndromes may be found in few patients. Histopathology and immunohistochemistry makes the diagnosis of ONB. Metastasis occurs in cervical and retropharyngeal lymphnodes. Consensus on management of ONB is not there, because of its rarity. Surgical resection followed by radiotherapy is currently treatment of choice, but there is no definite protocol for treatment [4] because of the rarity of the disease. However, the optimal treatment continues to be controversial because of the rarity of the disease. Small molecule or monoclonal antibody therapy is not supported by any report.

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2 CASE PRESENTATION:

A seven year female child reported to department of oral and maxillofacial surgery with complaint of swelling at left upper jaw since five months. Figure 1 She was not giving any history of trauma, pain, trismus, loosening of teeth, decrease in vision and epistaxis. On examination, she was afebrile. Glasgow coma scale (GCS) was E4V5M6 and her vitals were stable.

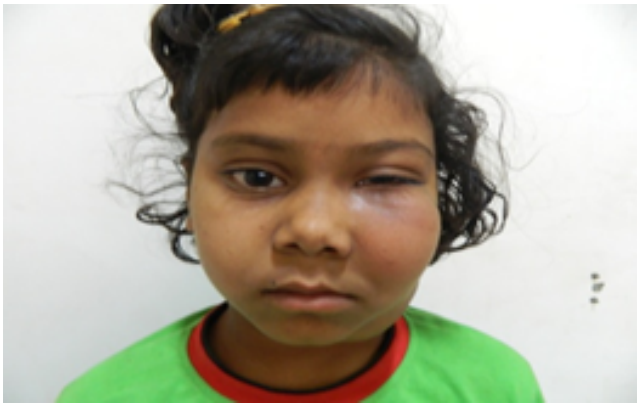

Figure 1. Showing swelling atface

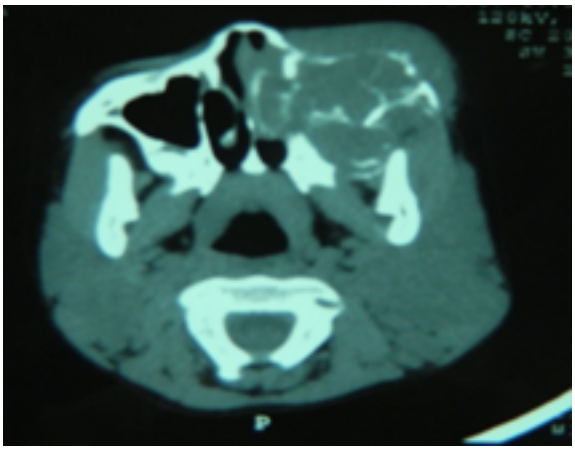

Figure 2. CT Axial View

On regional examination smooth and hard bony swelling that was involving left maxilla and neck was present while skin overlying it was normal. Provisional diagnosis of fibro-osseus lesion was made. Curettage was done, but patient again appeared with complain of swelling of left maxilla and neck after two months. On extra –oral examination two swellings were seen at maxilla and neck respectively. Maxillary swelling measuring 6x6cm in diameter, extending from extra –orbital rim to zygomatic bone and ala of nose to 8 cm anterior to tragus of ear. Swelling at neck were three enlarged lymph nodes measuring into 9 cm in diameter, extending lobule of ear to angle of mandible superior-inferiorly and body of mandible to anterior border or sterno-cleidomastoid muscle, anteroposteriorly. On palpation maxilla swelling was not tender and temperature over skin was slightly raised. Neck nodes were not tender, firm in consistency and mobile. All other systemic examinations were within normal limit. Lab investigations showed haemoglobin (Hb) of 13.4g/dl, total lecocyte count(TLC) 10300 cells/mm^3, neutrophils 78% and lymphocytes20%, sodium(Na) 138mmol/l, potassium(K) 3.87mmol/l. Renal parameters and liver enzymes were normal in range. CT scan face and neck Figures 2 and 3 was suggestive of osteodestructive, osteoexpansile mildly homogenously enhancing soft tissue lesion involving left maxilla with features and extension as described-neoplastic etiology. (? Rhabdomyosarcoma ?? lymphomatous infiltration)

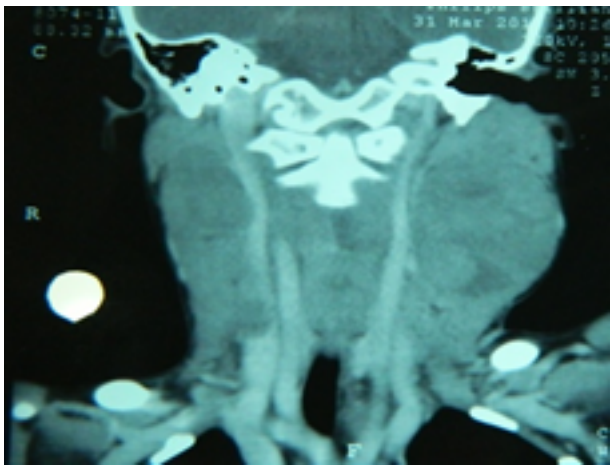

Figure 3. CT Coronal View

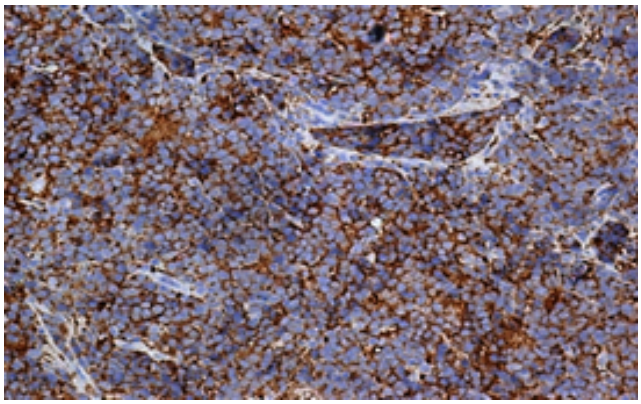

Figure 4. IHC positive for synaptophysin

Histomorphology and immunohistochemistry report revealed olfactory neuroblastoma with positivity of synaptophysin, CD-56 and FLI-1. Ki prolification index was 60-70%. On the basis of history, clinical evaluation, radiography and histological report we could reach on the diagnosis of olfactory neuroblastoma.Figure 4. Cisplatin + etoposide,+Endoxan-N + vincristin +doxorubicin based chemotherapy was given but patient did not respond and died after 10 days of chemotherapy

3 DISCUSSION:

ONB is a rare malignant tumor and among all nasal and paranasal malignancies it comprises of 4,65%. [5] About 1000 cases are reported since 1924, when it was first introduced. Peaks of occurance are in extremes of age with highest in the second, third , sixth and seventh decades of life. ONB is equally found in both sexes. Six month is the mean duration from appearance of first symptom to making of the diagnosis. [6] In our case it was 5 months. Unilateral nasal obstruction and epistaxis are the commonest initial symptoms, but in our patient these were absent and she was having non tender swelling at left maxilla only. We didn't have universally accepted diagnostic criteria for ONB. Small blue cell neoplasm with a lobular architecture is the characteristic of histopathology. Undifferentiated small cell carcinoma, rhabdomyosarcoma , melanoma, or other small blue cell neoplasms are the differentials for ONB. The mortality as whole in ONB is 32% while in patients with metastasis it is 60% [6] It is challenging task to find the optimal treatment scenario for recurrent ONB and the standard treatment is combination of endoscopic/open surgery, radiotherapy, and/or chemotherapy along with neoadjuvant and adjuvant chemotherapy [7] Outcome after the treatment remains unknown and it was also seen that with traditional treatment regimens there is high chances of failure in recurrent and metastatic ONB cases. Brain and bone marrow metastases are associated with poor prognosis. Although orbital involvement is a serious prognostic factor we can save eyes if there is only periosteal involvement and not intraocular structures. A study was published by The Institute of Laryngology and Otology, University College London, where 42 patients treated for 23 years and craniofacial resection was done in all patients. 57% patients treated with combination therapy of surgery and radiation therapy. Survival rates were 77 and 61% at 5 years, and 53 and 42% at 10 years, respectively. Worse outcome was there with intracranial extension and orbital involvement. Cervical metastasis was a having better prognosis in comparison to other distant metastases. Few patients may have intolerance of radiation and/or chemotherapy. Genome-based precision medicine is the most recent therapeutics dealing the attention of oncologists. Targeted medicines may have complete or significant improvement in ONB cases which was seen in so many studies and it was because of their high specificity and low toxic effects. Sunitinib and cetuximab are two small molecule-targeted drugs. Vascular endothelial growth factor (VEGF) receptors (VEGFR2/KDR), fibroblast growth factor receptors (FGFR1 and FGFR2), platelet-derived growth factor receptors (PDGFRA and PDGFRB), fetal liver tyrosine kinase receptor (FLT1, FLT3, and FLT4), RET, and c-Kit receptors of tyrosine kinases are inhibited by sunitinib which was approved by [8] Food and Drug Administration (FDA) in 2011 for treatment of unresectable or metastatic pancreatic neuroendocrine tumors, metastatic renal cell carcinoma, metastatic or unresectable gastrointestinal stromal tumors (GIST). [9] Head and neck cancers are treated by cetuximab, a monoclonal antibody which was approved by US FDA in 2004. There is no any report of personalization of treatment of ONB with targeted drugs based on the patient's genomic variations. Combination regimen of cetuximab plus sunitinib showed a drastic response in treatment of ONB. In conclusion ,ONB is a very rare tumor. It should be early diagnosed and treated. Surgery combined with radiotherapy is the proffered mode of treatment in ONB cases. In conclusion ONB is a very rare tumor. It should be early diagnosed and treated. Surgery combined with radiotherapy is the proffered mode of treatment in ONB cases.

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