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Otorhinolaryngology

A CASE REPORT OF SOFT TISSUE EWING'S SARCOMA OF NASAL SEPTUM

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ABSTRACT Ewing's sarcoma, an aggresive malignant round cell tumour of bones , arising most commonly in long bones and in young age group. This is a case report of Ewing's sarcoma of extra osseous origin arising from cartilagenous septum and in atypical age group, A 51 year old male, presented to ENT opd with 2 years history of right sided nasal obstruction and reccurent unprovoked anterior epistaxis. Biopsy taken from the mass in right nasal cavity revealed small round blue cell tumor on HPE , and, NKX2.2, CK and Synaptophysisn are positive. PET CT confirmed a local metastatic node (level 1B). Post surgery(excision of tumour with free margins) , Patient is on chemotherapy and is on regular follow up. No evidence of reccurence and metastases were observed so far.

KEYWORDS : Ewing's sarcoma, Extra skeletal, Nasal septum

INTRODUCTION

Ewing's sarcoma(ES) is a malignant round cell neoplasm originating from both skeletal and extra skeletal structures. In general, skeletal forms are more common. Long bones of the extremities are most common areas in skeletal forms while soft tissue of lower extremities, para vertebral tissues, chest wall and retroperitoneum are common areas in extra skeletal forms.[1] Primary sino-nasal location is rare. Only 1-4% of Ewing's sarcoma's are primarily located in head and neck region,10 case reports of sinonasal ES of the nose or paranasal sinuses have been reported.[6]The term Ewing's sarcoma is a part of Ewing family tumour (EFT) used in soft tissue neoplasms which was originally described as primitive neuroectodermal tumours (PNET).[8]

Multiple differential diagnoses of round cell neoplasms and specific tumours for the site makes diagnosis challenging. Immunohistochemistry(IHC) confirms the diagnosis.

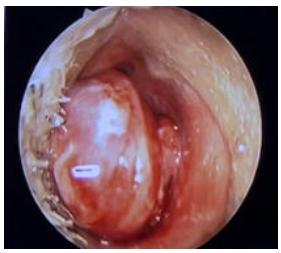

Figure-1: Endoscopic image showing mass in right nasal cavity

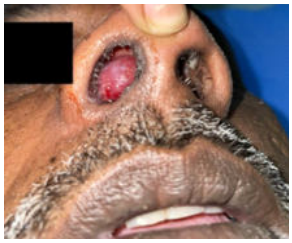

Figure-2: Tip raise test showing mass in right nasal cavity

CASE REPORT

A 51 year old male presented to ENT OPD at NRI medical college, chinakakani, mangalagiri, Andhra pradesh, a tertiary care referral hospital, with 2 years history of right sided nasal obstruction and intermittent unprovoked anterior epistaxis. A pinkish firm lobulated mass seen in right nasal cavity anteriorly just behind the vestibule, attached to cartilagenous part of the right side of nasal septum extending from caudal end till bony cartilagenous junction, not attached to roof and lateral wall, not bleeding on touch and cannot be probed all around and the mass was attached to nasal septum. CT scan of nose and paranasal sinuses was done- which suggested well defined, soft tissue density lesion of 2.3x4x2.5cm obliterating the right nasal cavity and causing erosion of cartilagenous septum on right side with no evidence of calcifications.

Endoscopic assisted excision of mass with free margins was done. And sent for HPE which revealed, sheets of small blue round cells with clear vacuolated cytoplasm, intervening stroma showing fibrocollagenous tissue with increased vascularity. Considering possibility of Ewing's family tumour, IHC Was done which showed tumour cells positive for synaptophysin, CK, NKX2.2.

EWSR1 gene re-arrangement studies done. PET CT revealed one metastatic node (level 1B) on same side. Patient was referred to department of medical oncology for further management. Patient is on chemotherapy (cyclophosphamide, etoposide) and is on regular followup without any residual disease, and reccurence.

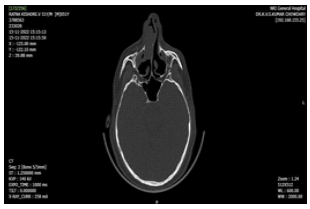

Figure-3: Computed Tomogram axial cut showing mass in right nasal cavity

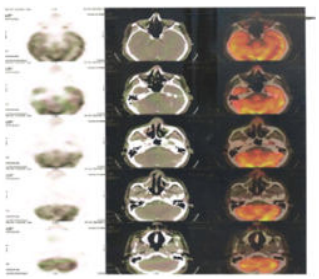

Figure-4 Image of post operative PET-CT SCAN

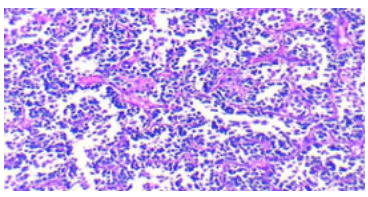

Figure-5: Low power view (100x magnification) show tumour cells in sheets and vague alveolar pattern interspersed by delicate fibrous septa

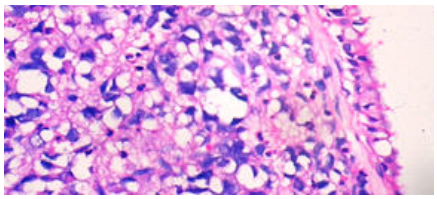

Figure-6 : High power view (400x magnification) showing tumour cells with relatively monomorphic round nuclei, coarse and illdefined clear cytoplasm. Perivascular rosette-like configuration noted.

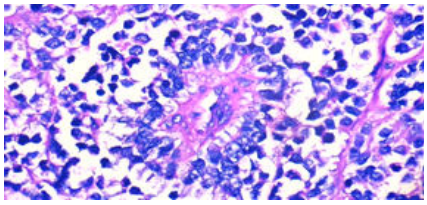

Figure-7: High power view (400x magnification) show tumour cells reaching close but doesnot involve overlying respiratory epithelium

DISCUSSION

ES is a rare aggresive tumour that typically involves the long bones of extremities (skeletal form). The less common variety (extra skeletal form) involves soft tissue and rarely manifests in head and neck region.[1],[2] Usually occurs in patients younger than 30yrs of age with a peak incidence in those aged 10 and 15 yrs.[7] In the sinonasal tract, the differential diagnosis includes melanoma, rhabdomyosarcoma, sino nasal undifferentiated carcinoma, lymphoma, esthesioneuroblastoma, Ewing's sarcoma, poorly differentiated carcinoma. It is difficut to differentiate these tumours based on clinical and radiological examination alone; hence it requires a HPE, IHC and cytogenetic analysis to reach a definitive diagnosis.[8]

Clinical manifestations of sino nasal Ewing's sarcoma include, nasal obstruction, rhinorrhea and epistaxis. Most of the patients present with metastasis at the time of diagnosis[9],[10]. Most common sites of distant metastasis are lungs and bones. Positron-emission tomography should be performed to stage the disease and to look for evidence of metastatic disease. According to literature, the metastatic rate at diagnosis is 12.5% for Ewing's sarcoma of head and neck; 20% to 30% for Ewing's sarcoma of all sites[9],[10] .

Treatment of Ewing's sarcoma involves multi drug chemotherapy followed by surgical resection and or radiotherapy[2],[11].

The prognosis depends on the site of the primary tumour, presence of distant metastasis at presentation and the age of patient. According to literature, patients younger than 15 years of age and patient with axial and sinonasal tract involvement have better prognosis. While the 5-year survival of patients with metastasis is around 22% , it is 55% in those without metastasis[2],[9],[10]. However, the effective treatment of Ewing's sarcoma has improved the survival rate upto 86% in patients without metastatic disease.

CONCLUSIONS

Ewing's sarcoma is a small round cell tumour that originates from primitive neuroectodermal cells and typically involves the long bones of extremities. Primary origin of Ewing's sarcoma in sino nasal tract is a rare presentation. Diagnosis of the disease is challenging by HPE and it always requires IHC and Cytogenetic studies. Treatment includes a multidisciplinary approach with Chemotherapy followed by surgery as first line and or radiotherapy.

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