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Rare Presentation of Extra Skeletal Ewing's Sarcoma of the Sinonasal Tract - A Case Report

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ABSTRACT

Ewing's sarcoma (ES) is a rare and aggressive malignant tumor that typically involves the long bones of the extremities (skeletal form). The less common extra skeletal form involves soft tissues and rarely manifests in the head and neck region. Genetically, the abnormal t (11:22) chromosome translocation is a hallmark of diagnosis. Ewing's Sarcoma of the nose or paranasal sinuses is a rare subset of this type of tumour to occur in Nose and Paranasal Sinuses and treatment consisting primarily of chemotherapy followed by surgery and/or radiotherapy.

Case details- A very rare case of 37-year-old female with an extra skeletal form of Ewing's sarcoma who was diagnosed and operated in the department of ENT- Head & Neck Surgery, Agartala Government Medical College is presented here because of its rarity.

Keywords: Ewing's sarcoma (ES), Paranasal sinuses (PNS), Extraskeletal sarcoma (ES)

INTRODUCTION

Ewing's Sarcoma (ES) is a form of very aggressive tumours which is closely related to family of small round cell tumours [1]. It is a rare and aggressive tumour which is classified as peripheral primitive neuro ectodermal tumor. It was first described by James Ewing, an American pathologist in 1921 [2]. Ewing's sarcoma commonly occurs in early childhood and adolescence, but rarely it can occur in adults [3]. Most of the Ewing's sarcoma are arised from long bones and extremities. Extra skeletal Ewing's sarcoma is a relatively uncommon primary tumor of the soft tissues, which accounts for 20-30 % of all reported case of Ewing's sarcoma among them head and neck region accounts for only 1-4 % of all Ewing's sarcoma. ES of the nose or paranasal sinuses is a rare subset of this type of tumours and treatment consisting primarily of chemotherapy followed by surgery and/ or radiotherapy.

CASE HISTORY

A 37-year-old female patient from remote area of state of Tripura who presented with a history of left nasal obstruction, bleeding from nose on and off for 3 months. On clinical Examination- A friable, fleshy, reddish mass was visualised in left nasal cavity (Fig 1). On Diagnostic nasal endoscopy – mass was filling up the whole left nostril extending to left Osteomeatal complex and choanae. Medial wall of Left maxillary sinus was destroyed (Fig 2). Computed tomography of PNS showing gross nasal septal deviation to the right side with a soft tissue attenuation of the whole left nasal cavity, maxillary sinus and ipsilateral sphenoidal sinus (Fig- 3). MRI of PNS reveals heterogenous solid mass lesion

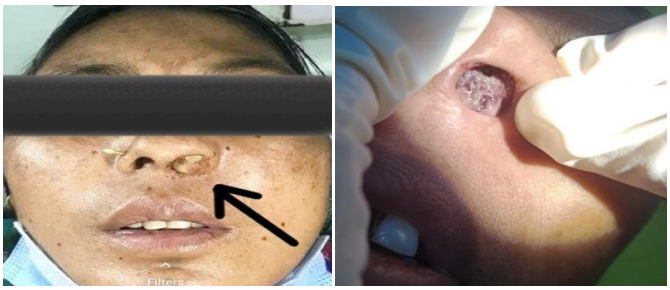
(5.84cmx4.22cm) in the left nasal cavity that appears hyperintense in T2W1, hypointense in T1W1. Mass lesion is displacing the nasal septum laterally, extending laterally into left maxillary sinus causing obstruction of ostium of left maxillary sinus and fluid collection, mucosal thickening of left maxillary sinus, posteriorly upto choana, superiorly upto sphenoid sinus. (Fig 4). Histopathological examination was inconclusive.

Through transnasal endoscopic approach mass removed in completely (Fig 5). It was friable and encapsulated mass involving whole left nasal cavity and maxillary sinus and also sphenoid sinus. Nasal septum was grossly deviated to right and thinned out. Tissue was sent for Histopathological examination.

After surgery Post op was uneventful. Nasal pack removed after 48 hr. Patient was discharged after 7 days

Histopathological report suggestive of Sections from all the & specimen shown uniform round cells with central round nucleus with stippled chromatin and moderate amount of vaculated cytoplasm. Tumor cells are PAS positive. No lymphovascular invasion is seen. The malignant cells have circumferentially infiltrated the margins. IHC findings: - Chromogranin negative, CK Positive focally. Impression:- Extra-skeletal Ewings sarcoma (Nasal Polyp (left) (Fig6).

After receiving the Histopathological report of nasal mass patient was referred to regional cancer centre for further treatment. Patient has received 6 cycles of radiotherapy.



A **B**

Fig -1: Showing friable mass in left nasal cavity.

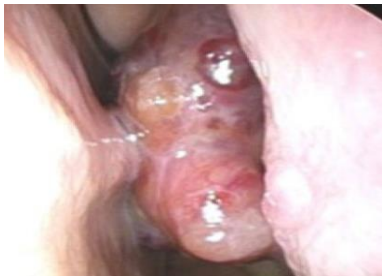


Fig-2: Nasal endoscopy image showing a mass in left nasal cavity.

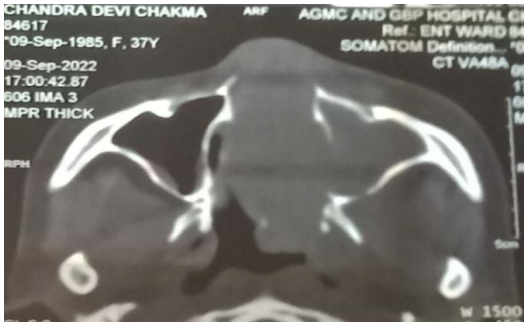


Fig- 3: Computed tomography of nose and paranasal sinuses showing gross nasal septal deviation to the right side with a soft tissue attenuation of the whole left nasal cavity, maxillary sinus and ipsilateral sphenoidal sinus.

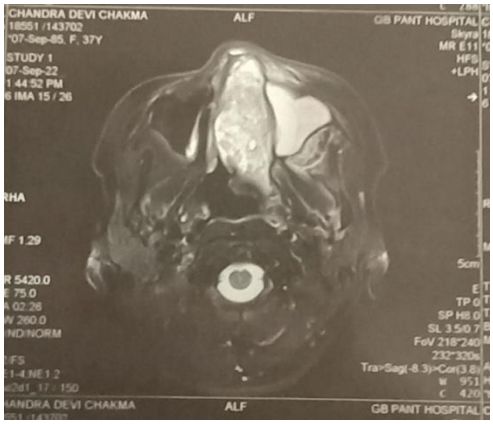
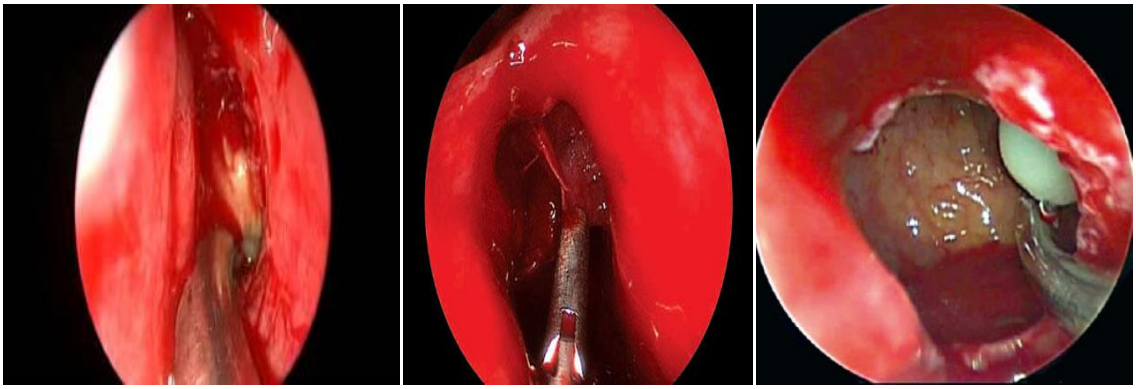
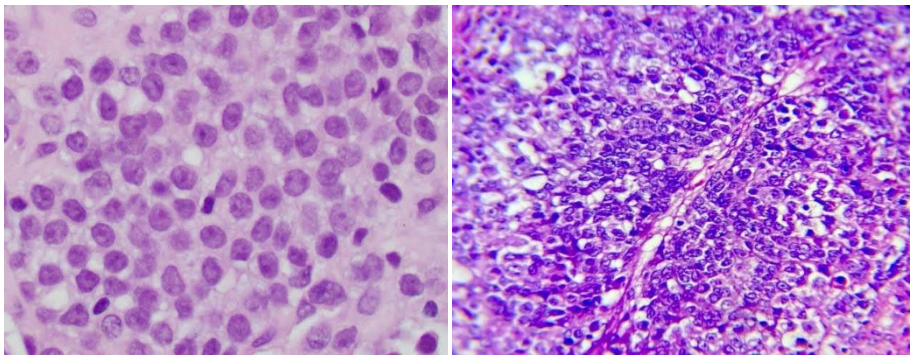


Fig-4: MRI of PNS reveals heterogenous solid mass lesion in the left nasal cavity. Mass lesion is displacing the nasal septum laterally, extending laterally into left maxillary sinus causing obstruction of ostium of left maxillary sinus and fluid collection, mucosal thickening of left maxillary sinus, posteriorly upto choana, superiorly upto sphenoid sinus.



A **B** **C**

Fig-5: Showing the removal of tumor by transnasal endoscopic sinus surgery.



A **B**

Fig-6: Histopathological report suggestive of Sections from all the & specimen shown uniform round cells with central round nucleus with stippled chromatin and moderate amount of vaculated cytoplasm. Tumor cells are PAS positive. No lymphovascular invasion is seen

DISCUSSION

ES is a rare disease, making up only 4% to 6% of all primary bone tumors [2,3]. Furthermore, ES involves the head and neck region in only 1% to 4% of cases, and tumors with a sinonasal origin form another rare subset [3]. Tumors arising in the nasal cavities and paranasal sinuses can present with nonspecific symptoms such as nasal obstruction, rhinorrhea, and epistaxis making the diagnosis of Ewing sarcoma difficult [2]. In this case, although the patient presented with symptoms mimicking chronic rhinosinusitis. The effective

treatment plan includes combined surgical excision and modern chemotherapy or radiotherapy [5]. Combined treatment has increased the overall 5-year survival rate 20 to 70% [4].

CONCLUSION

Ewing's sarcoma originating from the sinonasal tract is very rare. Although the diagnosis of the disease is challenging, it is feasible using histopathological examination, immunohistochemical study and cytogenetic analysis. Treatment includes a multidisciplinary approach with surgery followed by chemotherapy and radiotherapy.

Declaration by Authors

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