

Extraskelatal ewing sarcoma in a 62years old male smoker: a rare case presentation

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Abstract

Extraskelatal Ewing Sarcoma is rarely found in Head and Neck region. Its extraskelatal morphology is similar to the Ewing Sarcoma originating from the bone. Its occurrence in head and neck as a primary tumor is very unusual. We report a Case of Ewing Sarcoma in 62 years old male in Nasopharynx who has been a chronic smoker. It occurs predominantly in adolescents and young adults between 10-30 years of age and it follows an aggressive course with a high rate of recurrence.

Keywords: Extra Skeletal Ewing Sarcoma, Neck, Nasopharynx, Bikaner

Introduction

Extaskelatal Ewing's sarcoma (EES) first described by Tefft *et al.*,⁽¹⁾ is a very rare, rapidly growing malignant round cell tumor of uncharacterized mesenchymal origin which histologically resembles Ewing's sarcoma. It is a highly malignant tumor found mainly in children and young adults, most commonly arising from skeletal structures, especially in long bones. Primary Ewing's sarcoma of the head and neck is very rare, accounting for only 2-3% of all Ewing's sarcomas, and even rarer in the nasal cavity and/or paranasal sinuses^(2,3).

Ewing's sarcoma of the long bone is known to be a well-enhancing soft tissue density mass without calcification on the CT examination. On the MRI examination, it is known to be hypointense to isointense on T1 weighted imaging (WI) and various signals intense on T2WI with heterogeneously marked enhancement by gadolinium. Invasions of subcutaneous tissue and ill-defined bony destructive changes are common^(2,4,6). We provide an overview of Ewing's sarcoma and present a case of the Ewing's sarcoma in the nasopharynx.

Case Report

A 62 year old male presented to our OPD with complaints of Nasal obstruction, headache, occasional nasal discharge and diminished hearing for 2 months. Patient did not give any history of epistaxis. Patient has been a Chronic Smoker for last 40 years taking 1 bundle of bidis per day. On Endoscopic examination, a fleshy lobulated mass measuring about 3cm by 2cm coated with discharge found in nasopharynx extending to choana of both nasal cavities. There was no neck lymphadenopathy. Contrast Enhanced Computed Tomography Scan of Nose and PNS was performed using submillimeter thin contiguous axial plain scans. Bony window and coronal and saggital reformatted images were obtained. Hyperdense soft tissue is seen in

posterior part of bilateral nasal cavity and nasopharynx (Fig. 1) causing complete obstruction of bilateral nasopharyngeal air-way. Bilateral pterygopalatine fossa are not widened (Fig. 2). There is obliteration of bilateral torus tubaris with possible bilateral Eustachian tube obstruction and opacification of bilateral mastoid air cells.



Fig. 1: Axial Computed Tomography Scan showing homogenous non enhancing mass in Nasopharynx obstructing both choanae (10mm *6mm* 4mm)



Fig. 2: Coronal Computed Tomography Scan Showing Mass in Both Choane. Note Sphenopalatine Foramen bilaterally is not widened

The mass in nasopharynx with invasion into adjacent structures and bony destructive changes suggested malignant nature of the tumor. So, our differential diagnosis included Squamous Cell Carcinoma, Malignant lymphoma, Rhabdomyosarcoma or poorly differentiated carcinoma.

An Endoscopic assisted biopsy was taken. Grossly, specimens were irregularly fragmented myxoid and necrotic mucosal tissues.

On Microscopic examination, section shows massive areas of necrosis with infiltration by neutrophils, some tissue representing viable tissue showing infiltration by uniform size small round cells around blood vessels (pseudorosette) and shows areas of ischaemic coagulative necrosis away from blood vessels. The individual neoplastic cells are small uniform round cells having scanty cytoplasm and was PAS positive (Fig. 3, 4). The nuclei were round to ovoid having inconspicuous nucleoli. These cytological features were suggestive of malignant small round cell tumor (Fig. 3) with possibility of Ewing's sarcoma, PNET, Neuroblastoma or Rhabdomyosarcoma. Immunohistochemical panel for vimentin, desmin, chromogranin and CD99 was suggested. The present case showed strong positivity for CD99 (Fig. 5) and vimentin but was however negative for desmin and chromogranin. On the basis of histological, immunohistochemical, clinical and radiological findings a final diagnosis of Extraskelatal Ewing's sarcoma was made. Following the diagnosis, the patient was immediately put on chemotherapy and radiotherapy as EES is a radiosensitive tumor.

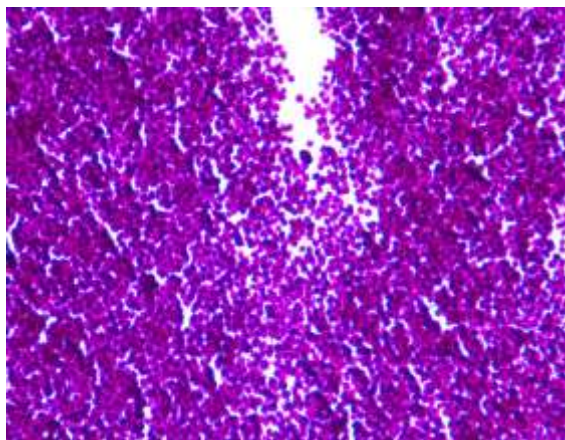


Fig. 3: showing Uniform round cells having scanty cytoplasm (PAS positive) with round to ovoid nuclei having inconspicuous nucleoli

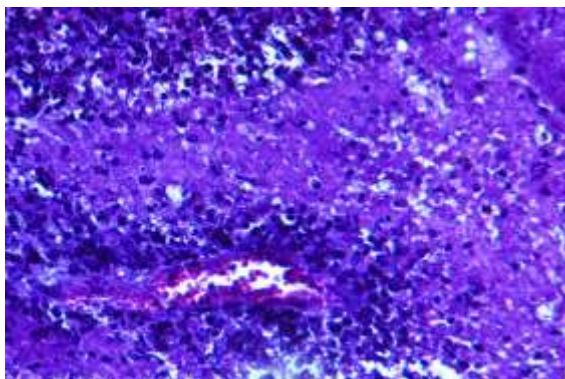


Fig. 4: Round Cell Tumor (40X) showing Pseudo Rosette

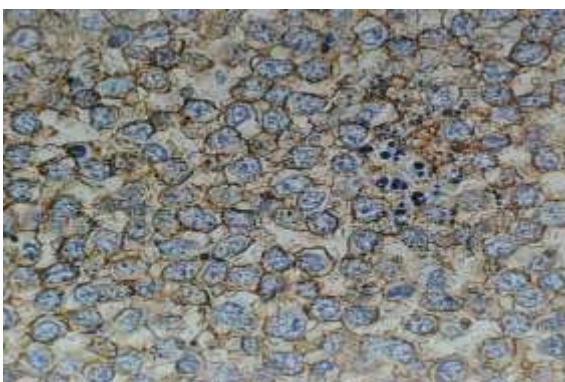


Fig. 5: Showing CD 99 positivity in Tumor Cells

Discussion

Ewing's sarcoma is highly malignant, small, round cell tumor, originated from primitive neuroectodermal cells, as first described by James Ewing in 1921. Primary Ewing's sarcoma is represented commonly in early childhood, adolescence but rarely in adulthood. There is slightly male predominance as 1.5: 1 male to female ratio^(5,7,8). Majority of the patients have t(11;22)(q24;q12), i.e., fusion between the 5' end of the EWS gene from chromosome band 22q12 with the 3'

portion of the 11q24 FLI1 gene, a member of the ETS family of transcription factor^(10,11). This EWS/ETS fusion protein blocks the differentiation of pluripotent marrow stromal cells. Rest 10-15% of the cases have t(21;22)(q22;q12) fusing EWS to a closely related ETS gene, ERG from chromosome band 21q22. In less than 1% of cases, t(7;22), t(17;22), t(2;22) and inv(22) have been found that give rise to fusions between EWS and the ETS genes like ETV1, E1AF, FEV, and ZSG, respectively. Mutations associated with P53 or P16/p14 ARF has high aggressive behavior and poor chemotherapeutic response.

Ewing's sarcoma is distinguished by two types: skeletal type and extraskelatal type. Most commonly, Ewing's sarcoma arises from skeletal structures, especially long bones (35%), and pelvis (24%). The extraskelatal type of Ewing's sarcoma usually occurs in the soft tissue of the lower extremities and the paravertebral region⁽¹⁰⁾. Thirteen cases of primary Ewing's sarcoma in the nasal cavity and/or paranasal sinuses have been reported in the otolaryngology literature^(2,8,10). In the radiology literature, only a single case report from 2003 by Harman et al.⁽⁴⁾ has been described. The prognosis for Ewing's sarcoma depends on the presence of metastases, because Ewing's sarcoma is highly malignant and metastasizes early to bones and lungs. Recently, it is accepted that prompt chemotherapy is necessary to treat occult metastasis, and a combination of surgical excision, radiotherapy and chemotherapy has significantly improved the 5-year survival rates, now reaching to 75%^(2,10). Microscopically, Ewing's sarcoma shows uniform, small, round cells with round to elongated nuclei, scanty cytoplasm and indistinct cytoplasmic borders. Hemorrhagic areas and extensive necrotic lesions are common. The essential diagnostic examination for Ewing's sarcoma among many small round neoplasms is CD99 (O13) marker, the specific immunohistochemical examination. In addition, molecular studies using PCR to detect characteristics of chromosomal translocations are definitive for the diagnosis of Ewing's sarcoma. Specific genetic hallmarks of Ewing's sarcoma is a gene sequence t(11;22)(q24;q12), which results in the fusion of the EWS gene with the FLI gene^(2,10,11).

On the CT examination, Ewing's sarcomas show similar findings regardless of the primary site. It is known to be a well-enhancing, soft tissue density mass without calcification. If a tumor includes hemorrhage or necrosis, it may be detected as heterogeneous pattern. Invasions of subcutaneous tissues and ill-defined bony destructive changes are common, and these reflect the aggressive nature of the tumor^(4,6).

Conclusion

In conclusion, primary Ewing's sarcoma must be considered, when the expansile nasal mass is detected with extensive invasion into adjacent structures and

bony destructive changes, although which is a very rare condition. Realizing the nature of Ewing's sarcoma and understanding its diagnostic significance can lead to the approach of appropriate management.

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