Abstract:
Extraosseous Ewings Sarcoma is a rare malignant small round cell neoplasm of undifferentiated mesenchymal origin. Most commonly affects children and age group less than 20. Ewings sarcoma, Extraosseous Ewings saroma, Primitive neuro ectodermal tumour(PNET), Aesthesioblastoma, Askin tumour(PNET of chest wall) are all belongs to Ewings family of tumour (EFT). Less than hundred cases are reported all over the world. t(11 22)(q24 q12) translocation is common. A 27 year male presented with swelling in the left arm for past 6 months.1 year before excision biopsy done for swelling in the same region. MRI shows swelling in the subcutaneous plane, Biopsy shows EwingsPNET. CT chest normal. Wide local excision with latismus dorsi flap and split skin graft done followed by chemotherapy. Histo-pathology report came as small round cell with scanty cytoplasm and hyper-chromatic nuclei with mitosis,CD 99 positivity suggestive of Ewings Sarcoma. On follow up for 1 year with no evidence of recurrence metastasis. 

Keyword : Recurrent Extraosseous Ewings Sarcoma - upper limb - EWS-FLI translocation - CD 99 positivity - wide local excision - chemotherapy

INTRODUCTION
Ewings sarcoma is a rare malignant small round cell neoplasm of undifferentiated mesenchymal origin. Most commonly affects children and age group < 20. Ewings sarcoma, Extraosseous Ewings saroma, Primitive neuro ectodermal tumour, Aesthesioblastoma, Askin tumour(PNET of chest wall) are all belongs to Ewings family of tumour (EFT). Extra osseous Ewings are a rare tumour entity mostly arise from paravertebral nerve plexuses, extremities, head and neck region. Only few cases are reported all over the world. Clinically Ewings sarcoma/PNET represents a spectrum of presentation of the same tumour entity. 

CASE REPORT
A 27 year old male patient, presented with compliants of swelling in the left arm
Swelling was gradual in onset and slowly progressive, not painful, ulcerates and bleeds for past 20 days. No weakness/ altered sensation of the limb. History of swelling in the same region 1 year ago for which excision biopsy done and no details available, not on follow up.

On examination, an ulceroproliferative growth of size 12x8 cm seen in the left arm, not fixed to underlying structure. Incisional biopsy shows Ewings sarcoma/PNET. MRI left arm shows tumour in the subcutaneous plane with no evidence of infiltration to the underlying deep fascia, isointense on T1w image and hyperintense with post contrast enhancement on T2w image. CT chest was normal.

WIDE LOCAL EXCISION WITH L D FLAP & SSG
Case discussed in tumour board and proceeded with surgery. Wide local excision of tumour with latissimus dorsi flap and split skin graft was done for the patient. Histopathology Report came as small round cells with scanty cytoplasm and irregularly shaped hyperchromatic nucleus with prominent nucleolus, 2-3 mitoses/HPF, large necrosis with vascular emboli – features suggestive of small round cell tumour mostly Extraosseous Ewings Sarcoma/ PNET. Immunohistochemistry shows CD 99 positivity.

Post operative adjuvant chemotherapy started for the patient – 6 cycles of vincristine, adriamycin, cyclophosphamide completed. On follow up for past 1 year, no evidence of recurrence/distal metastasis.

DISCUSSION
Extraosseous Ewings Sarcoma is a rare malignant small round cell tumour of undifferentiated mesenchymal origin belongs to Ewings Family of Tumour (EFT) which also includes Ewings sarcoma, PNET, Aesthesioblastoma, Askin tumour(PNET of chest wall). Less than hundred cases are reported all over the world so far. First reported by Tefft et al in 1969. Mostly occurs in younger age group less than 20. With males are more commonly affected than females (2:1). t(11 22)(q24 q12) translocation known as EWS-FLI translocation is present in most of the cases. Most cases have CD 99 and S 100 positivity.

Mostly involving the paravertebral nerve plexuses, extremities mostly lower extremity, head and neck region. Slow growing tumour often presents with swelling and also with pain, fever. MRI is the investigation of choice among imaging modalities. Trucut/incisional biopsy if tumour is > 3cm and excisional biopsy if tumour < 3 cm. Xray/CT chest should be taken to rule out lung metastasis.
metastasis, which is the most common site of distal metastasis if occurs. Primary surgery followed by chemotherapy / radiotherapy, if needed is the common modality of management. Large bulky/ high grade tumour needs neoadjuvant chemotherapy/ radiotherapy followed by surgery. Vincristine, adriamycin, cyclophosphomaide are commonly used. Extraosseous Ewings Sarcoma have good prognosis with 5 year survival rate of 70-80 % for stage I disease ,50 – 60% for stage II, 30–35 % for stage III, less than 10 % for stage IV disease.

CONCLUSION

Extraosseous Ewings Sarcoma is a rare slow growing round cell malignant mesenchymal tumour occurring in younger age group with good prognosis on early detection and treatment. Surgery is the main modality of treatment. Antisense oligodeoxy nucleotide drug to down regulate EWS-FLI translocation, Fenritide, a retinoid derivative to produce cell death are the newer drugs under trial for Ewings/PNET tumours.

REFERENCES


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