Abstract:

Extraskeletal Ewings sarcoma (EES) is a rare soft tissue tumor that predominantly involves the soft tissue of trunk and extremities which is morphologically indistinguishable from Ewing’s sarcoma of bone. We report a case of Extraskeletal Ewing’s sarcoma in a 14 year old adolescent female in the Paravertebral region with lymph node metastasis. Histopathological examination, Fine Needle Aspiration Cytology, Enzyme histochemistry and Immunohistochemistry for CD99 confirm the diagnosis.

Keyword: "Extraskeletal Ewing’s sarcoma, CD 99, Lymphnode metastasis"

INTRODUCTION:

Ewing’s sarcoma is a malignant small round cell tumor first described by James Ewing in 1921. Extraskeletal Ewing’s sarcoma is a rare malignant soft tissue neoplasm histologically similar to skeletal Ewing’s sarcoma first described by Tefft et al in 1969. It occurs mainly in adolescents and young adults. It accounts for approximately 5% of all soft tissue sarcomas in childhood. Extraskeletal Ewing’s sarcoma is an uncommon mesenchymal tumor and has to be differentiated from other small round blue cell tumors on the basis of histology, Immunohistochemistry and Electronmicroscopy. Extraskeletal Ewing’s sarcoma usually involves soft tissue of trunk, paravertebral region and lower extremity. We report a case of Ewing’s sarcoma in the paravertebral region with lymph node metastasis.

CASE REPORT:

A 14 year old female presented with history of swelling in the back for one month with history of rise in temperature for 15 days which was high grade, spiking in nature and she also had loss of weight. Patient complained of weakness and loss of sensation in both lower limbs for 2 days. Clinical examination revealed a swelling in the paraspinal region at D8 – D12 vertebral level which was warm, tender, firm to hard in consistency and immobile. X-ray revealed no bony lesion.
MRI SPINE:
Intraspinal, intradural-extradural mass measuring 3.3cm involving D10 –D11 vertebral body with cord displaced and compressed to right side. Mass was extending to paraspinal region measuring 7.13x11x8.16 cm through D12 –D11 & D 11 – D 10 nerve foramina on left side .Mass had a heterogeneous texture. (Fig - 1)

GROSS:
The tumor was received in multiple pieces, largest one measuring about 3 x 2 cm and the smallest one was measuring about 1 x 1cm. Cut section revealed gray white and gray brown hemorrhagic areas with areas of necrosis.

Fig 1 magnetic resonanace imaging of soft tissue mass in para vertebral region
A clinical diagnosis of left paraspinal tumor involving D10 –D12 vertebra with paraplegia was made . Patient underwent laminectomy and excision of the mass. Peroperatively the mass was found to be firm, vascular arising from paraspinal muscles measuring 8x6 cm spread to D10 –D 11 vertebral spine compressing cord. Complete excision was not possible and the mass sent for histopathology.

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fig 2 Solidly packed uniform round cells in sheets
Fig 3 Individual cells with round to oval nuclei, distinct nuclear membrane , fine chromatin , pale scanty cytoplasm
Enzymehistochemistry for PAS stain showed a cytoplasmic positivity for glycogen(Fig -4).
Fig 4 PAS stain show cytoplasmic positivity for glycogen

Immunohistochemistry for CD99 showed a diffuse membranous positivity (Fig -5) and LCA was negative (Fig -6).

Fig 5 IHC for CD 99 Sshow diffuse membranous positivity
Fig 6 IHC for LCA is negative

The diagnosis of extraskeletal Ewing's sarcoma was confirmed and chemotherapy was started. During follow up, the patient revealed a lymph node enlargement involving the upper cervical group. Fine needle aspiration cytology of the lymph node was done. Cytology of the lymph node revealed a highly cellular smear composed of a mixture of two population of cells in single and loosely cohesive clusters (Fig-7). Larger cells had abundant pale cytoplasm, round to ovoid nuclei with finely granular chromatin and one to three small nucleoli. Smaller cells had a scanty cytoplasm, irregular nuclei and dense chromatin (Fig -8).

DISCUSSION:
Extraskeletal Ewing's sarcoma is a rare malignant neoplasm of uncharacterized mesenchymal origin was first described by Tefft et al in 1969. Recent evidences from IHC, cytogenetics and molecular genetics indicate a primitive, pluripotent, neural crest cell line of origin for Ewing's sarcoma.
Extraskeletal Ewings sarcoma constitutes 1.1% of all soft tissue neoplasms indistinguishable from more common Ewing sarcoma of bone. Extraskeletal Ewings sarcoma usually involves soft tissue of trunk (chest wall) 28%, paravertebral region 22%, lower extremity 25%, pelvis and hip 14%, retroperitoneum 8%, and upper extremity 8%. Extraskeletal Ewings sarcoma is a member of Ewing family of tumors which constitute a group of small, blue round cell tumor occur in bone and soft tissue characterized by presence of translocation t (11:22) (q24:q12). The fusion of EWS gene on chromosome 22q12 to FLI-1 gene on chromosome 11q24 results in a EWS-FLI-1 chimeric protein. Extraskeletal Ewings sarcoma usually present as a rapidly growing painful tumor which spreads locally, infiltrating the muscle planes and bone. Tumor appears as a fleshy mass with areas of necrosis and hemorrhage. Light microscopy reveals solid sheets of small uniform primitive cells with round nuclei, scanty clear cytoplasm, containing PAS positive glycogen. Necrosis is common with viable cells frequently in perivascular distribution. In more differentiated ES/PNET, Homer-Wright rosettes may be identified. Immunohistochemistry for CD99 is expressed in almost all cases of Ewing sarcoma in a diffuse membranous fashion. In Summary this is a case of Extra skeletal Ewings sarcoma, a rare soft tissue tumor presented with lymph node metastasis. The differential diagnosis include other small round cell tumors like PNET, lymphoma, Neuroblastoma, Embryonal Rhabdomyosarcoma. Immunohistochemistry helps in the confirmation of the diagnosis.

References:
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