

Case Report

Cytological Diagnosis of Ewing Sarcoma – Clavicle

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Abstract

Ewing sarcoma is a rare malignant round cell tumor of the bone. It is the second most common primary malignant tumor of bone found in children. It commonly affects long bones, pelvis and ribs. An 11 year old boy presented with painful swelling at the medial end of left clavicle. X-ray revealed a diaphyseal lesion of the medial end of the left clavicle with destruction of bone and mottled appearance. FNAC was done and reported as Malignant round cell tumor suggestive of Ewing sarcoma. Histopathologically the diagnosis of Ewing sarcoma was confirmed with special stains. Ewing sarcoma affecting clavicle is uncommon. Clavicle can be often resected with no need of reconstruction in young children.

Keywords

ewing sarcoma, clavicle, cancer in children

Introduction

Ewing sarcoma is an uncommon primary malignant round cell tumor of bone, named after Ewing, who first described it in 1921¹. Despite being the second most common primary bone tumor found in children with highest frequency of the disease between 10-15 years, Ewing sarcoma is still rare^{2,3}. It accounts for 2-3% of childhood tumors and can occur in any bone, most often found in long bones of extremities, pelvis and ribs but, Ewing sarcoma affecting clavicle is uncommon, < 5%^{3,4,5,6,7}. We report a case of cytological diagnosis of Ewing sarcoma of clavicle in an 11-year old boy.

Case Report

A 11-year old boy presented with a progressively increasing swelling at the medial end of left clavicle since 1½ month. There was H/O trauma 1½ month back, after which he developed a painful swelling which gradually increased to attain the present size. The patient was sent to cytopathology section for FNAC, with no definite clinical diagnosis. On examination,

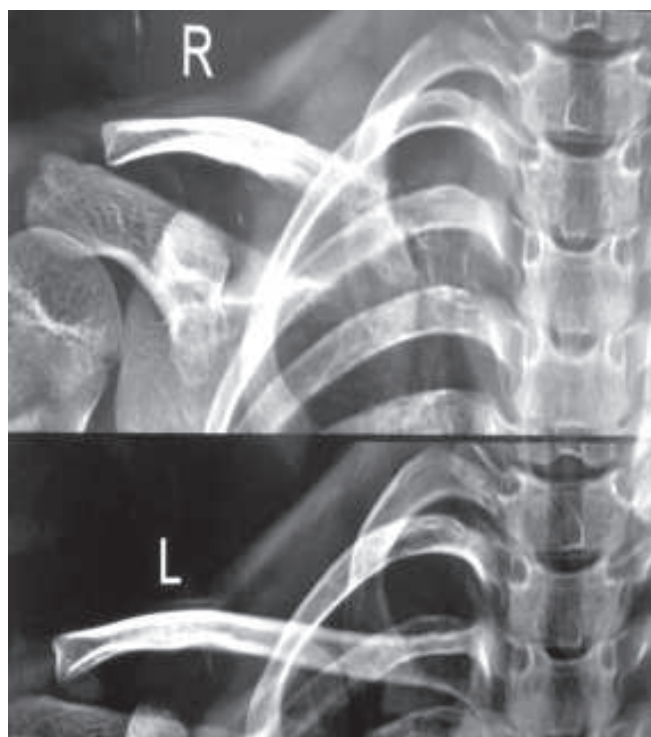


Fig. 1

X-ray showing diaphyseal tumor of the clavicle with destruction of bone and mottled appearance

a firm to hard swelling measuring 4x3 cm was noted at the medial end of the clavicle. No sign of inflammation was noted.

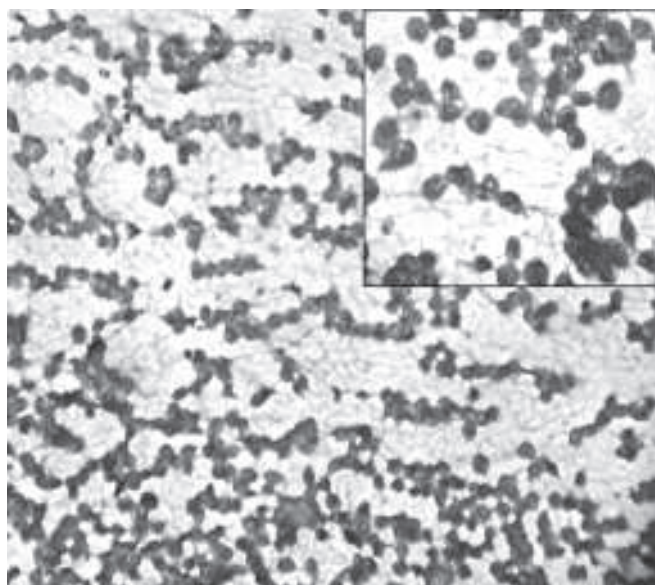


Fig. 2

Cellular smear composed of dissociated and clusters of loosely cohesive monotonous round cells with few bare nuclei. (H and E, 10X) Inset shows the rosetoid arrangement of the cells and the detailed cell morphology. (H and E, 40 X)

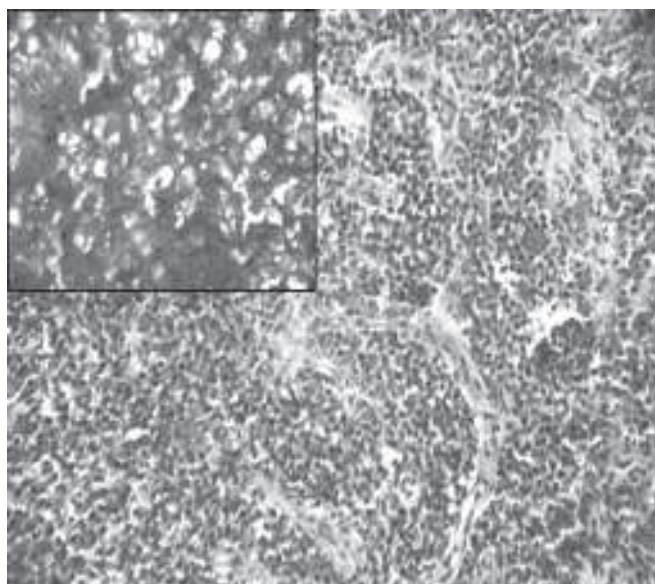


Fig. 3

Tissue section showing diffusely arranged uniformly small round tumor cells with little intercellular stroma. (H and E 10X) The inset shows the diastase sensitive PAS positive dense material demonstrating intracytoplasmic glycogen in the tumor cells (H and E 40x)

FNAC was done and the patient was sent for X-ray with a suspicion of the lesion involving the bone. X-ray revealed a diaphyseal tumor with destruction of medial end of the clavicle and mottled appearance (**Fig. 1**). Radiologic diagnosis offered was Osteoclastoma vs. Chondrosarcoma. Two H&E stained FNAC slides revealed a cellular lesion of dissociated monotonous round cells having round to oval nuclei with finely granular chromatin and inconspicuous nucleoli (**Fig. 2**). The cytoplasm was pale eosinophilic and at places vacuolated. Occasional rosette like structure was noted. Numerous round naked nuclei were seen. Considering the X-ray picture, FNAC was reported as Malignant round cell tumor suggestive of Ewing sarcoma. Biopsy was advised for confirmation of the diagnosis. The biopsy showed diffuse arrangement of uniformly small round cells with little intercellular stroma (**Fig. 3**). The cells had round to oval nuclei with finely divided chromatin and scanty ill defined cytoplasm. Presence of intracytoplasmic glycogen was demonstrated in form of dense, diastase sensitive PAS positive material. This helped to confirm the diagnosis of Ewing sarcoma. The patient was sent for radiotherapy but, we lost the follow-up of the patient who was not affording for the treatment.

Discussion

Ewing sarcoma has long been recognized as a very lethal tumor⁷. Despite being the second most common primary malignant bone tumor found in children between 10-15 years of age, Ewing sarcoma is still a rare tumor with an annual incidence of 0.8/ million of population^{2,3}. The most common mode of presentation is swelling that enlarges gradually along with pain^{4,8}. Ewing sarcoma of upper extremity is seen less frequently. Most common site in upper extremity is humerus. Clavicle is rarely involved with the incidence of <5%.^{3,4,6,7}. In our case the boy is 11 years old who presented with gradually increasing painful swelling, after trauma. Patient associated delay in diagnosis is due to symptom associated with pain and doctor associated delay is due to incorrect diagnosis of osteomyelitis^[2]. Cytological diagnosis of malignant round cell tumor of bone - suggestive of Ewing sarcoma was given on the basis of X-ray findings and morphological features⁹. Histologically the differential diagnosis of metastatic neuroblastoma, malignant lymphoma and embryonal rhabdomyosarcoma were considered. We came to make the diagnosis of Ewing sarcoma with confidence in this case because the tissue was processed

in formalin, i.e. without special precaution to provide preservation of glycogen and demonstrated the diastase sensitive PAS positive dense material; also the X-ray findings supported the diagnosis⁴. The consistent finding with Ewing sarcoma shows immunohistochemical positivity with vimentin, NSE and MIC-2 antigen^{4,5,10}. There is clonal translocation in the malignant cells between long arm of chromosome 11 and 22, which forms the fusion protein viz. EWS-FL11. This protein can be analyzed by using a small piece of tissue for PCR^{4,5}.

The treatment of Ewing sarcoma currently involves a multidisciplinary approach. In young children claviclectomy can be done with no need of reconstruction. In upper extremity, surgery along with chemotherapy has revolutionized the prognosis of Ewing sarcoma². Radiotherapy should be an adjunct to chemotherapy and surgery^{2,1,5}. Our case is unusual for the reason that clavicle is rarely involved in Ewing sarcoma. The diagnosis of Ewing sarcoma was not at all thought of clinically but we suspected it on FNAC with the collective support of other features and then it was confirmed on histopathology.

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