■ *Case Report*

Extra-skeletal (periosteal Ewing's sarcoma) Ewing's sarcoma of distal femur of right leg in a 10 year old boy- A case report

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Abstract

Extra- skeletal Ewing's sarcoma is quite uncommon.The most common locations of Ewing's sarcoma of bone include metaphysis and diaphysis of long bones and flat bones of shoulder and pelvic girdles. Here, we present a case of a 10 year-old school-going boy who had presented to the Orthopaedic Depatment of B.P.Koirala Institute of Health Sciences, Dharan, Nepal with the complaint of pain and swelling of pasterior aspect of disfal fourth of right thigh . Considering its rarity, we thought that this case needs to be reported.

Keywords: Skeletal, Periosteum, Tumor, Sarcoma, Wide excision

Introduction

Ewings sarcoma is the 4th common primary malignancy of bone, but it is 2nd most common in patients. younger than 30 years of age and most common in patients. younger than 10 years of age. Extra-skeletal Ewing's sarcoma is very uncommon.Ewing's sarcoma is a small malignant round cell tumour that arises from mesenchymal cells, predominantly in the medullary cavity of bone. In exceptional cases, it originates in the soft tissues and subsequently invades the underlying bone. A (sub) periosteal origin of Ewing's sarcoma is a very rare condition: only a few cases have been published so far.

Classically Ewing's sarcoma appears roentgeno graphically as destructive lesion in diaphysis of long bones with an onion skin periosteal reaction

Case report

A 10 year old school going boy from remote hilly area of Nepa reported to the Department of Orthopaedics, B.P.Koirala Institute of Health Sciences, Dharan with progressive painful swelling of posterior aspect of distal fourth of right thigh for 4 months. On examination, there was indistinct palpable mass in the posterior aspect of right thigh with warmth of overlying tissues.Superficial veins were dilated. Swelling was soft to firm in consistency which is fixed to underlying structures. A routine blood investigation was done. MRI scanning of mass was planned twice but couldn not be done because the child was moving during the procedure.So,FNAC of the mass was performed twice in two different centers which was suggestive of skin adnexal tumor in one center and some features of malingancy in another center but couldn't confirm that it was a case of Ewing's sarcoma. After doing pre anaesthetic check-up, mass was approached posteriorl and wide excision of tumor was done under general anesthesia and sent for histopathological examination.

Tumor was arising from periosteum of posterior aspect of distal femur which was extending into the surrounding soft tissues. Incision was closed in layers and above knee POP slab was applied. Sutures were removed on the 14th post-operative day.

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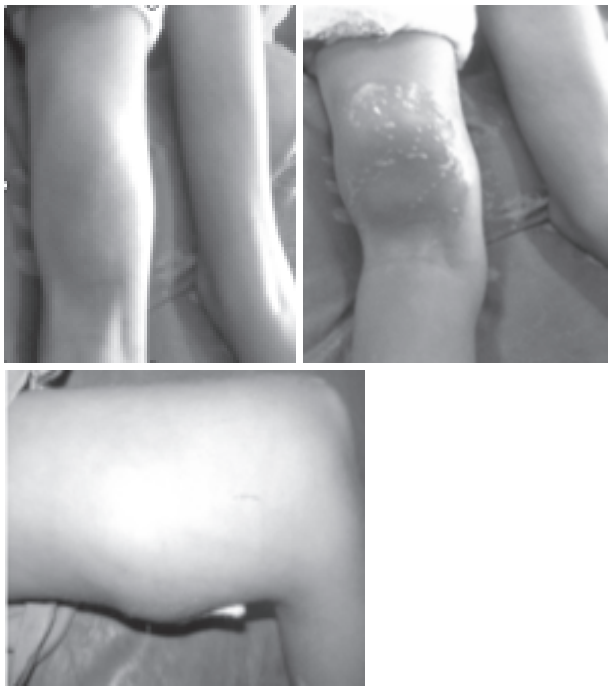


Fig:-1 Pre-operative Photographs

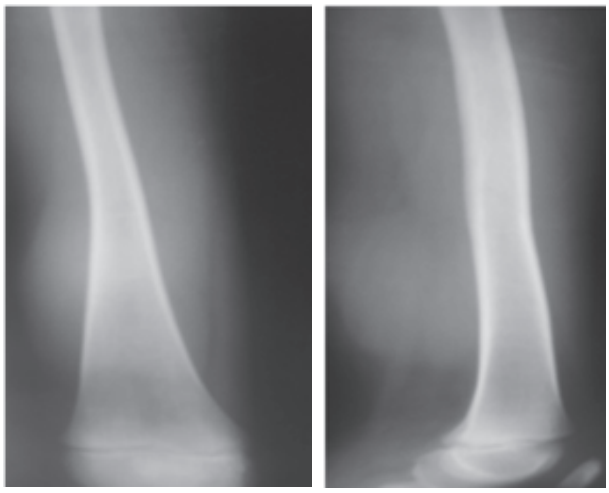
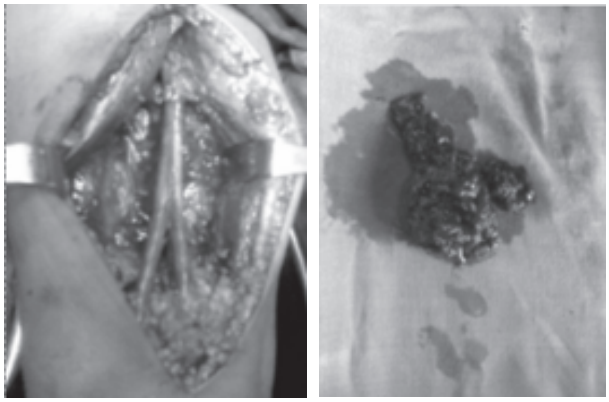


Fig:-2 X-ray findings



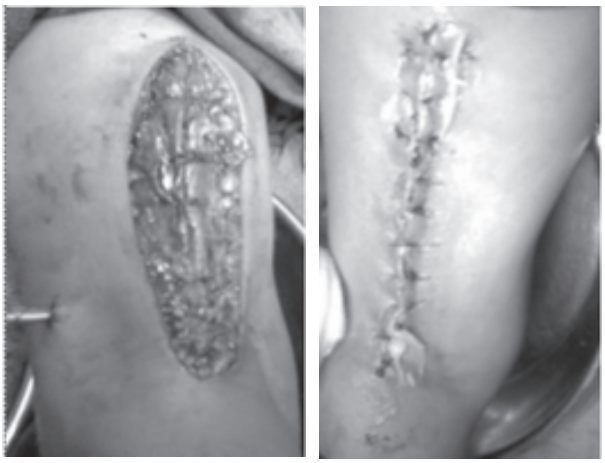


Fig:- 3 Intra-operative Photographs

Biopsy report revealed periosteal Ewing's Sarcoma. At 6th month follow-up, there was recurrence of tumor

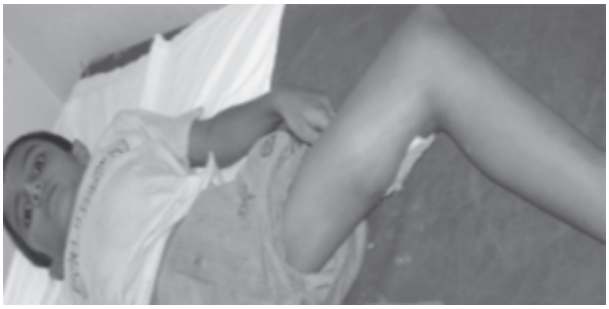


Fig:-4 Recurrence of tumor

Discussion

Ewings sarcoma is the 4th common primary malignancy of bone, but it is 2nd most common in patients. younger than 30 years of age and most common in patients. younger than 10 years of age. Extra-Skeletal Ewing's sarcoma is very uncommon. The most common locations include metaphysis and diaphysis of long bones and flat bones of shoulder and pelvic girdles. A (sub) periosteal origin of Ewing's sarcoma is a very rare condition: only a few cases have been reported so far.

Classically, Ewing's sarcoma appears roentgeno graphically as destructive lesion in diaphysis of long bones with an onion skin periosteal reaction. Skip metastases are not reported in Ewing's sarcoma. regardless of location, an MRI of the entire bone should be ordered to evaluate the full extent of lesion as well as extension into soft tissues. All patients should have a base line roentgenogram and CT scan

of chest because the lungs are the most common site of metastases. If possible, bone scan should be performed

The worst prognostic factor is presence of distant metastases. Even with aggressive treatment, the patients with metastases have only a 20% chance of long-term survival. Fever, anaemia and elevated laboratory values(WBC, ESR, LDH) have been reported to indicate more extensive disease and therefore worst prognosis. Older age at presentation and male gender also have been reported with worst prognosis.

There are various treatment modalities described in literature. The treatment of Ewing's sarcoma must include neoadjuvant or adjuvant chemotherpy, or both to treat distant metastases. Before the use of multiagent chemotherapy, long term survival was less than 10%. Today most centers are reporting longterm survival rates between 60-70%.

Local treatment of the primary lesion is more controversial. Ewing's sarcoma is very sensitive to both chemotherapy and radiotherapy. It melts like ice with chemotherapy. There is incrased rate of overall survival with wide resection of the primary tumor. Large, central, unresectable tumors are treated with radiation, whereas smaller, more accessible tumors have better prognosis when treated surgically.

Disease relapse is associated with a poor prognosis despite aggressive treatment of the relapse with further surgery, radiation and chemotherapy. Patients who have relapsed within first year of surgery have a worst prognosis than those who have an extended disease- free interval.

Summary

Ewings sarcoma of bone is the 4th common primary malignancy, but it is 2nd most common in patients. younger than 30 years of age and most common in pts. younger than 10 years of age. Extra-Skeletal Ewing's sarcoma is very uncommon.

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