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A Case of Distal Femur Primary Osteosar coma in an adolescent female managed with neoadjuvant chemotherapy, Excision of tumor with Above knee amputation and post operative Adjuvant Chemotherapy

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Abstract

Introduction: Osteosarcoma is the most common primary bone cancer. It has bimodal age distribution having first peak in teen age group while next peak is in adults aged more than 60.

Osteosarcoma can occur in any bone in body, but most common site is around the knee joint, either at the femur, or the tibia. MRI is the investigation of choice while gold standard investigation is excision biopsy. Available modalities for treatment consist of combination of neoadjuvant and adjuvant chemo therapy, and excision of tumor.

Case report: An 16 years old female presented with swelling, pain and difficulty in walking since 3 months. MRI report suggested a heterogenous mass arising from distal metaphysic of femur most likely to be osteosar coma. Patient is managed by neo adjuvent chemotherapy with wide margin excision of tumour with above knee amputation.

Post excision specimen biopsy was sent for his to patho logical examination which confirmed the diagnosis of Osteosar coma.

Conclusion: Osteosarcoma is an aggressive bone tumour and should be detected early. Early MRI with detection of tumour plays an important role in prognosis of the patient. Chemo therapy is critical to achieve a cure for patients with osteosar coma.

Resection of the of the tumour mass and associated soft-tissue mass should be performed with the goal of adequate bone and soft tissue margins. It is clear that negative resection margins allow a higher chance of local control.

Wide local excision of tumour mass with amputation gives good outcome with results comparable with other

modalities of treatment. Amputation provides good margin resection and helps in preventing recurrence with reduced chances of secondary mutation.

Keywords: Osteosar coma, Above knee amputation, Bone tumour, Excision.

Introduction

Osteosarcoma, while rare, is the most common primary bone cancer and accounts for up to 10% of all new pediatric cancer diagnoses annually [(1)]. Osteosarcoma is a carcinoma of osteoid forming mesenchymal cells, whose etiology is not known.

It is the most common primary bone cancer of childhood. Less frequently, it occurs in adults where it represents secondary malignant degeneration of primary bone pathology. Osteosar coma has a bimodal age distribution. First peak is observed in pubertal age group and next observable peak is in adults older than 65.

The patient's age has been found to correlate with survival; the poorest survival is among older individuals. The incidence of osteosarcoma has historically been reported as higher in males than in females, with an incidence rate of 5.4 cases per year per million males and 4 cases per year per million females

Osteosarcoma often begins around the knee joint, either at the femur, or the tibia. The next most common place osteosarcoma begins is in the humerus.

It is highly hetero geneous in its manifestation, which permits division into several subtypes according to the degree of differentiation, location within the bone, and histological variation. These subtypes vary in imaging appearance, demographics, and biological behavior. [(2)]

MRI is needed to evaluate the lesion's invasion into the soft tissue and neurovascular structures [(3)]. PET scans can be used to assess the primary tumour and metastatic sin other bones and the lungs [(4)].

Available modalities for treatment consist of combination of neoadjuvant and adjuvant chemotherapy, and surgical resection[(5,6)].

The goal of surgery is complete resection of disease with wide excision of the tumour. Surgery may include either limb salvage surgery or amputation to remove the involved bone [(7)].

Case presentation

A 16 years old female presented with complaints of swelling over right distal third of thigh since last 4 months, pain and difficulty in walking since 3 months. Patient gave history of fall from standing height 6 months back for which patient did not seek any treatment.

Later, patient developed swelling over distal third of right thigh. Swelling was insidious in onset and gradually progressive in nature. Patient also complains of pain over distal thigh which was insidious in onset, progressively increasing in intensity, aggravates on walking and reduces on rest. Initially it used to subside on medication, but later it did not subside even with medication.

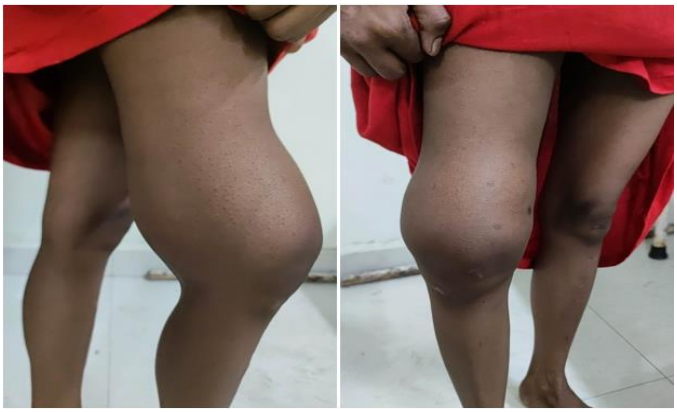
Patient was initially able to walk comfortably, but later patient started walking with limp and needed support to walk. Patient gives history of weight loss since last 4 months along with history of intermittent fever.

On examination patient looked pale, her built was thin, had generalized malaise, afebrile with pulse rate of 88beats/min, Blood Pressure 114/70 mmHg, temperature 37.8 C and SpO2 99% (in room air).

On physical examination, Patient was walking with a limp. Diffuse tender swelling was present circum ferentially over distal third of thigh, which was firm in consistency, non-mobile, arising from the underlying bone having rough surface and irregular margin (Figure

1). Range of motion at knee joint was painful and terminally restricted.

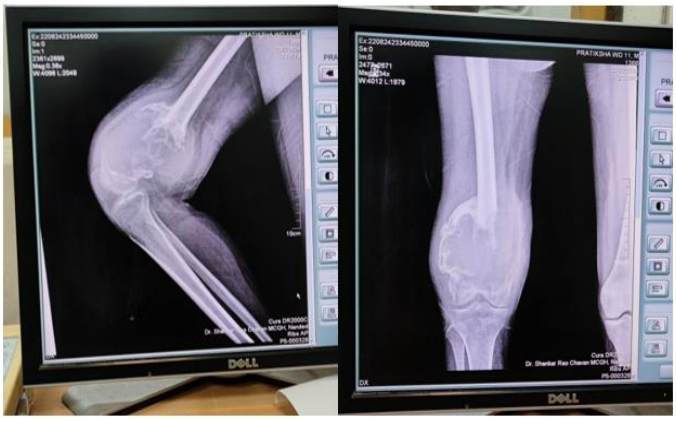
Figure 1



Investigations

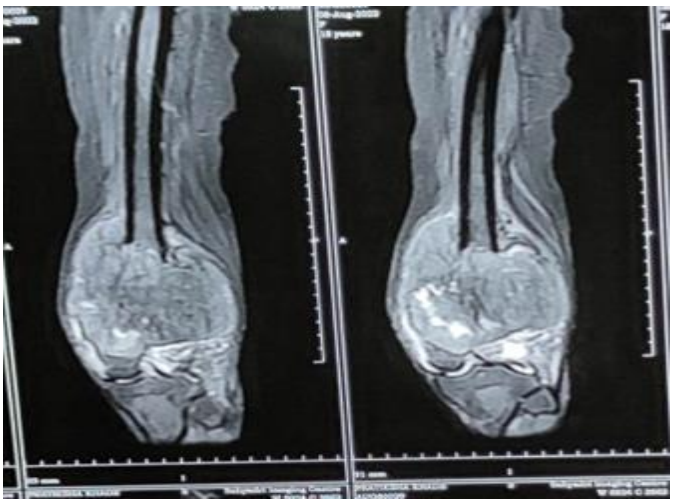
Blood investigations showed, Hemoglobin- 10.2gm%, Total leukocyte count- 19000(Lymphocytes 40 %), ESR-100mm/hr- CRP 106, Calcium 9.6 mg/dl, Alkaline phosphatase- 220 IU/L and Lactate dehydrogenase 460 IU/L.

Figure 2



Xray of the distal femur show Ed medullary and cortical bone destruction with typical moth-eaten appearance (Figure 2). There was aggressive periosteal reaction with sunburst appearance along with Codman's triangle.

Figure 3



Magnetic resonance imaging (MRI) (Figure3) showed A large well defined hetero geneously hyper intense eccentric cortex based osteolytic mass lesion involving anterior and medial cortex of distal metaphysis of femur protruding soft tissues on medial aspect.

This favours neoplastic etiology, most likely an osteosarcoma. The lesion is closely abutting distal third of superficial femoral neurovascular bundle on its posteromedial aspect.

Management

After completion of neo-adjuvant chemotherapy, and Anaesthetic fitness patient was posted for wide excision of tumour with above knee amputation (Figure 4). Biopsy specimen taken and sent for histopathological examination which confirmed the diagnosis of Osteosar coma.

Biopsy report showed tumour cells which were spindle, oval shaped with hyperchromatic nuclei with scanty cytoplasm with fibroblastic features (Figure 5).

Post-operative management and follow up

Post operatively patient was given adjuvant chemotherapy. Suture removal was done on 14th post operative day. There was no evidence of soft tissue involvement. Suture line was healthy and there were no

signs of infection or inflammation. Later, patient was advised to use Jaipur foot for rehabilitation.

Figure 4

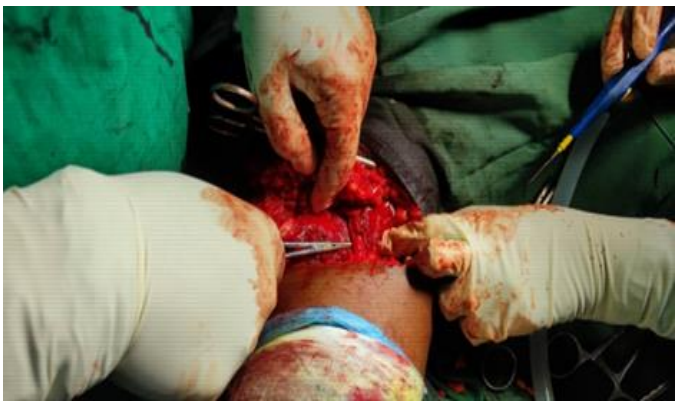
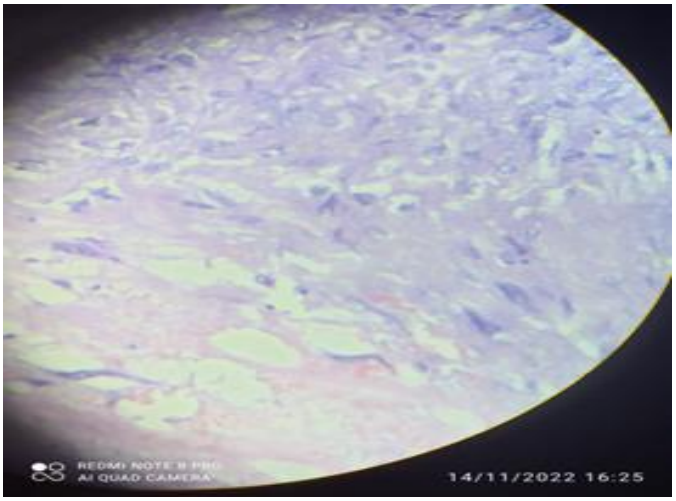


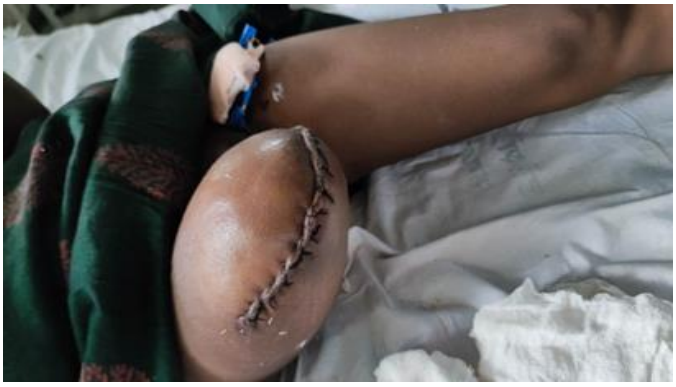
Figure 5



post operatively

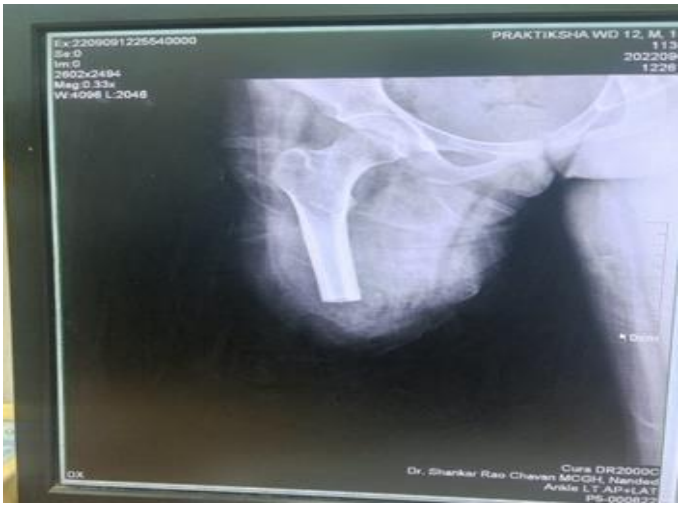
(Figure 6),The suture line is healthy ,no collection , no local rise of temperature.

Figure 6



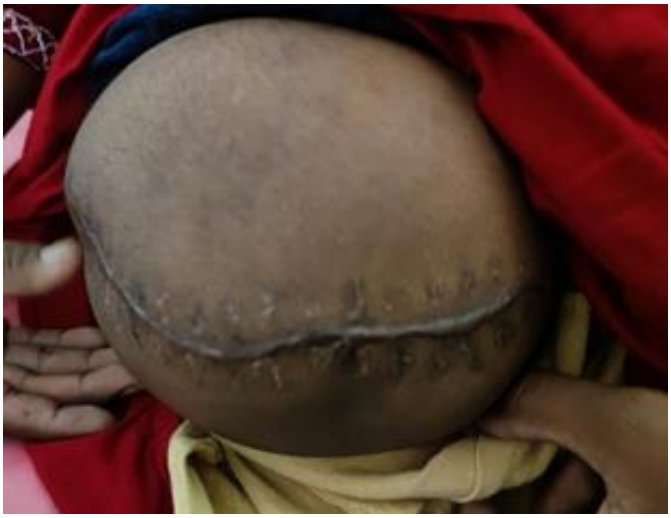
Post op x-ray,

Figure 7



On Follow up after 3 months, there is a healthy scar (Figure 8) with no collection and no any localised swelling with no local rise of temperature

Figure 8



Discussion

Osteosarcoma is an aggressive malignant neoplasm that arises from primitive transformed cells of mesenchymal origin and that exhibits osteoblastic differentiation and produces malignant osteoid [8]. Osteosarcoma is often high grade and originates in the intramedullary cavity. On x-rays Osteosarcoma can be osteolytic or osteoblastic, or both. Most of cases are located in the metaphysis of long bones, but Osteosarcoma can also

arise in the diaphysis of long bones as well as the axial skeleton [9].

Osteosar comas are diagnosed with the help of MRI (investigation of choice), CT scan which helps in delineating the tumour and biopsy confirmation after complete excision.

In this case patient gave history of trivial fall following which patient did not seek any medical advice. Patient did not have any complaints before this incident. Within few months patient started having swelling and pain in her leg. This suggests rapid growth and spread of the tumour and its aggressive nature. After MRI patient was advice chemo therapy [10,11]. Preoperative chemo therapy offers several advantages such as,

- Chemotherapy induces the development of a well-formed avascular, collagenous capsule and contributes to the improvement of the quality and adequacy of the surgical margin. Osteosarcomas which are pre-treated by chemotherapy are better demarcated against surrounding tissues and easier to operate on.
- Preoperative chemotherapy allows histological evaluation of the response to treatment, one of the most reliable prognostic indicators available to date [12].

After Chemotherapy patient was put up for wide local excision of tumour along with above knee amputation followed by replacement orthosis (Jaipur foot).

Other modalities of treatment for tumour were limb sparing surgery, resection with reconstruction.

Conclusion

Osteosarcoma is an aggressive bone tumour and should be detected early. Early MRI with detection of tumour plays an important role in prognosis of the patient. Chemotherapy is critical to achieve a cure for patients with osteosarcoma. Resection of the of the tumour mass and associated soft-tissue mass should be performed with the goal of adequate bone and soft tissue margins. It is clear that negative resection margins allow a higher chance of local control. Wide local excision of tumour mass with amputation gave results comparable with other good modalities of treatment. Amputation provides good margin dissection and helps in preventing recurrence with reduced chances of secondary mutation.

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