A Case Study On Post Operative Re-Occurrence Of Telangiectatic Osteosarcoma

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Abstract: *Telangiectactic osteosarcome is the rare variant of osteosarcoma. It has a major metastasis foe the lungs, brain and other vital organs which leads to fatal. The prognosis for this patient is very poor because of recurrence of osteosarcoma post operatively. Here this patient has the greater risk of metastasis for lungs. The prognosis rate for this patient is very compared to the conventional osteosarcoma. Adjuvant chemotherapy is advised with the patients with metastatic disease. With the clinical application of neoadjuvant chemotherapy and the clinical improvements in Limb Salvage Surgery, the tumor free survival rate of osteosarcoma has significantly improved. However, recurrence and metastasis occur in 1/3rd of affected patients and the treatment for this patient remains a challenge*

Key Words: *Osteosarcoma, Chemotherapy, Radiotherapy, Telangiectactic.*

1. INTRODUCTION:

Osteosarcoma is the most common primary malignant bone tumour that produces immature bone, usually found at the end of long bone often around the knee. Major sites of involvement of osteosarcoma are the metaphyseal areas (91%) of long bone of the the extremities with its occurrence in (descending order), lower end of femur, upper end of tibia, upper end of humerus and upper end of femur, diaphysis (uncommon)[1]. In some patients, the radiation therapy is advised, when patients doesn't respond to complete surgical excision[2].The neoadjuvant chemotherapy (NATC) plays a pivotal role in the improvement of clinical outcome in osteosarcoma individuals, the re-occurrence of osteosarcoma remains a common post-operative complication, often results in treatment failure.

The exact cause and mechanism of Telangiectatic Osteosarcoma (TOS) formation is unknown. Some of the causes of TOS are, the tumor can occur de novo or spontaneously (termed as a primary osteosarcoma), or due to some pre existing condition and abnormalities (termed as a secondary osteosarcoma). Such pre existing conditions could include radiation exposure, genetic anomalities, and any well established physical injury or trauma. A radiation influenced tumor takes many years to develop, after the radiation exposure has ceased. These radiation have been previously administered for treating other cancers.

2. CASE PRESENTATION :

The present study restrospectively analysis a case study with recurrent telangiectatic osteosarcoma (TO). Intially the has diagnosed and undergone surgical procedure (Limb salvage surgery, Debridement of necrotic skin and SSG) in Nizams Institute of Medical Sciences (NIMS) and recurrence of TO was detected in Sushrutha hospital where the patient is treated with Radiotherapy (RT).

A 14yr old attended in NIMS presented with complains of swelling of right lower limb just above the knee since past 2 months. With these complaints, he was diagnosed with MRI of knee, which was suggestive of osteosarcoma. Futher work up showed localized disease. He is admissioned for cycle I NACT, O/E PS1, vitals – stable.
L/E : 10x6cm swelling on upper part of right tibia, Biopsy scar +
No discharge, sinuses, heart and lungs – Normal, per abdomen – soft,
CNS – No focal neurological deficits

MRI

- Altered marrow signal in the proximal metadiaphyseal region of right tibia and reaching upto the proximal physeal plate with periosteal elevation in the posterior, medial and anterio medial aspects of the proximal metadiaphysis of tibia with subperiosteal soft tissue along with multiple cortical erosions as mentioned – possibily suggesting neoplastic etiology like osteogenic sarcoma.
- Focal marrow edema in the posterior – medial part of proximal tibial epiphysis without any obvious bony erosions.
- Soft tissues edema around the distal attachment of medical collateral ligament with its indentation by soft tissue lesion. Subcutaneous edema in the anterio medial aspect of upper 3rd of leg.

- Mild ill defined STIR hyperintense signal in midshaft of both right and left tibia without any signal changes in T_1 and T_2 – could be due to stress related changes.

Histopathology report

Sections from the biopsy submitted show fragments of a lesion along with few normal bony trabeculae. The lesion is comprising of polygonal to spinal shaped cells in osteochondroid matrix. There is a formation basophilic lacy osteoid with irregular contours lined by similar lesional cells. The lesion is seen to surrounding normal bony trabiculae suggest – **Osteosarcoma**

Bone Scintigraphy

Intense focal osteoblastic activity in right tibia as described – conistant diagnosis of osteosarcoma

NACT taken for 3 cycles

Cycle	No. of days	Drug	Dose	Dose based on BMI
Cycle I	2	Cisplpatin Andiamycin	75mg 45mg	60mg/m^2 37.5mg/m^2
Cycle II	2	Cisplpatin Andiamycin	75mg 45mg	60mg/m^2 37.5mg/m^2
Cycle III	2	Cisplpatin Andiamycin	75mg 45mg	60mg/m^2 37.5mg/m^2

- Reviewing the patient after 1month, the patient C/o of pain in right knee joint, limping present.

O/E : Patient conscious, oriented, GC – fair, PS – I , afebrile, PR – 74/min, BP – 110/70mmhg, No icterus/cyanosis/pedaledema/lymphadenopathy. No pallor.
Chest : clear
P/A : Soft, non tender, no mass/free fluid, no organomegaly, BS+.
P/R : NAD.
Local examination – 10x6cm swelling medial aspect right knee joint, upper end of tibia. Tenderness present, movement restricted.
Clinical Diagnosis : Osteosarcoma of right proximal tibia (post NACT).
Investigations : Blood group – A +ve, Hb – 13.9mg/dl, PCV – 40.9%, TLC – 15700/mm, PC – 2.3lakh/mm, N-88, L-6, M-5, E-1, Blood urea – 26.0mg/dl, Serum creatinine – 0.99mg/dl.
HPE – Features are consistent with osteosarcoma.
Treatment given (Surgical) – Limb salvage surgery
[Tumor resection and custom made endoprosthetic replacement right knee + medial gastronemous flap coverage].

- 15days after surgery, again the patient is experiencing severe pain, swelling in right knee. So, on examination of histopathological reports..,

Microscopic Description

Multiple sections studied from the specimen submitted. Sections show a lesion occupying the emtaphysis as well as diaphysis. The lesion is comprising of solid areas interspersed with large blodd filled cystic spaces. The solid areas show spindle to polygonal cells with marked pleomorphism and hyperchromasia. Abundant tumor osteoid production is seen which has eosinophilic and basophilic lace like appearance is rimmed by tumor cells. Entrapment of the pre existing bony trabeculae is also seen. Necrosis is not significant. The lesion is reaching upto the articular cartilage. There is also cortical breach with soft tissue extension. The posterior soft tissue margin is involved by tumor.
Impression: Features are consistent with telangiectatic osteosarcoma, yPT1N0.
Treatment given (Surgical) : Debridement of necrotic skin and SSG.
After 2 months due to monetary issues, they had visited sushrutha cancer hospital presenting C/o of pain and swelling with restricted in movements they had suggested X- ray

X-ray Right leg AP view

F/UC/O Telengiectatic osteosarcoma of tibia, post OP with post internal fixation.

3. DISCUSSION:

The patient's characteristics of telangiectactic osteosarcoma were summarized in this study. It has a higher incidence in males. Patients with late recurrent telangiectatic osteosarcoma are extremely rare, accounting for only 1% of the total osteosarcoma case. The higher rate of pathological fracture may be attributed to the massive bone destruction

associated with telangiectatic osteosarcoma. Currently, despite advanced imaging and biopsy techniques, the accurate and early diagnosis of telangiectatic osteosarcoma remains difficult, particularly regarding the differentiation of telangiectatic osteosarcoma from aneurysmal bone cyst. The need to improve the diagnostic accuracy of TOS is obvious and urgent. Pathological results have been regarded as the gold standard for diagnosing TOS.

With the clinical application of neoadjuvant chemotherapy and the clinical improvements in Limb Salvage Surgery, the tumor free survival rate of osteosarcoma has significantly improved. However, recurrence and metastasis occur in 1/3rd of affected patients and the treatment for this patients remains a challenge.

4. RECOMMENDATIONS:

Figure: 1 Bone scan showing osteosarcoma of right tibia

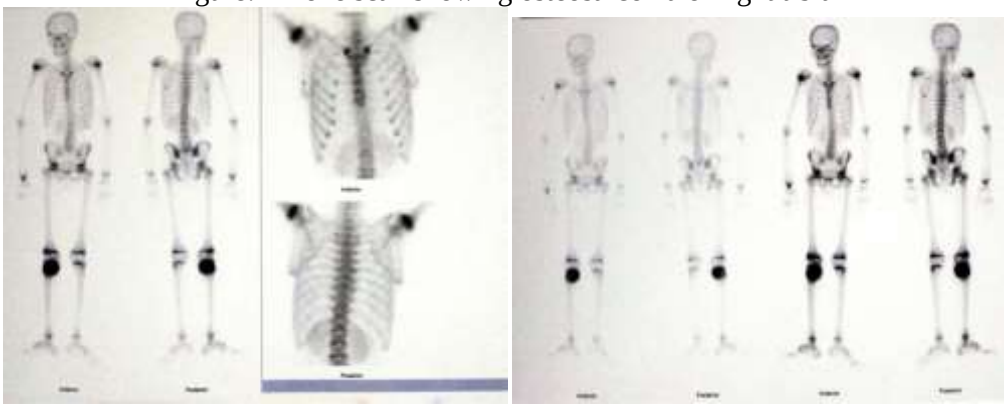

Figure 2: Bone scintigraphy showing intense focal osteoblastic activity of right tibia describing osteosarcoma with internal fixation

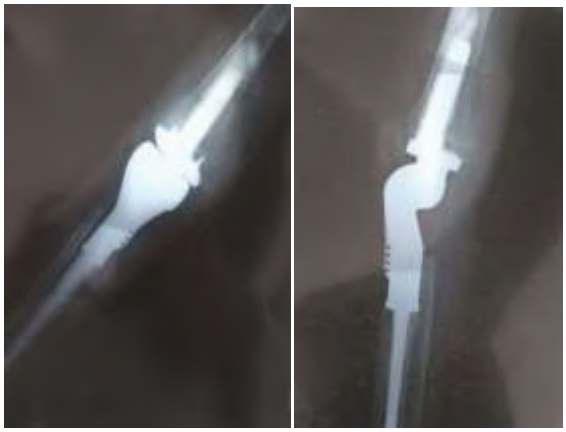

Figure 3: X-ray done 2 months after surgery showing Telangiectactic osteosarcoma, post OP with internal fixation

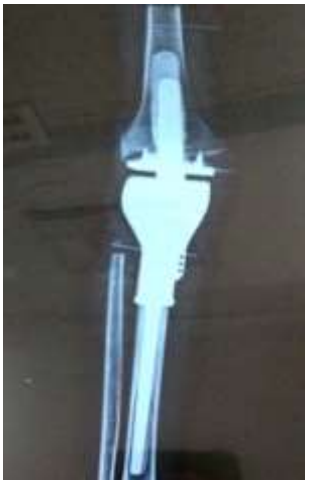

Table 1: Shows three cycles of neo adjuvant chemotherapy

Cycle	No. of days	Drug	Dose	Dose based on BMI
Cycle I	2	Cisplatin Andiamycin	75mg 45mg	60mg/m^2 37.5mg/m^2
Cycle II	2	Cisplatin Andiamycin	75mg 45mg	60mg/m^2 37.5mg/m^2
Cycle III	2	Cisplatin Andiamycin	75mg 45mg	60mg/m^2 37.5mg/m^2

5. CONCLUSION:

Most of the patients with TOS can be cured with neoadjuvant chemotherapy plus surgery (limb sparing surgery is possible and safe). Tumour responds to chemotherapy as induced necrosis was the only significant prognostic factors on survival, even if small tumour volume at diagnosis correlates with better prognosis at univariate analysis.

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