University Journal of Surgery and Surgical Specialities

ISSN 2455-2860 **2019, Vol. 5(9)**

A CASE OF TELANGIECTATIC OSTEOSARCOMA OF HUMERUS - A CASE REPORT

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Abstract : INTRODUCTION Telengiectatic osteosarcoma is an uncommon variant of osteosarcoma constituting 0.4- 12 of osteosarcomas. They were presumed to originate from transformed osteoblasts or stem cells of mesenchymal origin. Male female ratio of 21.Most common age group is 10- 30 years.CASE REPORT 34 year old male came with pain and swelling over the right arm for six months with insiduous onset increasing in size for the past 1 month. On inspection diffuse swelling noted over whole right arm, skin over swelling stretched and shiny, 201 cm surgical scar seen over the anterolateral aspect of right arm and enlarged veins seen .On palpation, Warmth, tender, Variable in consistency. Range of motion of shoulder and elbow painfully restricted with no distal neurovascular deficit. During time of presentation patient had undergone two previous surgeries done outside. Patient presented to our hospital after second surgery, 2 months later with increase in pain and swelling. Pre operative evaluation done with X ray Rt arm AP Lateral, CT chest CECT abdomen done to identify primary and to rule out metastasis which were negative. FNAC was positive for malignant cells. Bone scan showed no skip metastasis other than humerus. During the course patient develped bleeding from the tumor site spontaneously. Tumor has been staged Enneking Stage IIB. Since limb salvage is not possible at this stage Shoulder disarticulation was done. CONCLUSION With Neo- adjuvant Chemotherapy and Surgery ,survival rates for TO better than conventional OS. If treated only with surgery, prognosis worse than conventional OS. Early diagnosis and prompt treatment important for disease free survival. So staged work up is always essential in an osteolytic lesion. Always take a Biopsy before going for any definitive procedure.

Keyword : Telengiectatic osteosrcoma, Enneking staging, Limb salvage, shoulder disarticulation

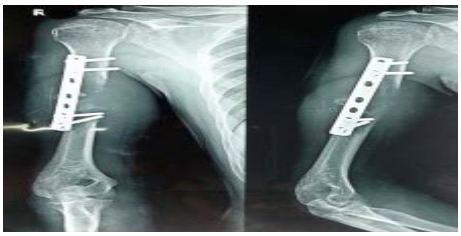
INTRODUCTION:

Telengiectatic osteosarcoma is an uncommon variant of osteosarcoma constituting 0.4- 12% of osteosarcomas. They were presumed to originate from transformed osteoblasts or stem cells of mesenchymal origin. Male : female ratio of 2:1.Most common age group is 10- 30 years. Clinical presentation resemble conventional osteosarcoma with local pain,soft tissue mass and Pathological fracture. Site of lesion may be osseous or extraosseous .Most common in metaphyseal region(90%) , diaphyseal(10%).Region of occurence in descending frequency are distal femur, proximal humerus ,proximal tibia, proximal femur, fibula, mid femur & mid humerus. Extra osseous sites are soft tissues of forearm, thigh and popliteal fossa. Radiological findings may be purely lytic, frequently permeative ,may have fluid filled spaces, cortical destruction and infiltrartion ,Periosteal bone reaction with codman triangle, pattern of parallell striations highly suggestive of it.Differential diagnosis is Aneursmal bone cyst. Histologic findings gross appearance of hemorrhagic mass, Multicystic channels filled with blood resembling ABC. Microscopic features include malignant cells in a background of blood and necrotic debris, large blood lakes alike ABC, lining of blood lakes show overt malignant cells differentiating from ABC and numerous giant- cells seen resembling Giant cell rich Osteosarcoma.

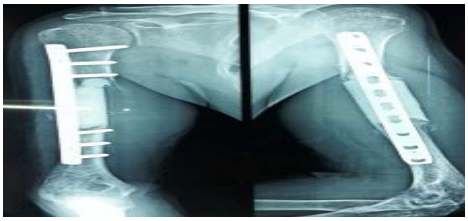
Case report:

34 year old male came with pain and swelling over the right arm for six months with insiduous onset increasing in size for the past 1 month. On inspection diffuse swelling noted over whole right arm, skin over swelling stretched and shiny,20*1 cm surgical scar seen over the anterolateral aspect of right arm and enlarged veins seen .On palpation, Warmth, tender, Variable in consistency. Range of motion of shoulder and elbow painfully restricted with no distal neurovascular deficit. During time of presentation patient had undergone two previous surgeries done outside.

INITIAL X-RAY WITH OSTEOLYTIC LYTIC OVER DIAPHYSIS OF HUMERUS



X -RAY 2 MONTHS AFTER SURGERY WITH EXTENSIVE OSTEOLYTIC LESION OVER HUMERU

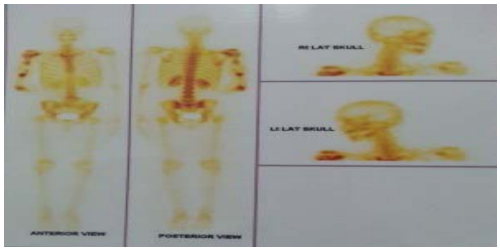


X-RAY SECOND SURGERY WITH CEMENT SPACER AND LOCKING PLATE

Initial X-ray shows expansile lytic lesion over diaphysis ,Cortical thinning, no cortical breach without periosteal reaction. Surgery has been done outside with Biopsy and fixation with Narrow DCP. Initial biopsy report shows, small sheets of malignant tumor cells with extensive areas of haemorrhage and necrosis suggestive of Malignant Neoplasm . Patient had no symptomatic relief of pain and during follow up after 2 months x-ray was taken which shows osteolyic lesion over diaphysis of humerus with plate in situ. Patient had undergone another surgery outside with implant exit, cement spacer and fixed the humerus with locking plate. Biopsy after second surgery shows, tumor composed of diffuse sheets of stromal cells with nuclear pleomorphism, abnormal mitoses, area of necrosis, spindle shaped cells and occasional osteoclastic giant cells with few nuclei suggestive of Malignant Giant cell Tumor Patient presented to our hospital after second surgery,2 months later with increase in pain and swelling. Pre operative evaluation done with X ray Rt arm AP & Lateral, CT chest & CECT abdomen done to identify primary and to rule out metastasis which were negative. FNAC was positive for malignant cells. Bone scan showed no skip metastasis other than humerus.

PATIENT AT THE TIME OF PRESENTATION TO US

X -RAY AT TIME OF PRESENTATION



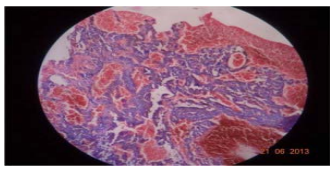
BONE SCAN AT TIME OF PRESENTATION

FUNGATING TUMOR AT TIME OF SURGERY

During the course patient develped bleeding from the tumor site spontaneously. Tumor has been staged Enneking Stage IIB. Since limb salvage is not possible at this stage Shoulder disarticulation was done.

GROSS SPECIMEN SHOWING DESTRUCTIVE OSLEOLYTIC LESION

Patient follow up after 1 year



MICROSCOPIC PICTURE SHOWING OSTEIOD AND SINUSOIDAL VASCULAR SPACES

Histopathology report of the amputated specimen, macroscopically shows the extent of tumor over diaphysis of humerus with two osteolytic lesions seen each measuring 5*3*2 cm over humerus proximal and distal to the cement spacer. The articular margins over the shoulder is free of malignant cells. Cut section appears haemorhagic and greyish white areas. Microscopy shows tumor tissue with increased vascularity ,tumor cells arranged in rows with marked pleomorphism and areas of haemorrhagic necrosis and osteoid formation seen, suggestive of Telangiectatic Osteosarcoma. Microscopy section of margins are negative for amlignant cells. Post operatively Medical oncologist opinion was obtained and

Adjuvant chemotherapy was started. High dose chemotherapy started with Cisplatin 120 mg and Doxorubicin 90 mg every 3 weeks for 6 cycles. Patient was followed up every 3 weeks. After 6 cycles of chemotherapy patient is free of disease and pain free , X- ray and CT scan of chest showed no metastasis and patient general condition was good. At 1 year follow - up patient was pain free and there is no evidence metastasis.

Discussion:

Matsuno et-al Criteria for Telangiectatic sarcoma includes Roentgenogram shows lytic destructive lesion with no area of sclerosis a cystic, cavitary gross specimen with little solid tumor tissue and no areas of dense bone. Histologically, a tumor with single or multiple cystic cavities, containing blood or necrotic tissue, with septa composed of anaplastic tumor cells. Radiographic findings of geographic margins, cortical bone destruction, and absence of matrix calcification may resemble benign lesions such as GCT or ABC. With the advent of Neo-adjuvant chemotherapy and Limb salvage surgery continuous disease free survival is better. Most recoomendations for osteosarcoma include 2-3 cycles Neoadjuvant chemotherapy,2-3 weeks later surgery (either biological reconstruction or non -biologicl reconstruction) followed by 2 -3 weeks later with 2-3 cycles of adjuvant chemotherapy. In our institution we follow IAP schedule (Ifosfamide,Adriamycin,Cisplatin),2-3 cycles of Neo-adjuvant Chemotherapy, followed by surgery and 2-3 cycles of Adjuvant chemotherapy post - operatively. Since Telengiectatic type of Osteosarcoma is a chemosensitive tumor, Limb salvage could be a viable option to this patient if patient presented to us early.

Surgical management depends on Tumor location, stage of disease, tumor response to neoadjuvant chemotherapy. With advent of Neo-adjuvant chemotherapy micrometastasis of tumor is prevented. Based on tumor necrosis factor after chemotherapy ,if tumor necrosis is >90% response is graded as good and we can do either biological reconstruction (vascularised graft, Non-vascularised graft, allografts)or with custom mega prosthesis like shoulder endo-prosthesis. Goal of surgery is complete resection of tumor with wide margins. In our case,the first surgery has been done without prior biopsy for an osteolytic lesion of humerus. For any osteolytic lesion or a suspected malignancy a complete and proper histopathology report is mandatory before going for definitive procedure. The first biopsy report was inconclusive suggesting malignant neoplasm and the surgeon has gone for fixation with plate without a proper diagnostic work-up. Limb salvage surgery is a multidisciplinary approach which includes the operating surgeon ,radiologist, pathologist and medical and surgical oncolgist. For any osteolytic lesion suspecting malignancy a core biopsy is must before any definitive procedure.

In our case if the patient had undergone a proper diagnostic work up with biopsy before fixation Limb salvage is still a viable option and radical amputation such as shoulder disarticulation could have been avoided. Limb salvage can be done in stage IA, IIA lesions, IIIA with good response to chemotherapy, skin should be uninvolved and free, feasibility of keeping a cuff of normal tissue surrounding the tumour, pathologic fractures and fungation are definite contraindications. In our case since the tumor is fungating, limb salvage cannot be done and shoulder disarticulation was done.

Conclusion:

With Neo- adjuvant Chemotherapy and Surgery ,survival rates for TO better than conventional OS. If treated only with surgery, prognosis worse than conventional OS. Early diagnosis and prompt treatment important for disease free survival. So staged work up is always essential in an osteolytic lesion. Always go for Core Biopsy before going for any definitive procedure. In our case case Limb salvage could have been done if patient patient had undergone a proper tissue diagnosis and staged work up before going for fixation and radical amputation could have been avoided.

An Initiative of The Tamil Nadu Dr. M.G.R. Medical University
University Journal of Surgery and Surgical Specialities