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# Case Report: Megaprosthesis replacement in proximal femur Ewing's sarcoma

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### Abstract

**Introduction:** Ewing's sarcoma develops in second decade of life with median age of 13 years. It is second most common primary pediatric bone malignancy and more common in males. Ewing's sarcoma has "triad" of findings (1) "diaphyseal" location, (2) "round-cell" or "permeative" lytic destruction of bone with periosteal elevation in lamellated fashion (3) large soft-tissue mass associated with tumor. The case is of 19 year male having Left proximal femur Ewing's sarcoma who presented to NCH, Surat with C/O pain in left hip since 9 months after trivial trauma. After 2 months, Patient had undergone FNAC which misdiagnosed it as Giant cell Tumour and treated with Curettage + Bone graft. Then patient was correctly diagnosed as Non-Metastatic Ewing's Sarcoma after MRI and PET scan. MRI is the best imaging study to assess the extent of medullary osseous involvement and soft-tissue extension. PET showed FDG avid lytic lesions in GT and proximal shaft femur.

**Methods and Materials:** Treatment of Ewing's sarcoma consists of neoadjuvant preoperative chemotherapy, followed by limb-sparing surgery and consolidation chemotherapy. Modern neoadjuvant chemotherapy regimen include Ifosfamide, Etoposide with vincristine, dactinomycin, cyclophosphamide, doxorubicin (VACA). After 6 cycles of preoperative chemotherapy, we chose surgically resecting the tumor with wide margin followed by Megaprosthesis consisting of proximal femoral component, tumor resection stem, femoral head, and cement.

**Results:** Patient had started complete weight bearing after 2 weeks postop and taken 1 cycle of chemotherapy postop. But patient developed infection and exposed implant stem. Hence VAC dressing was applied for 6 weeks and then patient was finally operated with Rotational flap surgery after infection was cleared.

**Conclusion:** Megaprosthesis Replacement is an excellent Limb salvaging surgery giving complete range of motion and functional outcome in Proximal Femur Ewing's sarcoma not involving vital structures.

**Keywords:** Ewing's sarcoma, Megaprosthesis, Chemotherapy, VAC dressing, Rotational flap.

### Introduction

Ewing's sarcoma is the second most common primary paediatric bone malignancy. It typically develops in the second decade of life (90%) with a median age of 13 years being more common in males. 15 to 20% of children with Ewing's sarcoma will present with metastatic disease.

Five translocations are associated with EFTs, with two of them, T(11;22) and T(21;22), being common. The clinical presentation is usually a painful mass in the shaft of the femur, tibia, humerus, ribs or flat bones (pelvis and scapula).

Ewing's sarcoma has a typical "triad" of findings on imaging, (1) a "diaphyseal" location, (2) a "round-cell" or "permeative" lytic destruction of the bone with periosteal elevation ("onion skin") in a lamellated or layer fashion and (3) an obvious or large soft-tissue mass associated with the tumor.

CT is an excellent aid to planning a biopsy with regard to cortical thinning. MRI is the best imaging study to assess the extent of medullary osseous involvement and the extent of soft-tissue extension of the tumor and its relationship to the major nerves and vessels. Positron emission tomography (PET) showed FDG avid lytic lesions in GT and proximal shaft of femur.

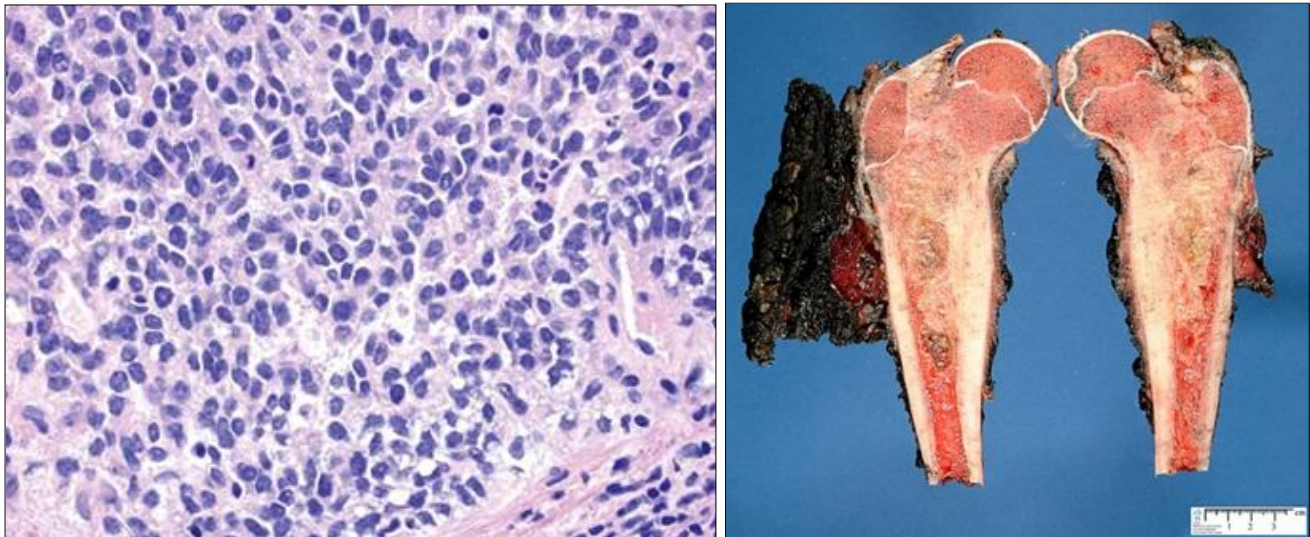
Incisional biopsy under a general anesthetic is the preferred biopsy technique which confirms diagnosis.

Grossly, Ewing's tumors can have a solid consistency or appear identical to pus. Microscopic appearance of Ewing's sarcoma is that of lobules or strings of small, round cells with clear cytoplasm and small nuclei [Fig-1].

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**Fig 1:** Grossly, Ewing’s tumors can have a solid consistency or appear identical to pus. Microscopic appearance of Ewing’s sarcoma is that of lobules or strings of small, round cells with clear cytoplasm and small nuclei

The modern treatment of Ewing’s sarcoma consists of neoadjuvant preoperative chemotherapy, followed by limb sparing surgery and consolidation chemotherapy. Modern neo-adjuvant chemotherapy regimens are variations of protocols that include Ifosfamide and Etoposide in addition to a combination of vincristine, dactinomycin, cyclophosphamide and doxorubicin (VACA).

**Methods and Materials**

The case is of 19 year male from Jalgaon, Maharashtra having Left sided proximal femur Ewing’s Sarcoma who presented to New Civil Hospital, Surat in 2021 with complaint of pain in left hip since 9 months after a trivial trauma. After 2 months of initial presentation [Fig-2, Fig-3,

Fig-4], Patient had undergone FNAC in Maharashtra where patient was misdiagnosed as Giant cell Tumour and treated with Curettage + Bone graft [Fig-5].

Then patient was correctly diagnosed as Non-Metastatic Ewing’s Sarcoma after MRI and PET scan in Surat.

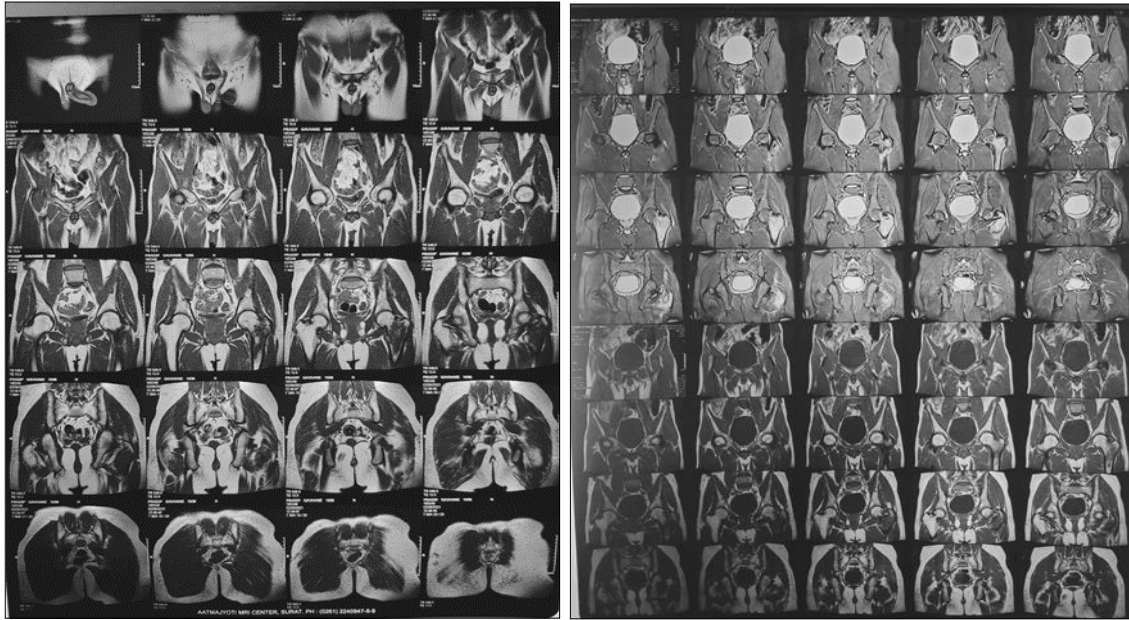
As per the neo-adjuvant chemotherapy regimen, he was started on Ifosfamide in addition to a combination of Vincristine, Dactinomycin, Cyclophosphamide, and Doxorubicin (VACA).

After a good response to 6 cycles of preoperative chemotherapy, we typically chose to respect the tumor with a wide margin followed by Megaprosthesis consisting of proximal femoral component, tumor resection stem, femoral head along with cement [Fig-6].

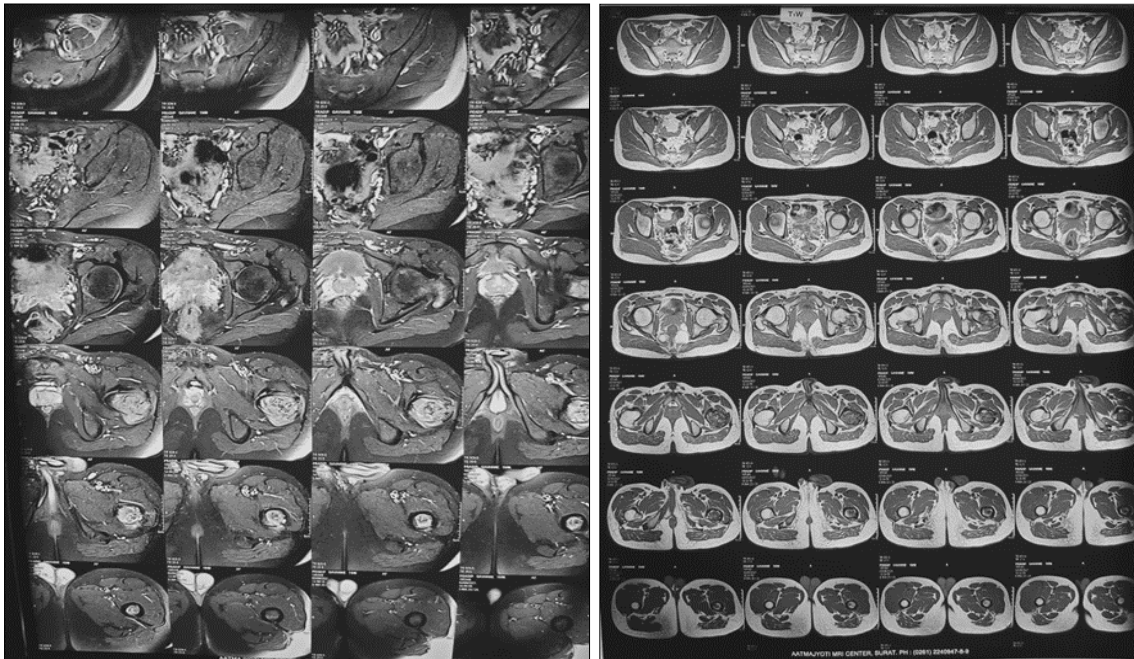


**Fig 2:** X-ray on initial presentation showing lytic lesion in left proximal femur trochanteric region.





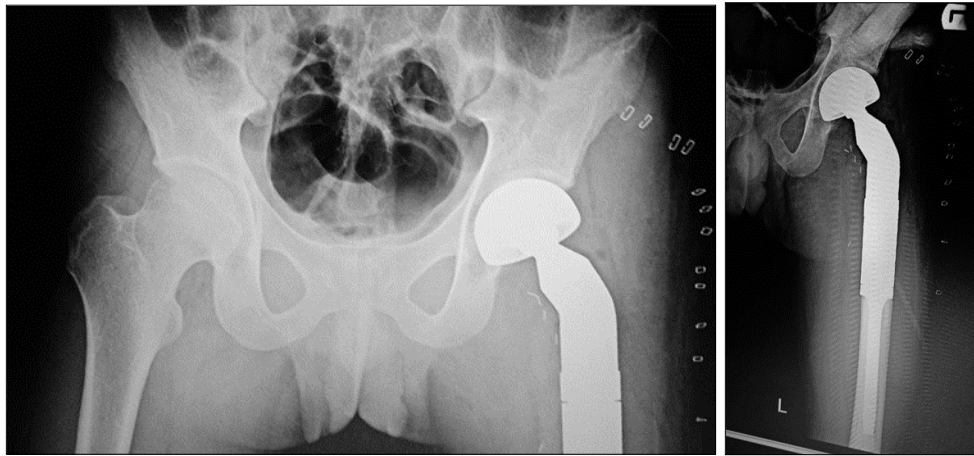
**Fig 3:** MRI on initial presentation coronal images



**Fig 4:** MRI on initial presentation sagittal & axial images



**Fig 5:** After curettage and grafting at previous hospital.



**Fig 6:** Post op X-ray showing Megaprosthesis consisting of proximal femoral component, tumor resection stem, femoral head along with cement



**Fig 7:** At 8 weeks postop, patient developed a sinus with pus discharge and the exposed implant



**Fig 8:** After sequential VAC dressing, patient was finally operated with Rotational flap surgery after infection was cleared.



**Fig 9:** Clinical picture at 4 months post op.



## Results

Patient had started complete weight bearing after 2 weeks postop with complete wound healing and taken 1 cycle of chemotherapy postop.

But after 8 weeks postop, patient developed a sinus at centre of stitch line which had pus discharge and the implant stem was exposed [Fig-7]. Hence VAC dressing was applied for duration of 6 weeks and then patient was finally operated with Rotational flap surgery after infection was cleared [Fig-8, Fig-9].

## Conclusions

Megaprosthesis Replacement is an excellent Limb salvaging surgery giving complete range of motion and functional outcome in Proximal Femur Ewing's sarcoma not involving vital structures.

Other implants that can be used are intercalary-diaphyseal allograft reconstruction fixed with locked intramedullary rod.

Surgical resection and reconstruction for Ewing's involves two basic choices: Limb salvage and amputation. Amputation is now nearly exclusively reserved for local recurrence or bulky pelvic primary tumors involving vital structures.

Radiation therapy is also used, depending on the location of the tumor.

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### How to Cite This Article

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