



CASE BRIEF

Name: [REDACTED]

Age: 9 years, Female

Address: [REDACTED]

Contact no: [REDACTED]

Diagnosis: Osteosarcoma right distal femur (Post NACT) with pathological fracture with Retinoblastoma post enucleation

Date of Diagnosis: 15/01/2023

Ref by: Dr. Jalila

Surgery/Surgeons: Wide local excision and reconstruction using K Nail cement spacer.
Dr. Pramod S Chinder and Dr. Suraj HP.

Date of Surgery: 28/03/2023

BRIEF SUMMARY OF THE EVENTS:

DATE	EVENTS	FINDINGS
2015	Known case of retinoblastoma. Underwent left eye enucleation in 2015.	Treated in India (thru TAC). Retinoblastoma Group C in RE and Neovascular glaucoma with Group E in LE. Status Post Chemo + LE enucleation with ball implant.
Past History: Retinoblastoma group C in RE and neovascular glaucoma with group E in LE. Initially she received 12 cycles of combination chemo of Crabo, Etoposide and Vincristine (CEV) followed by LE enucleation with ball implant on 03/11/2016. Histopathology revealed positive high risk, NM study and CSF were negative and she received another 6 cycles of CEV.		
Dec 2022	H/O trivial fall- Right distal femur-Complaints of pain & swelling.	Unable to walk after the fall. Sustained pathological fracture.



3 rd Jan 2023	CT Scan	(? Osteosarcoma)
3 rd Jan 2023	MRI Scan	Features suggestive of osteosarcoma.
3 rd Jan 2023	HRCT Thorax	No evidence of pulmonary or abdominal metastatic lesions noted.
15 th Jan 2023	Bone biopsy	Conventional Osteosarcoma of the right distal femur

Chemotherapy:

High dose METHOTREXATE regimen was started and 1st cycle was completed in Feb 2023.

Despite chemotherapy the swelling increased rapidly in size and involved the whole distal thigh region extending up to proximal middle 3rd junction of the right thigh. Due to which, the patient was not able to stand, not able to walk with support. The patient was referred to us after completion of 1st cycle of high dose METHOTREXATE for surgery.

On presentation patient was having high grade fever for which prophylactic IV antibiotics were started. After discussing the case with pediatric medical oncologist, plastic surgery and infectious disease team, the patient was planned for wide local resection and reconstruction using nail cement spacer. Due to the diffuse swelling and very large mass in the distal thigh, risk of amputation and skin flap related complications was also explained.

20 th Mar 2023	MRI	Suggestive of Osteosarcoma.
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MDT Discussion:

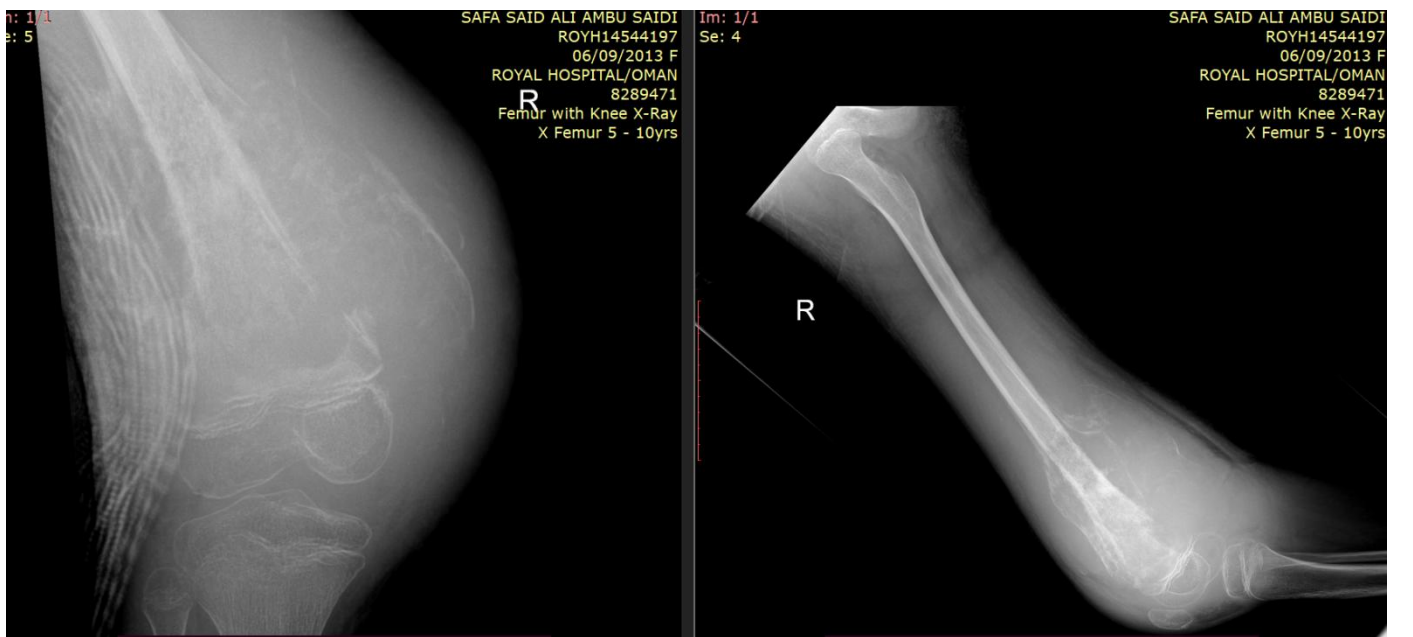
Patient was planned for surgery without chemotherapy as she progressed with HDMTX NACT. Initially patient was considered for resection + prosthesis replacement, but on repeat imaging since extensor mechanism could not be salvaged, she was planned for nail cement spacer application.

After 3-5 years after surgery, if the patient is disease free with good functional status, she can be considered for prosthesis reconstruction to get knee movements in the future.

27 th Mar 2023	Pre -op embolization	
28 th Mar 2023	Surgery	Wide local excision and reconstruction using K Nail cement spacer
28 th Mar 2023	HPE	Osteosarcoma.
4 th Apr 2023	Germ line Mutation study	REPORT AWAITED



X-RAY IMAGES: 27/02/2023



MRI IMAGES: 03/01/2023



Findings:

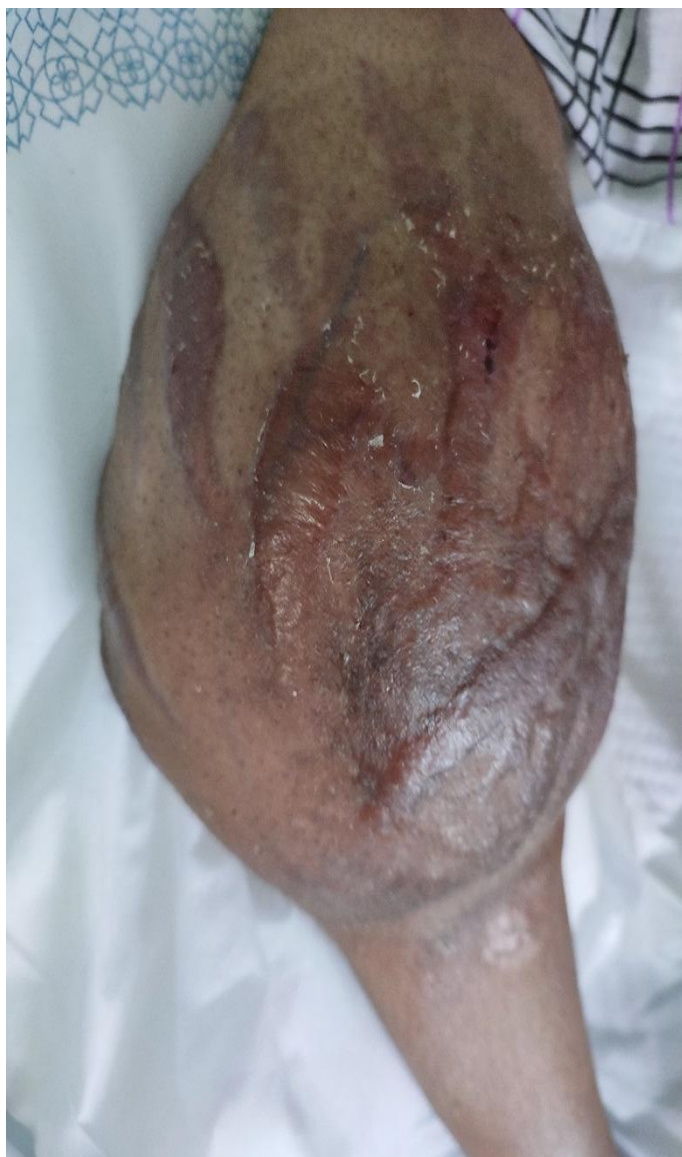
1	Large aggressive bone lesion affecting right distal femur metaphysis and diaphysis measuring 5x4x8 cm with heterogenous signals with haemorrhagic areas and aggressive periosteal reaction and postcontrast enhancement, features suggestive of Osteosarcoma.
2	The lesion is inseparable from the inner margin of the vastus intermedius muscle and posteromedial aspect of the vastus medialis muscle. Both these areas shows post contrast enhancement with raise of suspicion of muscular invasion.

CT: 03/01/2023

Findings:

1	No evidence of pulmonary or abdominal metastatic lesions noted.
2	An infiltrative meta-diaphyseal lesion is seen involving the distal femoral shaft with significant periosteal reaction (sunburst appearance) and areas of cortical thinning and disruption and large extra osseous soft tissue components (more at postero-medial aspect). Multiple sub-centimetric lymph nodes are seen at the popliteal fossa. Mild intra-articular synovial effusion. Opinion: Right distal femoral meta-diaphyseal infiltrative mass lesion, neoplastic (? Osteosarcoma).

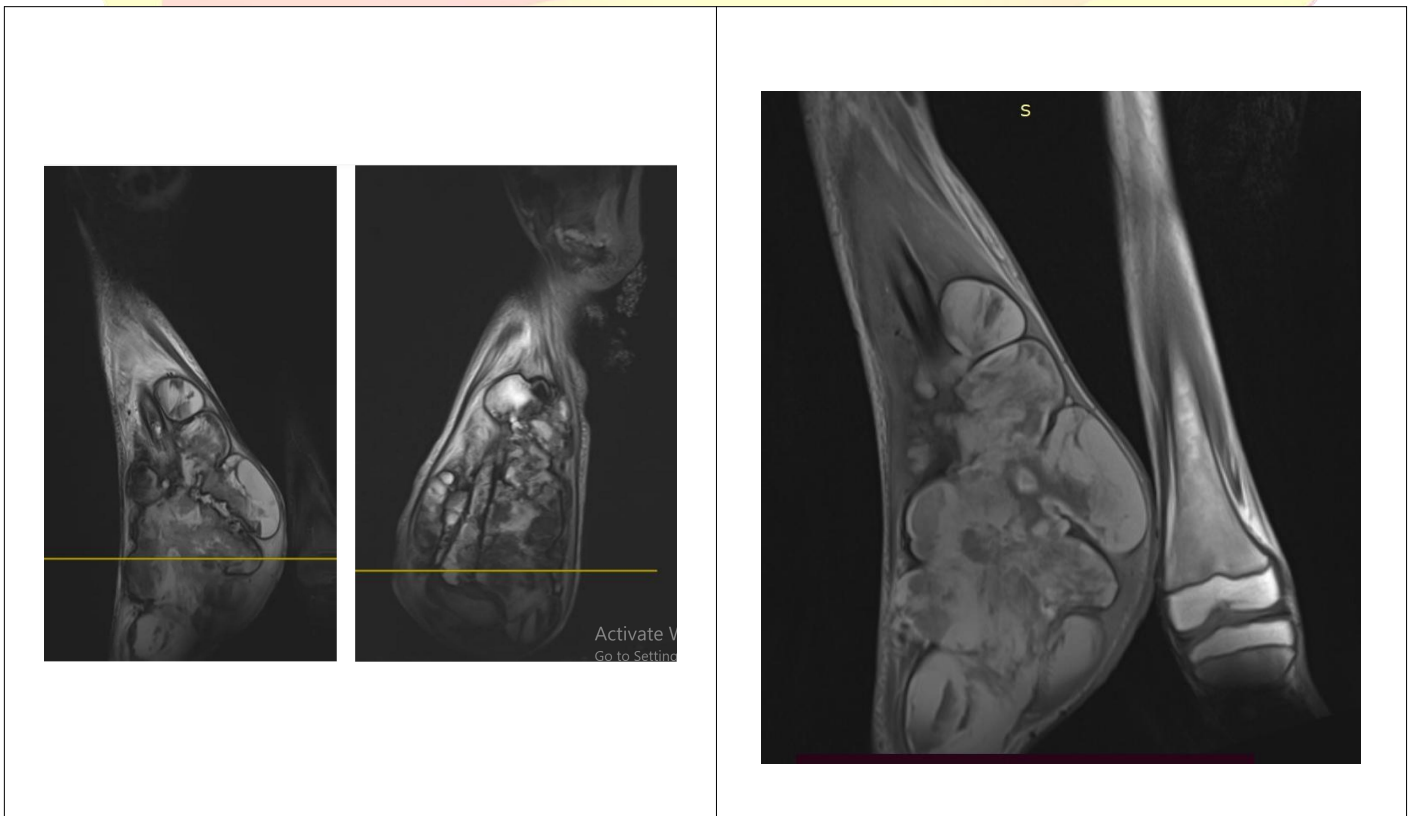
CLINICAL IMAGES:

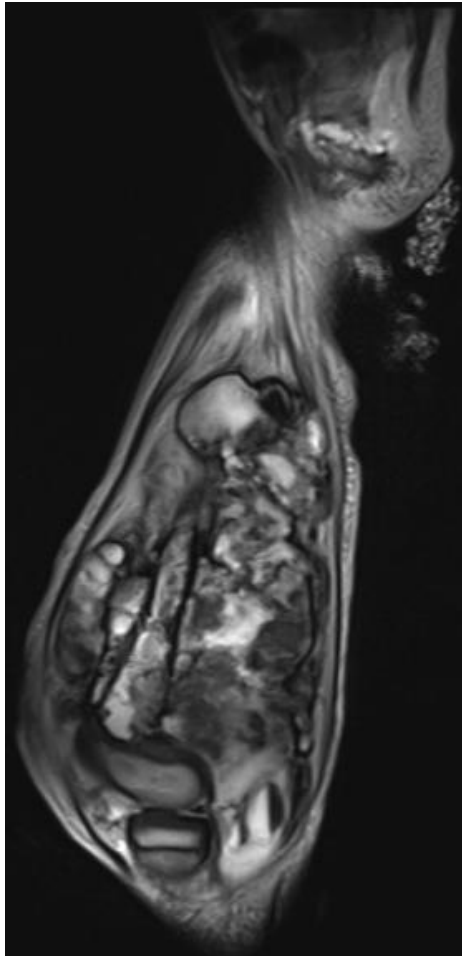


X-RAY IMAGES: Post Chemo 19/03/2023



MRI IMAGES: 20/03/2023 (Post Chemo)





Findings:

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|---|--|
| 1 | 26 x 13.6 x 12.9 cm expansile lytic lesion with solid and cystic component arising from the distal femoral metadiaphysis showing extraosseous soft tissue component- Suggestive of residual disease. |
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HISTOPATHOLOGY (Needle Biopsy) 15/01/2023:

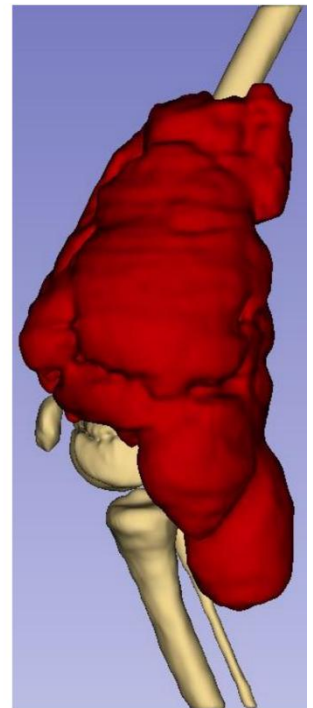
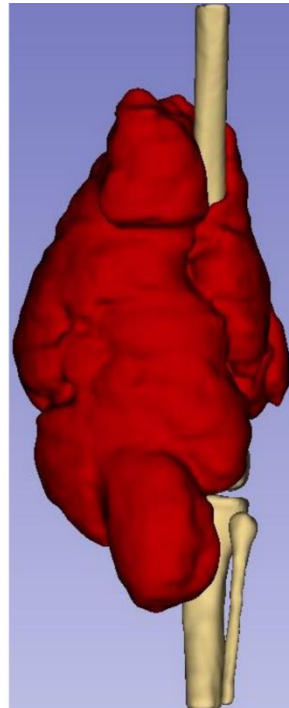
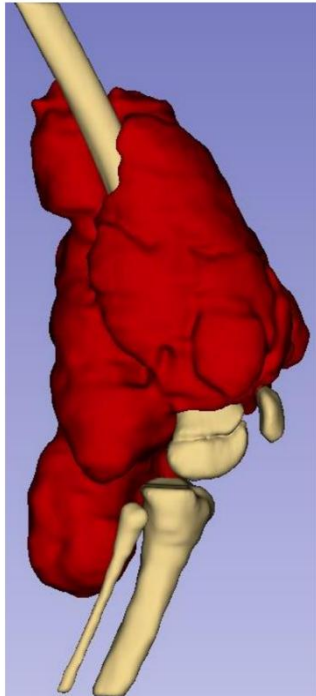
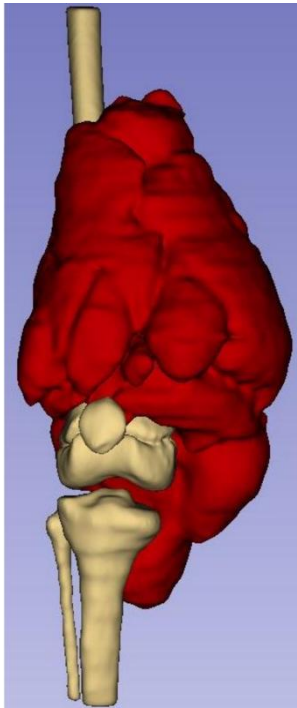
Findings:

- | | |
|---|---|
| 1 | Section shows bony tissue with a high-grade neoplasm composed of pleomorphic oval-to-spindle cells with hyperchromatic, bizarre nuclei and occasional multinucleated forms. Mitotic figures are present. The tumour cells are replacing the marrow and encasing and eroding pre-existing trabeculae. A few fragments show lace-like trabeculae matrix.
DIAGNOSIS: -RIGHT DISTAL FEMUR; BONE BIOPSY: CONVENTIONAL OSTEOSARCOMA. |
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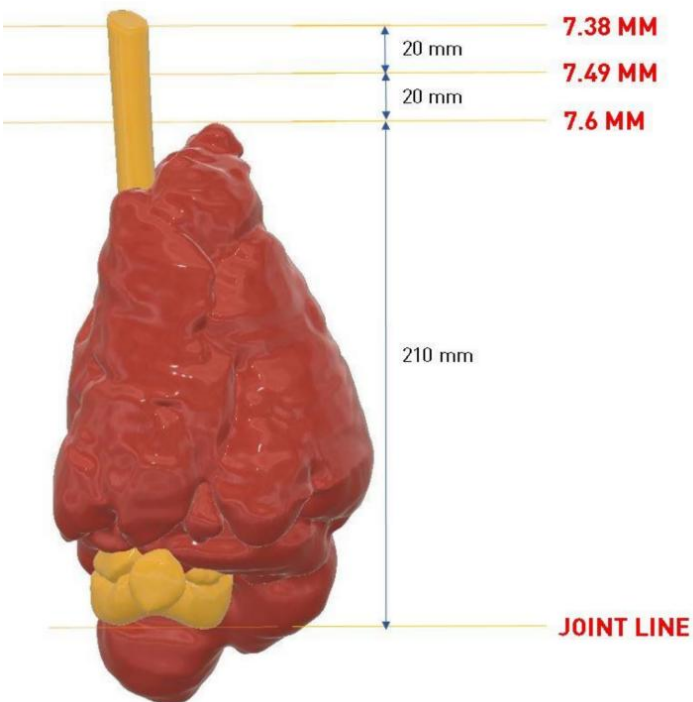
3D PRINTING AND PLANNING IMAGES:

Anterior, lateral, posterior, and medial views of the segmented ROI

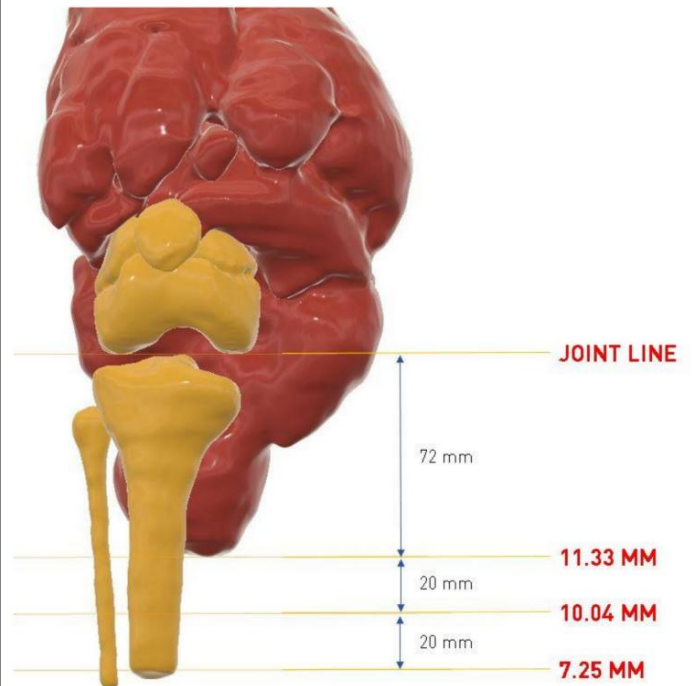


MEASUREMENTS

- Intramedullary canal widths of femur:

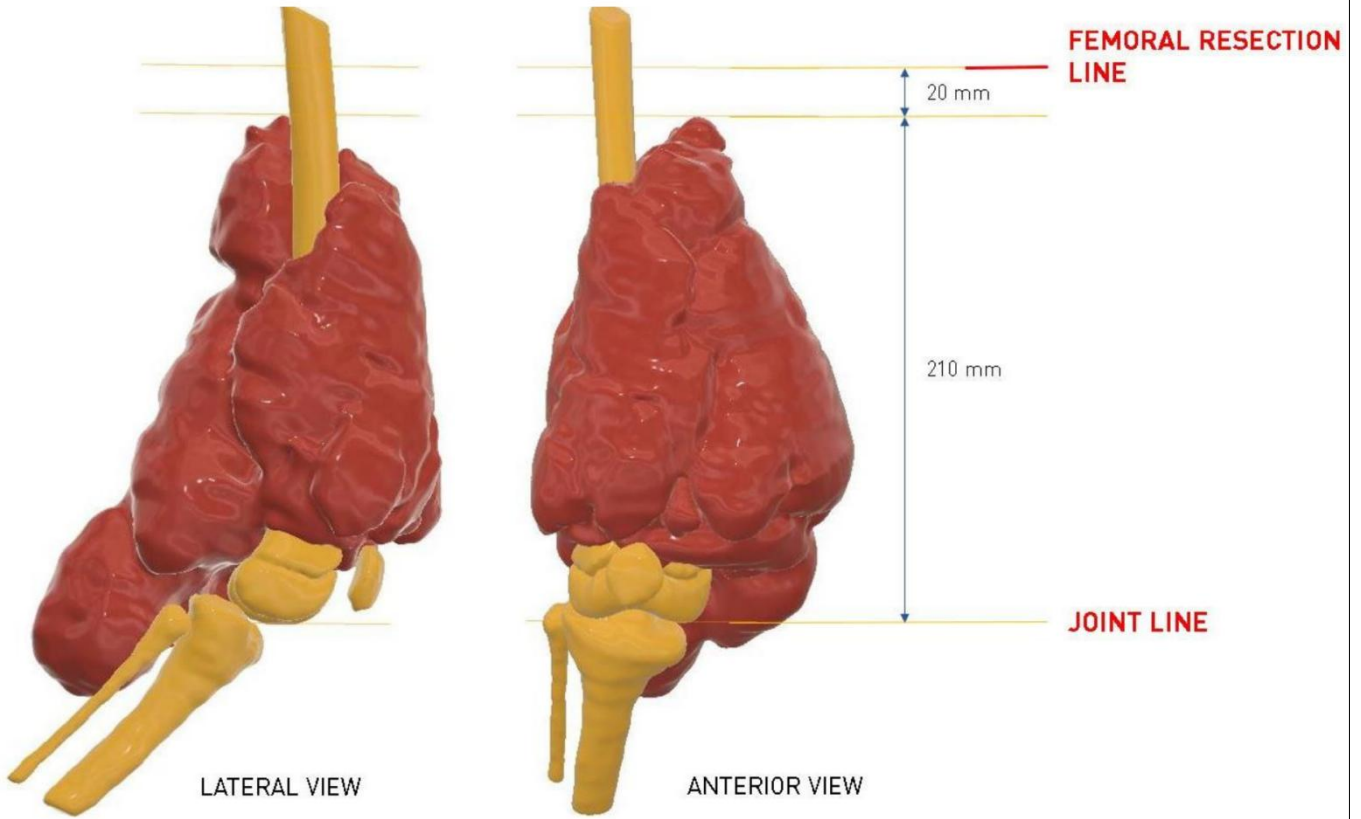


- Intramedullary canal widths of tibia:

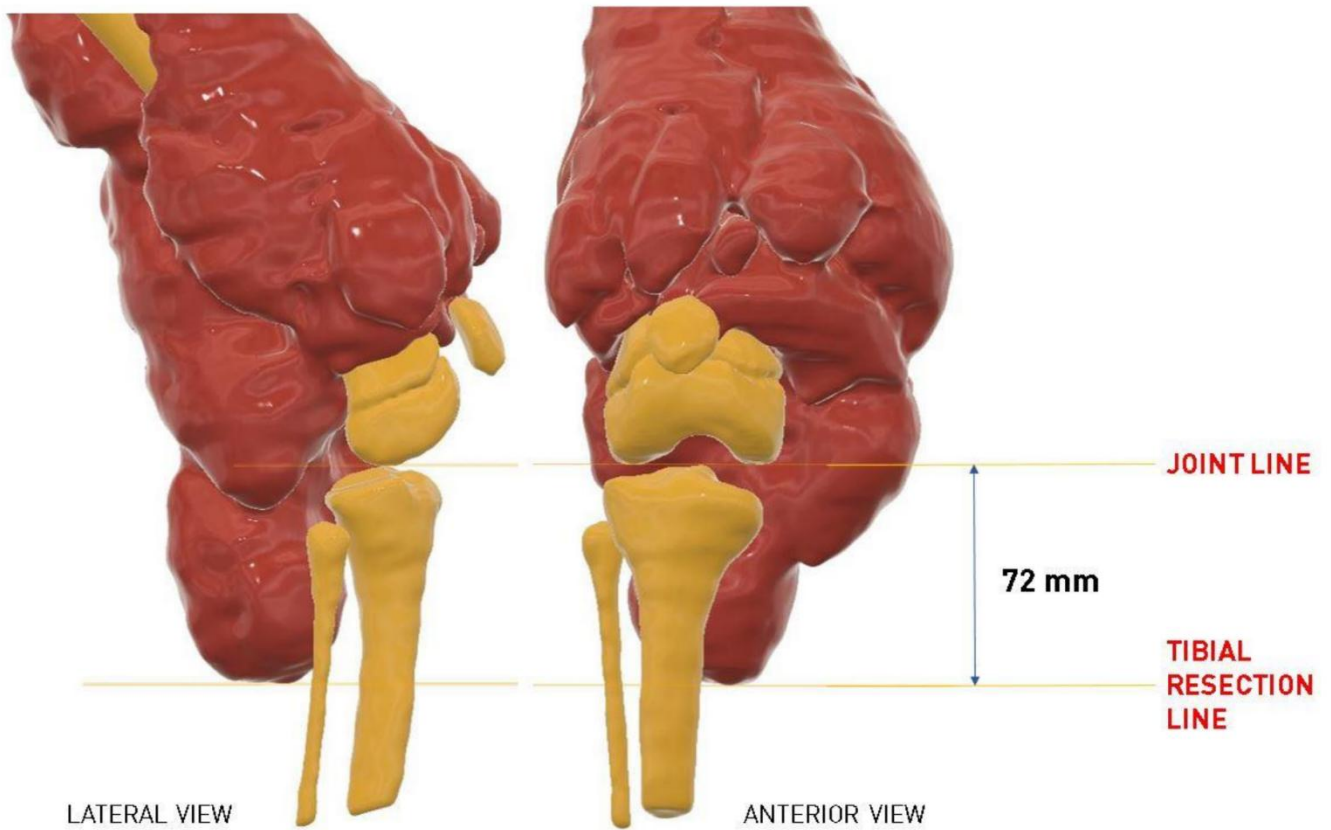




Resection line of femur (with respect to knee joint line):

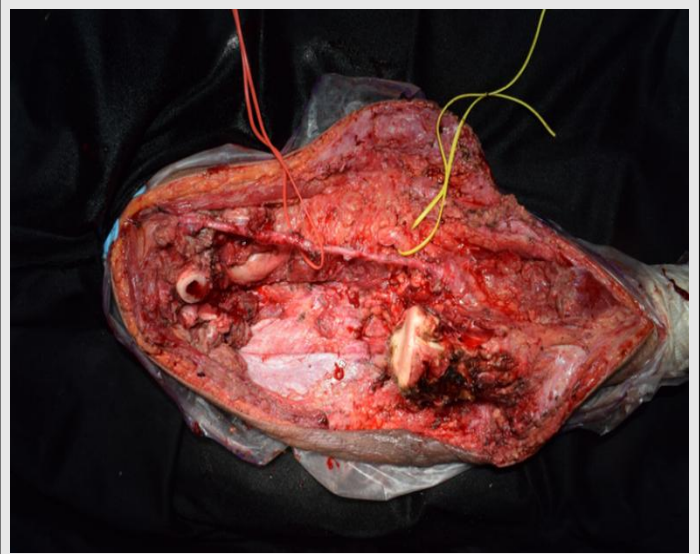
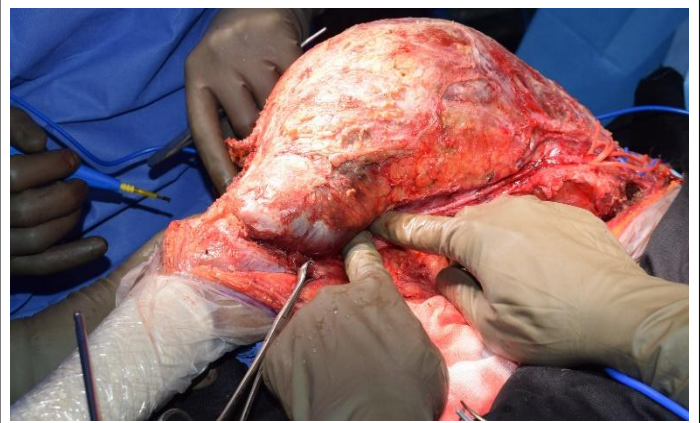
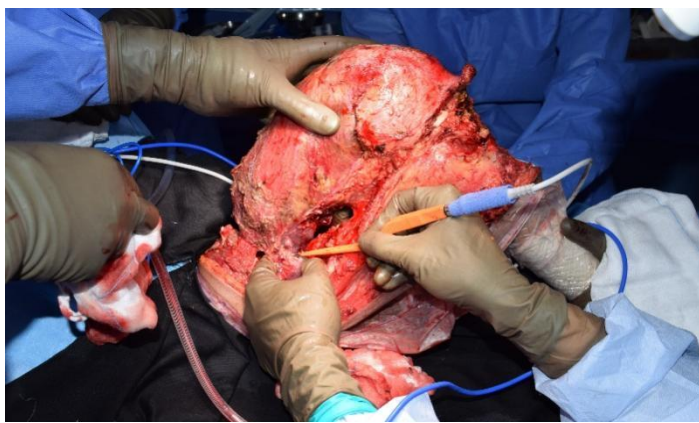


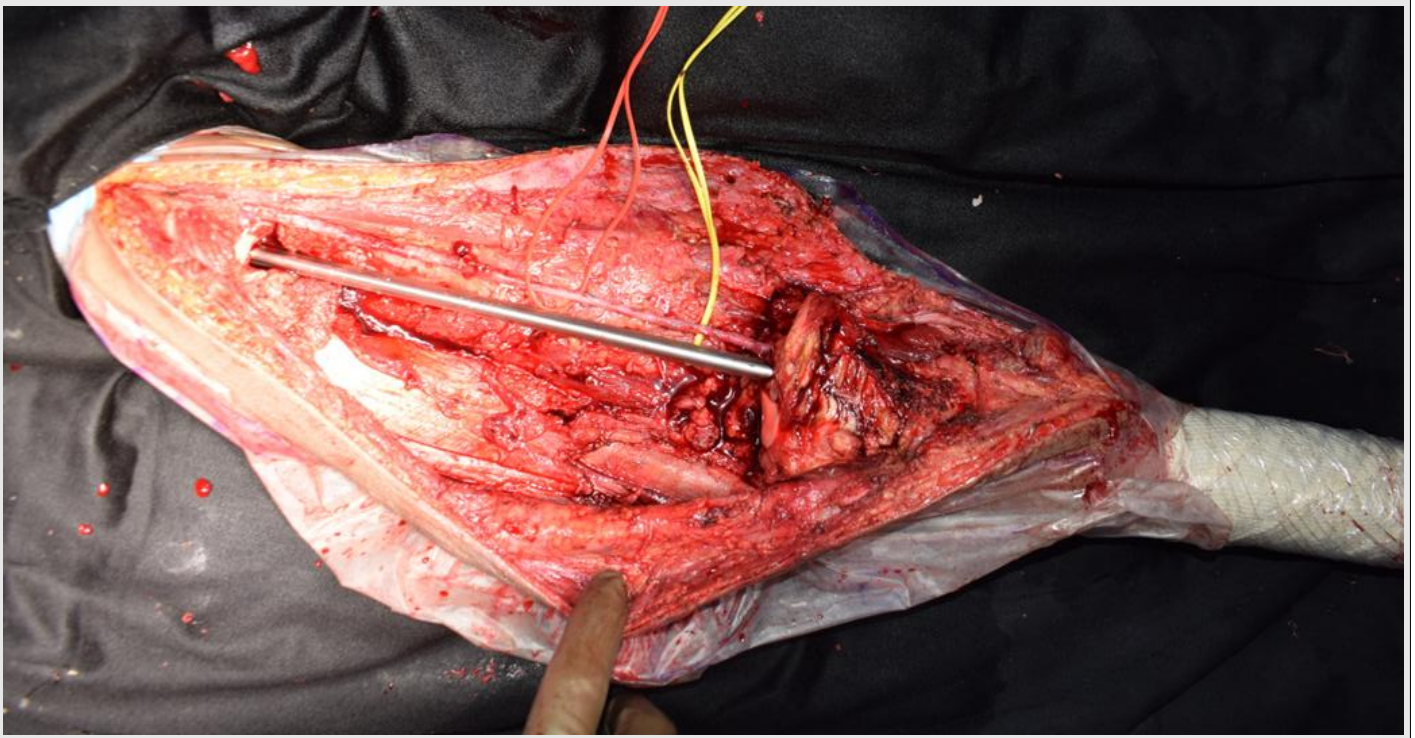
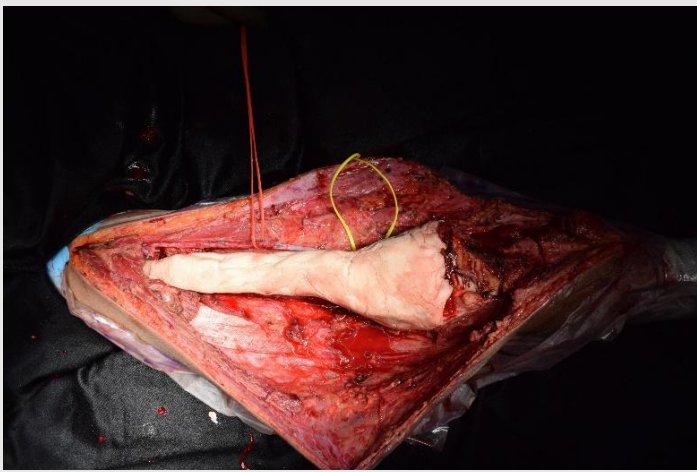
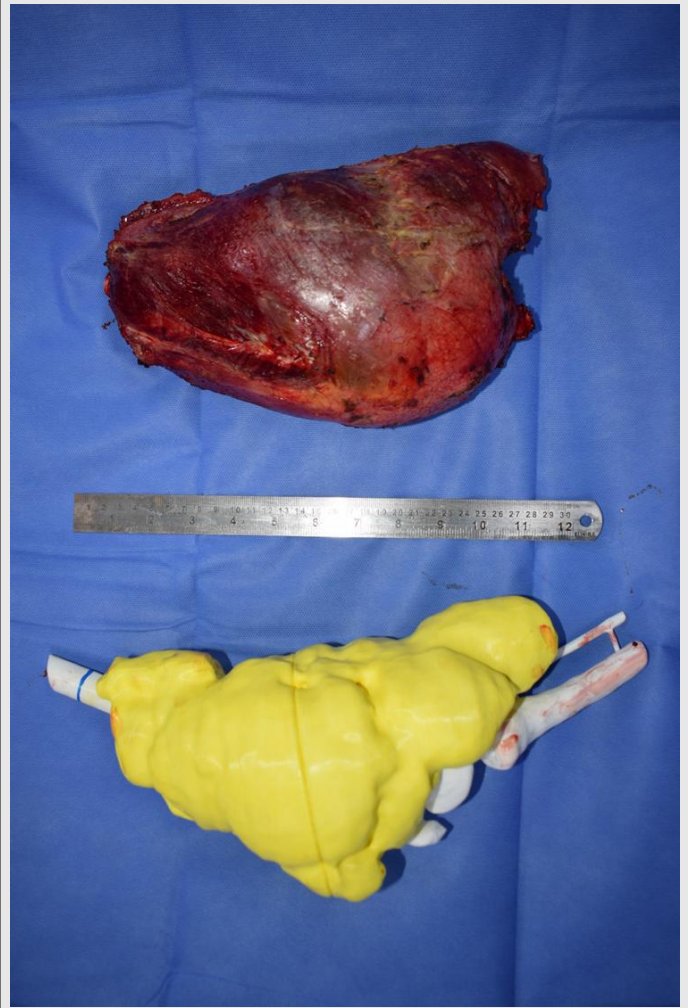
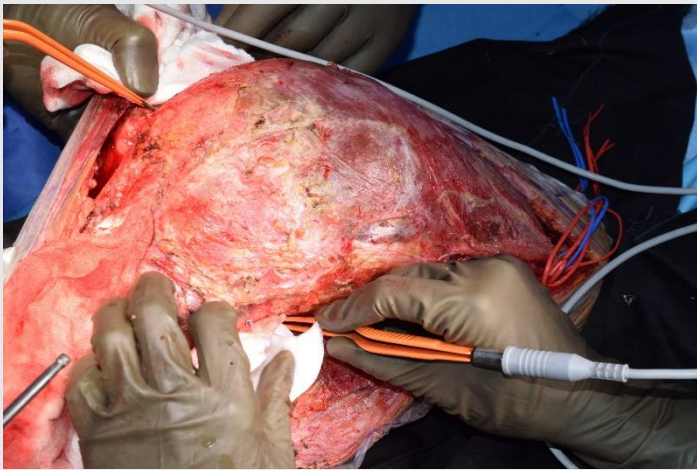
Resection line of tibia (with respect to knee joint line):





OPERATIVE IMAGES: 28/03/2023



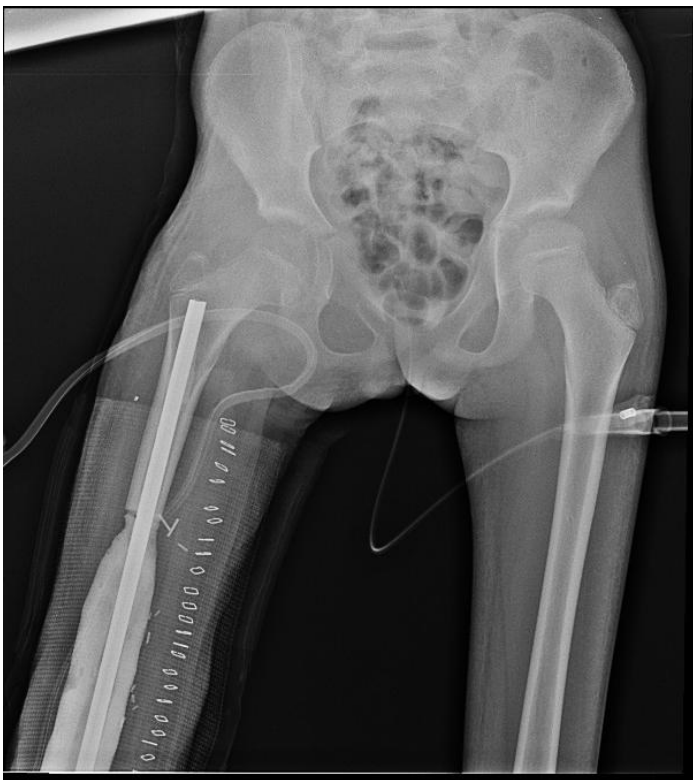




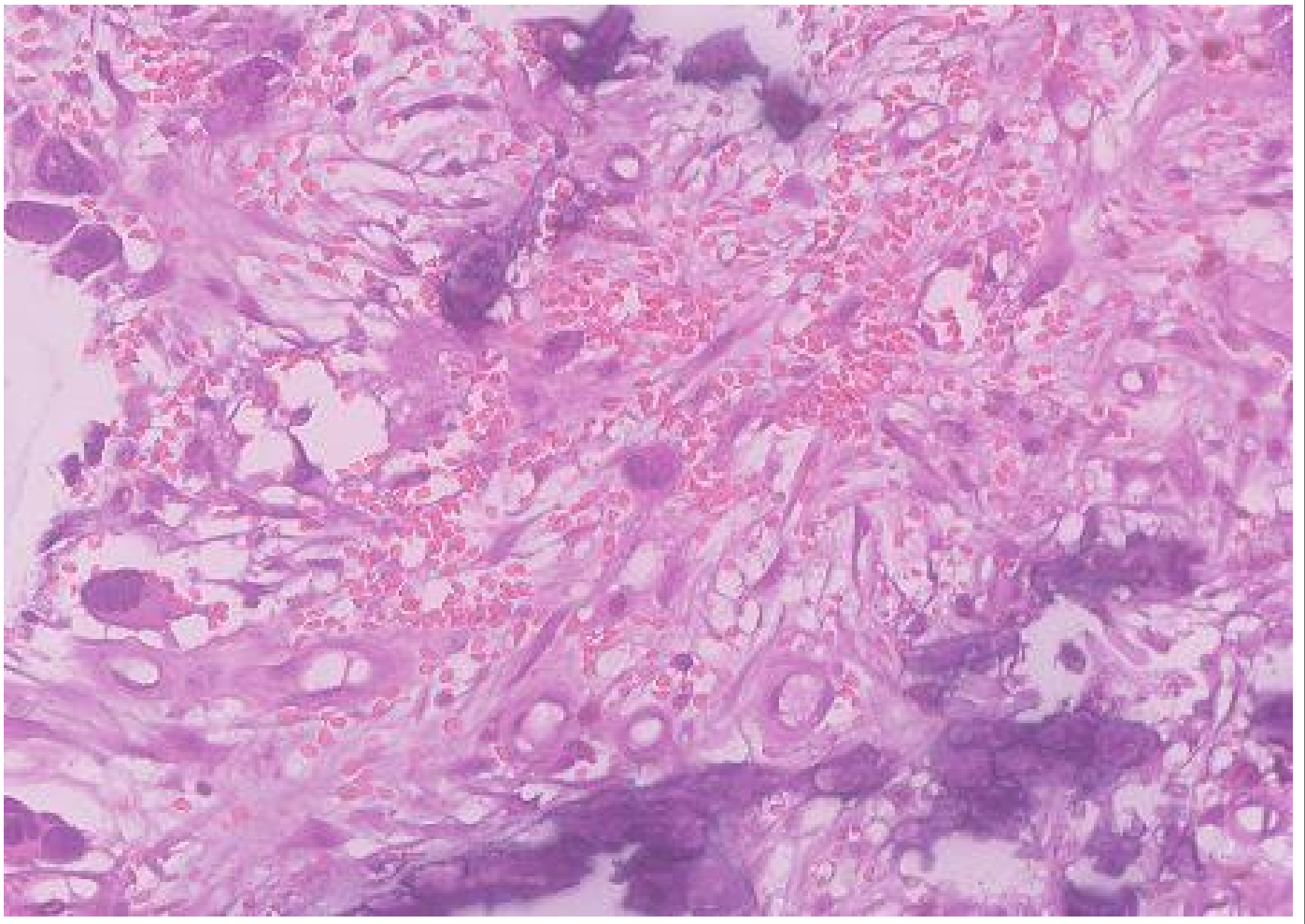
Findings:

1	Tumor removed En-bloc.
2	No neurovascular injury.
3	Reconstructed with nail cement spacer.

POST-OP X-Ray: 29/03/2023



HISTOPATHOLOGY IMAGES 28/03/2023:



Findings:

1	A small focus of Residual viable Osteogenic Sarcoma.
2	99% necrosis (HUVOS Grade III), Free Margins.



PROPOSED RECOMMENDATION AS DISCUSSED IN TYR MULTIDISCIPLINARY SARCOMA TUMOUR BOARD:

1	To continue remaining High dose methotrexate chemotherapy at Oman.
2	To keep close follow-up considering volume of disease and h/o Retinoblastoma.
3	Discussion: 99% necrosis. Role of Adjuvant Radiation.
4	<p>To review once in 3 months for every 2 years and for every 6 months for next 3 years. To review with Dr. Pramod every year at HCG to plan for further surgery. To follow up with genomic report accordingly. After discussion in our sarcoma tumor board MDT the child did not respond to chemotherapy, planned to start local radiation therapy after the wound is healed (Focused radiation) because the skin is very fragile- Please be very careful and also, to be planned for a targeted therapy like Pazopanib. The wound has to be reviewed regularly till the wound heals later the radiation has to be started.</p> <p>Continue walking with the walker support with (FCB) brace. Note: The implant used is not MRI compatible.</p>

Next 5-year plan for patient:

0-2 years:

No active intervention, close follow-up and monitoring once every 3months

2-4 years:

- Shoe raise
- Plan for epiphysiodesis of left distal femur.

4-5 years:

- Removal of nail cement spacer.
- Reconstruction using megaprosthesis and free functioning muscle transfer.



OUR MDT TEAM MEMBERS:

NAME	DESIGNATION
Dr.Pramod Chinder	Consultant Orthopaedic Oncosurgeon
Dr Suraj H P	Orthopaedic Oncosurgeon
Dr.Yoga Rakshith	Clinical fellow- Orthopaedic Oncology
Dr. Narendra	Clinical fellow- Orthopaedic Oncology
Dr. Kunal	Consultant Oncopathologist
Dr. Aparna	Consultant Oncopathologist
Dr. Imran	Consultant Oncopathologist
Dr. Shivakumar	Consultant Radiologist
Dr. Kumaraswamy	Consultant Radiation Oncologist
Dr. Vikram Maiya	Consultant Radiation Oncologist
Dr. Vijay Agarwal	Consultant Medical Oncologist
Dr. Intezar	Consultant Paediatric Medical Oncologist

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Mrs. Kavya	MSW	9632971901
Mrs. Veena	Coordinator	9148663925
Mr. Sukrit	Medical-Design Engineer	
Mr. Alit	Coordinator	9632419696

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Checked by: Dr.Pramod.S.Chinder