CASE BRIEF

Name:	[redacted]
Age:	2years, Male
Address:	[redacted]
Contact no:	[redacted]
Diagnosis:	Ewing's sarcoma right proximal fibula with vascular encasement
Date of Diagnosis:	19/05/2022
Ref by:	Dr. Jalila
Surgery/Surgeons:	Limb salvage surgery –wide local excision of tumor with proximal fibulectomy by Dr.Pramod S Chinder, Dr. Suraj.
Medical Oncologist:	Dr. Intezar
Radiation Oncologist:	Dr. Shridar P.S
Date of Surgery:	27/02/2023

Rationale of treatment

The patient was referred to us with the above-mentioned diagnosis after receiving about 6 cycles of neoadjuvant chemotherapy in his home country. Once the patient arrived to HCG, he was thoroughly evaluated clinically and radiologically with whole body PET scan and MRI scan. The scan revealed a moderately regressed tumor in the proximal fibula when compared to the previous scan which was performed in his hometown before neoadjuvant chemotherapy, with encasement of distal popliteal artery and bifurcation of anterior and posterior tibial artery. The case was then discussed in the sarcoma tumor board MDT and since we were planning for a definitive surgery with a curative intent. He was advised two more cycles of neoadjuvant chemotherapy with an intention that the chemotherapy would further reduce the size of the tumor and the popliteal vessels and nerve can be separated from the tumor mass and proximal fibula and the tumor can be resected with adequate margins without injuring the neurovascular structures. Hence, two more cycles of neoadjuvant chemotherapy was given by Dr. Intezar Mehndi (Consultant Paediatric Oncologist) and team.

After chemotherapy, the patient was re-evaluated and his case was extensively discussed in the tumor board meeting. The repeat scan revealed further regression in size of the lesion however, the

neurovascular structures was still found to be encased within the tumor. Hence, the MDT was of the consensus that neoadjuvant radiation would benefit in shrinking the tumor and preserving the posterior neurovascular structures. The possibility of performing adjuvant radiation after surgery was also discussed however it was not recommended considering the tumor bed would be significantly larger if radiation was given after surgery and it would increase the possibility of injury to the adjoining tibial epiphysis and tibial bone. Hence, we were of the consensus that neoadjuvant radiation would benefit the patient in performing limb salvage surgery without much injury to the neurovascular structures.

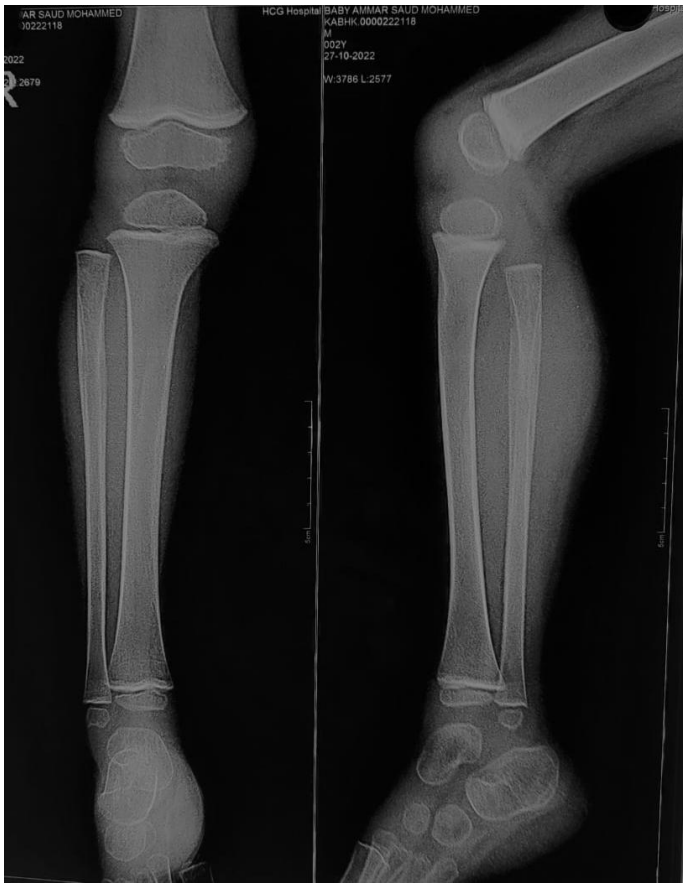
Hence, he received neo-adjuvant radiation and was planned for surgery after 4 weeks from end of radiation therapy.

BRIEF SUMMARY OF THE EVENTS:

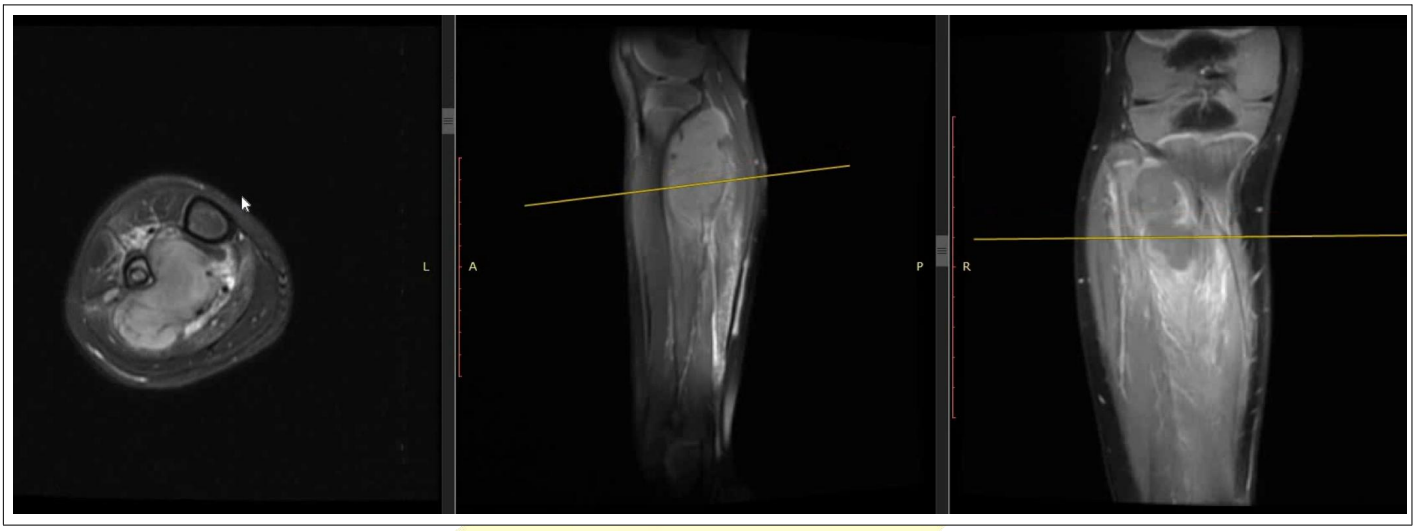
DATE	EVENTS	FINDINGS
Apr 2022	Right upper leg	Complaints of pain and swelling. Gradually increasing in size with inability to bear weight.
May 2022	MRI-Right leg	Expansile intramedullary lesion of meta-diaphysis of proximal fibula.
19th May 2022	CT guided biopsy	Small round cell tumor. IHC cells were positive for CD99 and occasionally positive for BCL2
16th June 2022	PET CT scan	No other lesions elsewhere in the body.
19th June 2022	Chemotherapy	Neo-adjuvant chemo-VDC/IE regimen.
Sept 2022	PICU admission	Possible febrile neutropenia with septic shock.
26th Nov 2022	MRI	Further regression in size of the lesion. The soft tissue measures 3.1 x 2.1 x 4.6cm. (Previously measuring 3.3 x 2.4 x 4.6cm) Complete encasement of the distal popliteal artery and the proximal tibial, peroneal arteries with luminal narrowing.
9th Dec 2022	PET CT Scan	3.1 x 2.2 x 4.4 cms lesion showing vascular encasement-Suggestive of residual disease.

		Total 11 course of neoadjuvant chemotherapy along with neoadjuvant RT were given.
23rd Jan 2023	PET CT Scan	Minimal interval regression in size of lesion- 2.4 x 1.0 x 4.2cms. (Previously measuring 3.1 x 2.2 x 4.4 cms)
22nd Feb 2023	MRI Scan	Further regression in size of the lesion.
27th Feb 2023	Surgery	Limb salvage surgery –wide local excision of tumor with proximal fibulectomy.
Mar 2023	CT Angiogram	Non-visualization of the right proximal anterior tibial artery
15th Mar 2023	HPE	Negative for malignancy. No evidence of necrosis seen.

X-RAY IMAGES: 27/10/2022



MRI IMAGES: 17/05/2022



Findings:

1	A large expansile intramedullary lesion of the metadiaphysis of the proximal fibula measuring 4.6cm in length with posteromedial cortical erosions and oedema of the epiphysis.
2	It shows thick periosteal reaction and significant extraosseous soft tissue component exceeding the size of the osseous component measuring 3.6 X 3.9 X 5.5cms.

PET CT IMAGES: 16/06/2022

Findings:

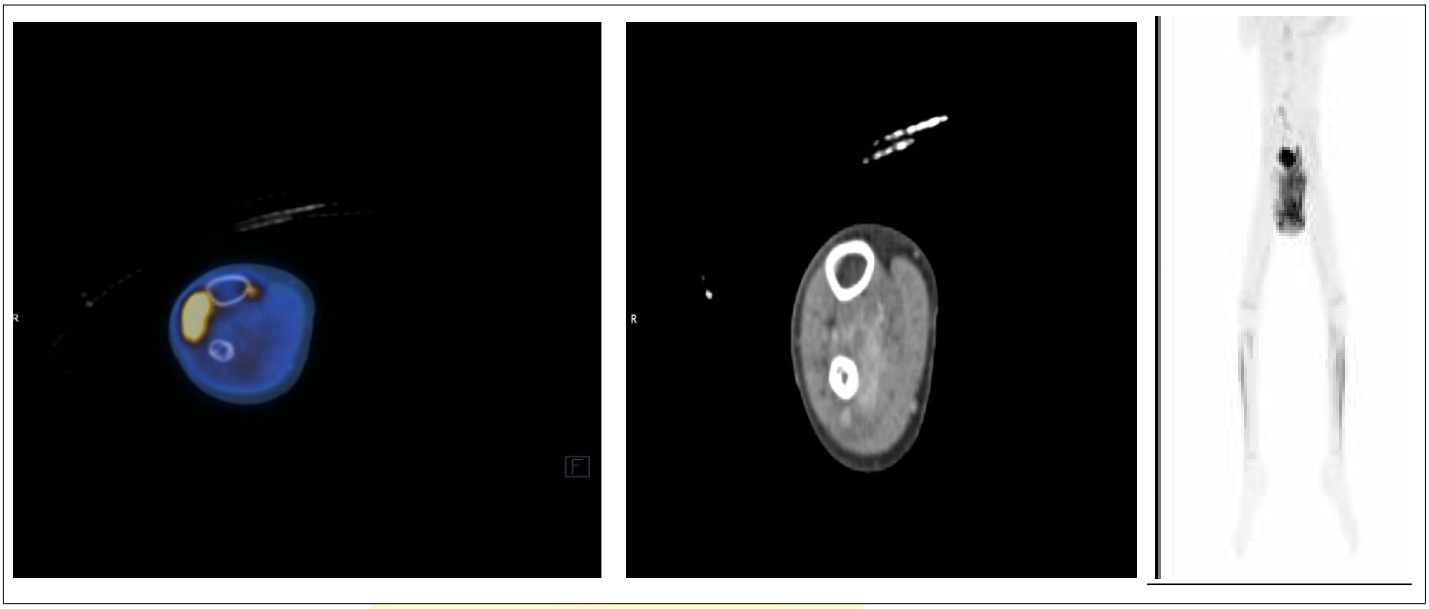
1	FDG avid permeative lytic lesion involving proximal right fibula with periosteal reaction associated with a large soft tissue component approximately measuring 49 X 53 X 65mm, surrounding the proximal fibula infiltrating surrounding muscles, encasing distal popliteal artery and tibioperoneal trunk with adjacent muscles appearing bulky and edematous- **Metabolically active primary Ewing's sarcoma.**

HISTOPATHOLOGY (CT Guided Biopsy) 19/05/2022:

Findings:

1	Small round blue cell tumor in favour of Ewing's sarcoma. IHC: The cells were positive for CD99, occasional positive for BCL2 and synoptophysin is seen. The neoplastic cells are immunonegative for AE1/AE3, chromogranin A, MyoD1, Myogenin, Tdt and LCA.

PET CT IMAGES: 09/12/2022



Findings:

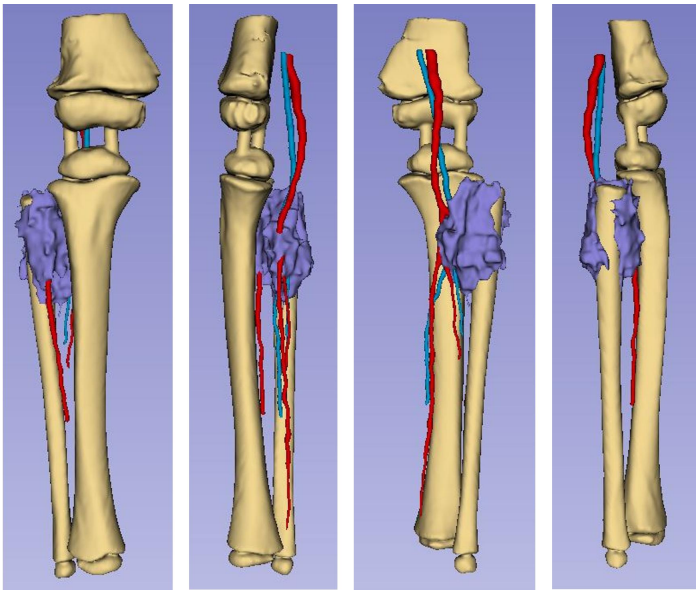
1	Metabolically active 3.1 x 2.2 x 4.4 cms irregular heterogeneously enhancing lesion along the right proximal fibular shaft with enhancing extraosseous soft tissue component showing vascular encasement- Suggestive of residual disease. SUV:1.4

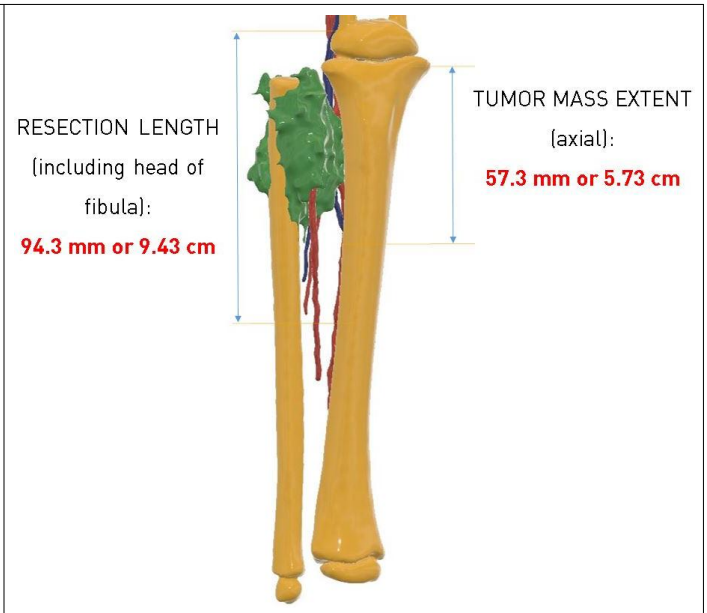
MRI IMAGES: 23/01/2023

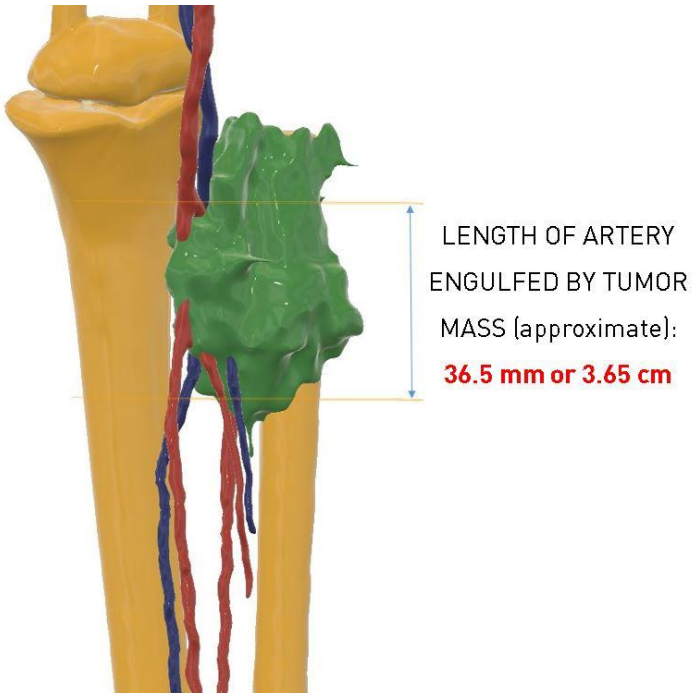
Findings:

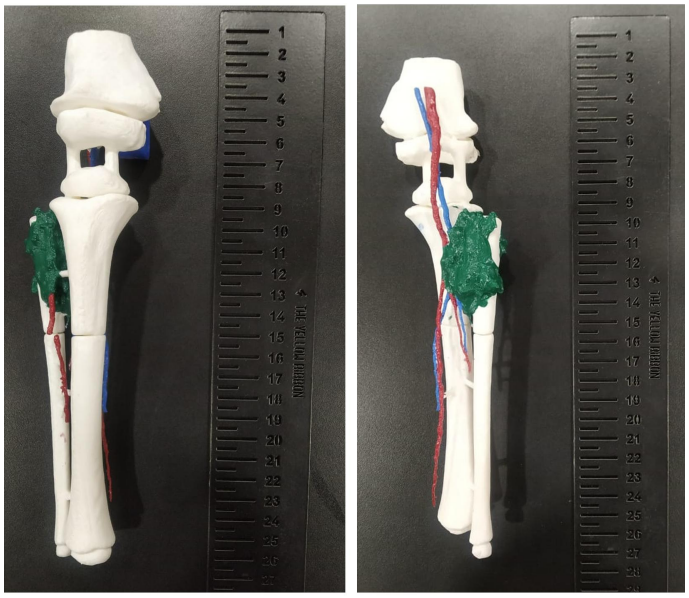
1	Minimal interval regression in size of lesion arising from the right proximal fibular metadiaphysis extraosseous soft tissue. The soft tissue now measures 2.4 x 1.0 x 4.2cm, previously measuring 3.1 x 2.1 x 4.6cm.
2	Persistent short segment complete encasement of the distal popliteal artery and the proximal tibial, peroneal arteries with luminal narrowing.

3D PRINTING AND PLANNING IMAGES







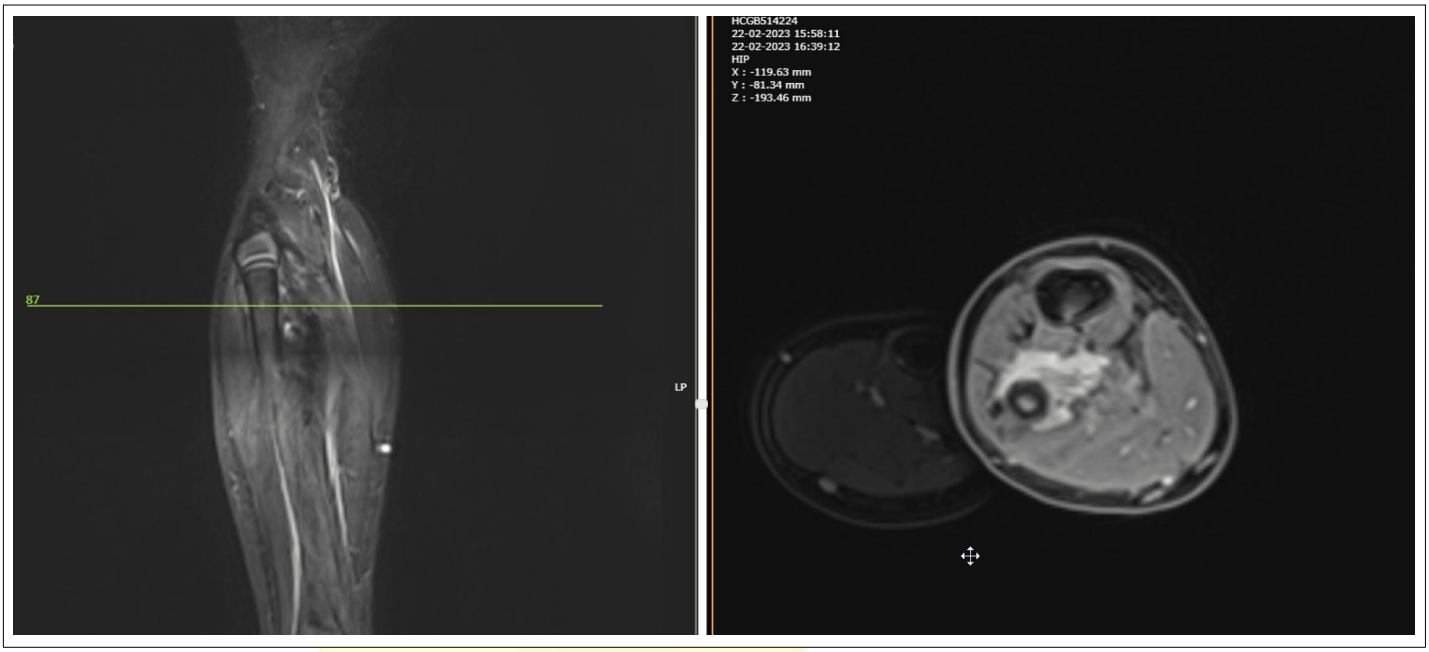


Anterior and posterior view of the 3D printed model

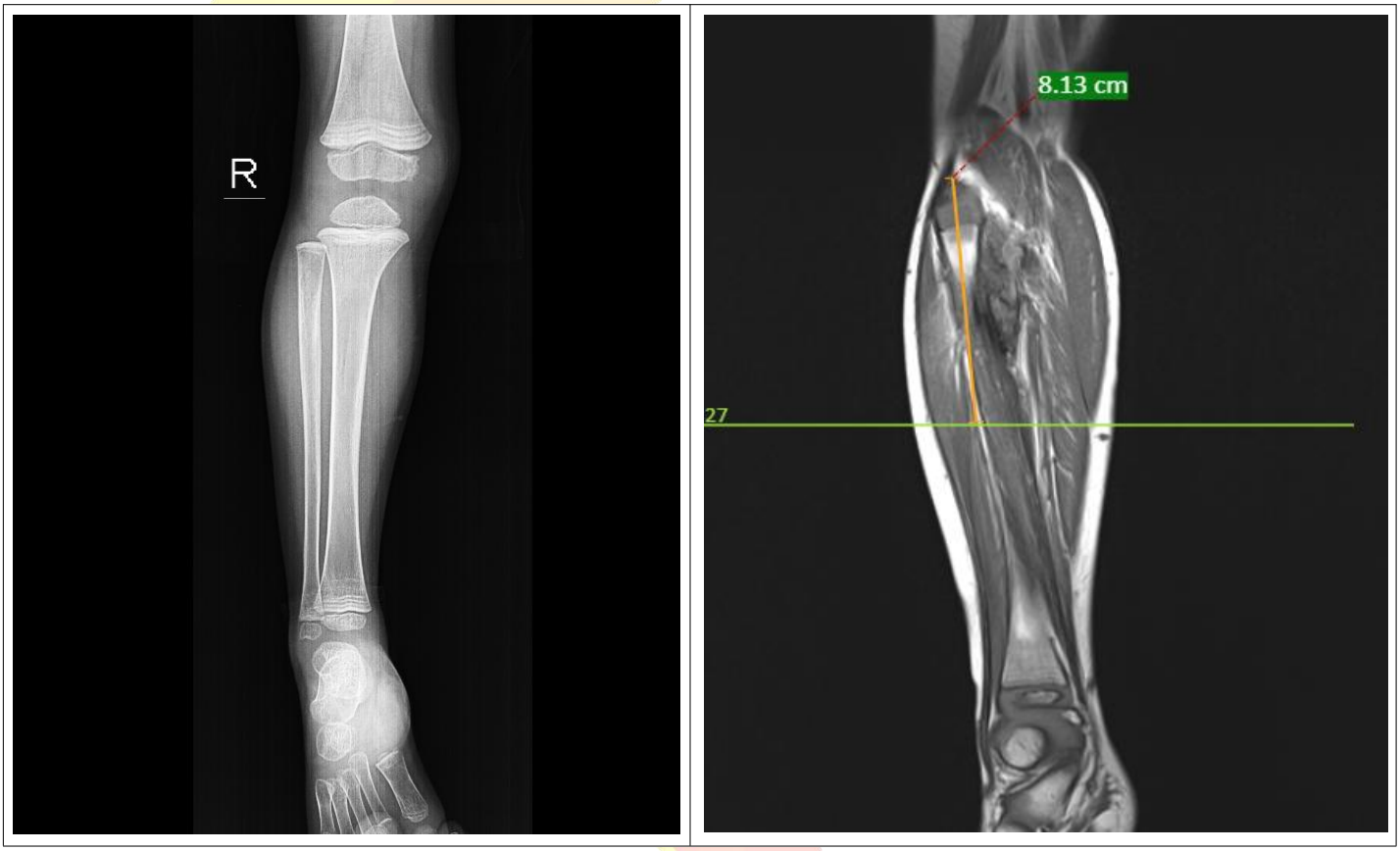
MRI IMAGES: 22/02/2023

Findings:

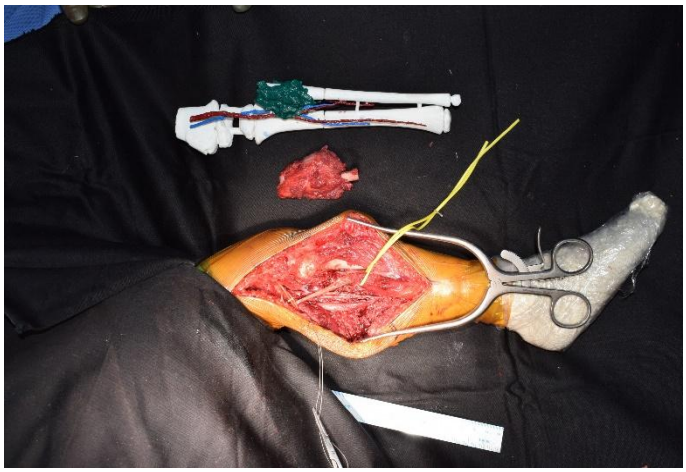
1	Further regression in size of enhancing lesion arising from the right proximal fibular metadiaphysis extraosseous soft tissue. The soft tissue now measures 2.8 x 2.1 x 4.6, previously measuring 3.1 x 2.1 x 4.6cm.
2	Persistent short segment complete encasement of the distal popliteal artery and the proximal tibial, peroneal arteries with luminal narrowing.

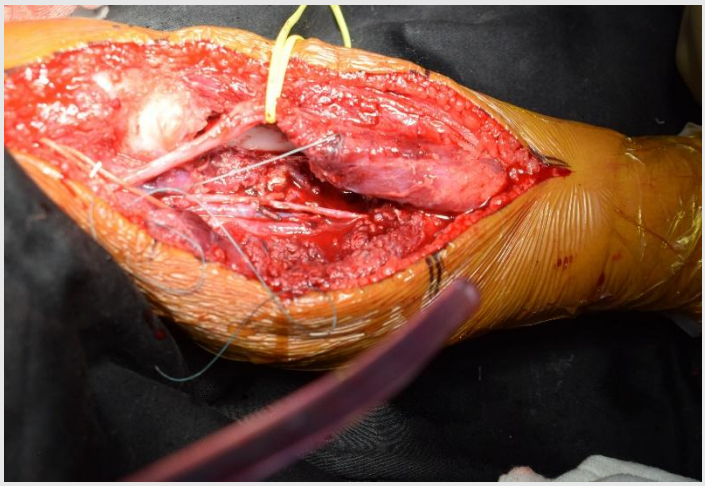


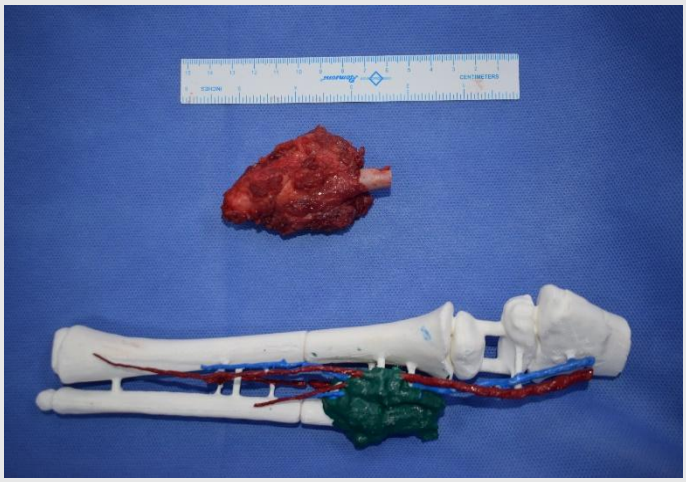
X-RAY IMAGES: 22/02/2023 **Resection length-8cms**

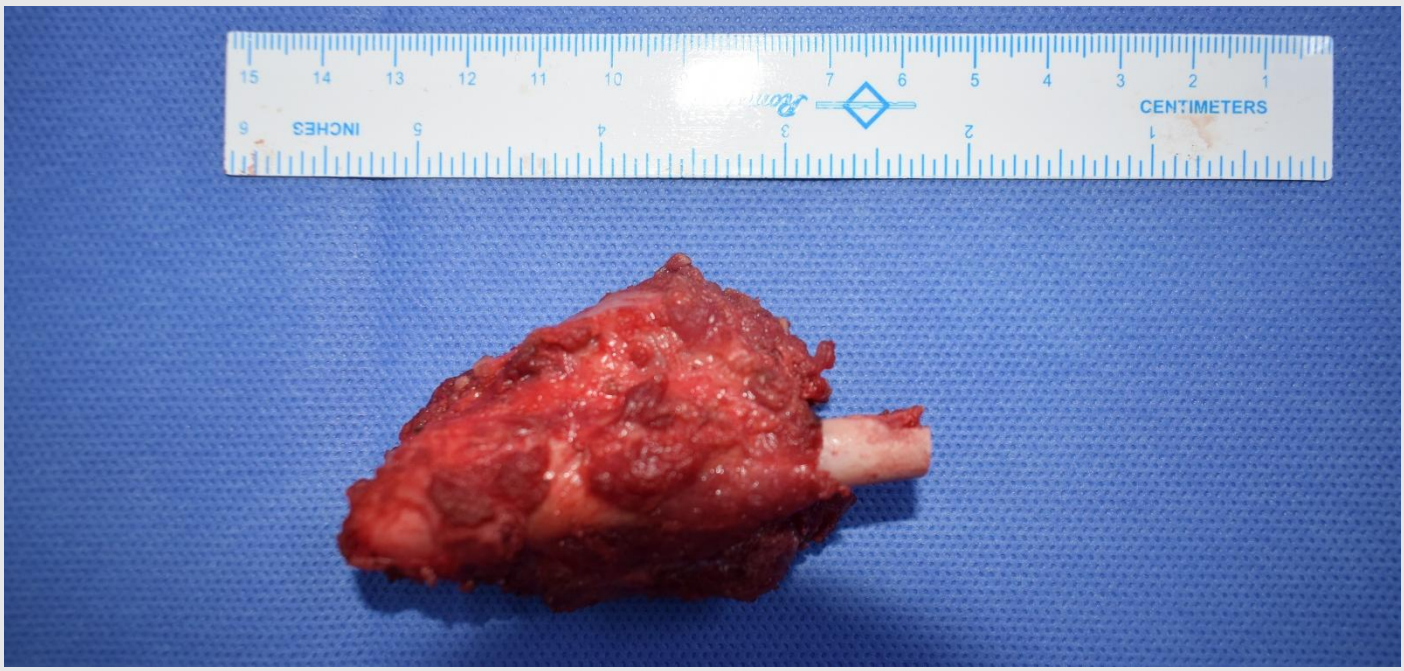


OPERATIVE IMAGES: 27/02/2023









Findings:

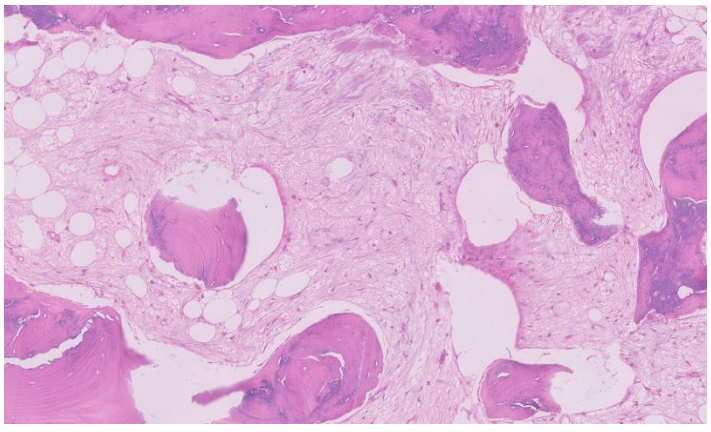
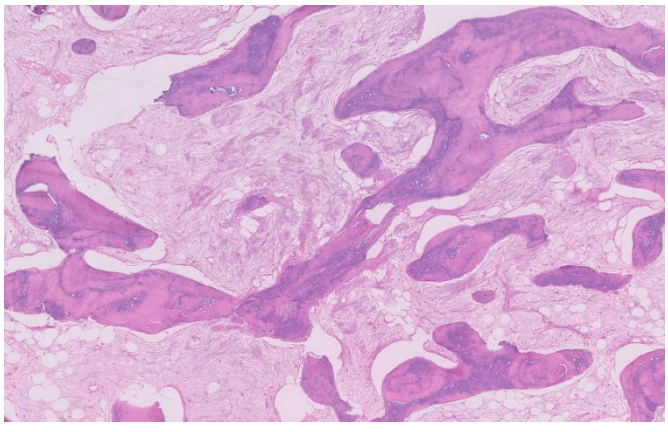
1	Patient put in left lateral position. An incision of 20cm taken lateral aspect of the knee and proximal leg, soft tissue dissected.
2	Enbloc dissection of the tumor done with wide margins.
3	Tibial nerve, common peroneal nerve, dissected out and preserved. Anterior tibial artery and posterior tibial artery identified and cut along with tumor because of encasement.
4	Leg vascularity was checked after deflating torniquet.
5	With plastic surgeon opinion intraop adequate flow to leg was confirmed. Haemostasis achieved. Soft tissue reconstruction-Biceps femoris, lateral collateral complex secured with suture anchor calf muscles reconstructed. Skin closed in layers after placing drain.

CT ANGIO- (POST OP)

Findings:

1	Status post resection of right proximal fibular ill defined soft tissue with post-operative changes.
2	Mild attenuation of the right distal popliteal artery.
3	Non-visualization of the right proximal anterior tibial artery with well opacified and normal caliber distal right ATA in the mid and lower leg.
4	Surgical clips limiting the evaluation of right common peroneal and proximal posterior tibial arteries. Well opacified and normal in caliber PTA and peroneal arteries distally.

HISTOPATHOLOGY IMAGES 15/03/2023:

Findings:

1	No active tumour cells, necrosis and therapy related changes seen.
2	Good response to chemotherapy and radiation therapy.
3	Impression: Negative for malignancy. No evidence of necrosis seen.

PROPOSED RECOMMENDATION AS DISCUSSED IN TYR MULTIDISCIPLINARY SARCOMA TUMOUR BOARD:

1	To continue adjuvant VAC-IE chemotherapy (remaining cycles)
2	Close follow-up in V/O location of the lesion, MRI once in 3months for 2years
3	Close monitoring for growth discrepancy around the operated knee
4	Continue physiotherapy as advised
5	To review once in 3 months for every 2 years and for every 6 months for next 3 years.

PHYSIOTHERAPY NOTES:

1	Gentle knee range of motion exercises; to achieve complete knee flexion and extension in the next 4 weeks (6weeks from surgery).
2	Static and dynamic quadriceps strengthening exercises; to achieve complete knee extension (Active) over the next 3-4 weeks.
3	Partial weight bearing ambulation with pediatric walker support. Gradually increase weight bearing by 20% every 2-3weeks. To achieve complete weight bearing and ambulation with support by 6-8 weeks and to achieve complete weight bearing and ambulation without support by 4-6 months (from surgery date).

OUR MDT TEAM MEMBERS:

NAME	DESIGNATION
Dr.Pramod Chinder	Consultant Orthopaedic Oncosurgeon
Dr Suraj H P	Orthopaedic Oncosurgeon
Dr. Rakshith	Clinical fellow- Orthopaedic Oncology
Dr. Narendra	Clinical fellow- Orthopaedic Oncology
Dr. Veena	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 Mehdi	Consultant Paediatric hematooncologist

OUR TEAM MEMBERS:

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Mr. Aasim Mohammed	Nurse Practitioner	9060178007 - **9632419696**
Ms. Tejashvini Anchan	Secretary	9535832969 – **7996907996**

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

Prepared by: Tejashvini.

Checked by: Dr.Pramod.S.Chinder