- 30	Irnal or P	OR	IGINAL RESEARCH PAPER	General Surgery	
Indian			E REPORT: EXTRASKELETAL MYXOID ONDROSARCOMA	KEY WORDS:	
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ACT	Extra skeletal myxoid chondrosarcoma is a rare variety low grade malignant soft tissue tumor that typically occurs in deep soft tissues of proximal part of lower extremity. ^{1,2} We had a case of 48 years old female patient having left thigh				

deep soft tissues of proximal part of lower extremity. We had a case of 48 years old female patient having left thigh swelling for 6 years. On investigation, MRI of the lower limb revealed soft tissue mass in left upper thigh and core needle biopsy reported as myxoid soft tissue sarcoma. Patient was planned for surgical management and pathological report confirmed after immunohistochemistry as extra skeletal myxoid chondrosarcoma. Patient was referred to medical oncologist for further plan of treatment and management.

CASE REPORT:

A 48 years old female patient came to surgery OPD with presenting complaint of swelling over left thigh for 6 years which was noticed first after history of trauma over the same site. Initially the swelling was of small size and then it gradually increased up to present size and associated with pain over swelling and difficulty in walking.

EXAMINATION



Fig 1 :- showing gross appearance of swelling

On examination swelling is 18*10 cm in size , firm in consistency, 15cm below anterior superior iliac spine, no redness, no tenderness, no local rise of temperature , surrounding skin found normal, no loss of hair, no any previous scar mark , multiple lymph node palpable at the left inguinal region which is hard in consistency.

INVESTIGATION 1) XRAY LOCAL PART:

soft tissue shadow not involving bone



fig 2 :- Xray local part

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2) MRI:

Well defined multiloculated solid lesion measuring 19*10*13 cm in size involving muscles and subcutaneous tissue at posterior and posterolateral aspect of mid and lower thigh with internal hemorrhagic component. Multiloculated solid lesion in femoral triangle p/o metastatic lymphadenopathy

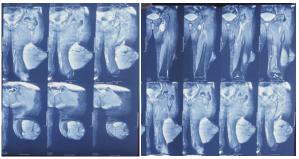


Fig 3 :- MRI local part

4) CORE NEEDLE BIOPSY

Core needle biopsy report suggestive of Myxoid soft tissue sarcoma.

TREATMENT:

After doing all radiological and histopathological investigation, we planned wide local excision of mass and Lymph node followed by post op chemotherapy and radiotherapy and follow up for recurrence.

SURGERY :- WIDE LOCAL EXCISION OF SOFT TISSUE SARCOMAWITH LYMPHNODE EXCISION.

Intra operative finding:- skin incision kept obliquely, tumor separated from surrounding tissue and muscle, tumor excised, transverse incision kept over left inguinal region to excise matted lymph node and sent for histopathological examination.



Fig 4 :- intra operative excised mass

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POST OPERATIVE STATUS



Fig 5 :- post operative wound at day 6

on post-operative day 7 suture removal done under aseptic precaution.

HPE: Mass was 20*18*11cm3 in size and firm in consistency. Overlying skin, circumferential skin margin and base is free from tumor. Immunohistochemistry, desmin,S-100,CK,CD-117,SYNAPTOPHYSIN.Markers were positive confirming

EXTRASKELETAL MYXOID CHONDROSARCOMA

FNCLCC GRADE 2 (T2 N1 M1)¹

POST OPERATIVE CHEMO-RADIOTHERAPY:- after

wound healing we started chemotherapy in form of sunitinib 25mg for 4 week along with 50Gy IMRT.

DISCUSSION:

Extra skeletal myxoid chondrosarcoma is rare soft tissue sarcoma of lower extremities in proximal part.

Predominantly occur in male, more than 40 yr. Incidence rate 0.0001-0.001%. Associated with underlying reciprocal translocation and fusion of orphan nuclear receptor **NR4-A3** (**Ch.9q22**)¹

FNAC AND CT/MRI ARE IMAGING MODALITY, Diagnosed by HPE AND IMMUNOHISTOCHEMISTRY.¹

And genetically in higher institutions by **FISH by NMB** (NEUROMEDIN-B) diagnostic marker.¹

(MC) NR4A3 + EWSR1 (Ewing sarcoma a receptor 1) TAF-15(TATA binding protein asso.factor) TCF-12(transcription factor 12) TGF

Wide local excision of sarcoma and Lymph node excision and postoperative chemotherapy in form of tyrosine kinase inhibitor-SUNITINIB.^{1,2} And post-op radiotherapy IMRT to reduce recurrence.²

RECURRNCE RATE: LOCAL: 37-48%¹ METASTATIC: 50% (**MC** pulmonary)¹

CONCLUSION:

Extra skeletal myxoid chondrosarcoma is rare variety of soft tissue sarcoma requires wide local excision of mass and Lymph node followed by post op chemotherapy and radiotherapy and follow up for recurrence.

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