Abstract: Extra skeletal chondrosarcoma is rare tumour of occurrence than the chondrosarcoma of the bone. Extra skeletal chondrosarcoma has certain unique features different from the skeletal chondrosarcoma. Extra skeletal chondrosarcoma occurs in middle aged men, whereas skeletal chondrosarcoma has peak incidence in 5th to 7th decades of life. Extra skeletal chondrosarcoma has site predilection, mainly in the extremities rather than primary chondrosarcoma which most commonly occurs in the axial skeleton. Extra skeletal chondrosarcoma is slow growing tumour, usually manifests as painless swelling contrary to primary skeletal chondrosarcoma which is a fast growing tumour presenting mainly with pain. Survival rate of Extra skeletal chondrosarcoma is 70% at 10 years, better than conventional which has 53% at 10 years. Extra skeletal chondrosarcoma has got 2 variants of tumours which having different natural history. 

Keyword: Extra skeletal chondrosarcoma, Soft tissue chondrosarcoma, Myxoid type, Mesenchymal type.

INTRODUCTION:
Extra skeletal chondrosarcoma is a relatively rare but well recognised clinicopathological entity. They usually develop in the deep soft tissues of the extremities. It has also been referred to as chondroid sarcoma. 90% of chondrosarcoma are conventional primary chondrosarcoma followed by other rare types like juxtacortical chondrosarcoma, Dedifferentiated chondrosarcoma, clear cell chondrosarcoma and extra skeletal chondrosarcoma.

CASE REPORT:
29 years old male patient presented with a painless swelling in

A RARE CASE OF EXTRA SKELETAL CHONDROSARCOMA - MYXOID TYPE
CASE REPORT
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the right leg for past 6 months. Patient noticed
the swelling after a trivial trauma. It was pro-
gressively growing in nature. On examination
patient had a swelling of size 8×6 centimetres
occupying the posterior aspect of the lower
third of the right leg. Skin over the swelling is
stretched and shiny. Firm in consistency. Mo-
bile only in horizontal direction.

INVESTIGATIONS:
Routine blood investigations were within nor-
mal limits. X-ray of the patient showed soft tis-
sue shadow soft tissue shadow in the right leg
with minor stippled calcification. Cortical border
of the tibia and fibula was normal.

BIOPSY:
We did incision biopsy which showed
greyish white friable soft tissue. Section stud-
ied showed a tumour composed of interlacing
fragments of cartilage with many closely
packed chondrocytes with many of them show-
ing mononucleate, binucleate, and trinucleate
lacunae with moderate nuclear anaplasia, sur-
rounded by clusters of short spindled cells with
similar nuclear morphology. These cells are
separated by variable amount of mucoid sub-
stance. Pathologist reported as myxoid chon-
drosarcoma.

TREATMENT:
We planned for wide excision of
the tumour. Through posterior approach we
opened the lower half of leg. There was a
mass of size 10×6 cm which was well capsu-
lated. Tendo achillis not involved. We sepa-
rated it from the surrounding soft tissue and
excised out. Histopathological report again
confirmed the diagnosis. Excised tissue mar-
gins were showed normal clearance. Postop-
erative period is uneventful. At 6 months fol-
lowup patient is symptom free. there was no
reccurence of the tumour. patient on regular
follow up.

DISCUSSION:
Extra skeletal chondrosarcoma is extremely
rare condition which has a predilection for mid-
dle aged men and most commonly affects the
lower extremity.

Two different varieties are described
1. Myxoid type
2. Mesenchymal type

Myxoid chondrosarcoma occurs
more frequently in lower limbs, deep
to the fascia in muscular compart-
ments. It has a very low fluid density
due to the richly mucinous material,
whereas mesenchymal type shows a
high density due to calcification. Mes-
enchymal chondrosarcoma may arise
in any mesenchymal tissue, some-
times in an irradiated area.

Painless slow growing soft
tissue swelling will be the chief com-
plaints in both groups. Sometimes it
may present with vague pain in the
swelling.

IMAGING:
A well defined soft tissue neoplasm,
often speckled with calcification, is
more indicative of a mesenchymal
type than myxoid chondrosarcoma
because in second type there is no
intrapomoural calcification or only mi-
nor stippling. Isotope scan show up-
take in both types of tumour but is
more pronounced in mesenchymal
chondrosarcoma. In angiography
mesenchymal chondrosarcoma
shows a hypervascularisation of the
neoplasm with many broadened capillaries.

MACROSCOPIC FEATURES:
Myxoid chondrosarcoma presented
as ovoid, soft, nodular masses, well
circumscribed by a distinct pseudo-
capsule. On section surface ap-
peared translucent, gelatinous and
grey to brown in colour with large
haemorrhagic and sometimes ne-
crotic areas. Mesenchymal chon-
drosarcoma were multilobulated and
circumscribed, firm in
consistency with no well defined pseudocapsule. Cut section showed a pale grey, fleshy surface with scattered cartilaginous foci, which creaked on cutting. Areas of necrosis were abundant while haemorrhagic zones were scanty.

HISTOLOGICAL FEATURES:
Myxoid chondrosarcoma were multilobulated and well circumscribed by a condensed connective tissue capsule, which had resulted from the expansile growth of the tumour. Fibrous septae of variable thickness with a few blood vessels extended in from the pseudocapsule. There was a narrow rim of deeply eosinophilic and sometimes vacuolated cytoplasm around the small, ovoid hyperchromatic nucleus. These cells were separated by variable amount of mucoid ground substance which was weakly basophilic with Haematoxylin and eosin

Mesenchymal chondrosarcoma showed a characteristic pattern of two basic cellular elements. They are undifferentiated round and small nodules of well differentiated tumour cartilage. Mesenchymal cells had ovoid or elongated hyperchromatic nuclei and scanty, poorly outlined cytoplasm. PAS stain was always negative. Silver stain showed a well defined pattern of reticulin fibres around small groups of undifferentiated cells and vessels.

TREATMENT:
- Large tumour with intra-articular invasion
- Osseus erosion
- Major neurovascular involvement

Local recurrence varies from 20% to 60% with a median of 49%. There is no role for chemotherapy and radiotherapy. Rate of metastases of myxoid is less than mesenchymal. Most common site of metastases is pulmonary. It is treated by pulmonary lobectomy or local excision. The one year survival rate was 82.0 percent and five year survival rate was most likely 81.2 percent.

CONCLUSION:
Extra skeletal chondrosarcoma includes two histologically well defined and distinct subtypes. They differ both clinical and radiographic features and in prognosis. Of which myxoid chondrosarcoma has very good prognosis after surgery if detected and treated early. We present this case for its rarity and good prognosis after surgery if detected early.

REFERENCES:
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<thead>
<tr>
<th></th>
<th>MYXOID</th>
<th>MESENCHYMAL</th>
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<tbody>
<tr>
<td>Age</td>
<td>&gt;35 years</td>
<td>&lt;35 years</td>
</tr>
<tr>
<td>Clinical</td>
<td>Small mass</td>
<td>Large mass</td>
</tr>
<tr>
<td>Radiograph</td>
<td>Little or no calcification</td>
<td>Irregular calcification</td>
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<tr>
<td>CT scan</td>
<td>Low homogeneous density</td>
<td>Irregular density</td>
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<tr>
<td>Bone scan</td>
<td>Faint uptake</td>
<td>High uptake</td>
</tr>
<tr>
<td>Angiography</td>
<td>Hypervascular margin</td>
<td>Avascular centre</td>
</tr>
<tr>
<td></td>
<td>Diffuse hypervascularity</td>
<td></td>
</tr>
<tr>
<td>Invasion</td>
<td>Wide</td>
<td>Radical</td>
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<tr>
<td>Chemo/Radiotherapy</td>
<td>Useless</td>
<td>Probably useful</td>
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<tr>
<td>Local recurrence</td>
<td>Rare after wide excision</td>
<td>Frequent</td>
</tr>
<tr>
<td>Metastases</td>
<td>Rare or late</td>
<td>Common &amp; early</td>
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