MAGICAL OF SOFT TISSUE TUMOR

Associate Professor May Thwe Lin

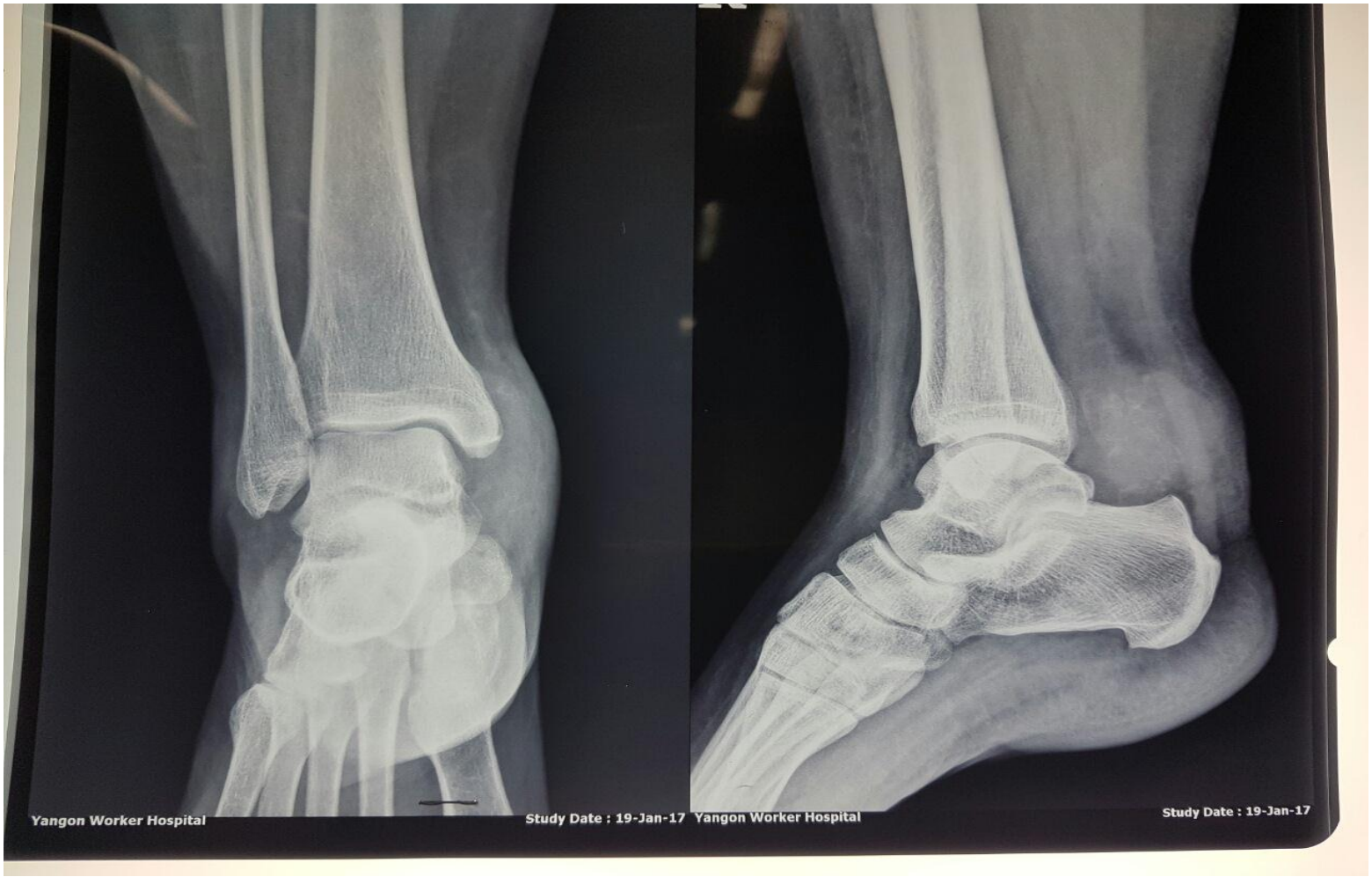
Professor Myint Myint Nyein

Professor Myat Mon

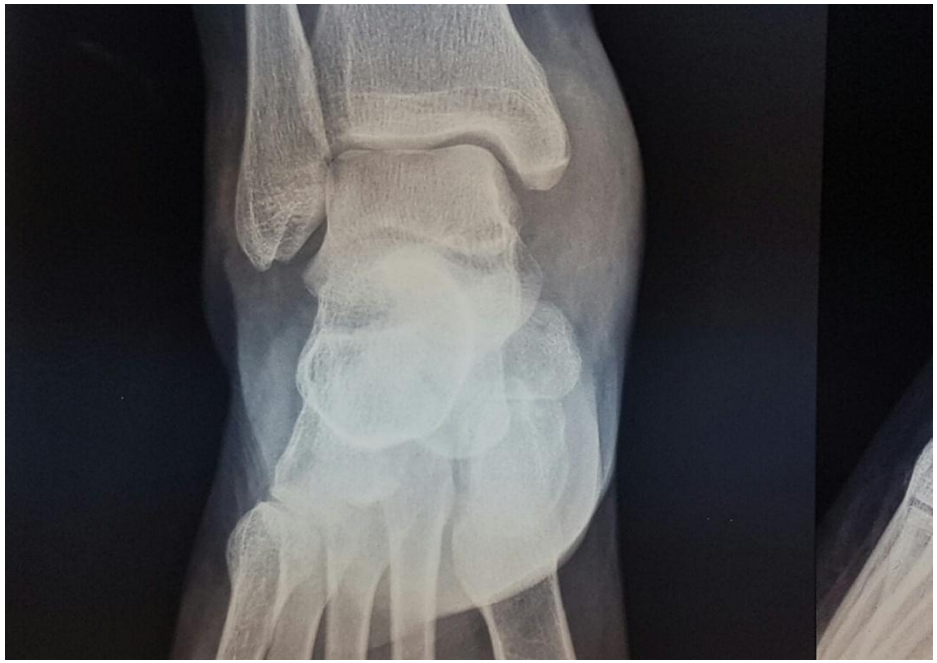
Case History

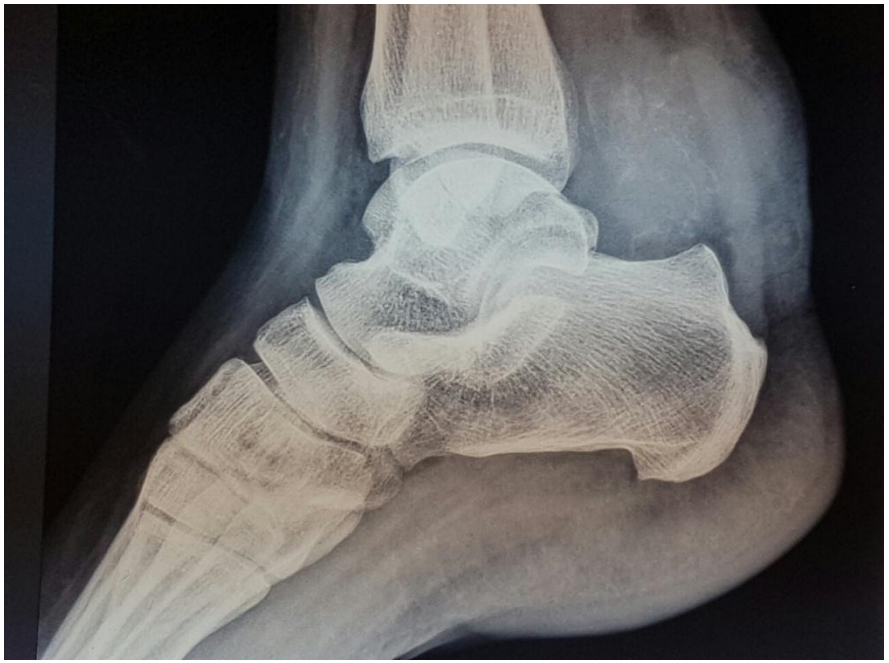
- 35 years old female, presenting with swelling over right ankle for 4 months duration.
- Not painful
- No previous history of trauma

XR of right ankle joint



XR of right ankle joint



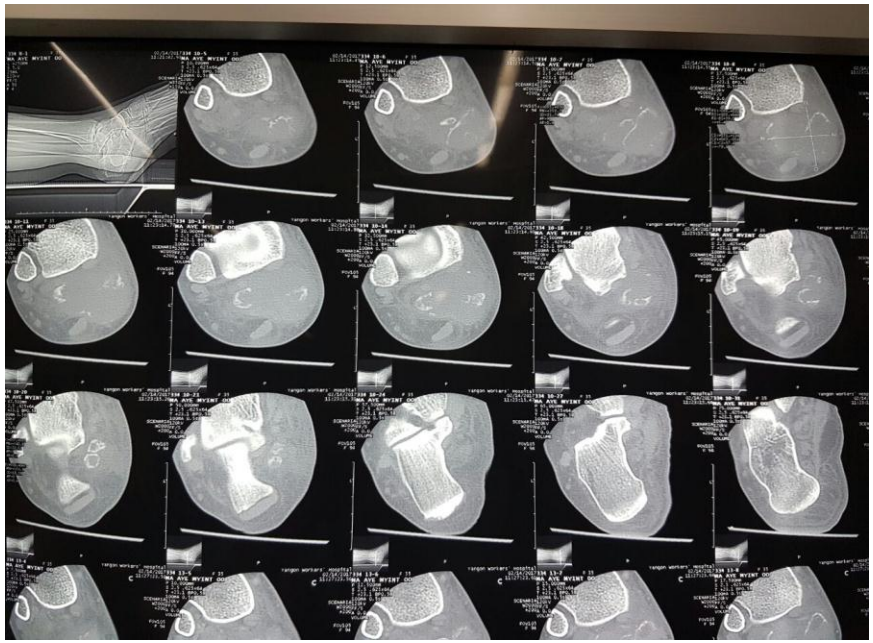


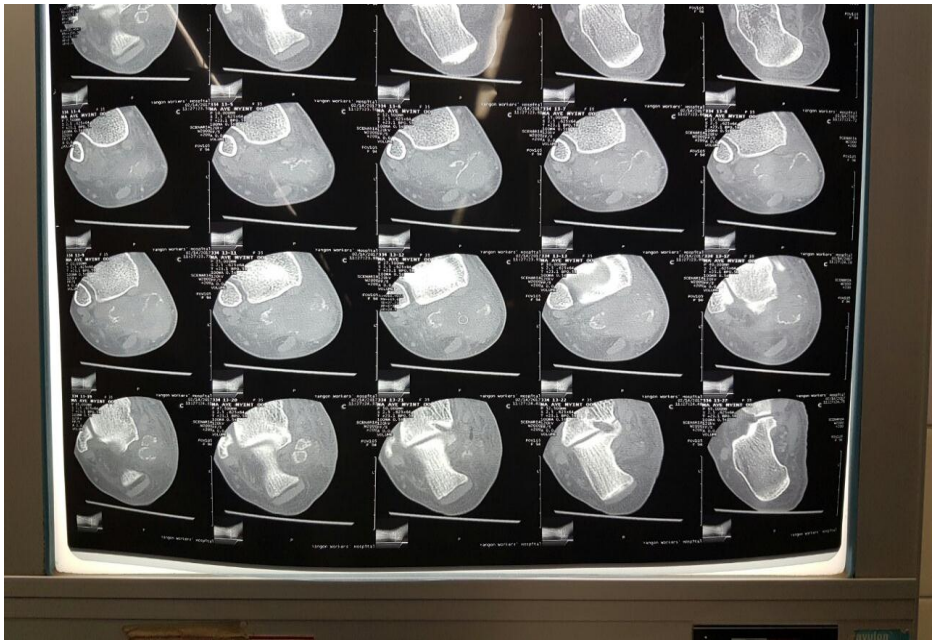
No obvious bony (or) joint lesion is seen apart from calcaneal spurs.

XR of right ankle joint

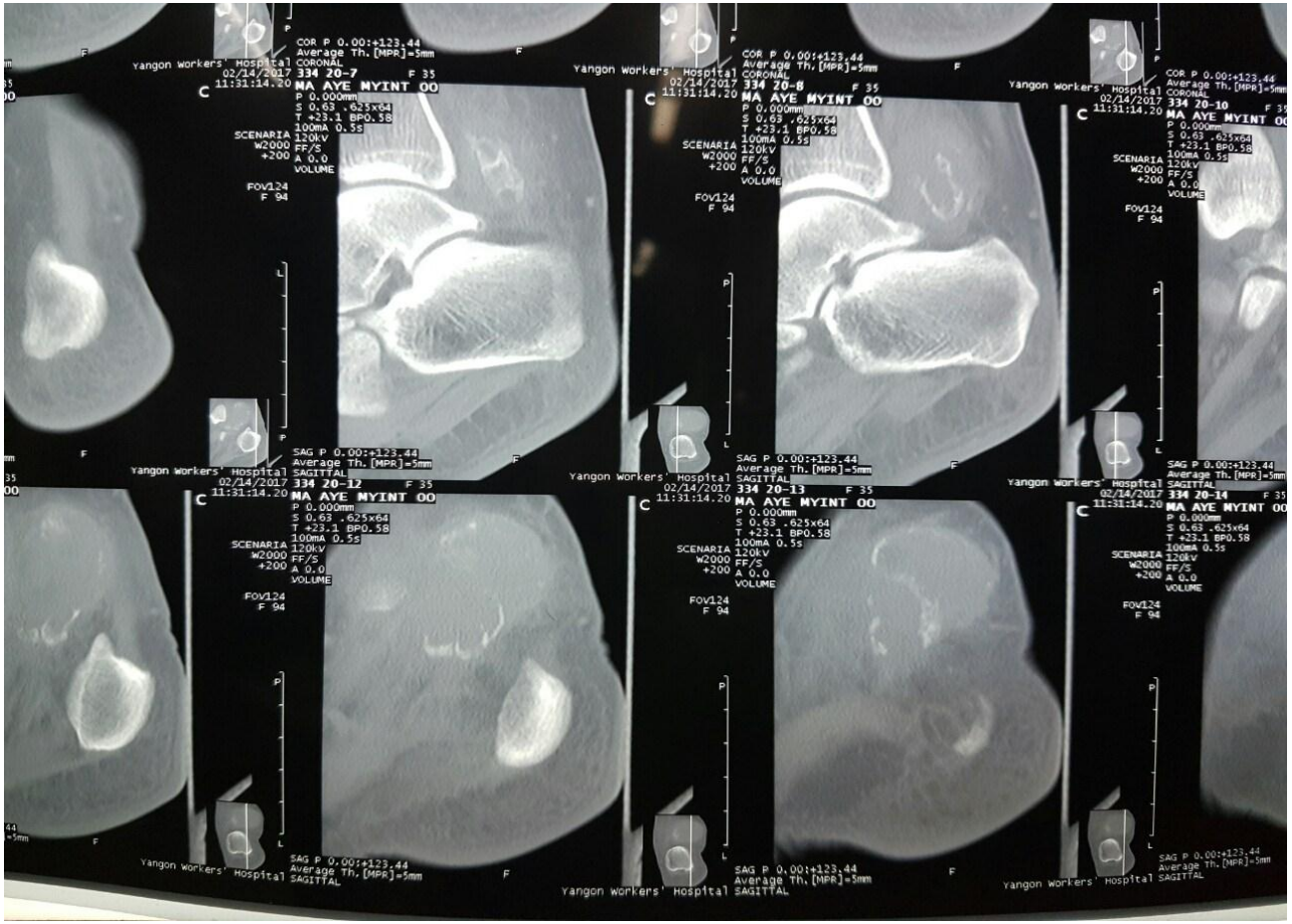
- showed no osteolytic lesion and no expansile tumor mass on adjacent bone.

CT (Right ankle joint)

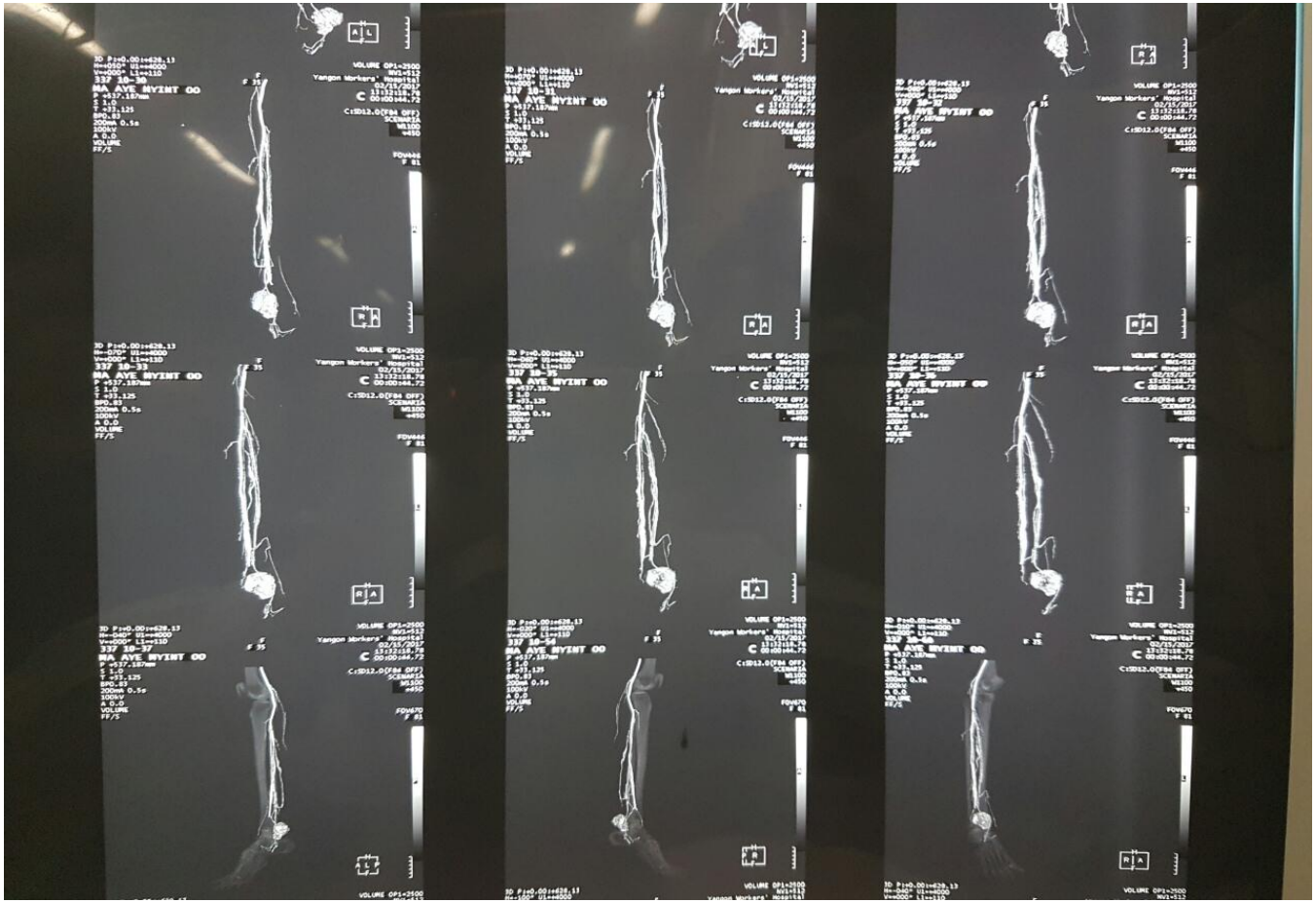




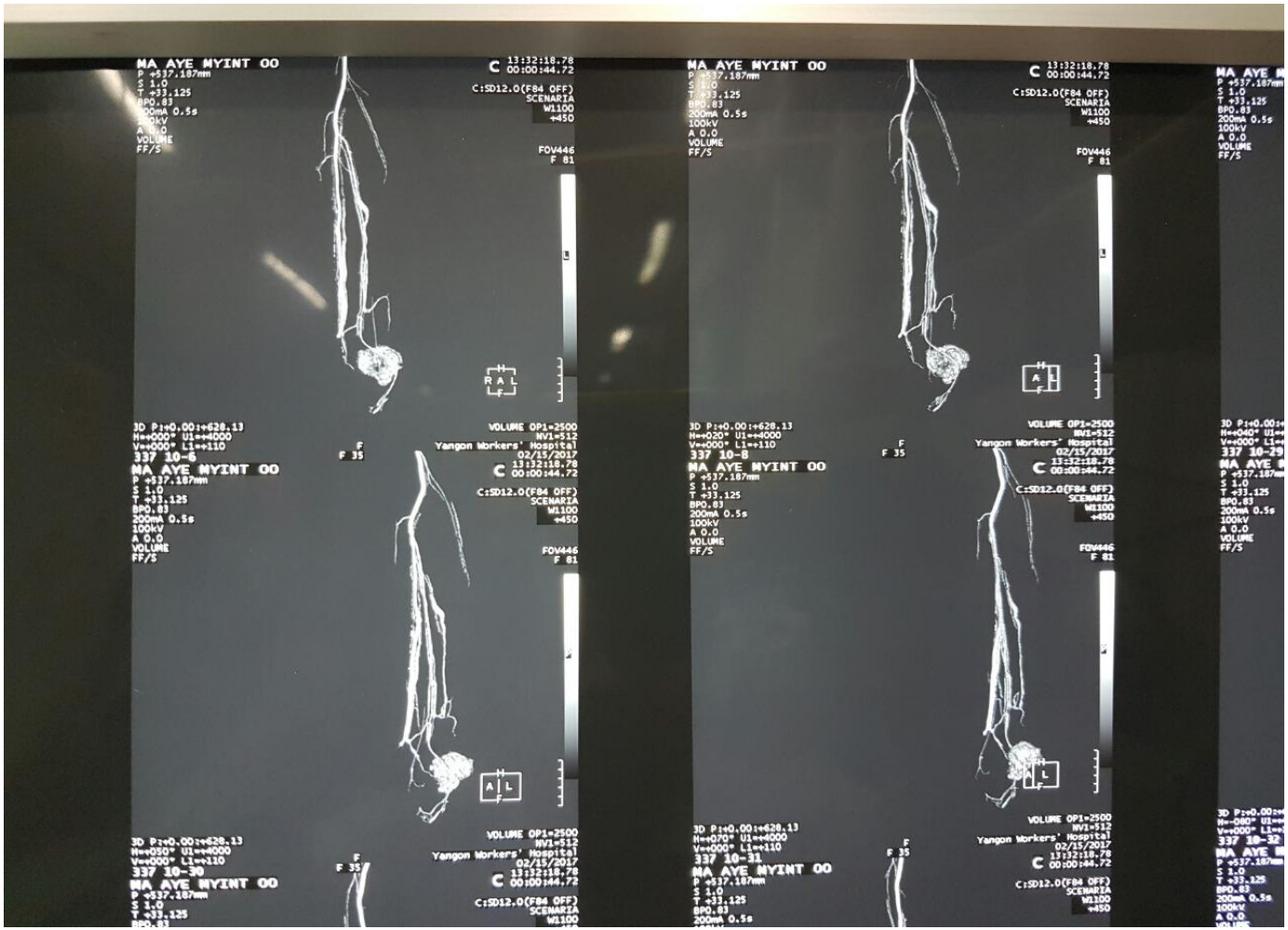
CT (Right ankle joint)



Rt lower limb angiogram



Rt lower limb angiogram



CT & Angiogram Report

- The study was done without and with IV contrast with 128 slices CT scanner.
- About 4.5x 3.9x 4 cm in size , **a soft tissue mass** noted in just below the medial malleolus of right ankle joint, calcifications are noted within the lesion.
- Marked contrast enhancement is seen.
- **No bony erosion or periosteal reaction** is seen in adjacent bone.

CT & Angiogram Report

- No aneurysm is seen in right lower limb angiogram. Small vessels from anterior and posterior tibial vessels noted within the lesion.

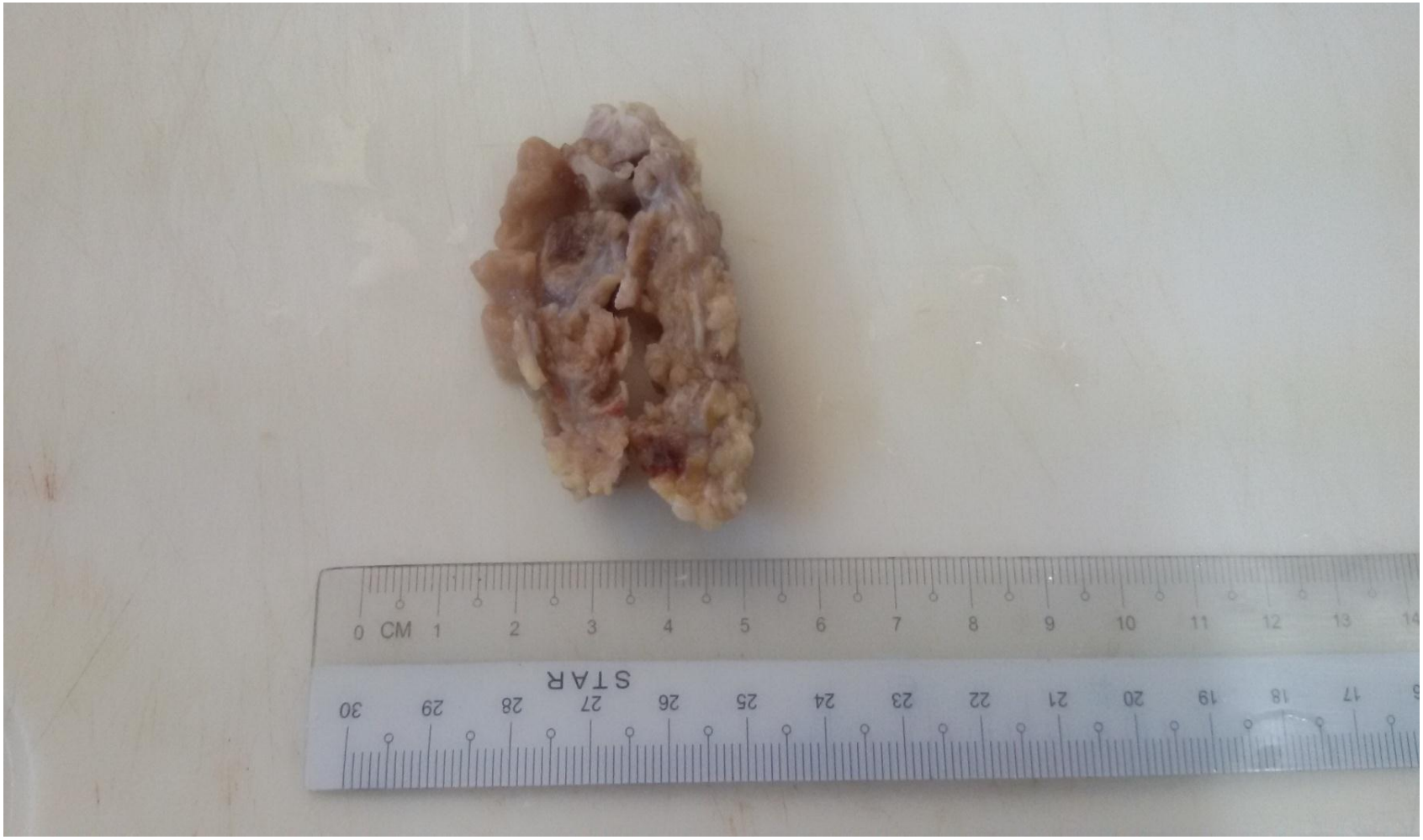
CT & Angiogram Result

- **Features are suggestive of Arteriovenous haemangioma.**

- Excisional Biopsy taken from swelling over right ankle joint.

Gross

- Resected soft tissue mass measured 5x3.5x1.2 cm.
- Cut section showed friable tumor with surrounding fragments of bony tissues and haemorrhage.

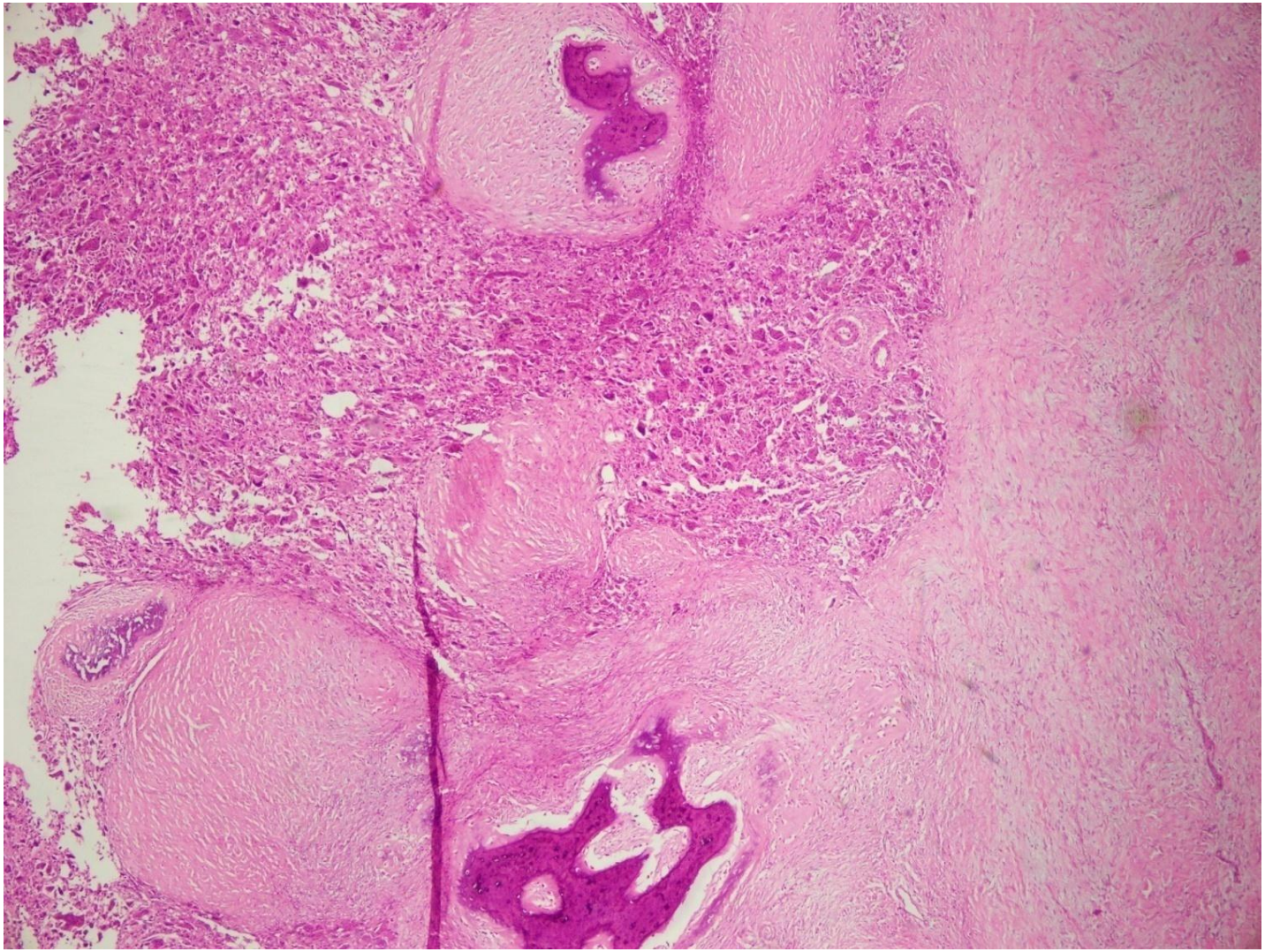

0 CM 1 2 3 4 5 6 7 8 9 10 11 12 13
STAR
30 29 28 27 26 25 24 23 22 21 20 19 18 17

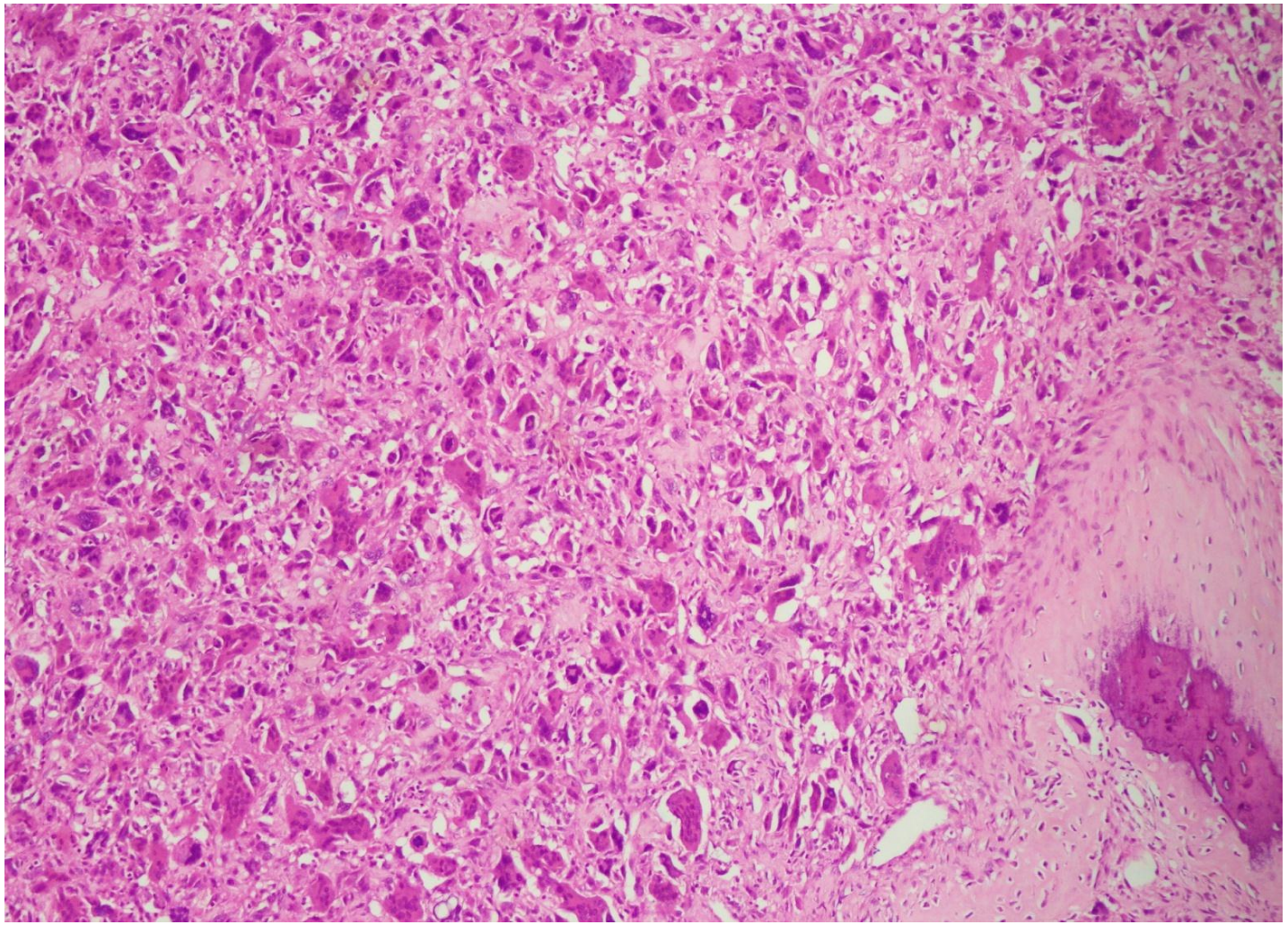
Histology

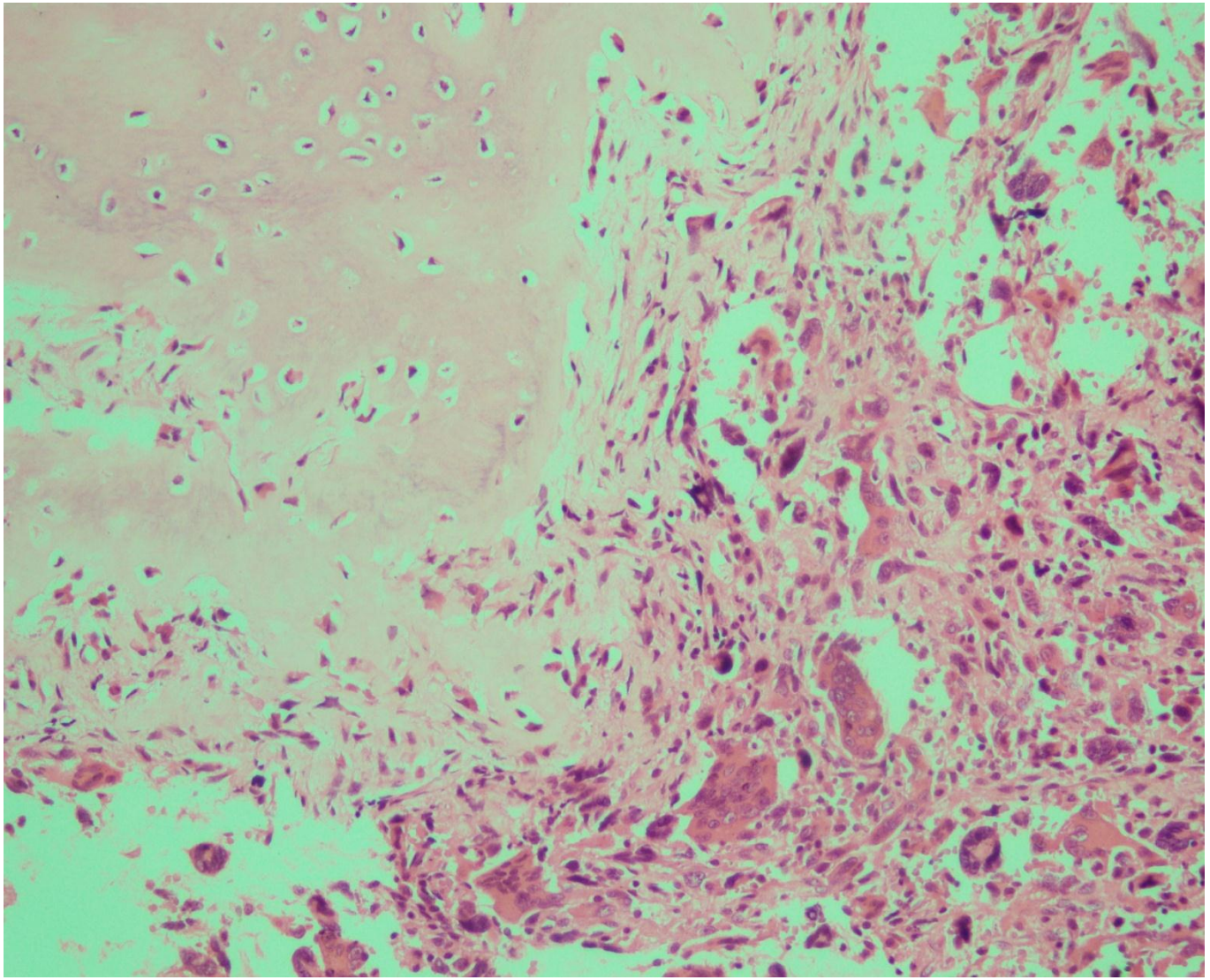
- Histological sections taken from soft tissue swelling of right ankle region show an invasive tumor composed of highly cellular sheets of pleomorphic oval to spindle shaped neoplastic cells showing marked cellular and nuclear atypia. These cells are infiltrating into surrounding normal bony trabeculae and adjoining fibrocollagenous tissues.

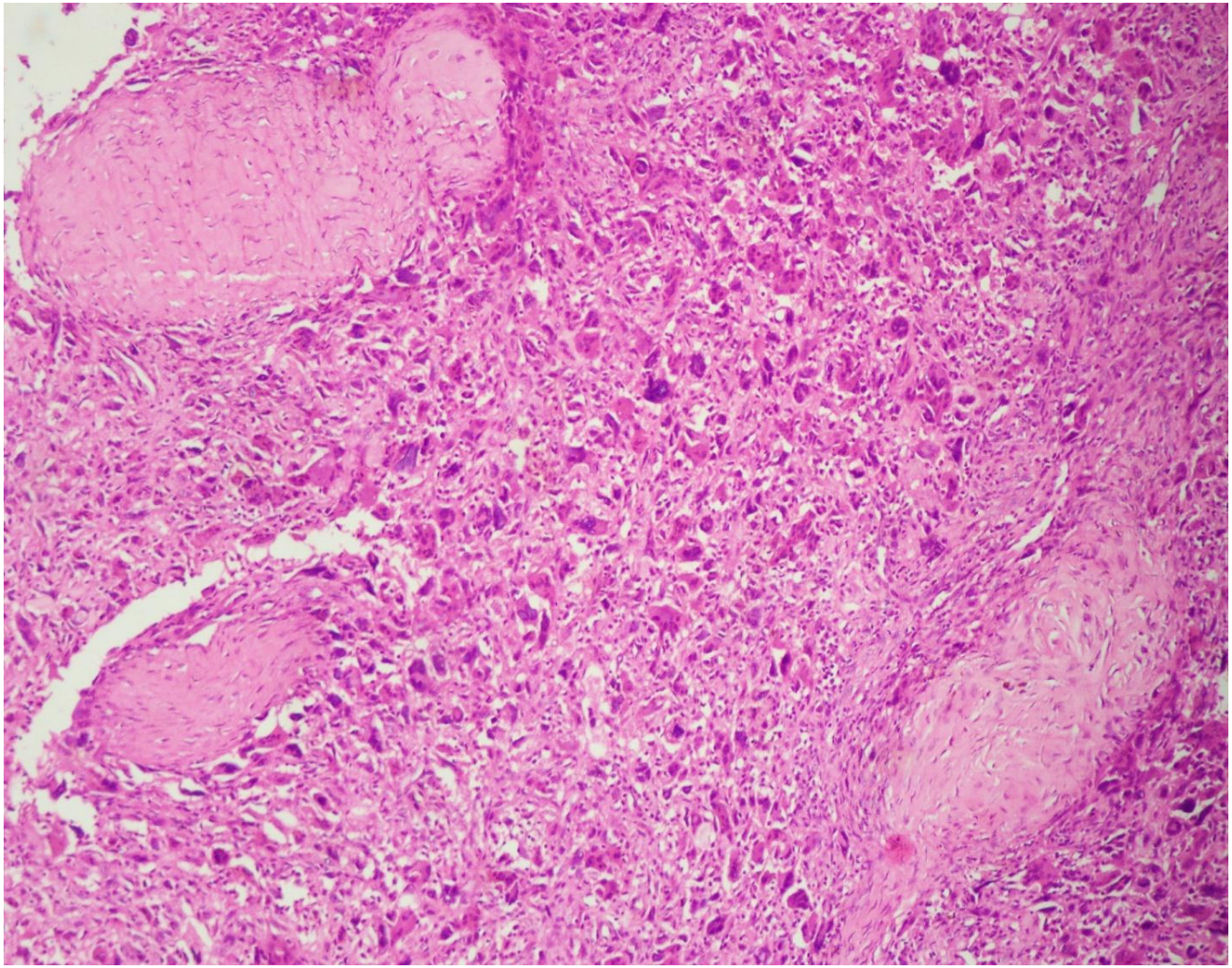
Histology

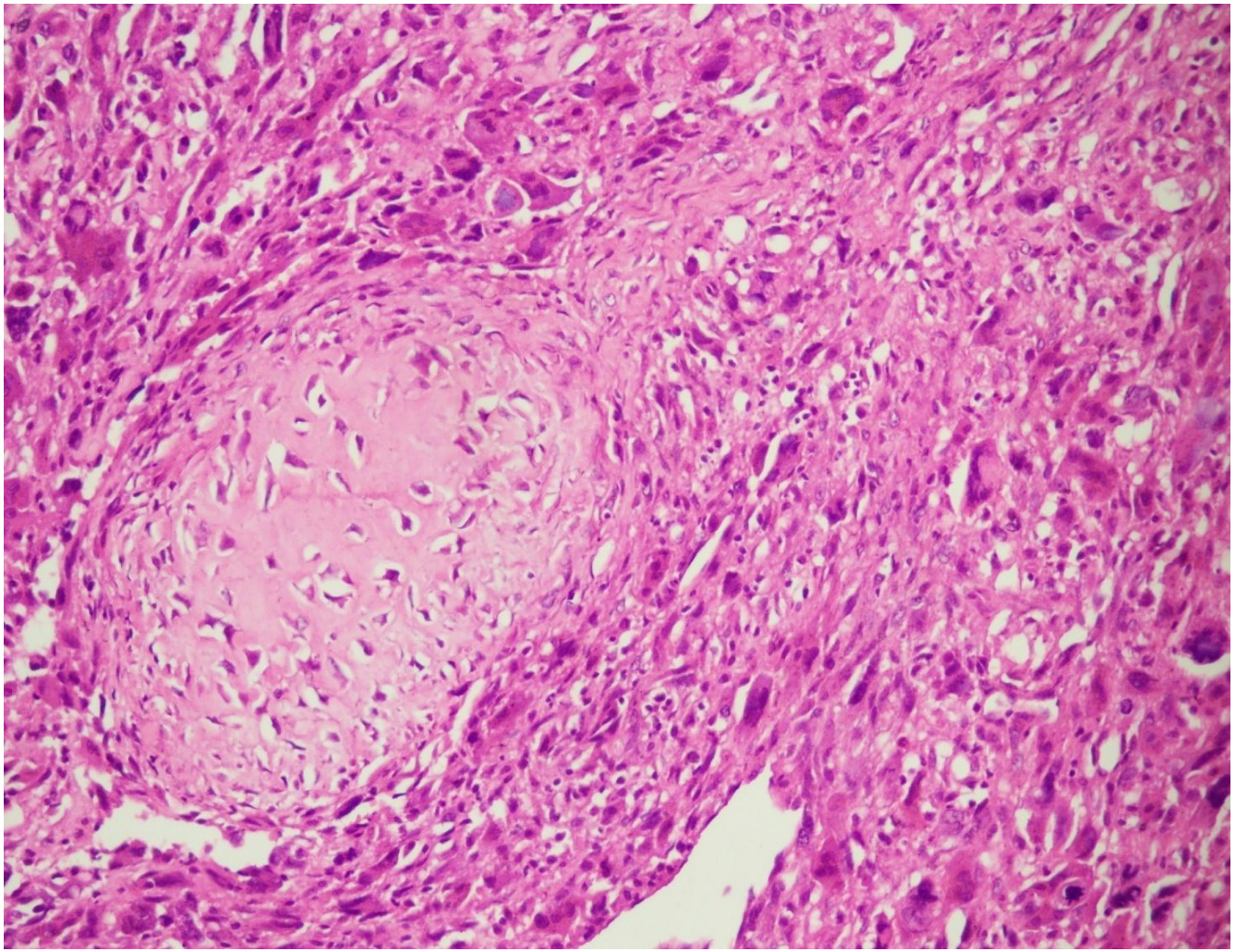
- Numerous multinucleated osteoclast like giant cells and bizarre tumor giant cells are also seen. Mitoses are frequent. There is no tumor osteoid. Areas of haemorrhage and necrosis are also present. Resection margins are not free from tumor.

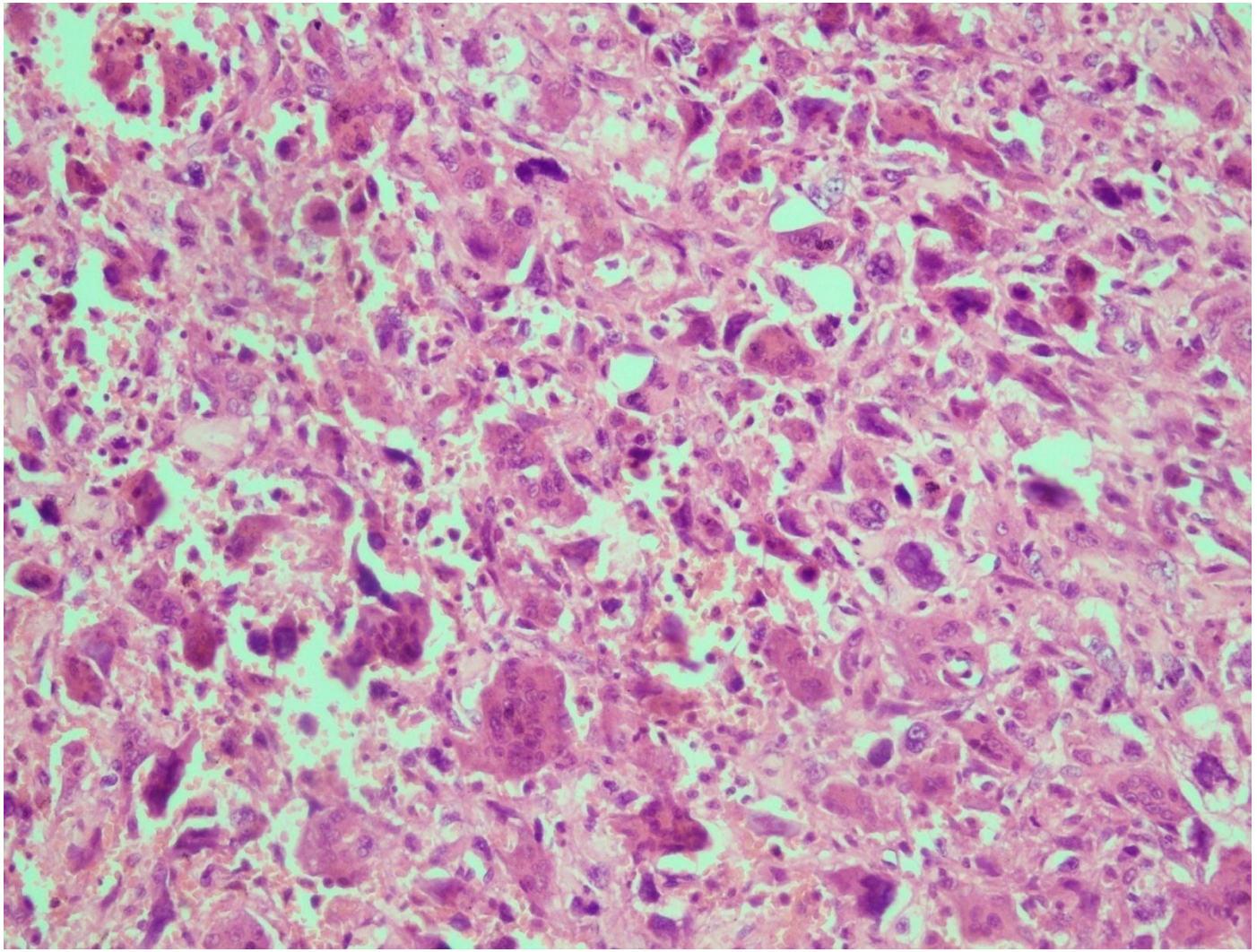


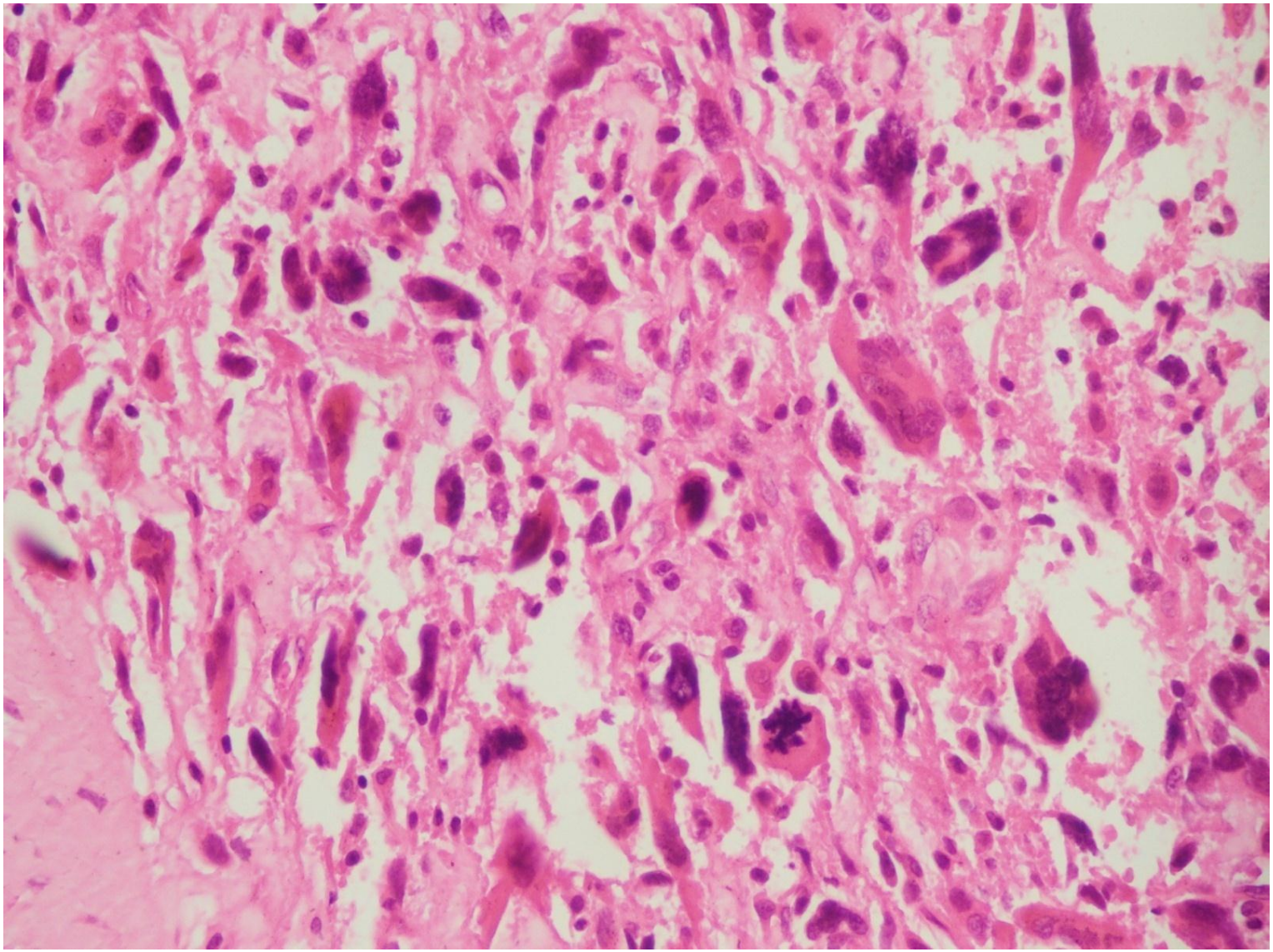


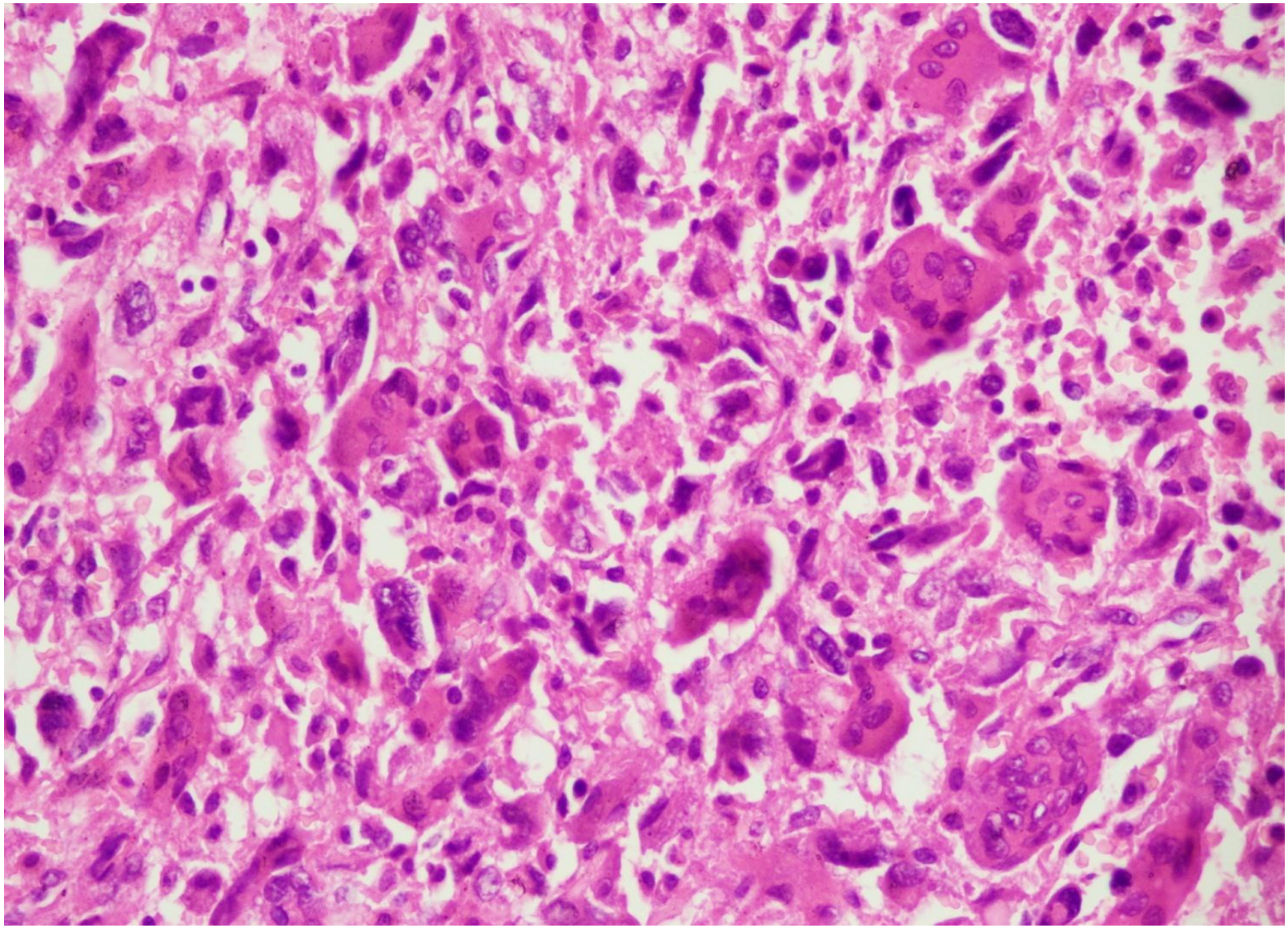


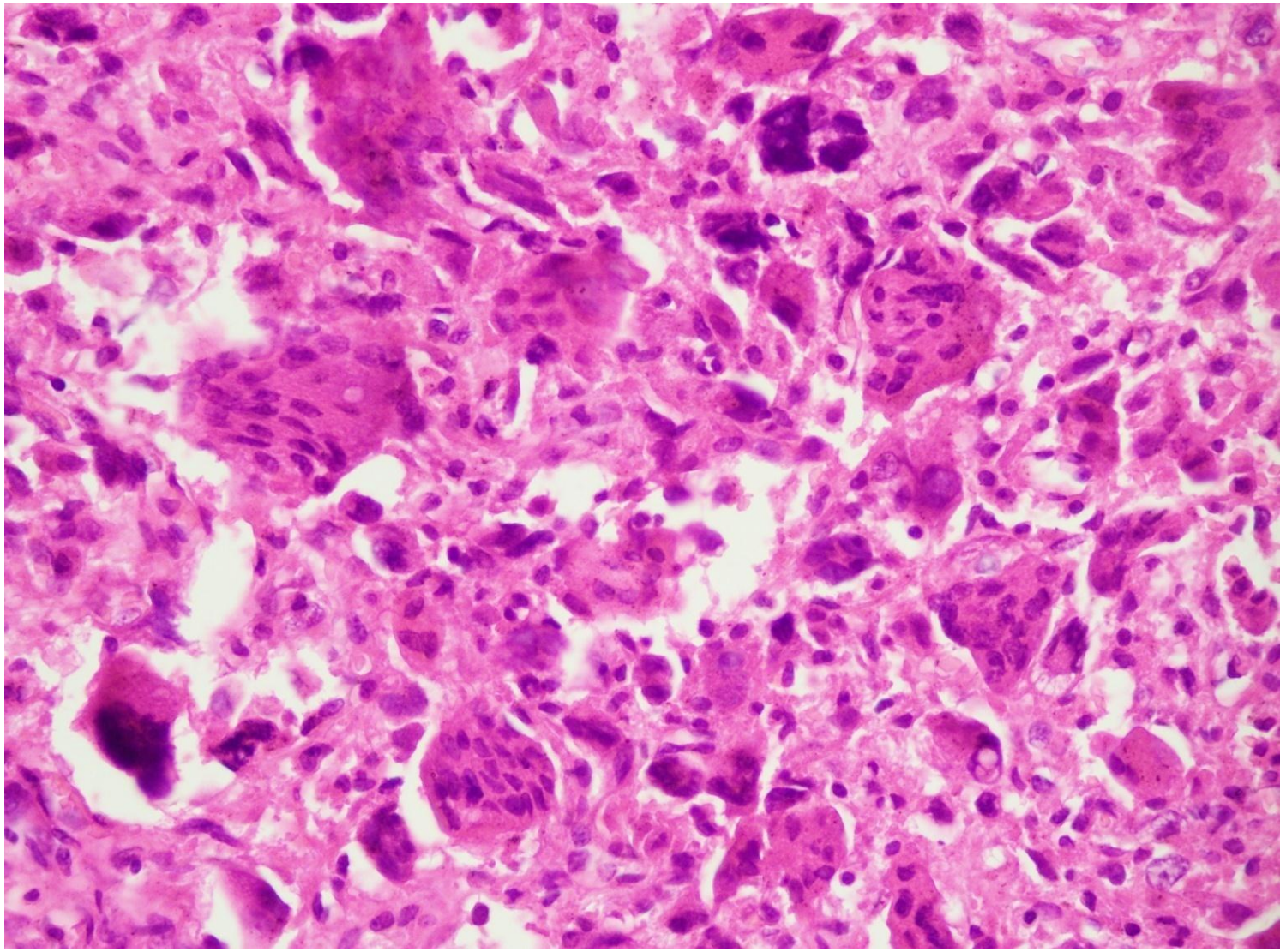












Diagnosis

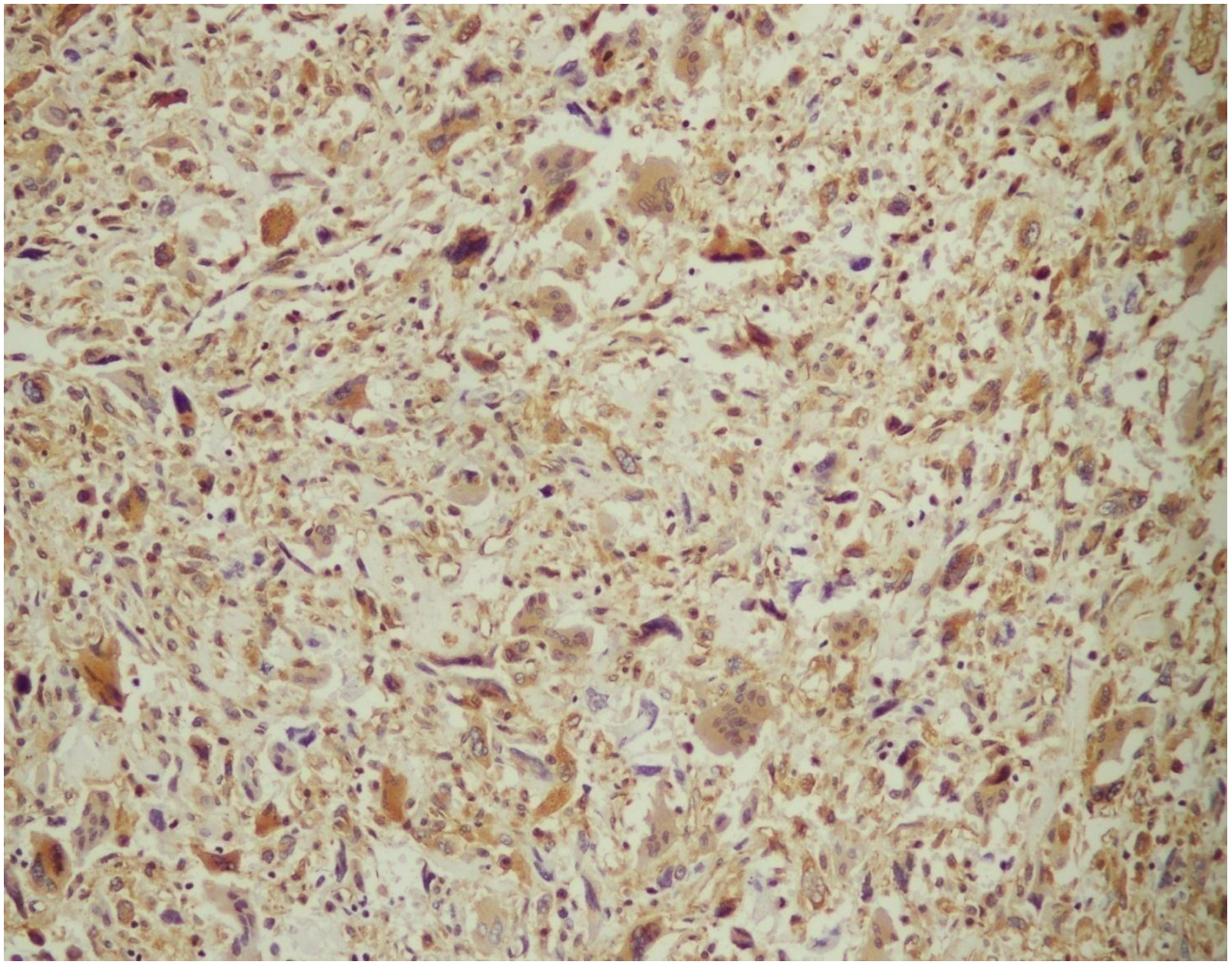
- Compatible with **giant cell rich high grade pleomorphic spindle cell sarcoma** (Right ankle region)

Differential Diagnosis

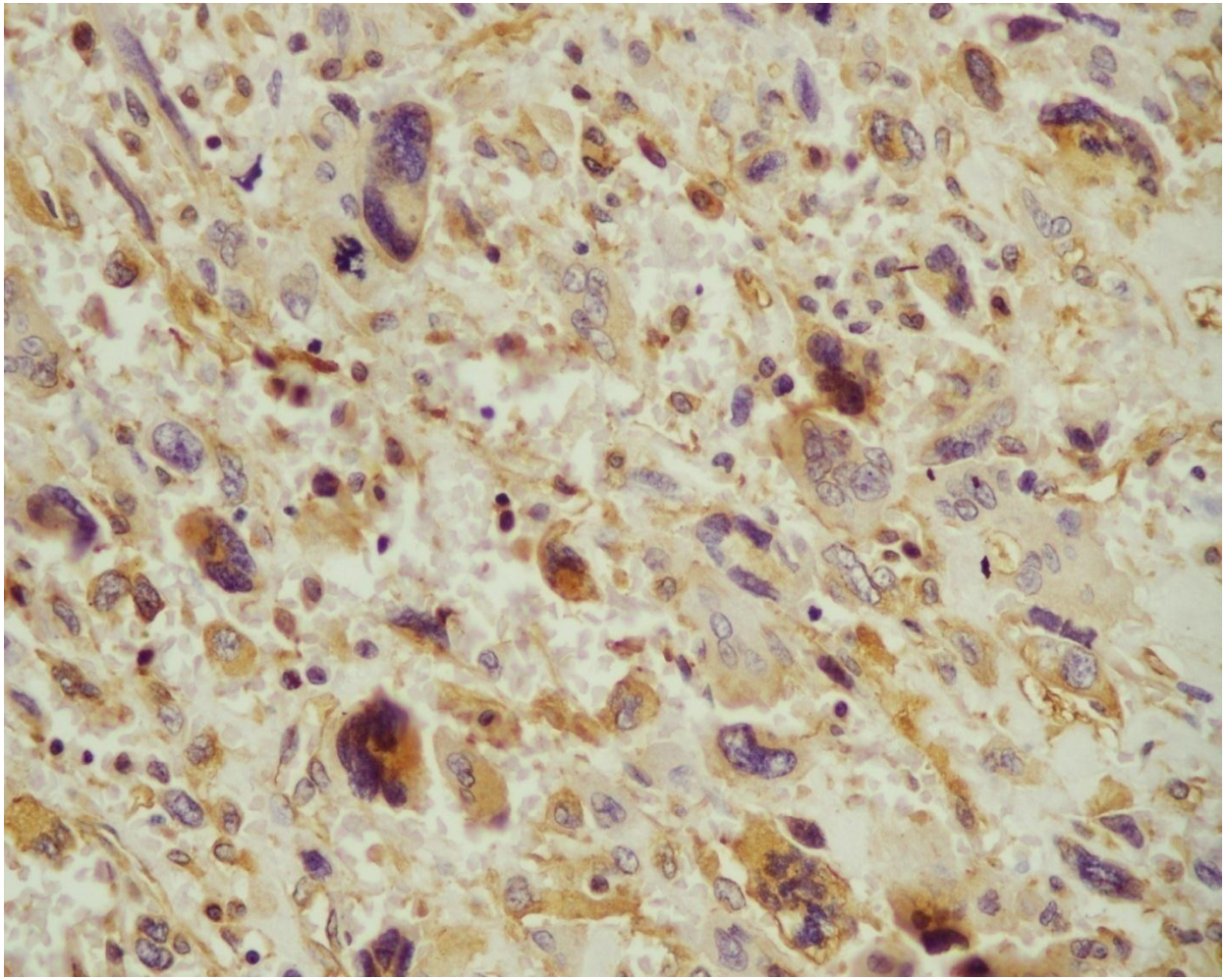
1. Malignant GCT of bone
2. Extraskeletal osteosarcoma
3. Giant cell rich amelanotic melanoma
4. High grade leiomyosarcoma
5. Anaplastic spindle cell carcinoma
6. UPS storiform/ pleomorphic

Need to do IHC for confirmation

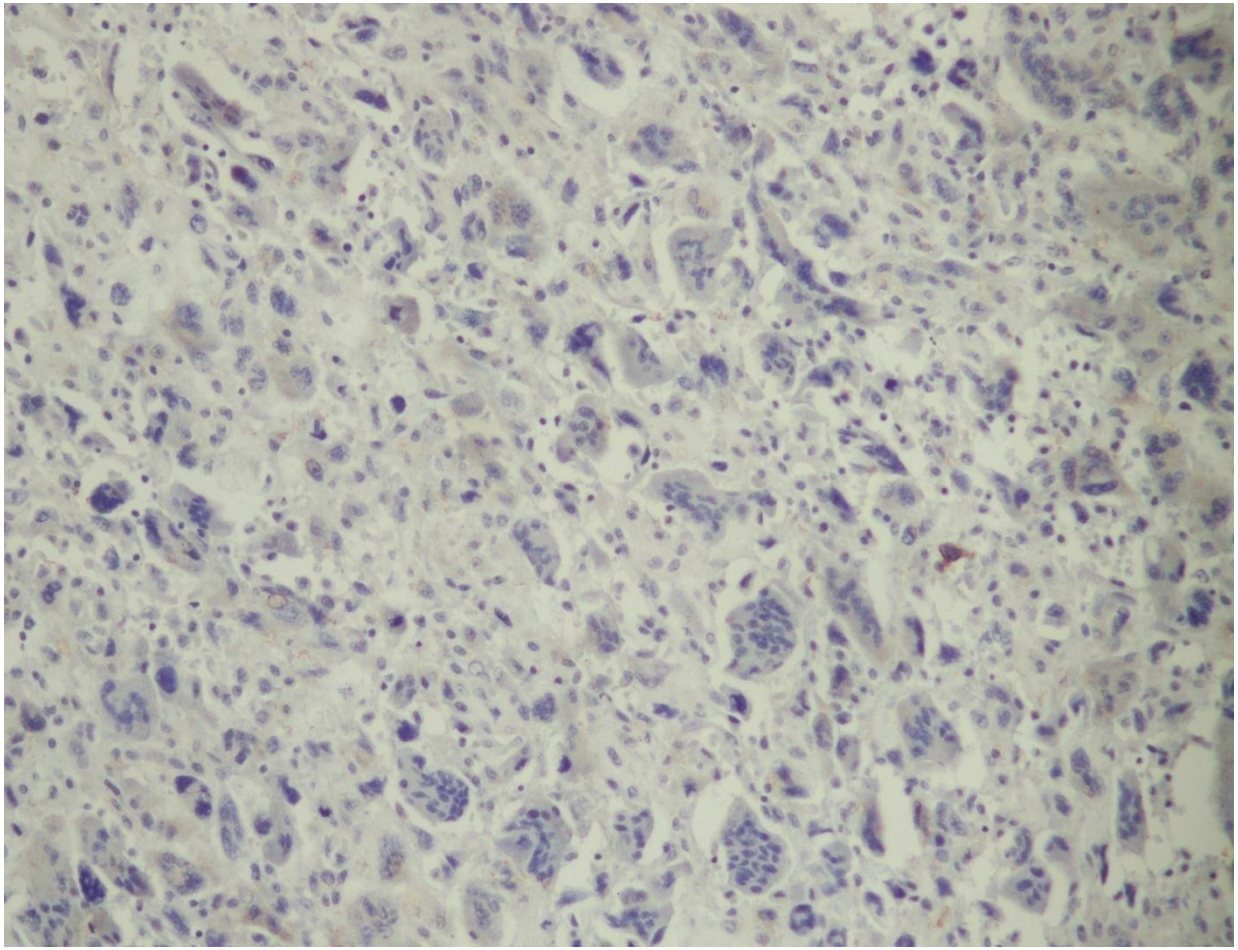
Vimentin



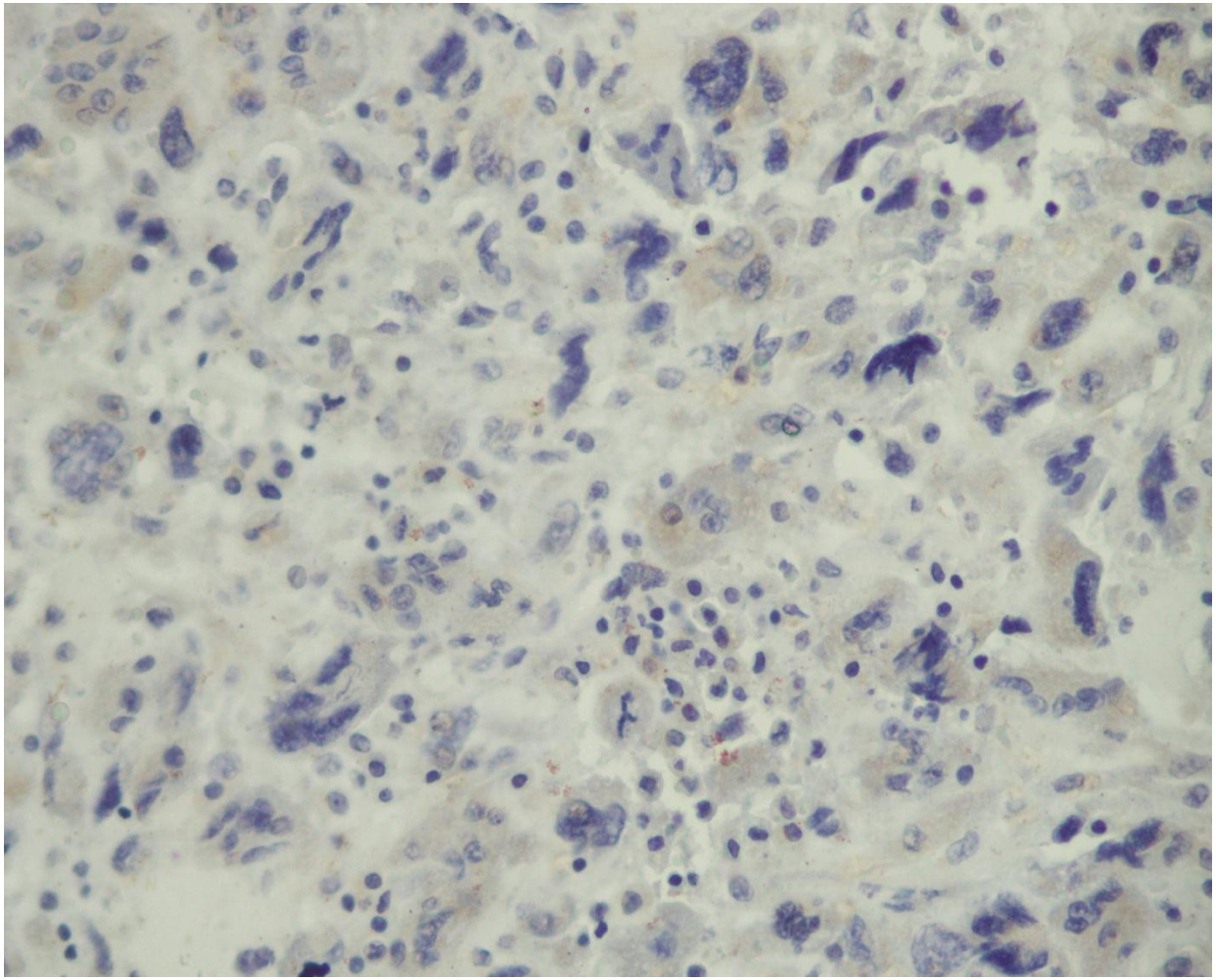
Vimentin



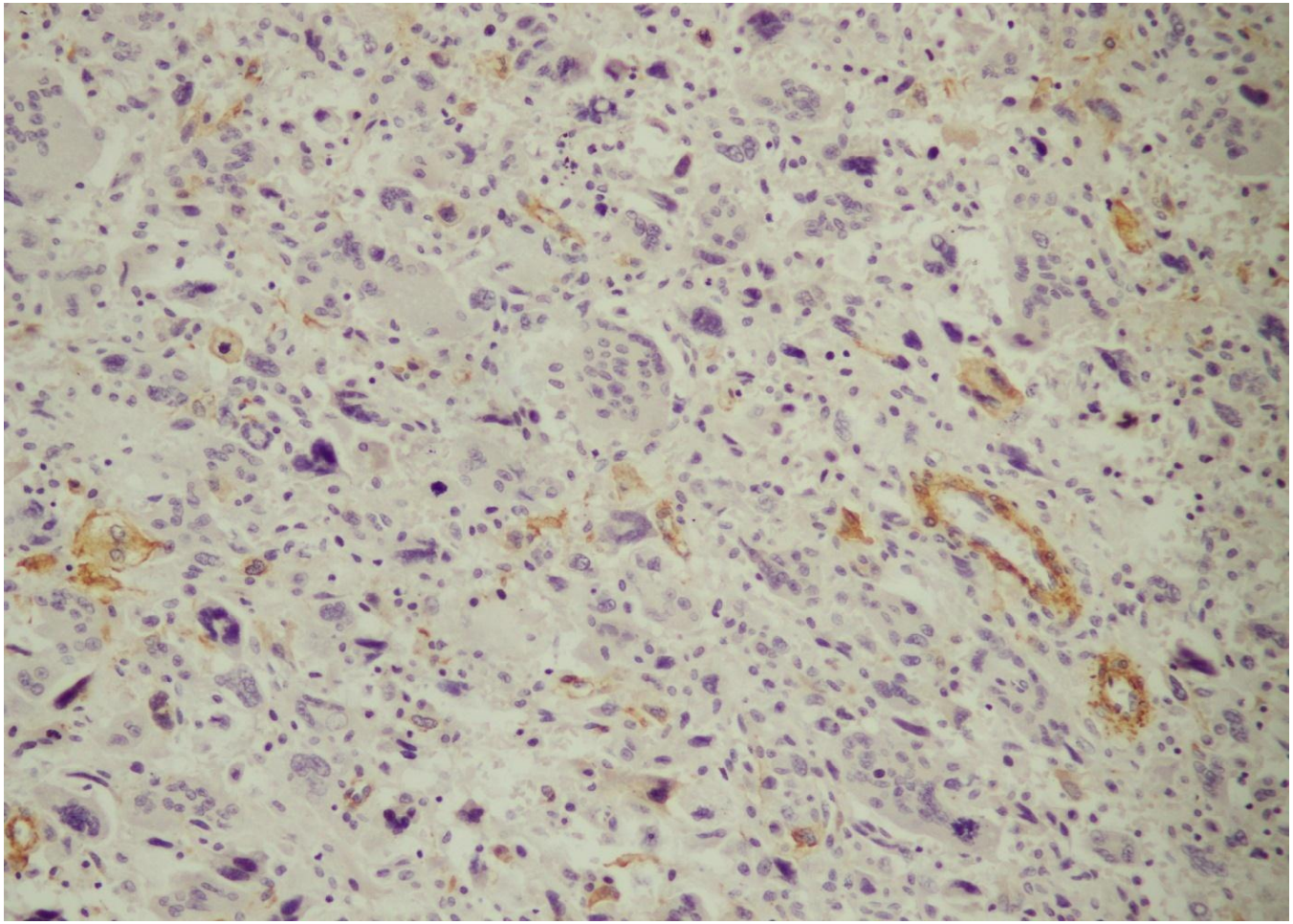
Desmin



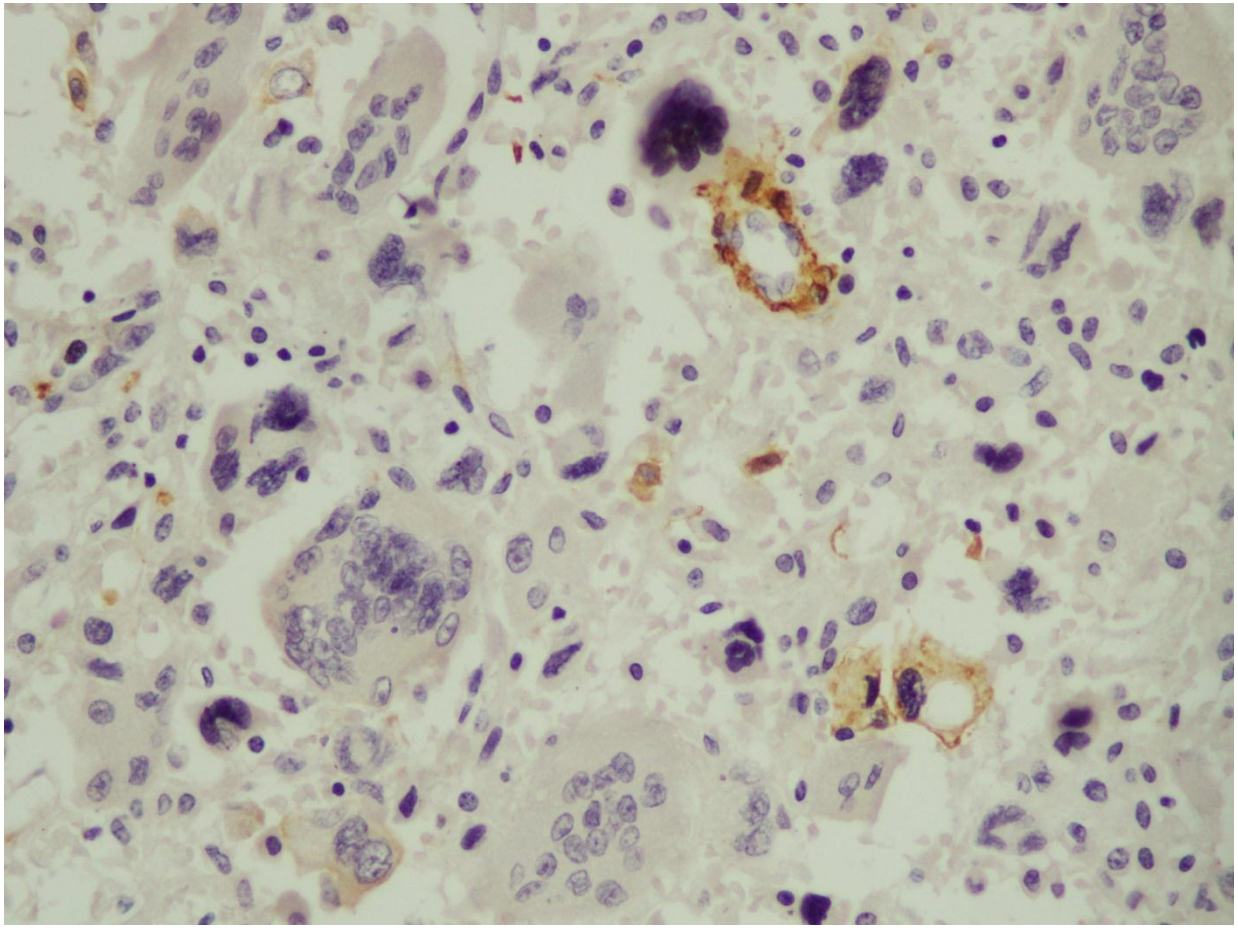
Desmin



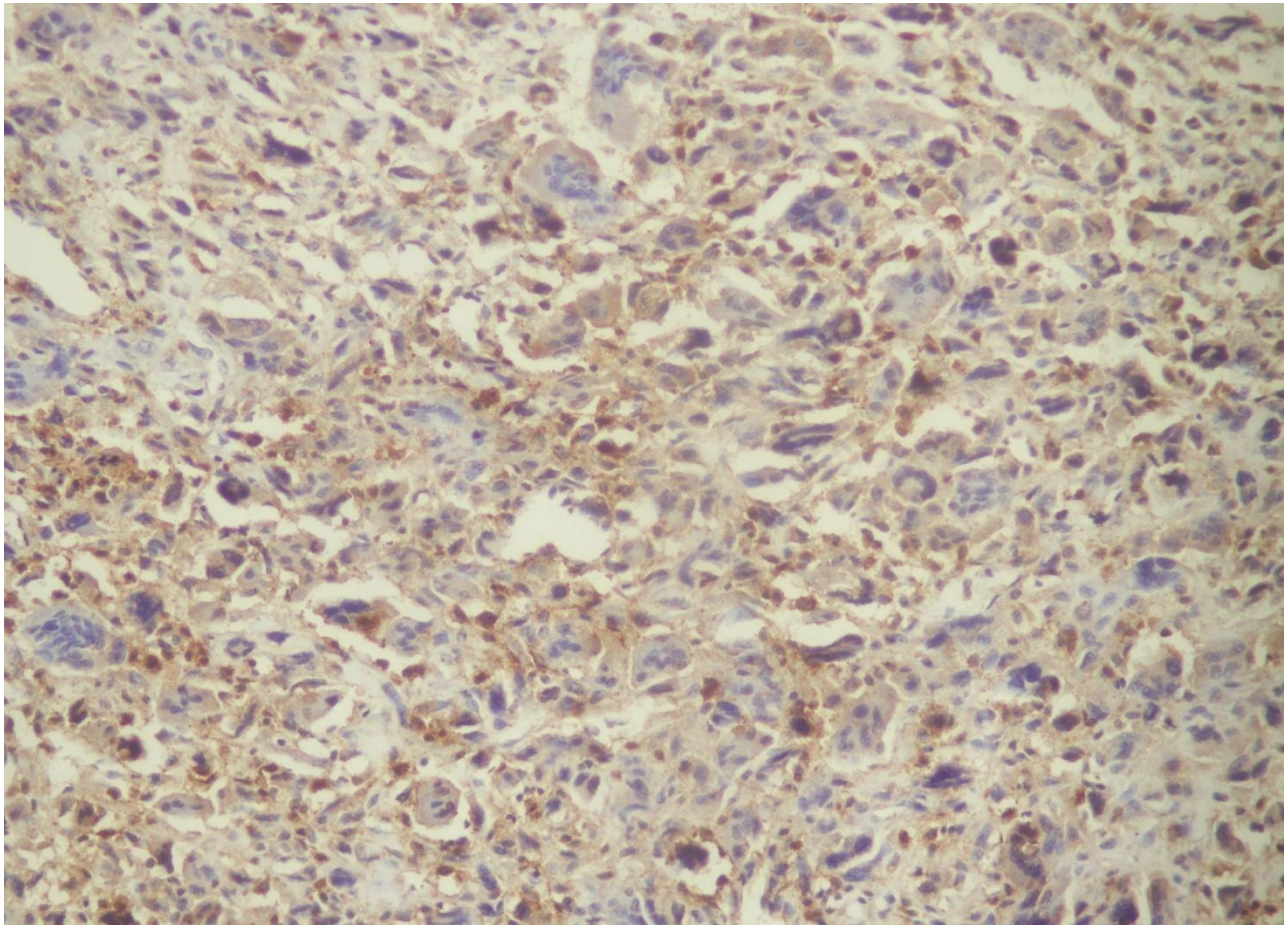
SMA



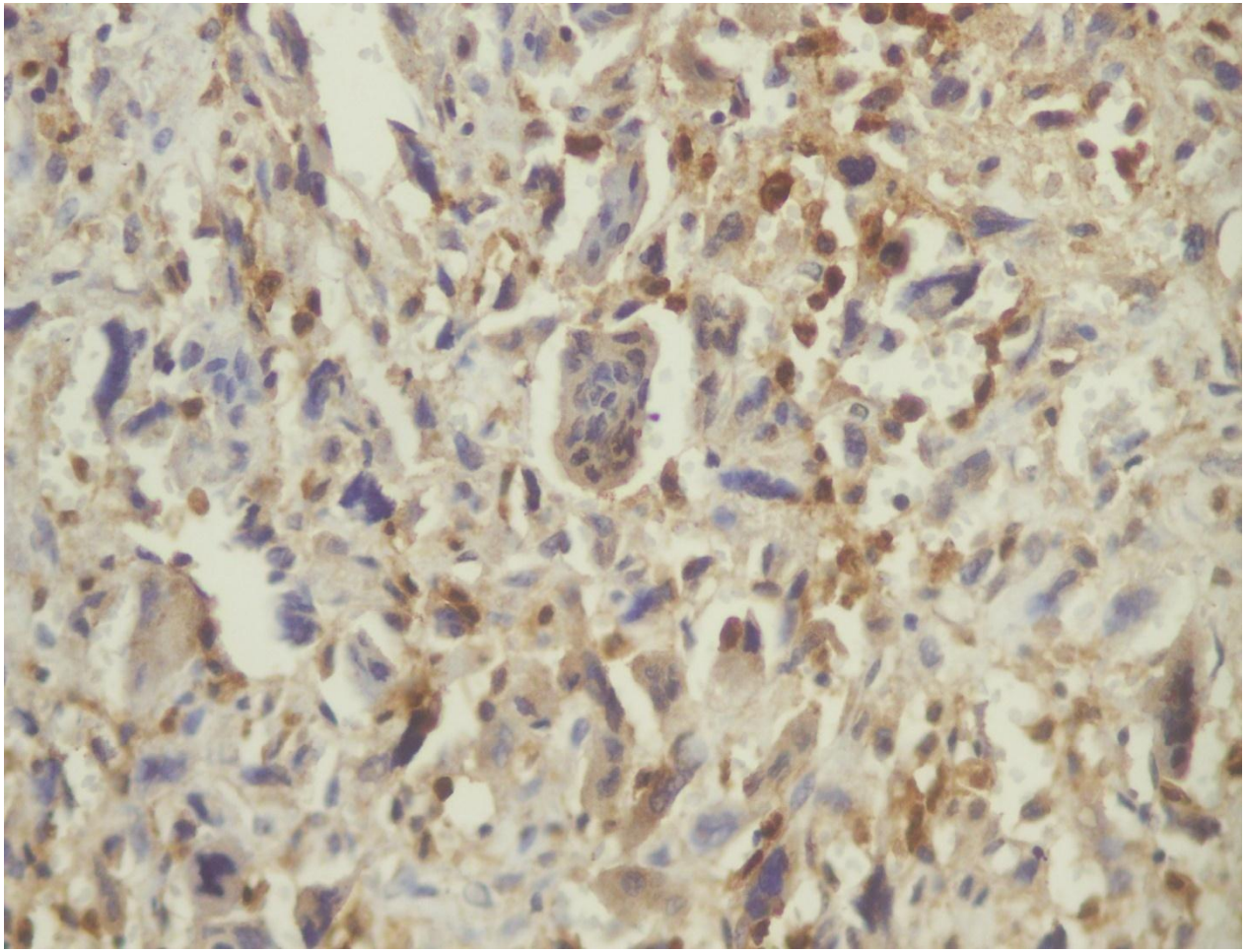
SMA



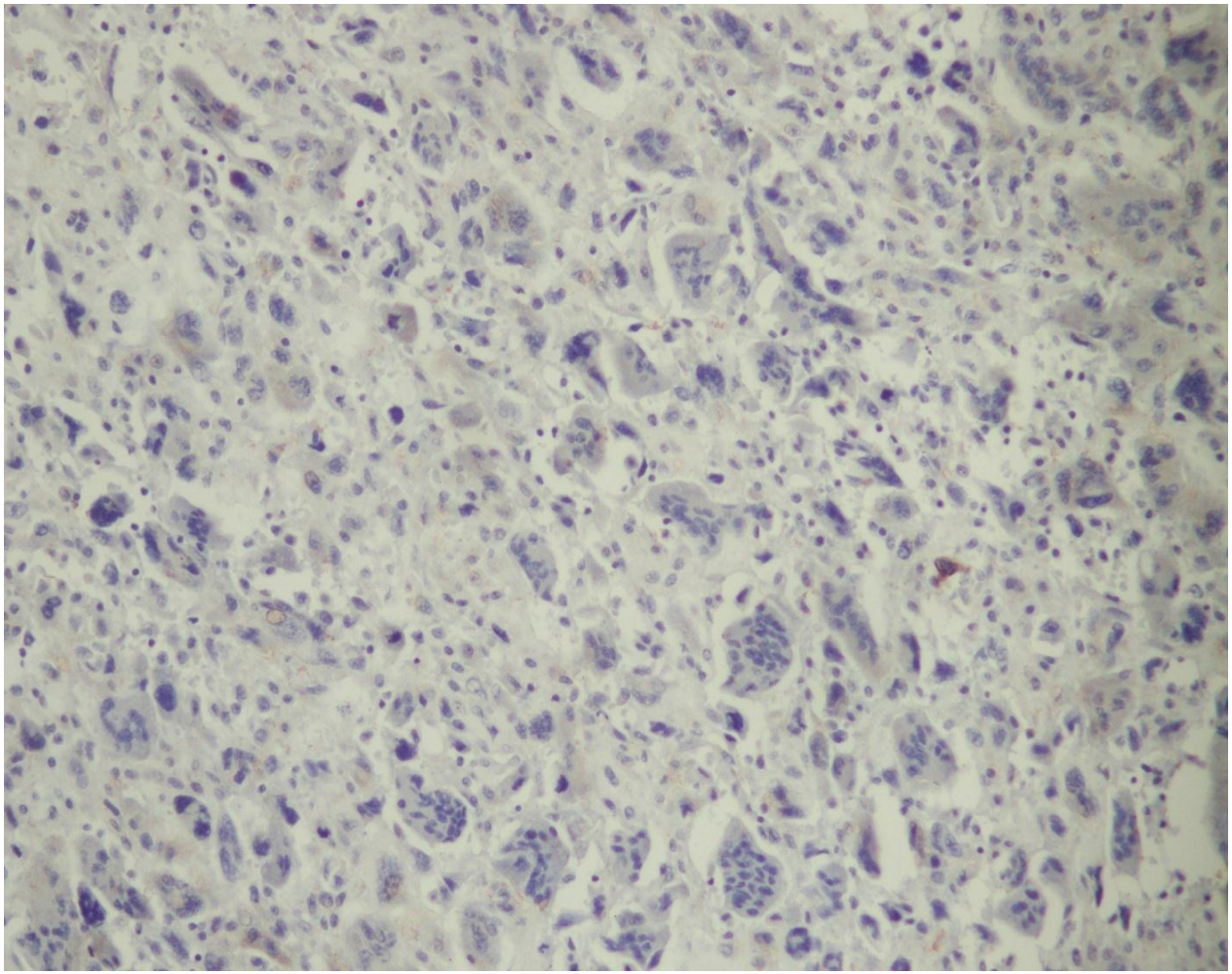
S 100



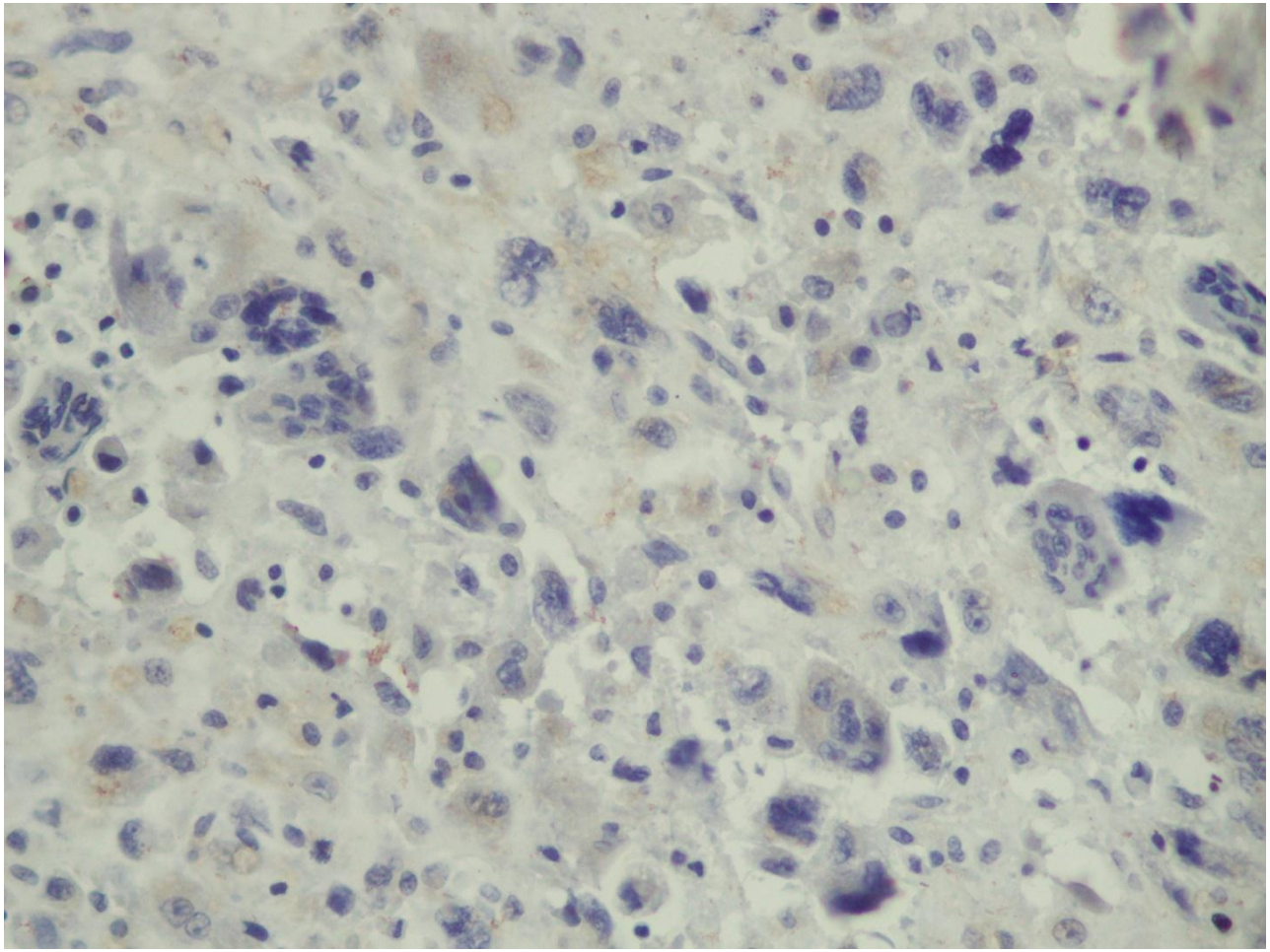
S100



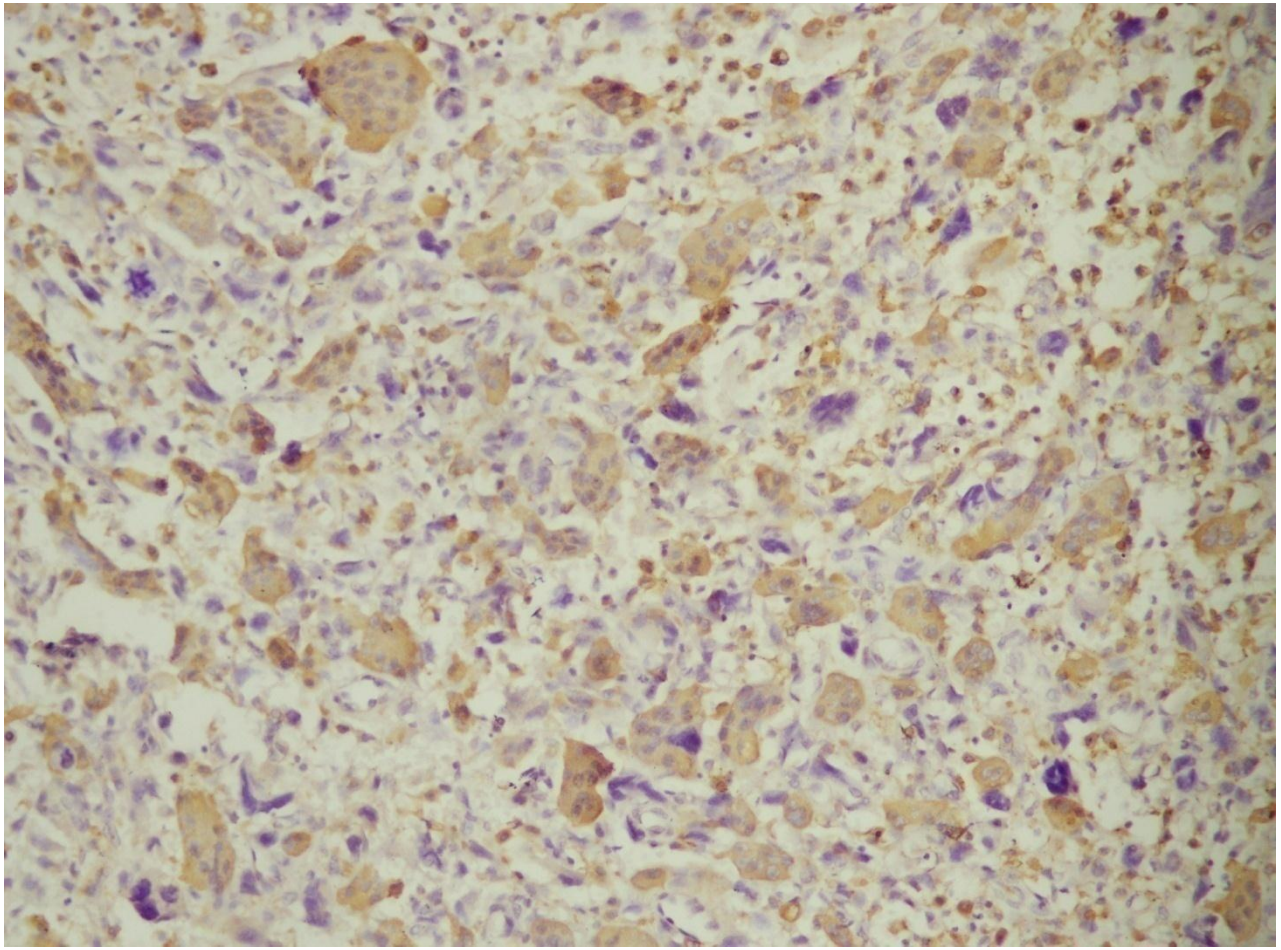
HMB 45



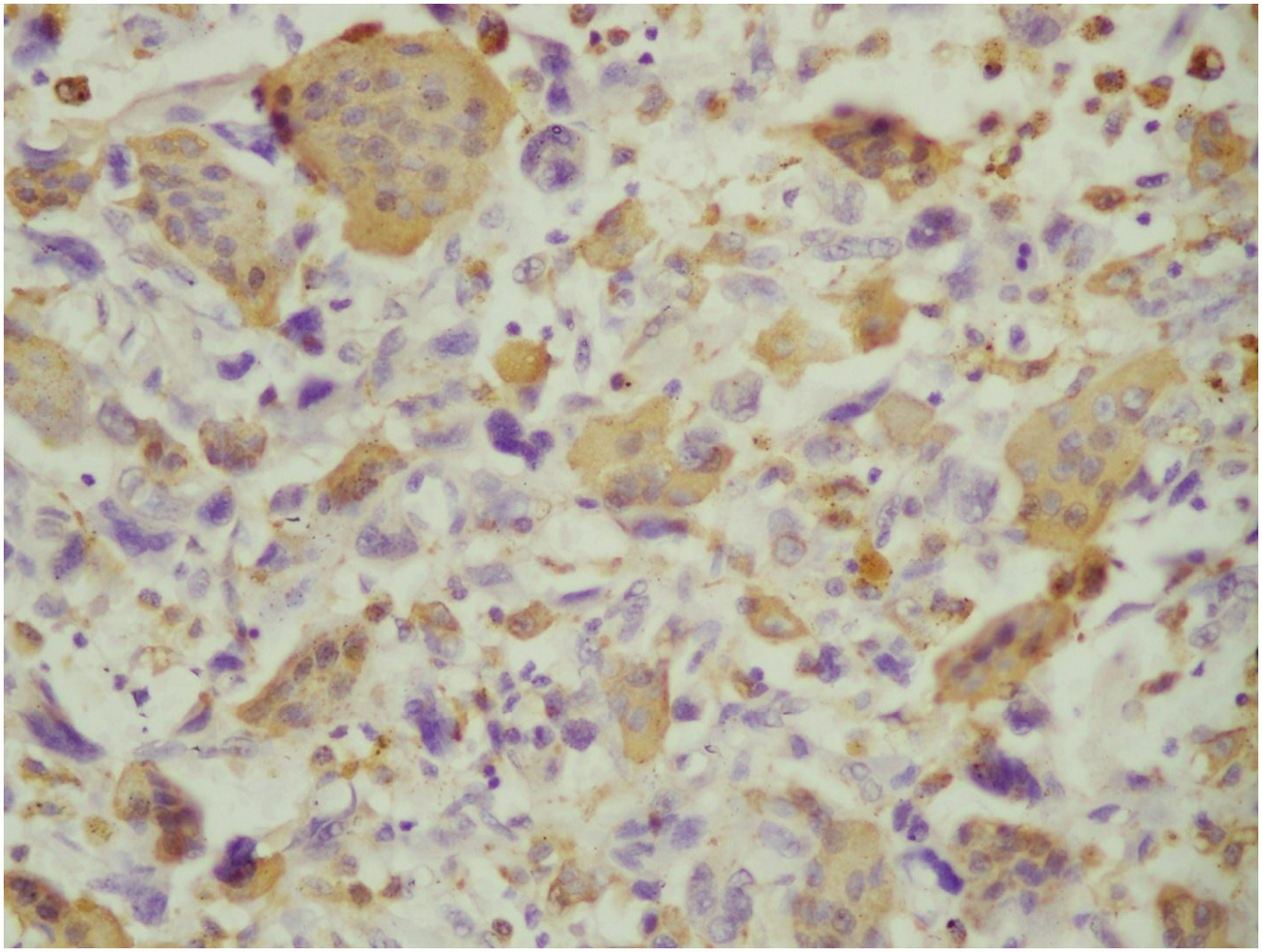
AE1/AE 3



CD68



CD 68



Immunohistochemical profile

IHC markers	
Vimentin	**Positive**
Desmin	Negative
SMA	Negative
S 100	**Weakly Positive**
HMB 45	Negative
AE 1/AE3	Negative
CD 68	**Positive in osteoclast like giant cells**

Final Diagnosis

- **Undifferentiated pleomorphic sarcoma (giant cell rich type) (Right ankle region)**

UPS (Giant cell type)

- An undifferentiated pleomorphic sarcoma with prominent osteoclast-like giant cells.
- Previously called **Malignant Fibrous Histiocytoma (Giant cell Type)**
- High-grade soft tissue sarcomas that fail to show any specific line of differentiation using currently available ancillary techniques.
- **Diagnosis of exclusion-**

Epidemiology

- 3-15% of malignant fibrous histiocytomas
- Number of cases is declining as specific sarcoma types are identified.
- Age - > 40 years of age
- Average age – 56 years
- M/F – 1.3:1
- Rare in children

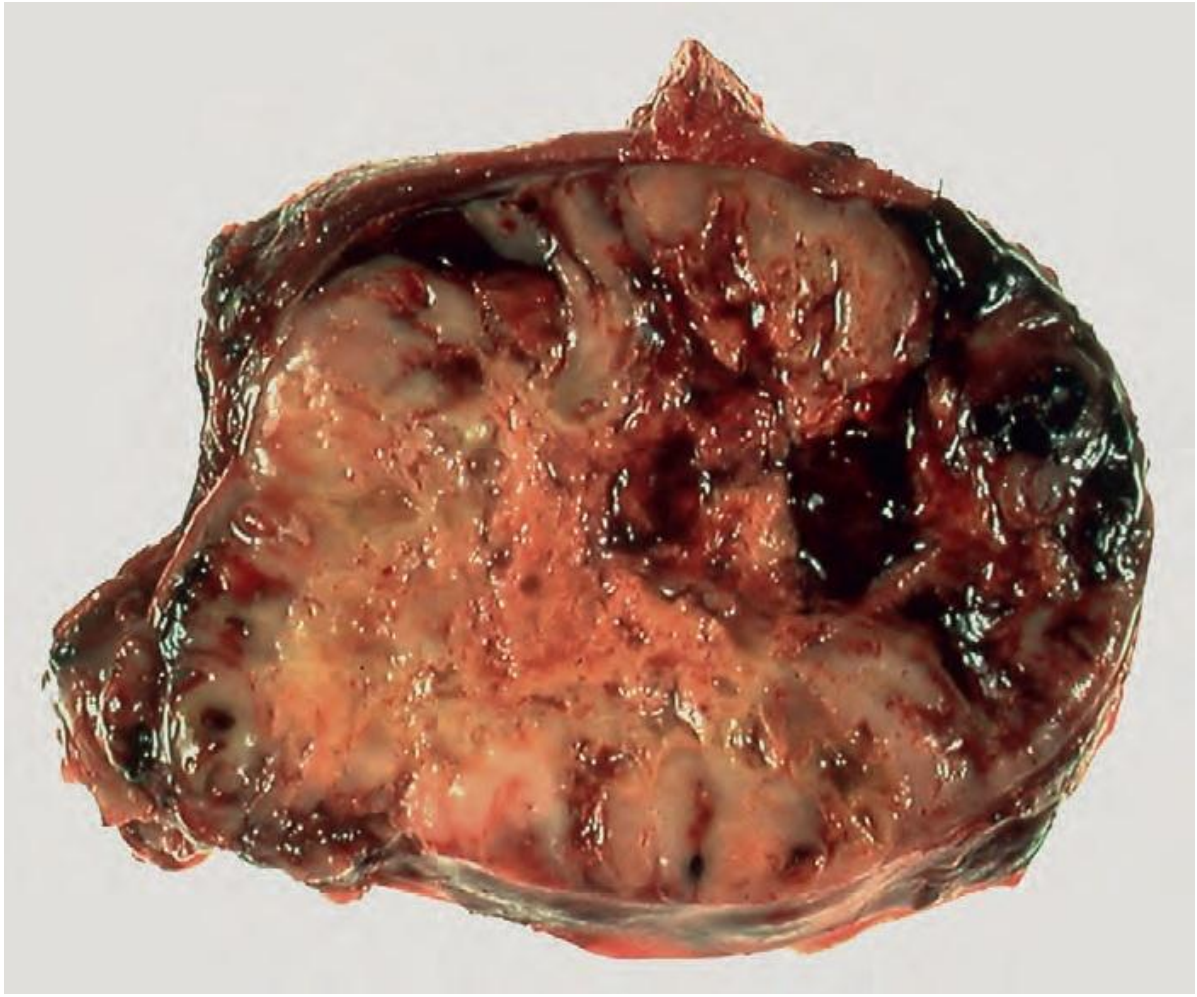
Clinical feature

- In deep soft tissue of the limbs
- Leg (thigh)
- Trunk
- Subcutaneous tissues (small portion)
- Large, often necrotic tumors
- Rapidly growing tumors may be painful

Morphology

Gross –

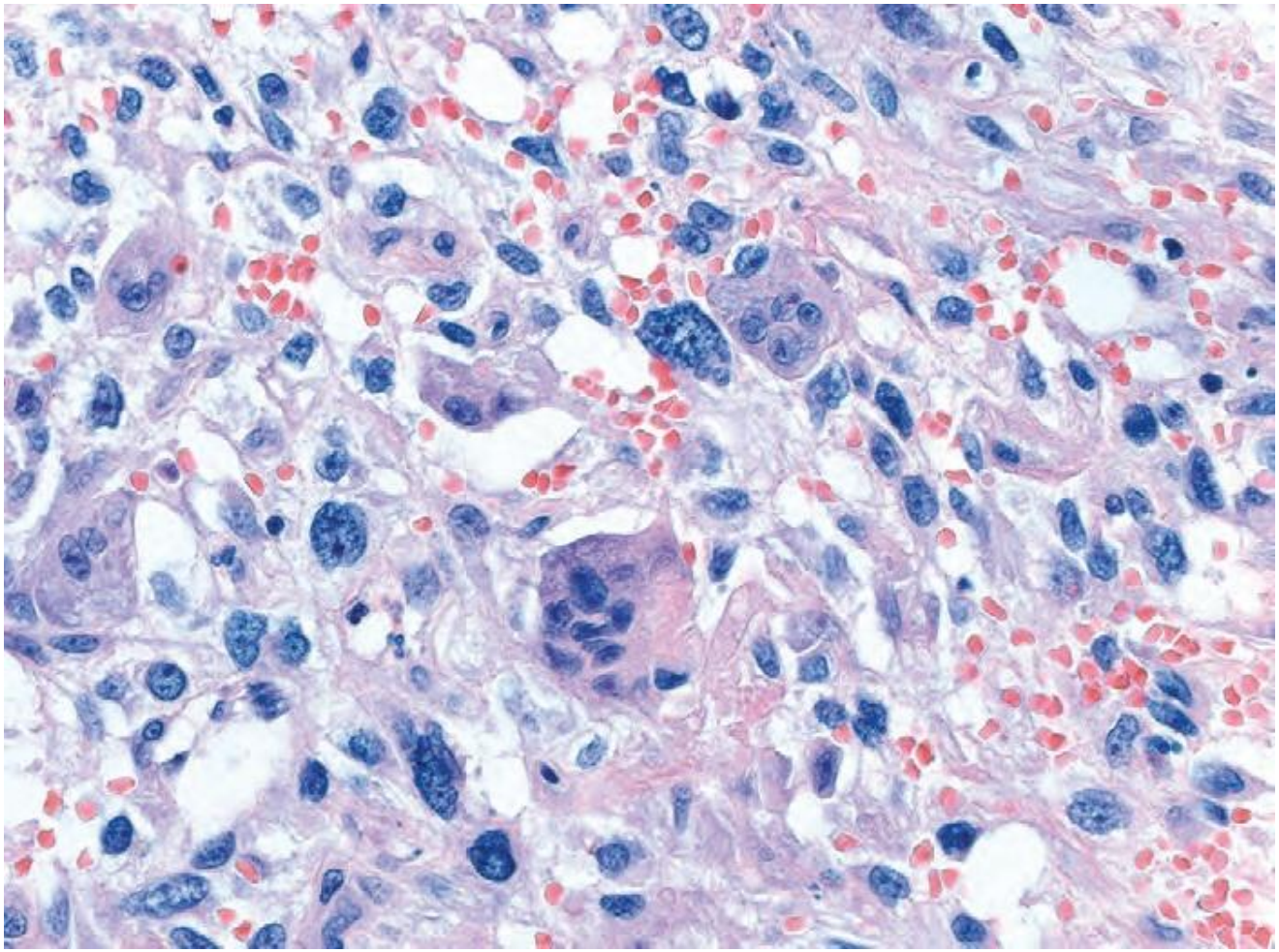
- tumors vary in size from 5 to 15 cm
- usually firm with a grayish color
- usually well circumscribed with infiltrative margins
- visible areas of haemorrhage & necrosis



Histology

- Tumors are composed of a haphazard, storiform, or fascicular arrangement of highly pleomorphic and spindle-shaped cells with a variable amount of eosinophilic or amphophilic cytoplasm and numerous typical and atypical mitoses.
- **Hall mark of UPS-GC type is that it contains two types of giant cells- namely, the osteoclast-like type and the more characteristic pleomorphic hyperchromatic bizarre cell (UPS/MFH cells)**
- **Although these pleomorphic cells may be outnumbered, they are always present.**

- May have angiolymphatic invasion
- Rarely small foci of neoplastic bone or cartilage (some designate these tumors as osteosarcoma or chondrosarcoma)



Positive IHC stains

- Vimentin,
- CD 68,
- S100 (variable),
- SMA (Variable)

Molecular genetics

- Nonspecific complex karyotypes
- Extensive intratumoral heterogeneity
- Aneuploidy (haploid, triploid, tetraploid)
- Genomic imbalances, mutations in TP53, RB1, CDKN2A

Treatment

- Wide surgical excision with free margins and adjuvant radiotherapy is the primary therapeutic modality of choice

Prognosis

- Worst Prognosis like UPS/SP
- Highly aggressive and rapidly spreading
- Recurrence – 53%
- metastases- 53-60%,
- 5 yr survival rate- 31-33%. (UPS/SP- 30-42%)

Take Home Message

- Diagnosis of soft tissue tumor is quite challenging for pathologists.

- Need to do ancilliary diagnostic technique like **immunohistochemistry** (**panels of IHC markers**) to get a correct diagnosis.

References

- Hornick JL (2013) Pleomorphic sarcoma. *In Practical Soft Tissue Pathology. A diagnostic approach* .Elsevier Saunders. 198-220.
- Mills SE et al (2010) Disorders of soft tissues. *In Sternberg's Diagnostic Surgical Pathology*. 5th edition. Lippincott Williams & Wikins. 124-197.

THANK YOU