CASE 2

SARCOMA MASQUERADING AS A SOFT TISSUE HEMATOMA :A MYSTERY OF FOOT SWELLING

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CLINICAL FINDINGS:

- 16 years, female
- Presented to a private hospital with c/o - pain and swelling in plantar aspect of right foot for 5 months.
- No history of trauma, significant past and family history.

RADIOLOGICAL FINDINGS:

- **ULTRASOUND LOCAL (Right foot swelling):**
- Oval hypoechoic area with minimal compression
- Mild fluid collection seen at IP joint
- No obvious deep extension
- Adjacent muscles appear normal
- Colour flow was seen on the doppler study

- **IMPRESSION: Possibility of muscle hematoma**


Surgical Excision
Specimen: Excised swelling at the fifth tarsometatarsal joint
Histopathological examination
(Outside lab)

HISTOPATHOLOGY REPORT(OUTSIDE LAB)

IMPRESSION:

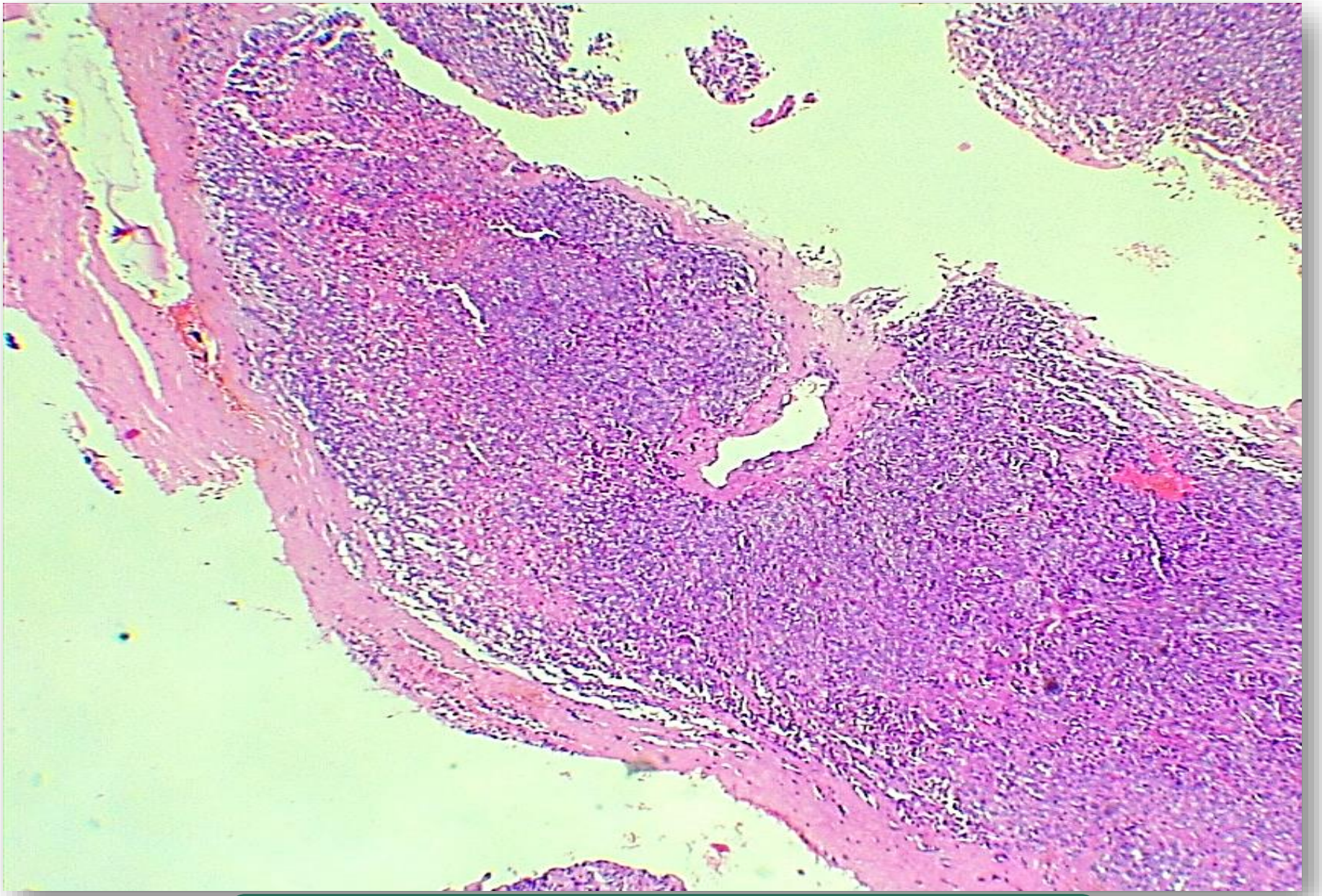
- Features suggestive of **poorly differentiated neoplasm with increased vascularity**

ADVISE:

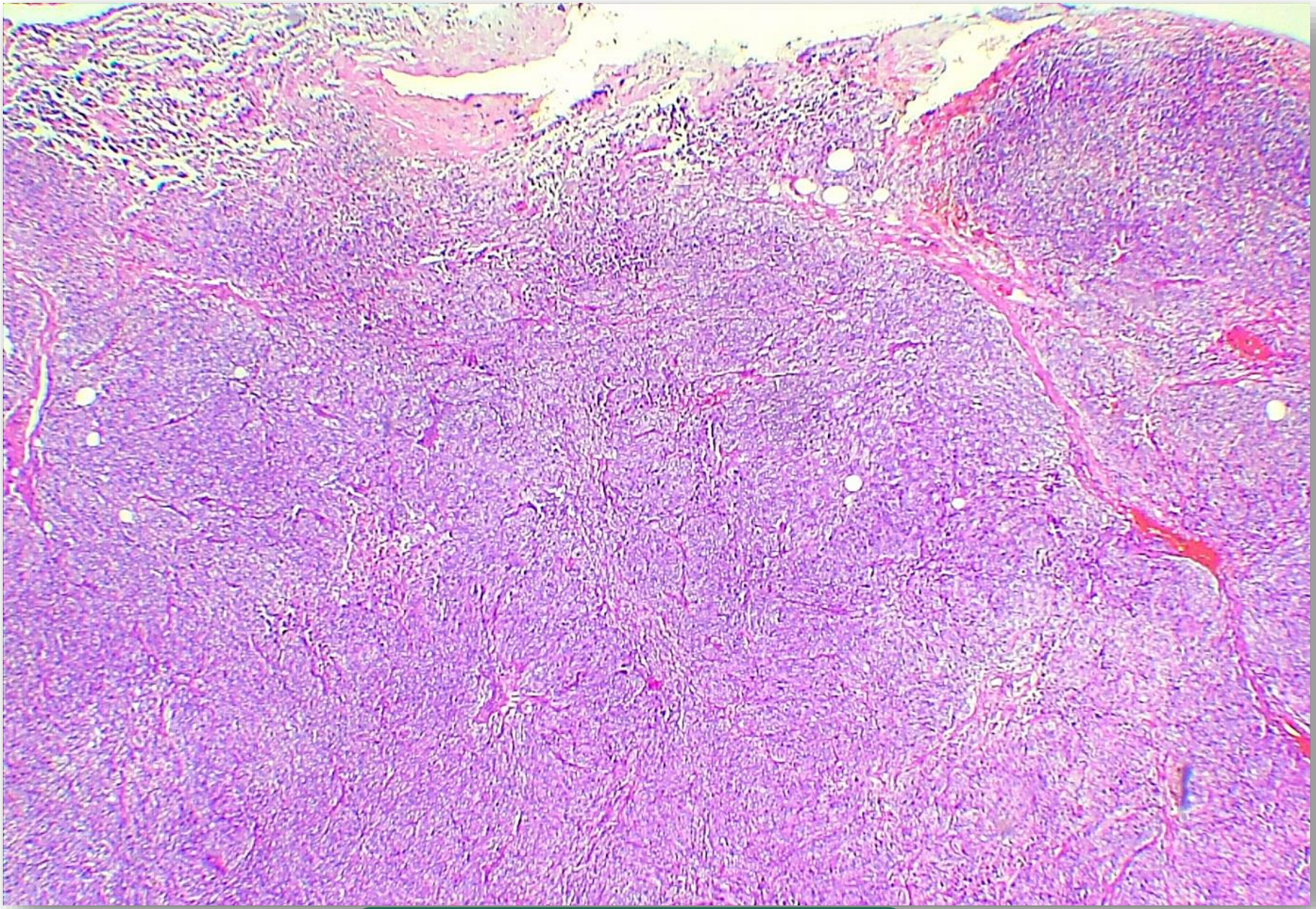
- **IHC for complete evaluation**

After surgery the patient was referred to our surgical-oncology department.

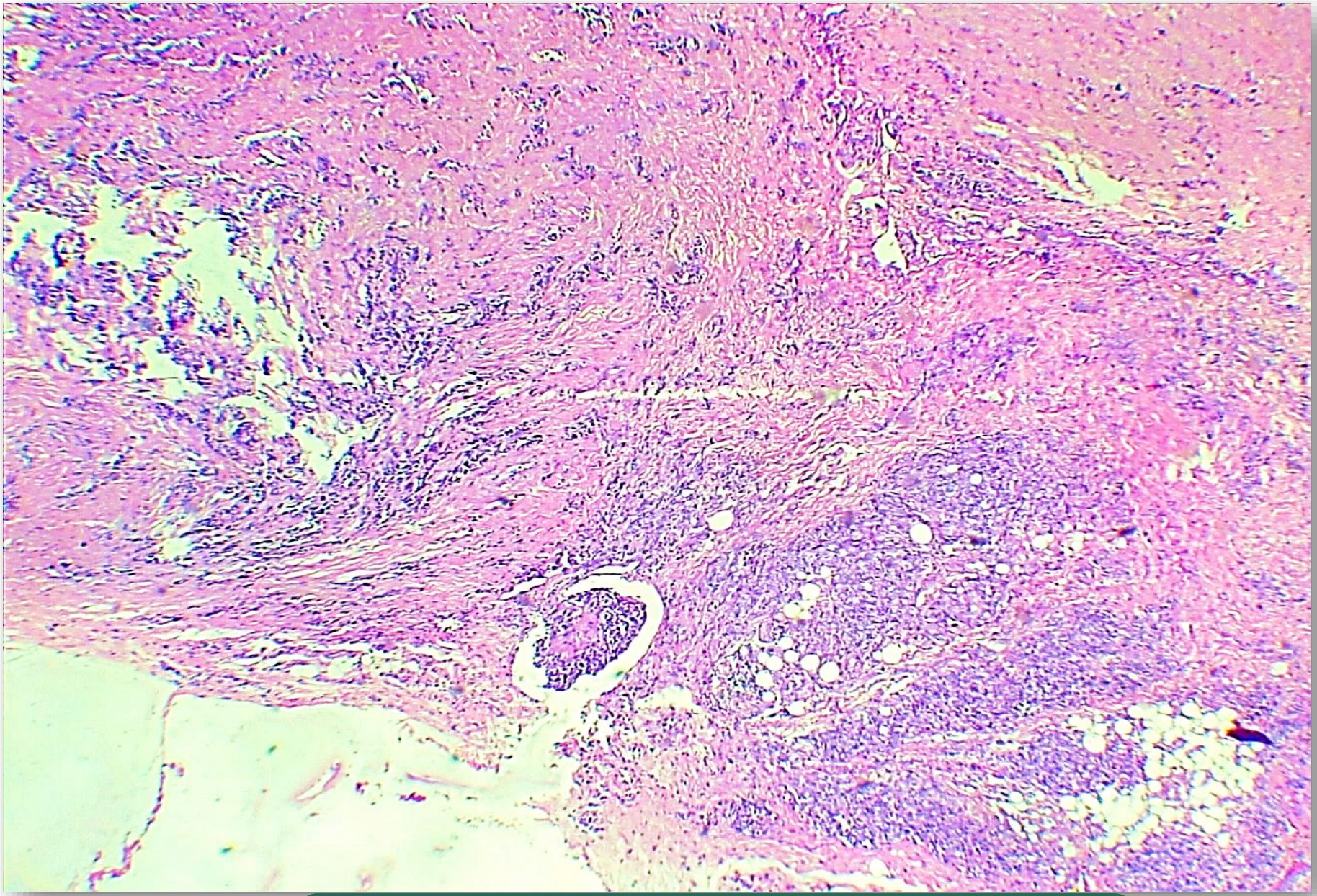
2 Paraffin blocks - REVIEW AND IHC



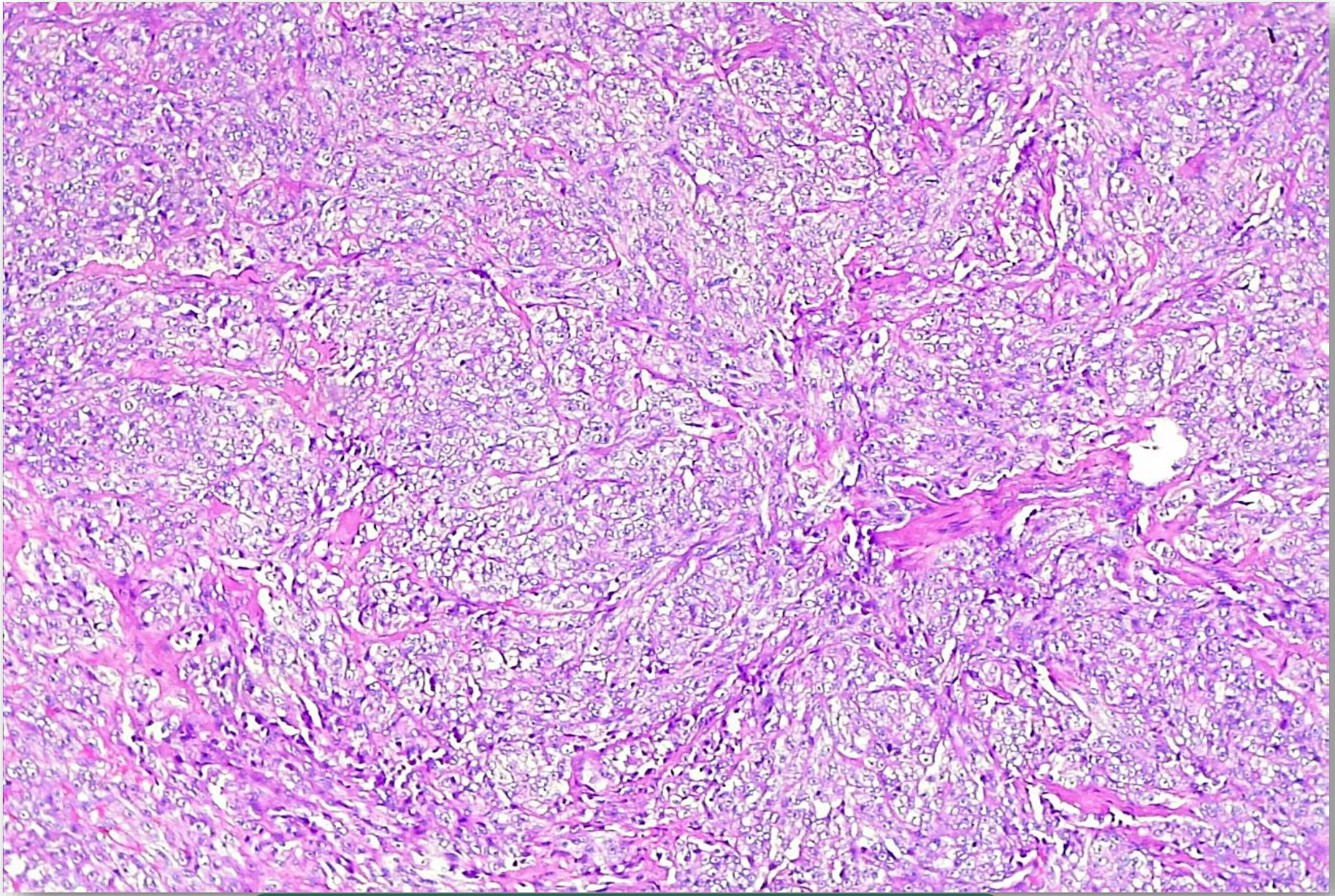
Fairly circumscribed and focally capsulated tumour



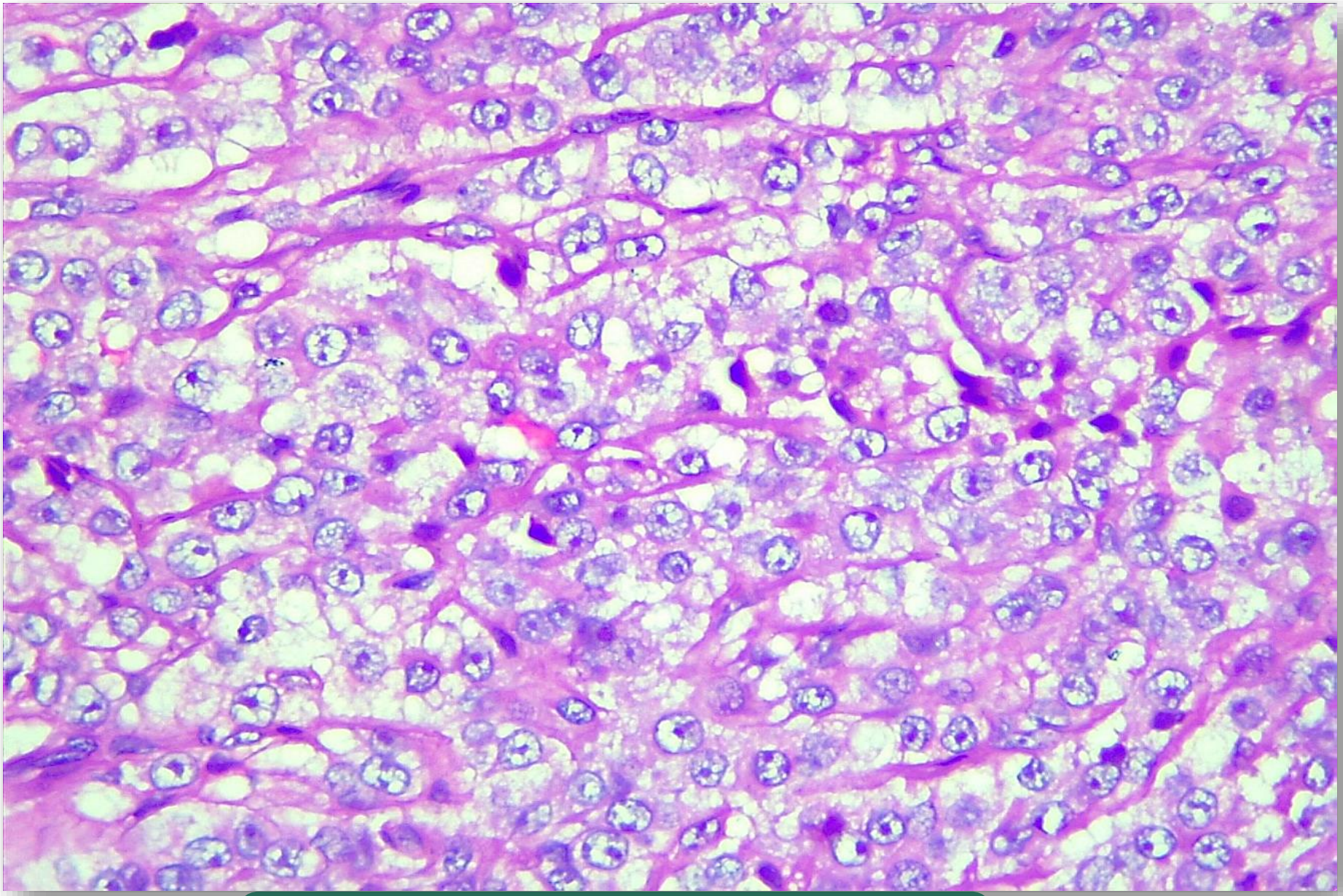
Sheets of tumour cells



Tumour is seen infiltrating into adjacent fat in few areas, stromal hyalinization



Alveolar/Nested pattern separated by thin fibrovascular septa

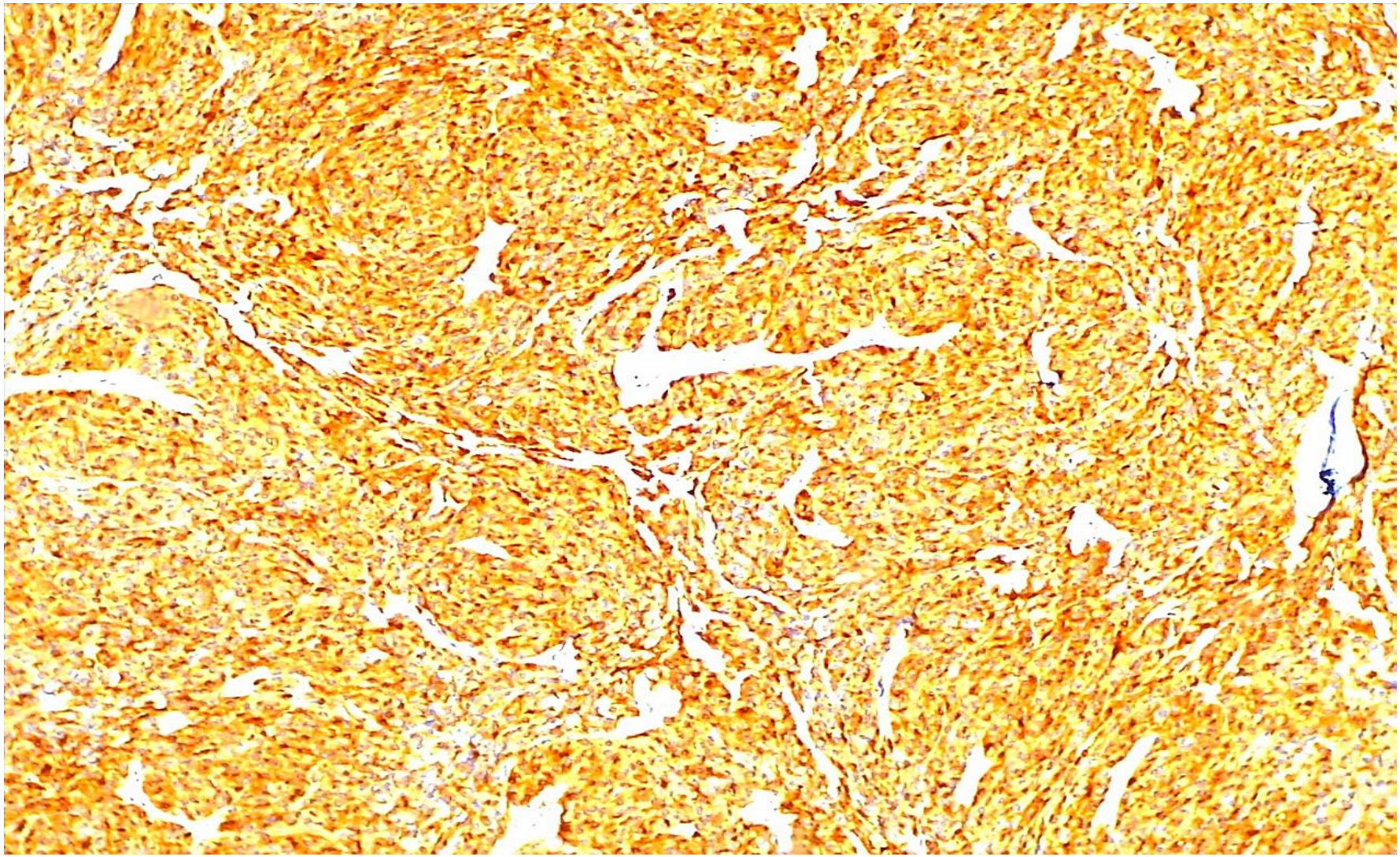


Clear cytoplasm, vesicular nuclei and prominent nucleoli

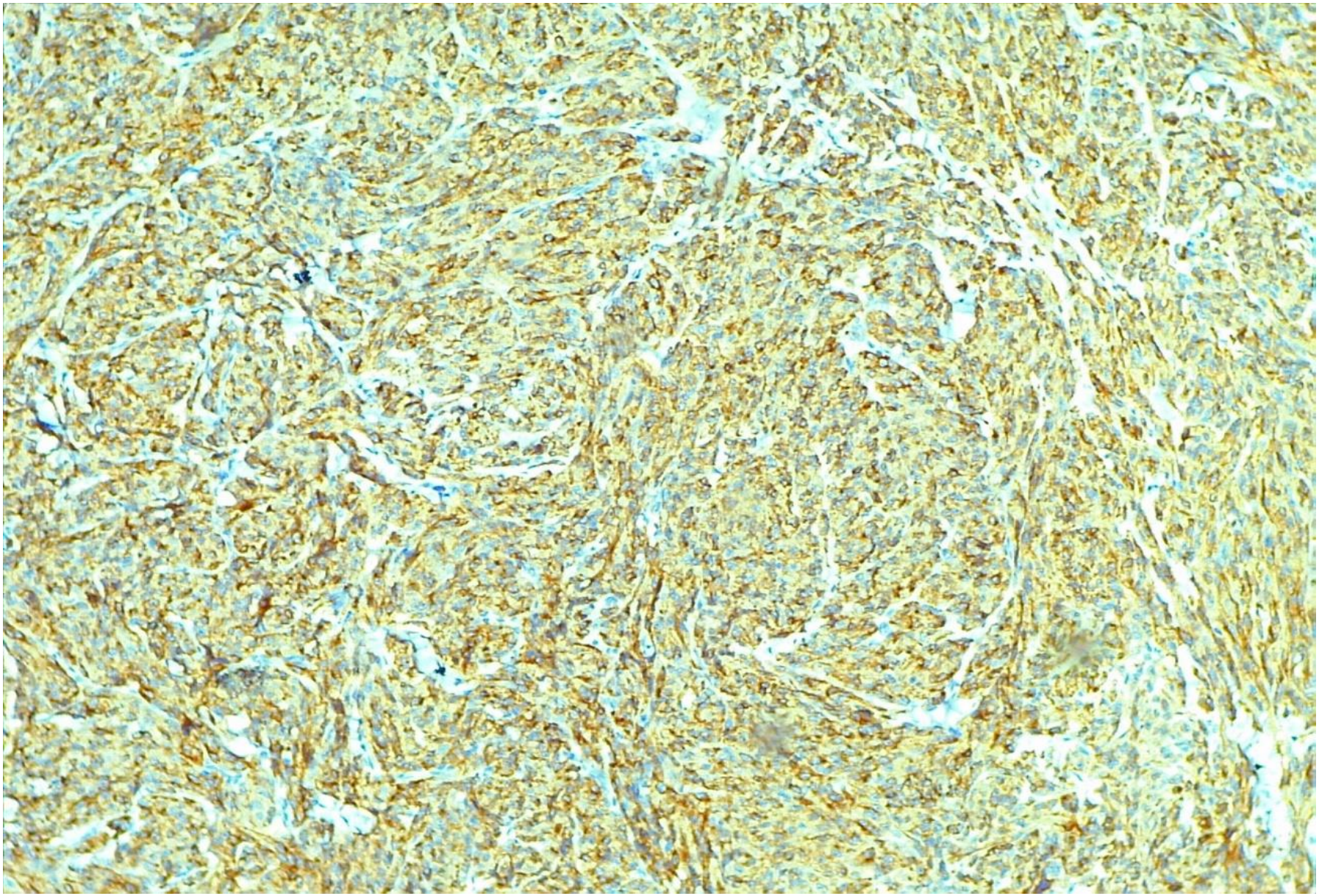
DIFFERENTIAL DIAGNOSIS

- Clear cell sarcoma
- Alveolar soft part sarcoma
- Alveolar Rhabdomyosarcoma
- Ewing’s sarcoma
- Neuroendocrine Tumours

IHC FINDINGS:



S-100-Diffusely positive



HMB-45 –Diffusely positive

NEGATIVE IHC MARKERS:

CD34,EMA- Alveolar soft part sarcoma

Desmin- Alveolar Rhabdomyosarcoma

CD99- Ewing's sarcoma

Synaptophysin and Chromogranin- Neuroendocrine Tumours

FINAL DIAGNOSIS:

CLEAR CELL SARCOMA OF FOOT

Margins could not be commented upon as blocks were received for second opinion.

Further workup was advised.

FNLCC GRADING SYSTEM FOR SOFT TISSUE SARCOMA

Histological grading according to FNCLCC	
Tumour differentiation	
Score 1	Closely resembling normal tissue
Score 2	Histological typing is certain
Score 3	Embryonal or undifferentiated sarcomas
Mitotic count (per 1.7 mm^2)	
Score 1	0-9 mitoses per 1.7 mm^2
Score 2	10-19 mitoses per 1.7 mm^2
Score 3	>19 mitoses per 1.7 mm^2
Tumour necrosis	
Score 0	No necrosis
Score 1	<50% tumour necrosis
Score 2	≥50% tumour necrosis
Histological grade	Grade 1: total score 2, 3 Grade 2: total score 4, 5 Grade 3: total score 6, 7, 8

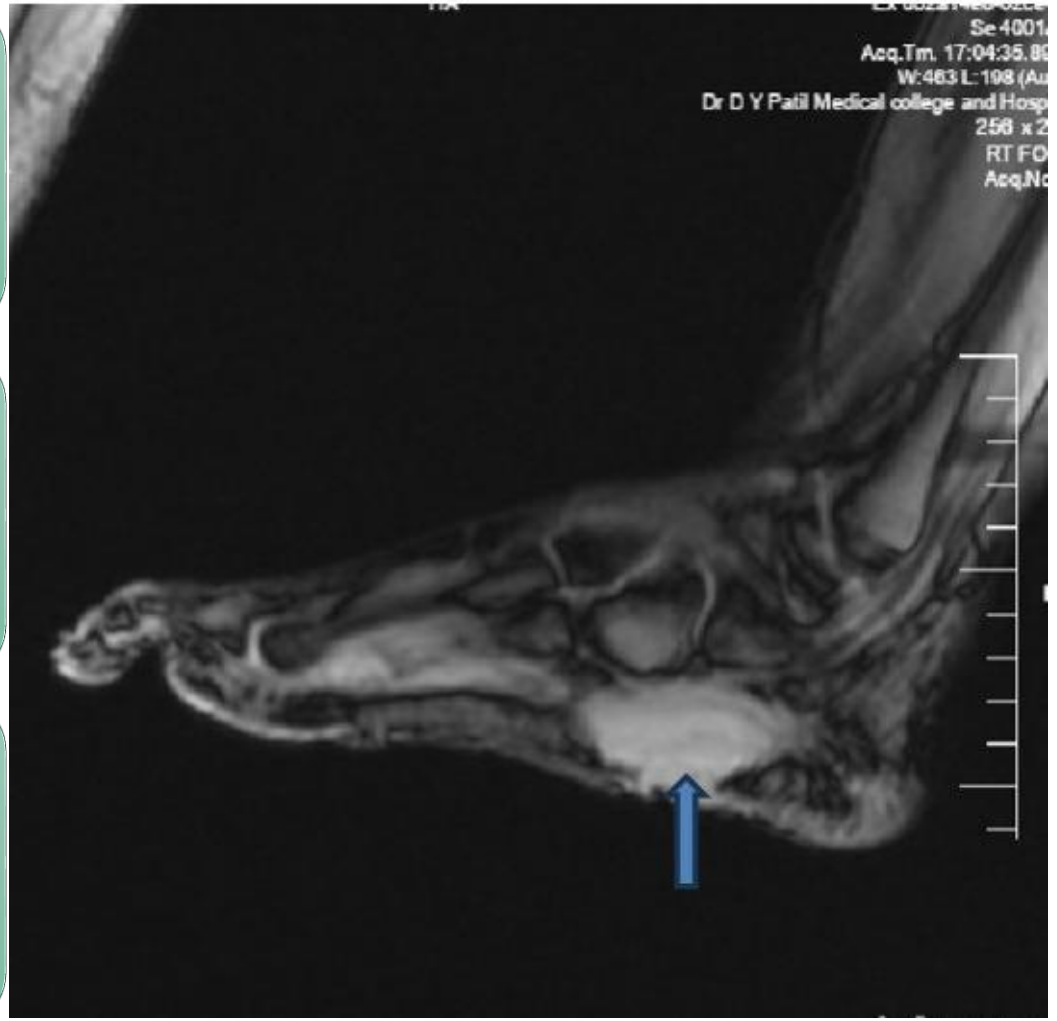
However clear cell sarcoma is invariably considered as high grade sarcoma

FOLLOW UP- MRI OF RIGHT FOOT (PLAIN)

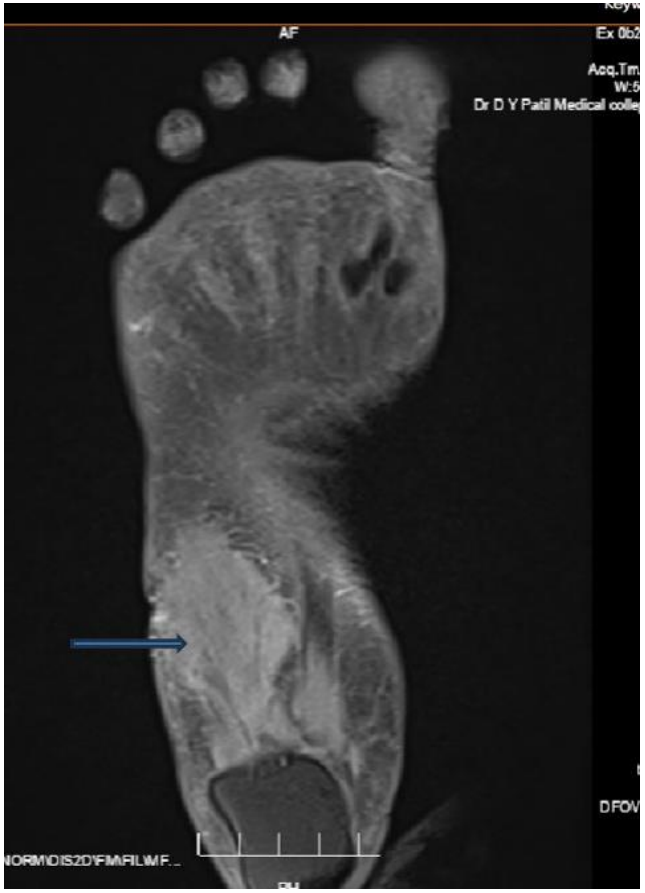
A fairly well defined lesion measuring 17x14x23 mm is noted in the planter aspect.

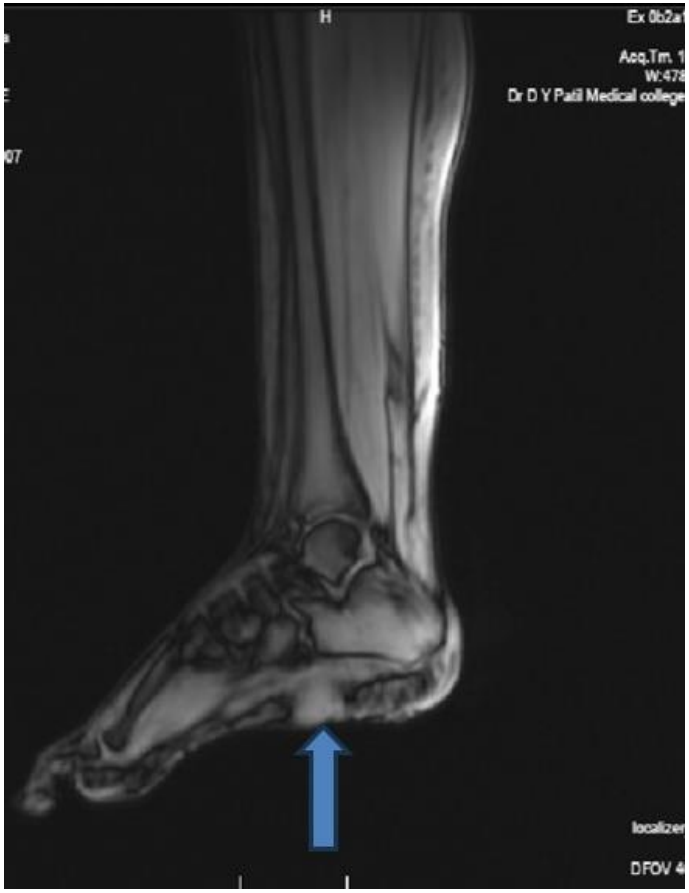
The lesion is in close proximity to cuboid and base of 5 th metatarsal

Bony outlines are maintained.



MRI Impression: A fairly well defined, altered signal intensity lesion in the planter aspect of right foot –likely residual tumour





DISCUSSION:

Clear cell sarcoma (CCS) of soft tissue :

- Malignant mesenchymal neoplasm, typically involving deep soft tissue
- Young adults, third and fourth decades, male predominance
- **Location:** deep-seated sites in the extremity, ankle, or foot, It has also been reported in the head/neck, trunk, and viscera including the lung and gastrointestinal tract.
- **Clinical features:** patients present with a palpable mass of months' to years' duration. Pain and tenderness
- **Macroscopic appearance:** 2–5 cm, but tumors, > 15 cm have been reported. Circumscribed mass with a lobulated tan to greyish-white appearance, sometimes with a coarse or gritty texture.

- **Essential and desirable diagnostic criteria(WHO)**
- *Essential:* characteristic nested or fascicular low-power architecture; plump spindle or ovoid cells with palely eosinophilic cytoplasm and prominent nucleoli; multinucleated wreath-like giant cells are common.
- **Melanocytic markers:** S100, SOX10, Melan-A and HMB45
- **Diagnostic molecular pathology:** *EWSR1* gene rearrangement or *EWSR1-ATF1* gene fusion
- **Prognosis and prediction**
- Aggressive malignancy
- **Recurrence rates:** 40%
- **Metastasis:** Pulmonary or lymph node
- **Survival rates** at 5, 10, and 20 years are approximately 60%, 35%, and 10%, respectively
- **Unfavorable prognostic factor:** Tumor size > 5 cm, necrosis, and regional lymph node involvement

NCCN GUIDELINES- Clear cell sarcoma

Tumour < 5 cm	**Wide local excision with negative margins**
Tumour upfront resectable >5cm	**Radiation before surgery**
N+ and **M+** status positive	**Chemotherapy may precede the surgery**

Except when tumours is near critical structures- Microscopic positive margin is acceptable.

TAKE HOME MESSAGE

For all clinically suspected soft tissue swellings – Complete radiological investigation must be done.

Histopathological examination is a gold standard to avoid incomplete excision and its consequences.

REFERENCES:

- WHO Classification of Tumours Editorial Board. Soft tissue and bone tumours [Internet]. Lyon (France): International Agency for Research on Cancer; 2020.(WHO classification of tumours series, 5th ed.; vol. 3). Available from: https://tumourclassification.iarc.who.int/chapters/33.

- Soft Tissue Sarcoma - Guidelines Detail (nccn.org)

THANK YOU