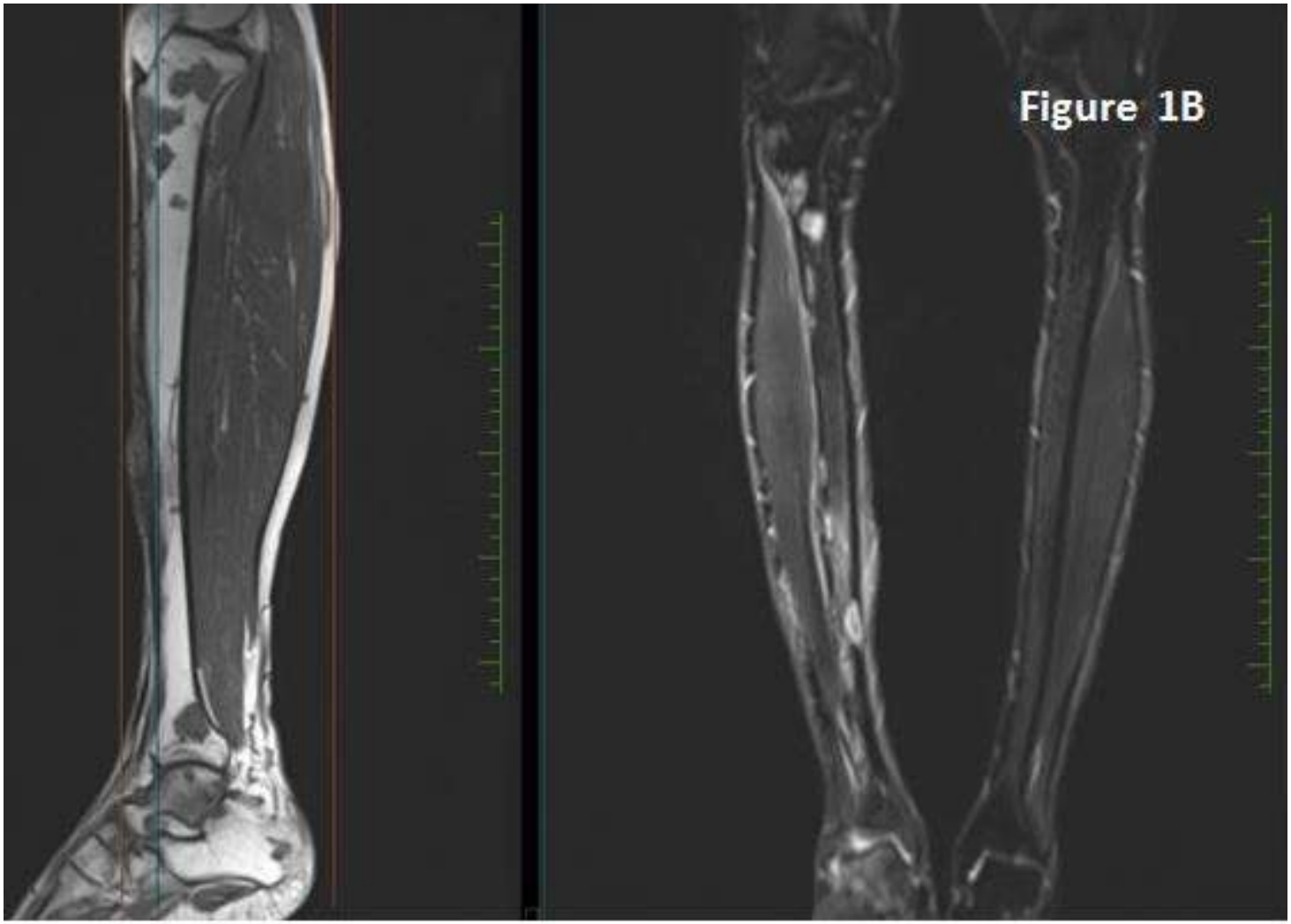
Clinical History

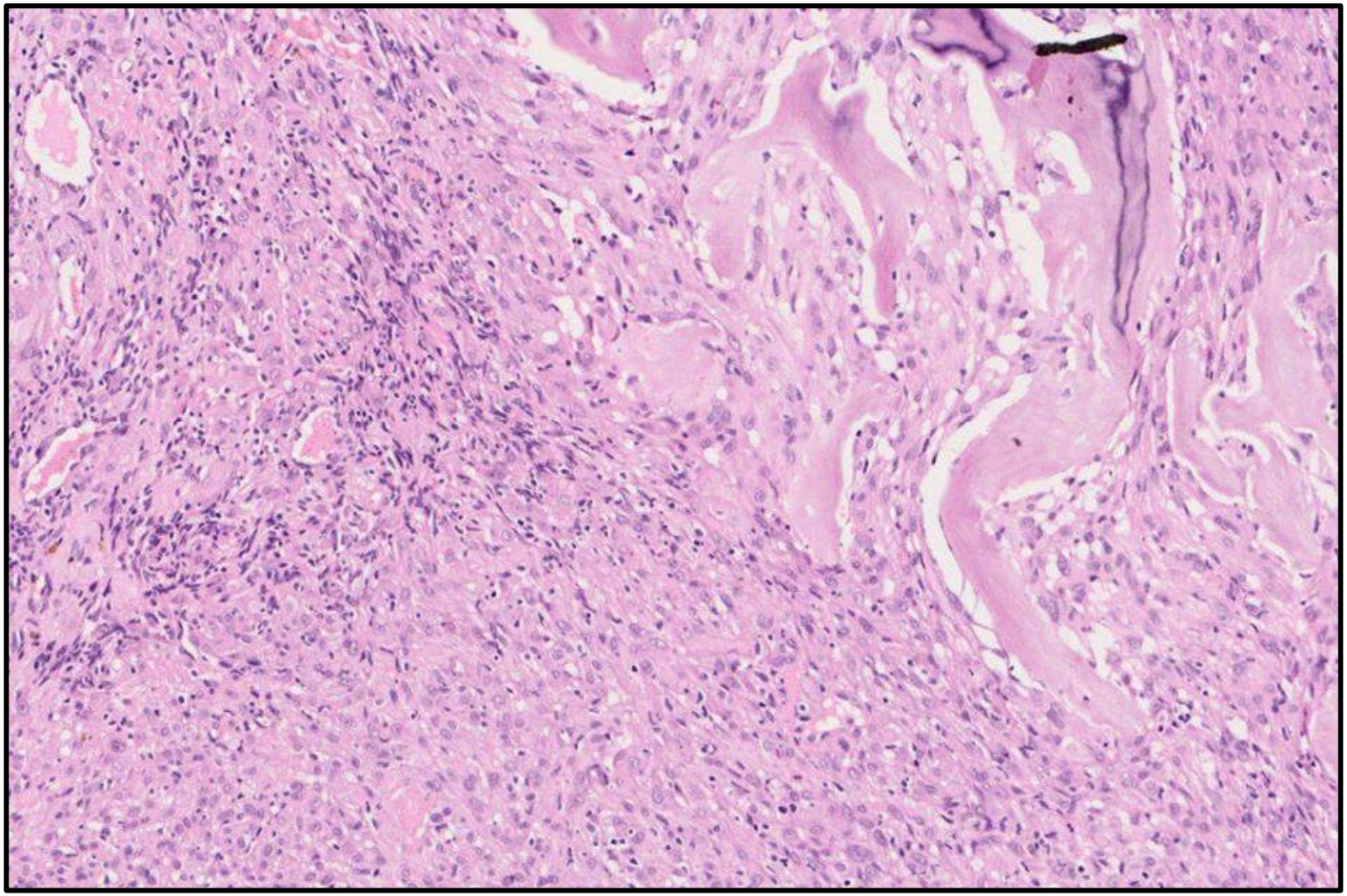
- Male 23 years
- No significant past/family history or addictions
- C/o of pain right leg for 1 year
- No difficulty in knee/ankle movements.

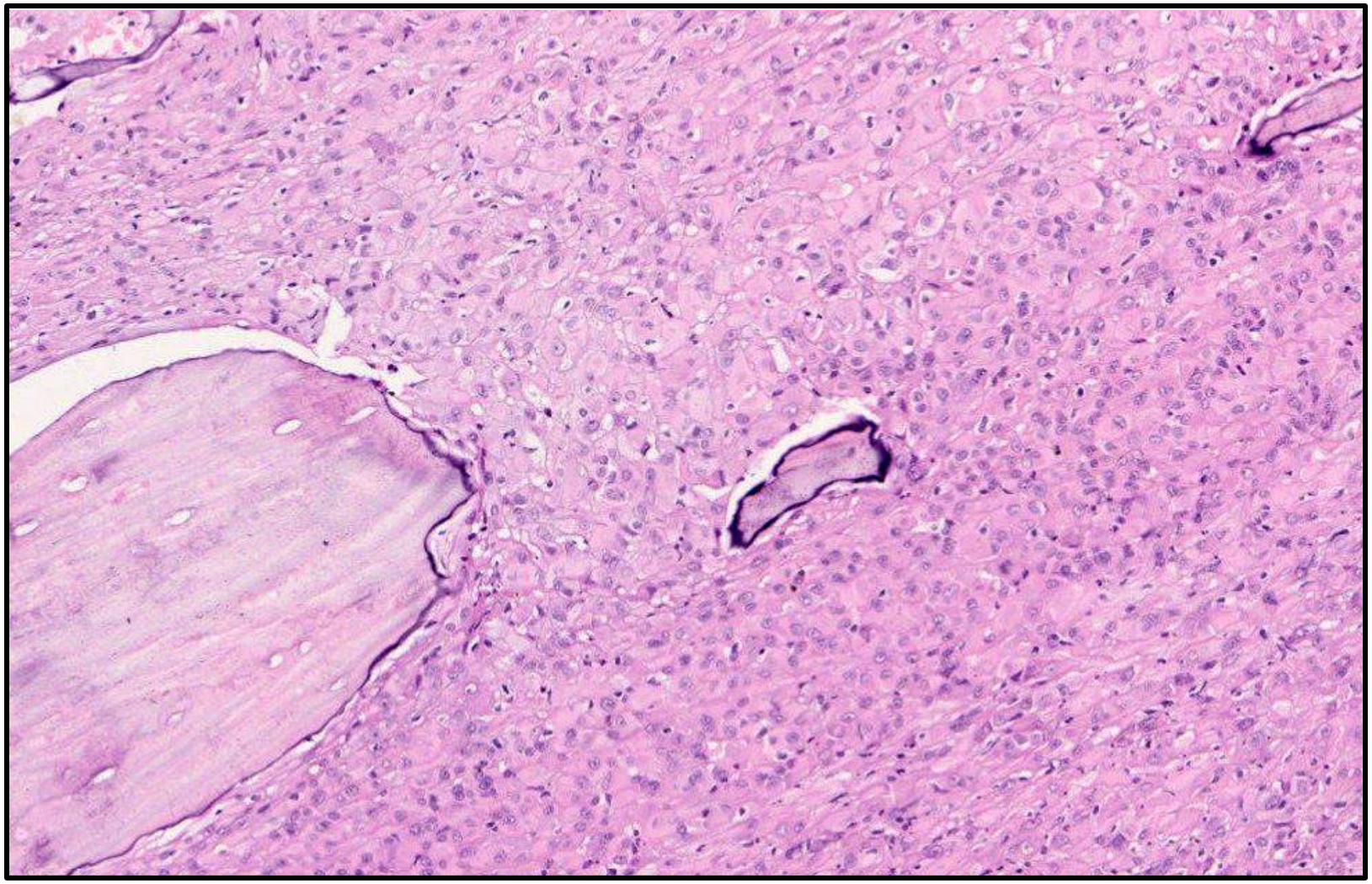
RADIOGRAPH: Multiple lytic lesions in right tibia & fibula with lesion of tibia causing cortical erosion

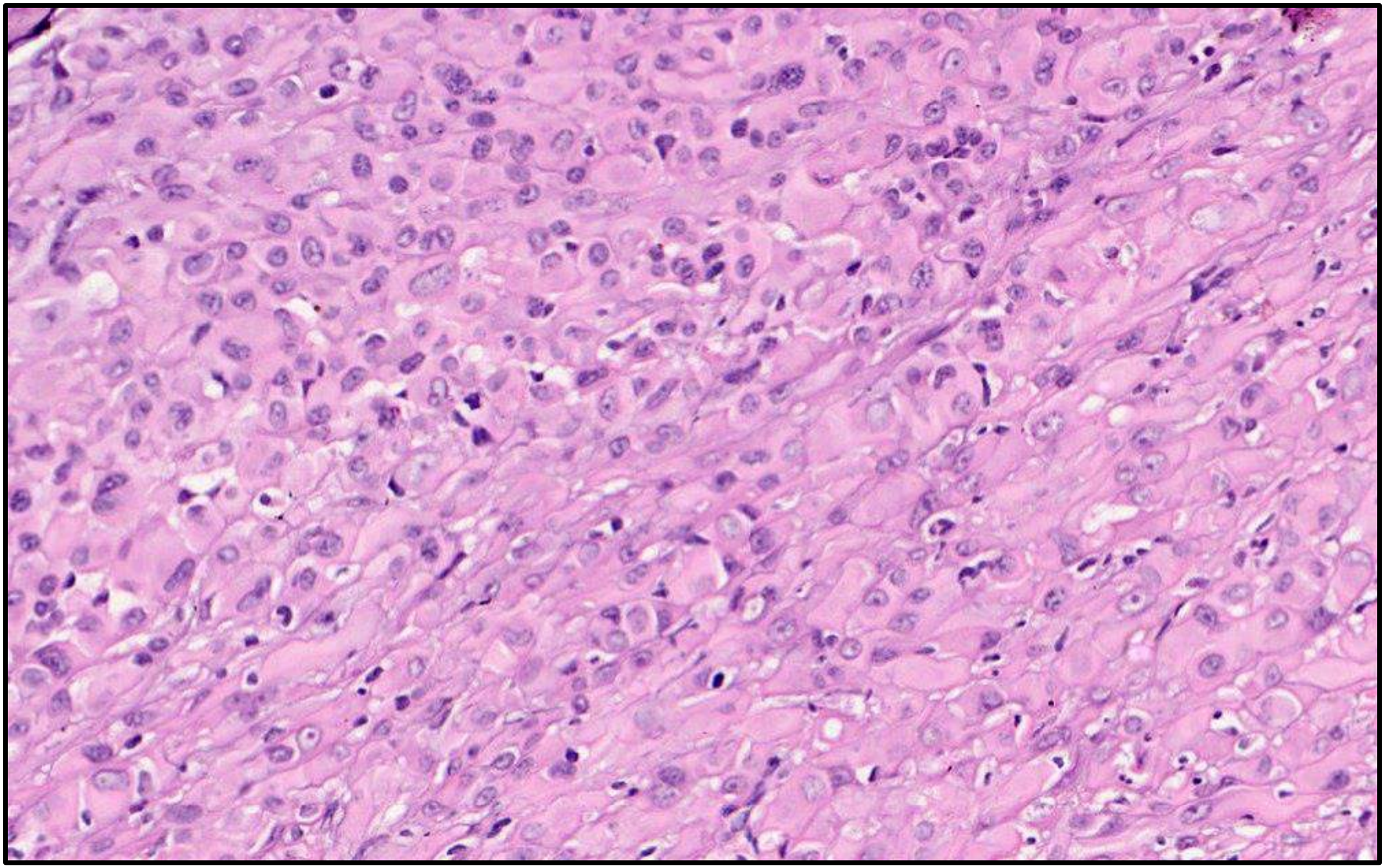
MRI revealed multiple altered signal intensity lesions in right tibia involving medulla with cortical destruction and adjacent soft tissue swelling

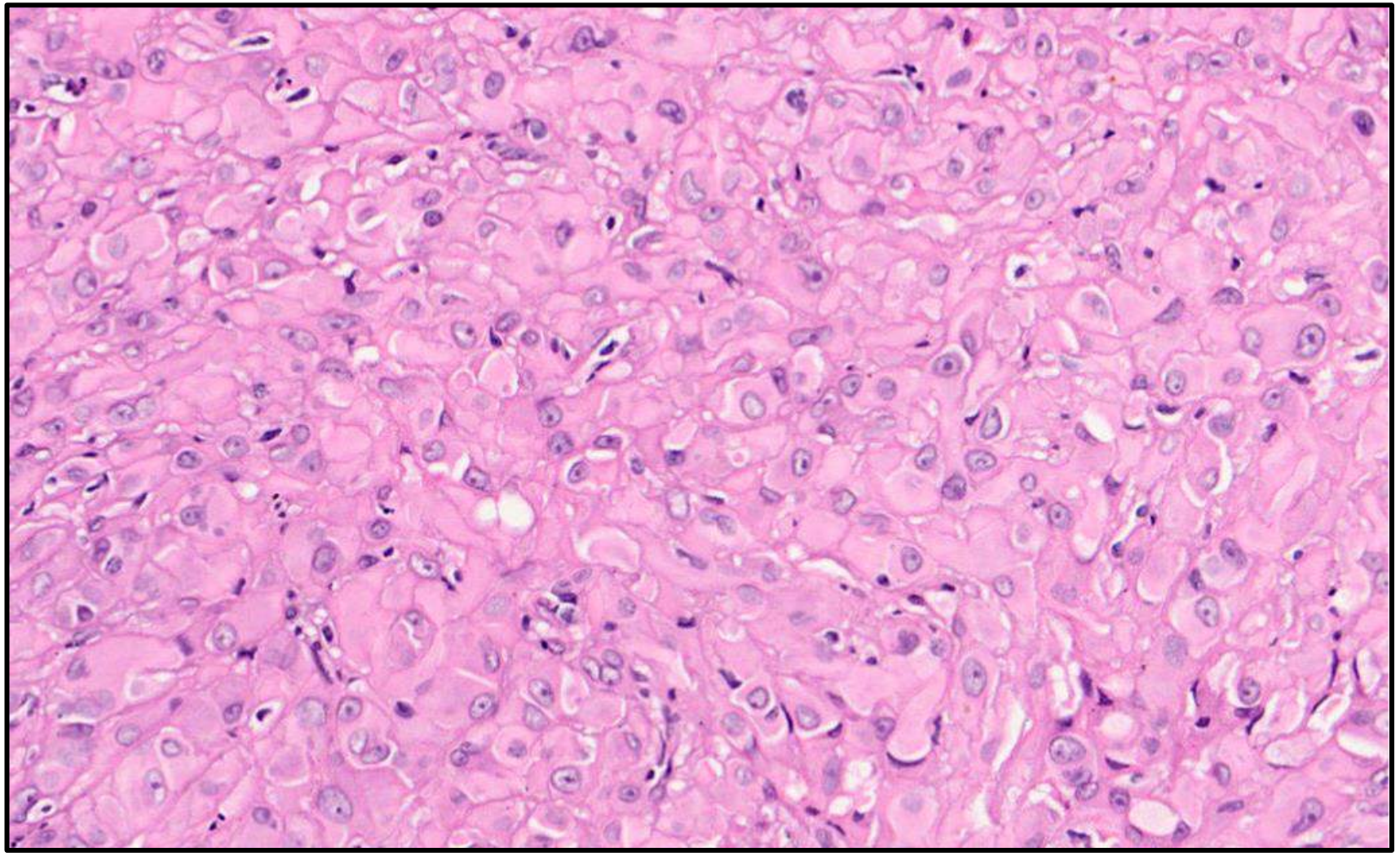


- Patient was initially evaluated outside and the H/P Diagnosis: Metastatic Carcinoma (CK +)
- Patient referred to RGCI&RC for further workup
- Blocks submitted at RGCI&RC for review

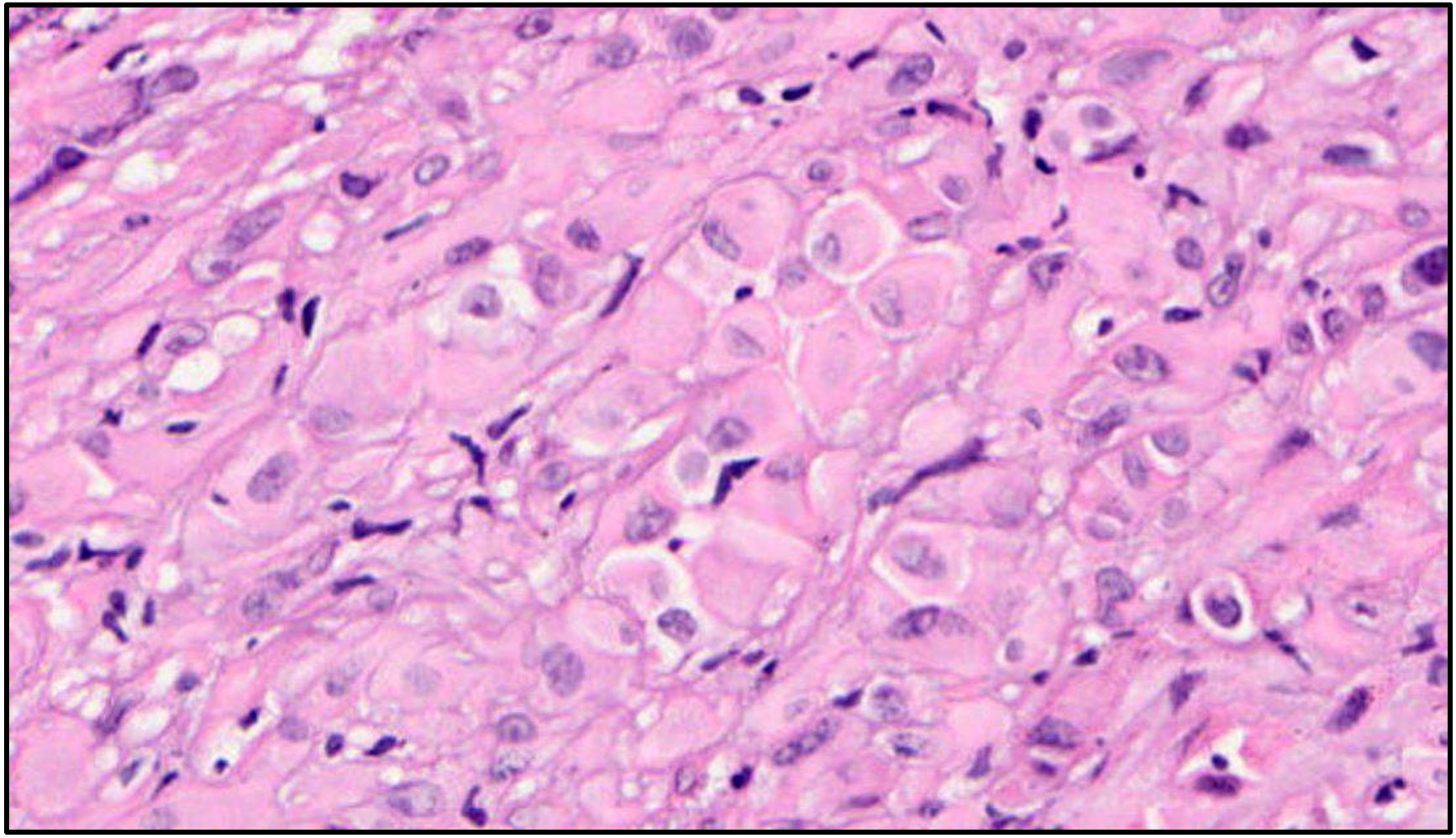








No Necrosis; Mitosis: Sparse

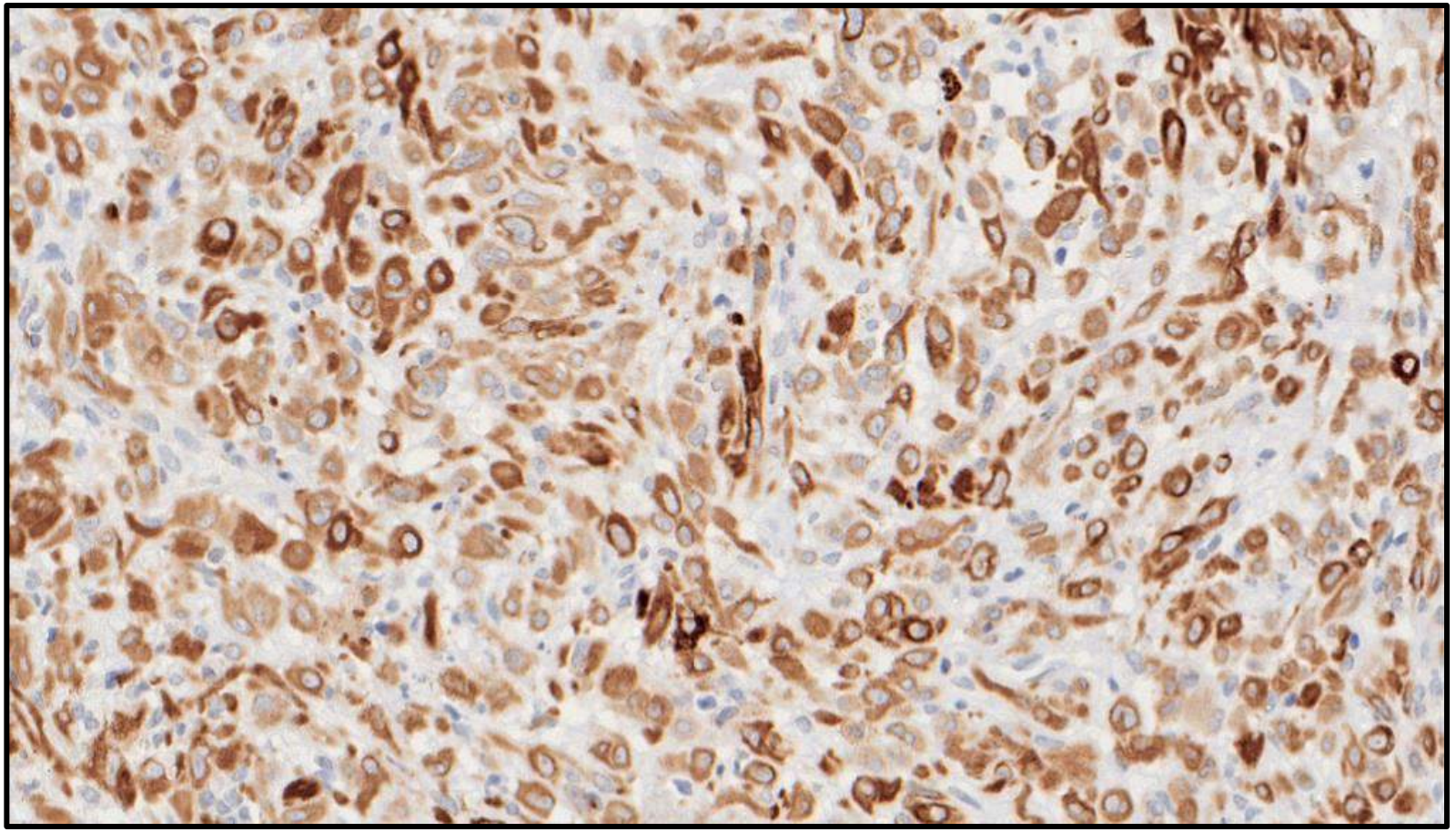


Summarizing the Histomorphological Findings

- Diffuse sheets of Plump spindle to epithelioid/polygonal tumor
- Vesicular nucleus and conspicuous nucleoli
- Dense eosinophilic glassy cytoplasm
- No Necrosis
- Mitosis: Sparse

D D: ?? In the given Clinico-radiological and pathological context

IHC: **Pan- CK**



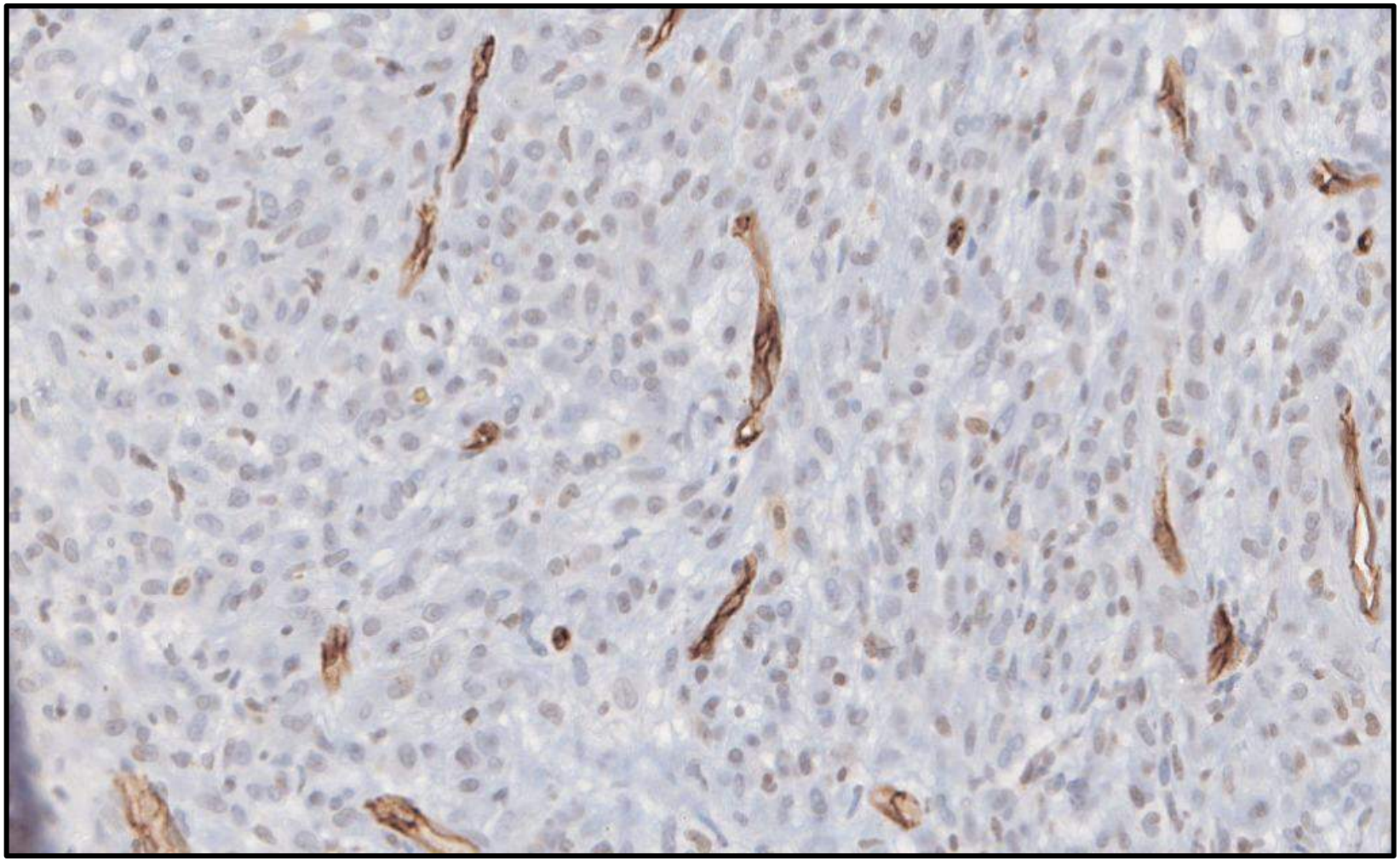
Differentials:

- Epithelioid Sarcoma
- Epithelioid Hemangioendothelioma
- Epithelioid RMS
- Metastatic Carcinoma
- Pseudomyogenic Hemangioendothelioma

Subsequent IHC findings

- CK 7 : Positive
- CK 20: Negative
- CD34: Negative
- S100 : Negative
- Desmin, MyoD1, Myogenin: Negative
- INI- 1: Retained

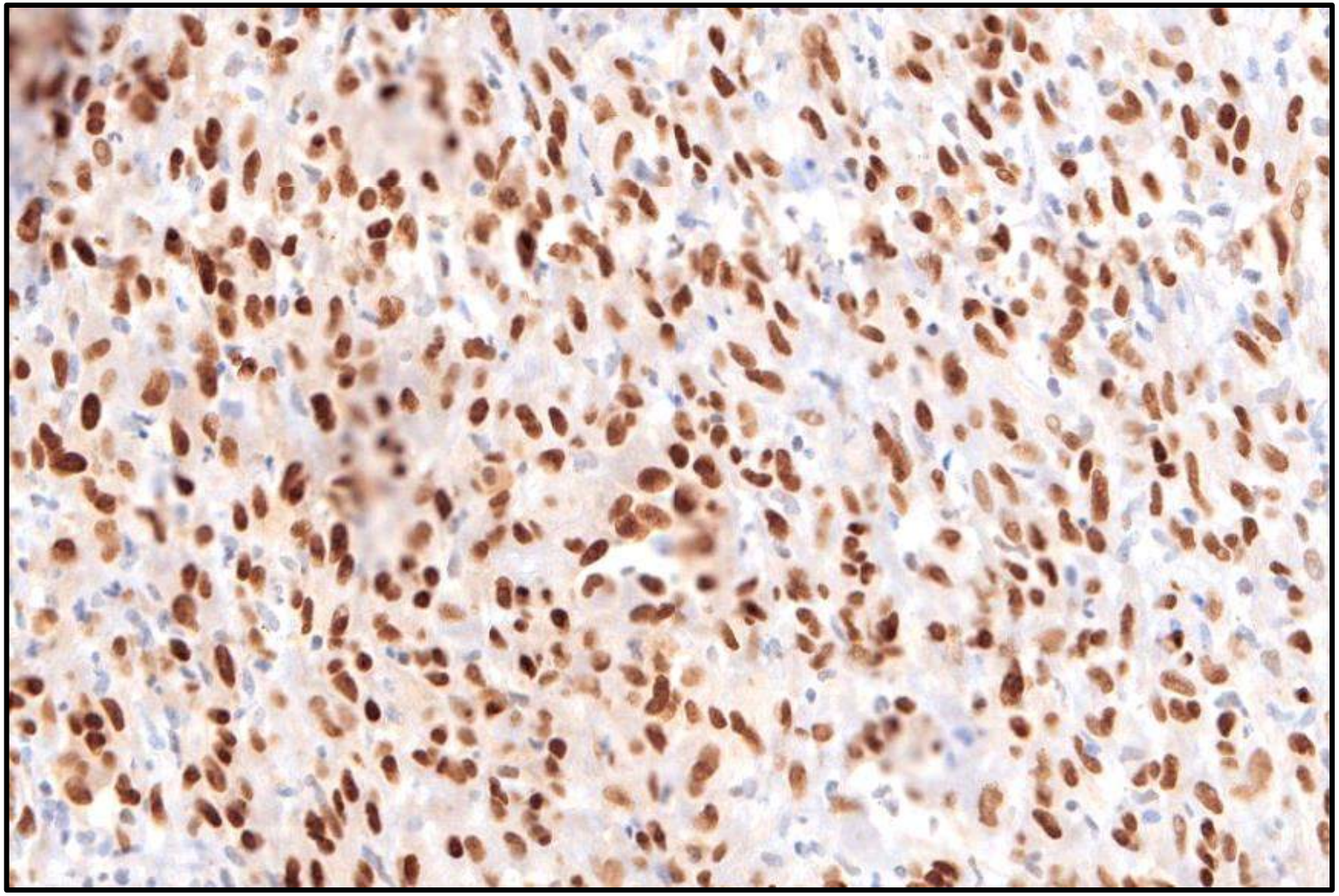
CD34



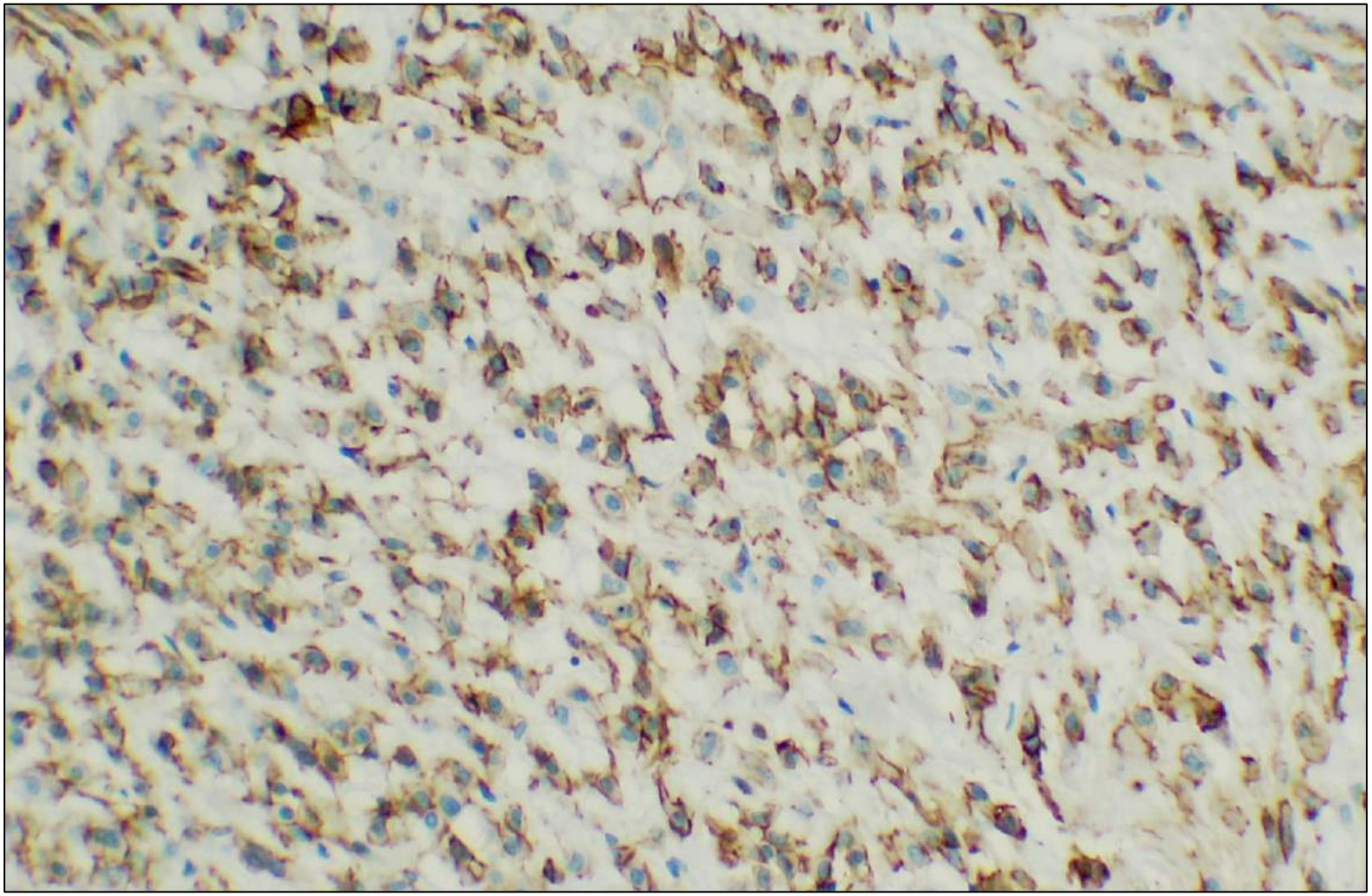
Narrowing the Differentials

- Pseudomyogenic Hemangioendothelioma
- Metastatic Carcinoma

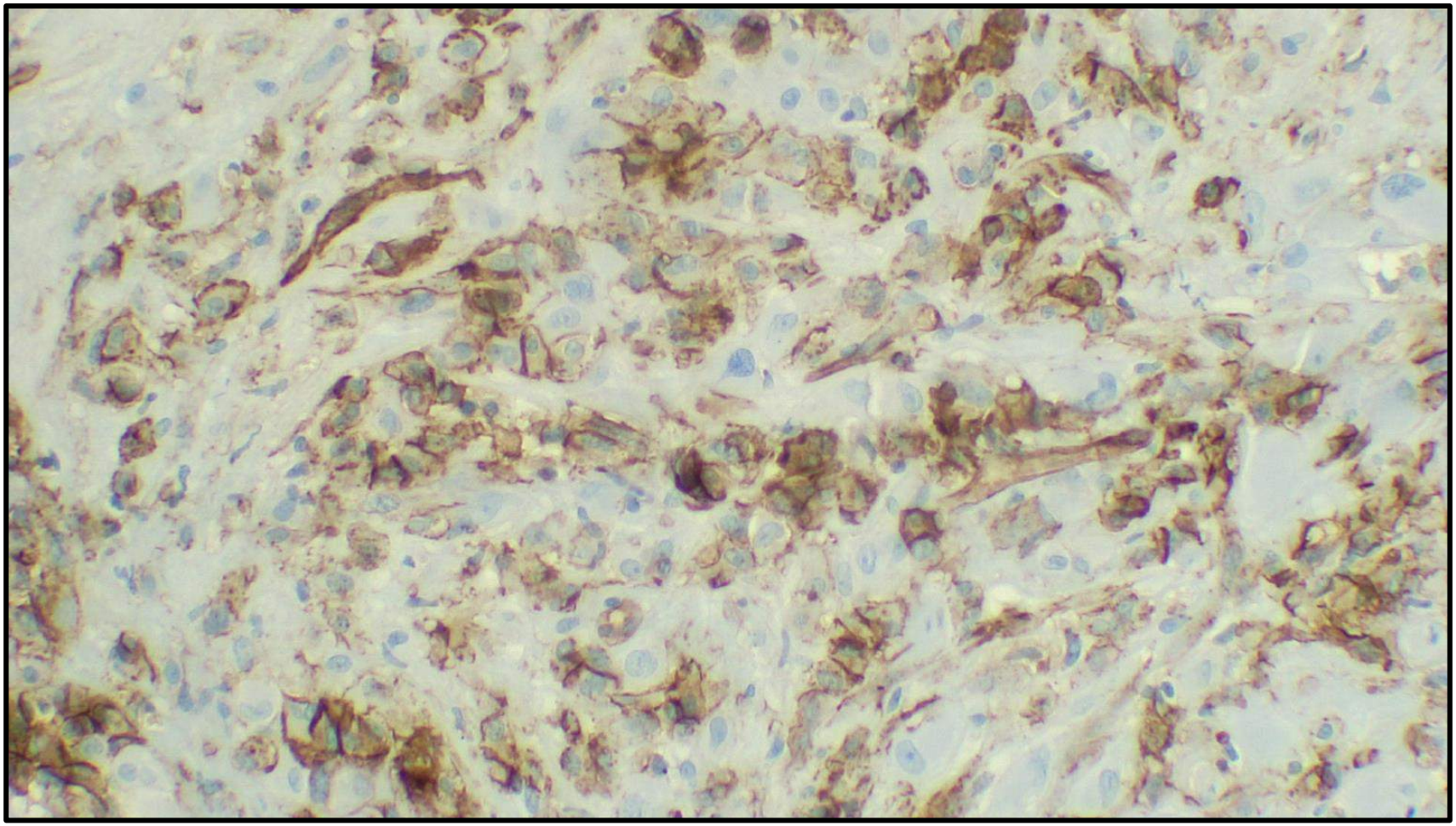
ERG-1



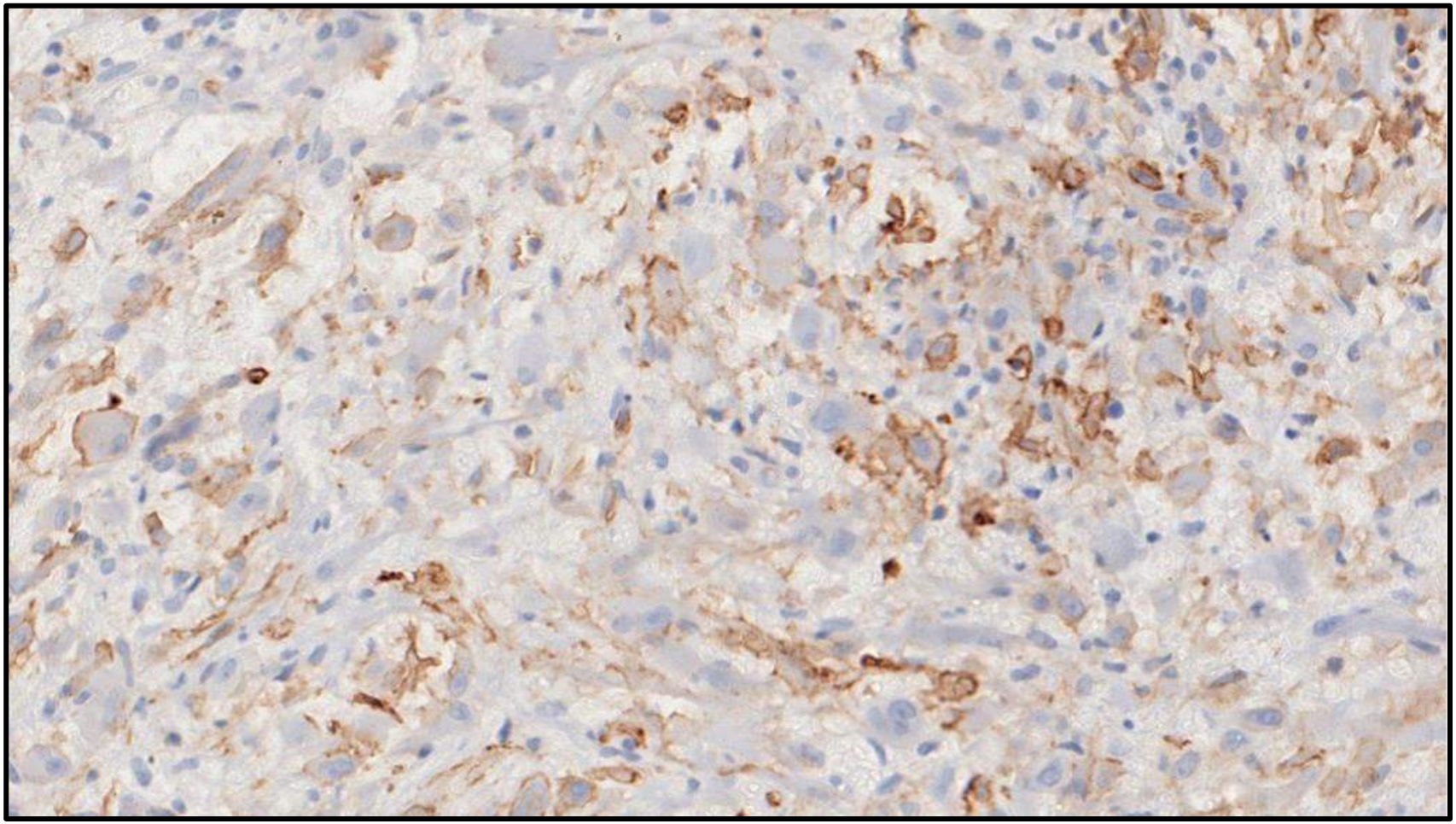
CD31



CD31



Thrombomodulin

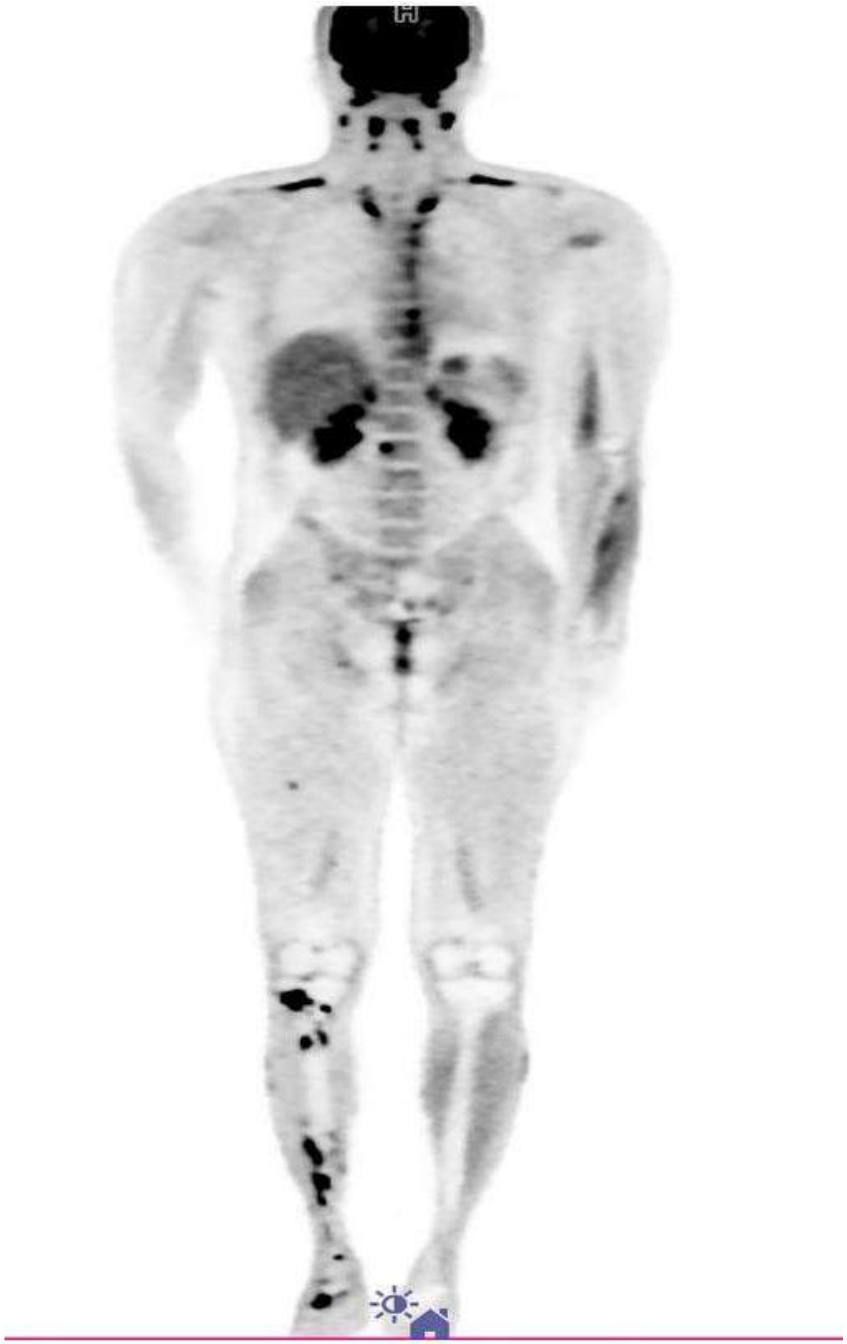


Summarizing IHC Findings in Tumor cells

Positive markers	Negative markers
CK	CD34
EGR-1	Desmin, Myo-D1, Myogenin
CD31	INI-1 Retained
Thrombomodulin	S100

Final Diagnosis

Pseudomyogenic Hemangioendothelioma



PET-CT

Multiple metaboloically active lytic/sclerotic lesions in:

- Multiple right tarsals
- Metatarsals bone
- Right tibia
- Right fibula
- Rght patella
- Right femur
- Right ischium
- Bilateral ilium
- Sacrum
- Multiple vertebrae
- sternum.
- Posterior compartment muscles of right thigh (1.0 x 0.9cm, SUV max 5.2)

Brief Discussion

- Pseudomyogenic hemangioendothelioma (PMH) is a rare vascular tumor of intermediate malignant potential
- Commonly presents as a subcutaneous & soft tissue mass with predilection for lower extremities
- Can present with combimed soft tissue and bony lesions
- PMH was previously recognized as fibroma like, fibrohistiocytic/myxoid variant of epithelioid sarcoma, due to its marked histological resemblance to epithelioid Sarcoma

- PMH is a distinct rarely metastasizing endothelial neoplasm which was adopted in 2013 in the 4th edition of WHO Classification of Tumors of soft tissue and bone

- PMH presenting as primary bone lesions is rare and can often mimick metastatic aetiology in view of multifocality

- PMH are typically characterized by balanced translocation t(7;19)(q22; q13) producing fusion of SERPINE1 and FOSB gene

- In terms of prognosis, PMH are locally recurrent and rarely metastasizing tumor

Management in the presented case

- Patient had disseminated bony disease but was primarily symptomatic for tibial lesion
- The data on systemic chemotherapy is limited
- It was decided to put patient on subcutaneous injections of Denosumab (120mg) 4 weekly
- After 8 months of follow up, the disease remained stable with marked symptomatic relief in pain.

THANK YOU