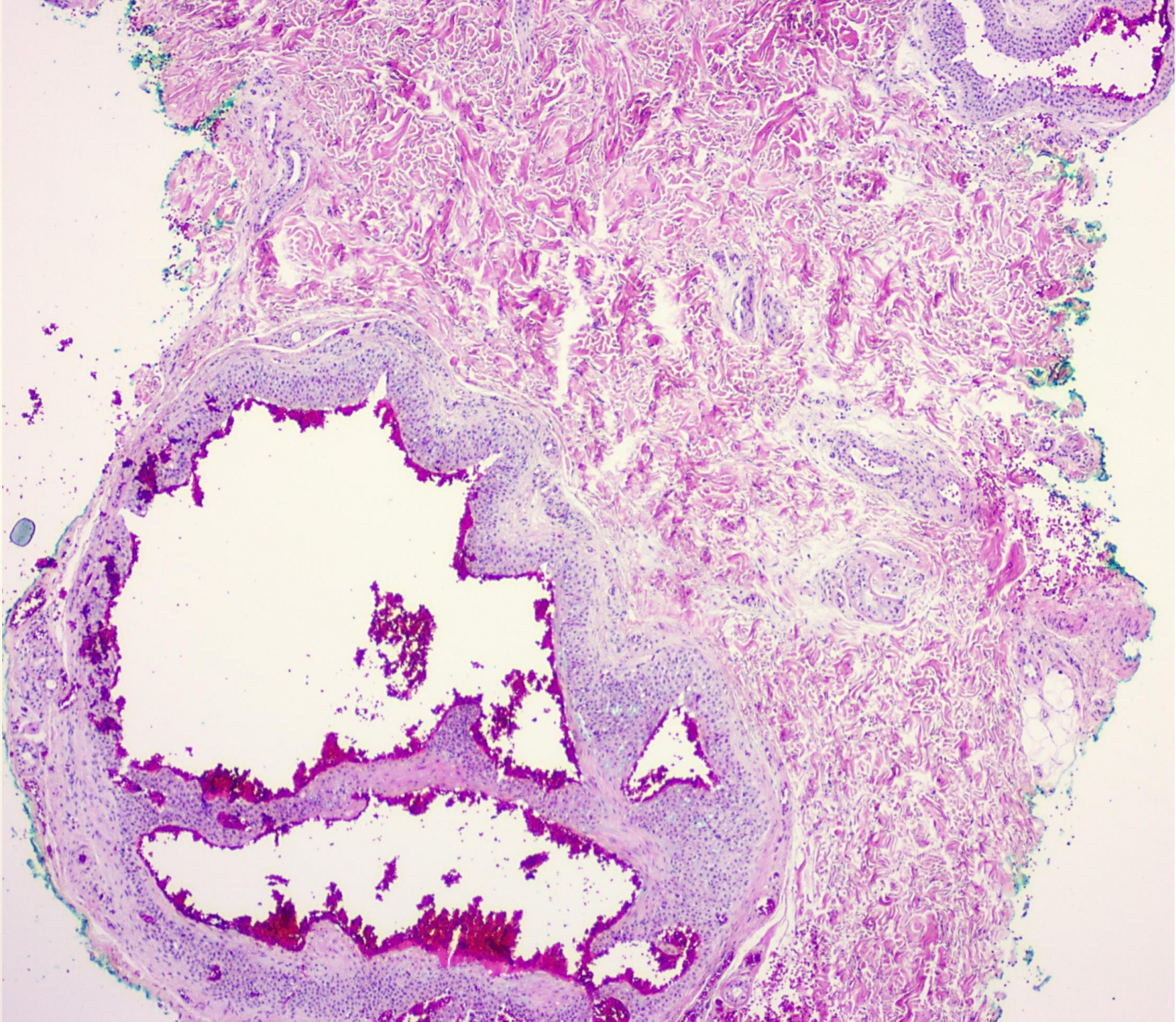
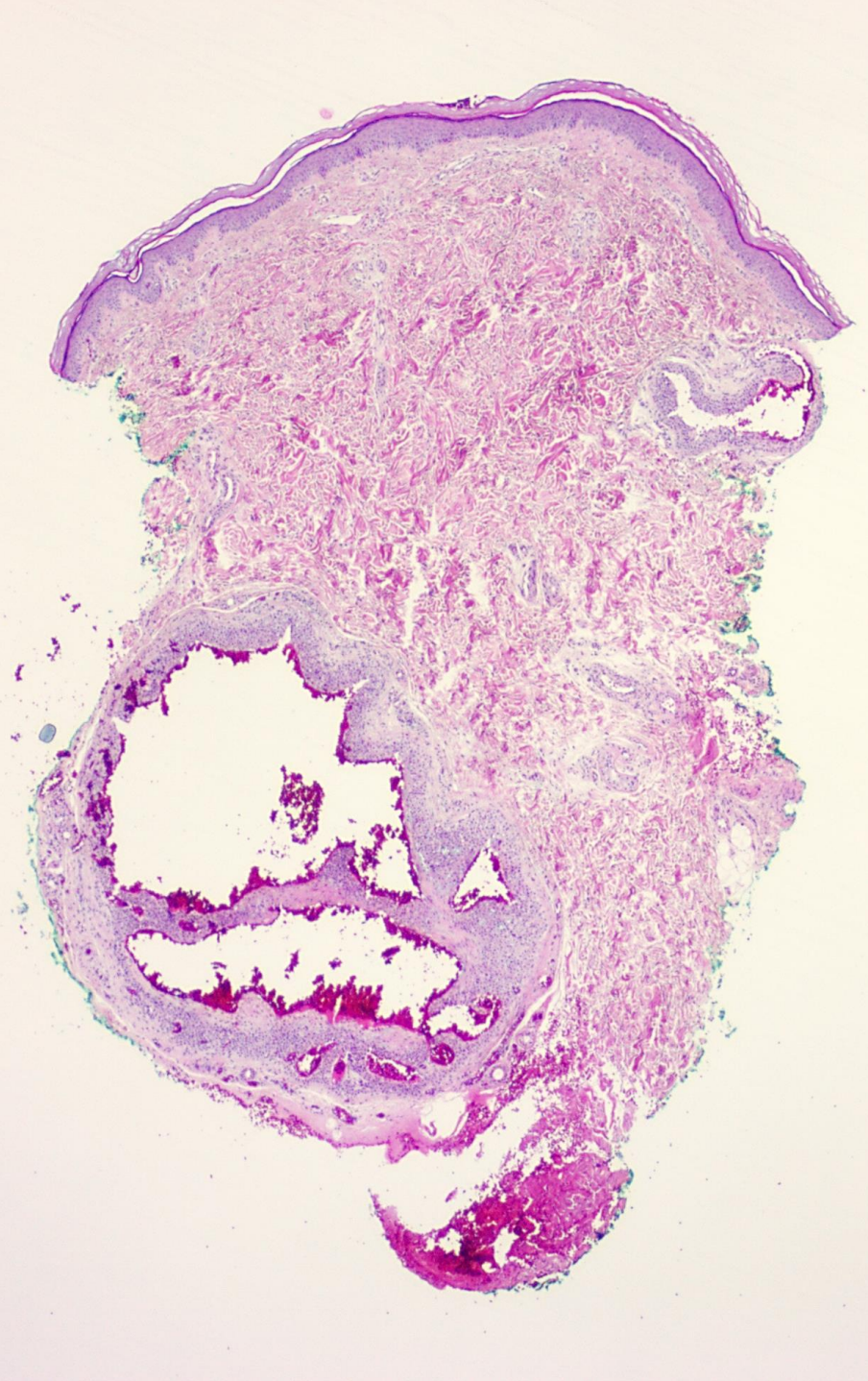


The background is a histological slide showing a large, irregularly shaped lesion. The lesion is filled with a dense collection of red-stained cells, likely erythrocytes, suggesting a hemorrhagic or vascular nature. The surrounding tissue is stained with hematoxylin and eosin (H&E), showing various cellular structures and nuclei in shades of blue and pink. The overall appearance is that of a skin biopsy specimen.

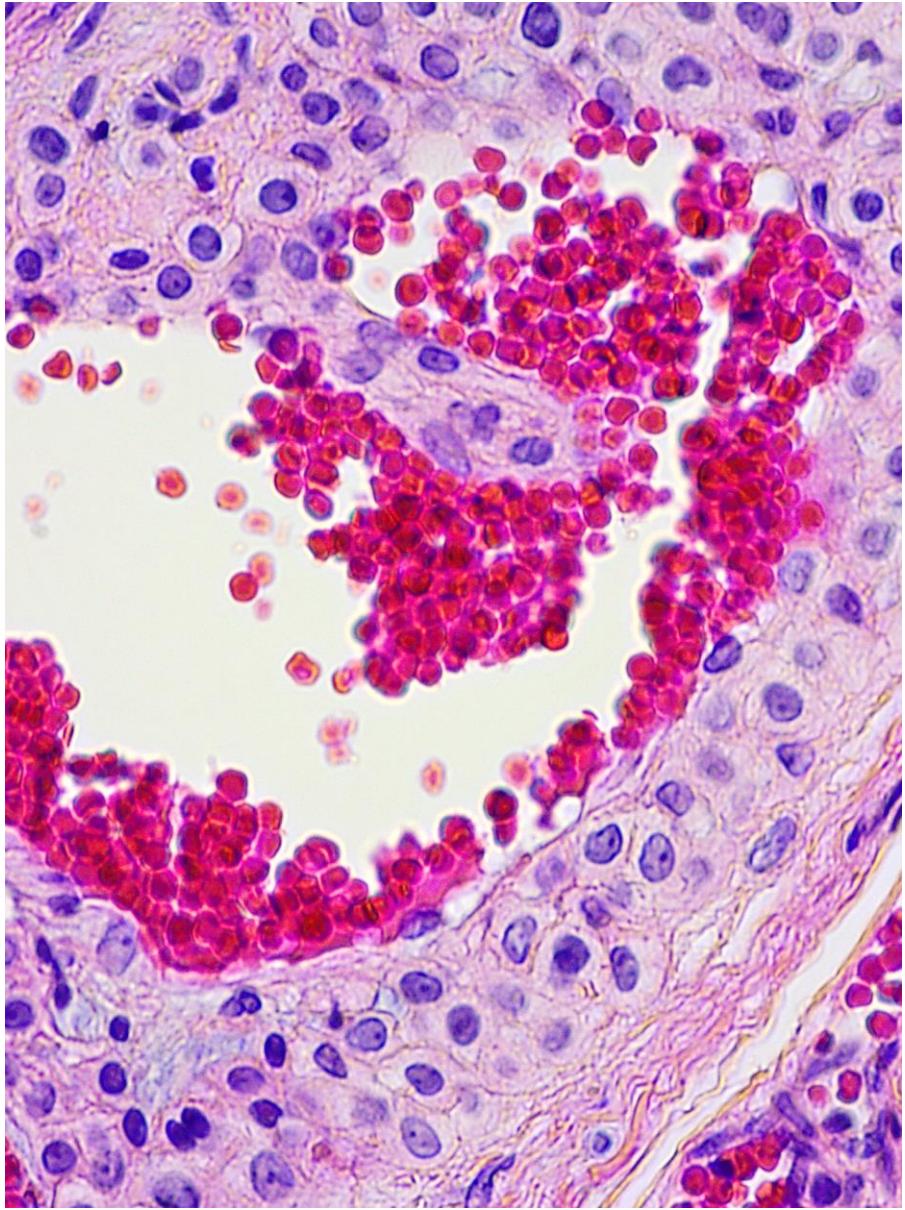
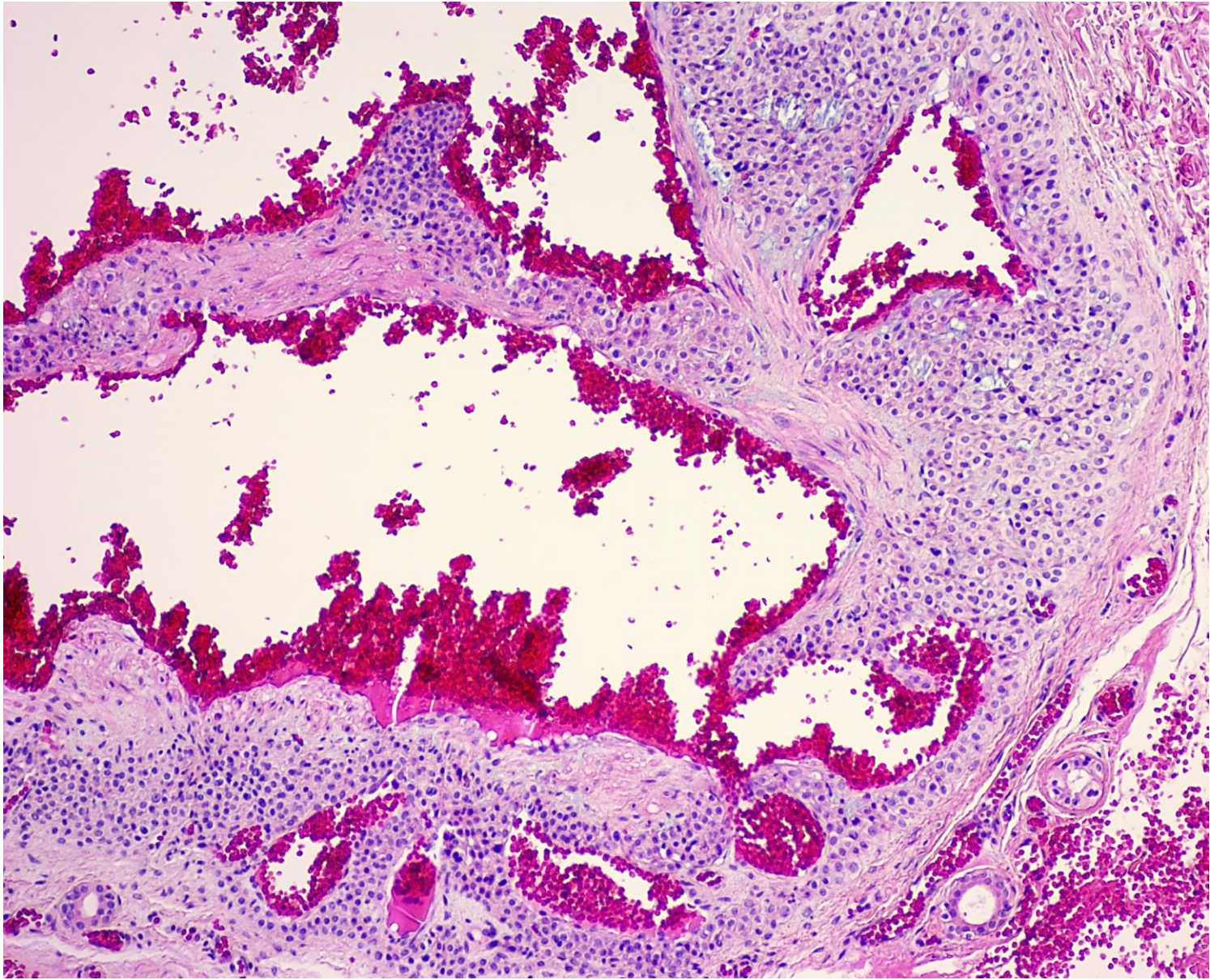
# 101 CASES DEMATOPATHOLOGY QUIZ CASES 77-86

Soheil S. Dadras MD-PhD

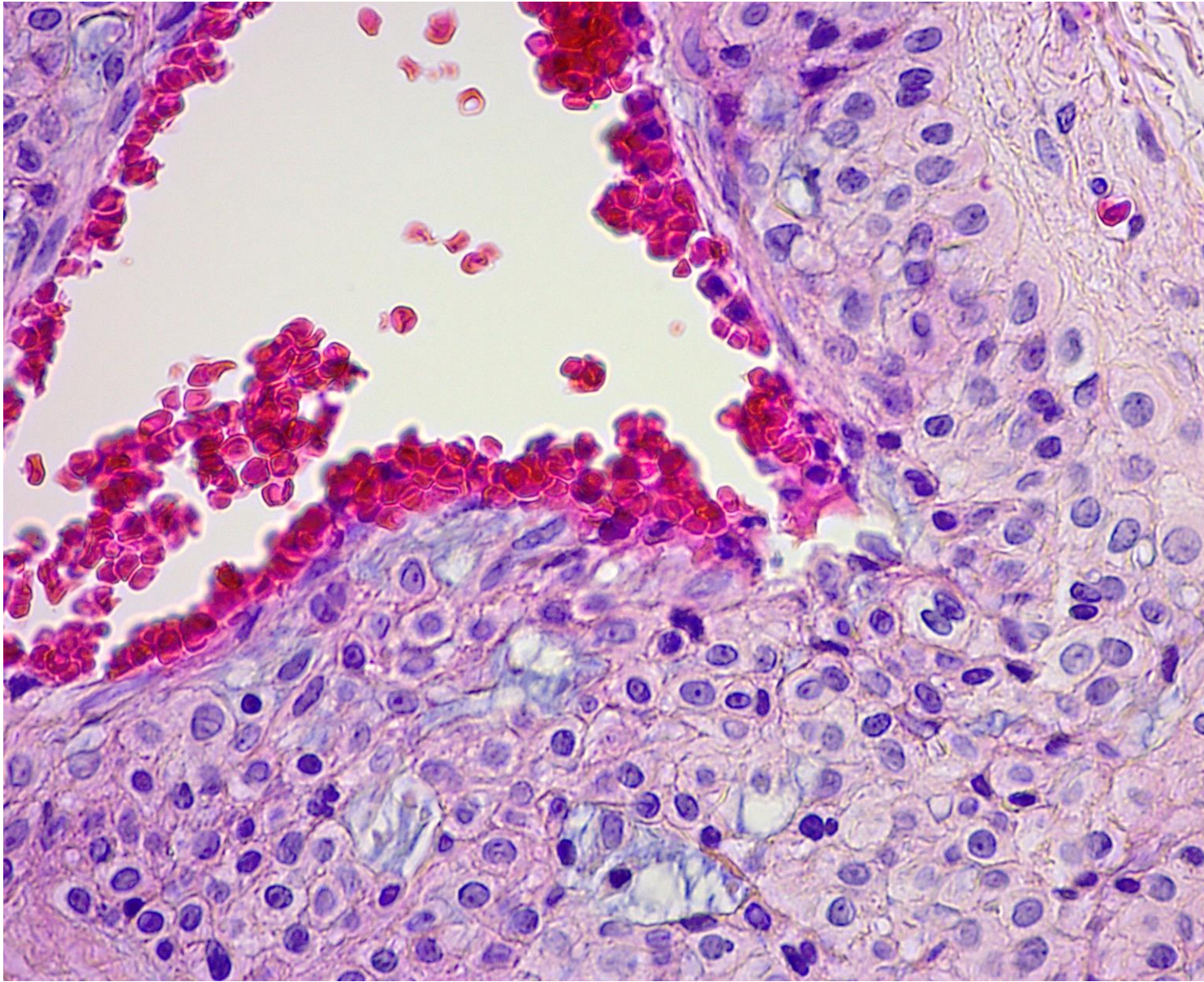














Case 77. 12F with blueish tender papules and nodules on the left foot. Family history of similar lesions. What is your diagnosis?

---

A. Kaposi's sarcoma, plaque stage

---

B. Intravascular papillary endothelial hyperplasia (Masson tumor)

---

C. Glomuvenous malformation (Glomangioma)

---

D. Glomus tumor

---

E. Venous lymphatic malformation

---



Case 77. 12F with blueish tender papules and nodules on the left foot. Family history of similar lesions. What is your diagnosis?

A. Kaposi's sarcoma, plaque stage

B. Intravascular papillary endothelial hyperplasia (Masson tumor)

C. Glomuvenous malformation (Glomangioma)

D. Glomus tumor

E. Venous lymphatic malformation



**Clinical Information:** 12F with blueish tender papules and nodules on the left foot (biopsied) and left knee. Family history similar lesions in mother. Ddx favor glomuvenous malformation, less likely venous malformation (25-15181).

## **DIAGNOSIS:**

Skin, Left Foot, Punch Biopsy:

- Glomuvenous malformation (Glomangioma), focally extending to the tissue edges and base.

**Teaching Points:** Boon LM, Mulliken JB, Enjolras O, Vikkula M. Glomuvenous malformation (glomangioma) and venous malformation: distinct clinicopathologic and genetic entities. Arch Dermatol. 2004 Aug;140(8):971-6. doi: 10.1001/archderm.140.8.971. PMID: 15313813.

## **Minimal Diagnostic Criteria:**

Small, round uniform epithelioid cell with distinct cell borders and punched out nuclei

Dilated, cavernous venous structures

Outer layer of thin smooth muscles

## **Differential Diagnosis:**

Venous lymphatic malformation

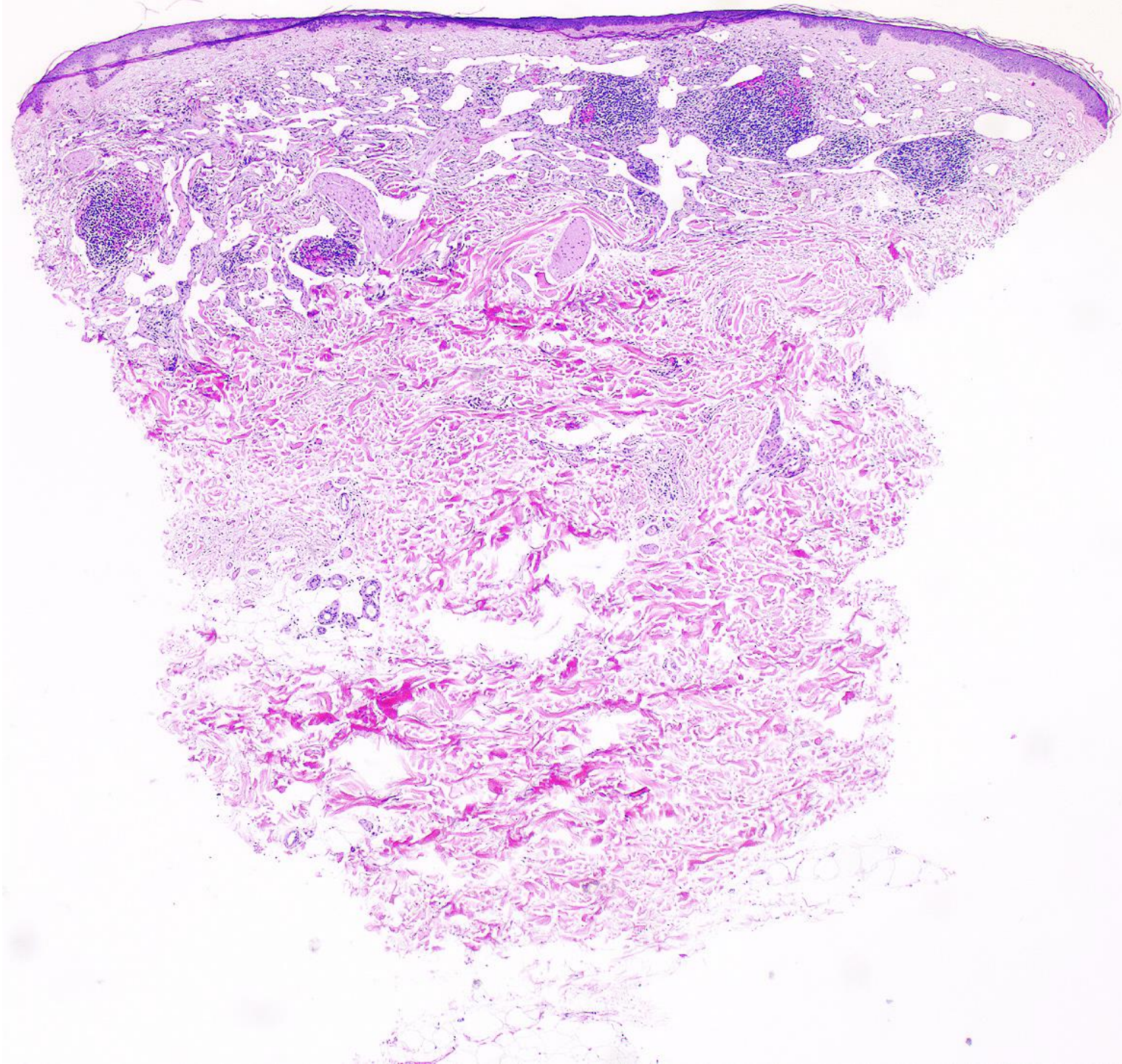
Glomangiomyoma/Glomangiopericytoma

Glomus tumor

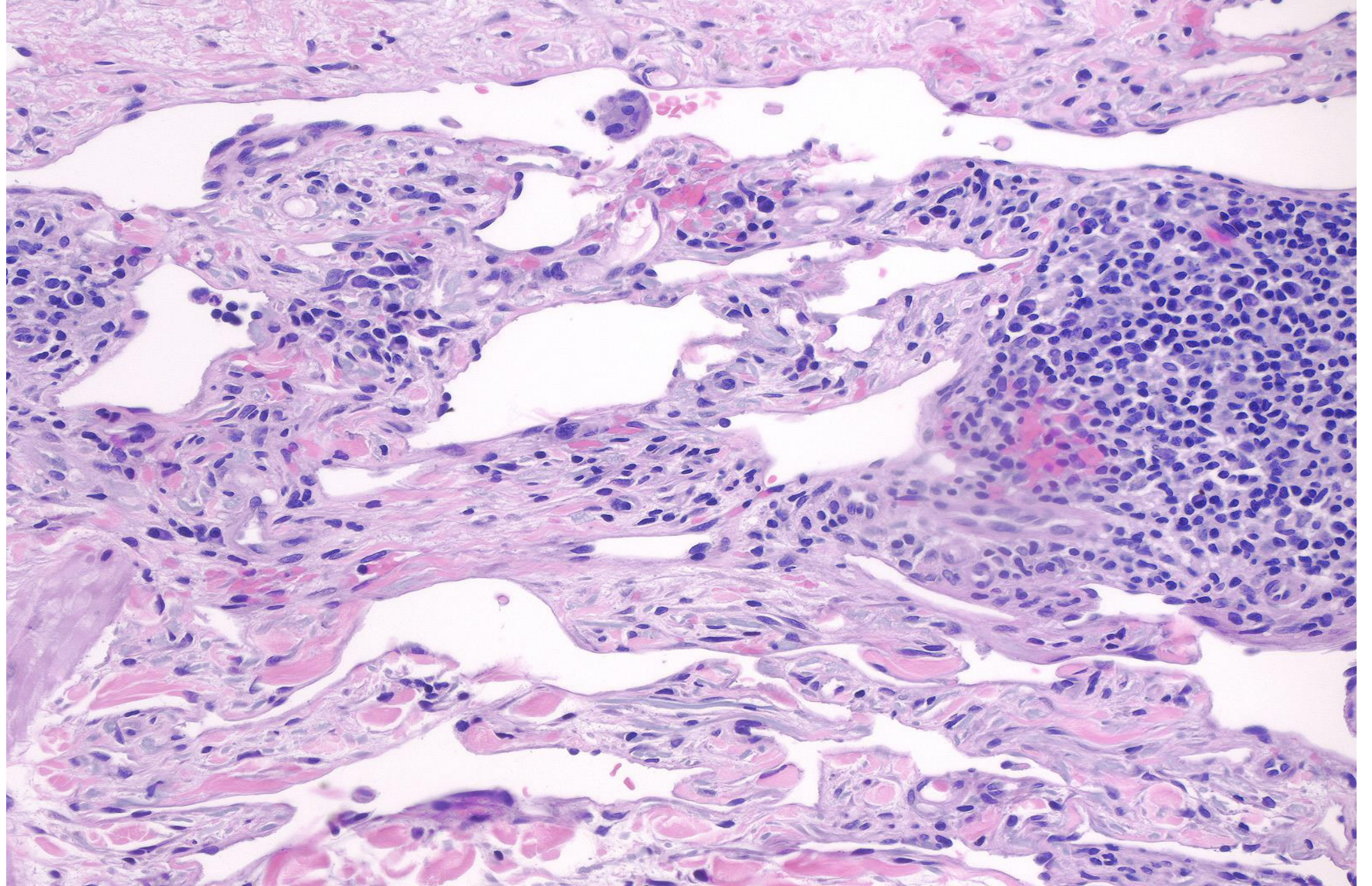
DIGITAL SKIN PATHOLOGY (DiSK)

Learn Histologic Diagnosis Case-By-Case











Case 78. 85M, arm, 4.5 cm red-purple-brown plaque. What is your diagnosis?

A. Kaposi's sarcoma, plaque stage

B. Intravascular papillary endothelial hyperplasia (Masson tumor)

C. Cherry angioma

D. Spindle cell hemangioma

E. Angiosarcoma



Case 78. 85M, arm, 4.5 cm red-purple-brown plaque. What is your diagnosis?

A. Kaposi's sarcoma, plaque stage

B. Intravascular papillary endothelial hyperplasia (Masson tumor)

C. Cherry angioma

D. Spindle cell hemangioma

E. Angiosarcoma

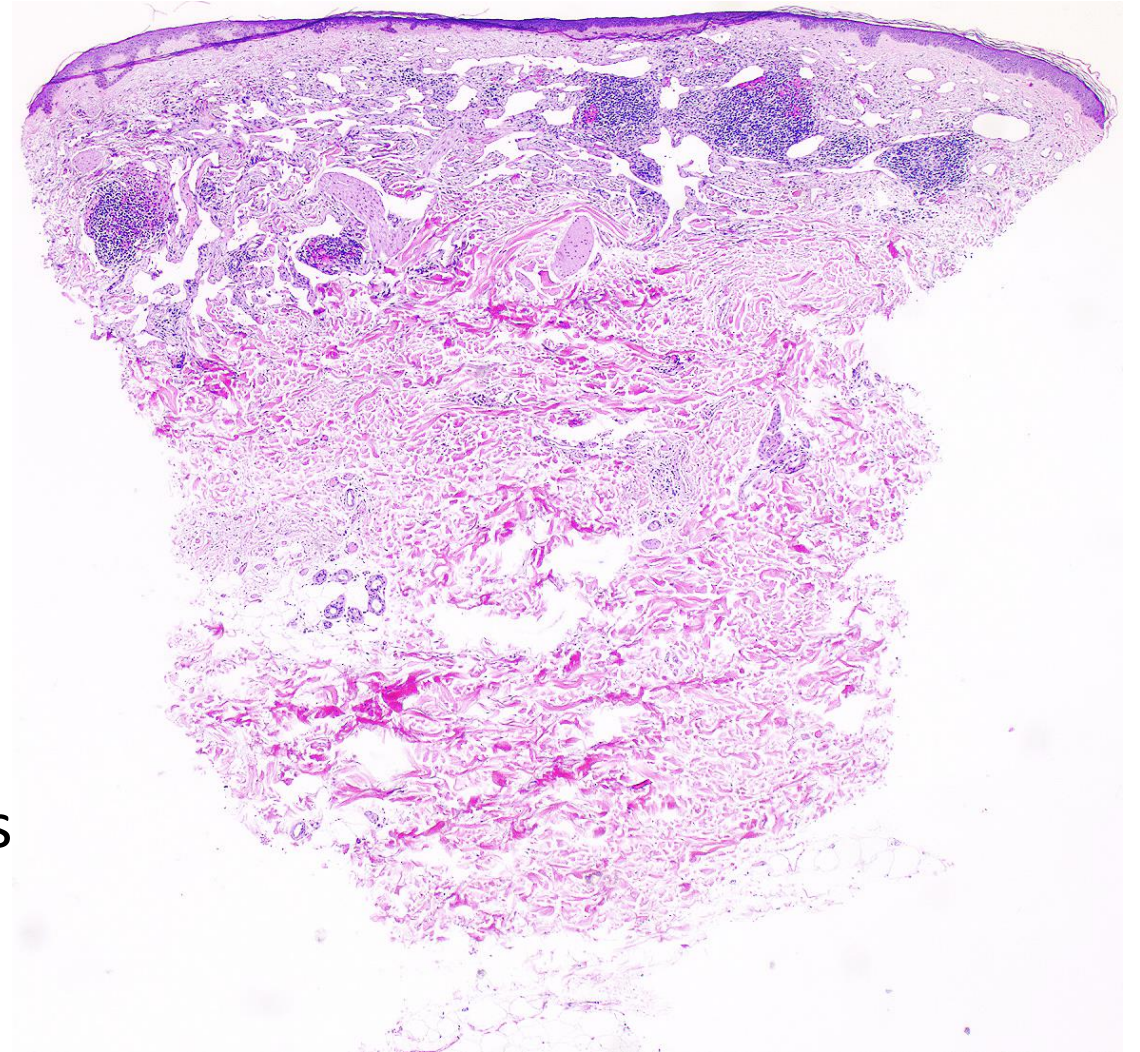


# KAPOSI SARCOMA

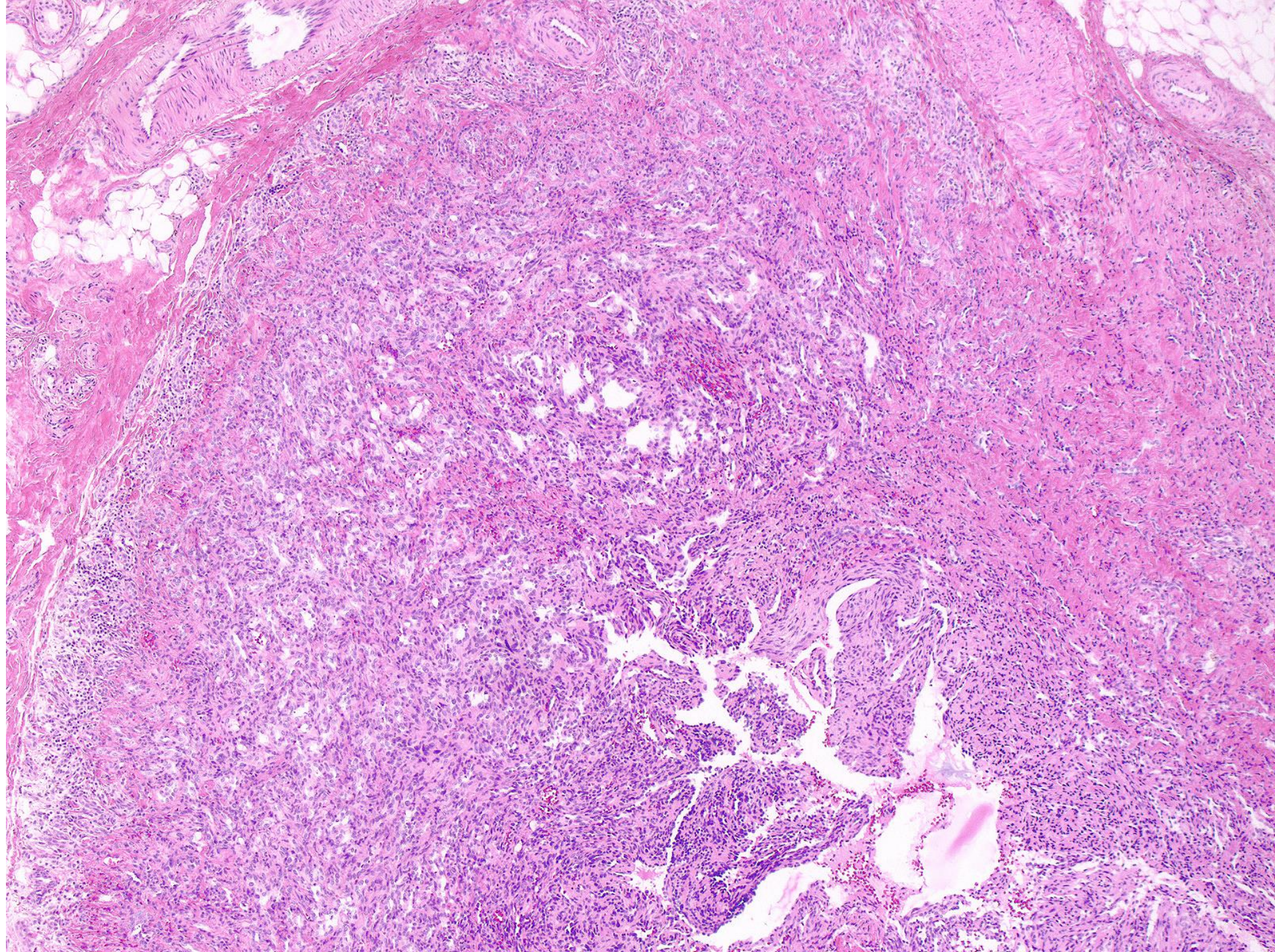
## Histologic features

### Plaque stage:

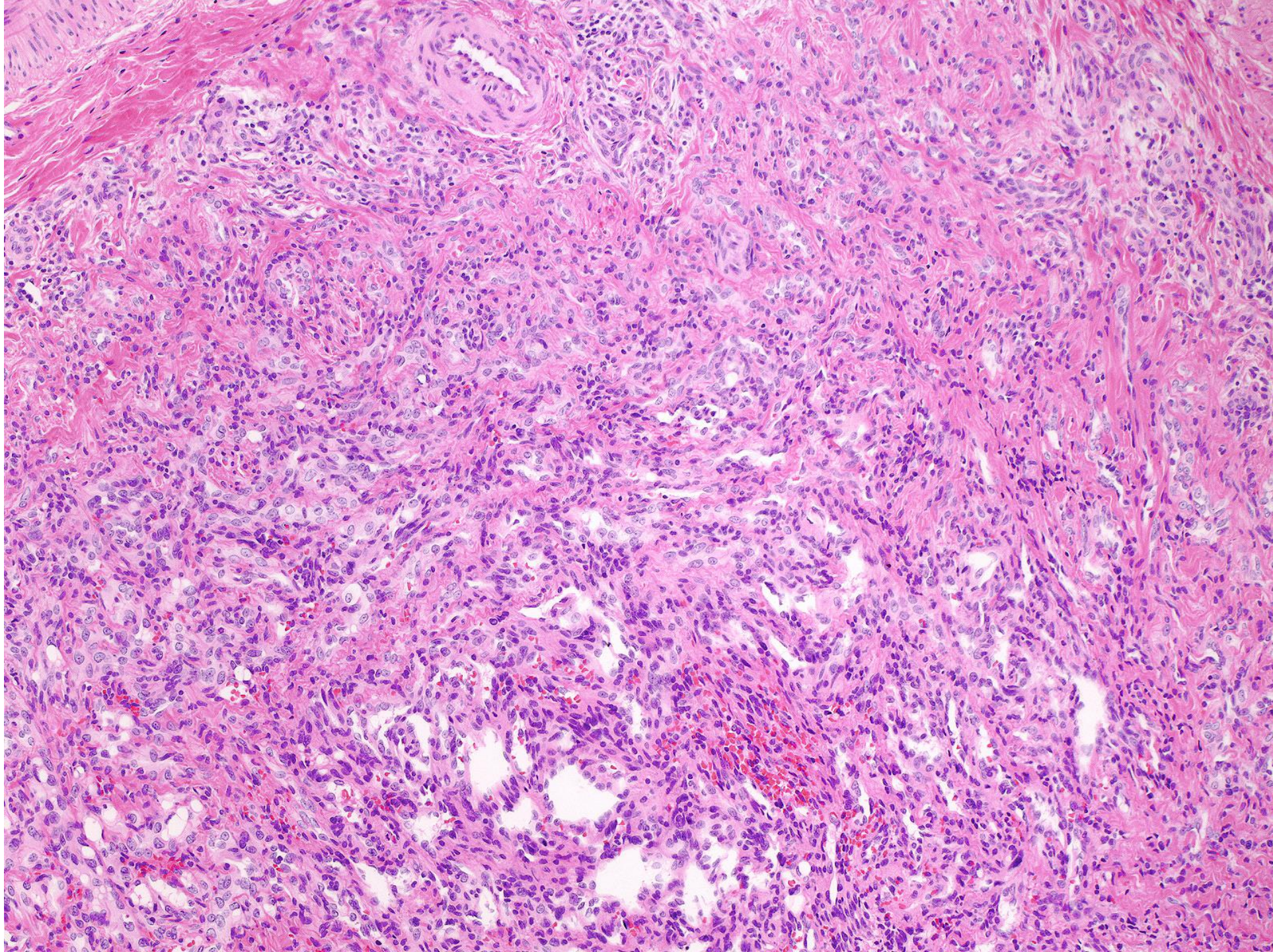
- Numerous ill-formed vessels in dermis
- Slit-like lumina
- The promontory sign: new vessels ensheath pre-existent ones
- Aggregates of lymphocytes, plasma cells
- Extravasated erythrocytes
- Hemosiderosis



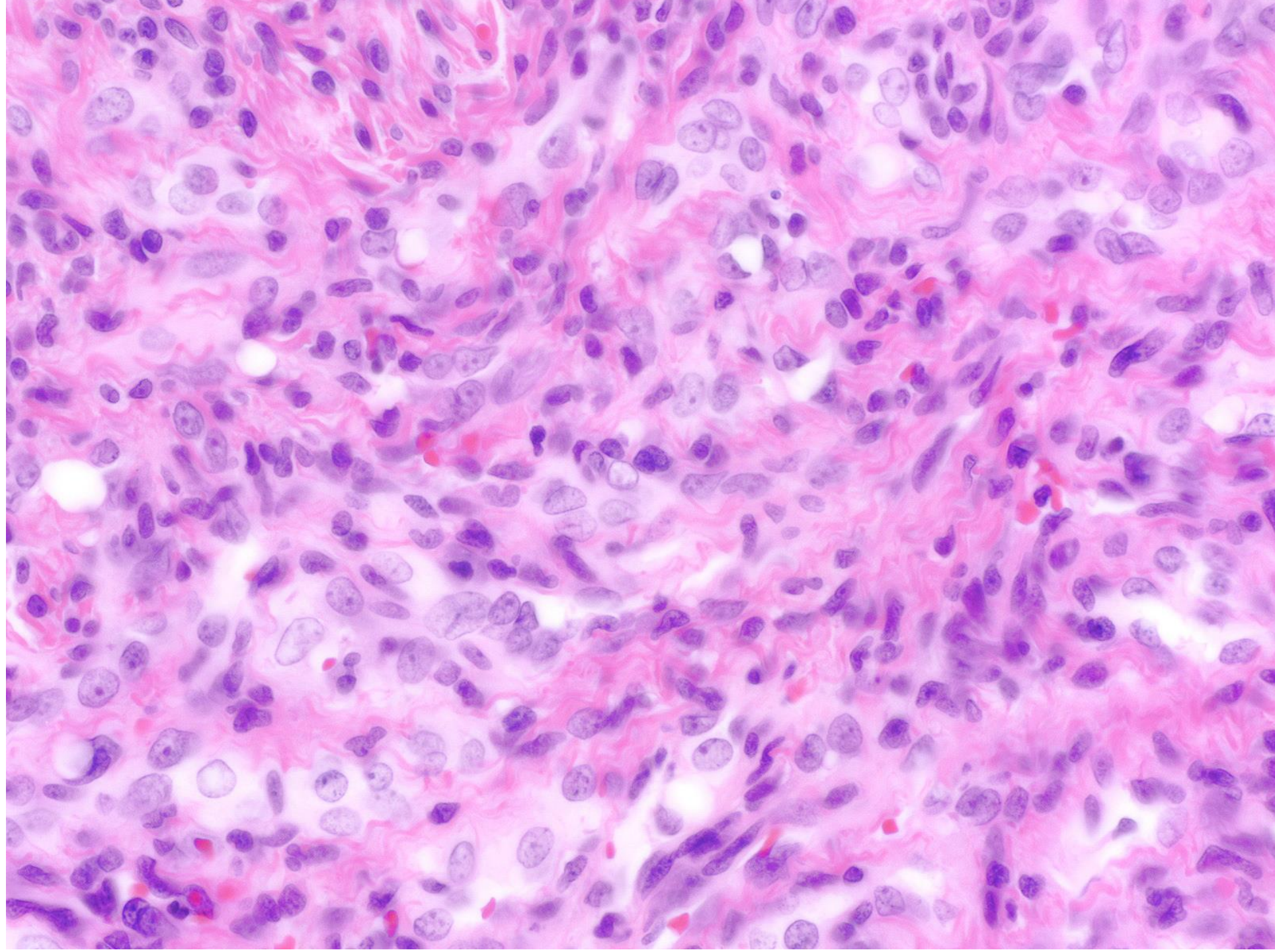














Case 79. 25F with a single red-blue, painful nodule on left foot. What is your diagnosis?

A. Kaposi's sarcoma

B. Intravascular endothelial hyperplasia (Masson tumor)

C. Non-involuting congenital hemangioma

D. Spindle cell hemangioma

E. Myopericytoma



Case 79. 25F with a single red-blue, painful nodule on left foot. What is your diagnosis?

A. Kaposi's sarcoma

B. Intravascular endothelial hyperplasia (Masson tumor)

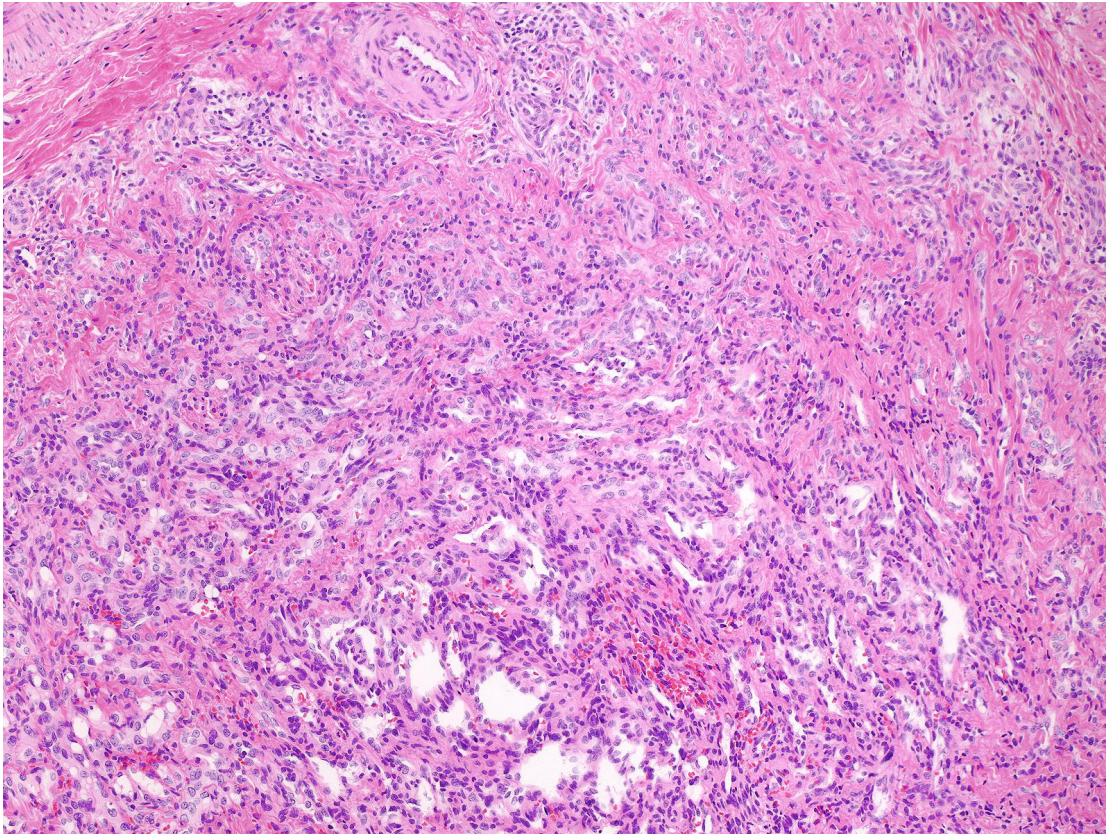
C. Non-involuting congenital hemangioma

D. Spindle cell hemangioma

E. Myopericytoma



# Spindle cell hemangioma



Age: Most common in young adults (20s–40s)

Location: Extremities (hands, fingers, feet) – most common

Presentation: Solitary or multiple red-blue nodules

Associations:

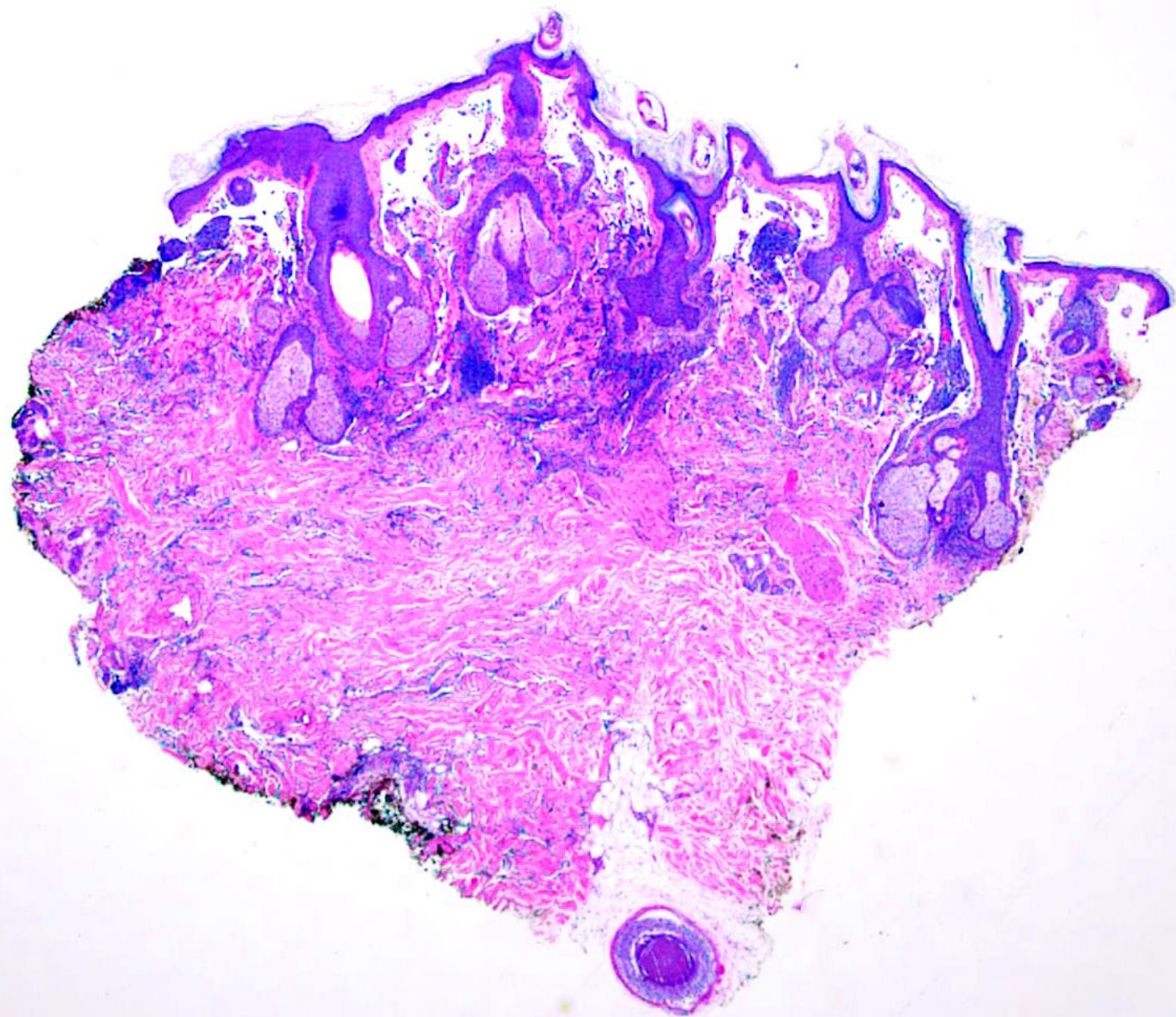
Maffucci syndrome (enchondromas + vascular tumors)

Klippel-Trenaunay syndrome (capillary-lymphatic-venous malformations)

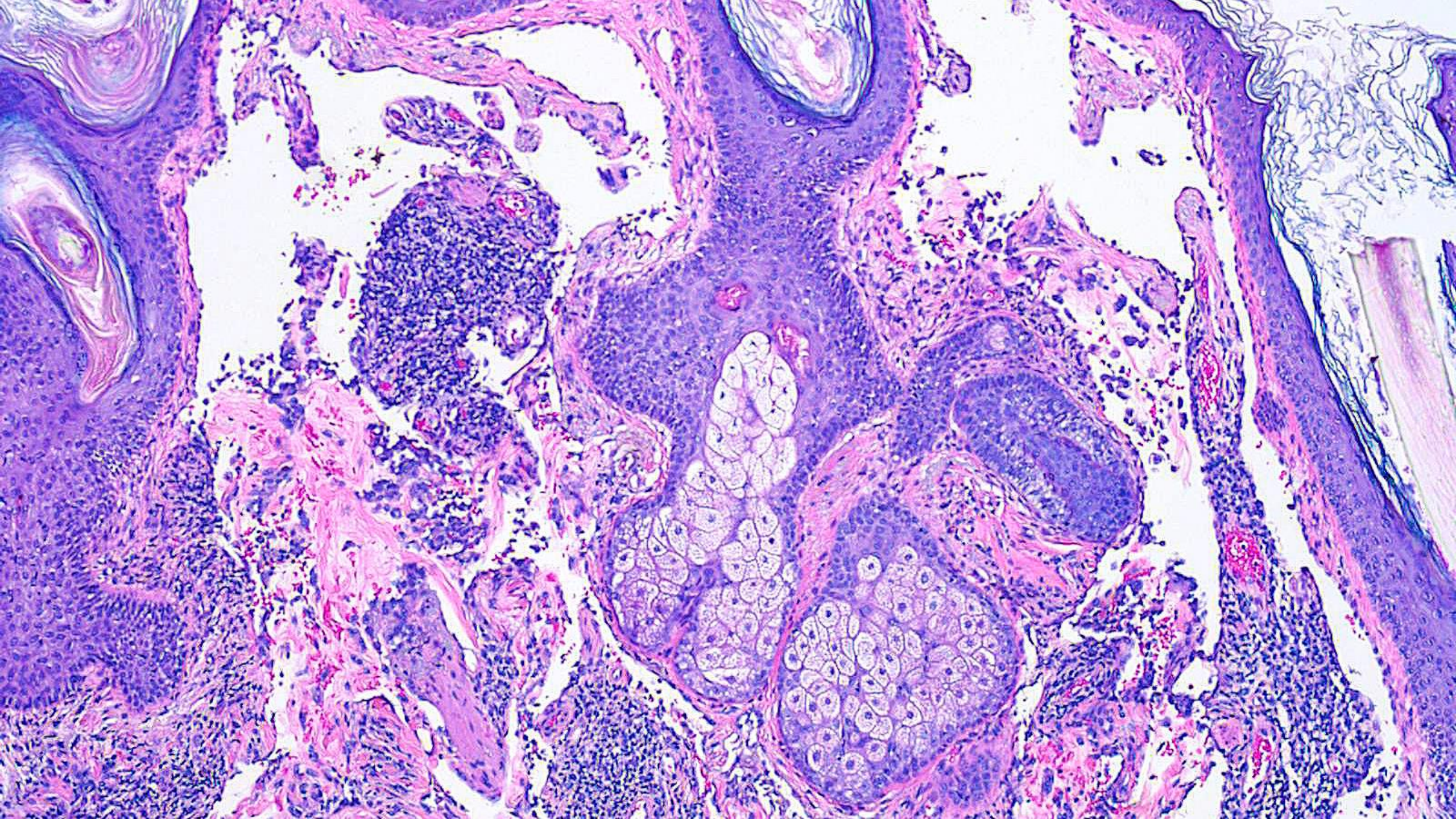
Key Histopathologic clues:

- Mixed cavernous vessels + spindle cell proliferation
- No true infiltrative growth or necrosis (helps distinguish from angiosarcoma)
- Negative for HHV-8 (unlike Kaposi sarcoma)

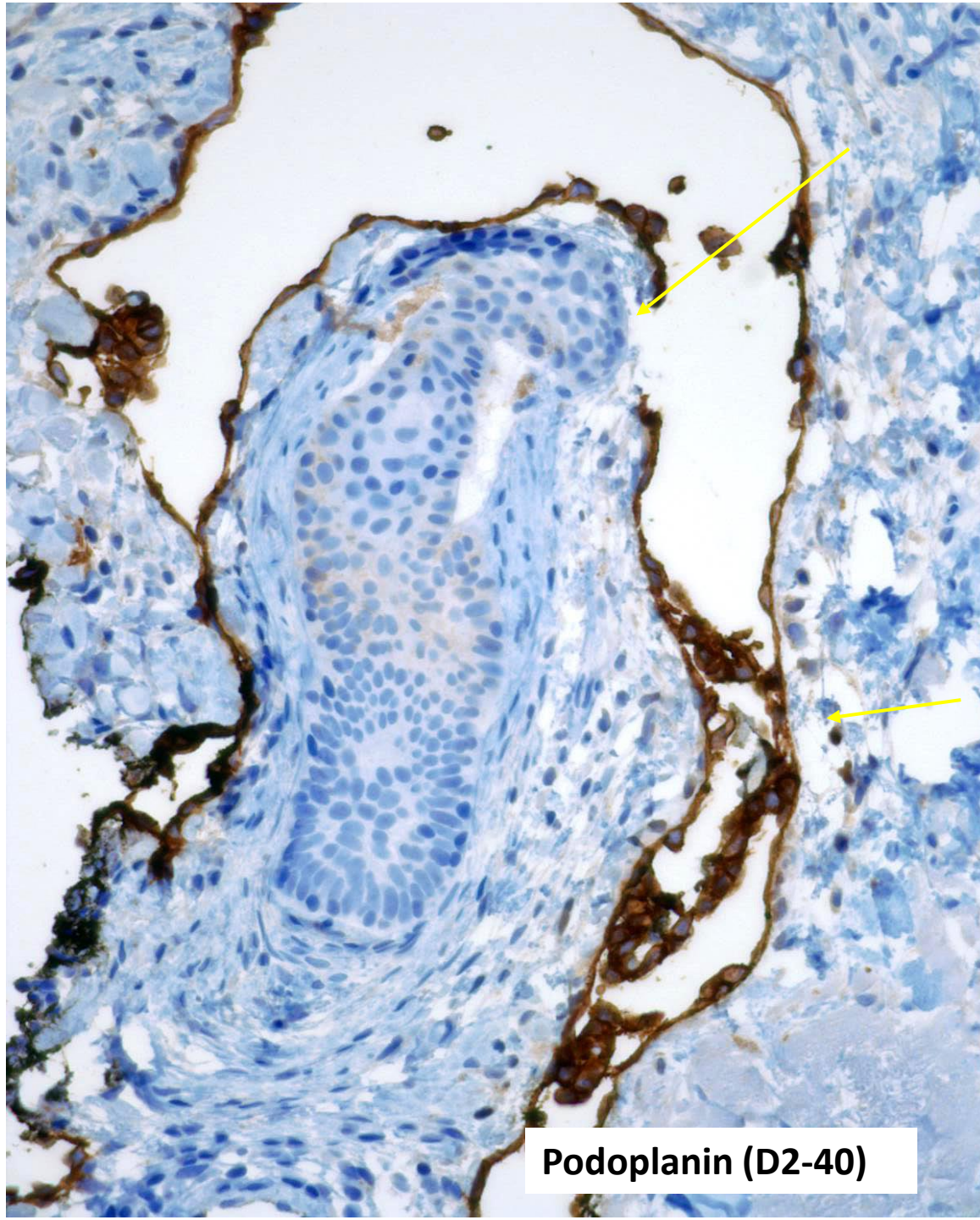




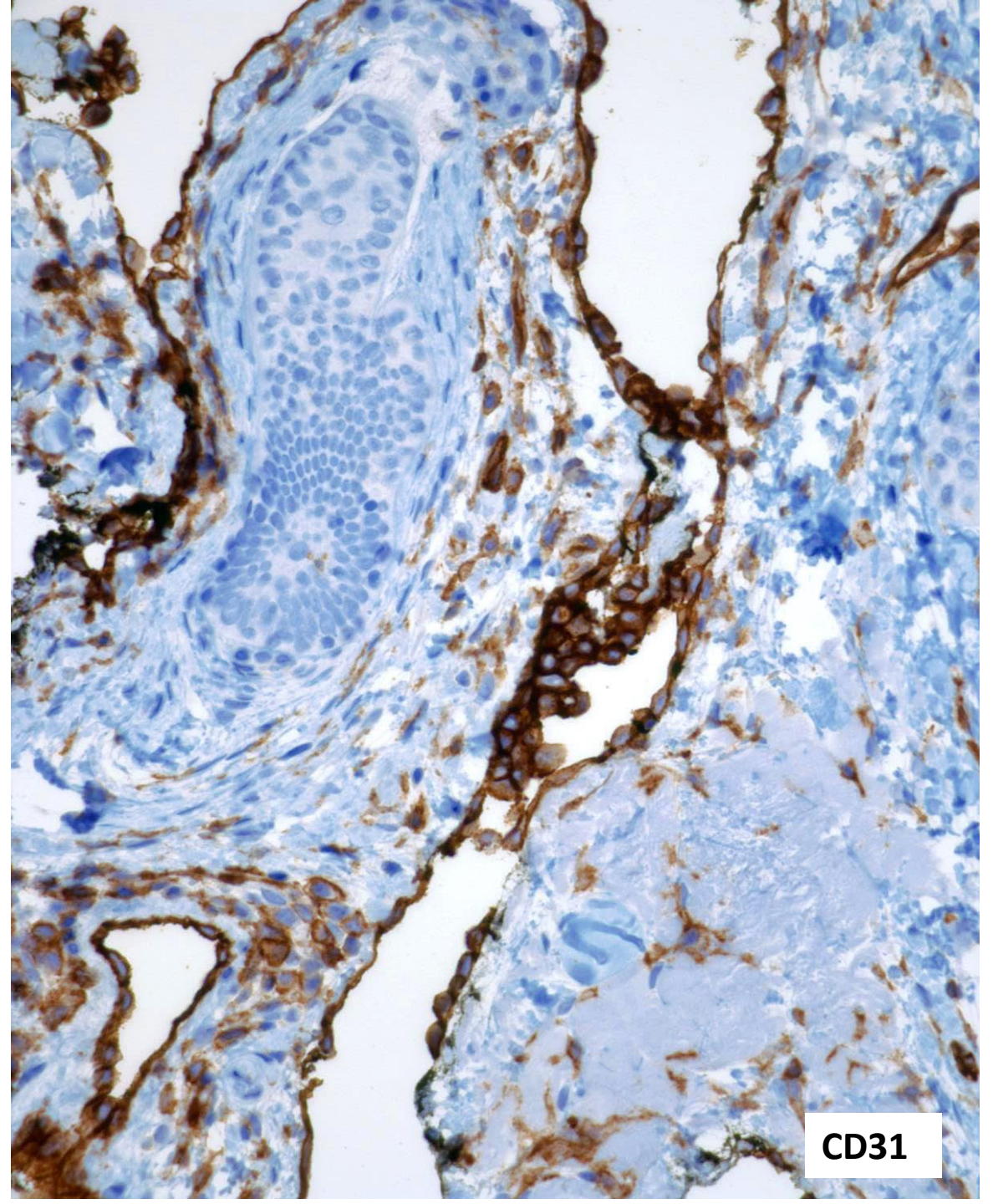








**Podoplanin (D2-40)**



**CD31**



Case 80. 76M with red-purple spreading rash with swelling of face near the ear. What is your diagnosis?

---

A. Kaposi's sarcoma

---

B. Intravascular endothelial hyperplasia (Masson tumor)

---

C. Angiosarcoma

---

D. Spindle cell hemangioma

---

E. Lymphangioma circumscriptum

---



Case 80. 76M with red-purple spreading rash with swelling of face near the ear. What is your diagnosis?

A. Kaposi's sarcoma

B. Intravascular endothelial hyperplasia (Masson tumor)

C. Angiosarcoma

D. Spindle cell hemangioma

E. Lymphangioma circumscriptum



**Clinical Information:** 76/M with red- purple spreading rash with swelling of face near the ear

**DIAGNOSIS:**

SKIN, LEFT FACE, EXCISION/BIOPSY:

ANGIOSARCOMA, EXTENDING TO THE TISSUE EDGES.

Comment: By immunohistochemistry, the lesional cells are positive for CD31, CD34, and Podoplanin while negative for HHV8. ki-67 highlights increased proliferative index in the endothelial cells (>5%).

**Teaching Points:**

Look for dissection of collagen

Look for multilayering of hyperchromatic nuclei with high proliferative index

Ancillary IHC: ERG (nuclear), CD31, CD34

Some angiosarcomas can be associated with lymphedema and exhibit lymphatic phenotype (Podoplanin+)

**Minimal Diagnostic Criteria:**

Dilated, irregular & anastomosing vascular channels

Channels dissect dermal collagen bundles

Vascular lumens are lined by multilayered nuclei, hyperchromatic and pleomorphic

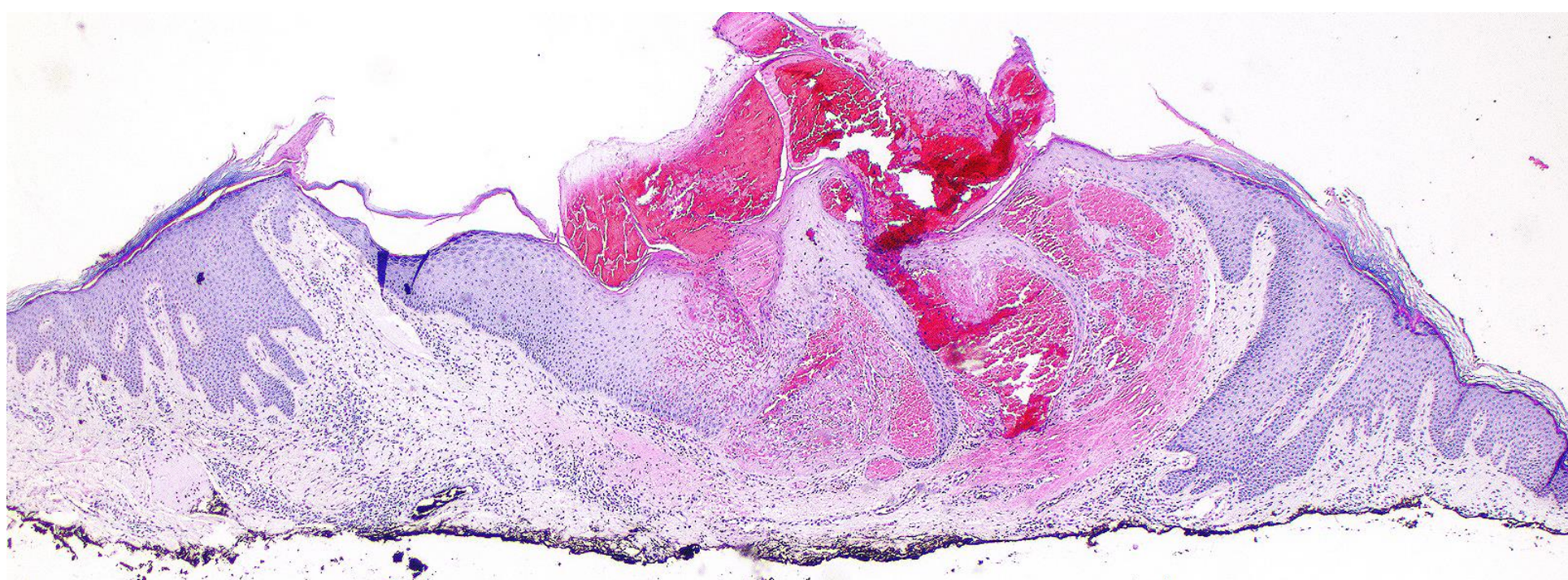
**Differential Diagnosis:**

Lymphangioma

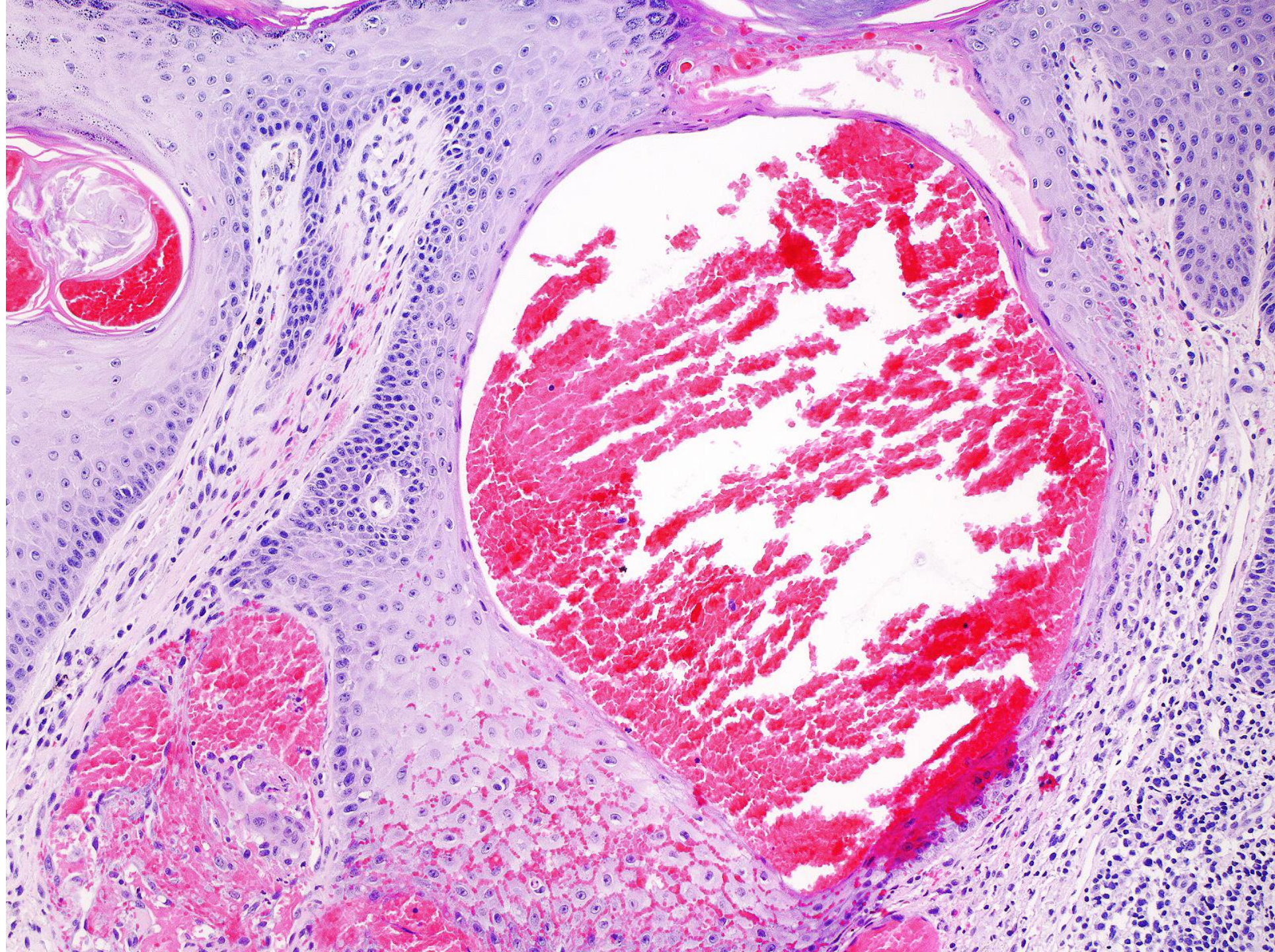
Kaposi sarcoma

Masson tumor











Case 81. 32F, left leg, a single purple crusted papule. What is your diagnosis?

---

A. Kaposi's sarcoma

---

B. Intravascular endothelial hyperplasia (Masson tumor)

---

C. Cherry angioma

---

D. Spindle cell hemangioma

---

E. Angiokeratoma

---



Case 81. 32F, left leg, a single purple crusted papule. What is your diagnosis?

A. Kaposi's sarcoma

B. Intravascular endothelial hyperplasia (Masson tumor)

C. Cherry angioma

D. Spindle cell hemangioma

E. Angiokeratoma



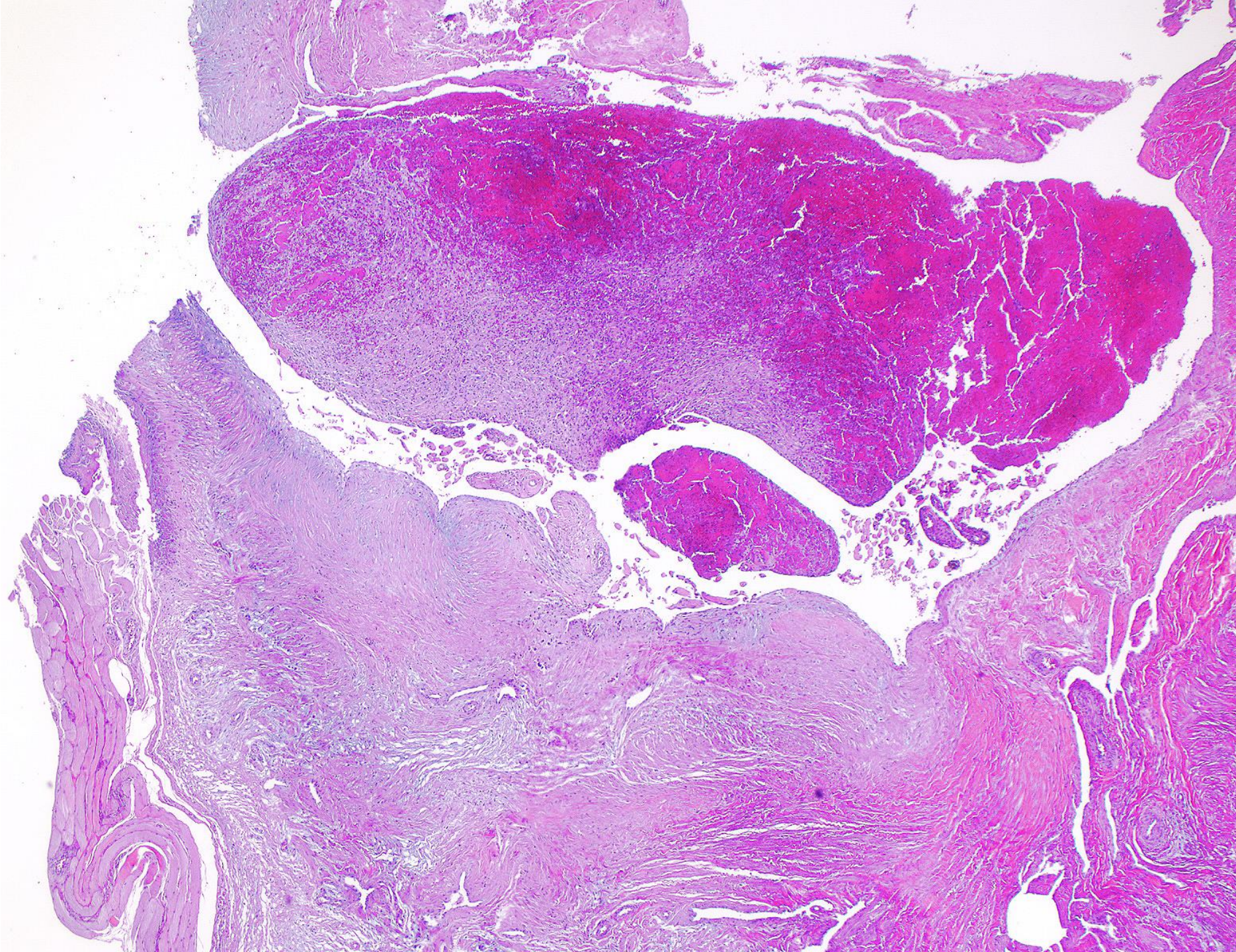
# ANGIOKERATOMA

- Histologic features:
  - Congested small dilated blood vessels in papillary dermis
  - Push up into epidermis (appear to be in the epidermis)
  - Overlying papillomatosis, acanthosis
  - Intracytoplasmic lipid vacuoles in endothelium pericytes, fibroblasts (in Anderson-Fabry's disease)

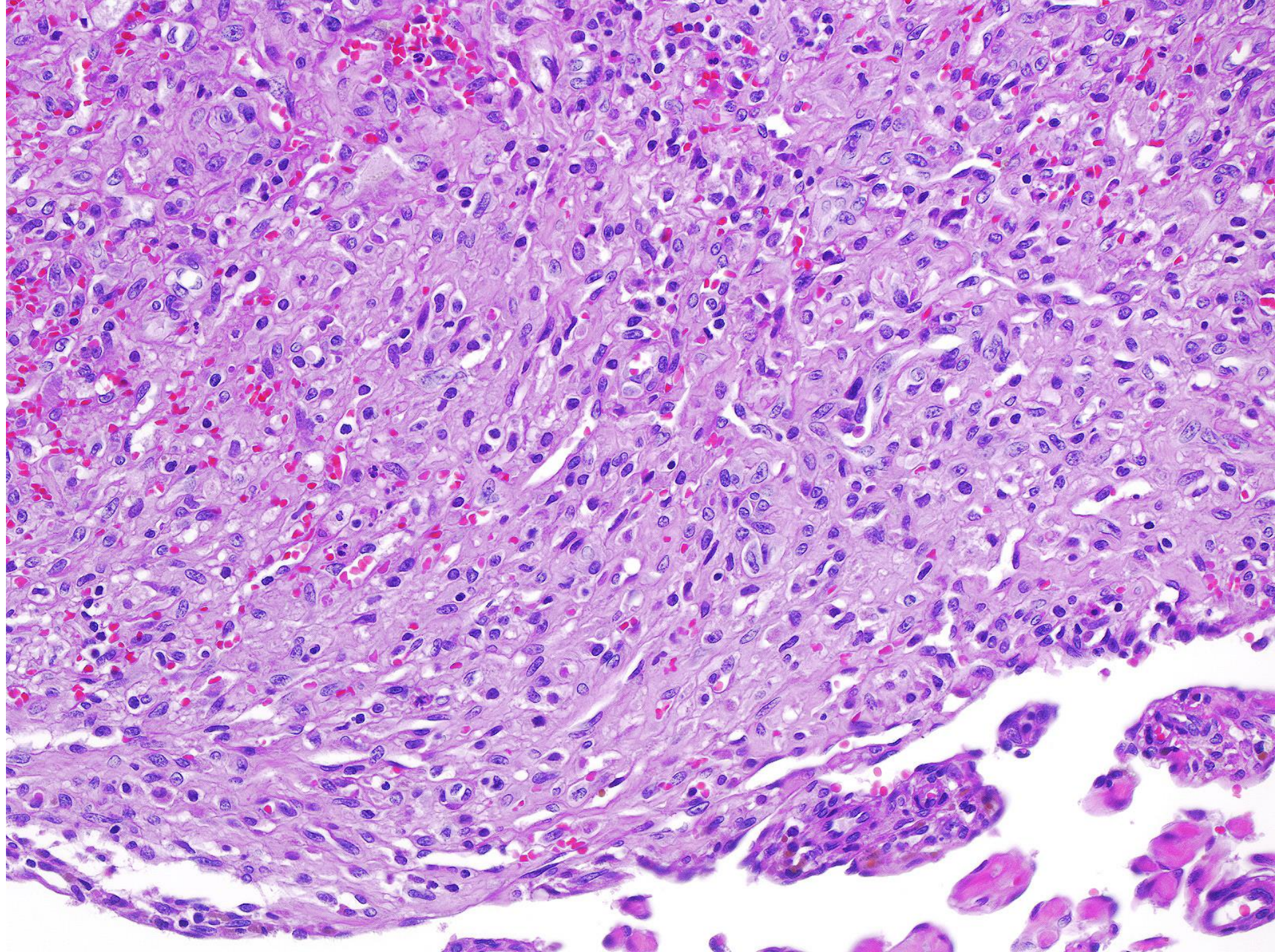


- Clinical features:
  - Of Fordyce: scrotum (old males), vulva (young females)
  - Of Mibelli: warty papules upper & lower extremities of children females
  - Circumscriptum: rare grouped papules, upper & lower extremities of children females
  - Corporis diffusum: red papules 'bathing-trunk' areas, associated with Anderson-Fabry's disease (deficiency of lysosomal  $\alpha$ -galactosidase A)
  - Solitary & multiple-wide age & anatomic site











Case 82. 57M, left arm, 4.5 cm red-purple painful mass. What is your diagnosis?

A. Kaposi's sarcoma

B. Intravascular papillary endothelial hyperplasia (Masson tumor)

C. Cherry angioma

D. Spindle cell hemangioma

E. Angiokeratoma



Case 82. 57M, left arm, 4.5 cm red-purple painful mass. What is your diagnosis?

A. Kaposi's sarcoma

B. Intravascular papillary endothelial hyperplasia (Masson tumor)

C. Cherry angioma

D. Spindle cell hemangioma

E. Angiokeratoma



# INTRAVASCULAR PAPILLARY ENDOTHELIAL HYPERPLASIA (MASSON TUMOR)

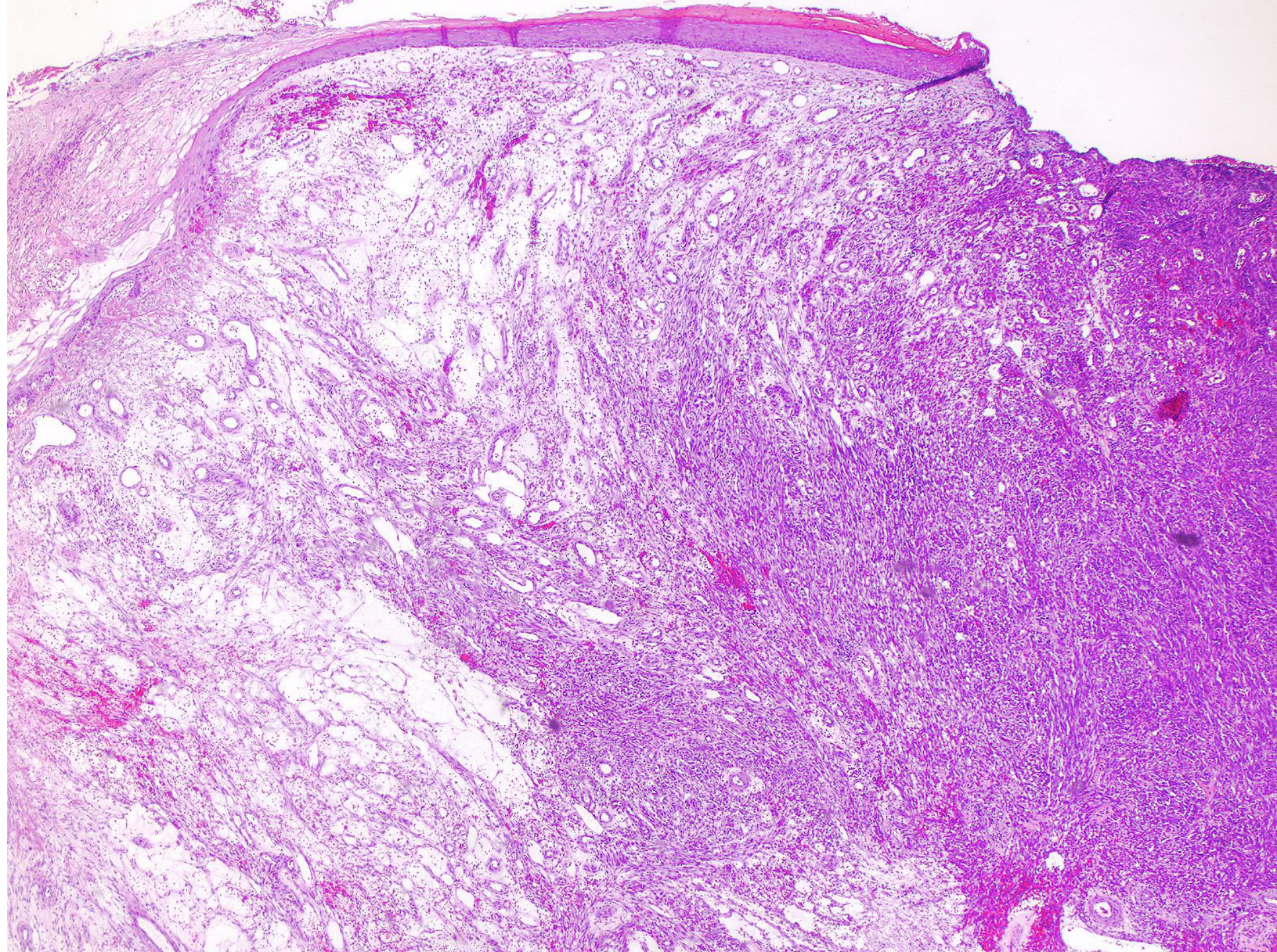
## Histologic features

- Organizing thrombus in a hemangioma (confined)
- No atypia or multilayering (angiosarcoma)
- Proliferation of plump endothelial cells forming papillary structures within a vascular lumen
- Eosinophilic hyaline material
- Lacks infiltrative growth (unlike angiosarcoma)
- Differential Diagnosis:
  - Angiosarcoma (infiltrative growth, nuclear atypia, necrosis)
  - Organizing thrombus (lacks endothelial proliferation)

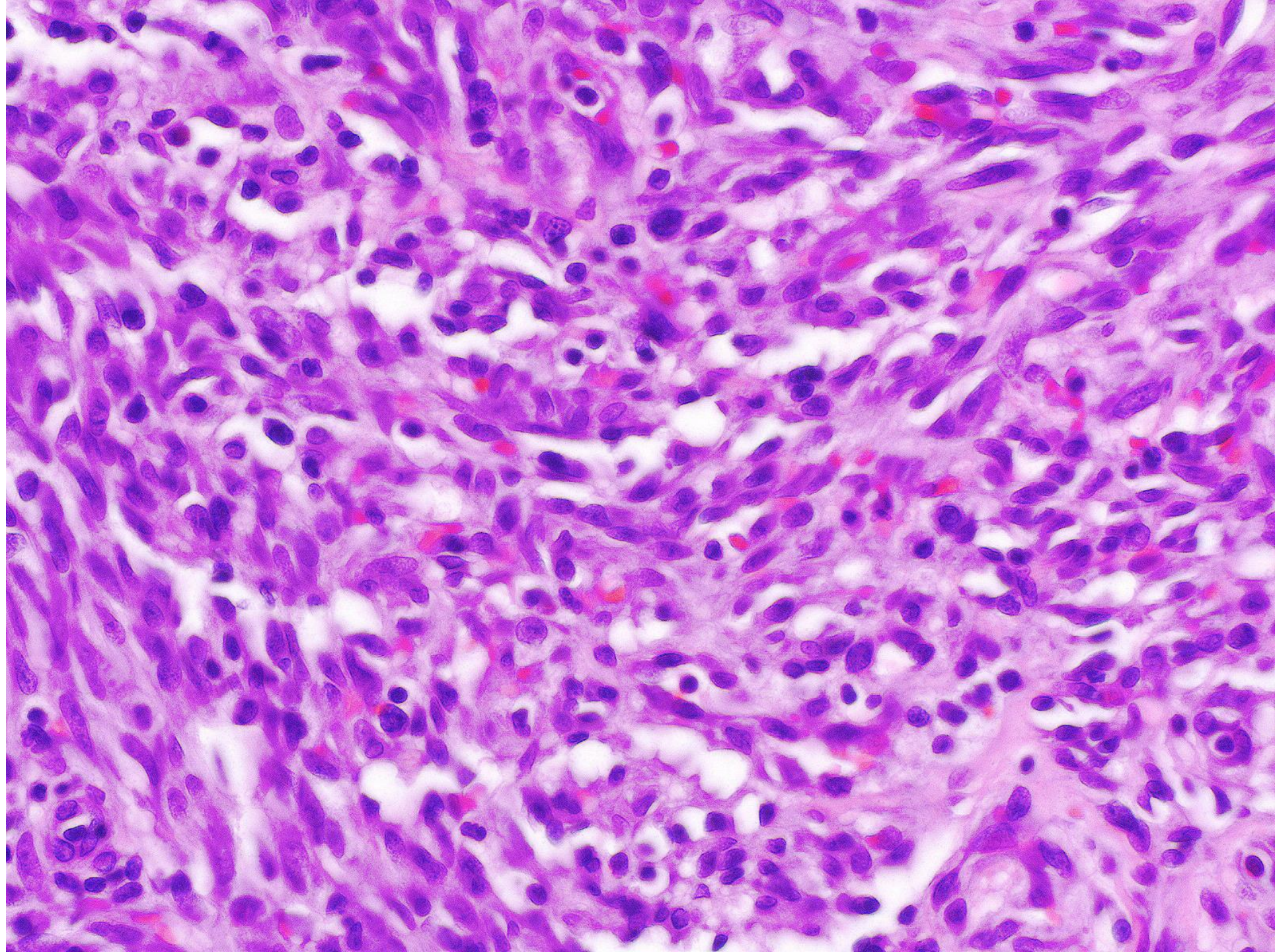
## Clinical features:

- young to middle-aged adults, associated with trauma
- Skin/subcutaneous tissues (especially fingers, head, and neck)
- May develop in deep veins, visceral organs, or pre-existing vascular malformations/hemangiomas
- Typically presents as a solitary, slow-growing, painless mass
- May mimic a thrombus, hemangioma, or malignancy clinically.











Case 83. 22M with numerous red papules, GI bleeding, and a single mass on left shin. What is your diagnosis?

---

A. Kaposi's sarcoma, nodular stage

---

B. Intravascular papillary endothelial hyperplasia (Masson tumor)

---

C. Non-involuting congenital hemangioma

---

D. Spindle cell hemangioma

---

E. Myopericytoma

---



Case 83. 22M with numerous red papules, GI bleeding, and a single mass on left shin. What is your diagnosis?

A. Kaposi's sarcoma, nodular stage

B. Intravascular papillary endothelial hyperplasia (Masson tumor)

C. Non-involuting congenital hemangioma

D. Spindle cell hemangioma

E. Myopericytoma

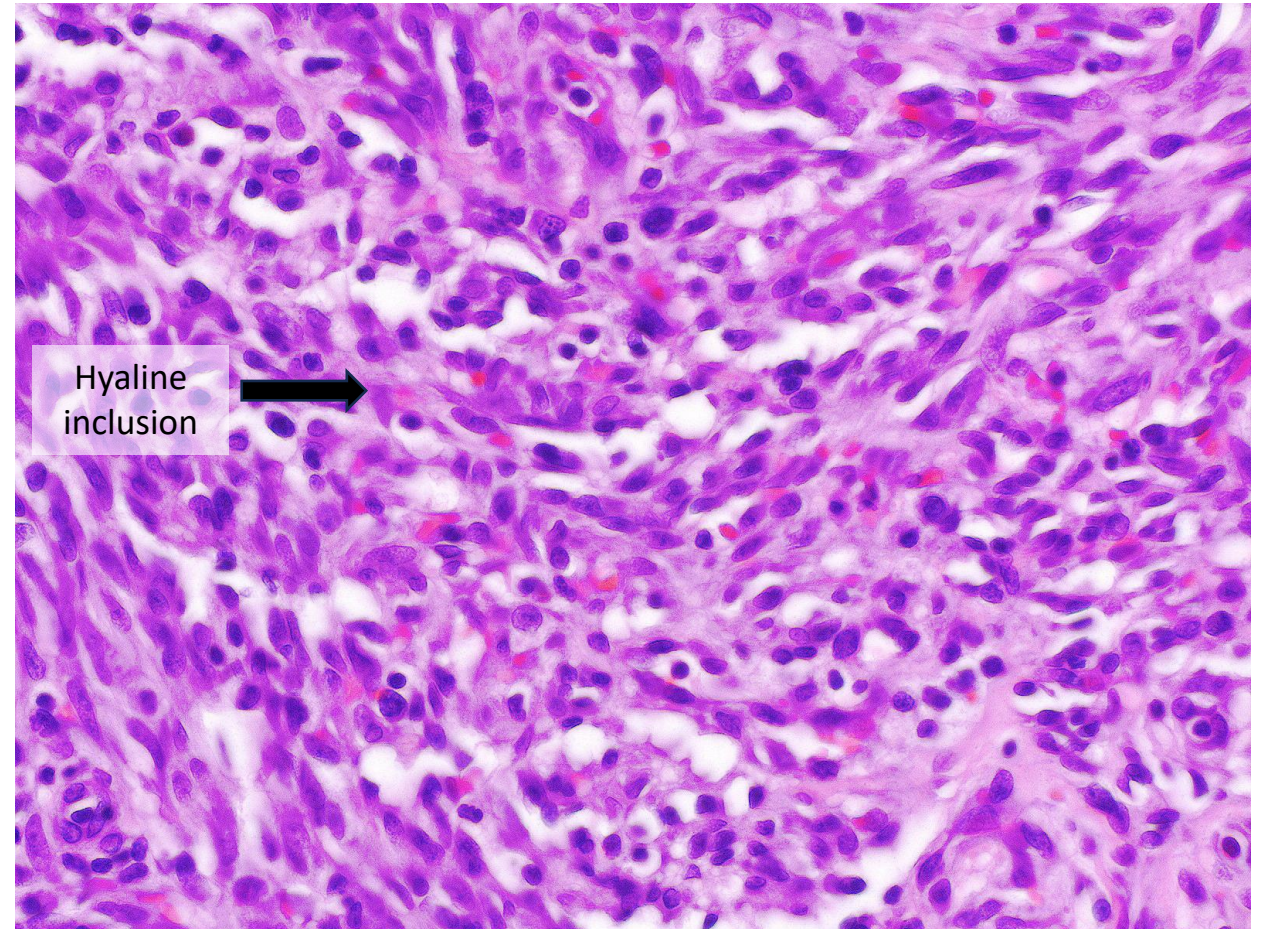


# KAPOSI SARCOMA

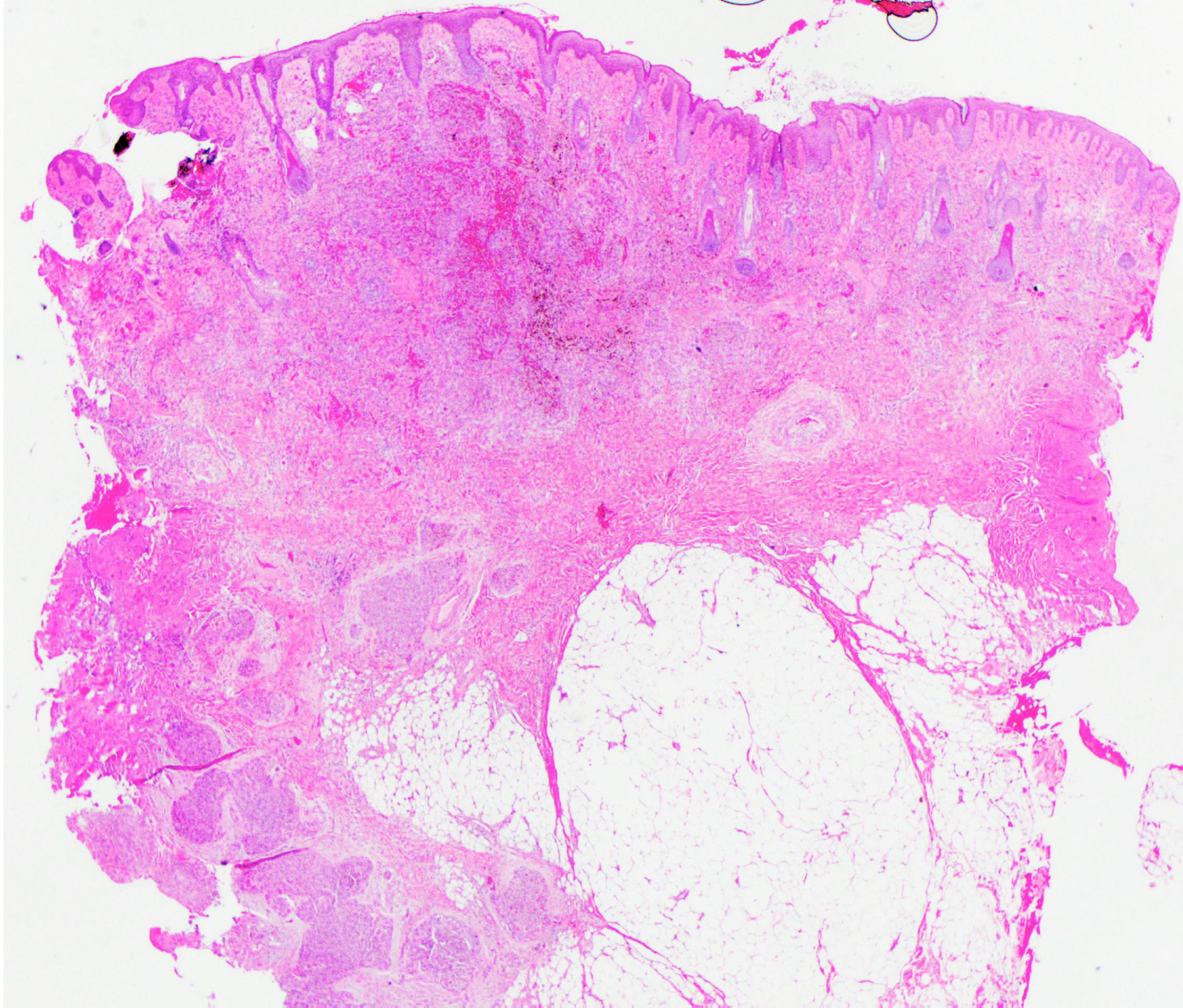
## Histologic features

Nodular stage:

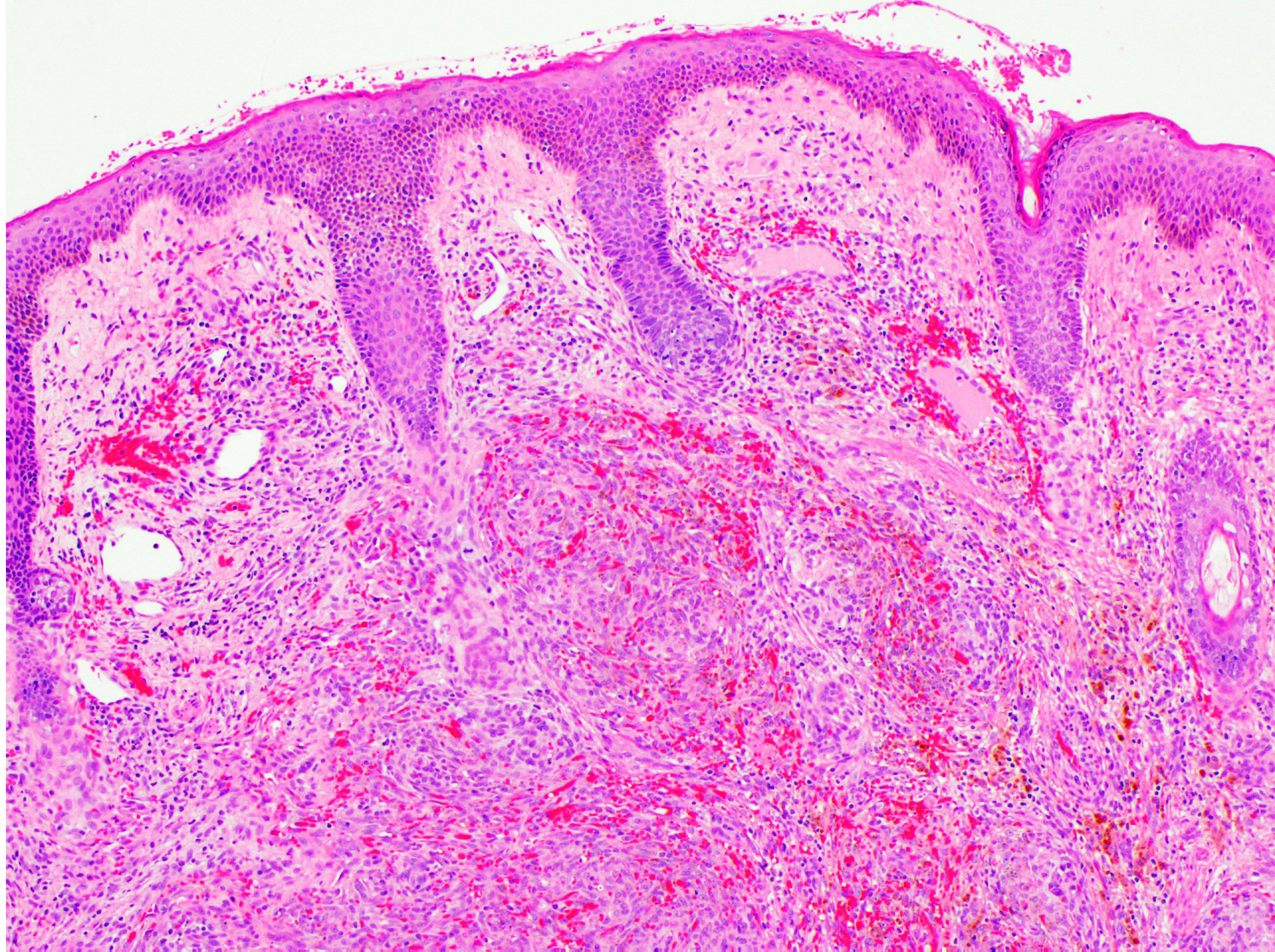
- Ill-defined dermal nodule
- Slit-like lumina
- Extravasated erythrocytes
- Numerous spindle cells with eosinophilic cytoplasm
- Scattered lymphocytes
- Extravasated erythrocytes
- Hyaline inclusion



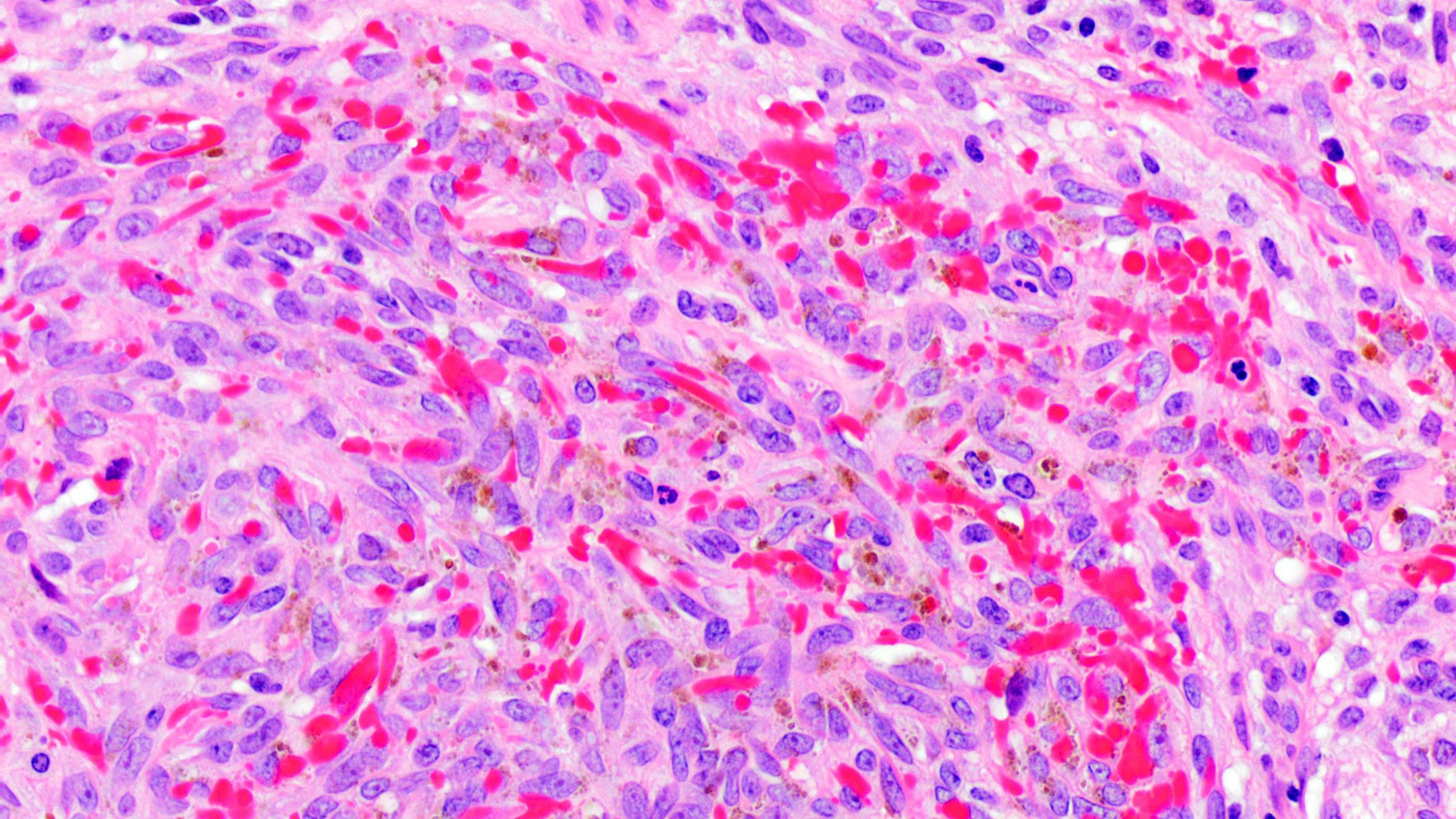














Case 84. 6-year-old girl with red-blue plaque on the face. What is your diagnosis?

---

A. Kaposi's sarcoma

---

B. Intravascular endothelial hyperplasia (Masson tumor)

---

C. Non-involuting congenital hemangioma

---

D. Spindle cell hemangioma

---

E. Kaposiform hemangioendothelioma

---



Case 84. 6-year-old girl with red-blue plaque on the face. What is your diagnosis?

A. Kaposi's sarcoma

B. Intravascular endothelial hyperplasia (Masson tumor)

C. Non-involuting congenital hemangioma

D. Spindle cell hemangioma

E. Kaposiform hemangioendothelioma



# KAPOSIFORM HEMANGIOENDOTHELIOMA

## Histologic features

Irregular, lobulated nodules infiltrating surrounding soft tissues

Sheets or fascicles of spindle-shaped endothelial cells (resembling Kaposi sarcoma)

Glomeruloid structures (rounded nests of tightly packed capillaries)

Differential Diagnosis:

Kaposi Sarcoma (HHV-8 positive, more common in adults, lacks glomeruloid structures).

Tufted Angioma (more organized "cannonball" clusters of capillaries).

Infantile Hemangioma (GLUT1 positive, well-circumscribed, regressing phase).

Angiosarcoma (more nuclear atypia, high mitotic activity).

## Immunohistochemistry (IHC)

Positive for:

CD31, CD34 (vascular endothelial markers)

FLI-1 (nuclear staining, endothelial marker)

PROX1 (highlights spindle cells)

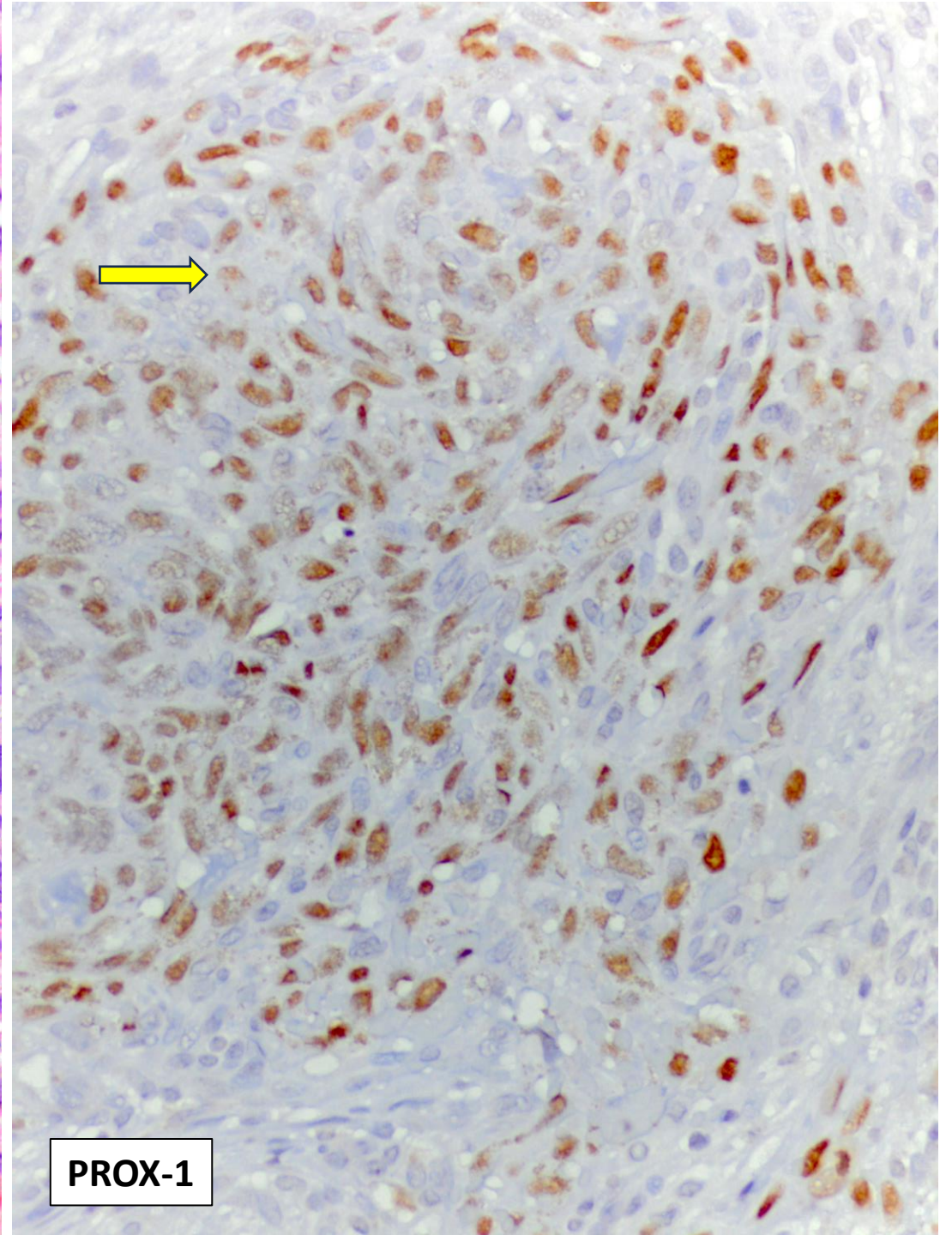
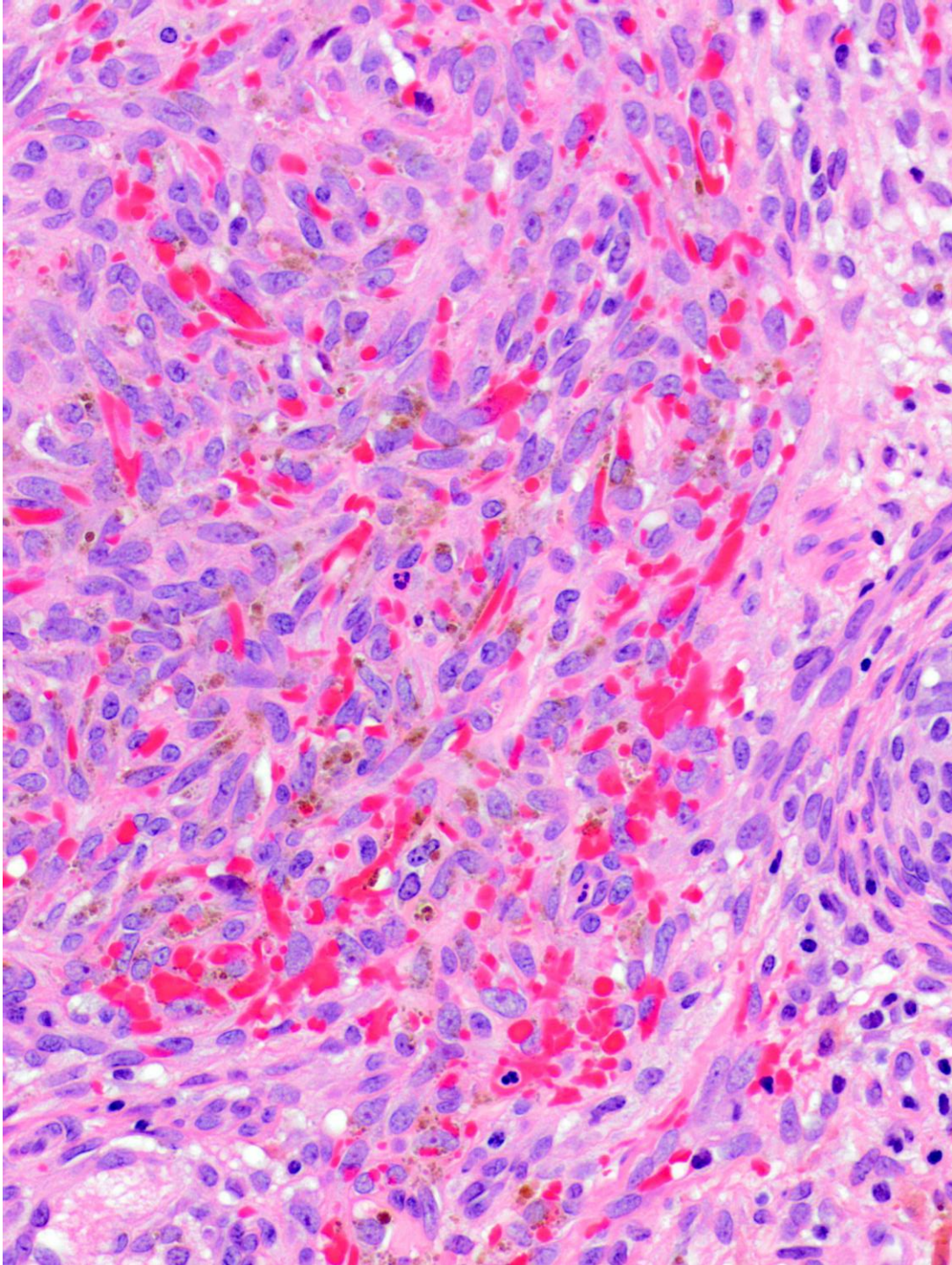
D2-40 (podoplanin, highlights spindle cells)

Negative for:

HHV-8 (helps differentiate from Kaposi sarcoma)

GLUT1 (negative, unlike infantile hemangioma)

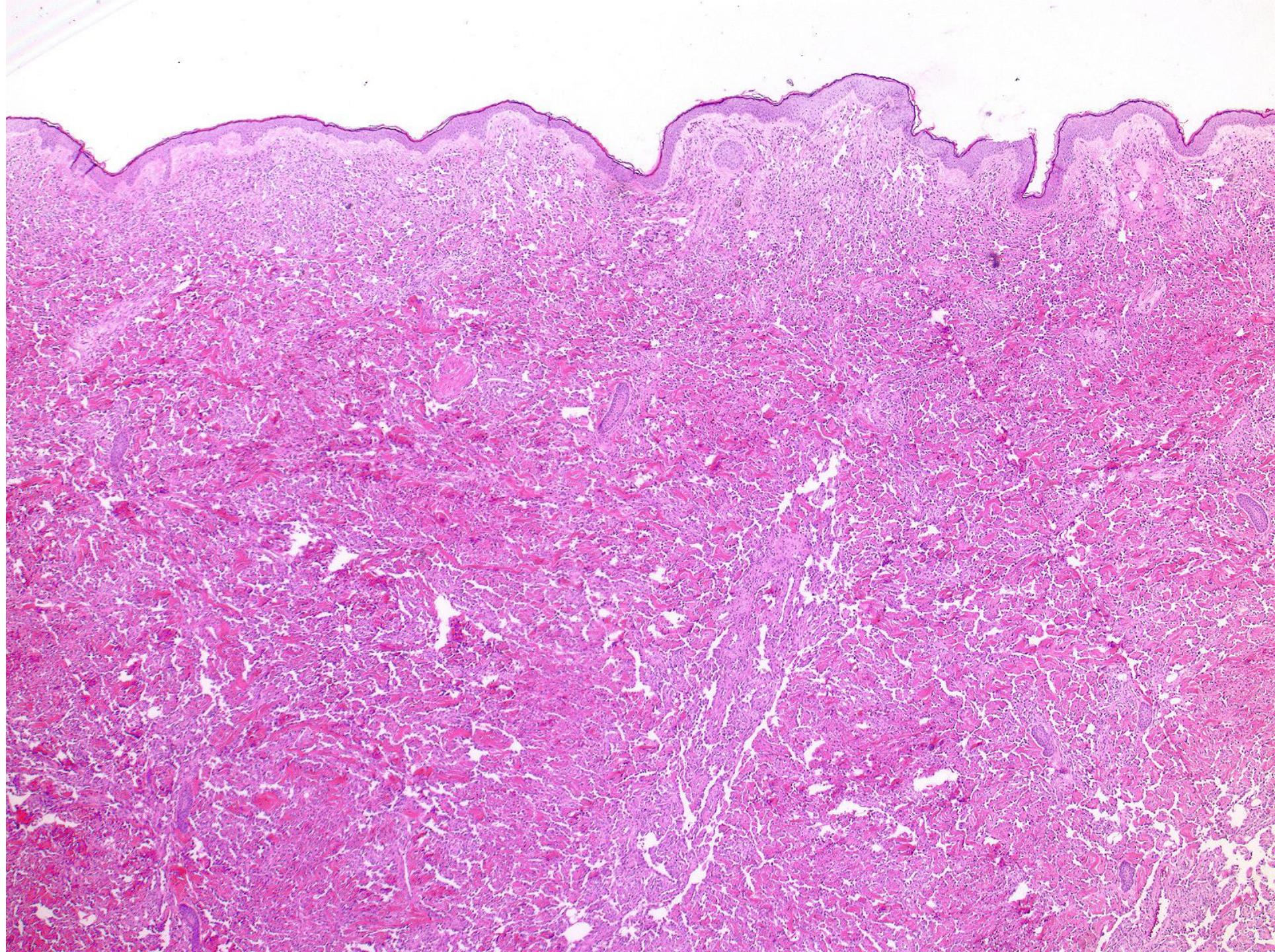




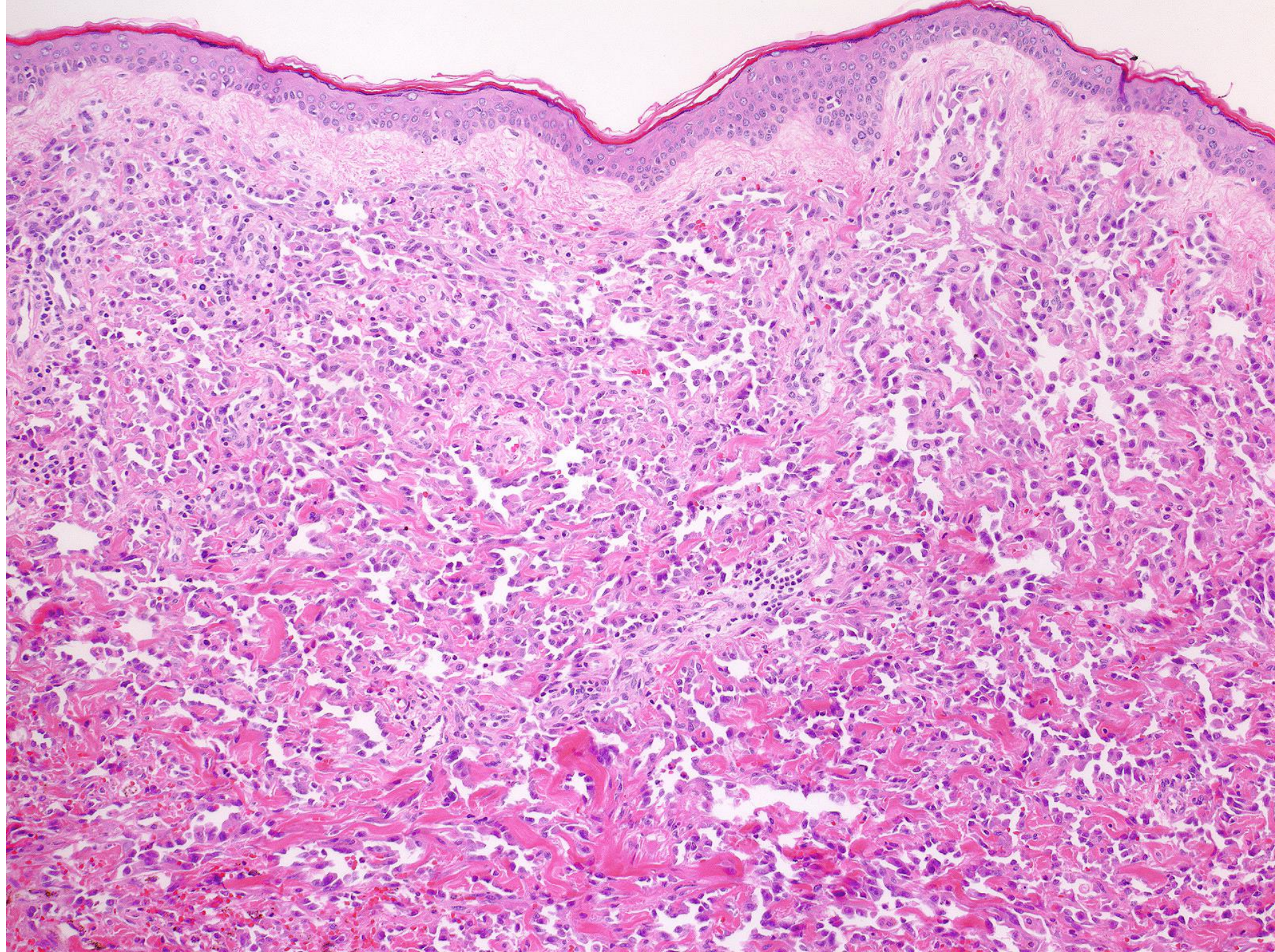
PROX-1

PROX-1-negative glomeruloid foci

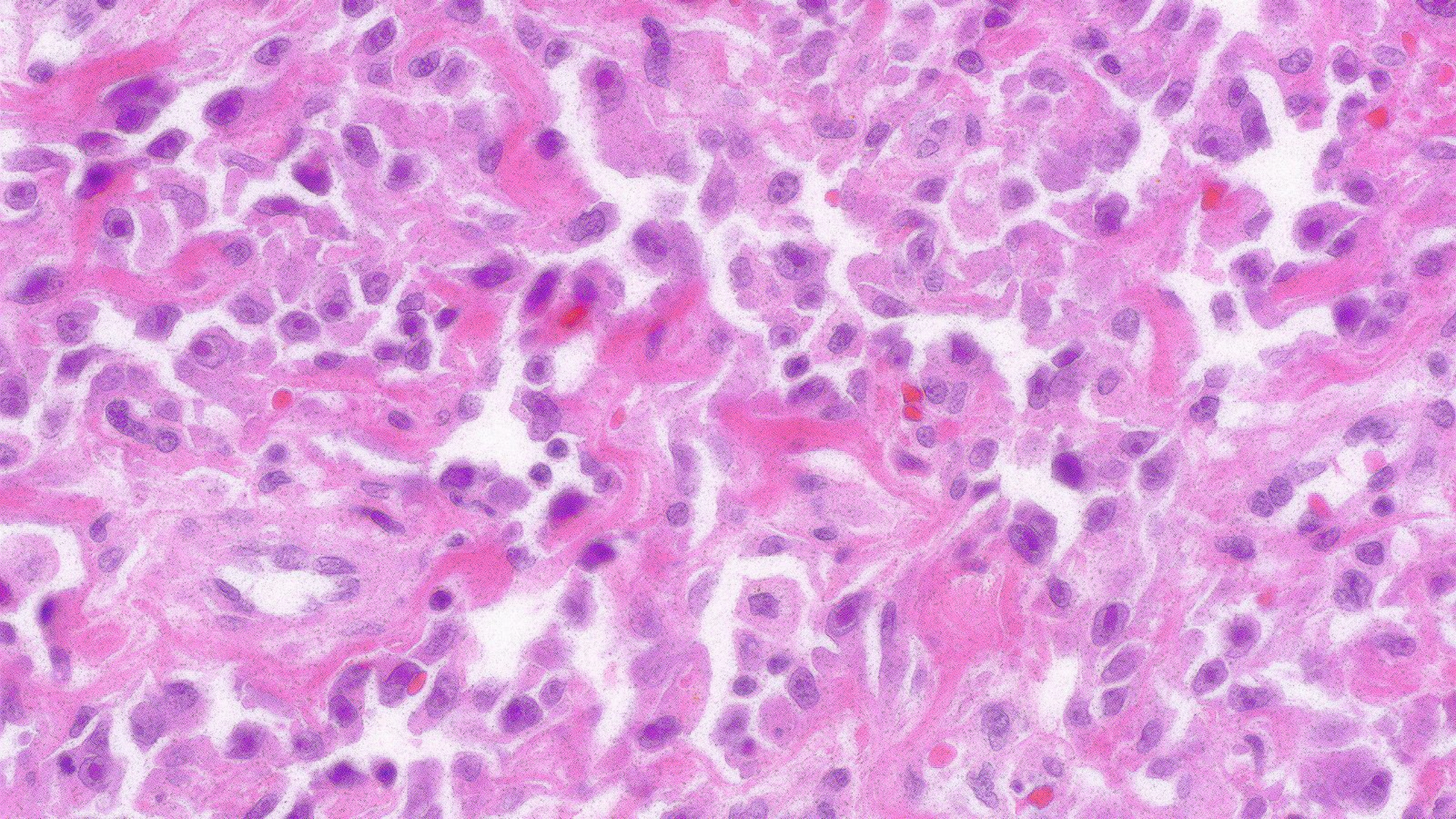














Case 85. 67M with a large red-blue, plaque on back. What is your diagnosis?

A. Kaposi's sarcoma

B. Intravascular endothelial hyperplasia (Masson tumor)

C. Angiosarcoma

D. Spindle cell hemangioma

E. Myopericytoma



Case 85. 67M with a large red-blue, plaque on back. What is your diagnosis?

A. Kaposi's sarcoma

B. Intravascular endothelial hyperplasia (Masson tumor)

C. Angiosarcoma

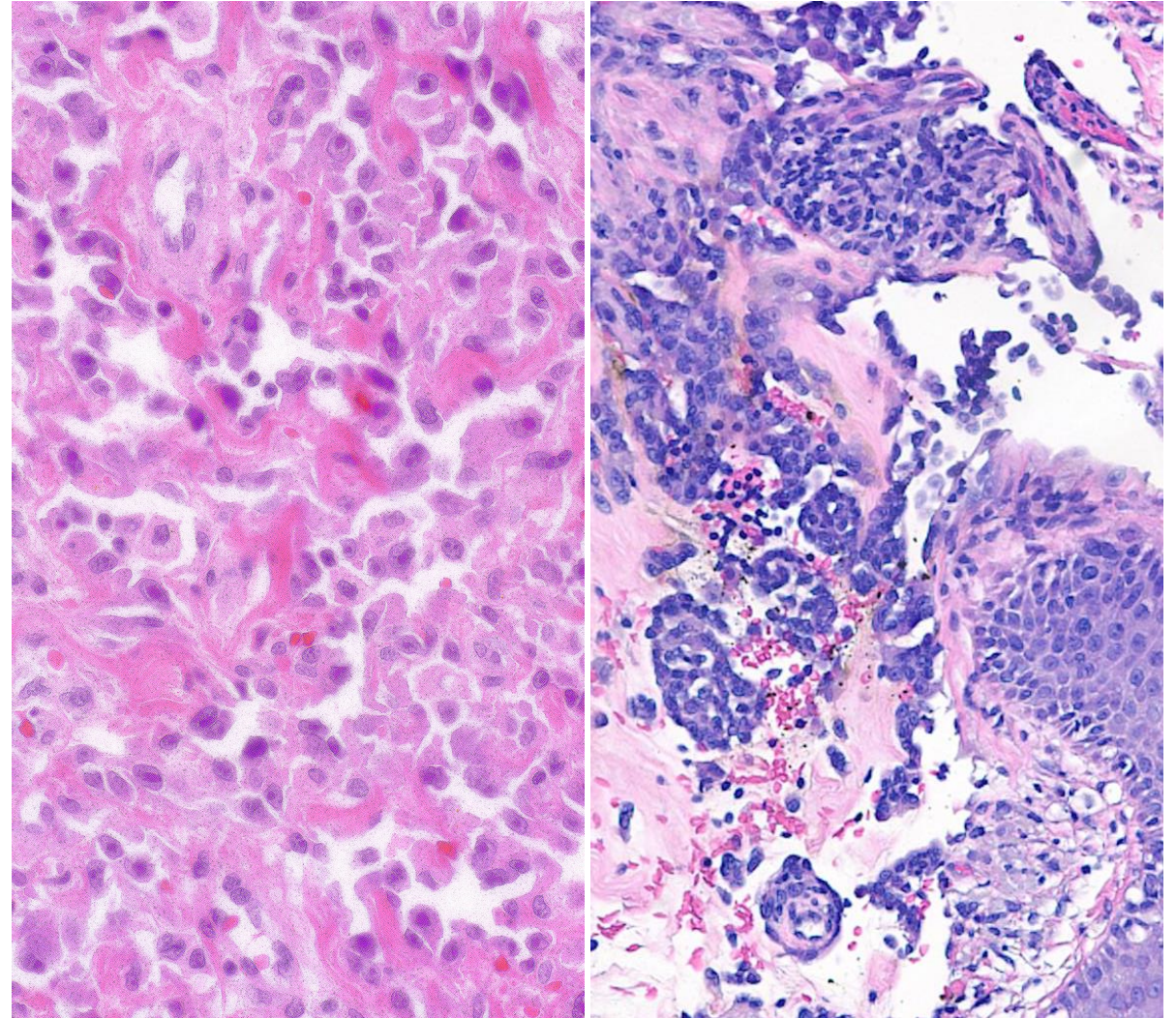
D. Spindle cell hemangioma

E. Myopericytoma

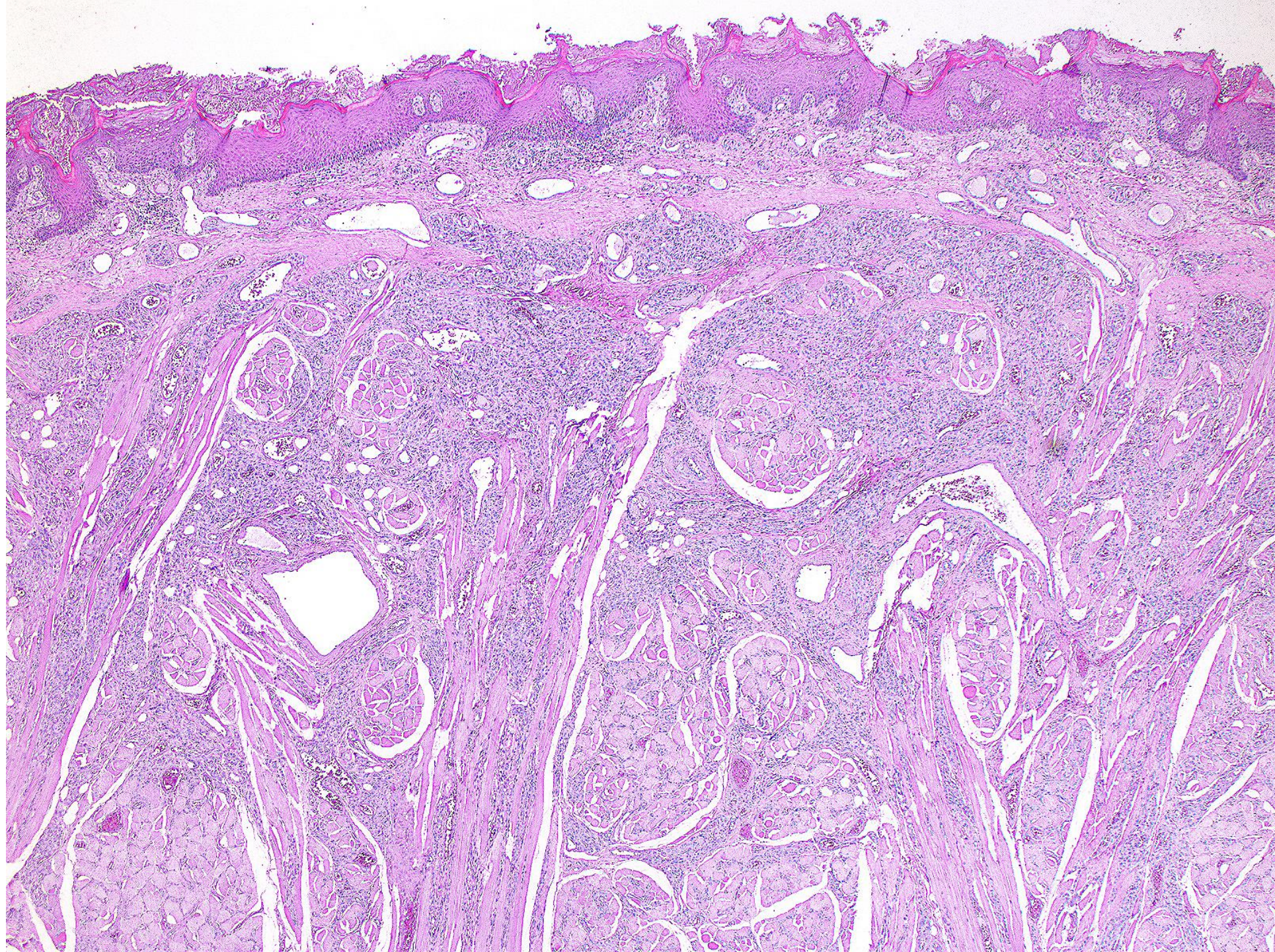


# ANGIOSARCOMA

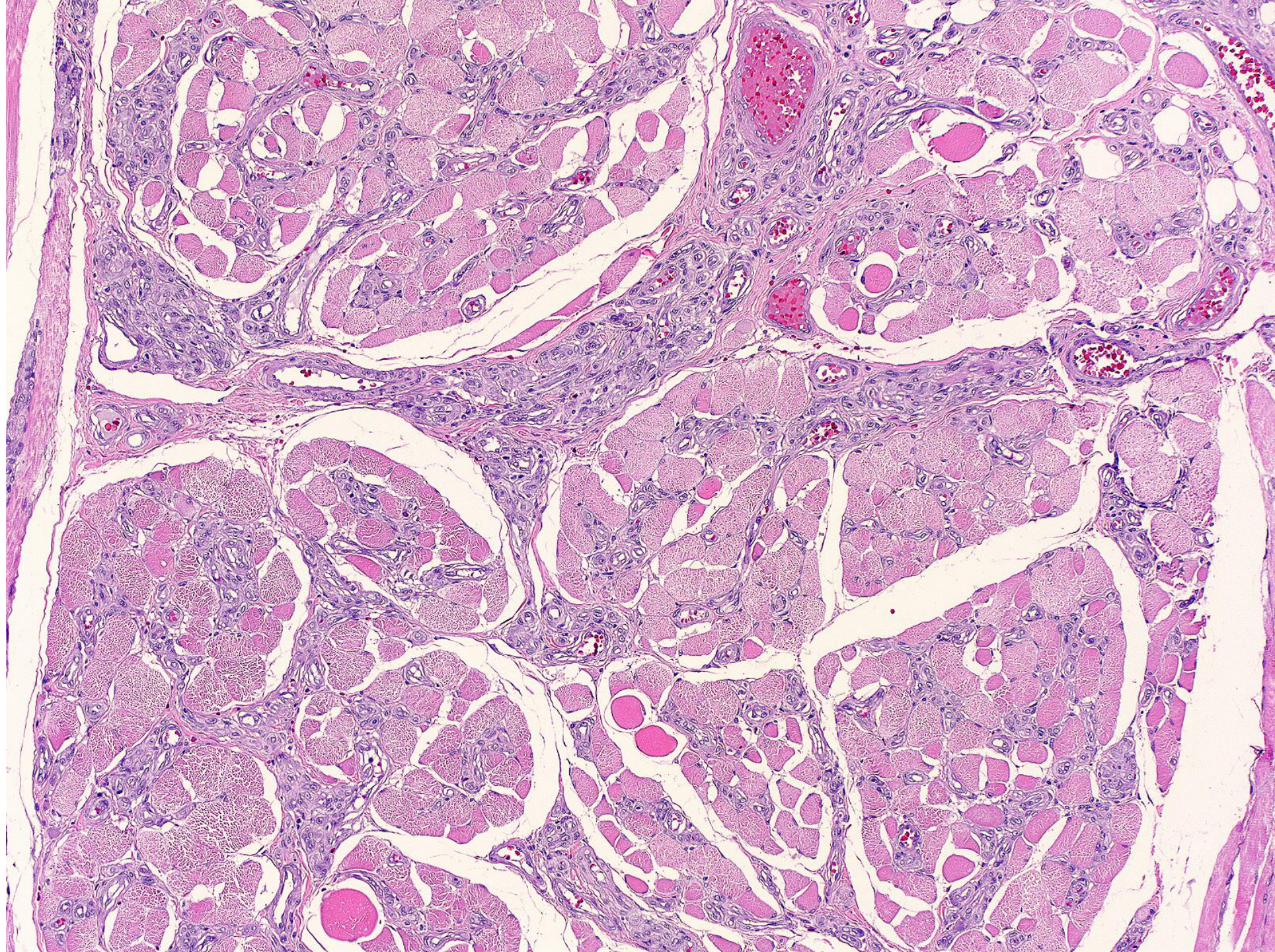
- Ill-defined, infiltrating dermal mass
- Destructive pattern, loss of adnexa
- Vascular channels of varying caliber
- Dissecting vascular collagen bundles
- Vascular space lined by multilayered endothelia
- Plump, pleomorphic endothelia, forming papillae
- Ample cytoplasm, round nuclei, open chromatin, prominent nucleoli
- Mitoses



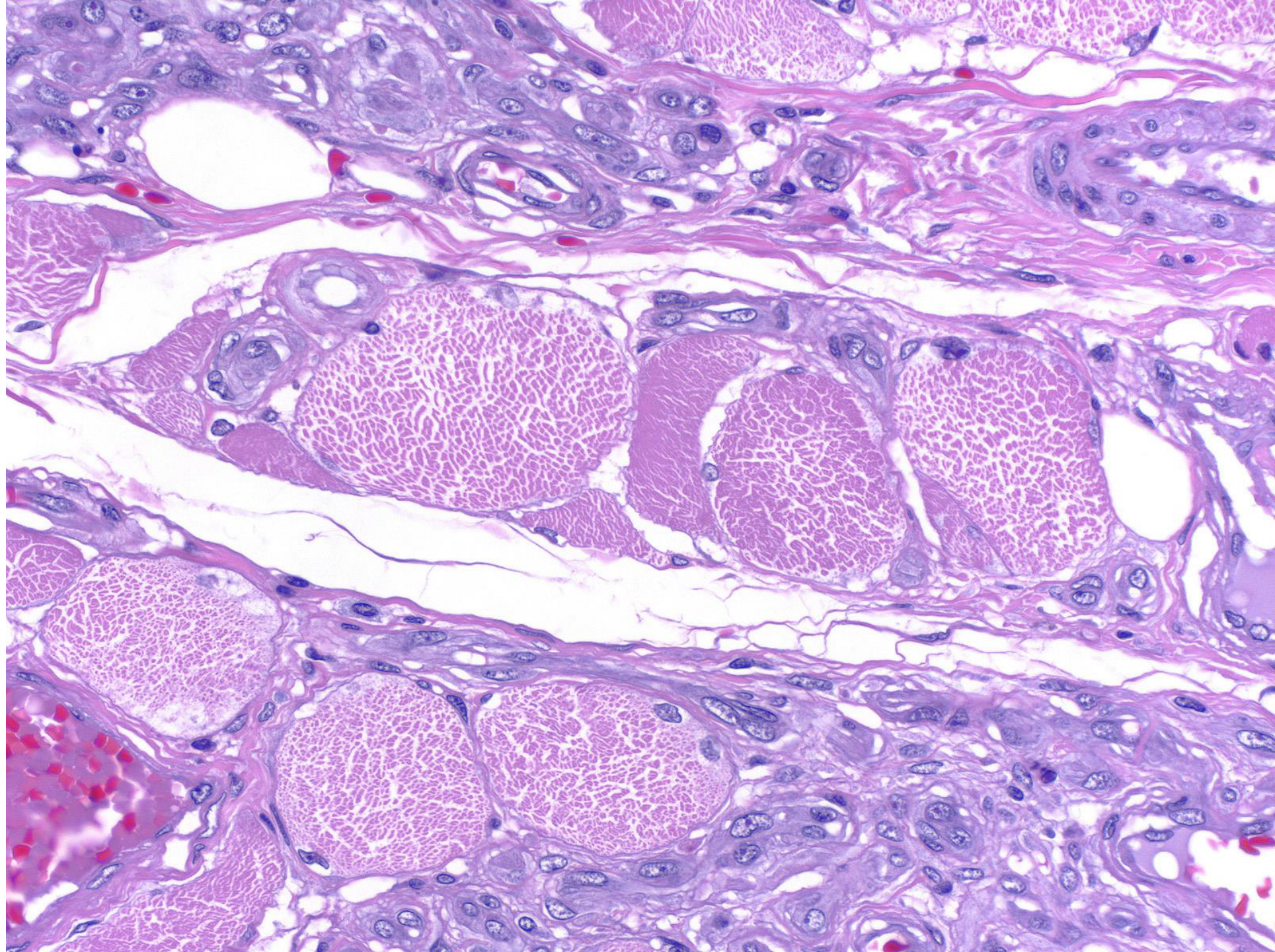














Case 86. 27F with a single large, slow growing soft tissue mass, left calf. What is your diagnosis?

A. Kaposi's sarcoma

B. Intravascular endothelial hyperplasia (Masson tumor)

C. Non-involuting congenital hemangioma

D. Infantile hemangioma

E. Intramuscular vascular malformation (capillary intramuscular hemangioma)



Case 86. 27F with a single large, slow growing soft tissue mass, left calf. What is your diagnosis?

A. Kaposi's sarcoma

B. Intravascular endothelial hyperplasia (Masson tumor)

C. Non-involuting congenital hemangioma

D. Infantile hemangioma

E. Intramuscular vascular malformation (capillary intramuscular hemangioma)



# INTRAMUSCULAR VASCULAR MALFORMATION (CAPILLARY INTRAMUSCULAR HEMANGIOMA)

## Key diagnostic points

- Maybe associated with thrombocytopenia
- Unclassified vascular anomaly (malformation)
- Lobules of tightly packed capillaries
- Infiltrating bundles of skeletal muscle
- Intact, not destroyed bundles of skeletal muscle
- Fibrous septae separate tumor lobules
- GLUT-1-

