

ADS Dermatopathology Meeting 2014 -
Selected Spindle Cell Tumors

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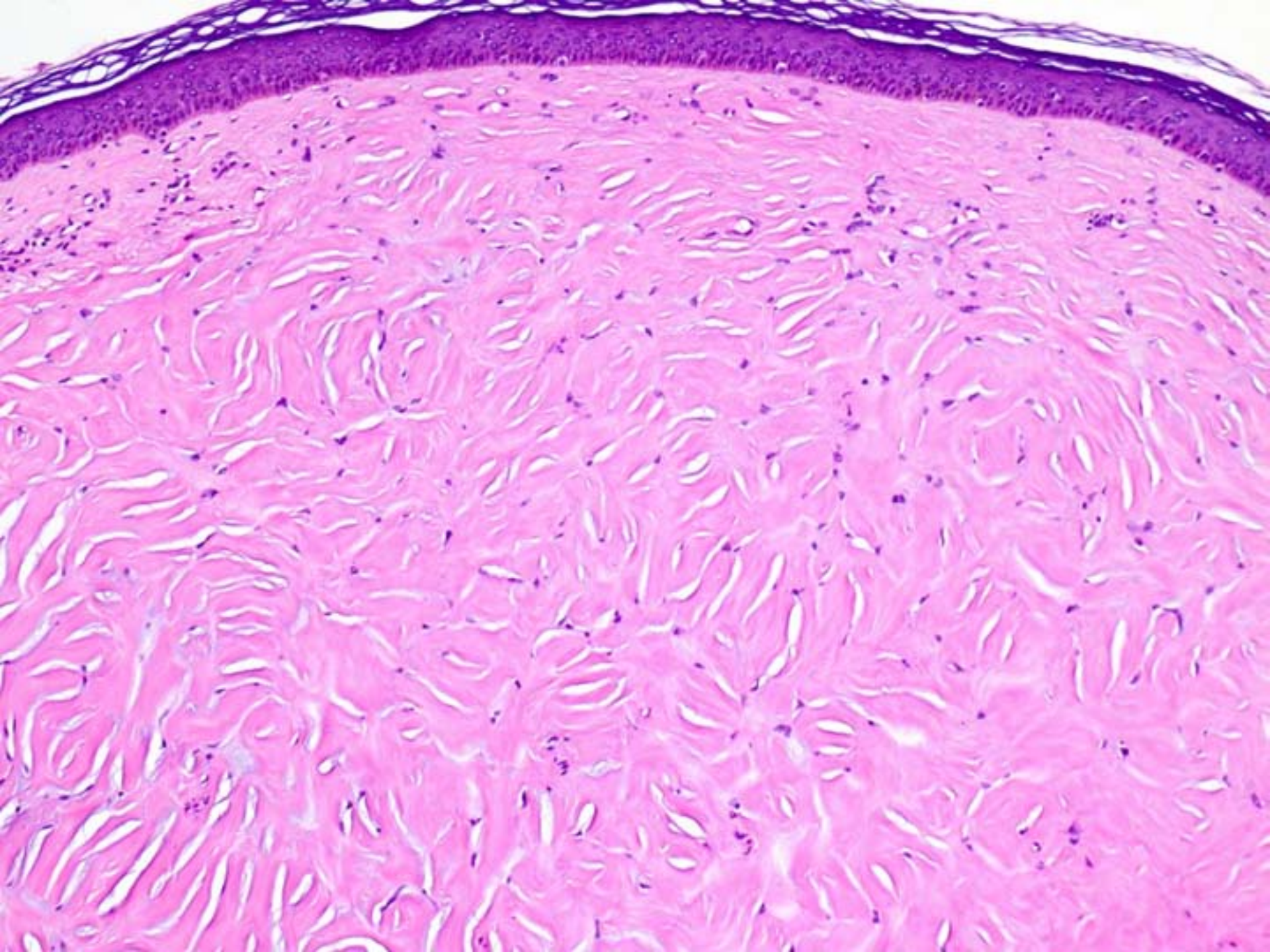
August 2014
Perth, Australia

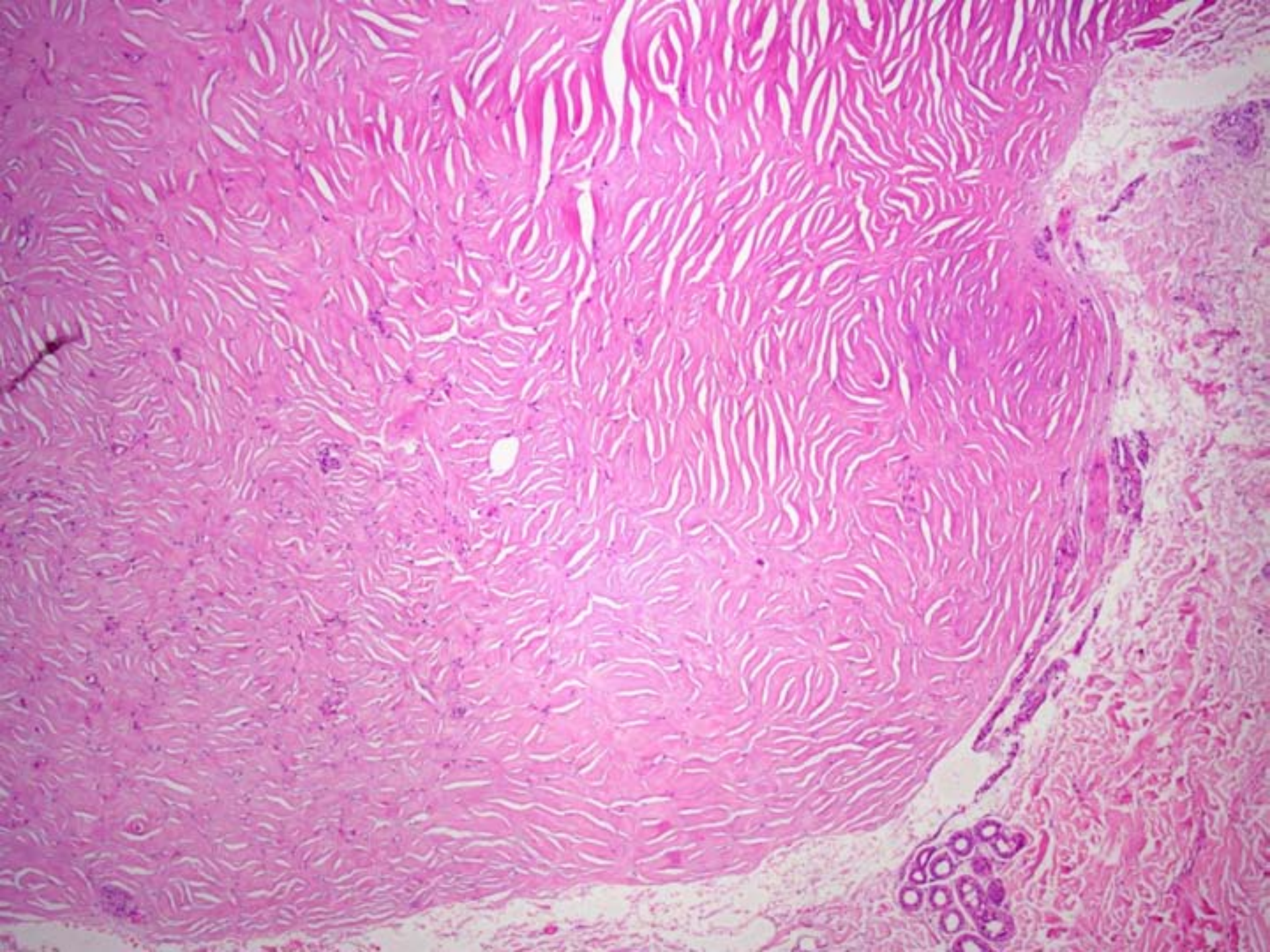
STORIFORM COLLAGENOMA

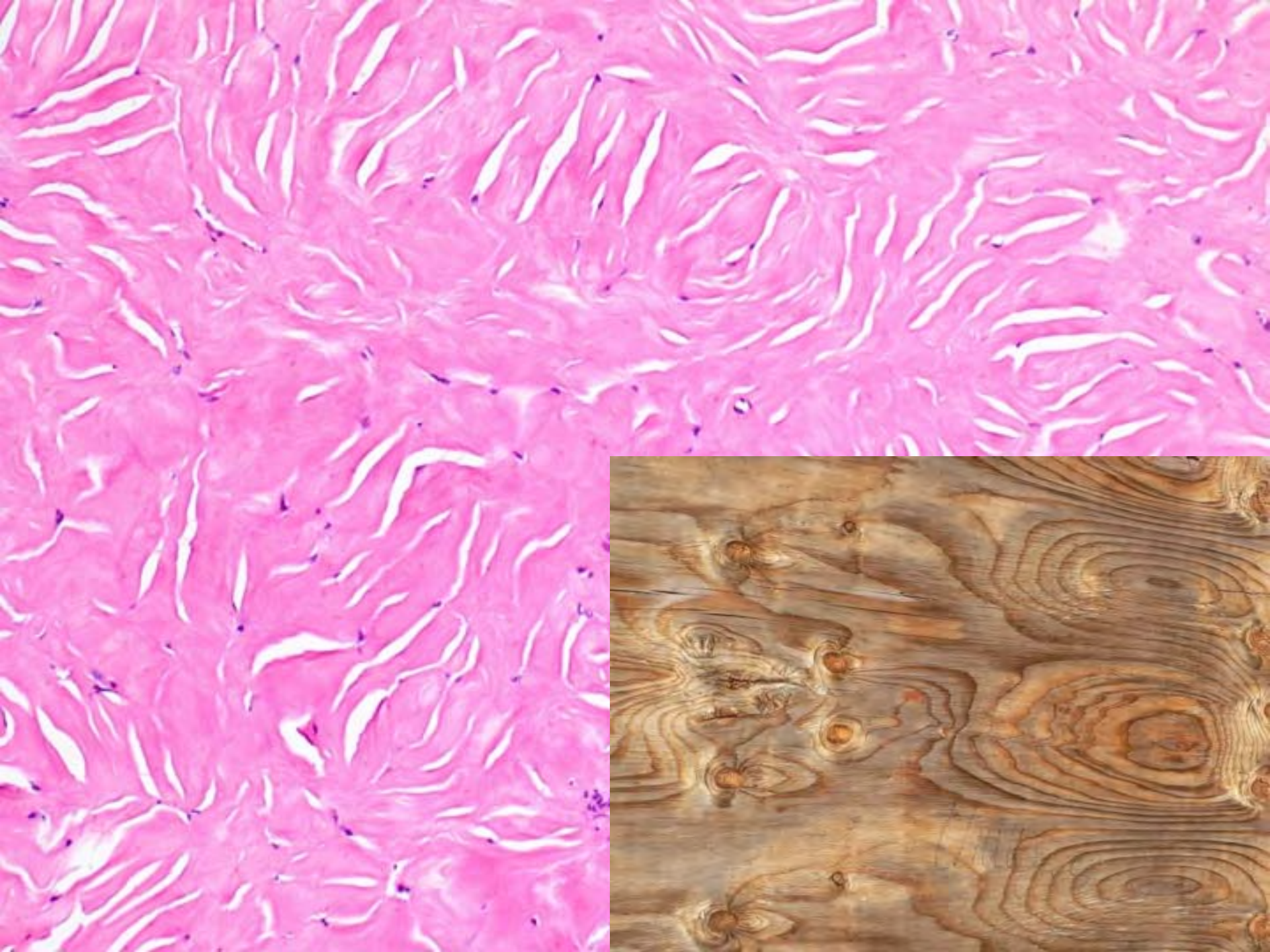
(Sclerotic Fibroma, Plywood Fibroma)

- Cutaneous & sometimes mucosal papules
- Single or multifocal (with Cowden's disease)
- Well-circumscribed, unencapsulated thickened and sclerotic collagen bundles
 - whorled and “plywood-like” pattern
 - paucicellular with few spindled fibroblasts and some accumulation of mucin
- May represent a final common endpoint









Sclerotic fibroma-like change in various neoplastic and inflammatory skin lesions: is sclerotic fibroma a distinct entity?

Abstract: Sclerotic fibroma was first described in association with Cowden's disease by Weary et al. in 1972. In 1989, Rapini and Golitz detailed 11 cases of solitary sclerotic fibroma (SFS) in the absence of Cowden's disease, suggesting the term SFS of the skin. Classic histological features include hypocellular, hyalinized bands of collagen sharply demarcated from the surrounding skin. Numerous authors have described sclerotic fibroma-like changes in other entities including melanocytic nevi, dermatofibromas, lipomas, tendon sheath fibromas, giant cell collagenomas, neurofibromas, angiofibromas, erythema elevatum diutinum, and folliculitis. Dissension has arisen, with some dermatopathologists asserting that sclerotic fibroma is just an evolutionary end-point of a previous lesion. Others contend that SFS is a distinct lesion and cite recurrent cases and/or proliferation marker studies to corroborate this view. We detail the histopathological findings of lesions consistent with the classic description of SFS and compare these to sclerotic changes observed in an intradermal nevus, blue nevus, erythema elevatum diutinum, neurofollicular hamartoma, angiofibroma, neurofibroma, accessory nipple, and dermatofibromas. Sclerotic fibroma-like change may be seen in a variety of lesions and may represent a common reaction pattern in the skin.

High WA, Stewart D, Essary LR, Kageyama NP, Hoang MP, Cockerell CJ. Sclerotic fibroma-like change in various neoplastic and inflammatory skin lesions: is sclerotic fibroma a distinct entity?

J Cutan Pathol 2004; 00: 1–6. © Blackwell Munksgaard 2004.

Whitney A. High¹, David Stewart¹, Lydia R. Essary¹, Nicole P. Kageyama¹, Mai P. Hoang², Clay J. Cockerell¹

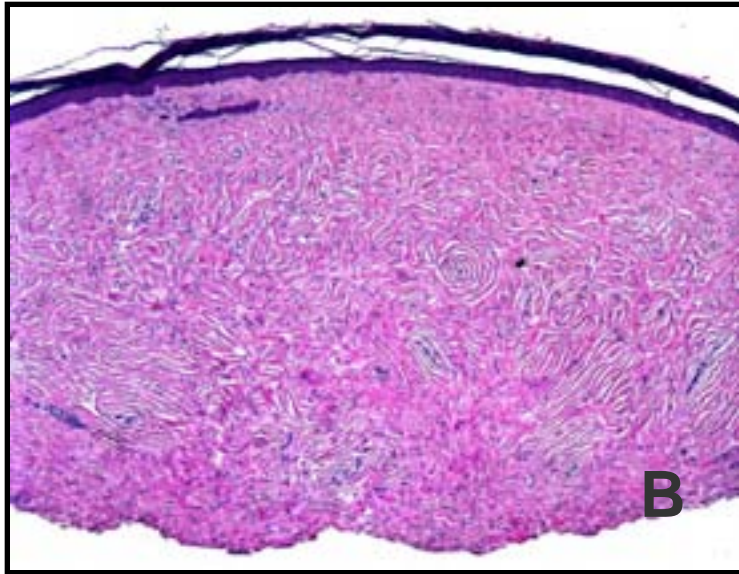
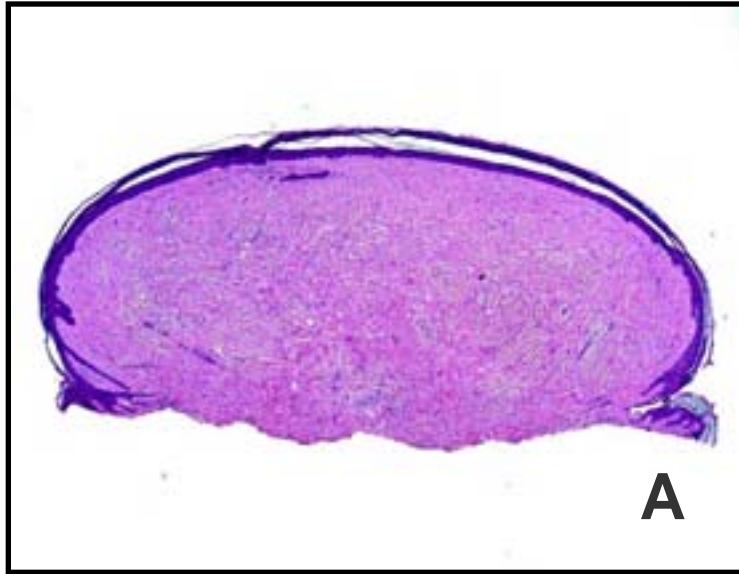
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SCLEROTIC FIBROMA



DERMATOFIBROMA

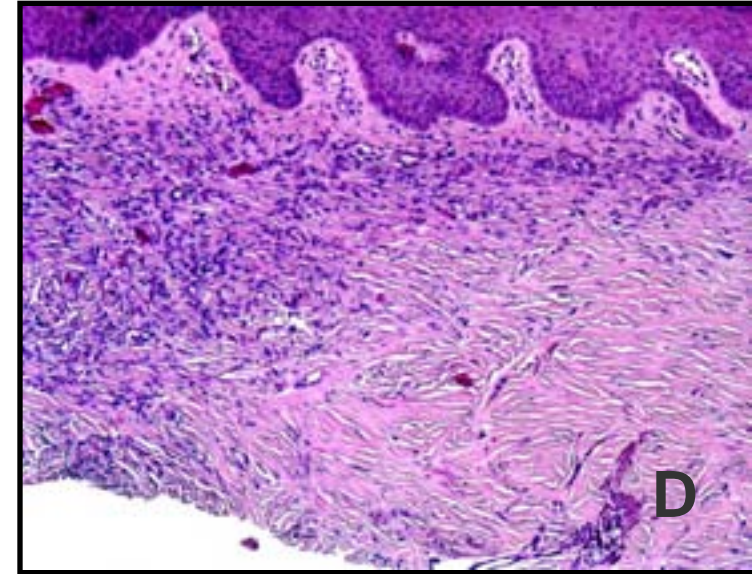
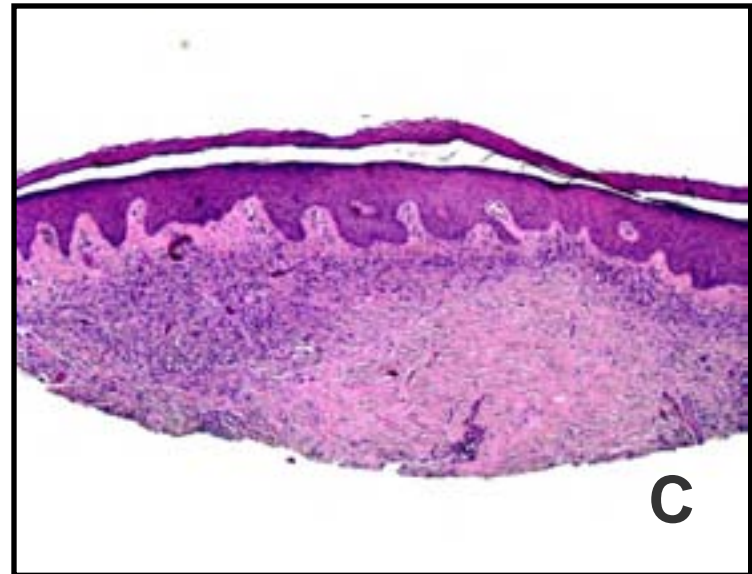


FIGURE 1 – Comparison of a classic sclerotic fibroma of the skin at low (A) and medium power (B) with a dermatofibroma with prominent central sclerotic change at low (C) and medium power (D).

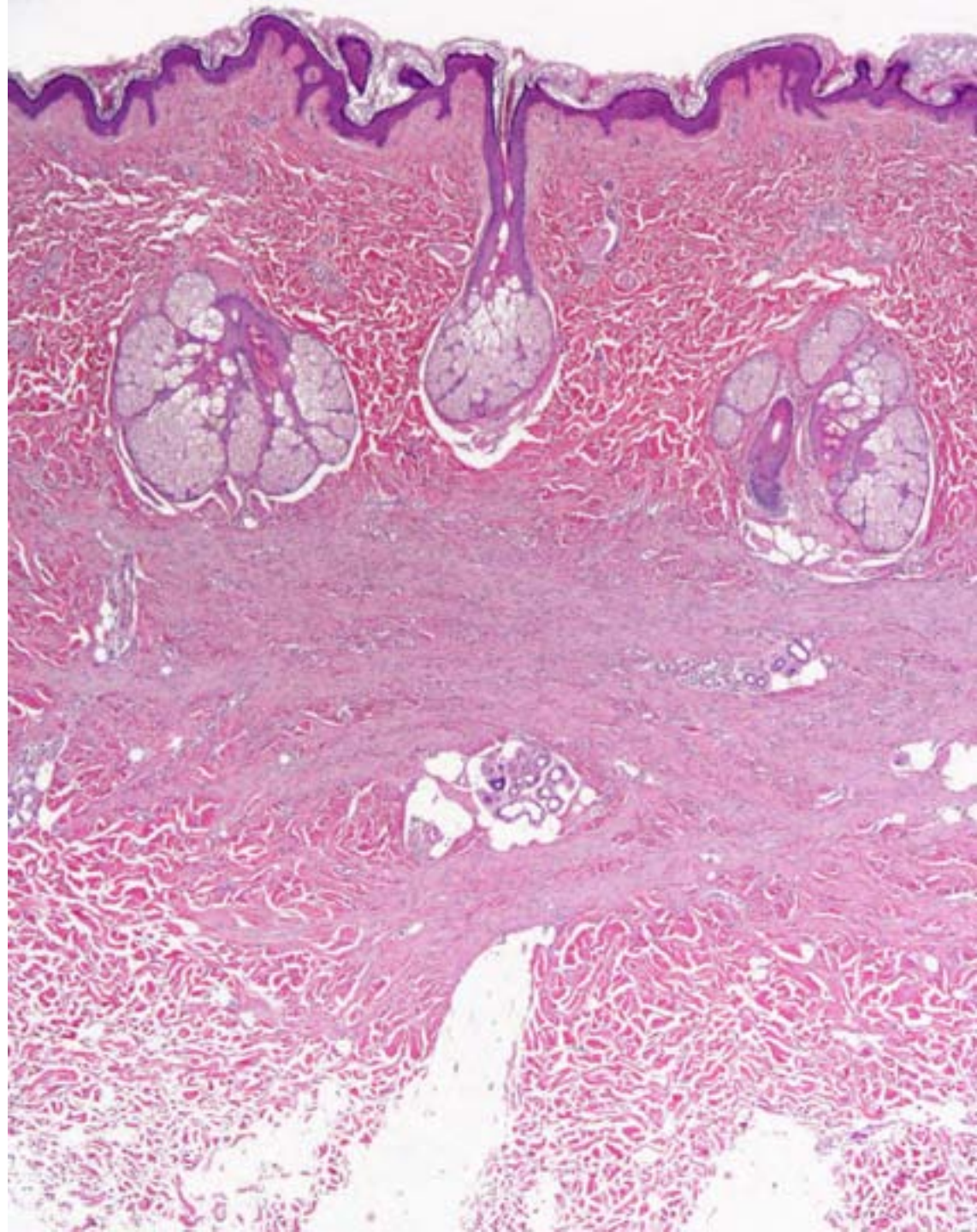
DERMATOMYOFIBROMA

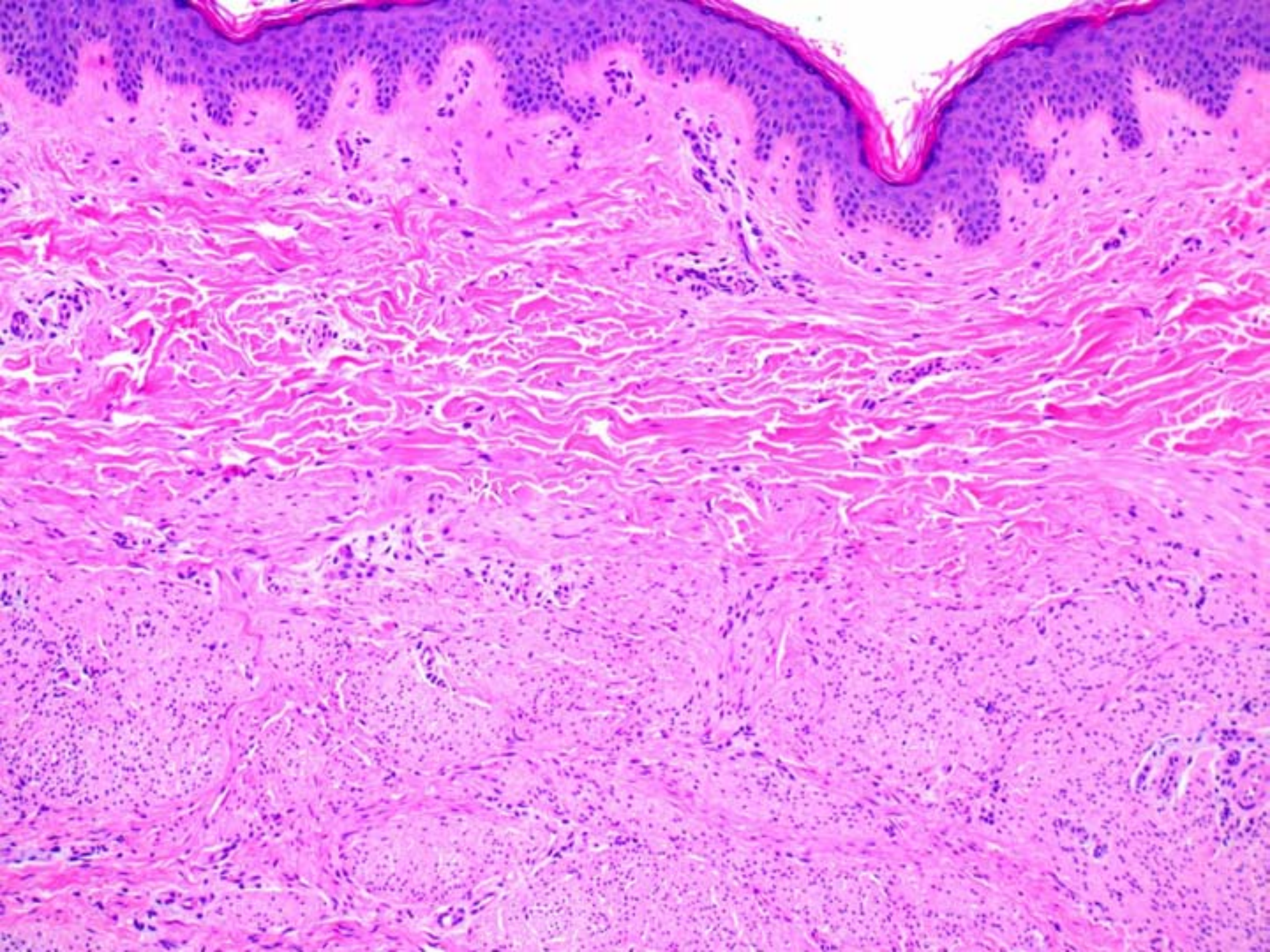
- Composed of fibroblasts/myofibroblasts
- Most common in adolescents & young adults, predilection for women
- Location: upper back, shoulder, axilla, upper arms, abdomen
- Primary lesion: asymptomatic, firm, skin-colored, tan/yellow plaque/nodule

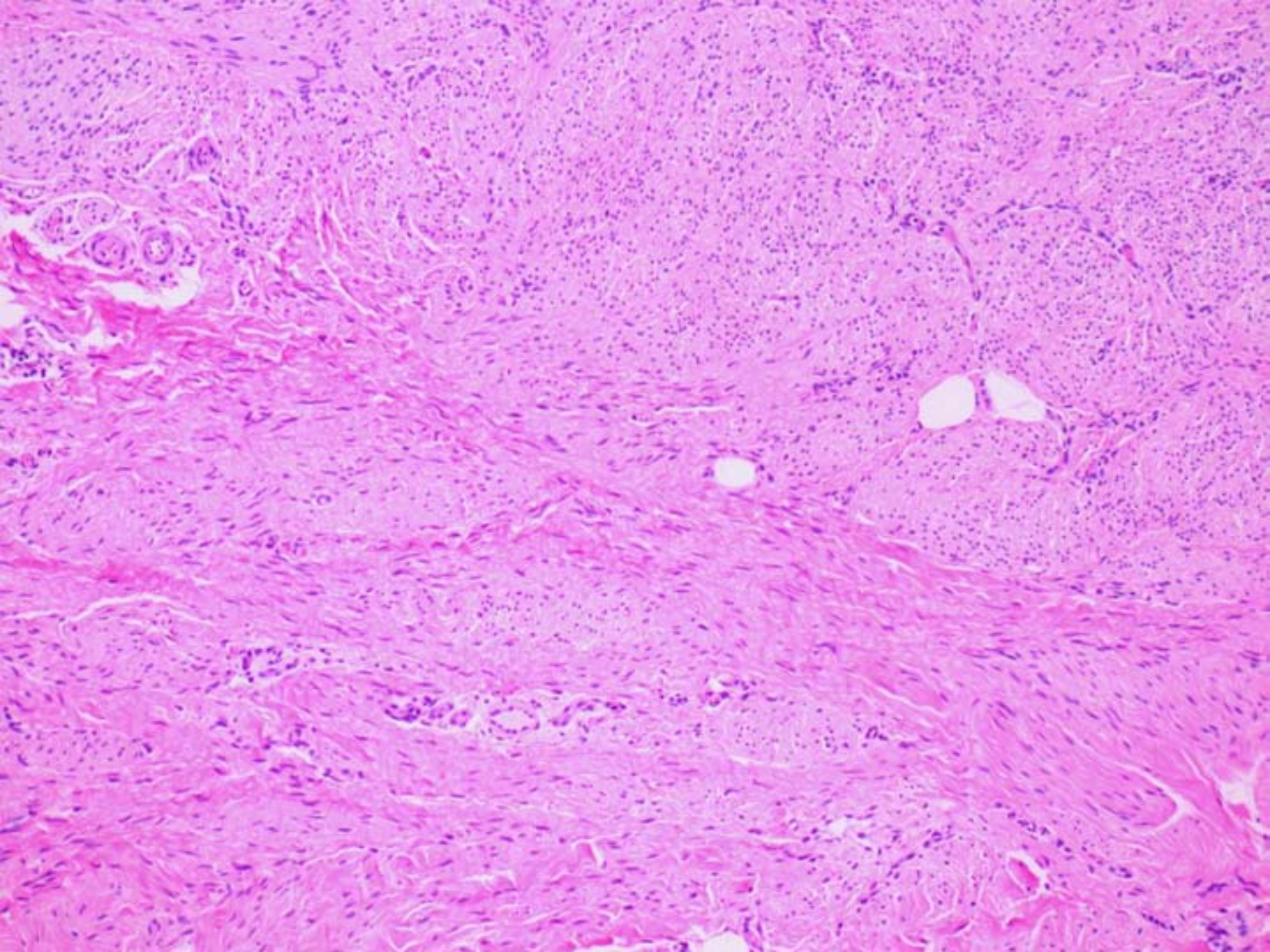
DERMATOMYOFIBROMA

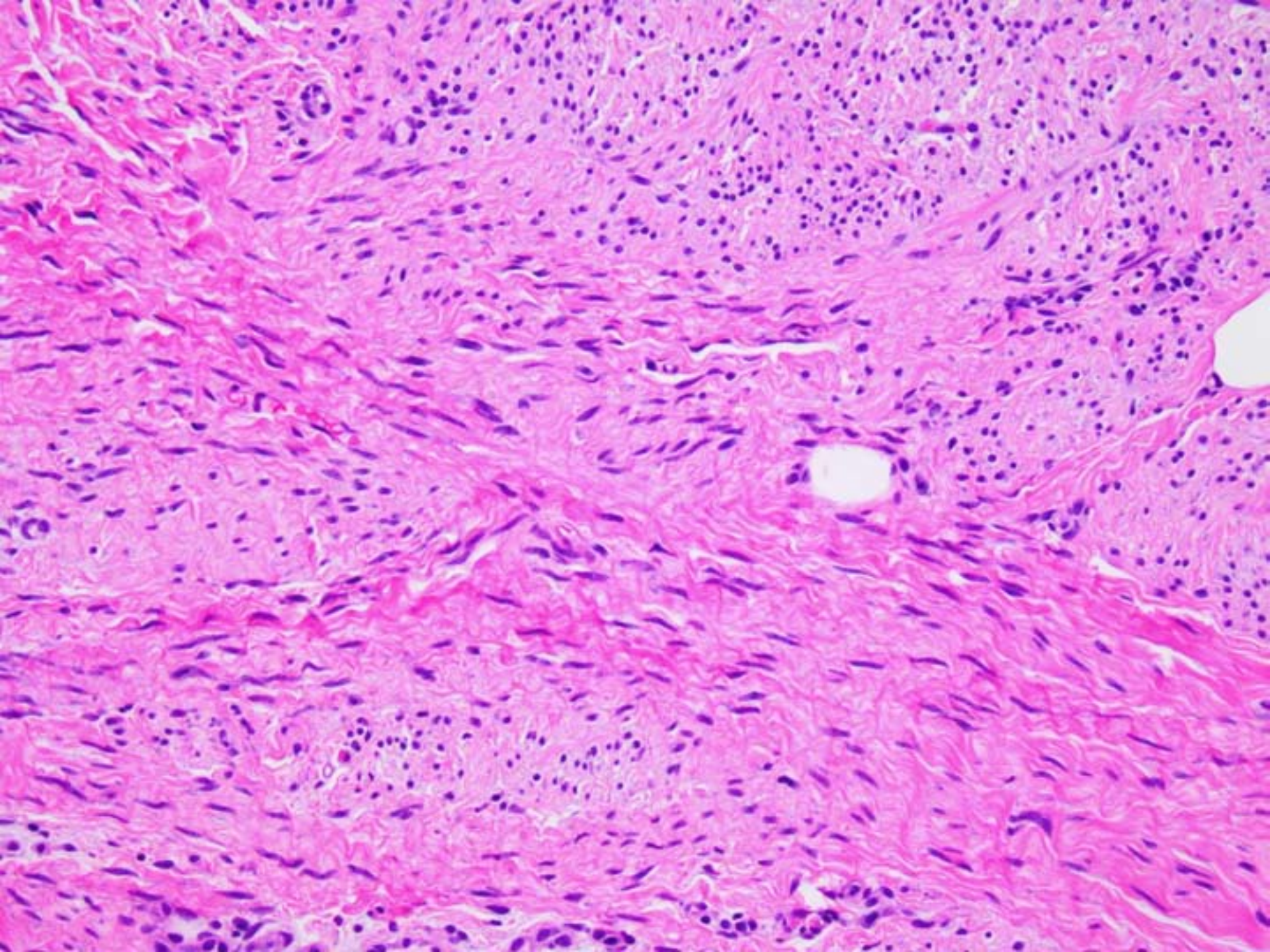
- Low Power:
 - well-circumscribed proliferation of spindle cells in reticular dermis, may involve fat
 - intersecting fascicles, resemble smooth muscle, oriented parallel to the epidermis
- High Power:
 - spindle cells with uniform nuclei, rounded/pointed ends but may be wavy
 - Embedded in a fine, collagenous stroma
- Immunohistochemistry
 - POSITIVE = vimentin, MSA, SMA (+/-)
 - NEGATIVE = desmin, factor XIIIa, S100

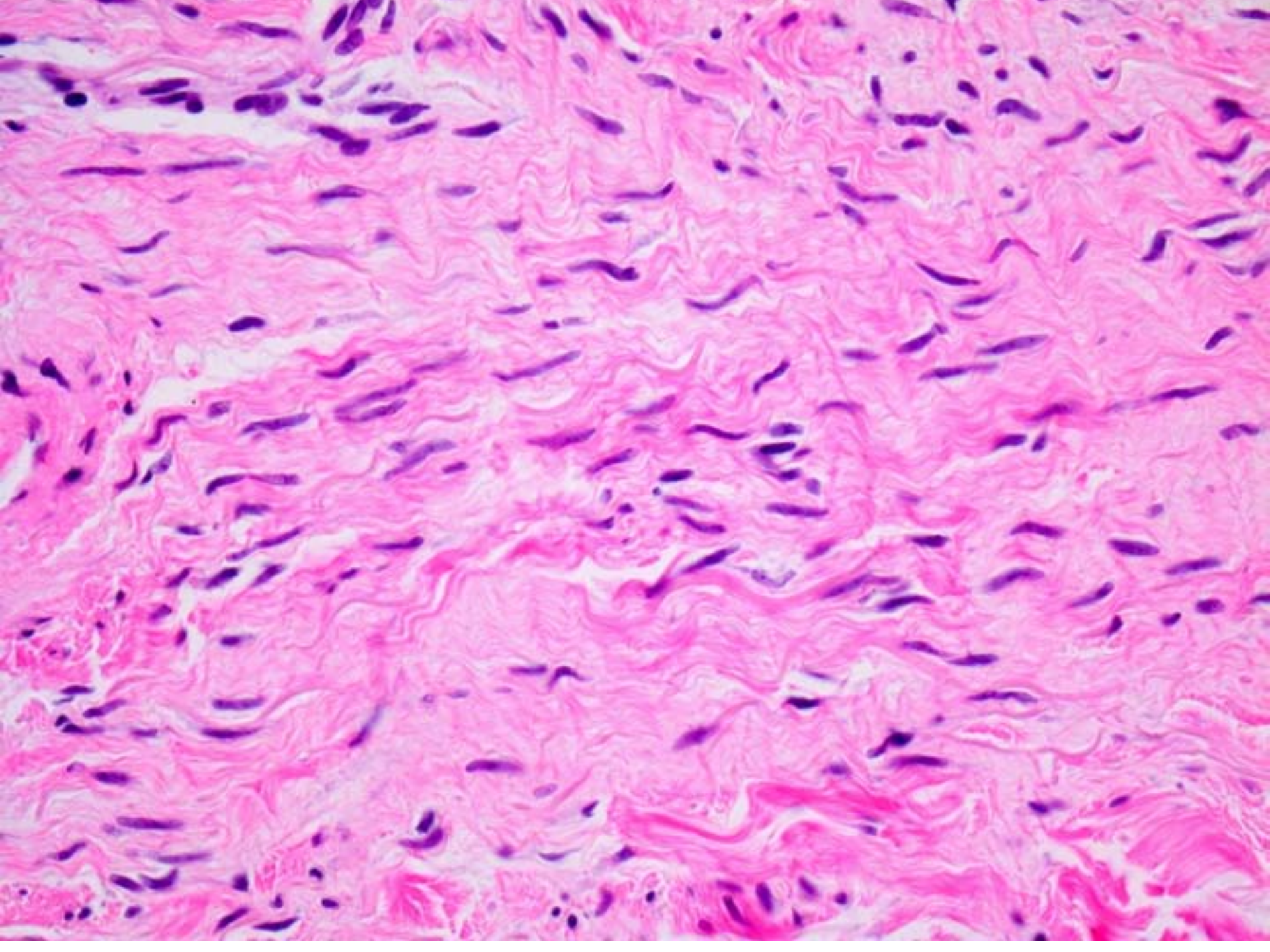












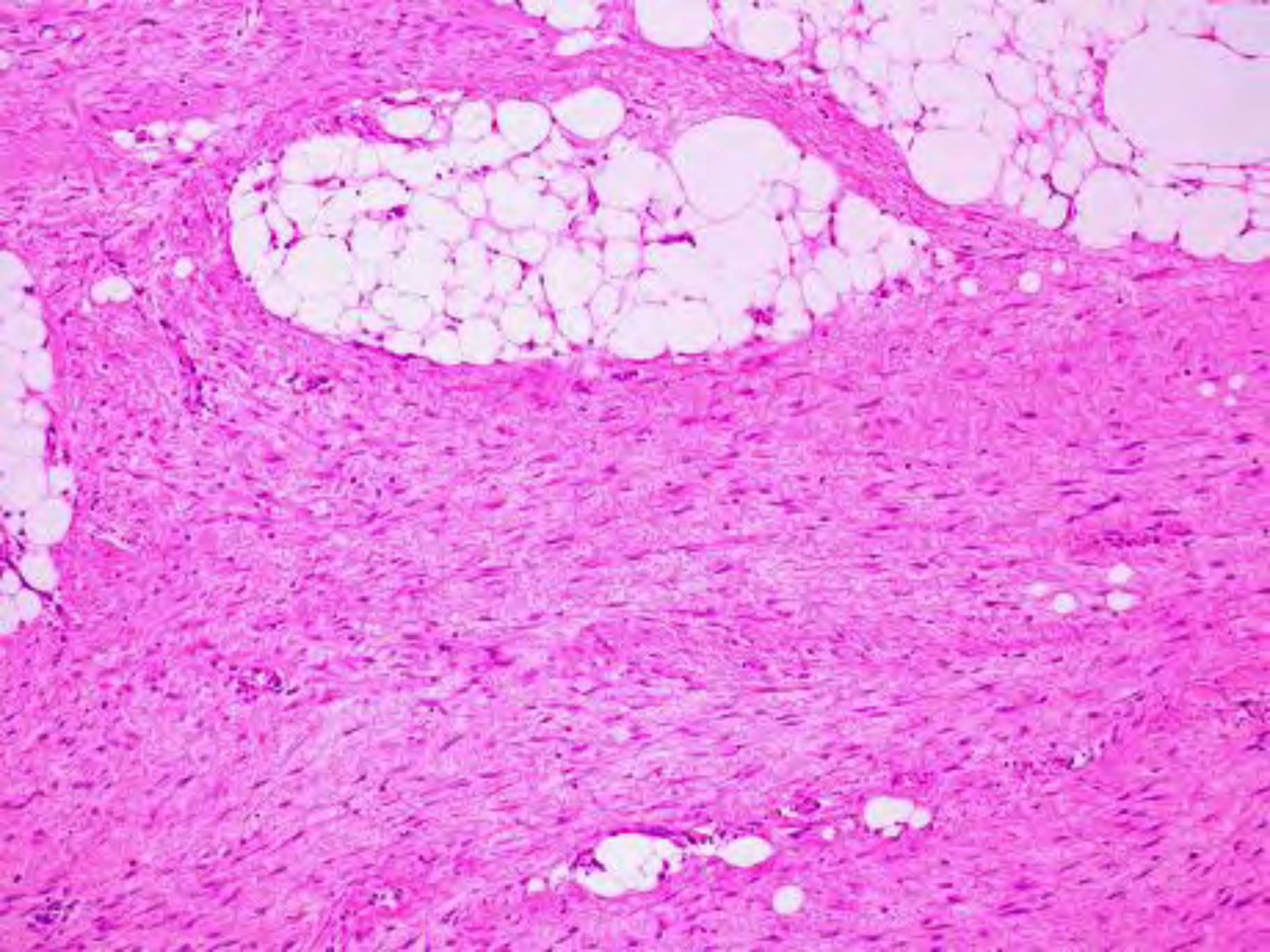
DESMOID TUMOR

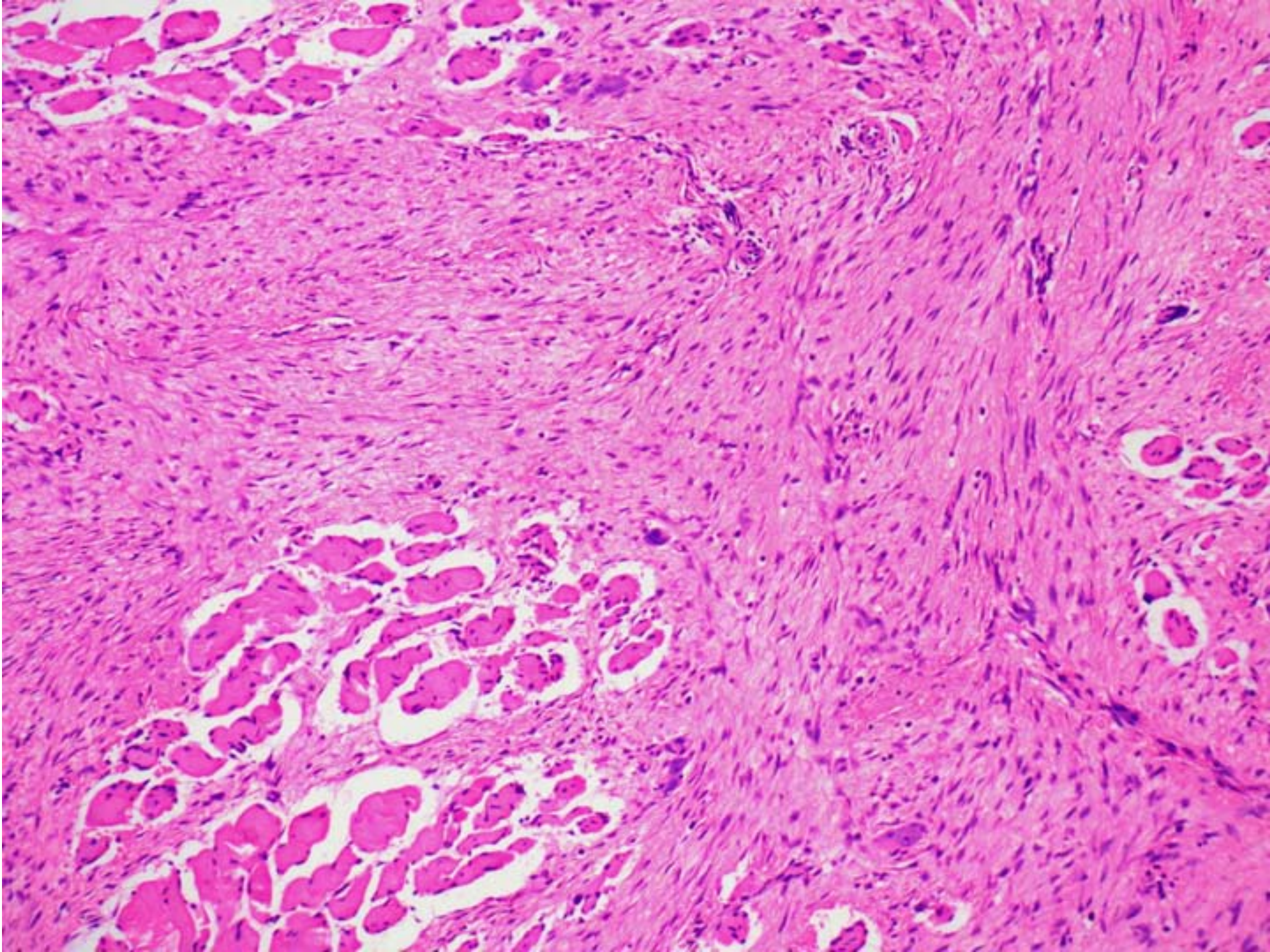
- Extraabdominal desmoid
 - Age - 10-40 years
 - Location - shoulder, chest wall, back, thigh
 - Deep-seated, firm, poorly defined mass
- Abdominal desmoid
 - Typically adult women
 - Location- fascial coverings of rectus and internal abdominal oblique muscles

DESMOID TUMOR

- Low power
 - poorly circumscribed tumor, ill defined fascicles
 - infiltrative growth pattern, often into muscle
 - necrotic muscle fibers common
- Cells - uniform, wavy, pointed
- Stroma
 - delicate, rarely sclerotic
 - mucoid stroma - focal, rare
 - chondroid or osseous metaplasia is rare







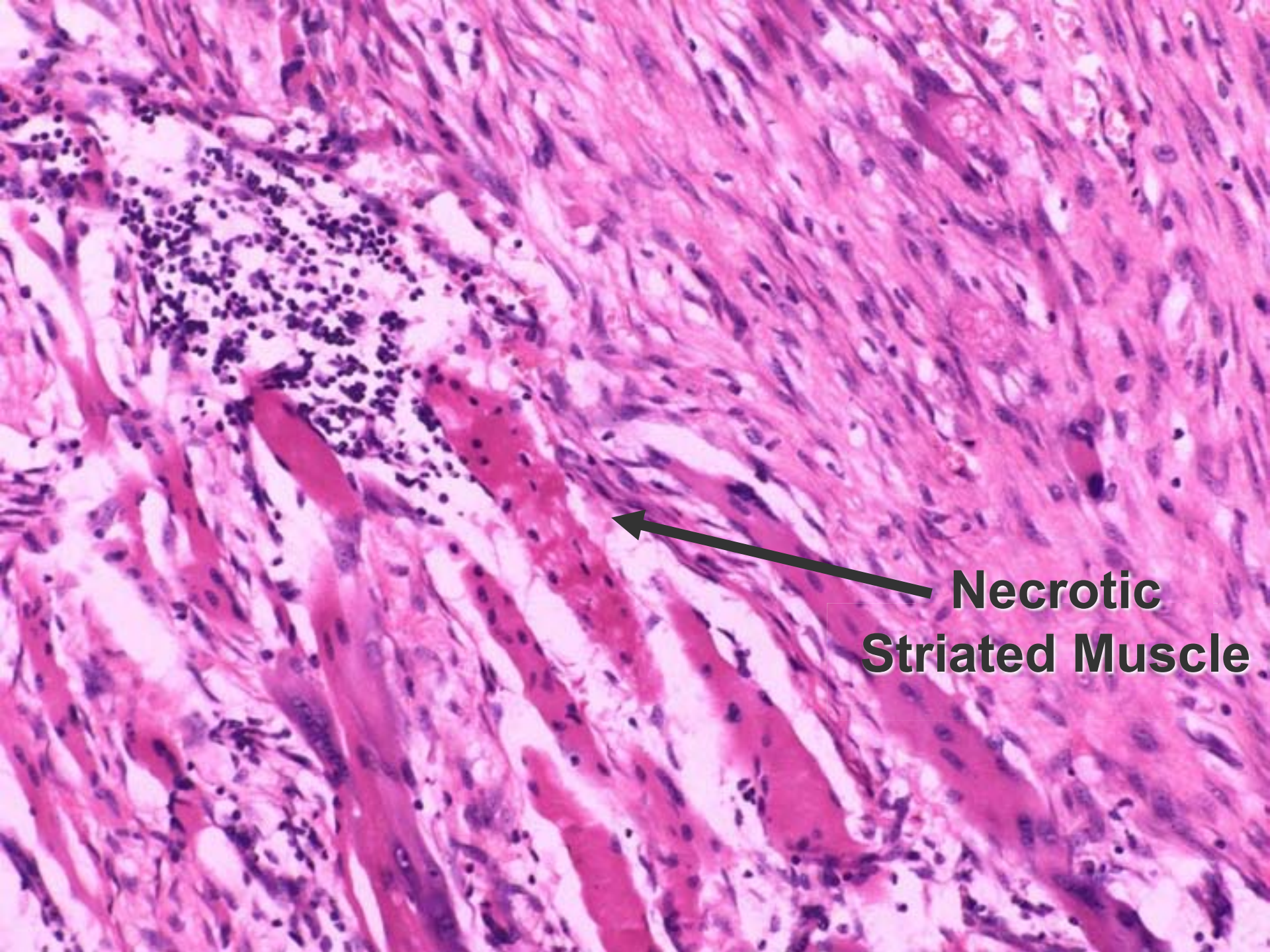


**Wavy Nuclei with
Pointed Tips** →

This histological image shows a section of smooth muscle tissue stained with hematoxylin and eosin (H&E). The tissue is characterized by elongated, spindle-shaped cells with prominent, wavy nuclei that have pointed tips. These nuclei are arranged in a regular, parallel pattern. The background is a dense, pink-stained extracellular matrix containing collagen fibers, which appear as thin, wavy lines. The overall texture is fibrous and organized.

**Collagen
Thin Tibers** →

This label points to the collagen fibers in the tissue, which are described as thin tibers. These fibers are visible as fine, wavy lines in the pink-stained extracellular matrix, providing structural support to the smooth muscle cells.

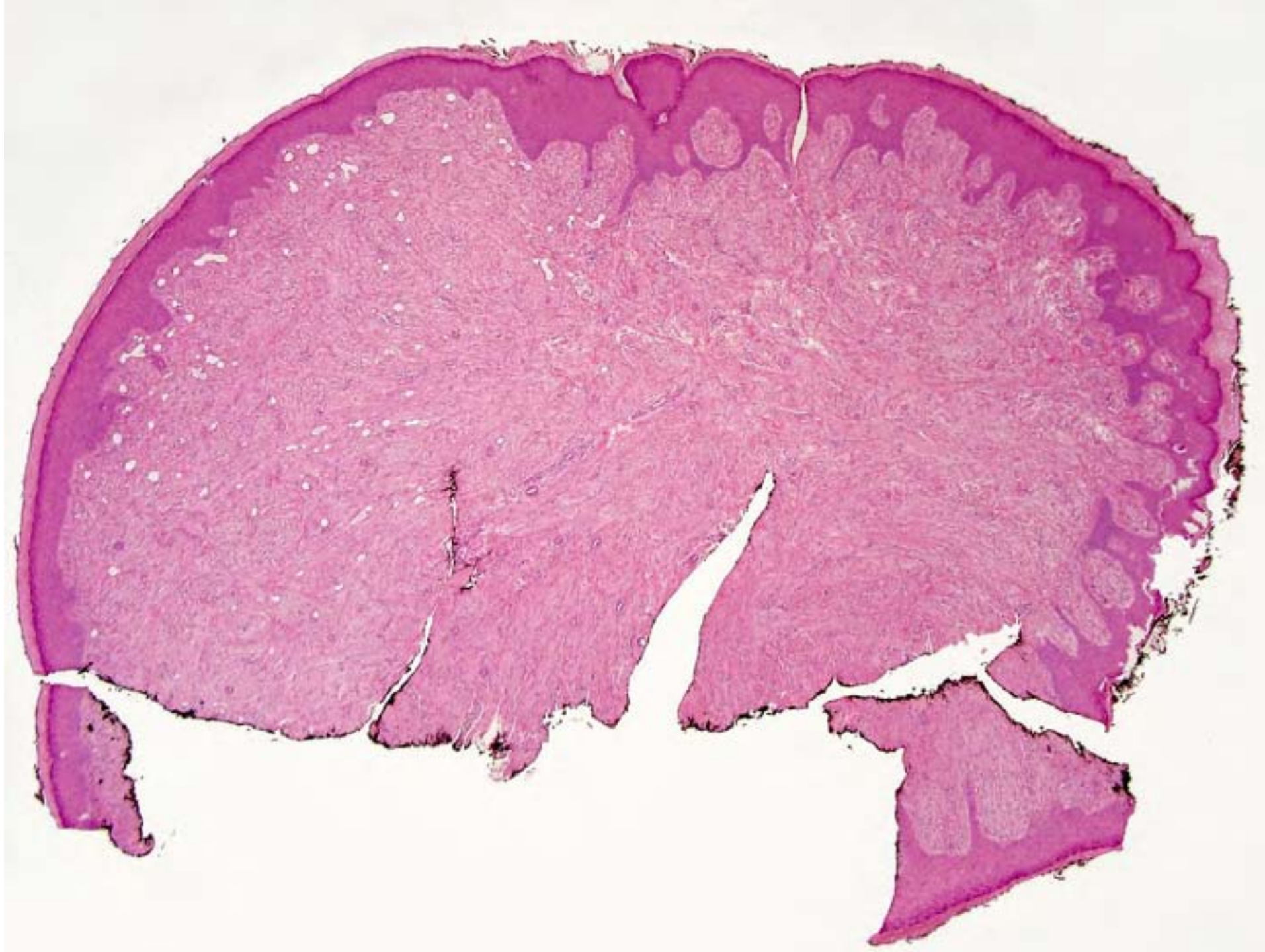


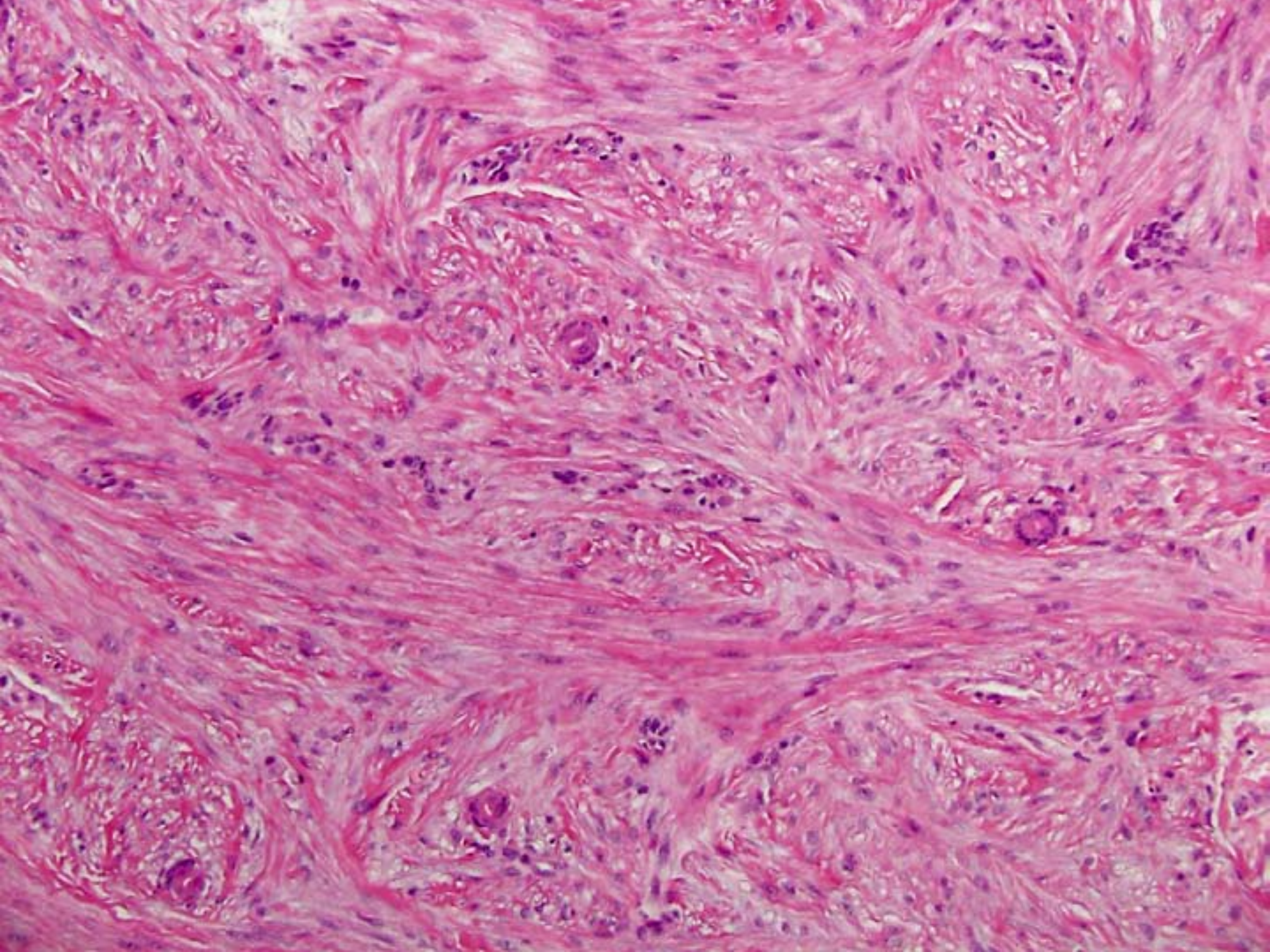
**Necrotic
Striated Muscle**

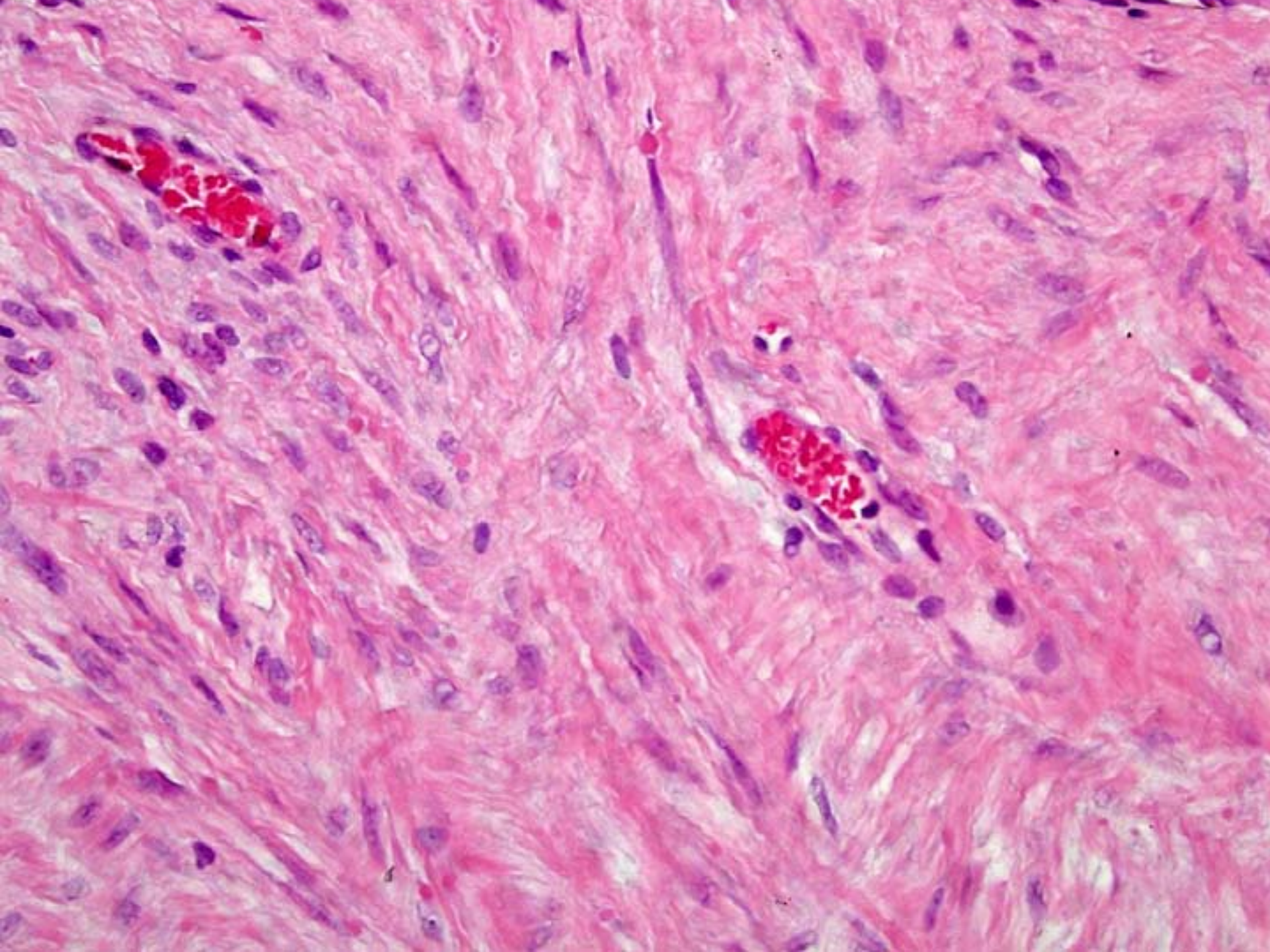
INFANTILE DIGITAL FIBROMA

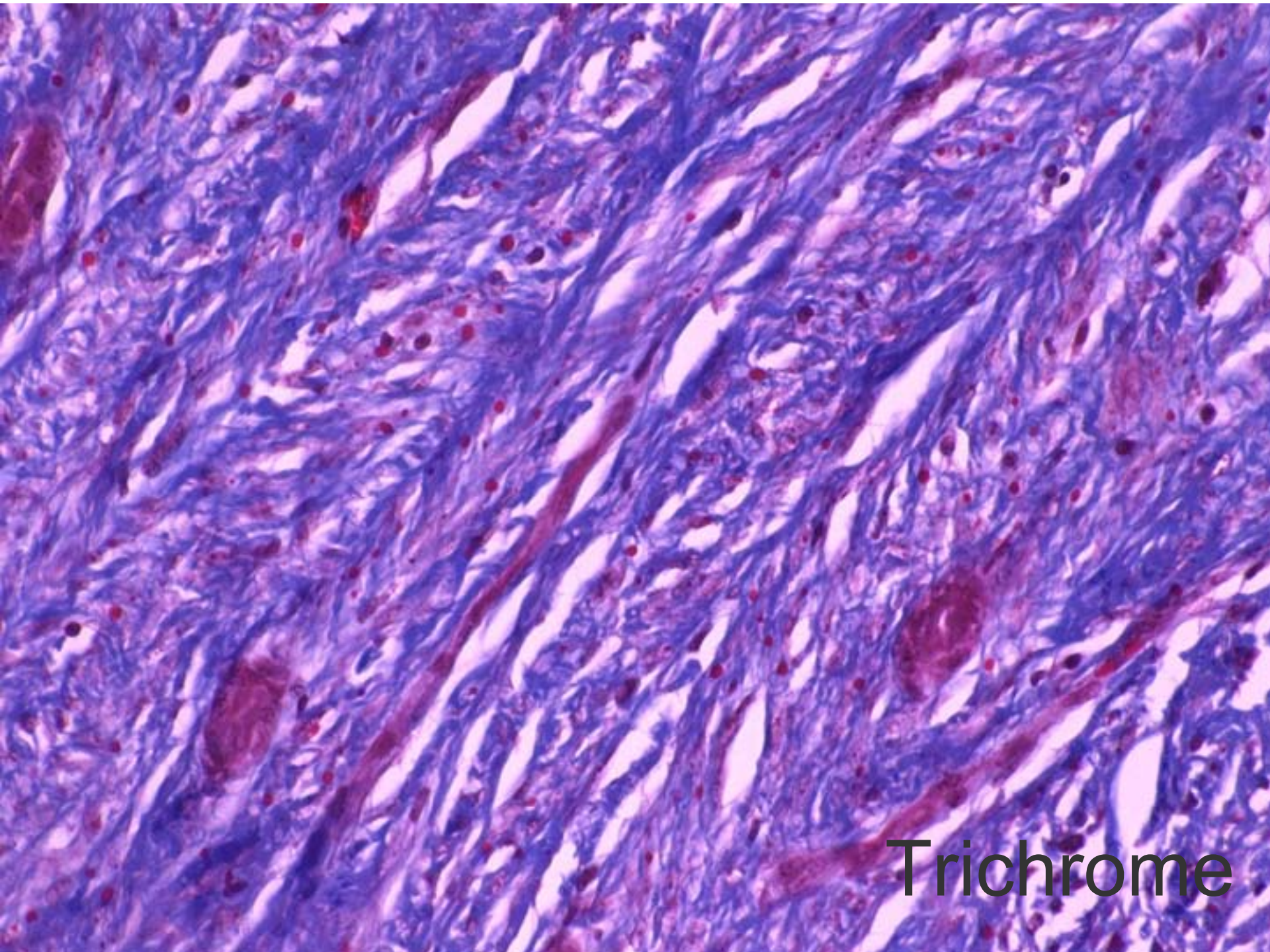
- Neonates & infants, rarely children and adolescents
- Skin - colored or erythematous nodule
- Location - digits of hands, occasionally feet
- May recur locally after excision
- Regress spontaneously over years





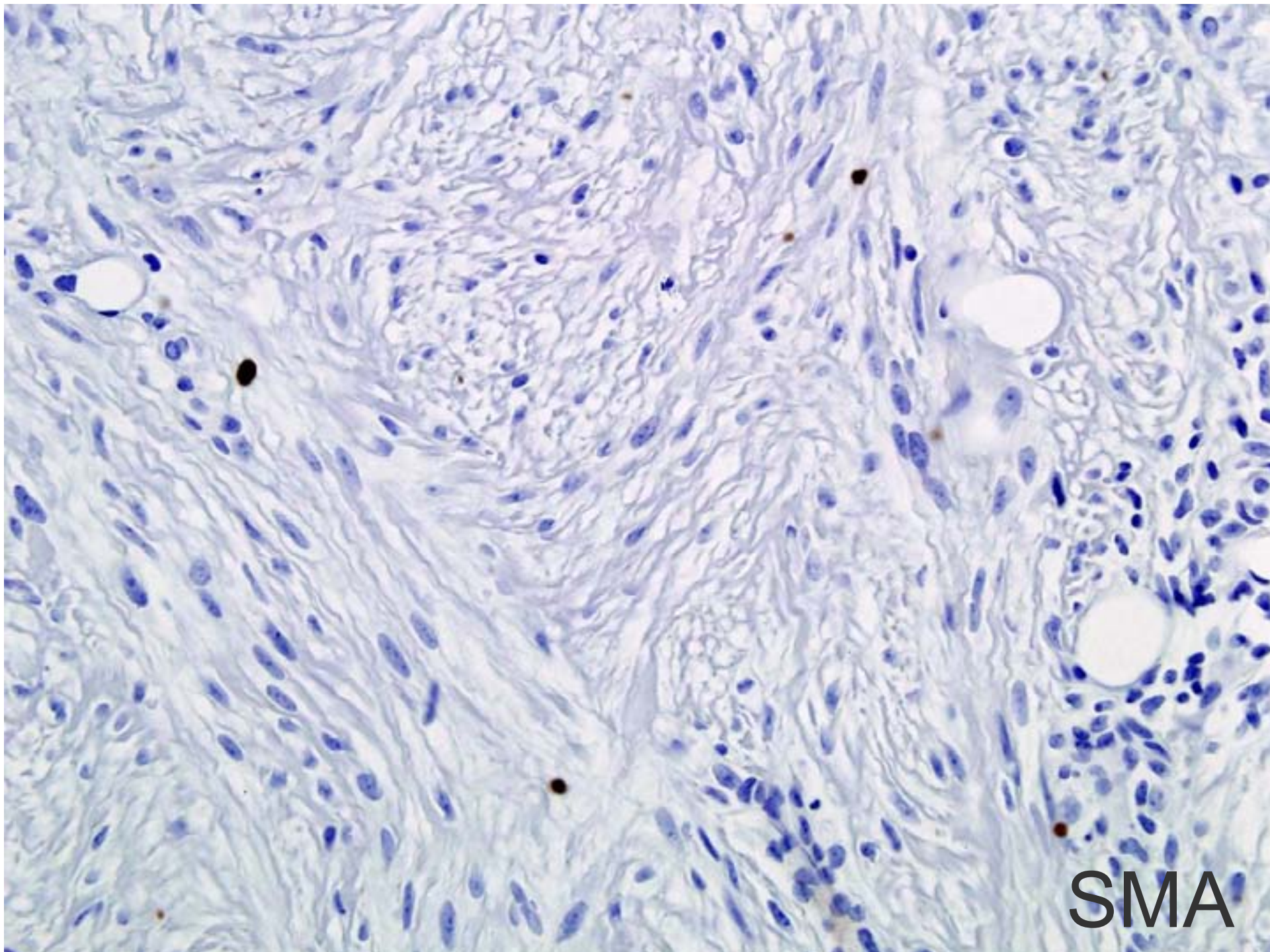






Trichrome



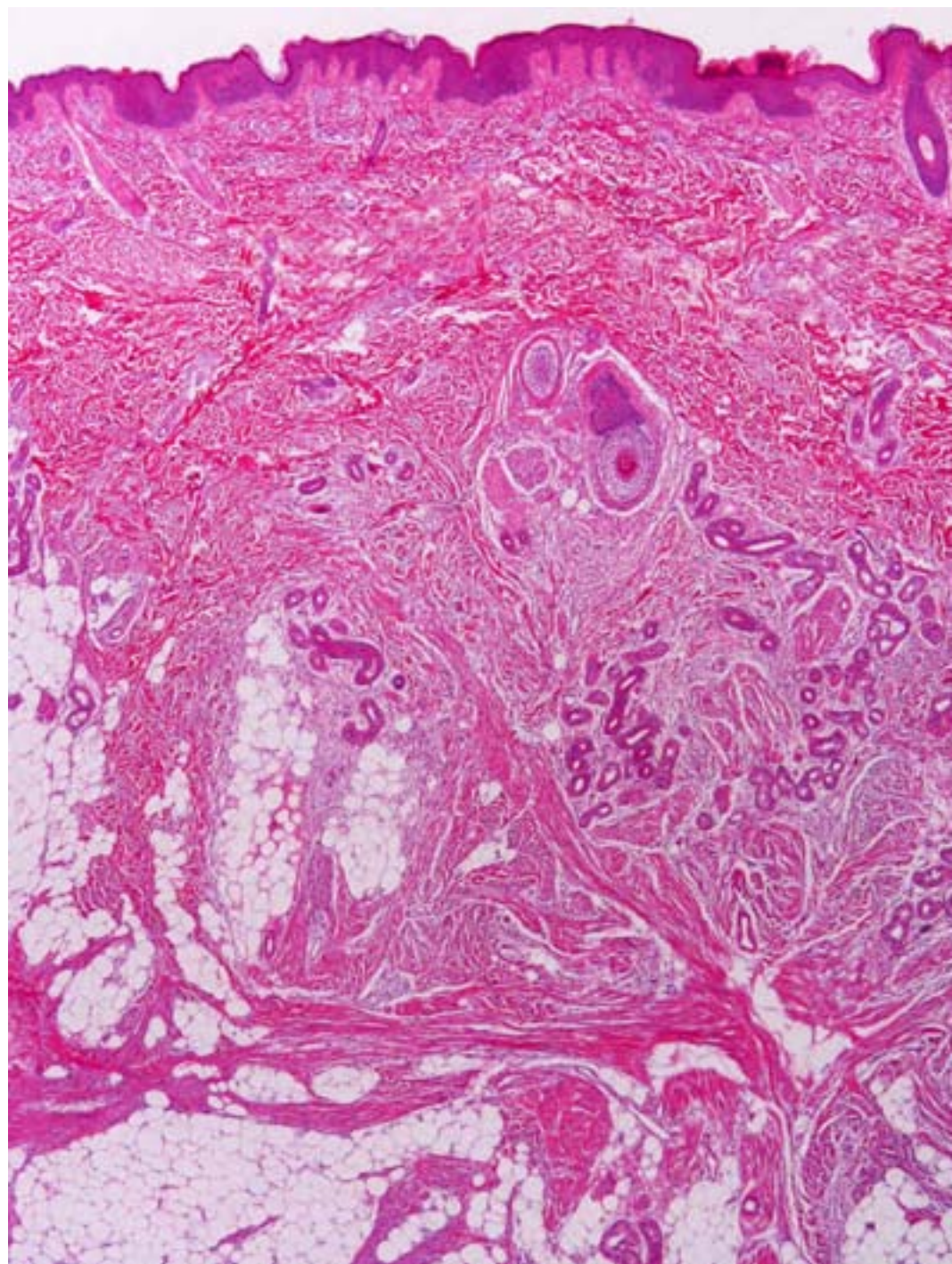


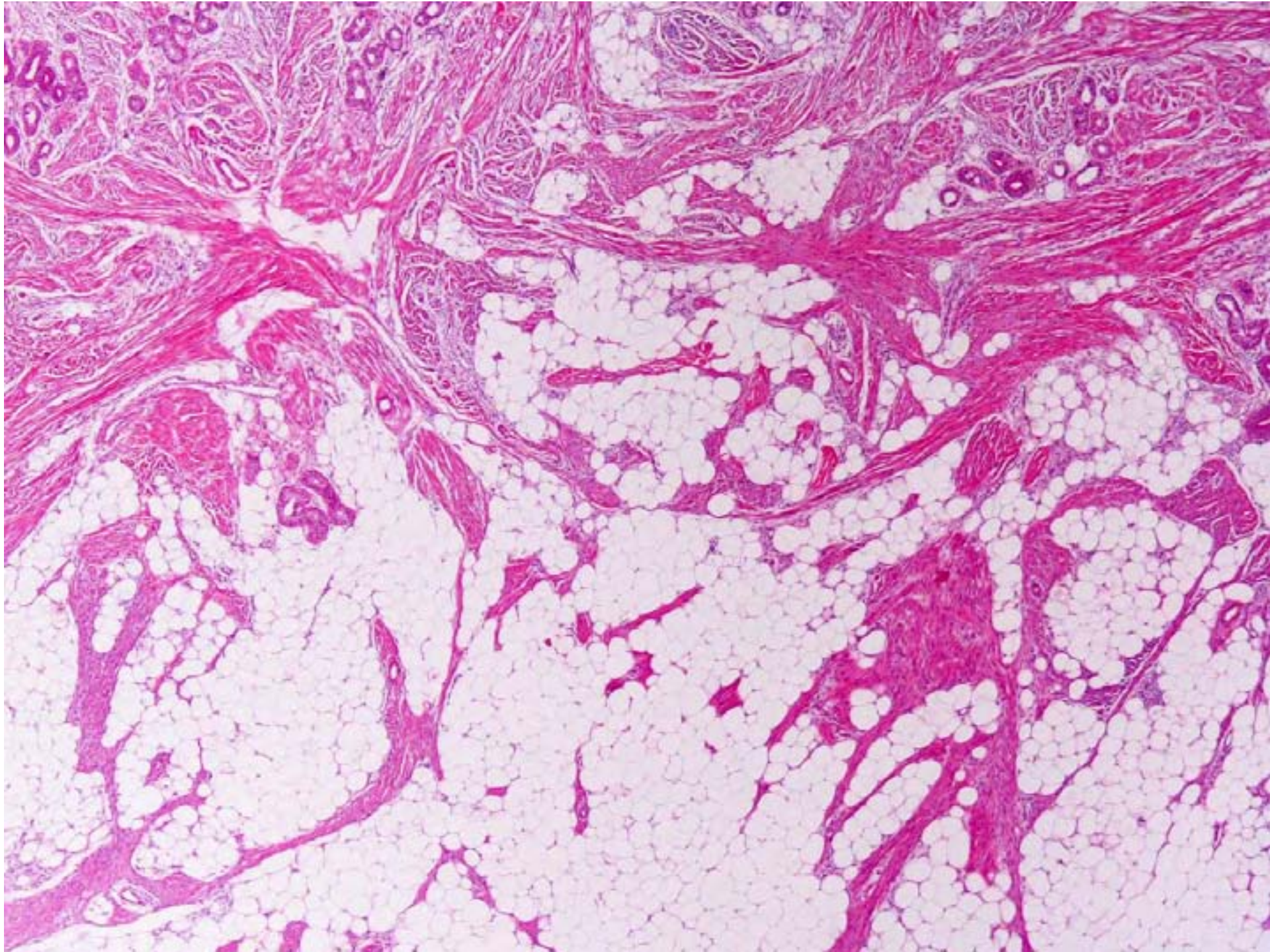
SMA

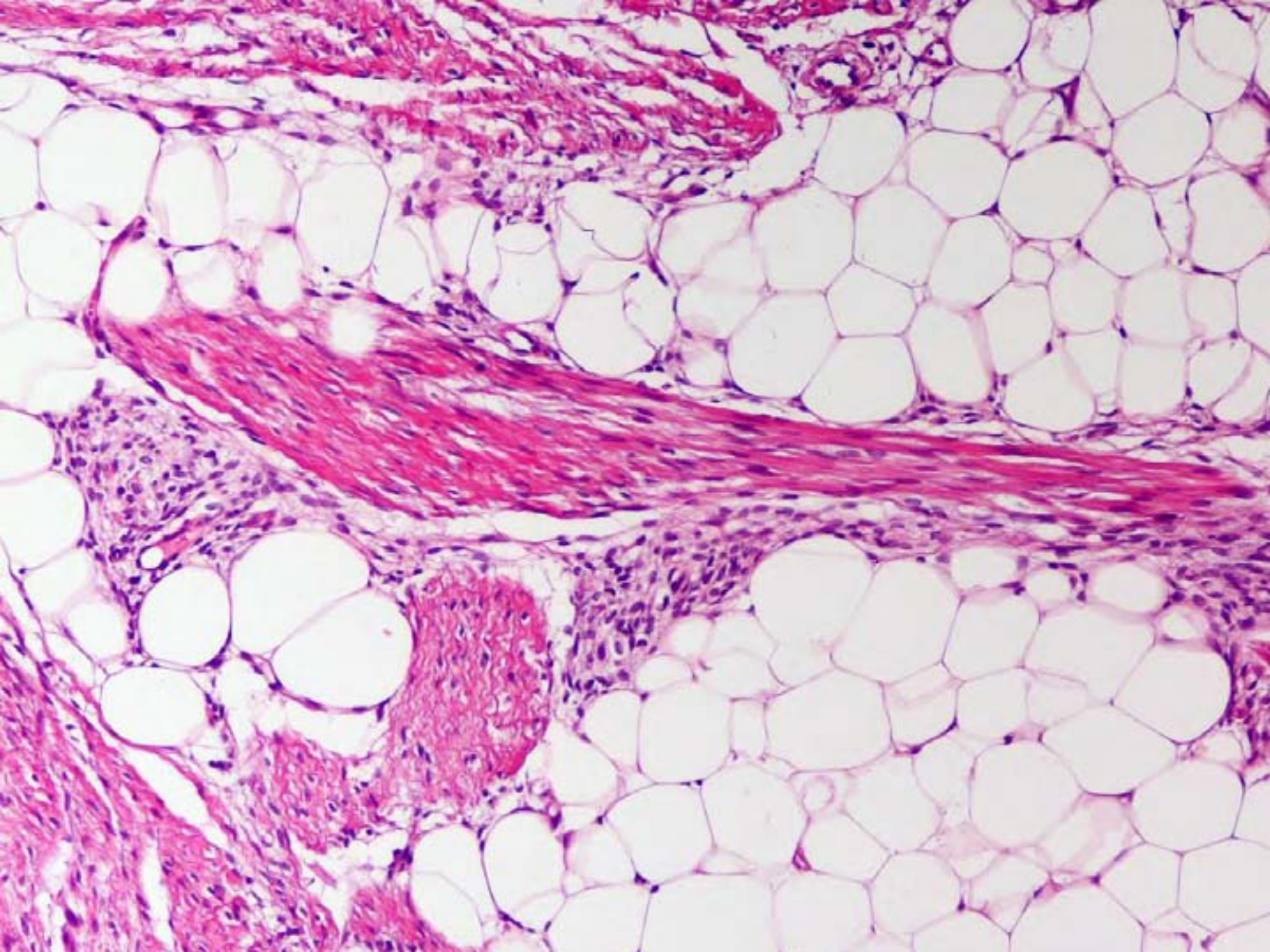
FIBROUS HAMARTOMA OF INFANCY

- Poorly circumscribed tumor in subcutis
- TRIPHASIC - three different components:
 - well-defined interlacing fascicles with abundant collagen
 - foci of immature ovoid cells with myxoid stroma
 - interspersed mature fat
- Less commonly fibrosis resembling neurofibroma

Fibrous Hamartoma of Infancy

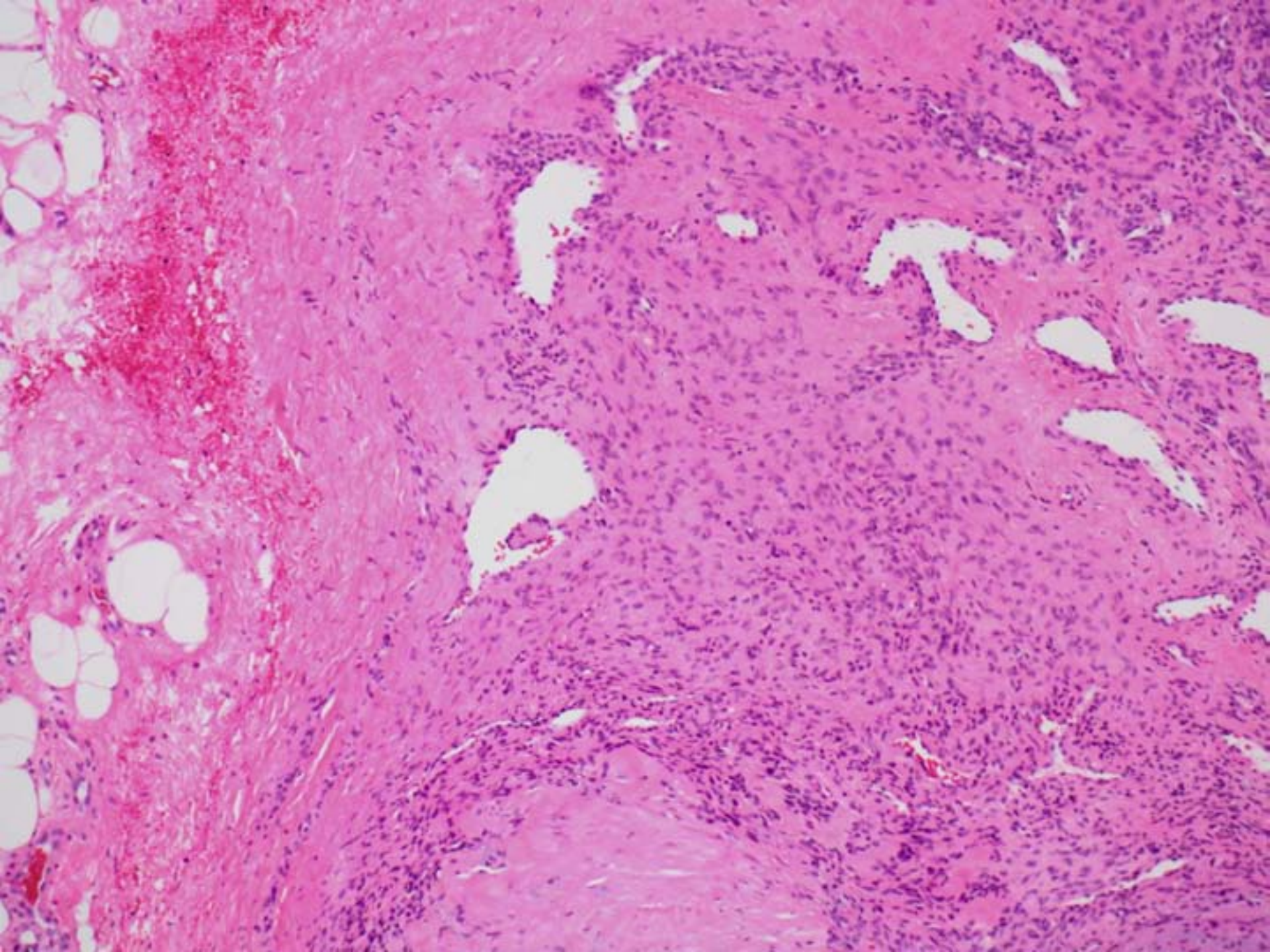


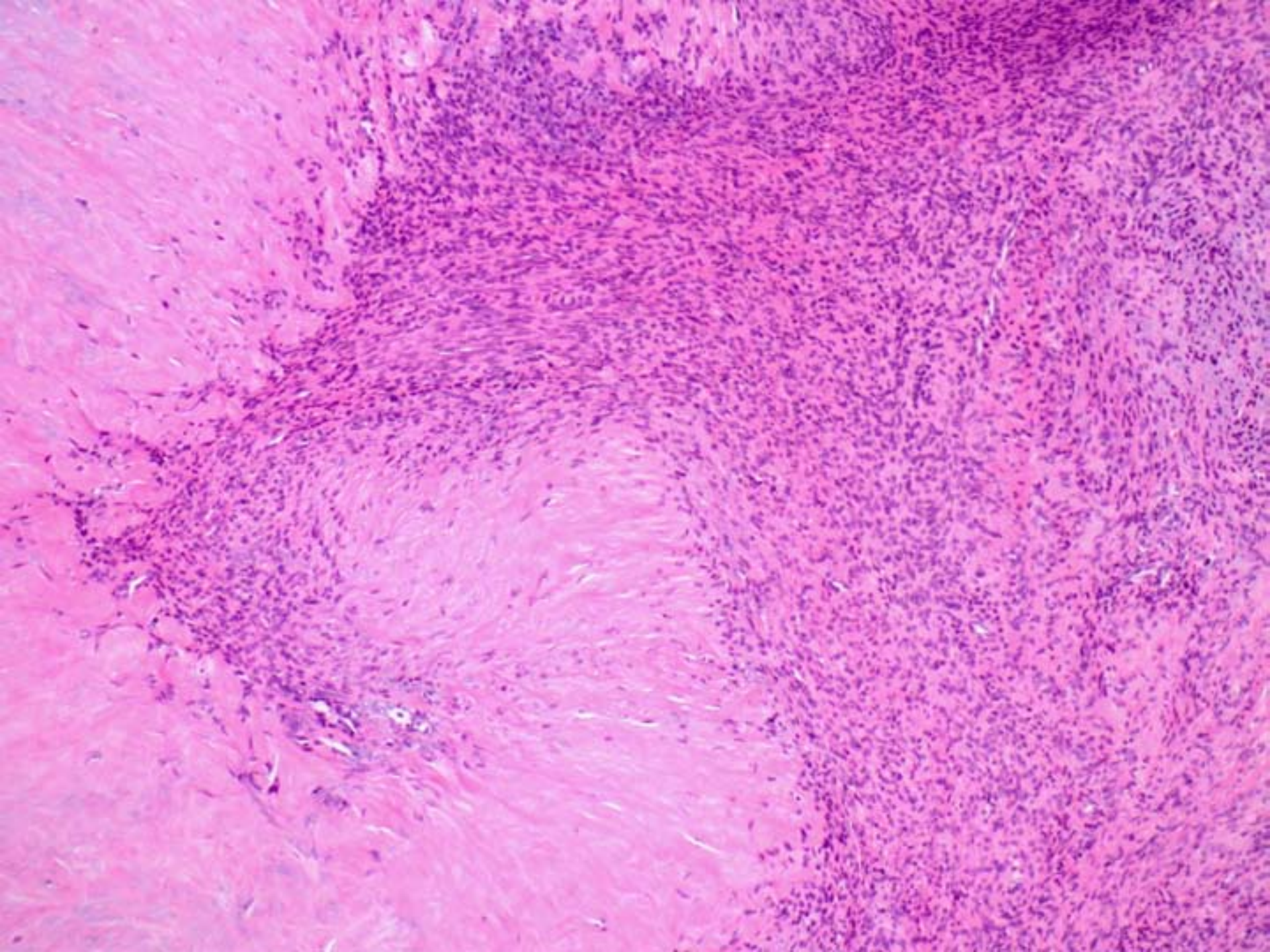


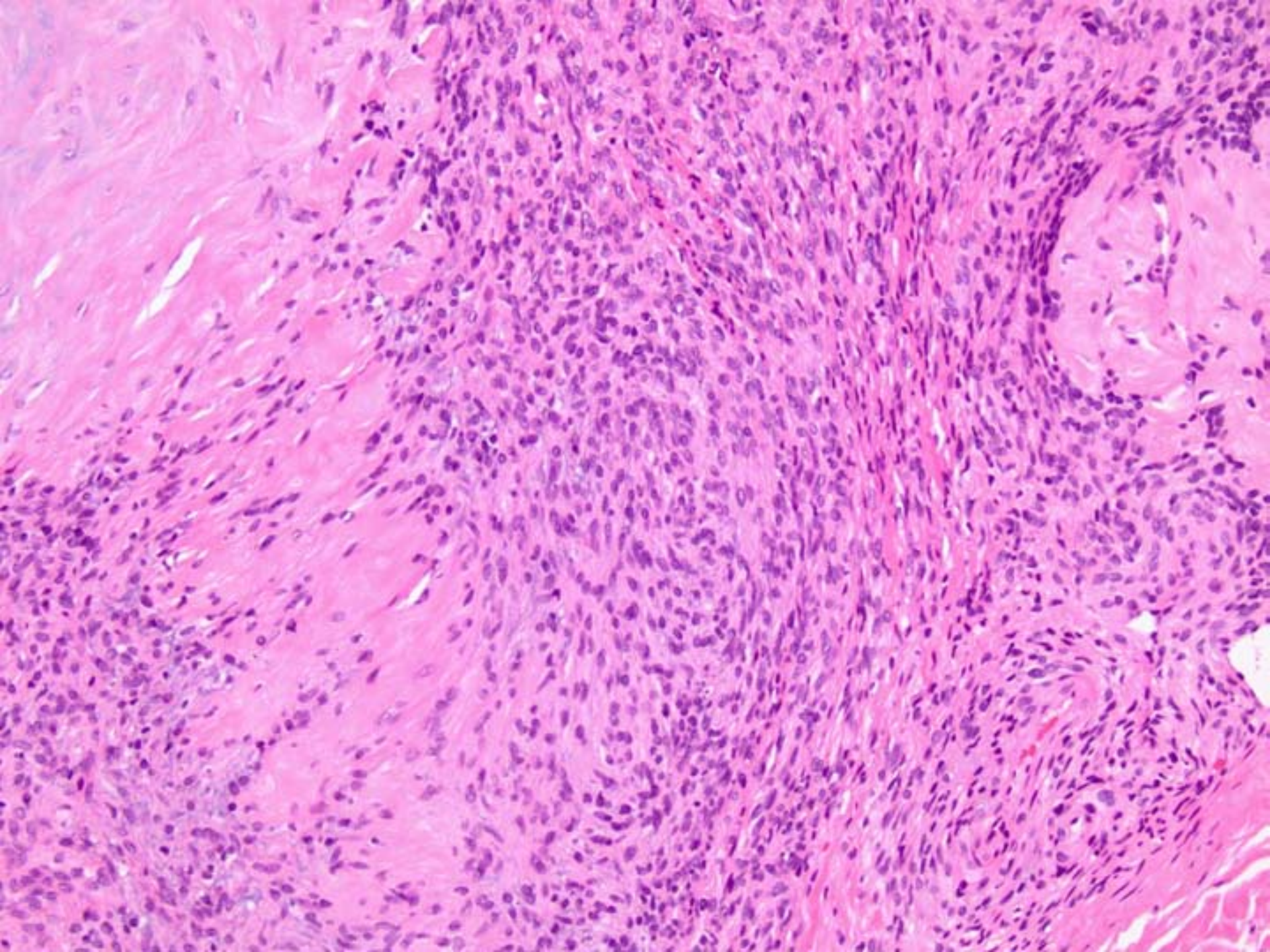


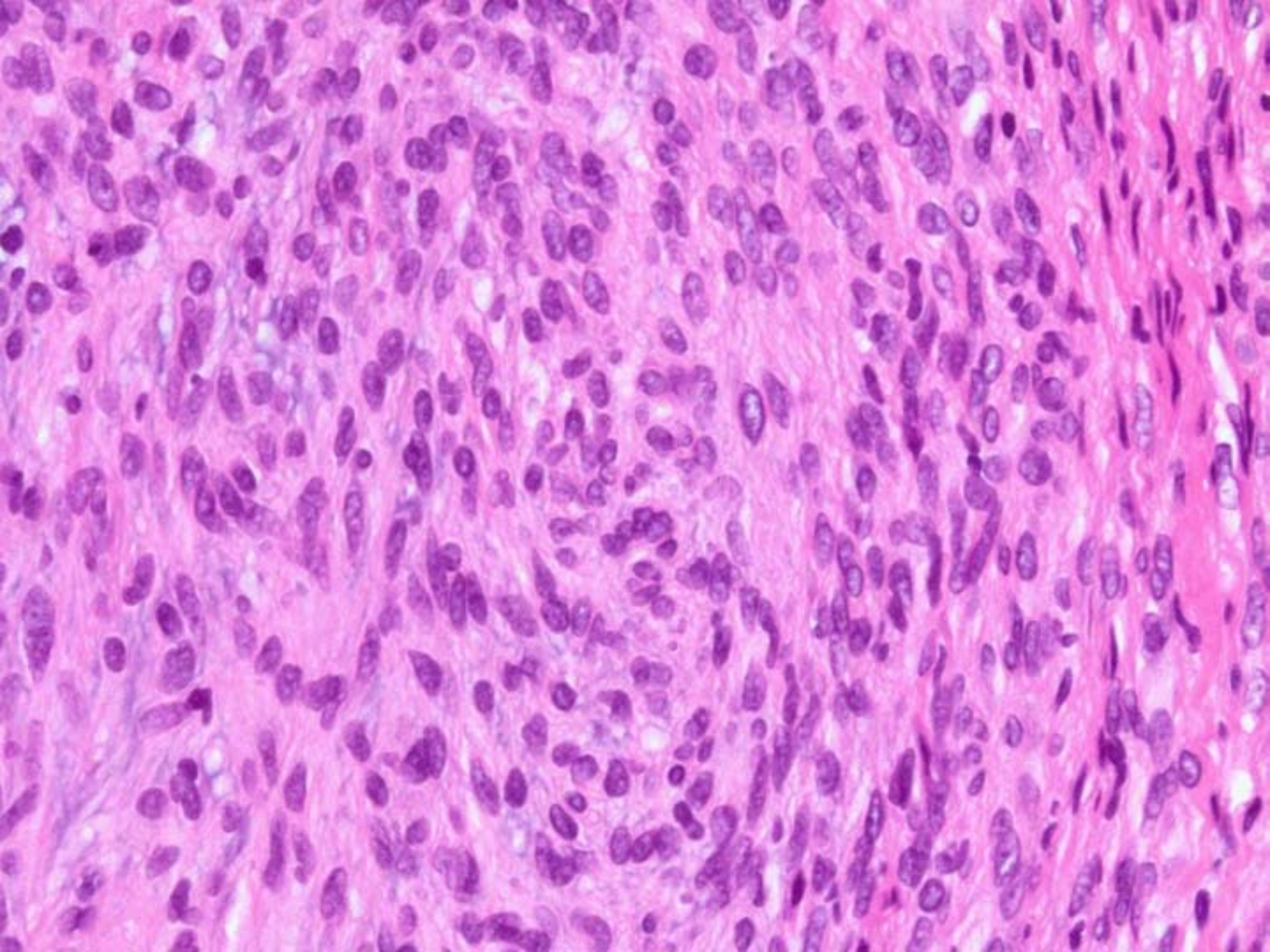
MYOFIBROMA

- Congenital variant (at birth or in early life)
 - 50% solitary, 50% multiple
 - usually SQ nodules
 - spontaneous regression (for pt with few lesions)
- Adult variant
 - single or rarely multiple
 - non-descript nodule, resembles DF or scar
- BIPHASIC histological pattern
 - spindled fascicles that resemble smooth muscle
 - hemangiopericytoma-like areas (vascular spaces)



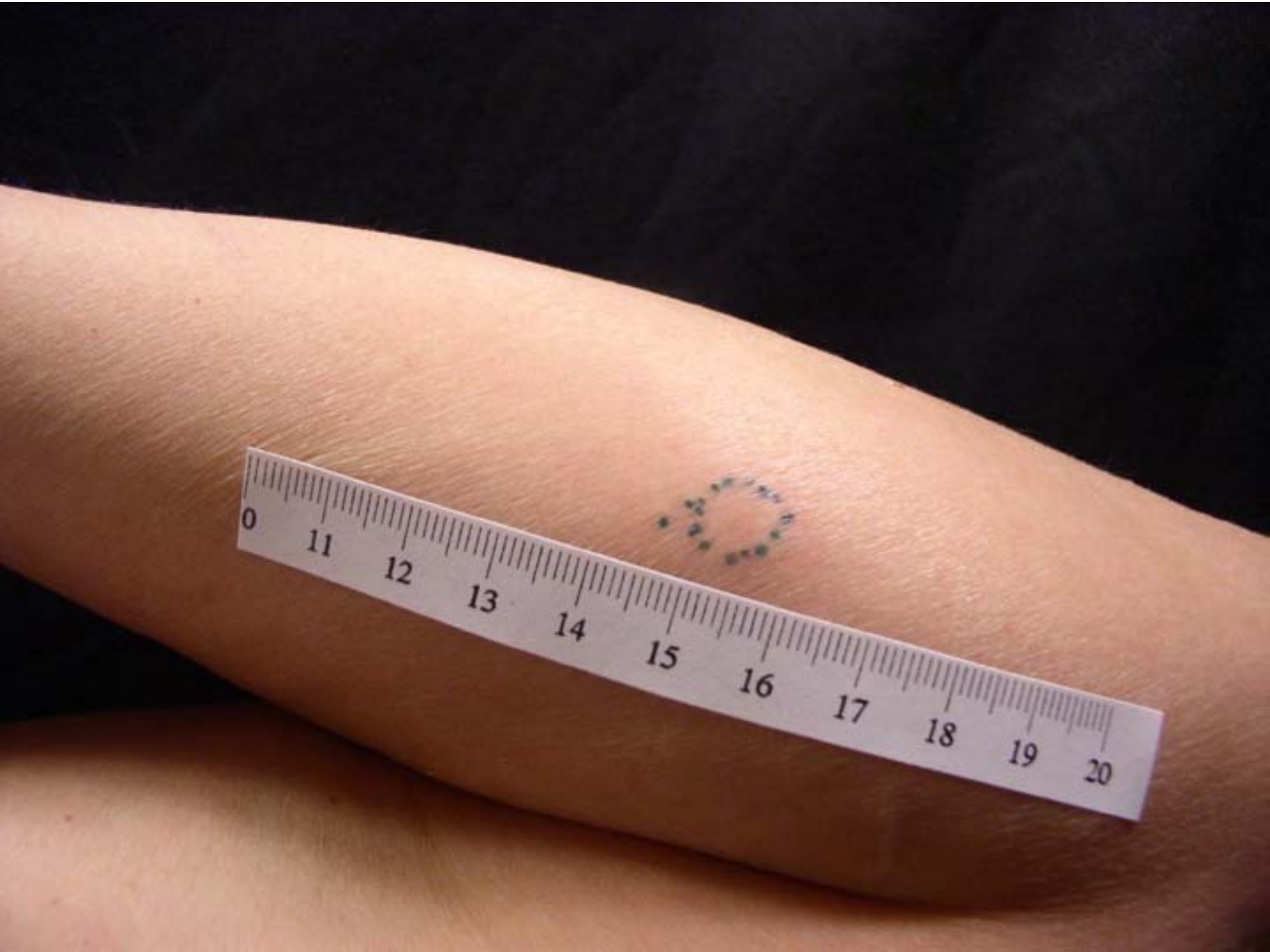






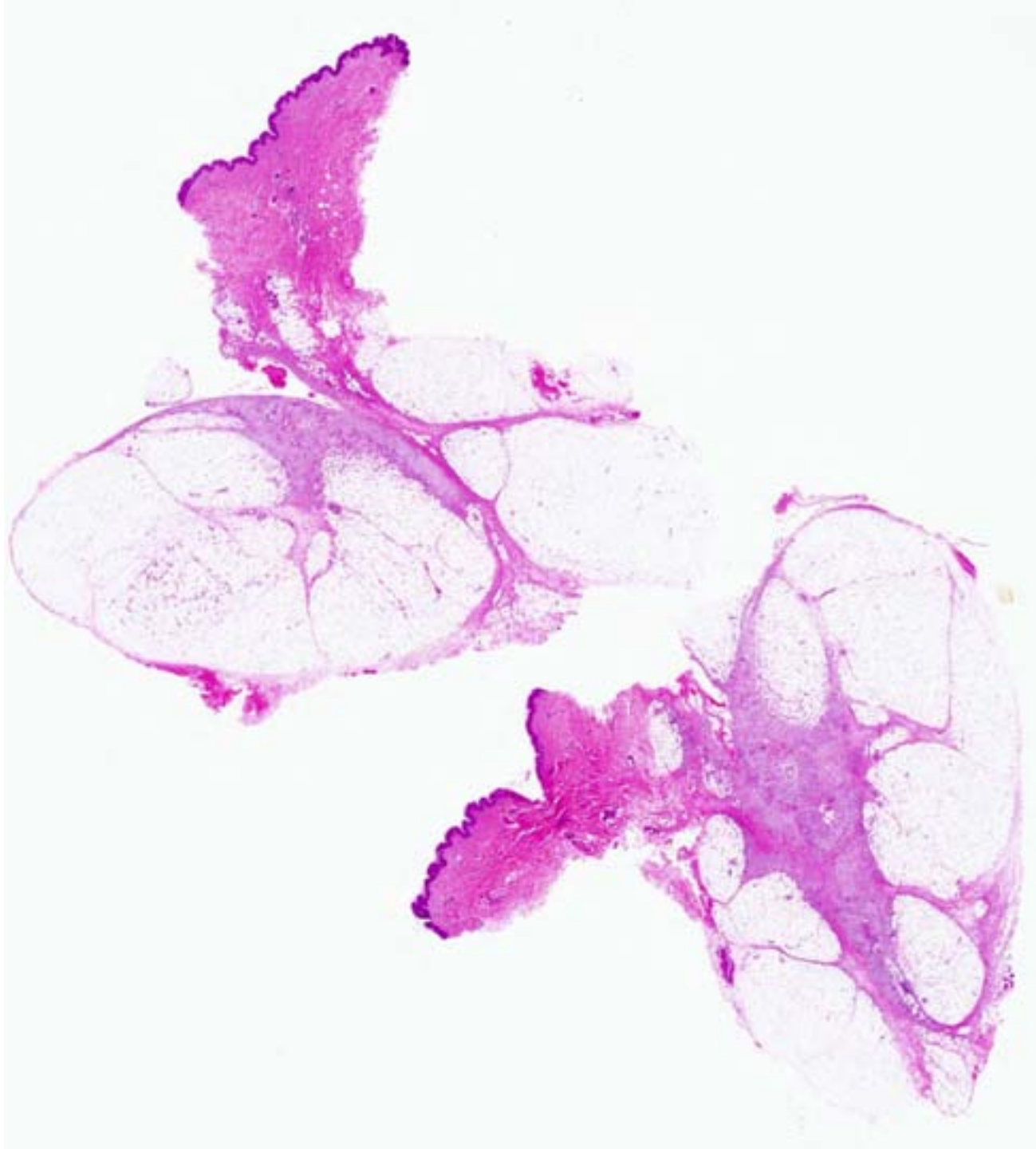
NODULAR FASCIITIS

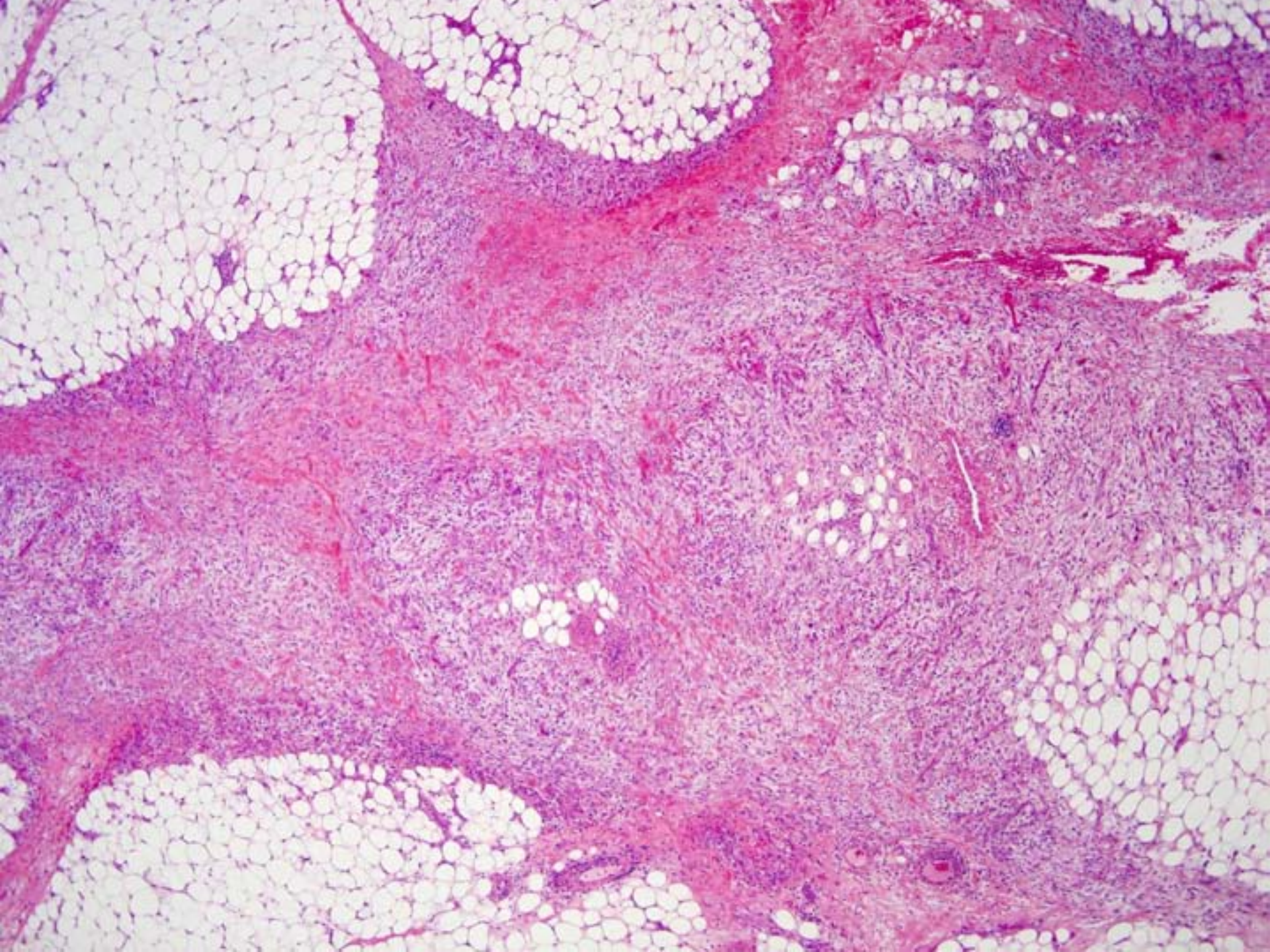
- Patients typically young adults
- Primary lesion – usually solitary, subcutaneous nodule, 1-3 cm
- Usually a history of rapid growth
- Most common locations
 - Adults - forearm
 - Children - head and neck (cranial fasciitis)

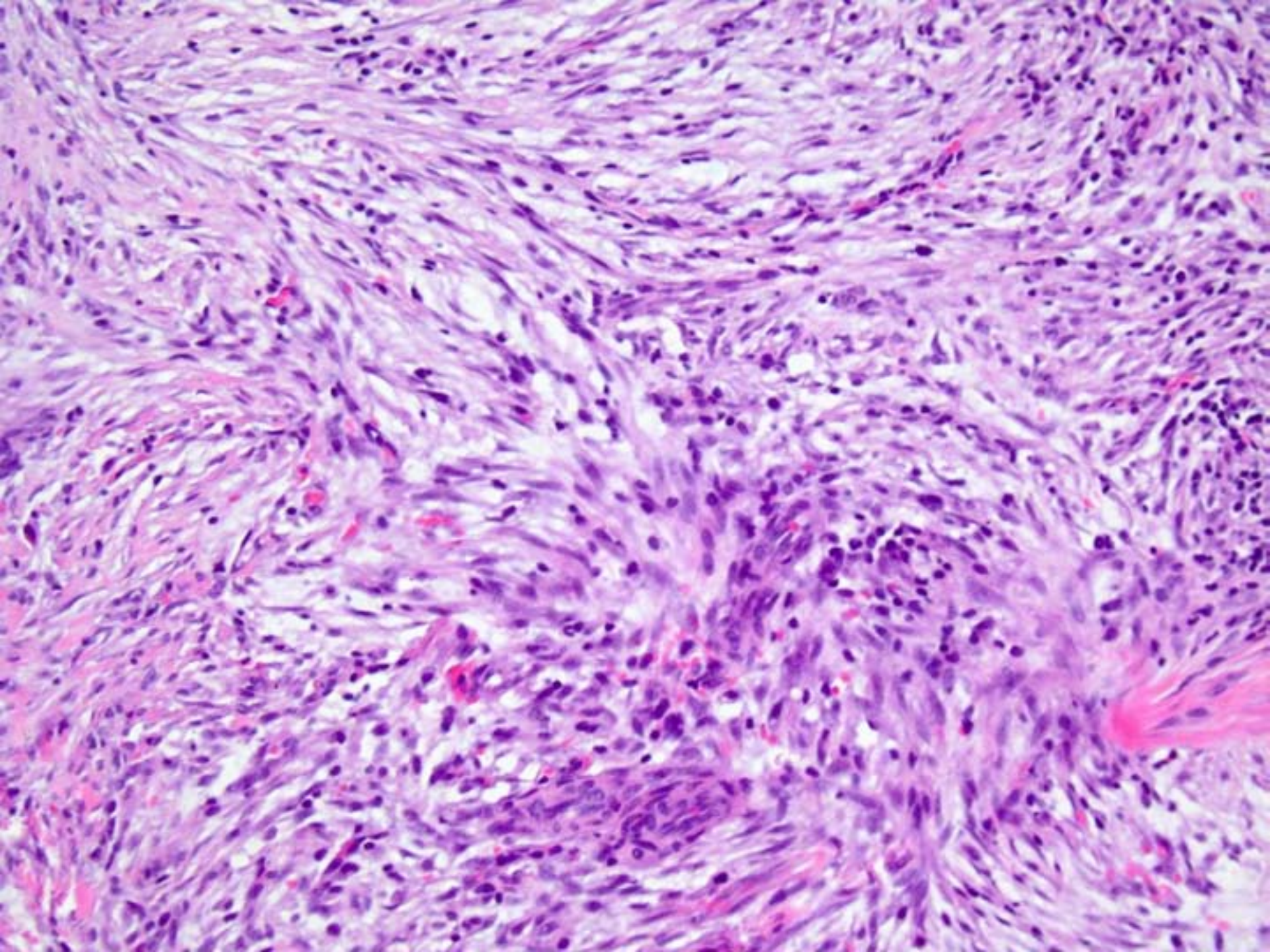


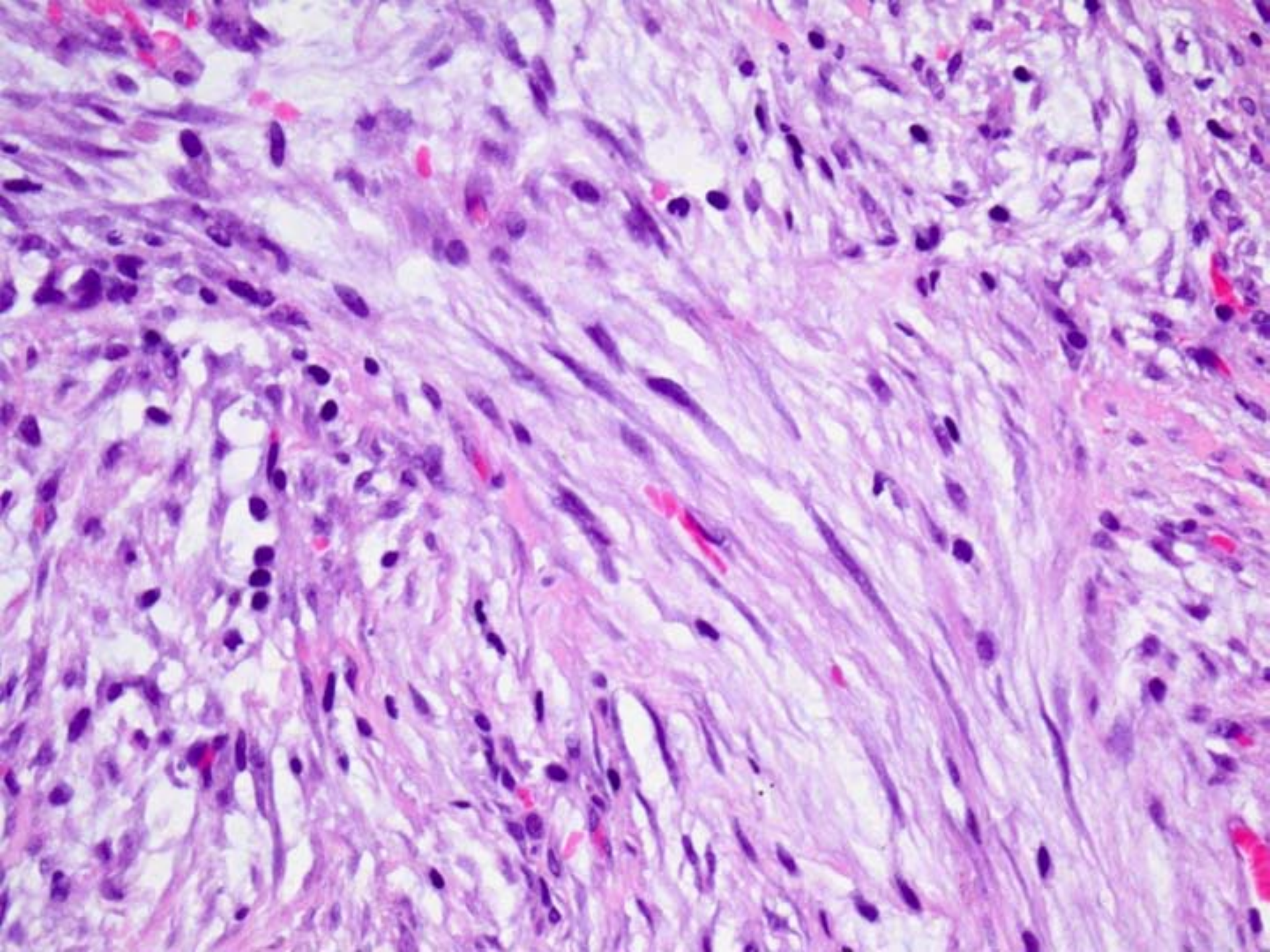
Histology

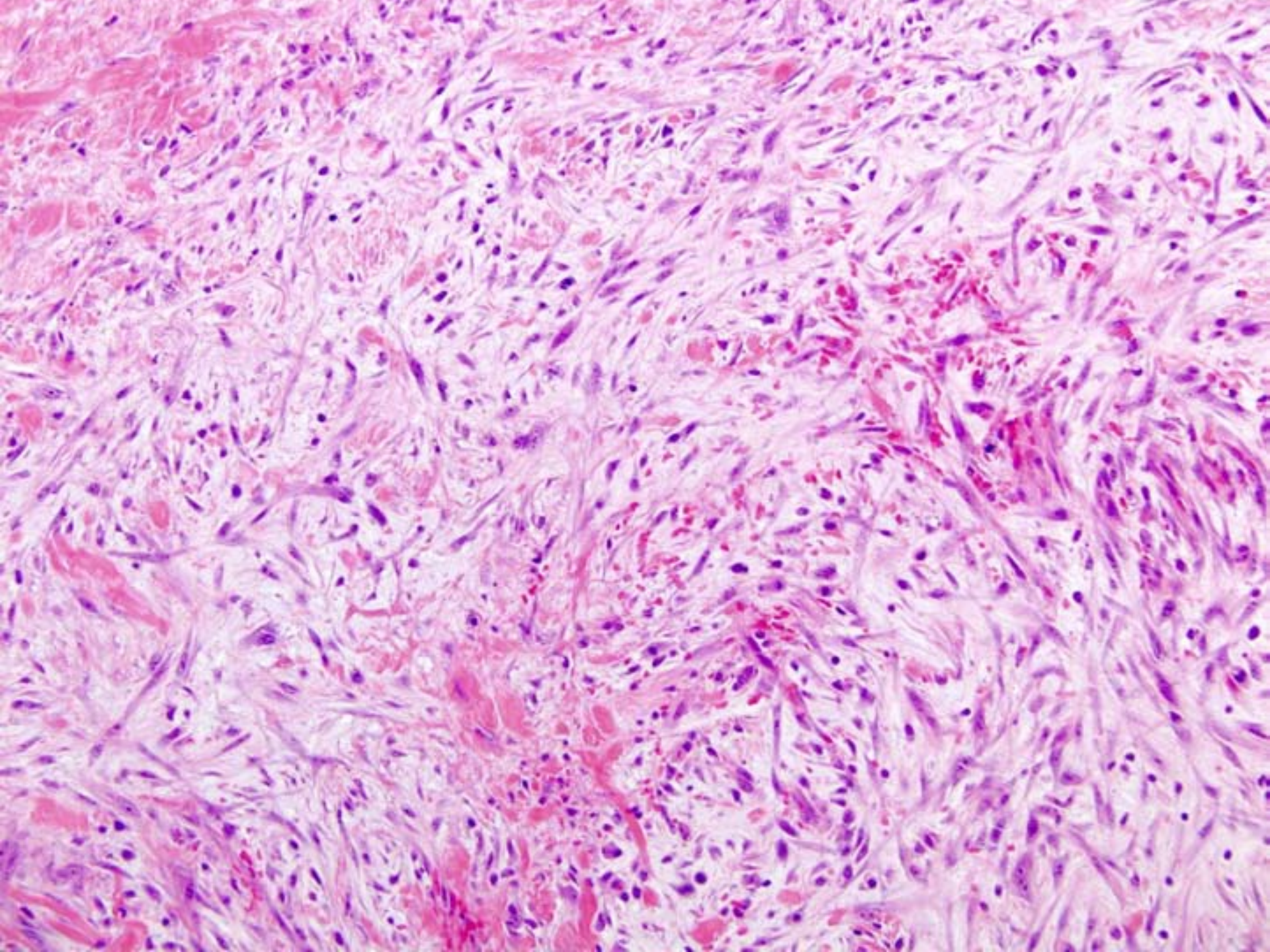
- Mass in the fibrous septa
- Cell arranged in fascicles
- Mucin/myxoid
- Chronic inflammation
- Extravasated RBCs
- Rare MNCs
- “Fibroblasts in tissue culture”

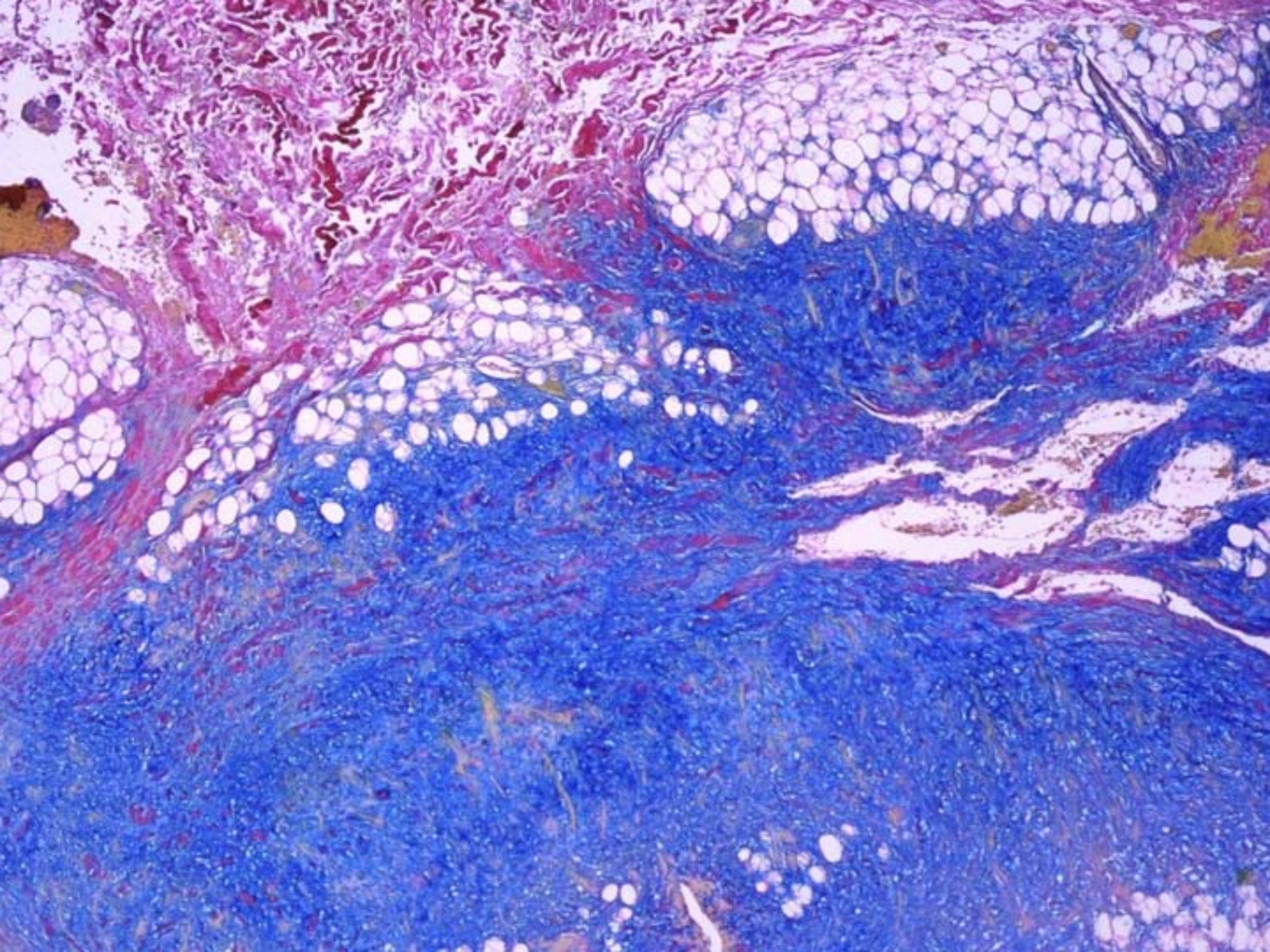












DERMATOFIBROMA

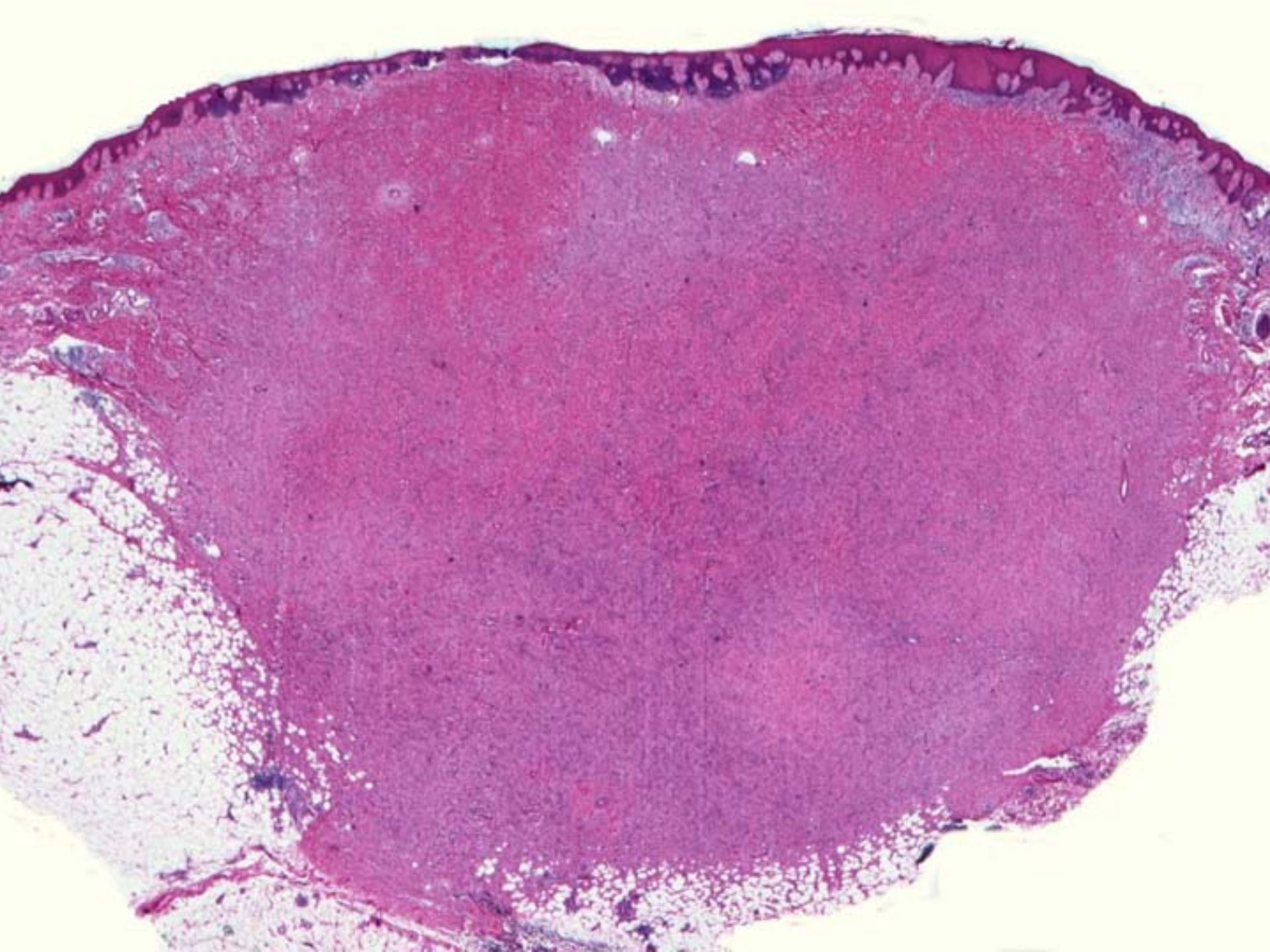
- Ubiquitous fibrohistiocytic tumor
- Typically arises in adults
- Women > men
- Most common location – legs > arms > trunk
- Primary lesion - firm nodule, exophytic or endophytic, positive dimple sign
- Eruptive lesions associated with SLE

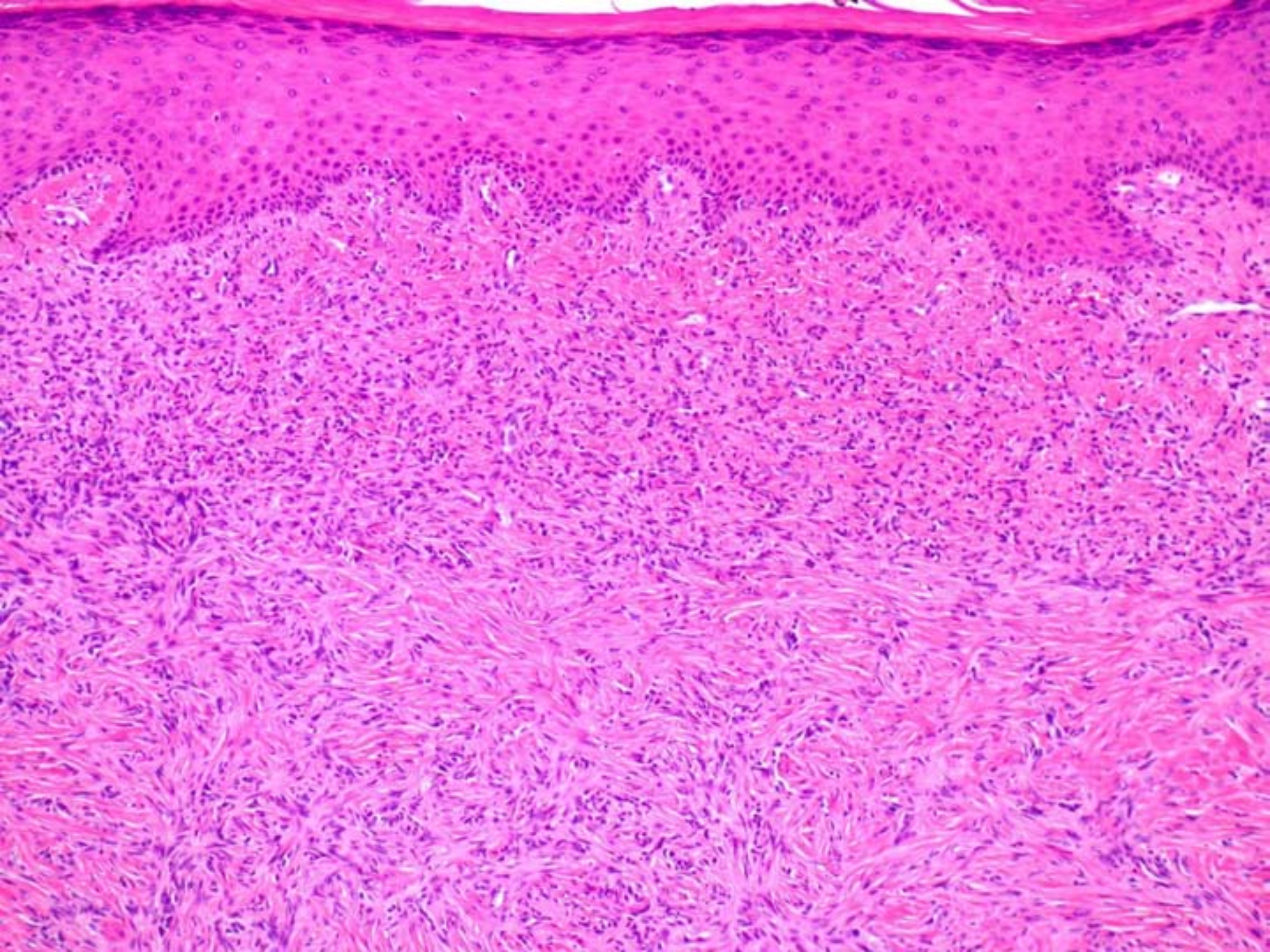
DERMATOFIBROMA VARIANTS

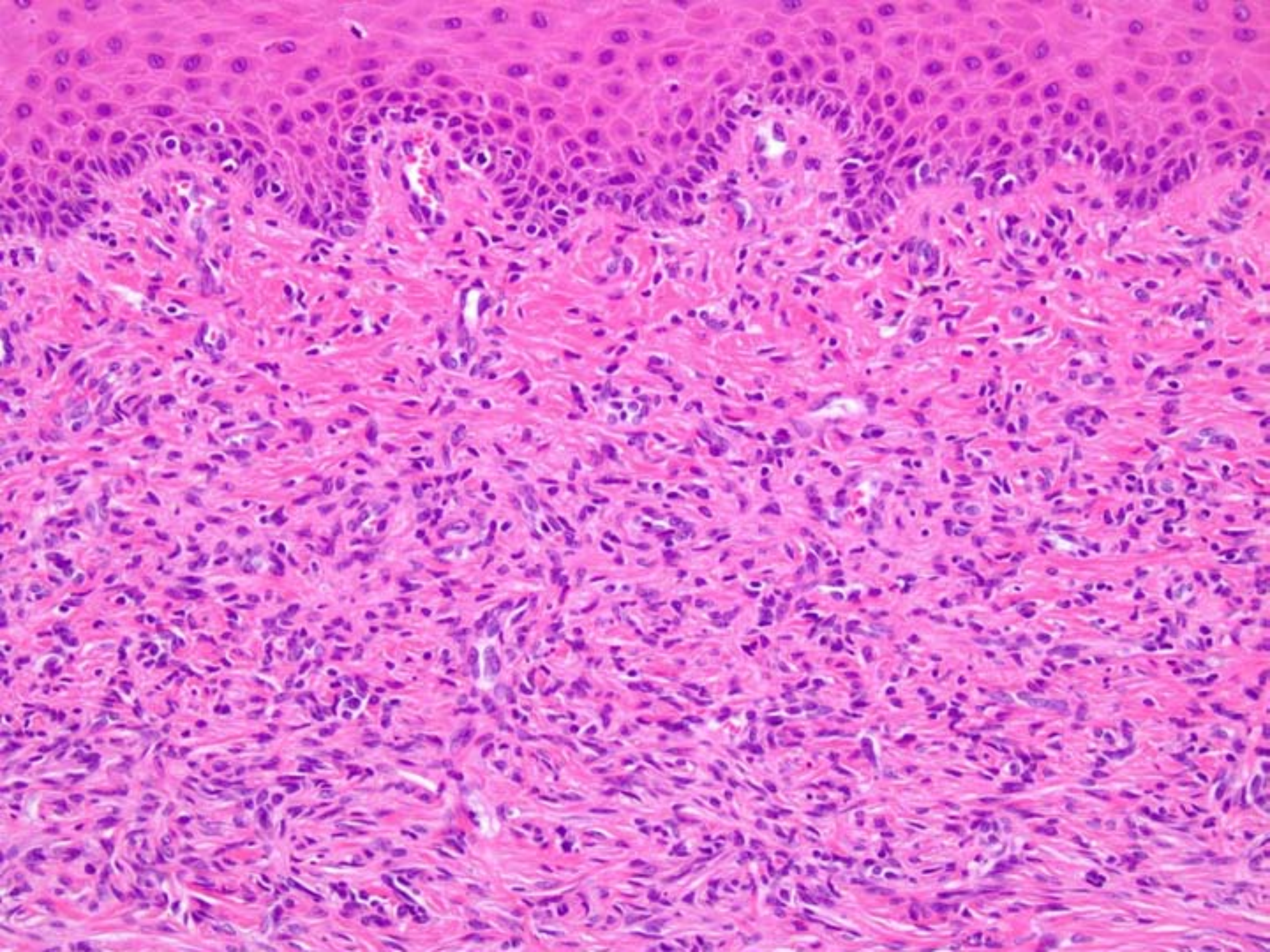
- Histiocytic (lipidized) variant - ↑ lipid, +/- Touton multinucleated giant cells
- Hemosiderotic variant - hemorrhage, increased hemosiderin
- Epithelioid cell histiocytoma - exophytic with stellate epithelioid cells
- Sclerotic fibroma - distinct tumor versus histologic variant controversy

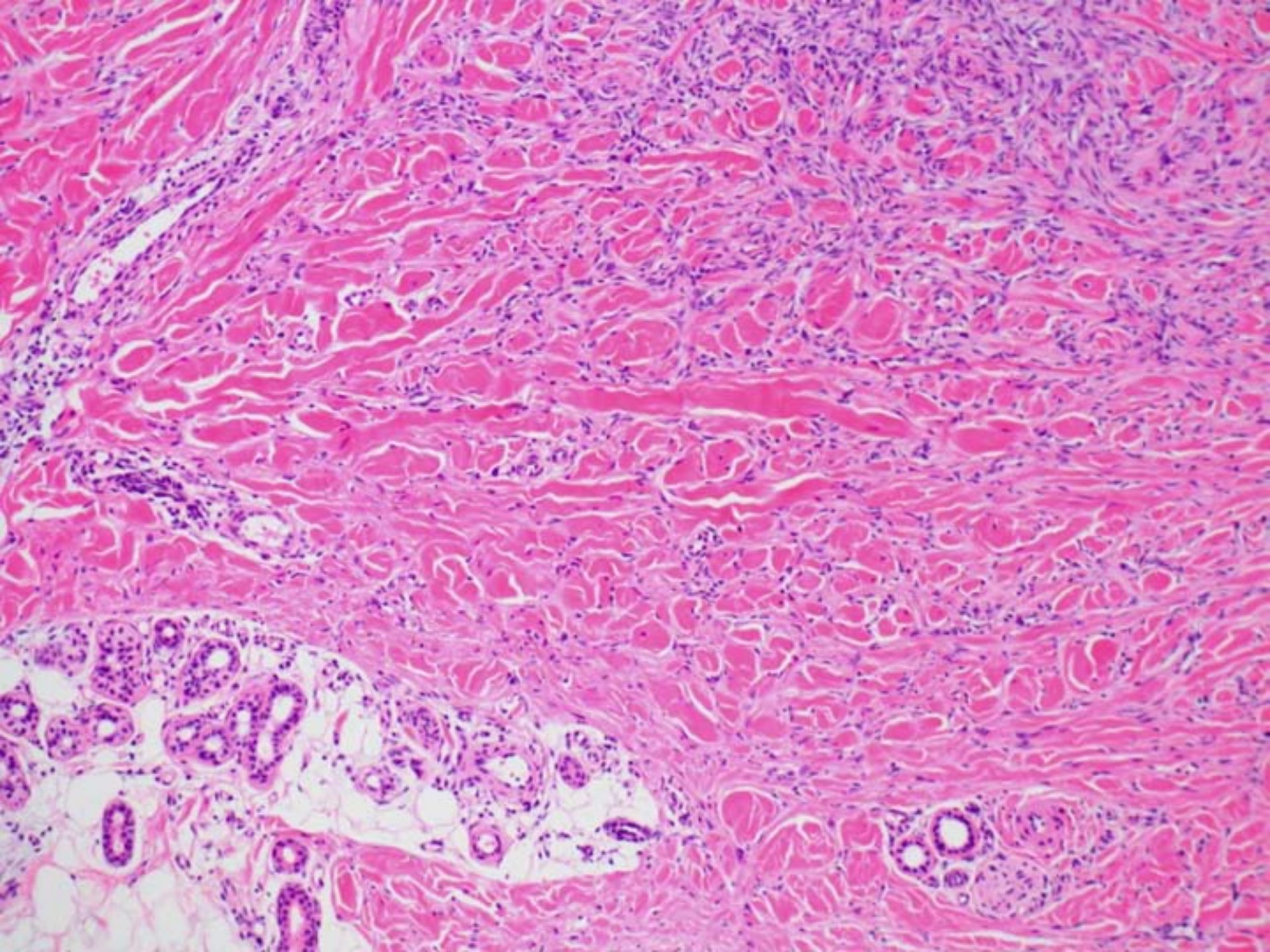


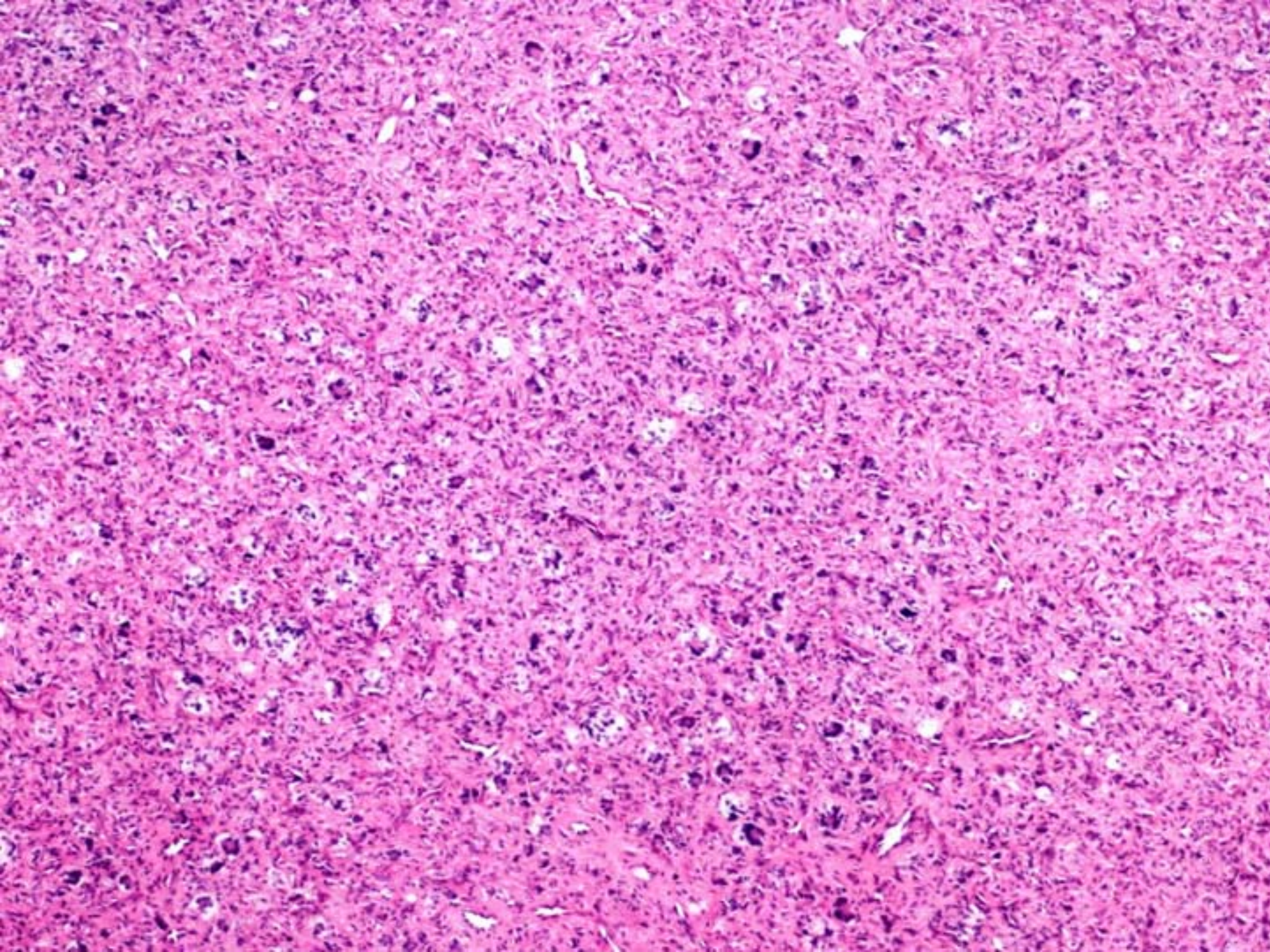


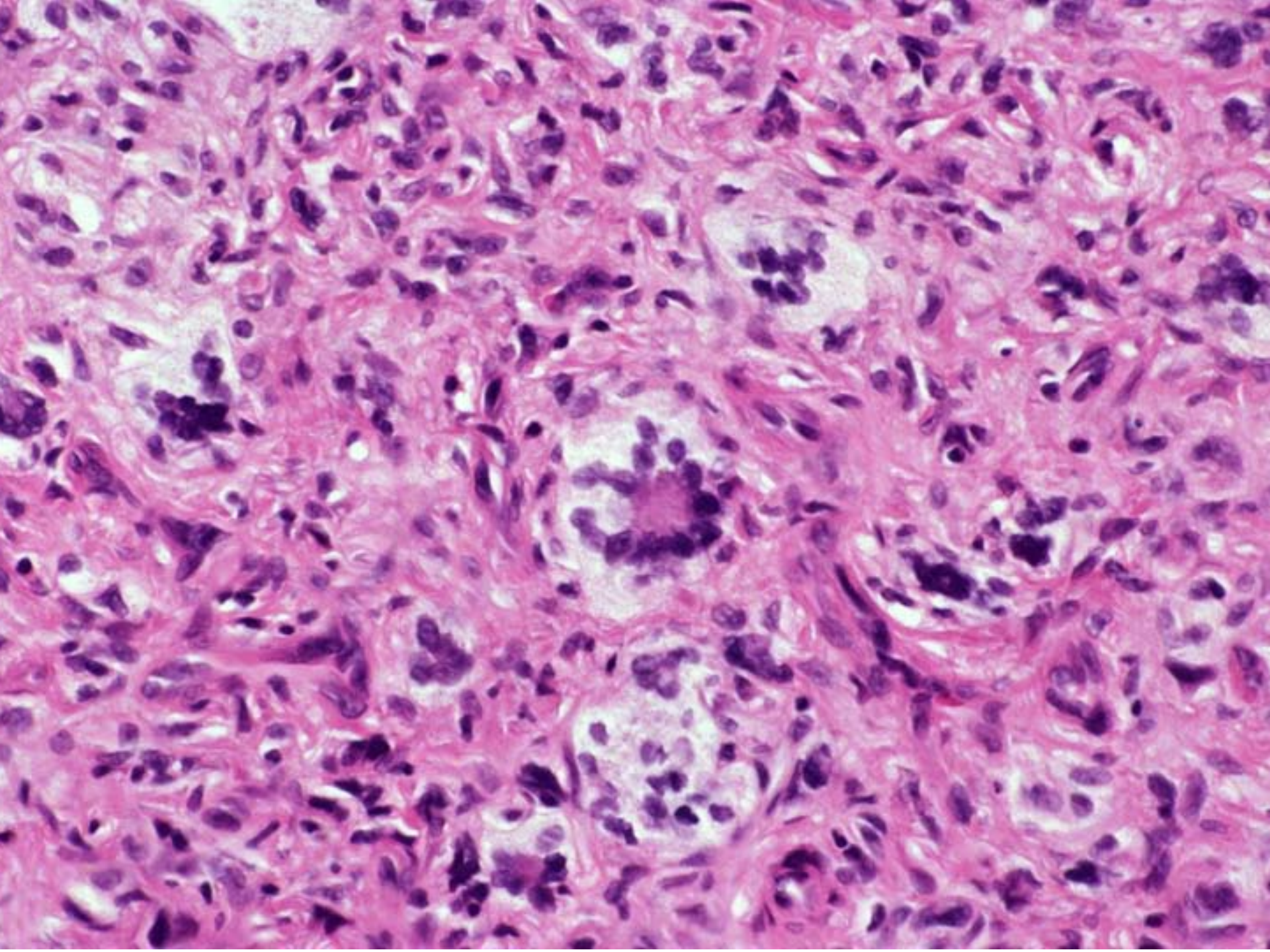


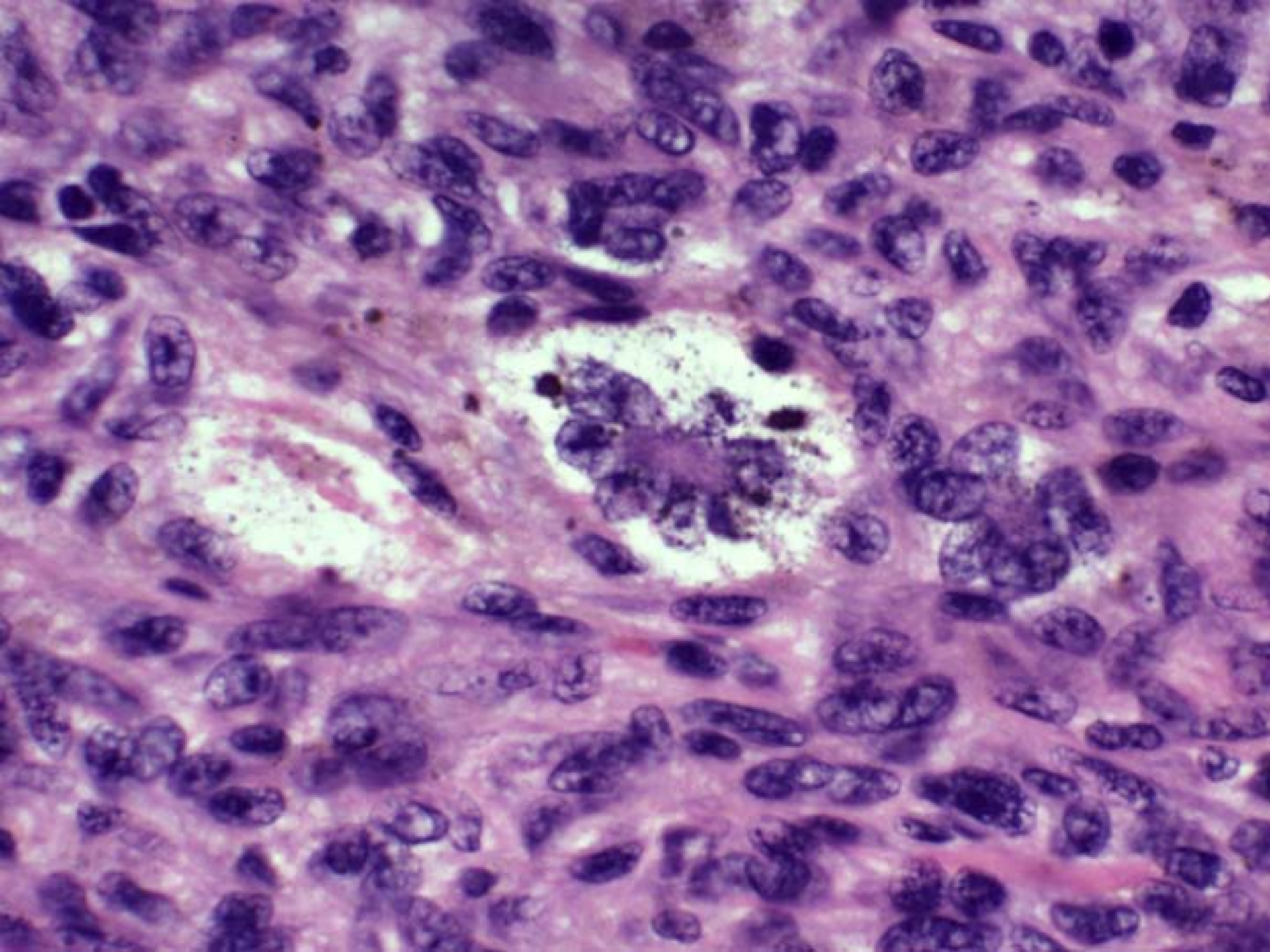


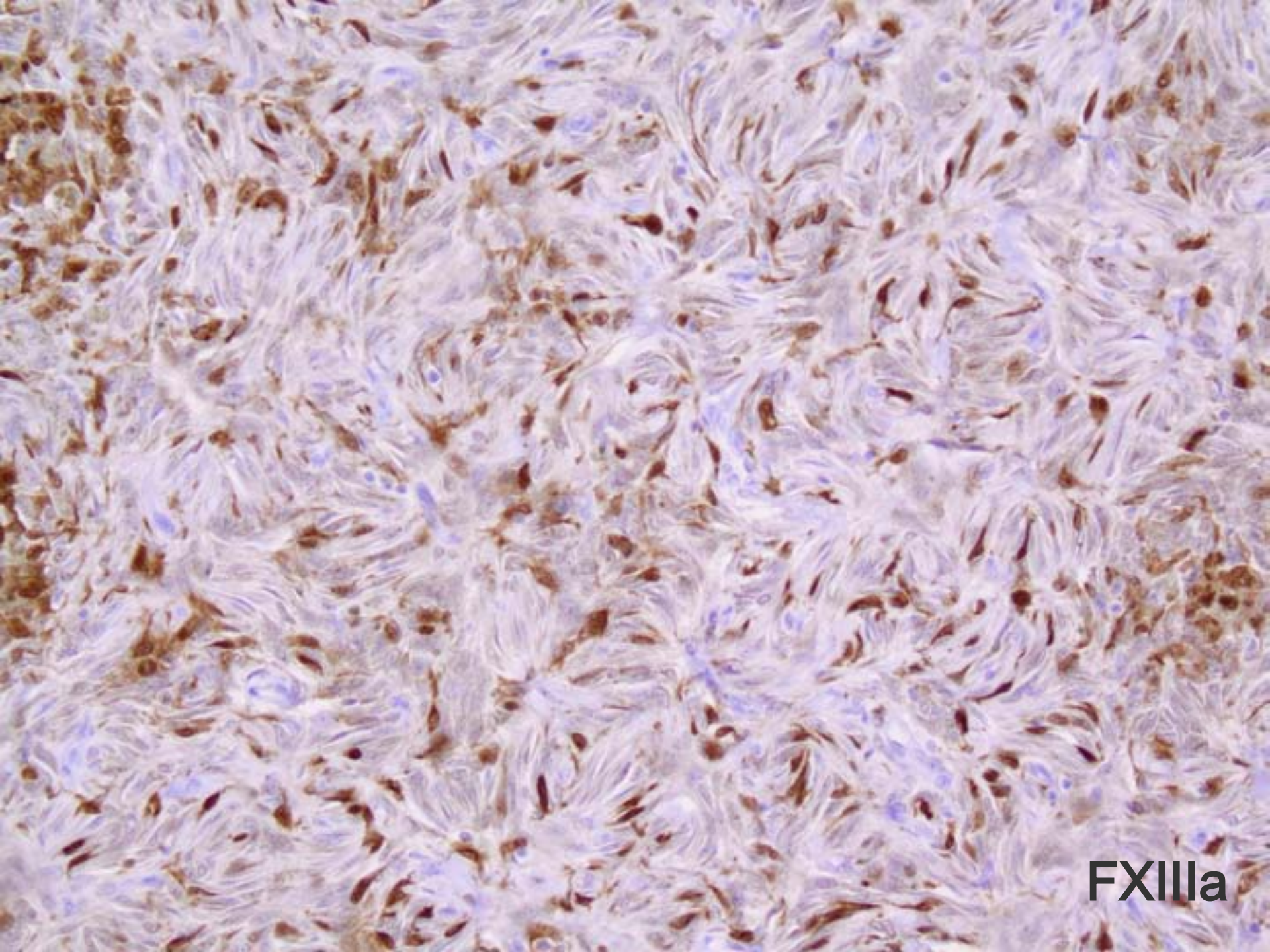




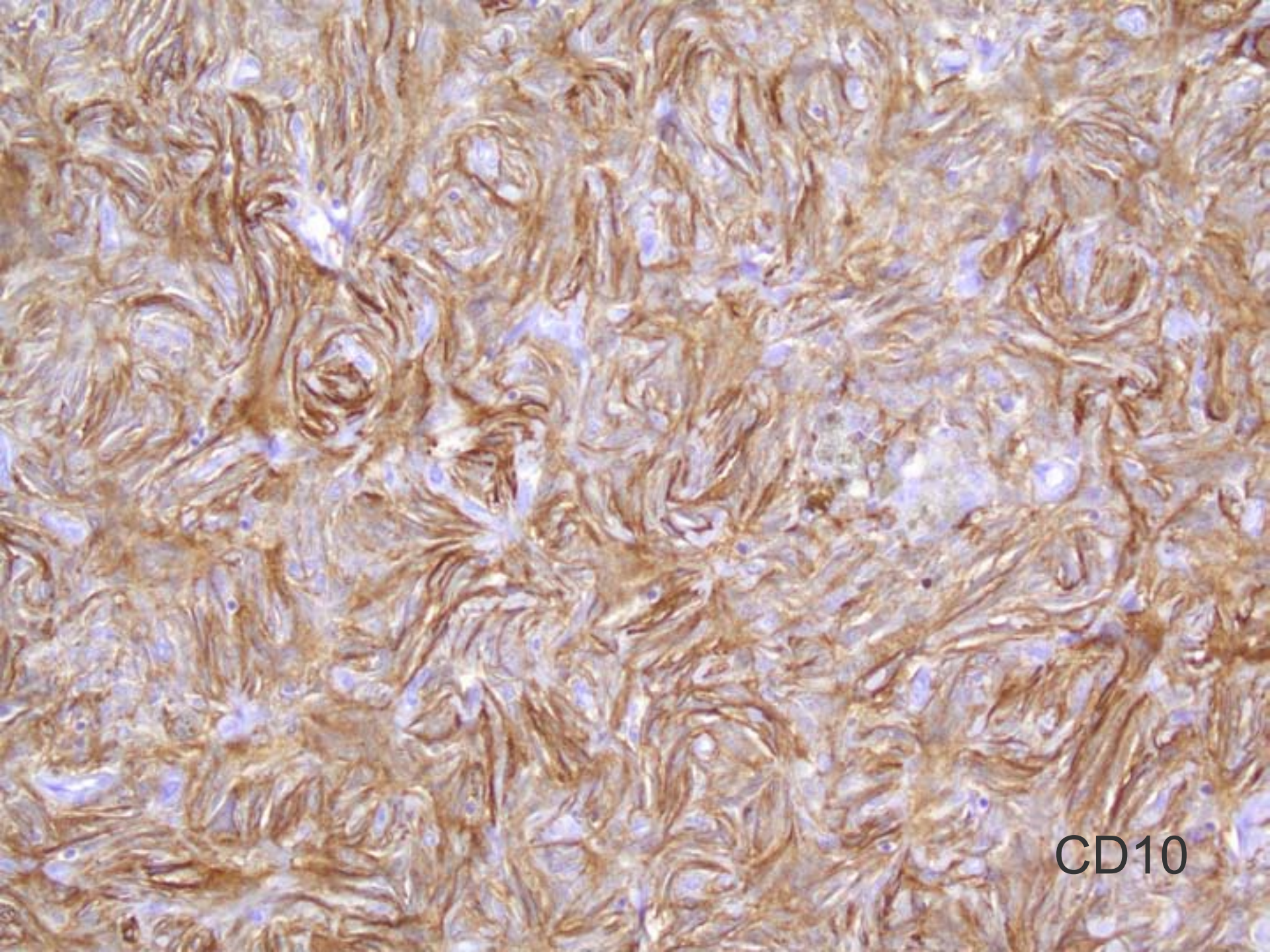








FXIIIa



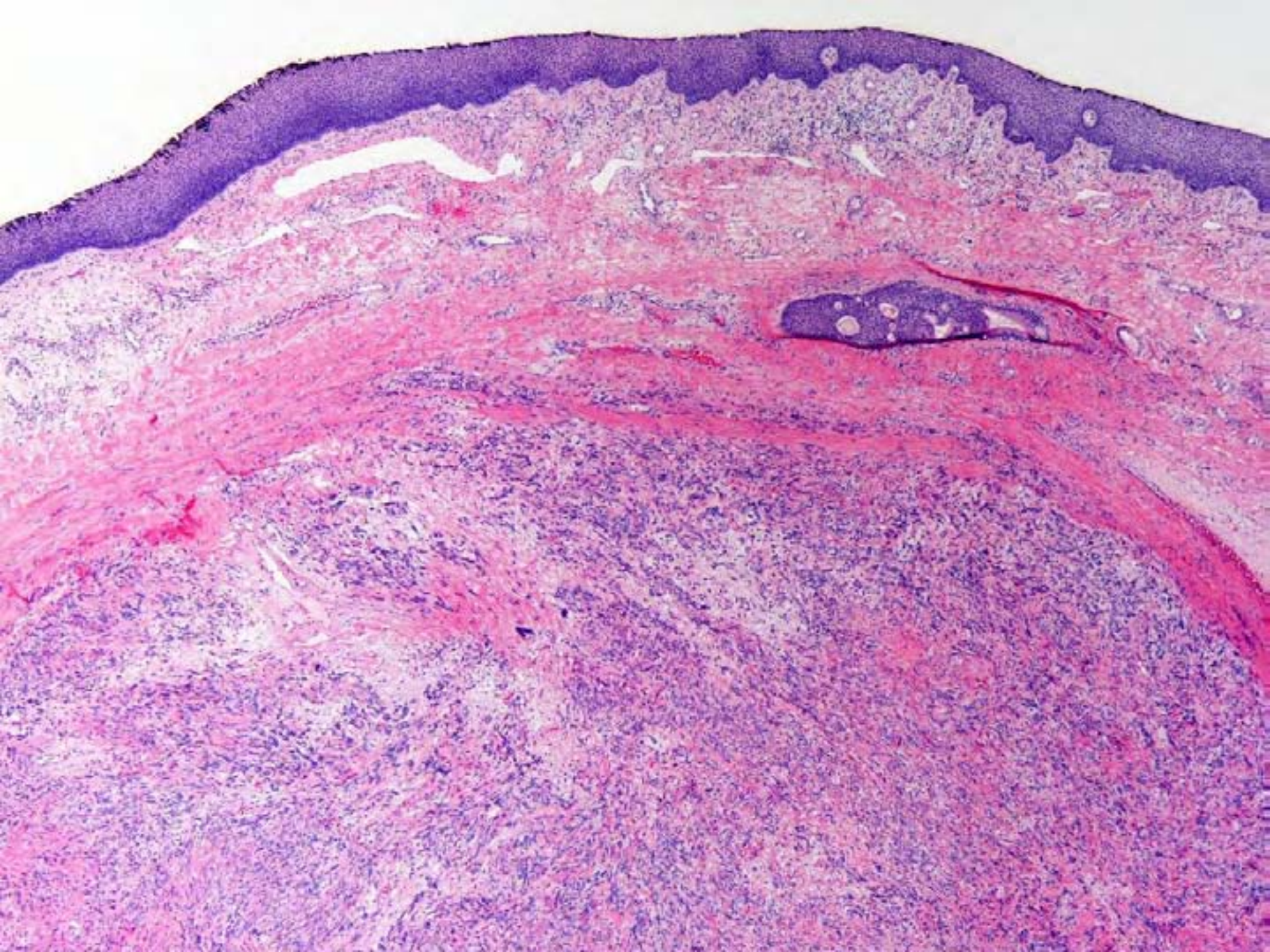
CD10

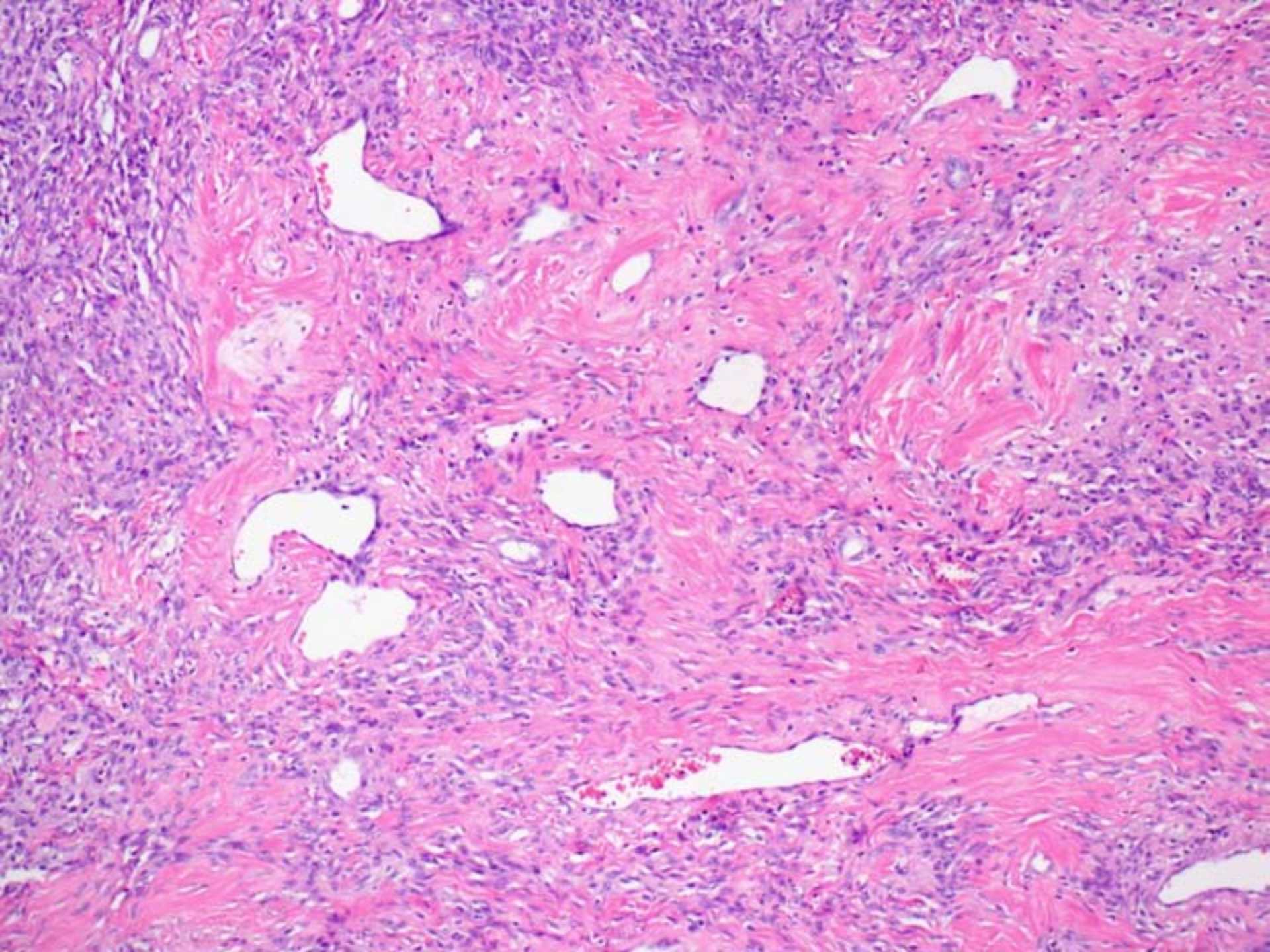
SOLITARY FIBROUS TUMOR

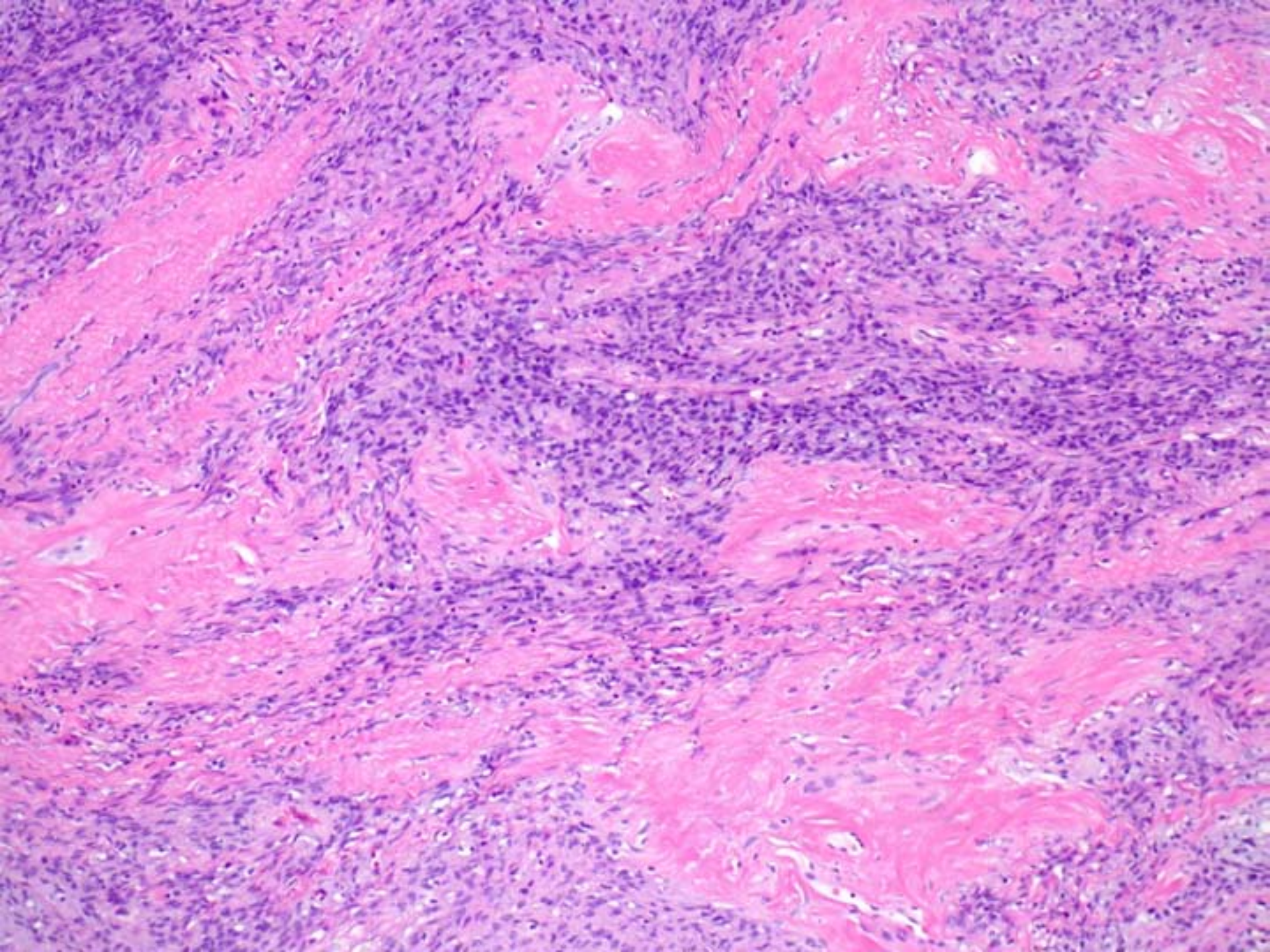
- Mesenchymal tumor originally described in pleural space
 - numerous extrapleural sites reported
- Rare tumor
- Typically middle-aged adults
- Site - primarily head & neck, extremities
- Slow-growing subcutaneous mass
- Malignant transformation in 10%

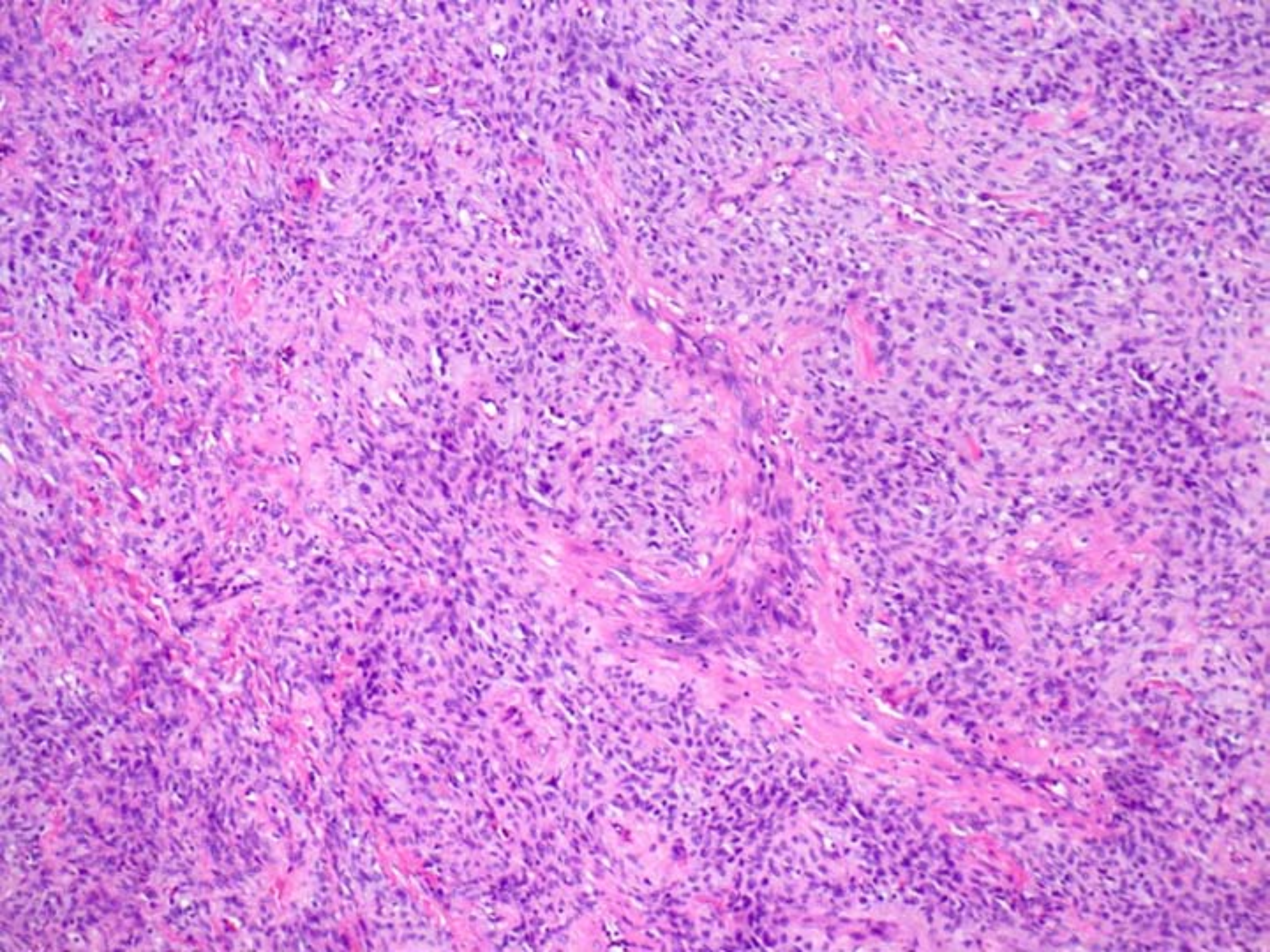
SOLITARY FIBROUS TUMOR

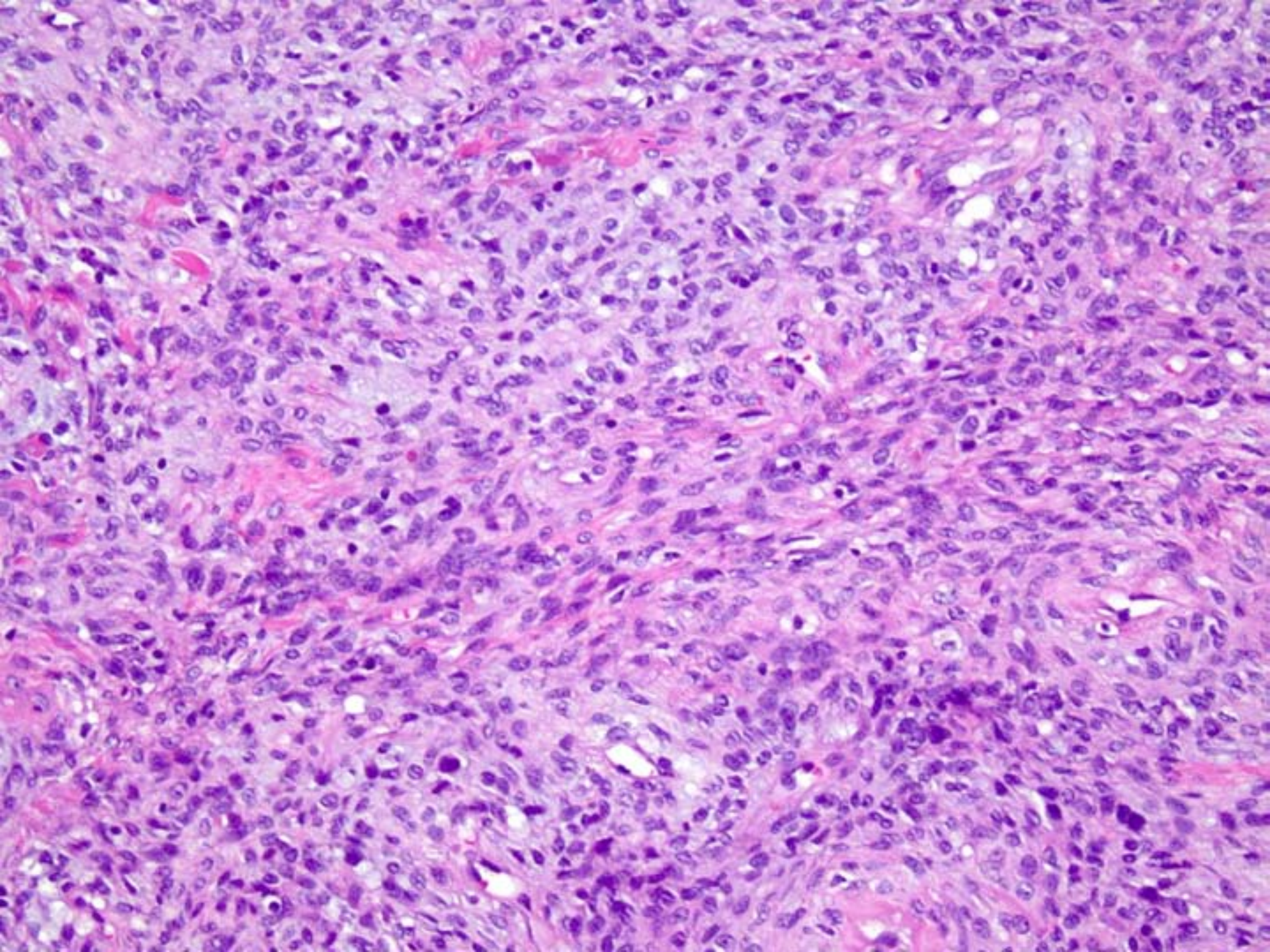
- Well-circumscribed, not encapsulated
- “Patternless Pattern Architecture”
 - alternating hypo and hypercellular areas
 - hyalinized collagen and keloidal fibers
 - myxoid changes & fibrosis variably present
- Branching vessels, round to spindle cells
- Variable numbers of plasma cells, mast cells, lipocytes, MNGC, CD34+

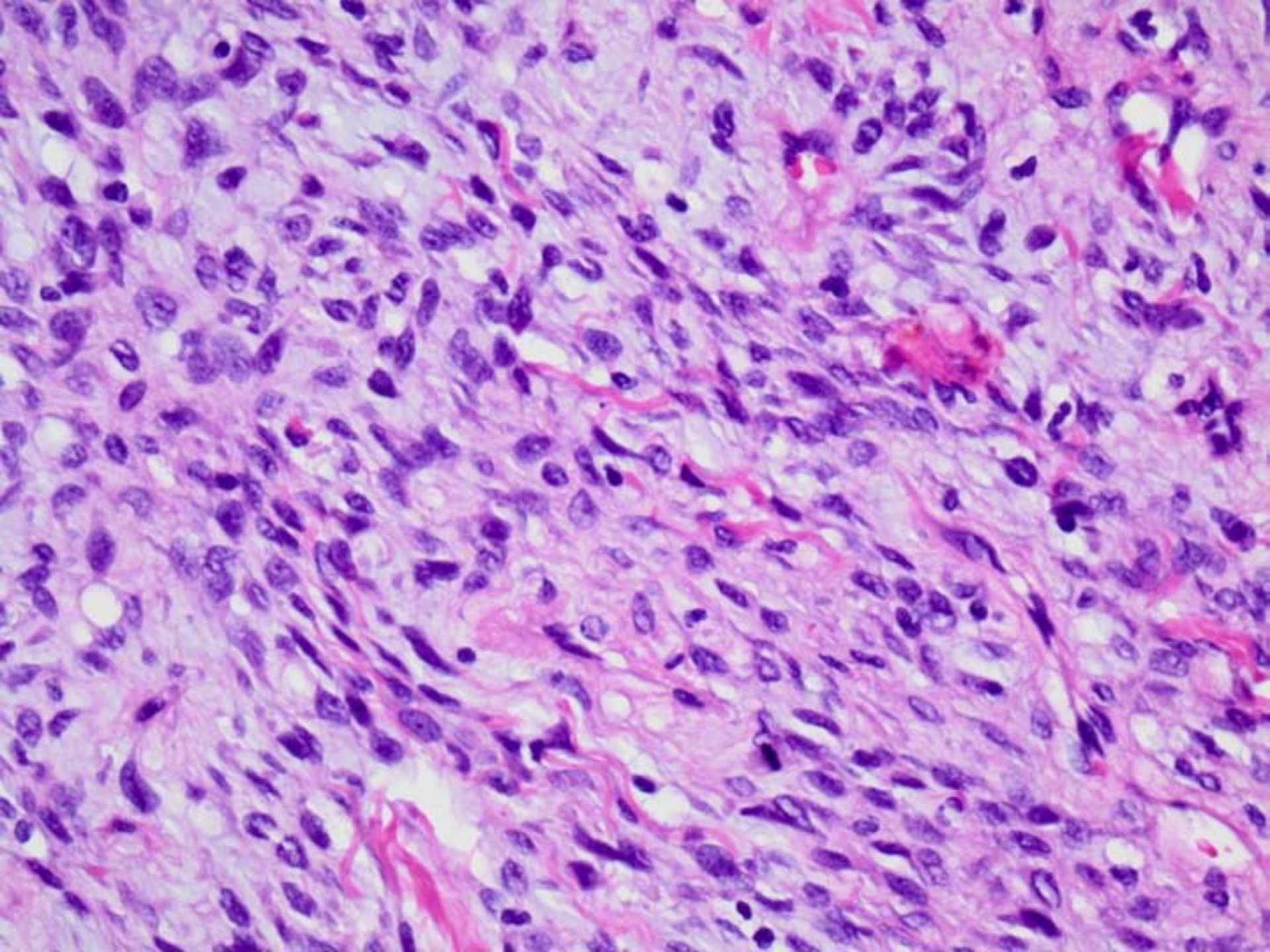










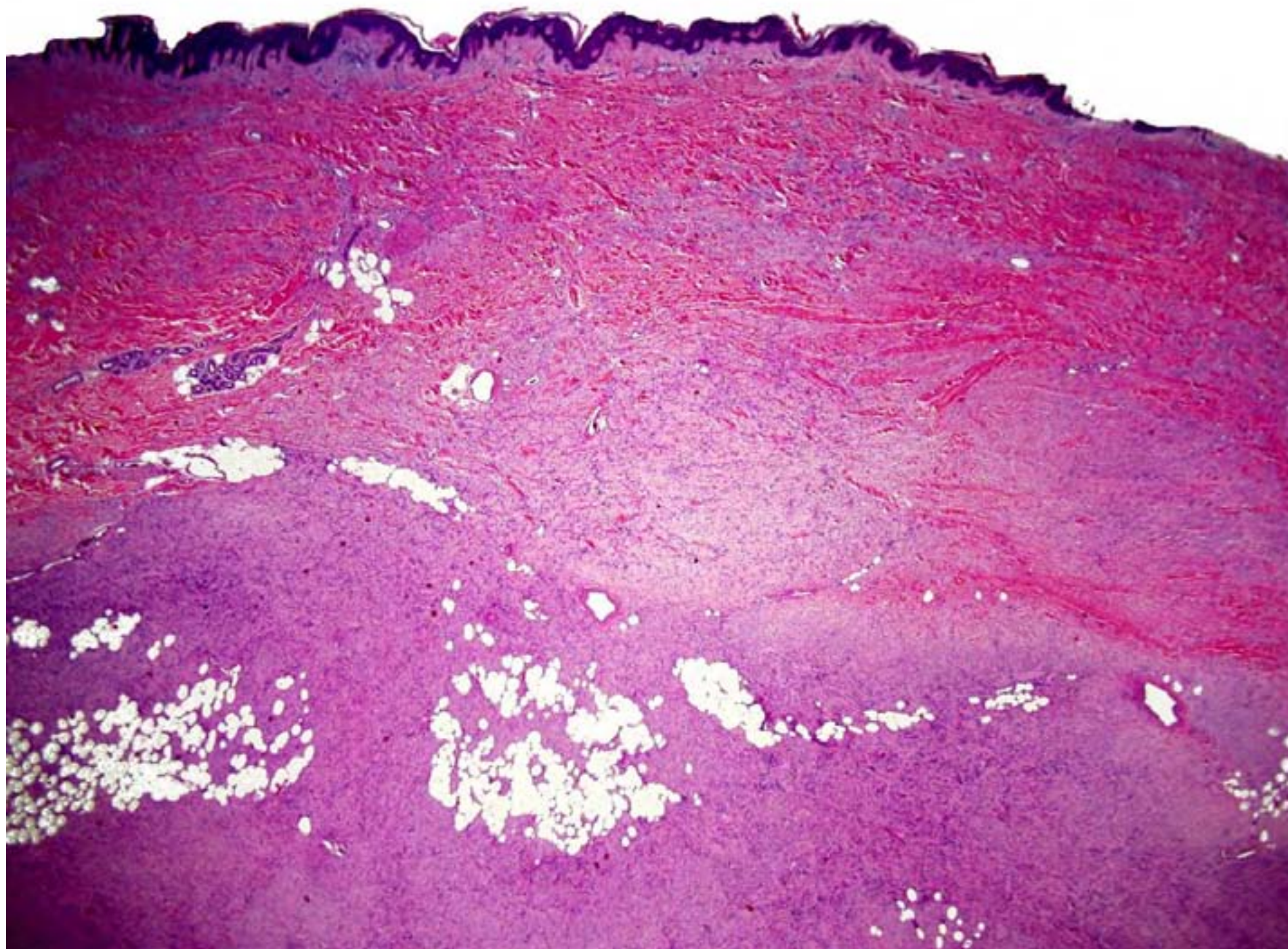


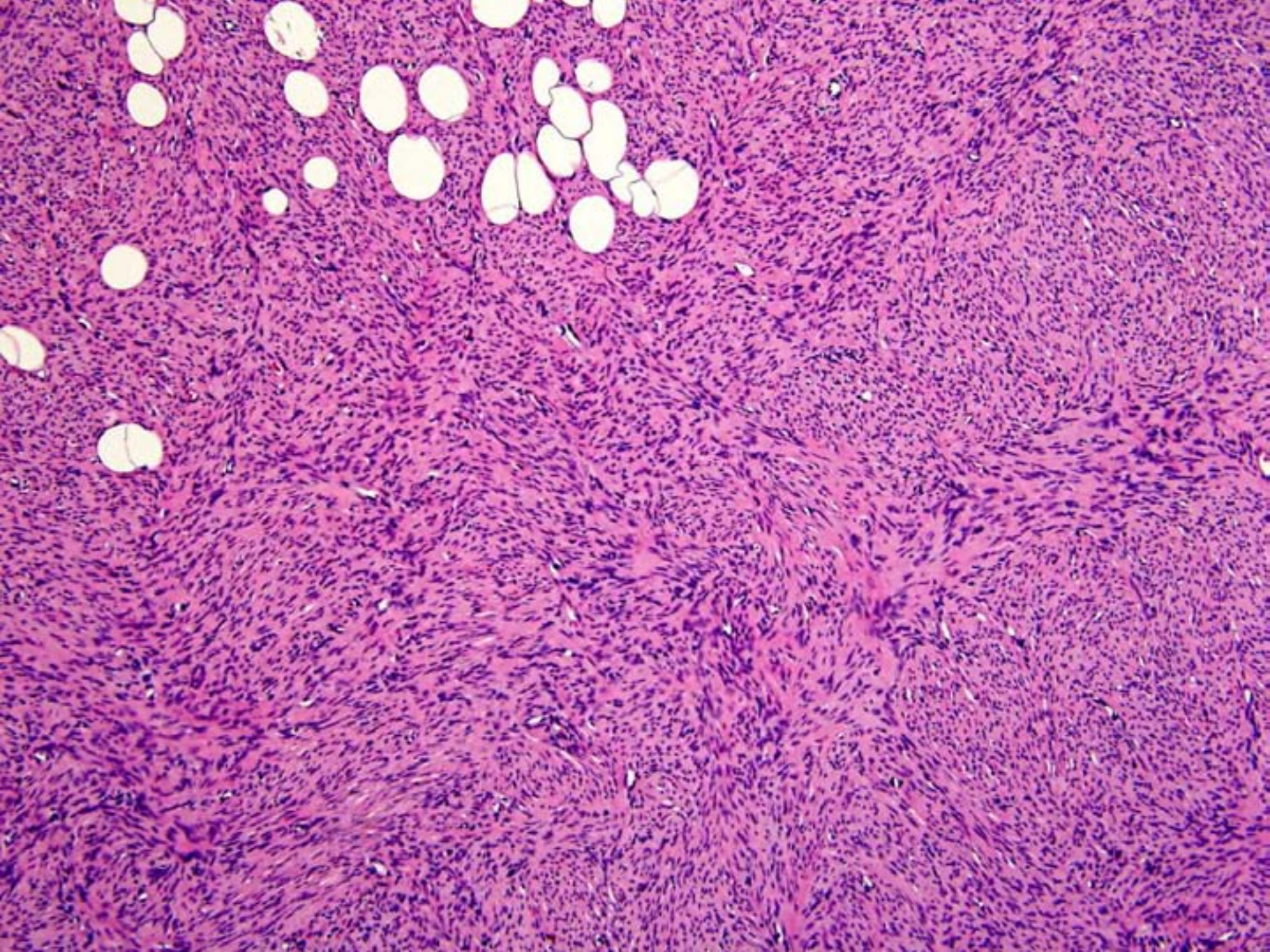
DERMATOFIBROSARCOMA PROTUBERANS

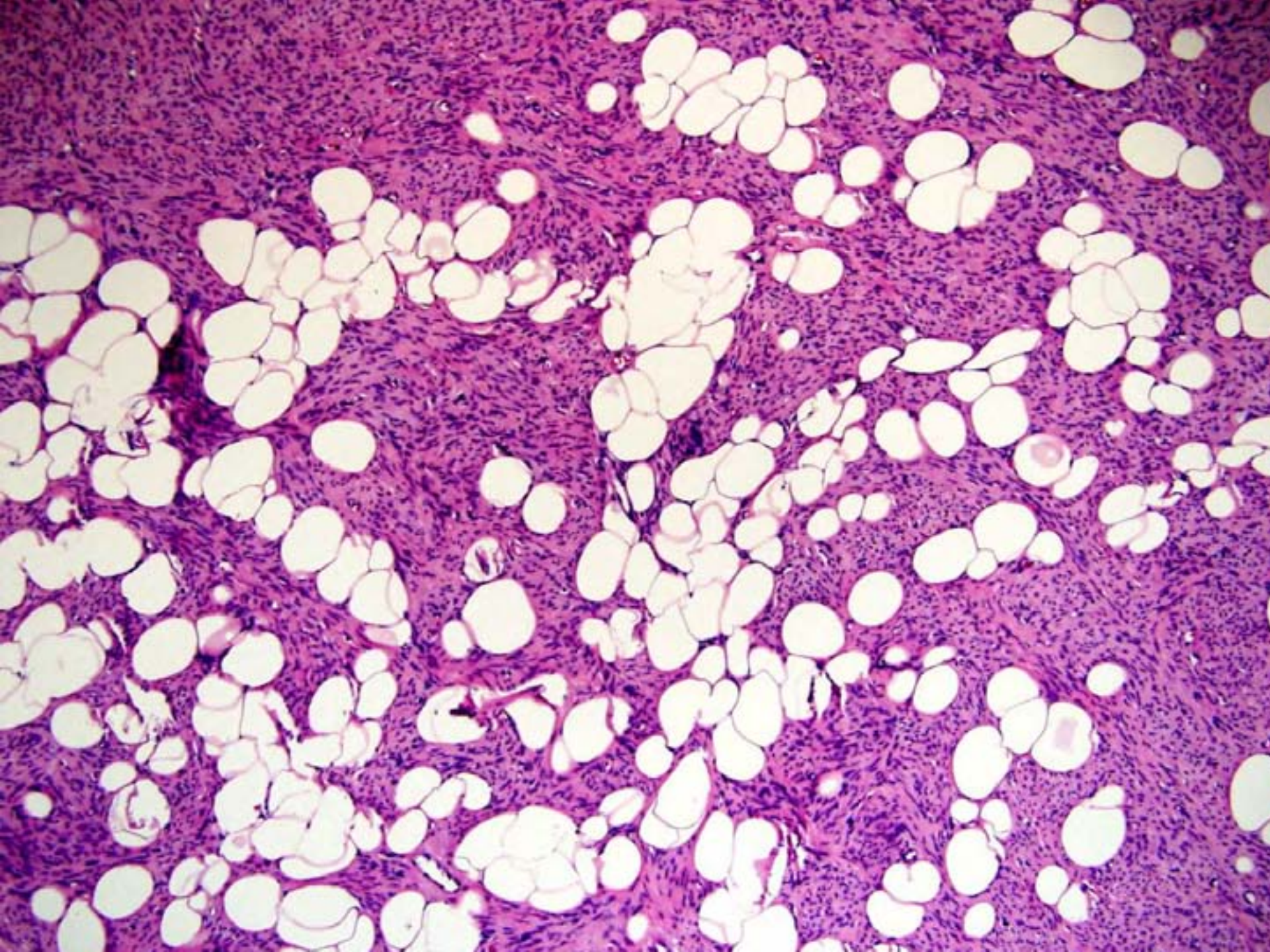
- Rare tumor (0.1% of all malignant neoplasms), but #1 soft tissue sarcoma
- Middle-aged persons, trunk and extremities
- Only 40% of cases are protuberant
- Atrophic variants can be particularly vexing
- Dense spindled cell proliferation with generally strong CD34+
- >90% with t(17;22) COL1A/PDGF translocation

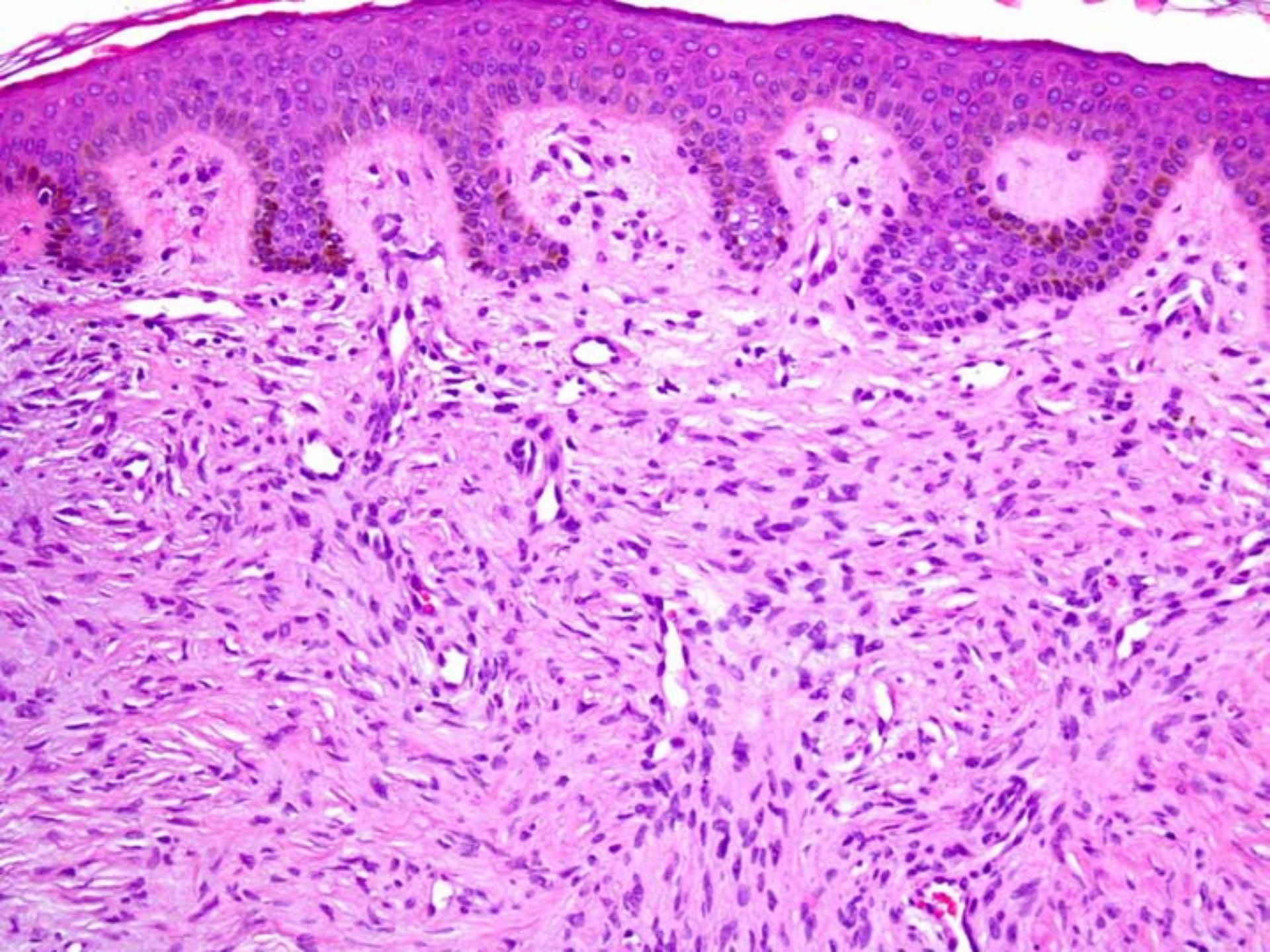
DERMATOFIBROSARCOMA PROTUBERANS

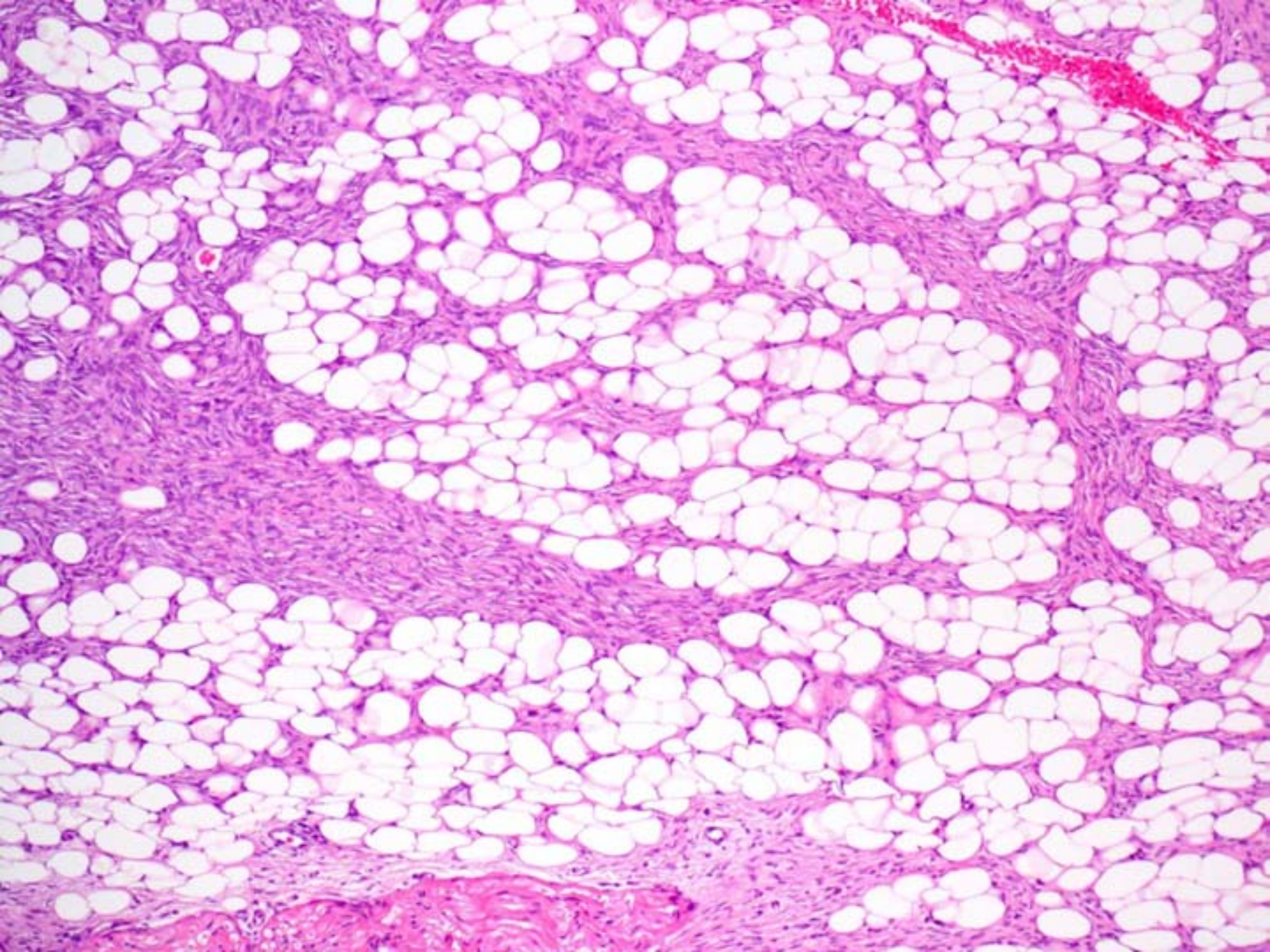


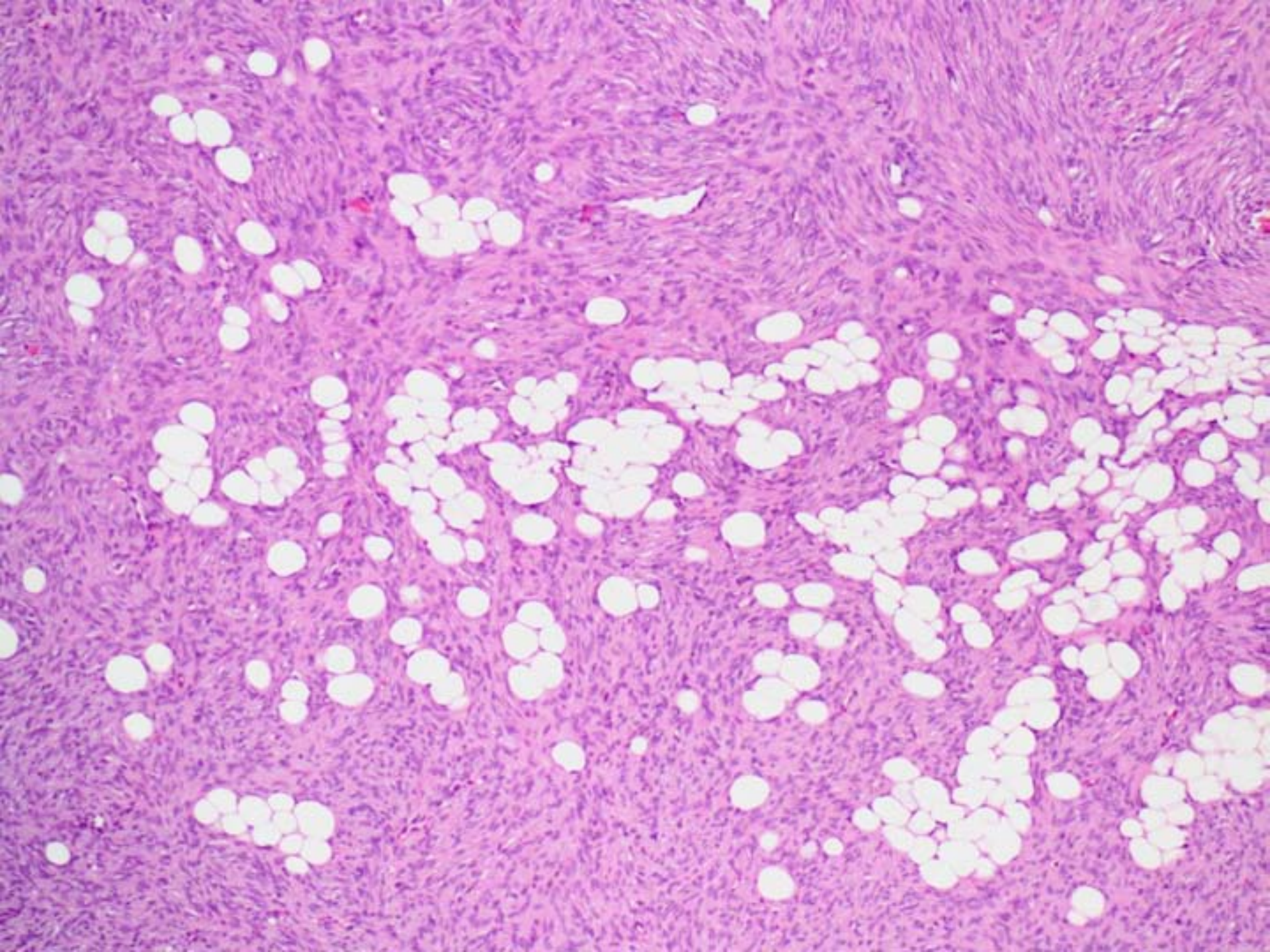


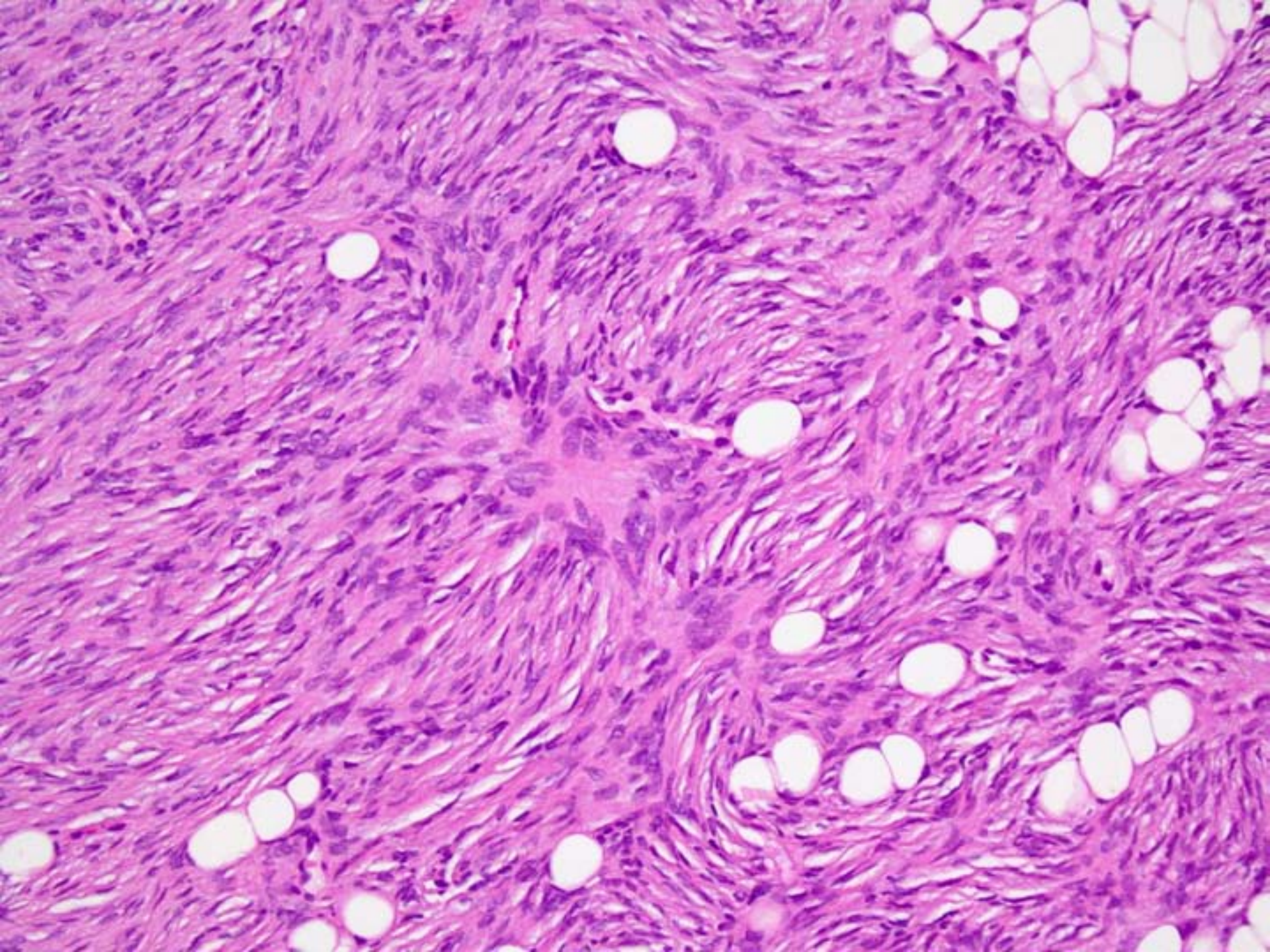


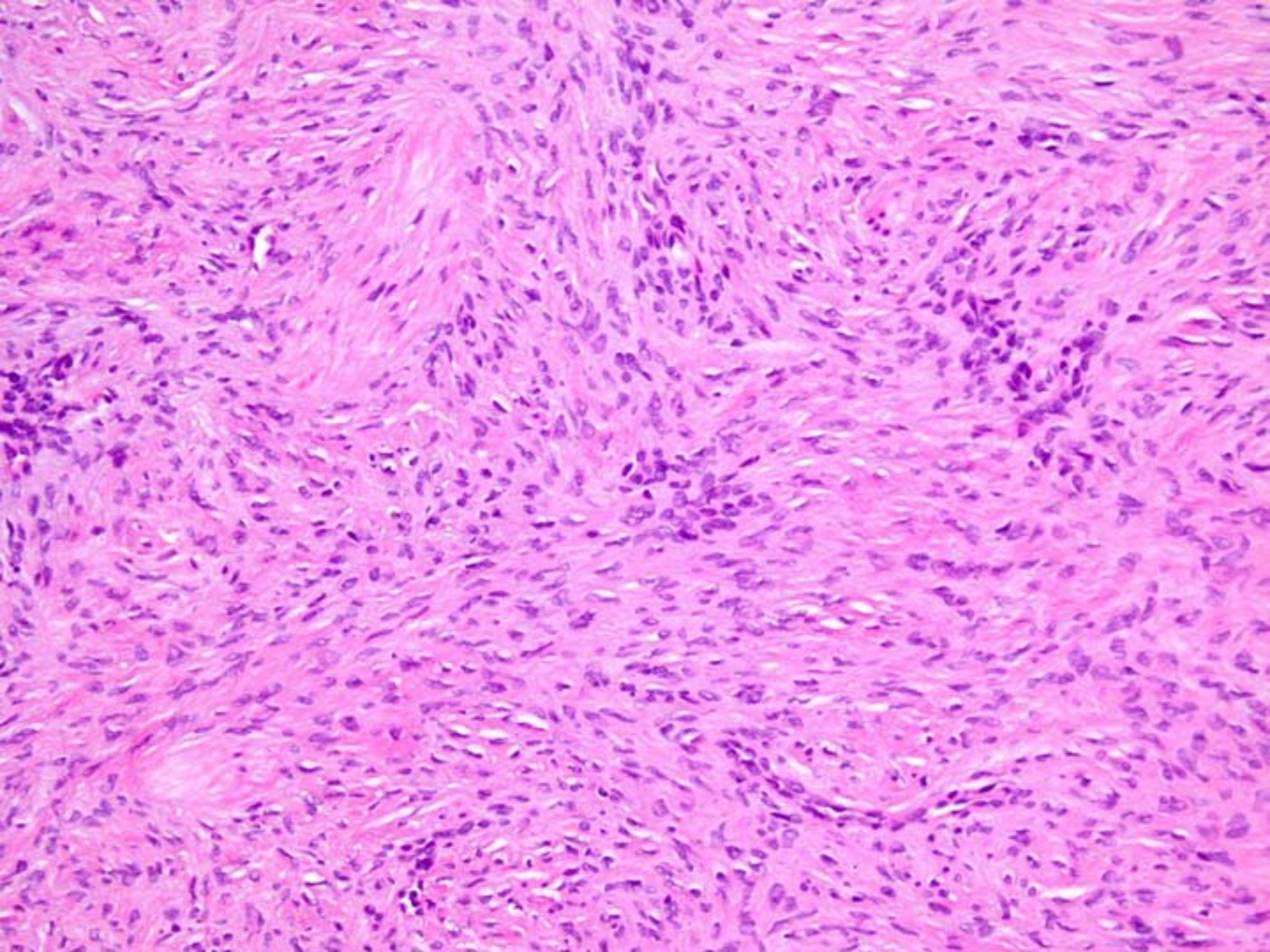




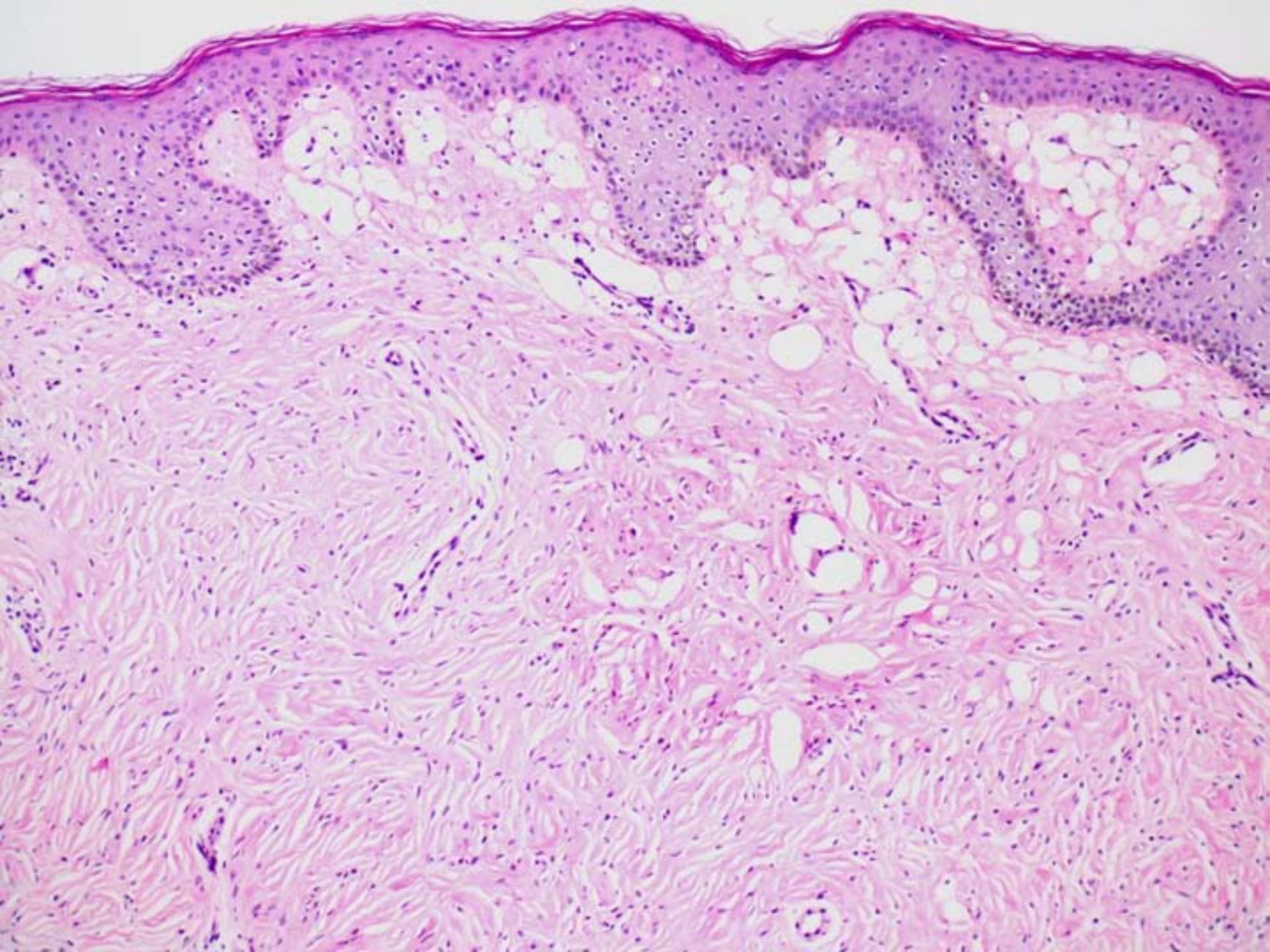


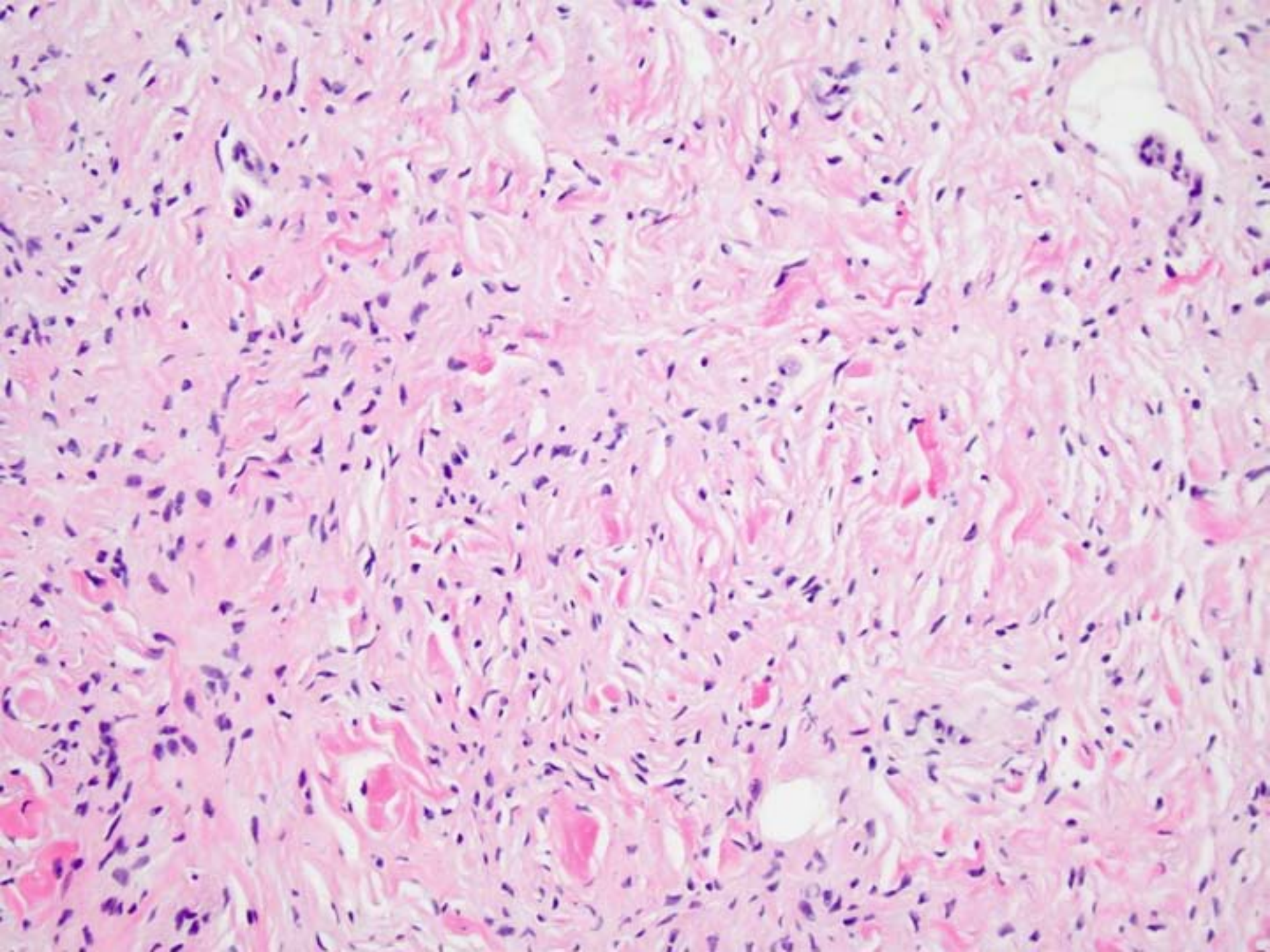


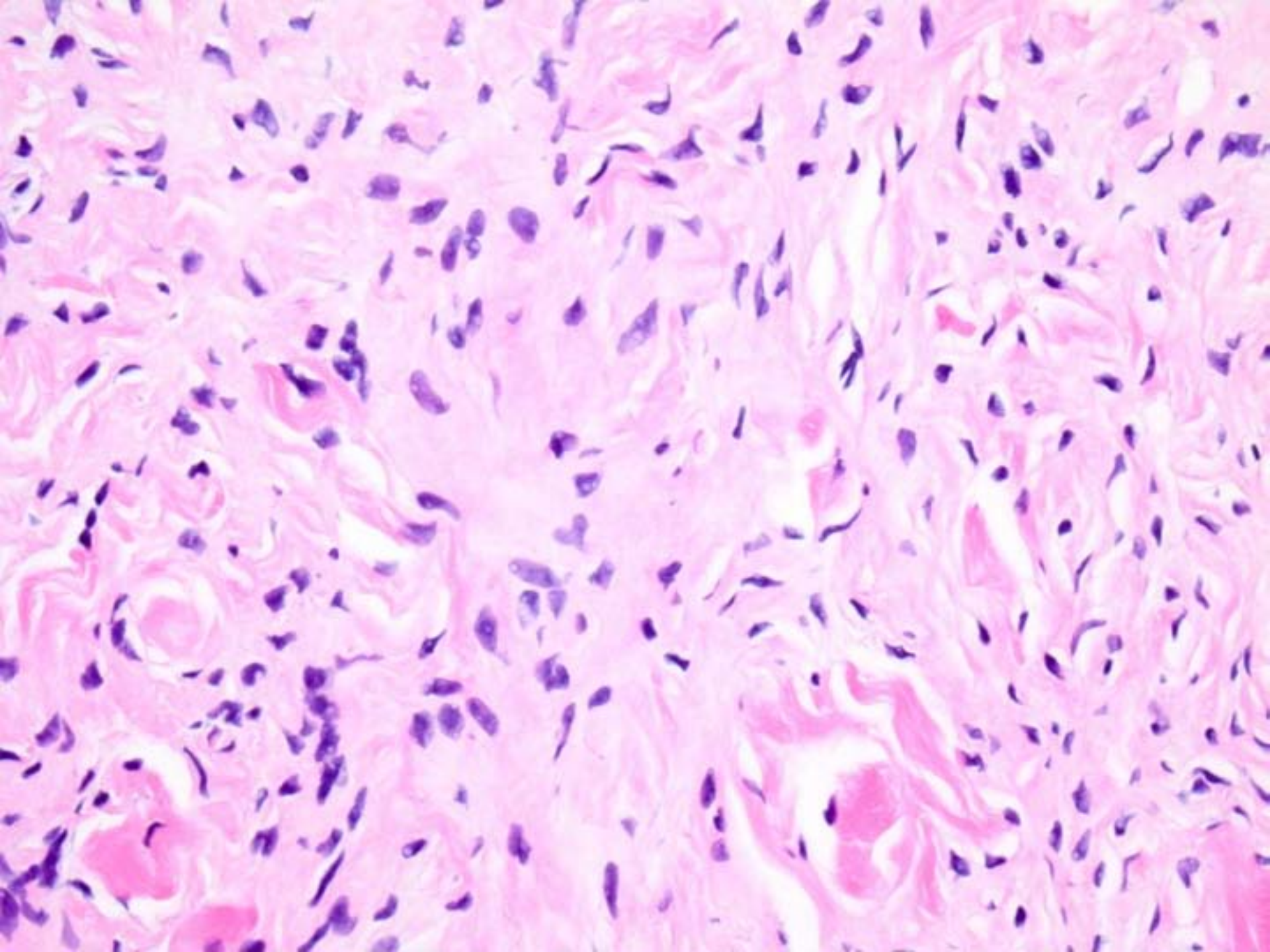








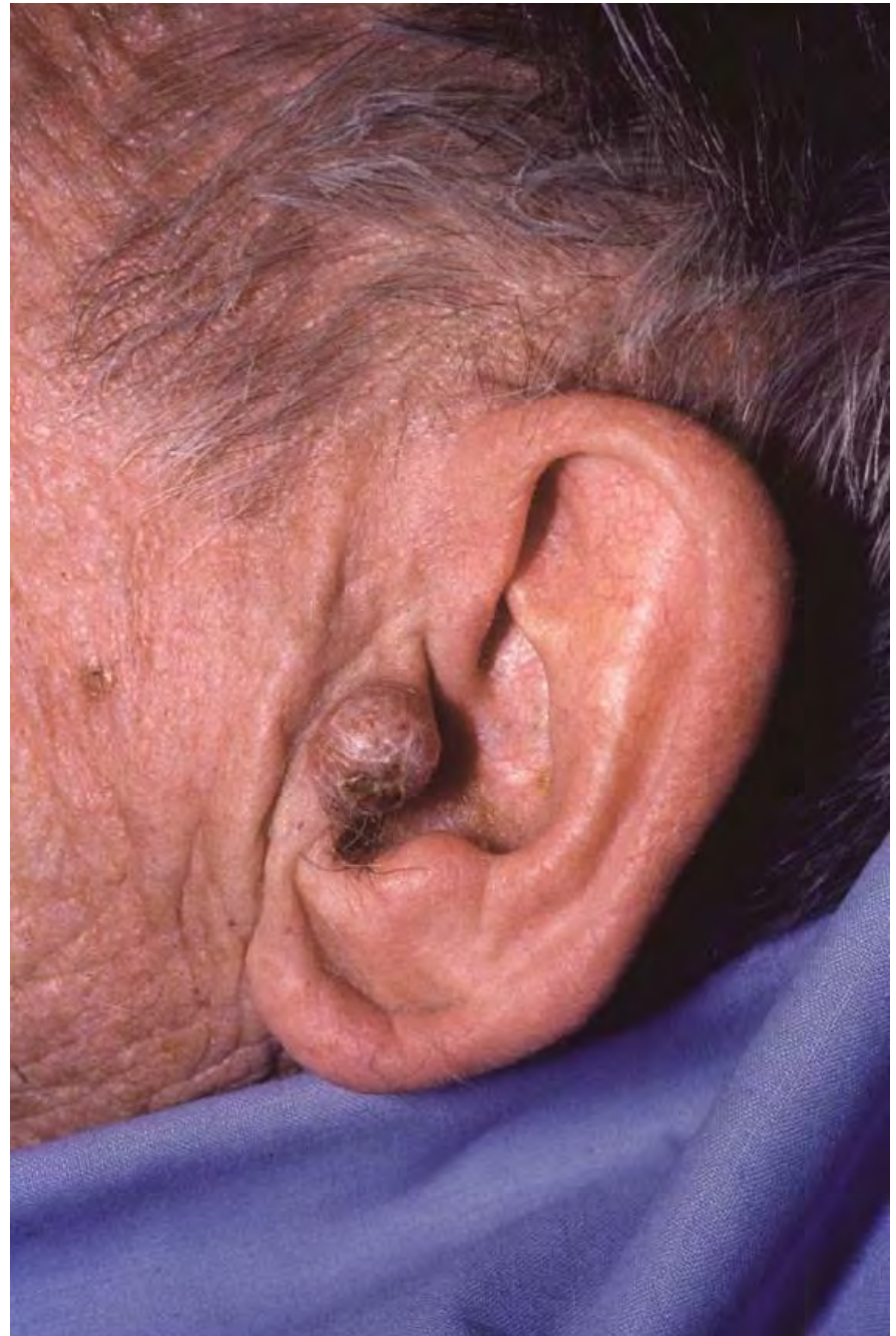


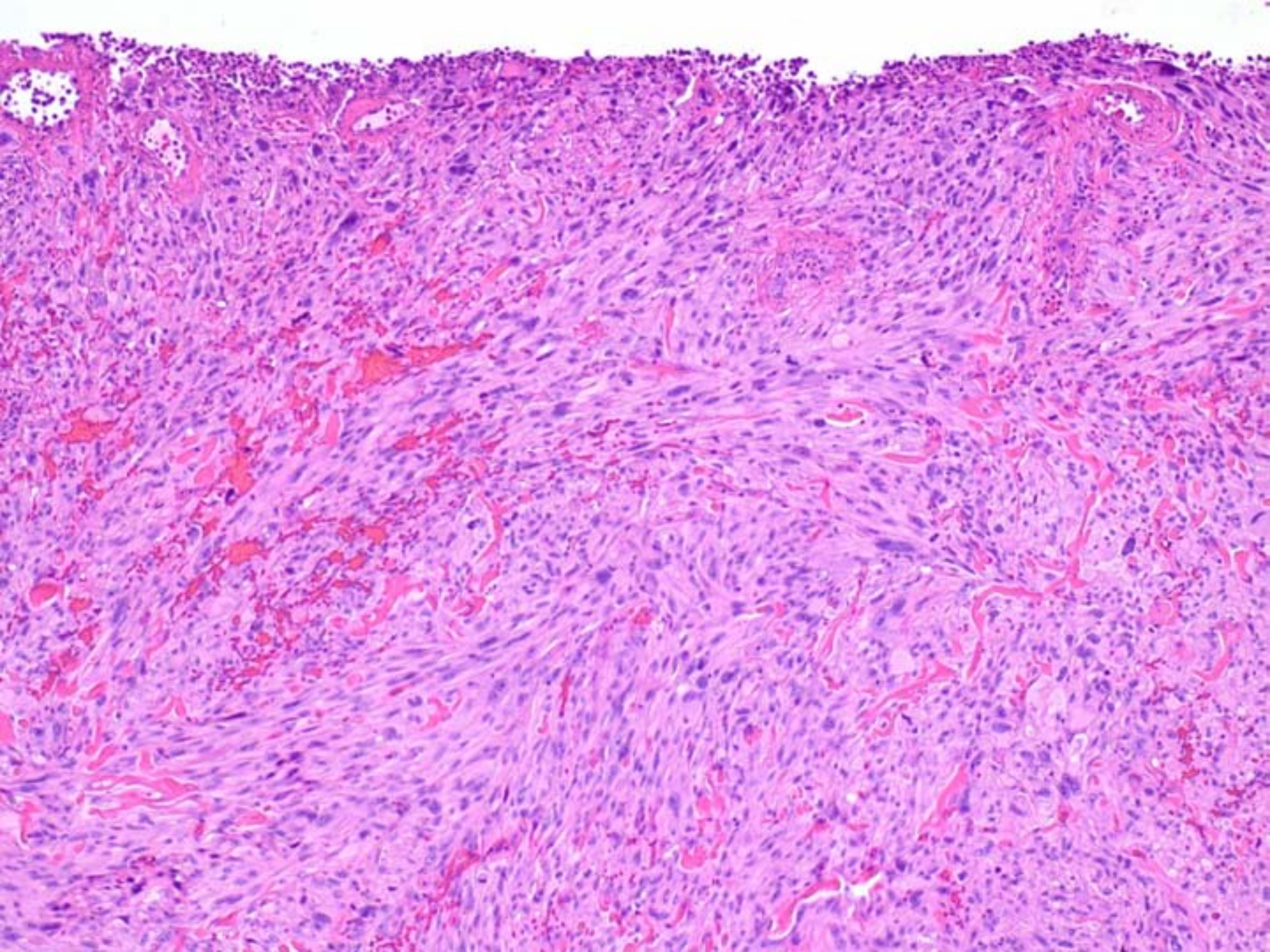


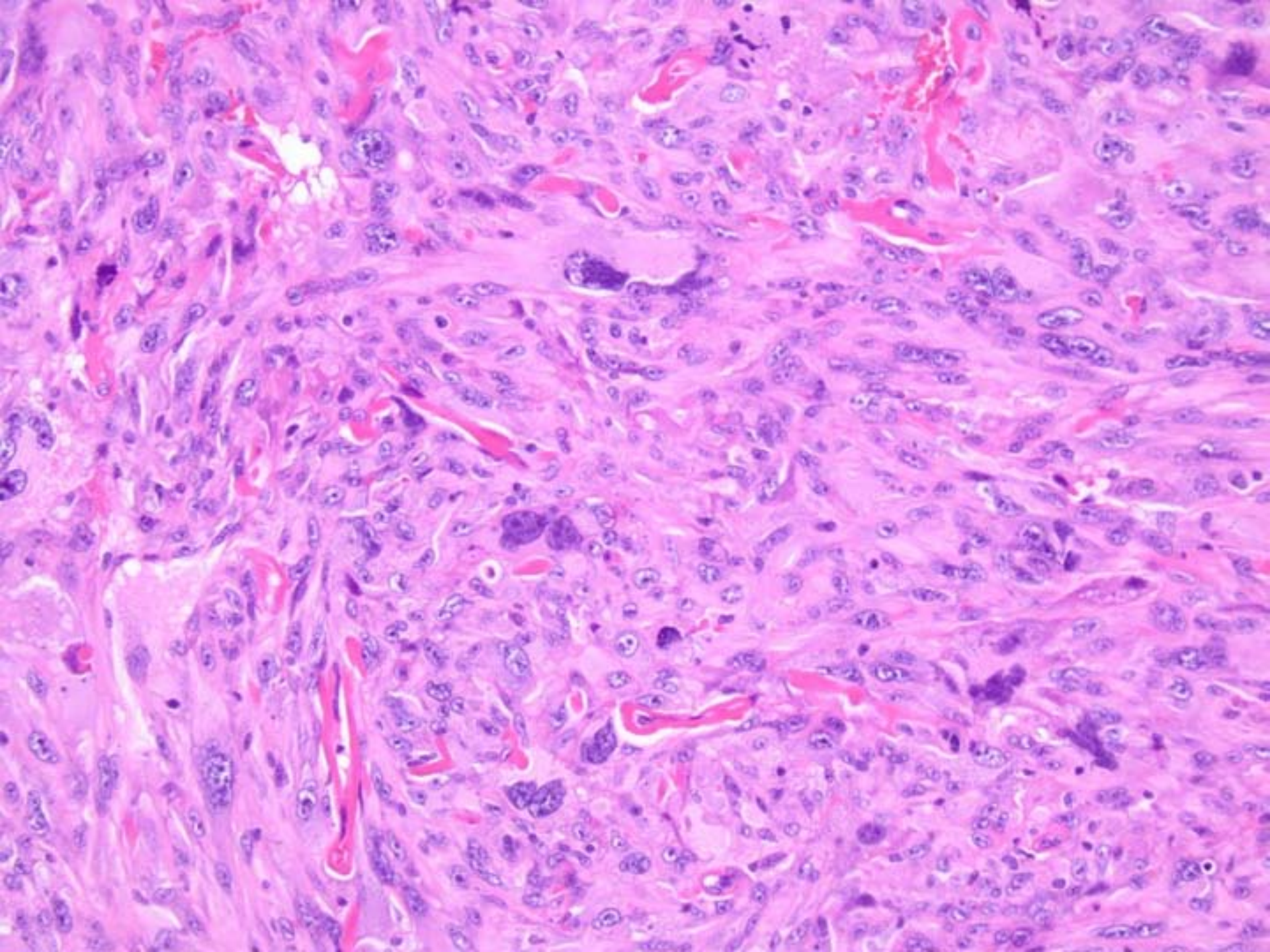
ATYPICAL FIBROXANTHOMA

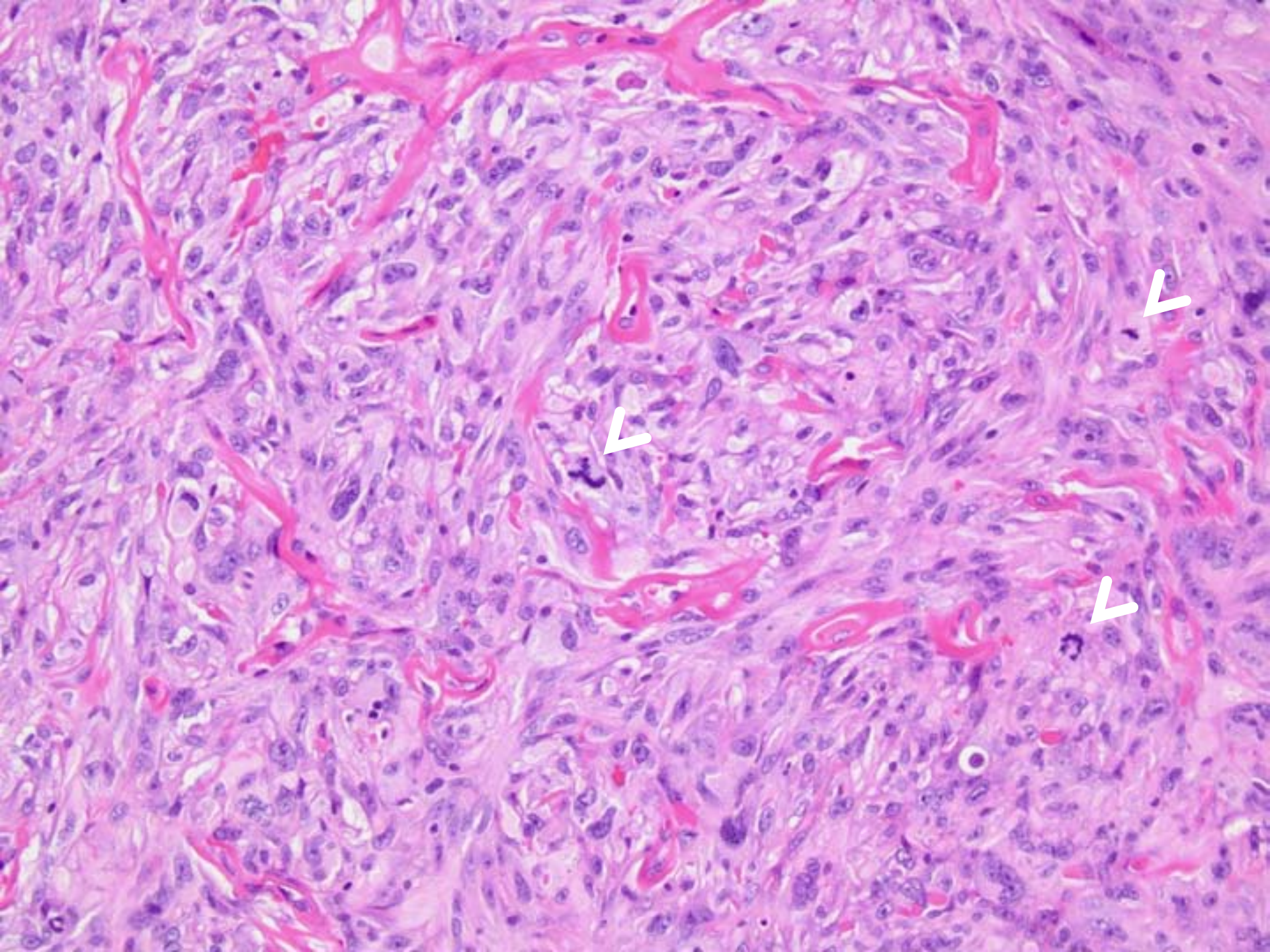
- Coined at AFIP in 1963
- Heavily sun-damaged skin of elderly
- Some low grade malignant behavior observed
- Aypically atypical
 (“too malignant to be malignant”)
- Immunostains employed
 - CD68, CD10, Procollagen I, P53

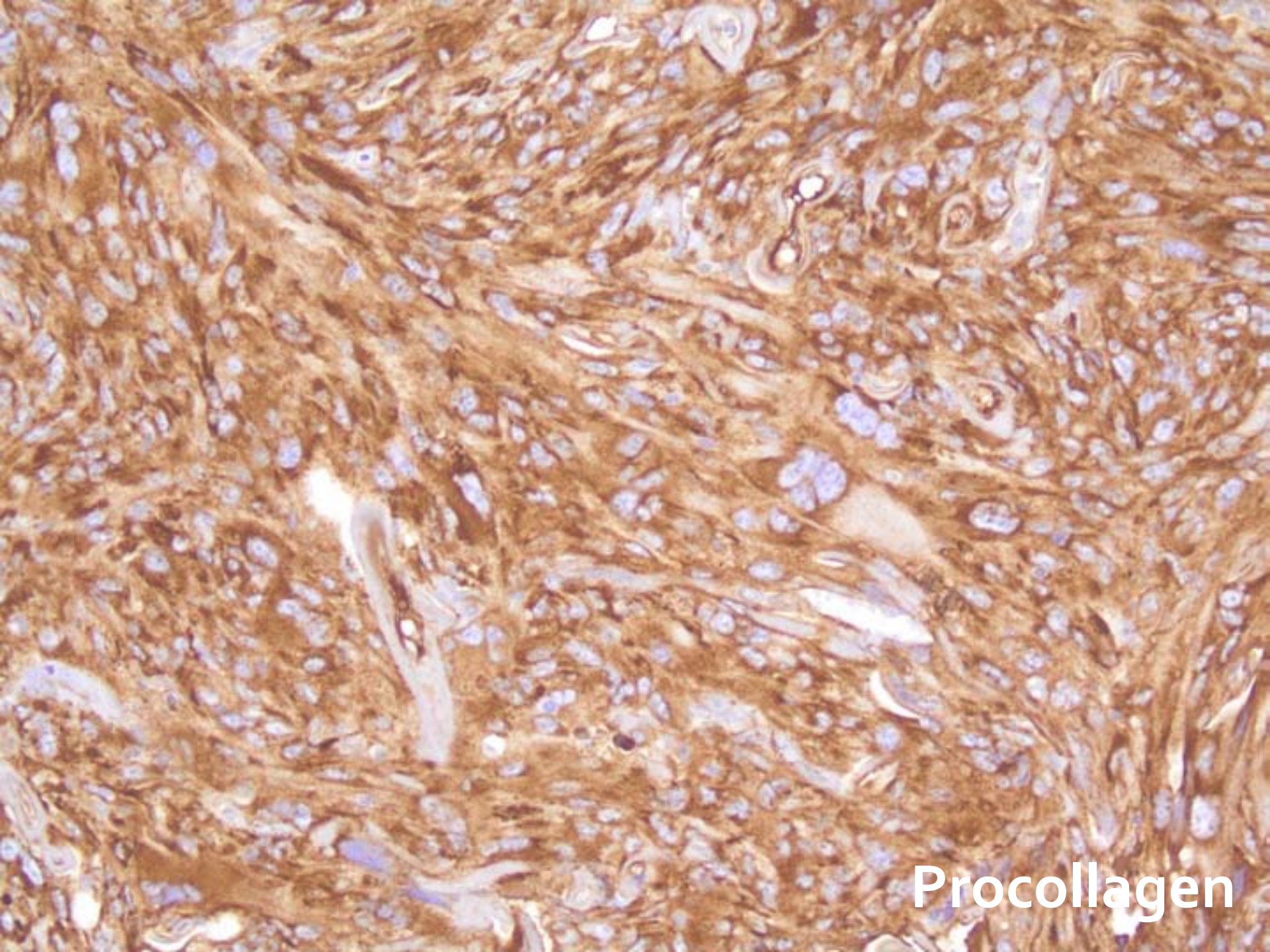
Atypical Fibroxanthoma



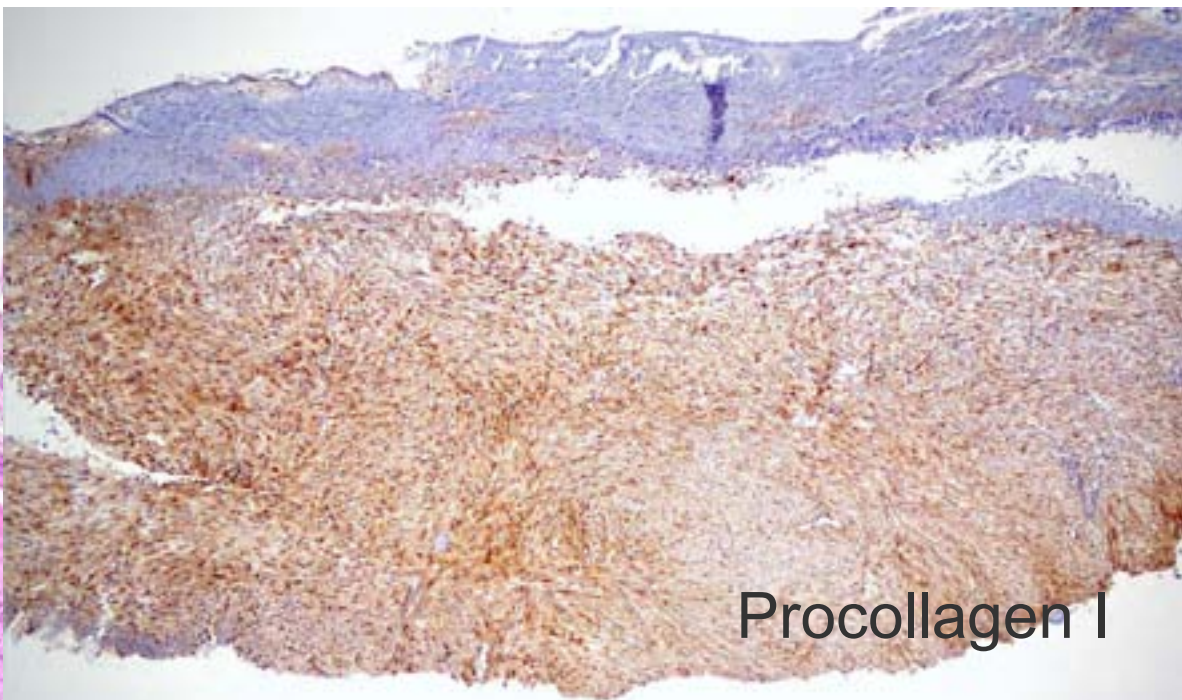
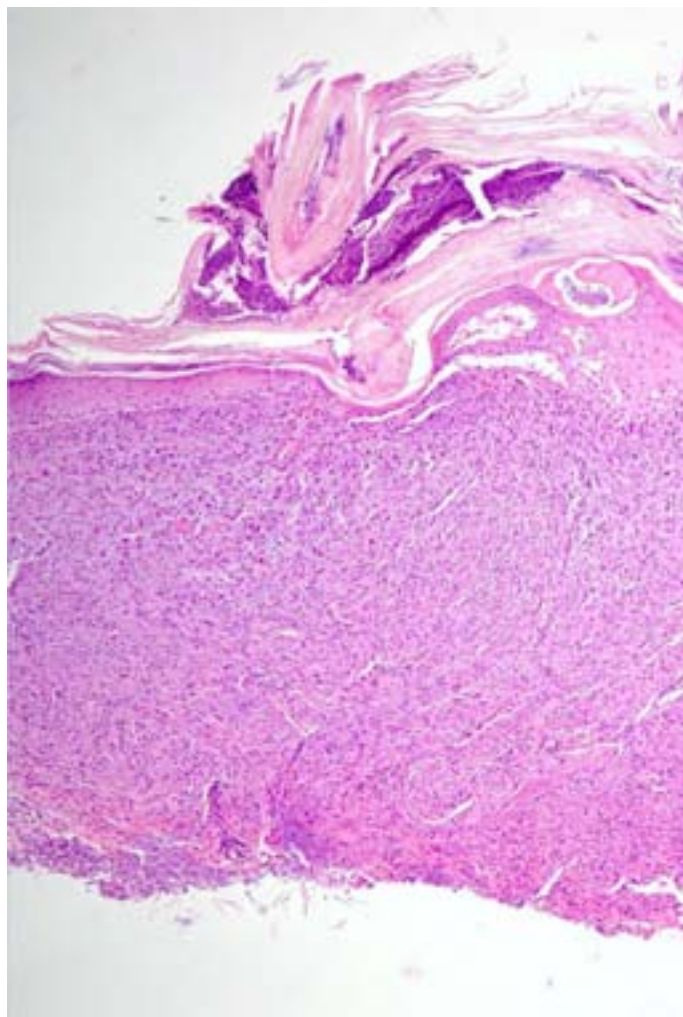




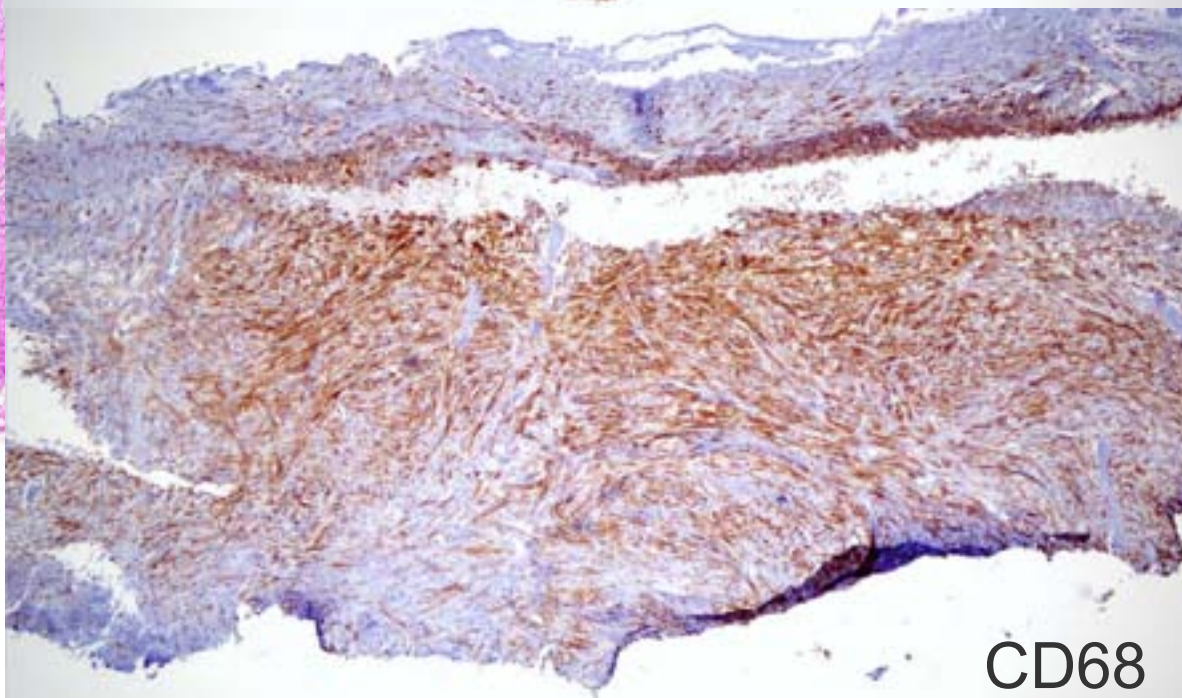




Procollagen



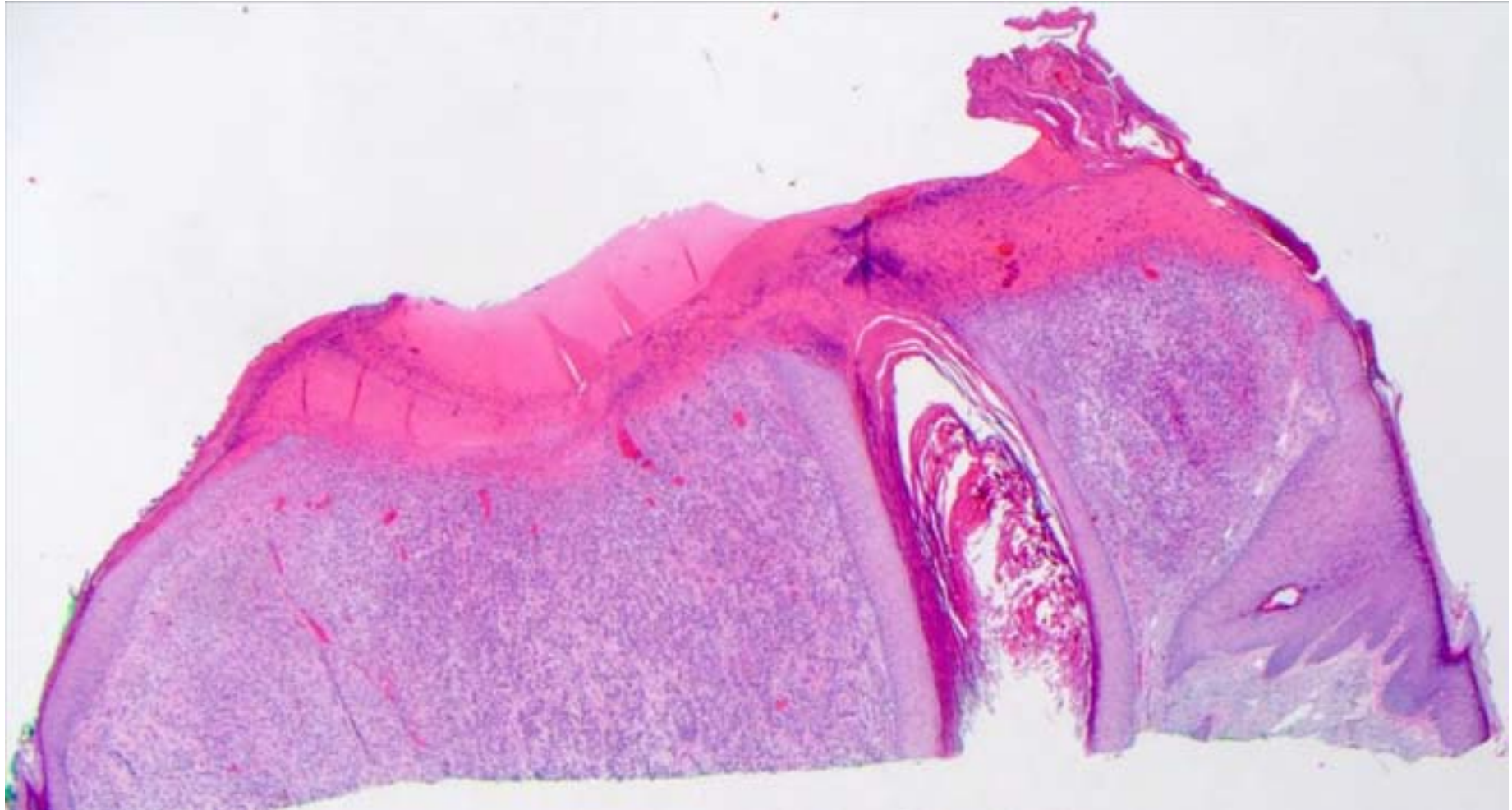
Procollagen I



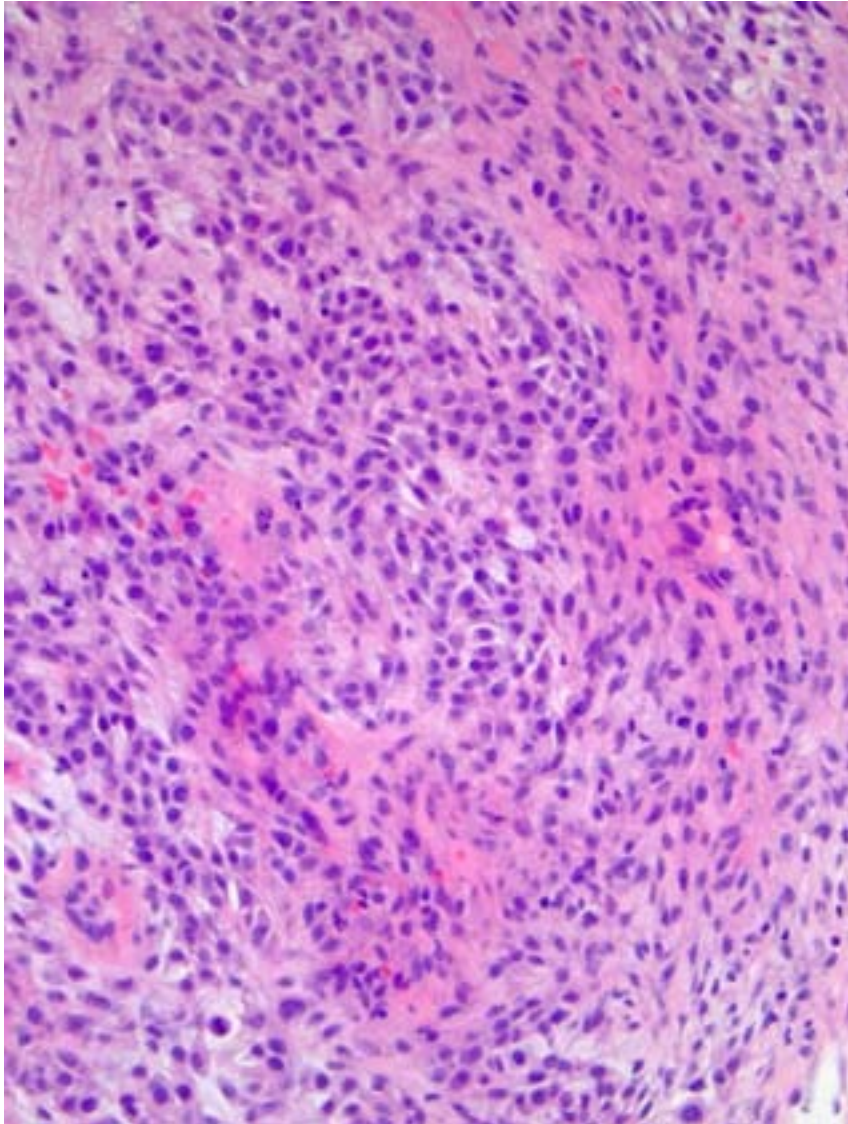
CD68

P53 is also
of utility...

- Eroded nodule
- Ear of 89 y/o woman



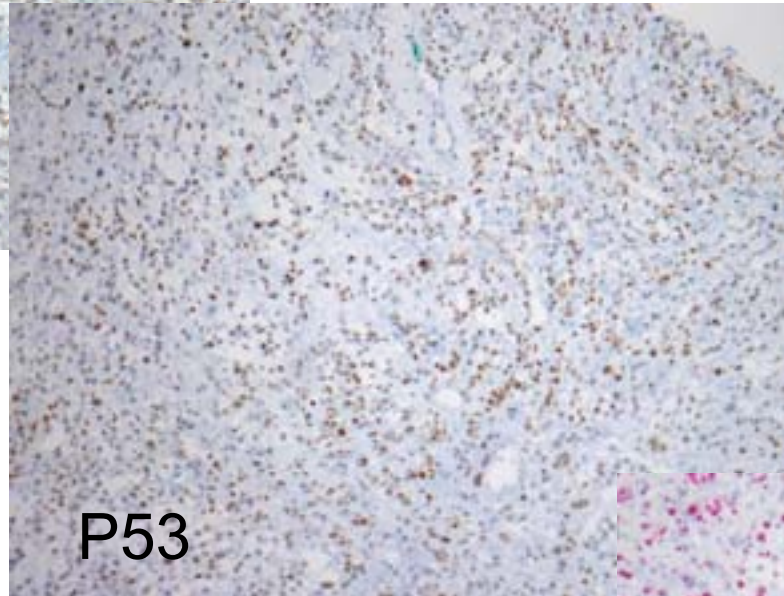
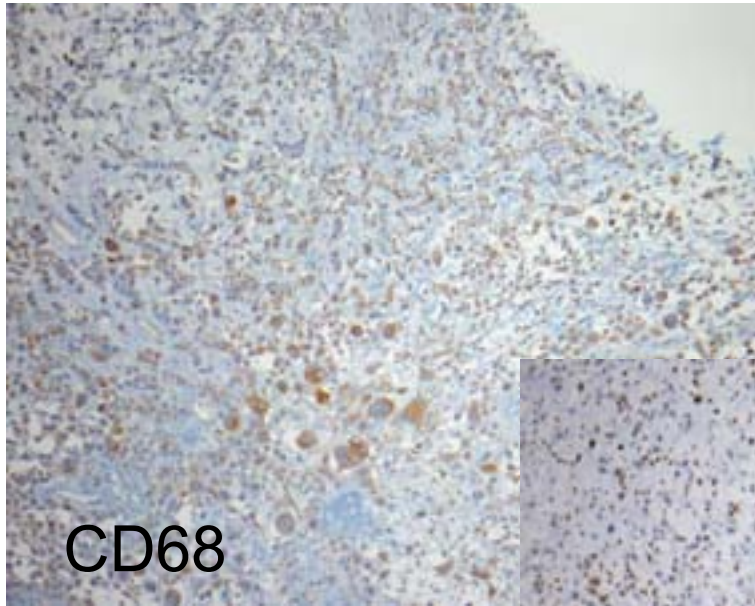
Atypical spindled cells...



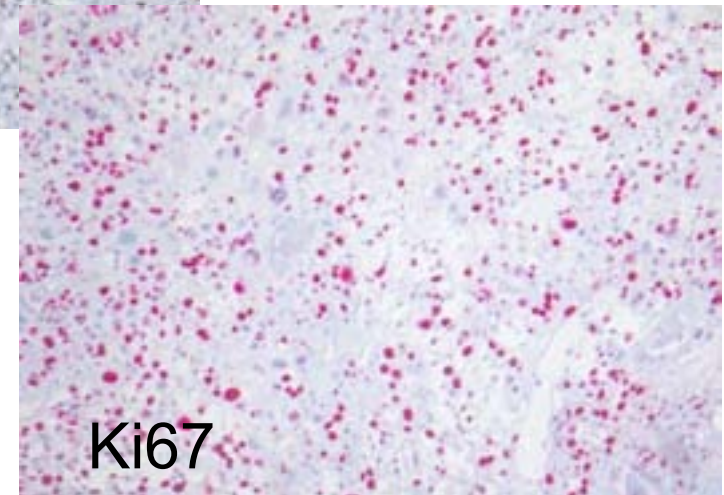
PanCK, P63,
34Beta12 all neg

S100 was similarly negative.

AFX



Combination of CD68/P53 has
been useful as confirmation of AFX

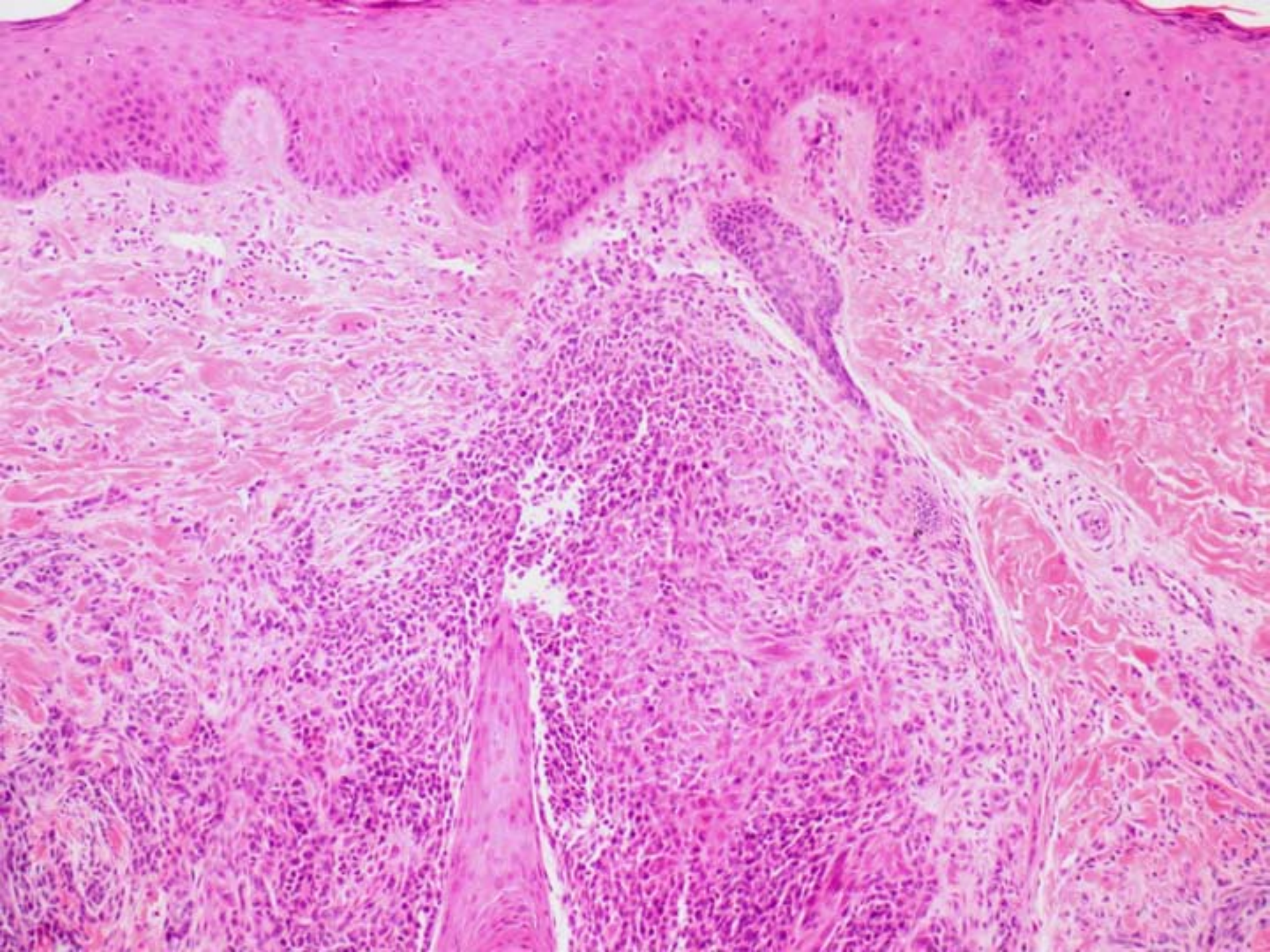


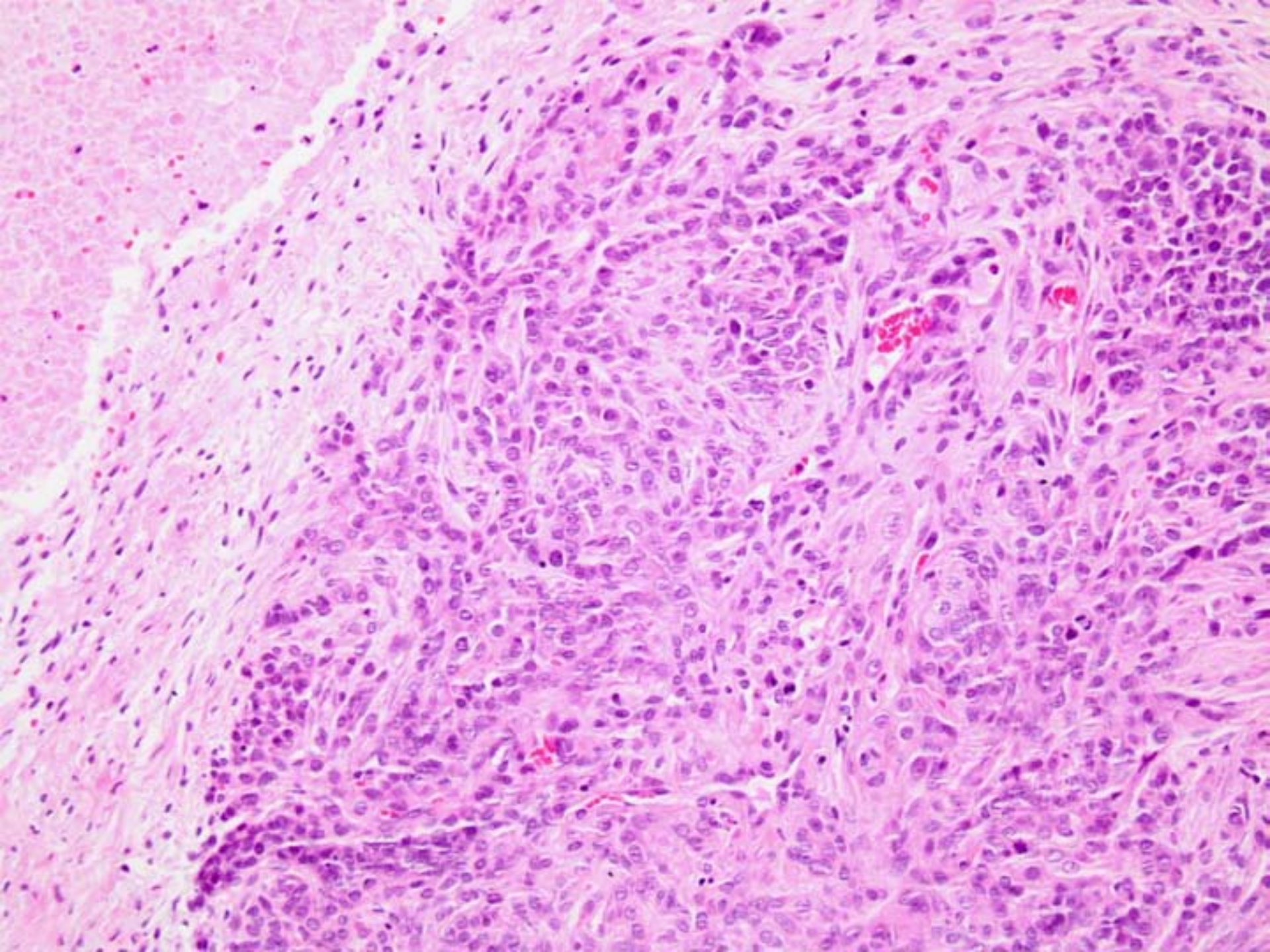
EPITHELIOID SARCOMA

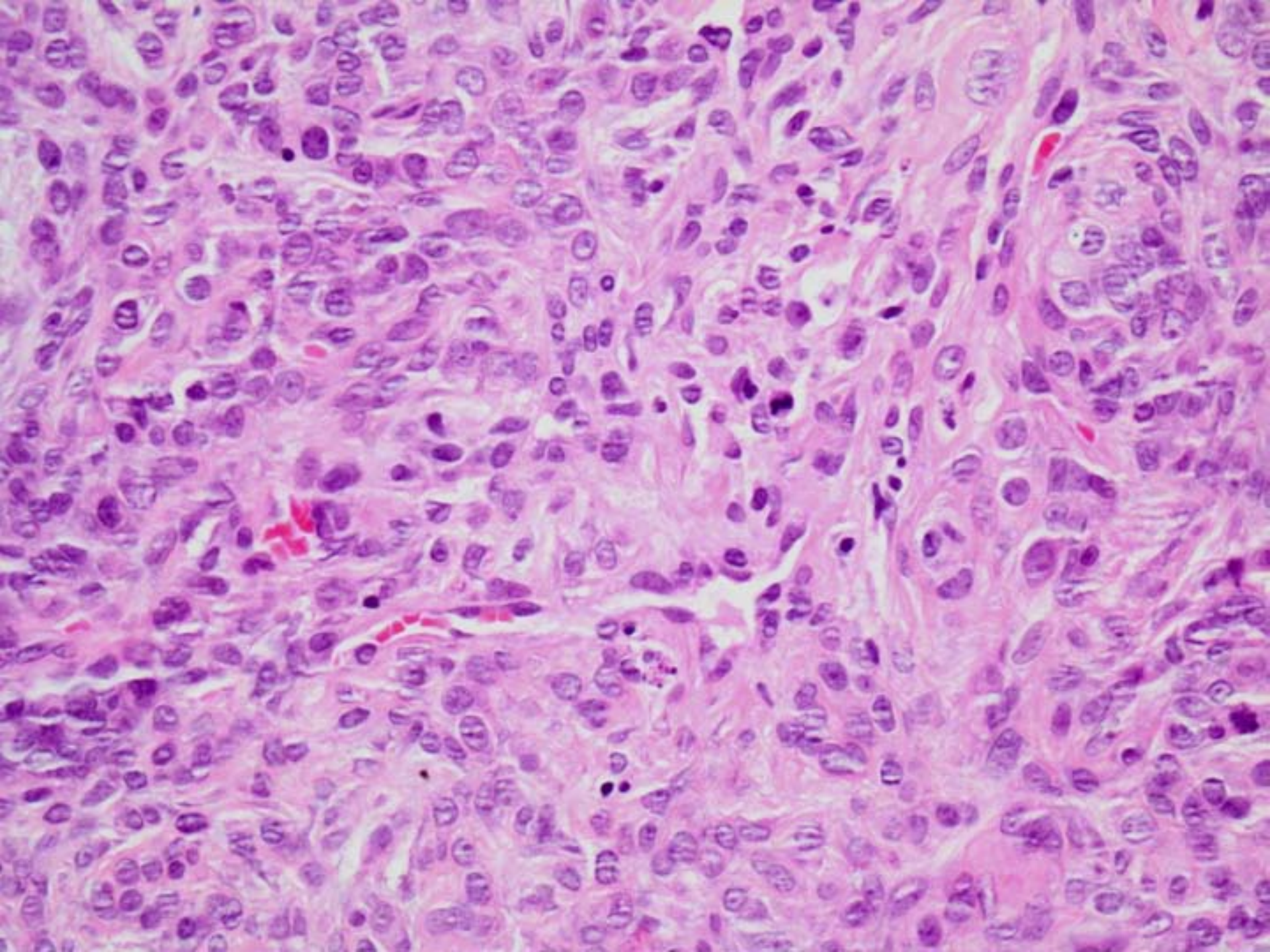
- Rare soft tissue tumor on extremities of young adults
- Confused clinically and histologically with granuloma annulare
- Aggressive (metastasis in 35-40% cases)
- Regional LN and lungs are most affected
- Disputed histiogenesis – synovial?

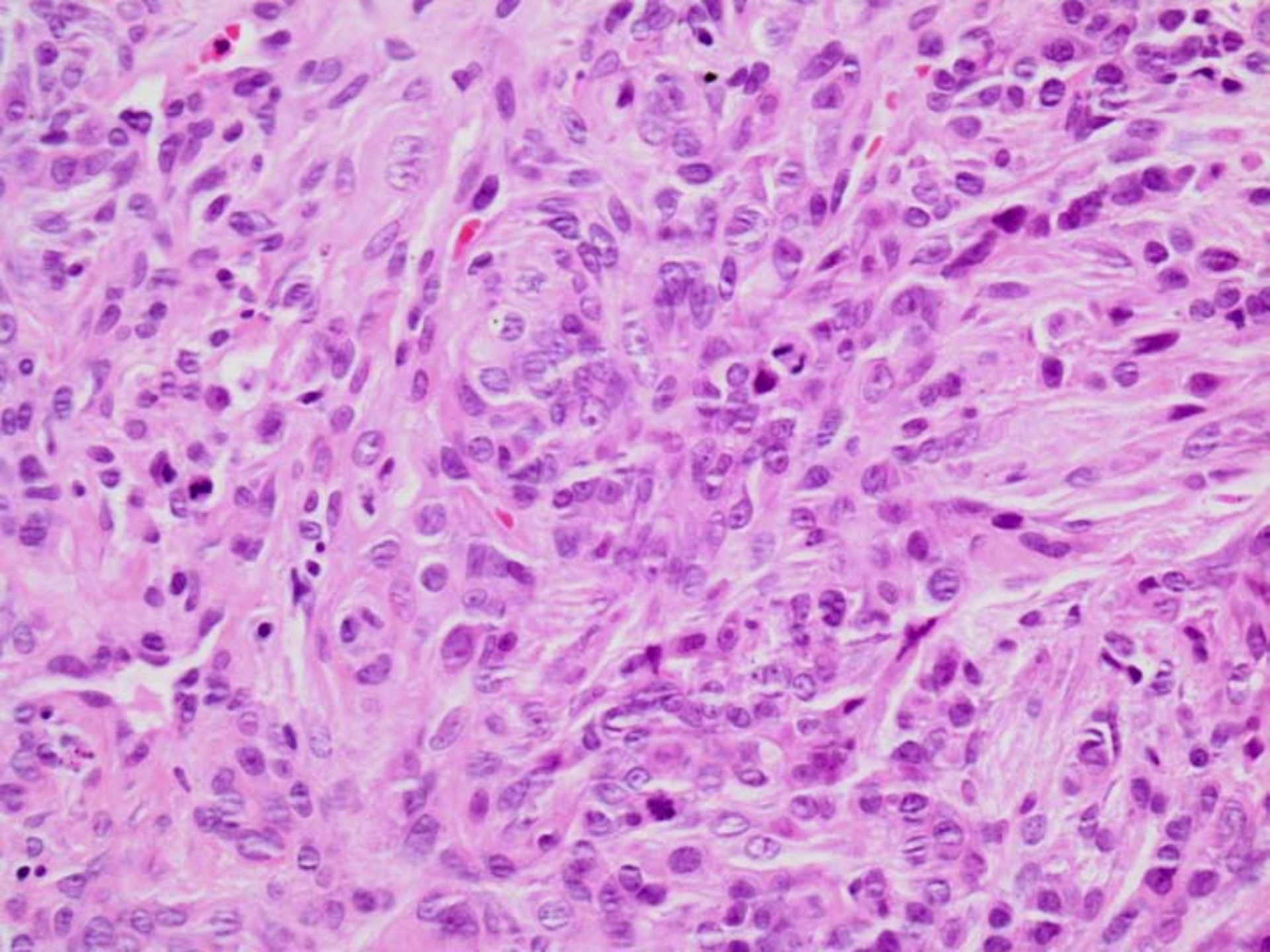
Epithelioid Sarcoma

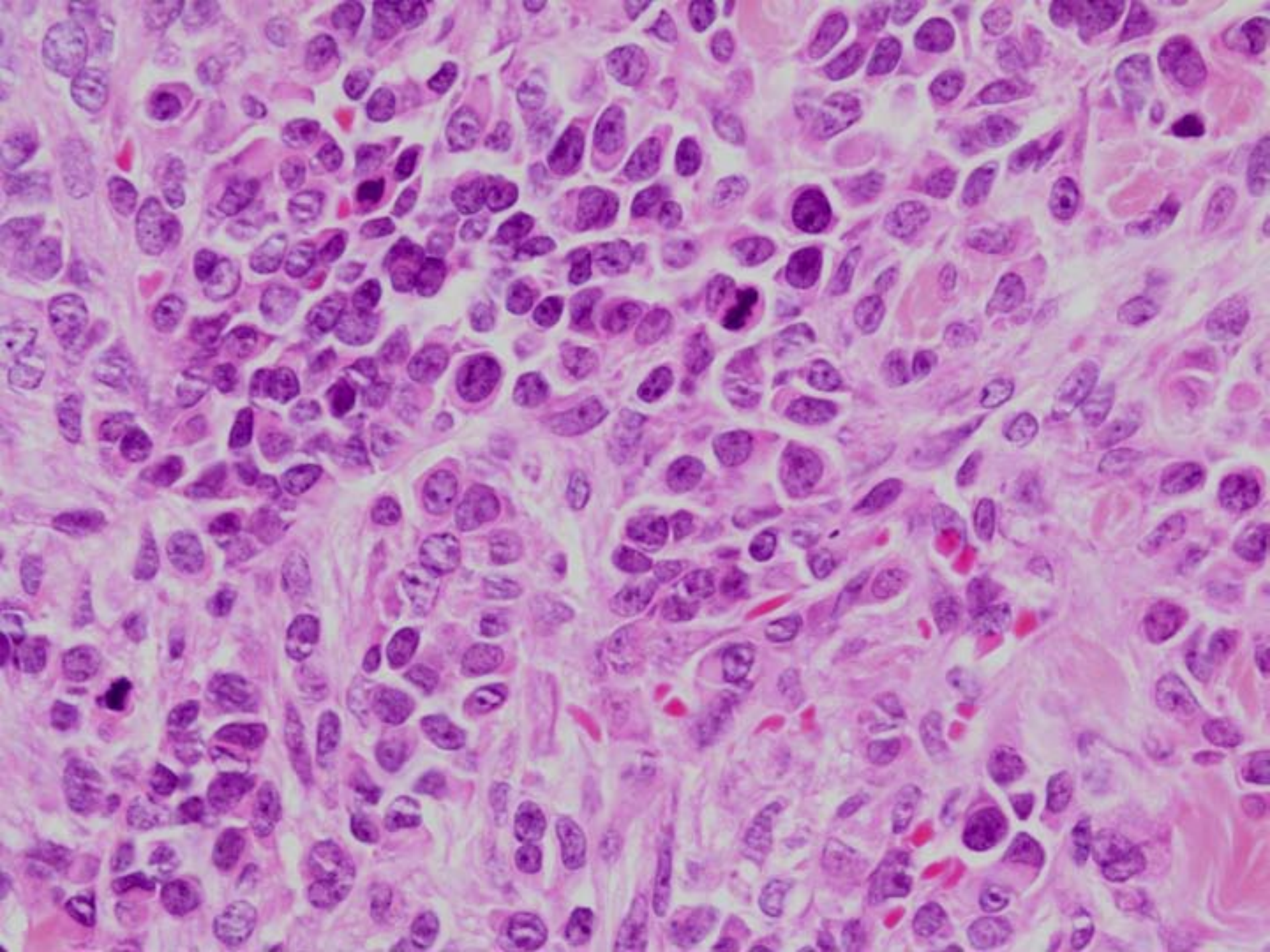












Staining Pattern

- Classic pattern:

POSITIVE = vimentin, PanCK, EMA

NEGATIVE = S100, CK20, CK5/6

Variable = 34Beta12, CK7