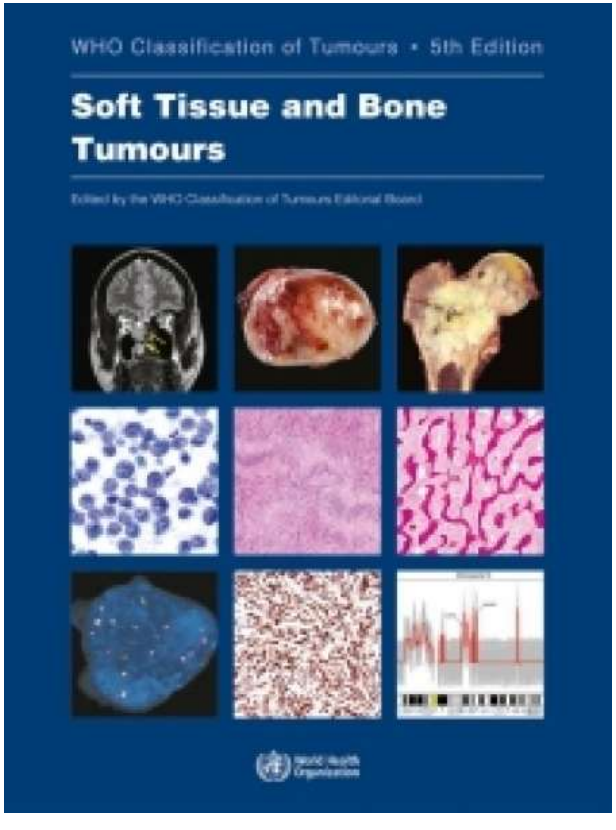
一般病理医・非専門医のための
紡錘形細胞腫瘍の病理診断

産業医科大学医学部第1病理
松山 篤二

- Adipocytic tumours
  - Lipoma
  - Lipomatosis
  - Lipomatosis of nerve
  - Lipoblastoma and lipoblastomatosis
  - Angiolipoma
  - Myolipoma of soft tissue
  - Chondroid lipoma
  - Spindle cell lipoma and pleomorphic lipoma
  - Hibernoma
  - Atypical spindle cell / pleomorphic lipomatous tumour
  - Atypical lipomatous tumour / well-differentiated liposarcoma
  - Dedifferentiated liposarcoma
  - Myxoid liposarcoma
  - Pleomorphic liposarcoma
  - Myxoid pleomorphic liposarcoma
- Fibroblastic and myofibroblastic tumours
  - Nodular fasciitis
  - Proliferative fasciitis and proliferative myositis
  - Myositis ossificans and fibro-osseous pseudotumour of digits
  - Ischaemic fasciitis
  - Elastofibroma
  - Fibrous hamartoma of infancy
  - Fibromatosis colli
  - Juvenile hyaline fibromatosis
  - Inclusion body fibromatosis
  - Fibroma of tendon sheath
  - Desmoplastic fibroblastoma
  - Myofibroblastoma
  - Calcifying aponeurotic fibroma
  - EWSR1-SMAD3-positive fibroblastic tumour (emerging)
  - Angiomyofibroblastoma
  - Cellular angiofibroma
  - Angiofibroma of soft tissue
  - Nuchal-type fibroma
  - Acral fibromyxoma
  - Gardner fibroma
  - Palmar fibromatosis and plantar fibromatosis
  - Desmoid fibromatosis
  - Lipofibromatosis
  - Giant cell fibroblastoma
  - Dermatofibrosarcoma protuberans
  - Solitary fibrous tumour
  - Inflammatory myofibroblastic tumour
  - Low-grade myofibroblastic sarcoma
  - Superficial CD34-positive fibroblastic tumour
  - Myxoinflammatory fibroblastic sarcoma
  - Infantile fibrosarcoma
  - Adult fibrosarcoma
  - Myxofibrosarcoma
  - Low-grade fibromyxoid sarcoma
  - Sclerosing epithelioid fibrosarcoma
- So-called fibrohistiocytic tumours
  - Tenosynovial giant cell tumour
  - Deep fibrous histiocytoma
  - Plexiform fibrohistiocytic tumour
  - Giant cell tumour of soft tissue
- Vascular tumours
  - Haemangiomas
    - Synovial haemangioma
    - Intramuscular angioma
    - Arteriovenous malformation/haemangioma
    - Venous haemangioma
  - Anastomosing haemangioma
  - Epithelioid haemangioma
  - Lymphangioma and lymphangiomatosis
  - Tufted angioma and kaposiform haemangioendothelioma
  - Retiform haemangioendothelioma
  - Papillary intralymphatic angioendothelioma
  - Composite haemangioendothelioma
  - Kaposi sarcoma
  - Pseudomyogenic haemangioendothelioma
  - Epithelioid haemangioendothelioma
  - Angiosarcoma
- Pericytic (perivascular) tumours
  - Glomus tumour
  - Myopericytoma, including myofibroma
  - Angioleiomyoma
- Smooth muscle tumours
  - Leiomyoma
  - EBV-associated smooth muscle tumour
  - Inflammatory leiomyosarcoma
  - Leiomyosarcoma
- Skeletal muscle tumours
  - Rhabdomyoma
  - Embryonal rhabdomyosarcoma
  - Alveolar rhabdomyosarcoma
  - Pleomorphic rhabdomyosarcoma
  - Spindle cell / sclerosing rhabdomyosarcoma
  - Ectomesenchymoma
- Gastrointestinal stromal tumour
  - Gastrointestinal stromal tumour
- Chondro-osseous tumours
  - Soft tissue chondroma
  - Extraskeletal osteosarcoma
- Peripheral nerve sheath tumours
  - Schwannoma
  - Neurofibroma
  - Perineurioma
  - Granular cell tumour
  - Dermal nerve sheath myxoma
  - Solitary circumscribed neuroma
  - Ectopic meningioma and meningothelial hamartoma
  - Benign triton tumour / neuromuscular choristoma
  - Hybrid nerve sheath tumour
  - Malignant peripheral nerve sheath tumour
  - Malignant melanotic nerve sheath tumour
- Tumours of uncertain differentiation
  - Intramuscular myxoma
  - Juxta-articular myxoma
  - Deep (aggressive) angiomyxoma
  - Atypical fibroxanthoma
  - Angiomatoid fibrous histiocytoma
  - Ossifying fibromyxoid tumour
  - Myoepithelioma, myoepithelial carcinoma, and mixed tumour
  - Pleomorphic hyalinizing angiectatic tumour of soft parts
  - Haemosiderotic fibrolipomatous tumour
  - Phosphaturic mesenchymal tumour
  - NTRK-rearranged spindle cell neoplasm (emerging)
  - Synovial sarcoma
  - Epithelioid sarcoma
  - Alveolar soft part sarcoma
  - Clear cell sarcoma of soft tissue
  - Extraskeletal myxoid chondrosarcoma
  - Desmoplastic small round cell tumour
  - Extrarenal rhabdoid tumour
  - PEComa, including angiomyolipoma
  - Intimal sarcoma
  - Undifferentiated sarcoma



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本日の内容

一般病理医・非専門医のための
紡錘形細胞腫瘍の病理診断

コンサルテーション症例 Case 1－4 の解説

↓

鑑別に挙げられた腫瘍、疾患概念や形態像から
関連のある腫瘍

ほぼルーチンで使用している抗体

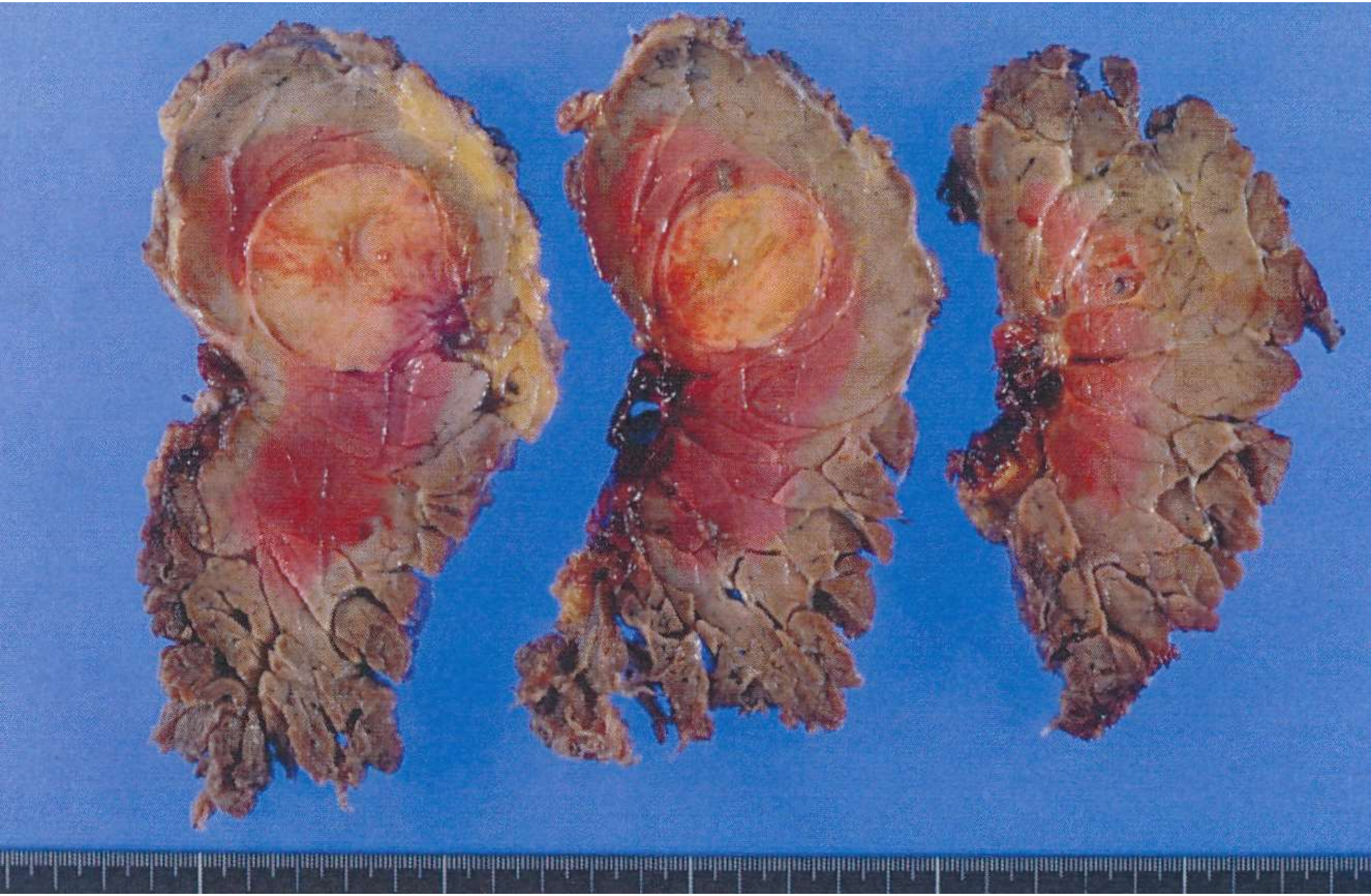
αSMA, desmin, CD34, S-100, AE1/AE3, CAM5.2, EMA

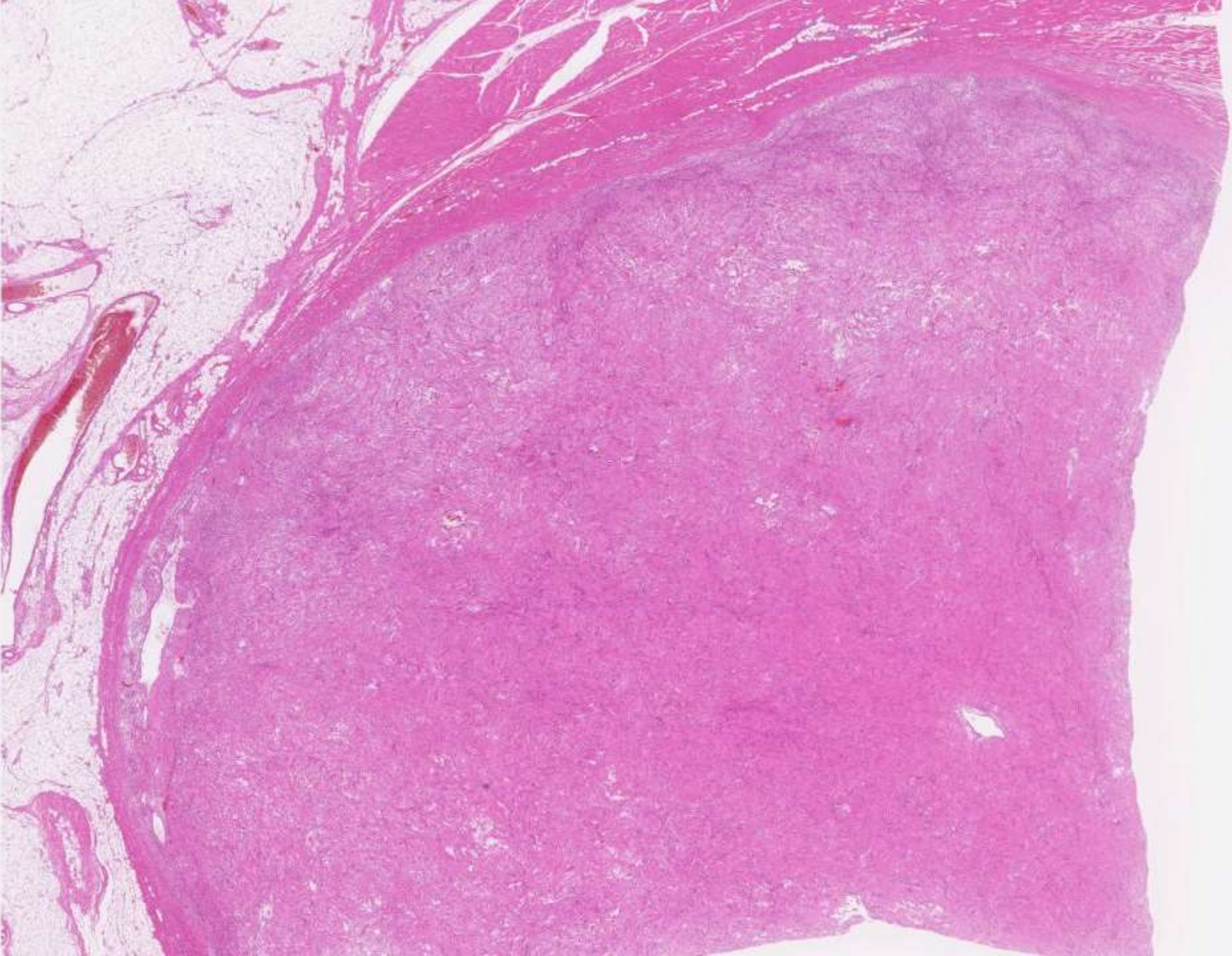
+ MDM2, CDK4

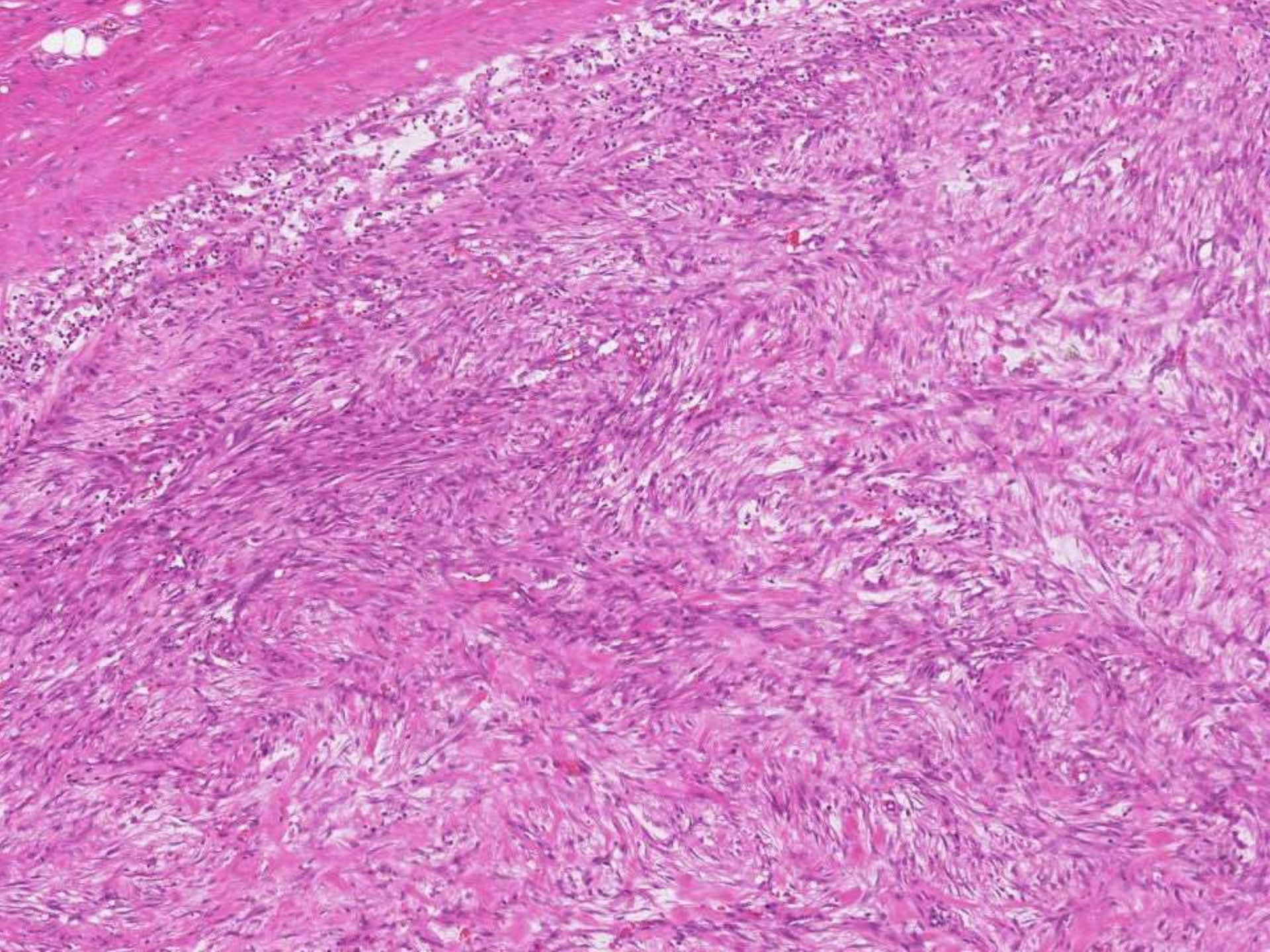
コンサルテーション依頼者が染めている頻度は高いが、我々はあまり用いない抗体

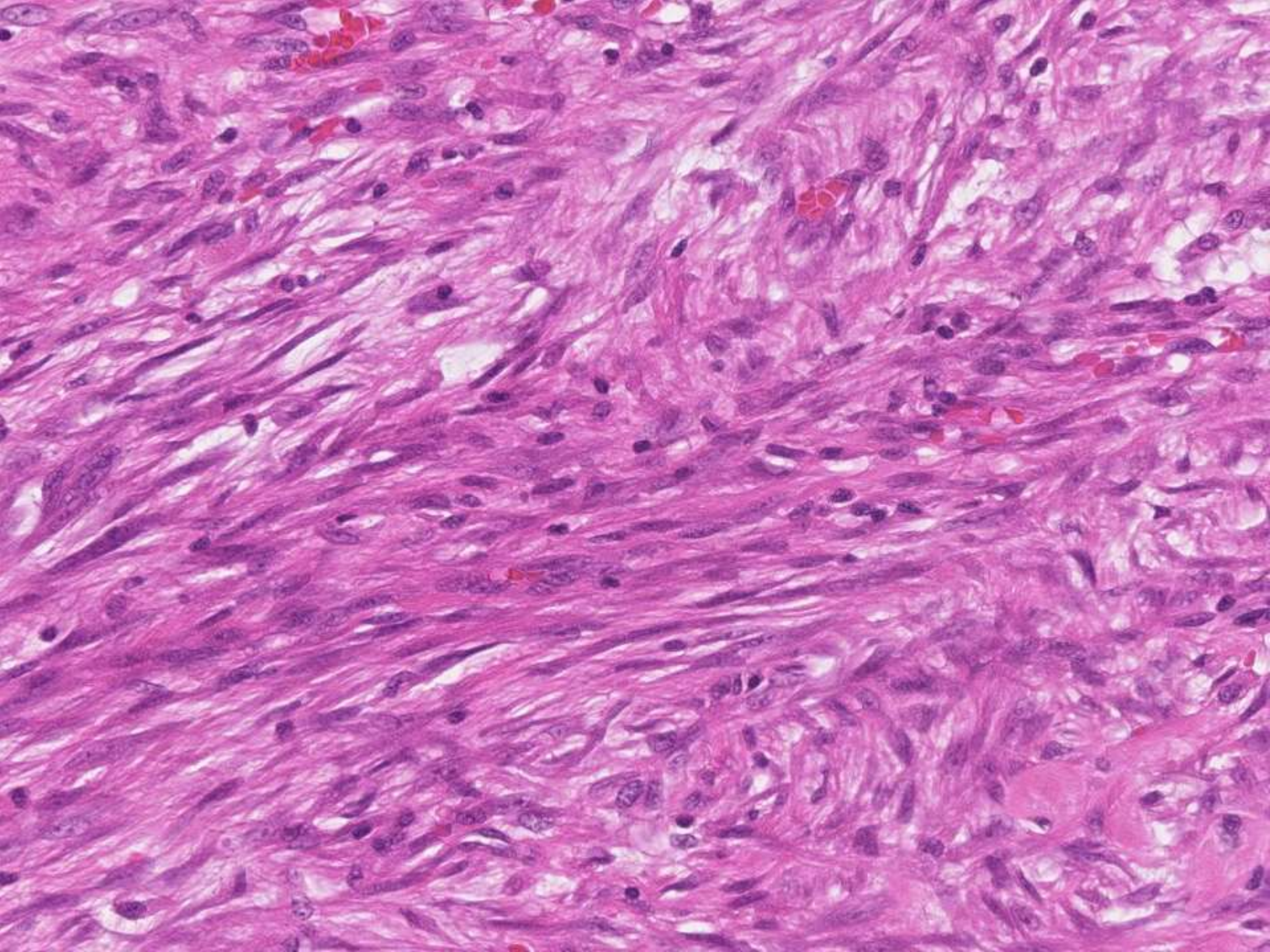
Vimentin, c-kit, CD68

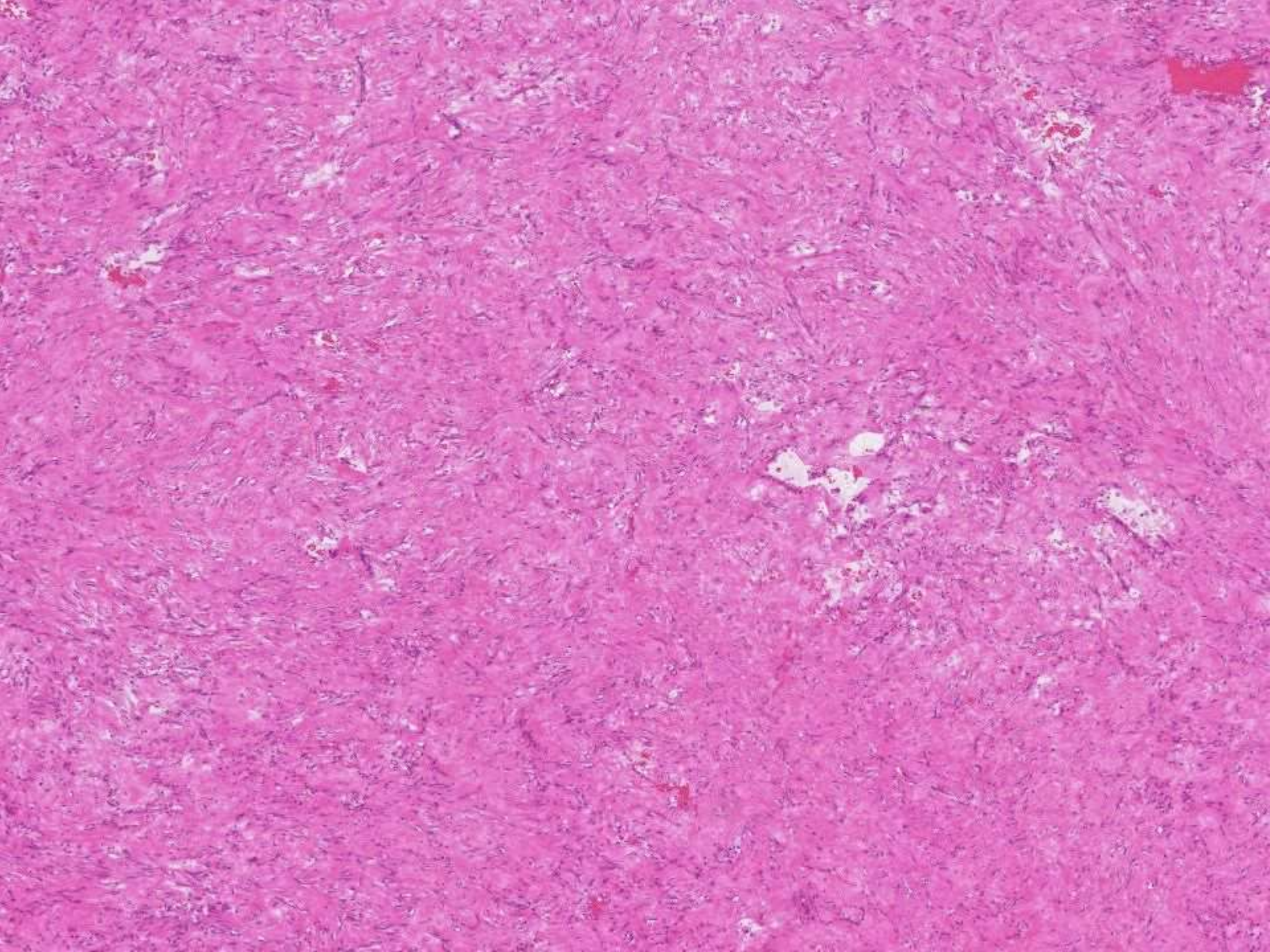
Case 1: 45M, buttock

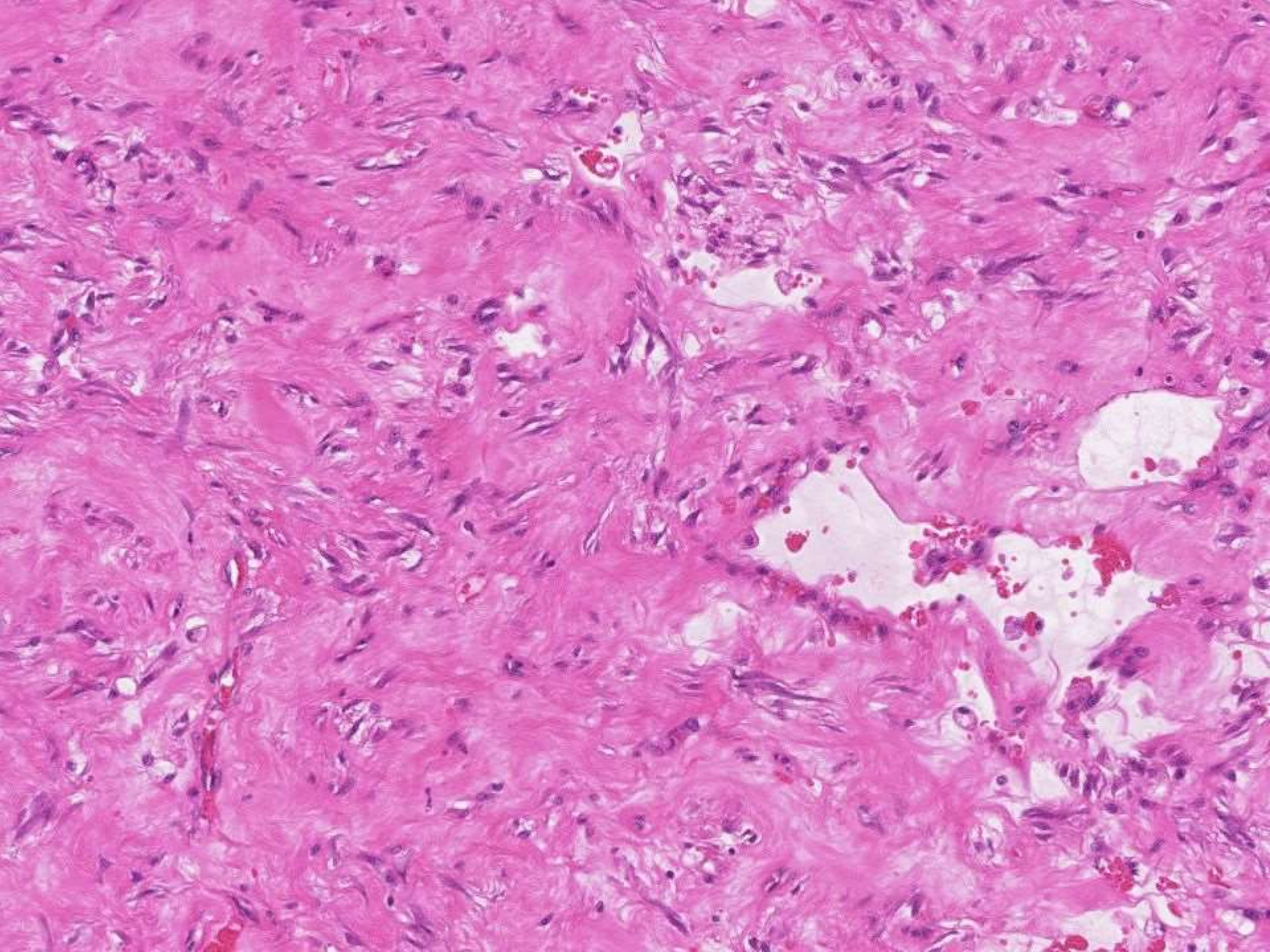


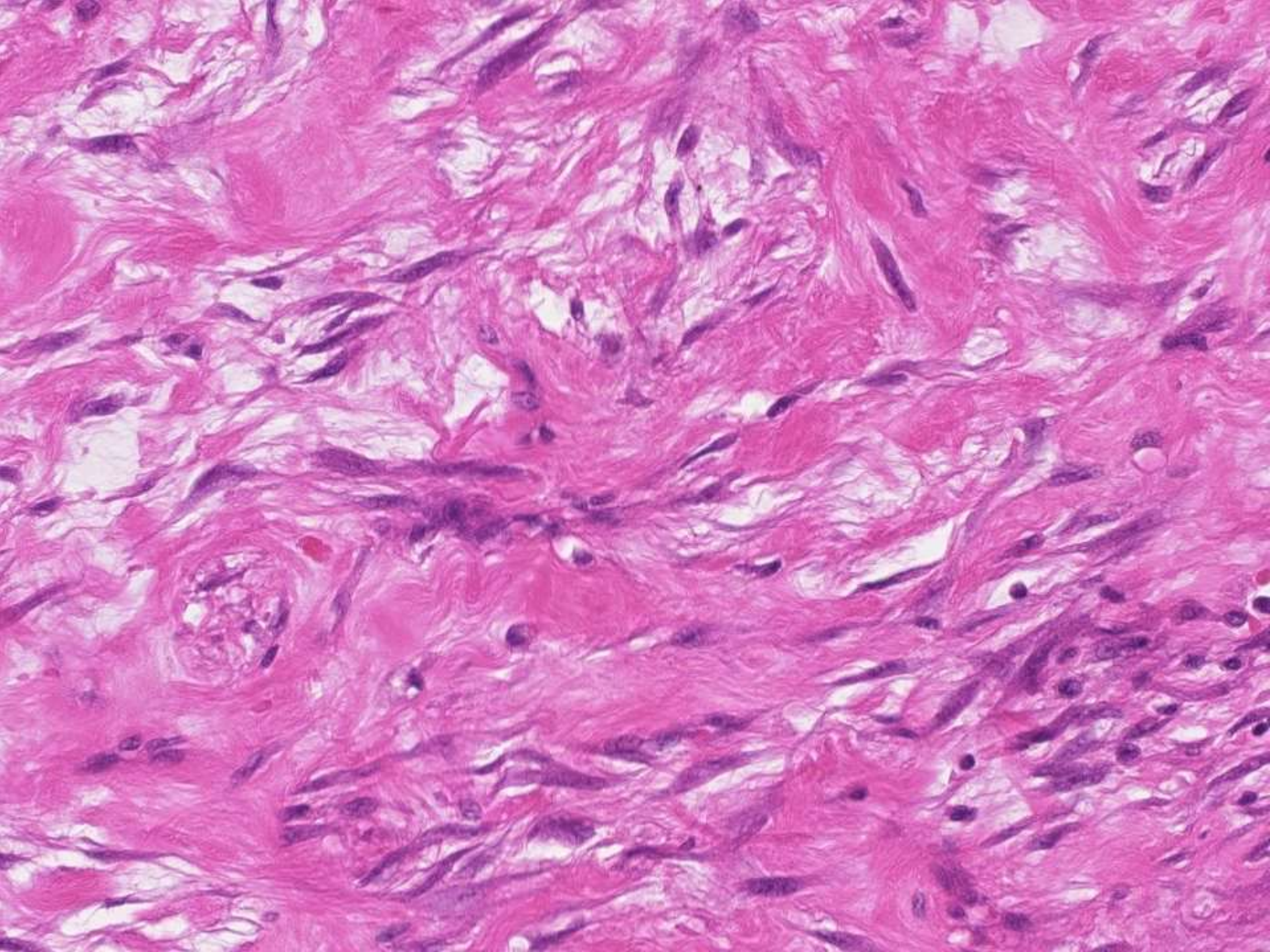


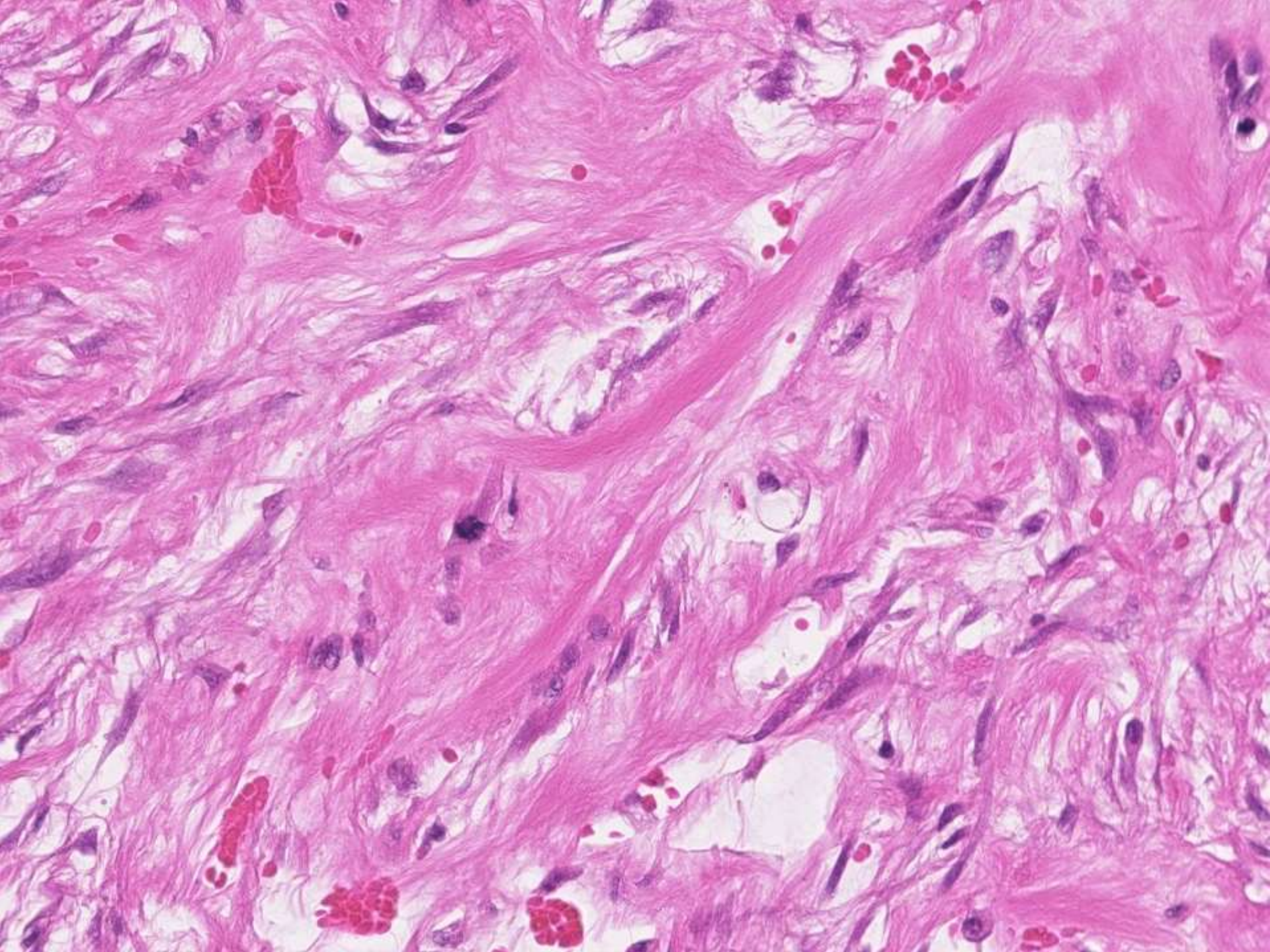




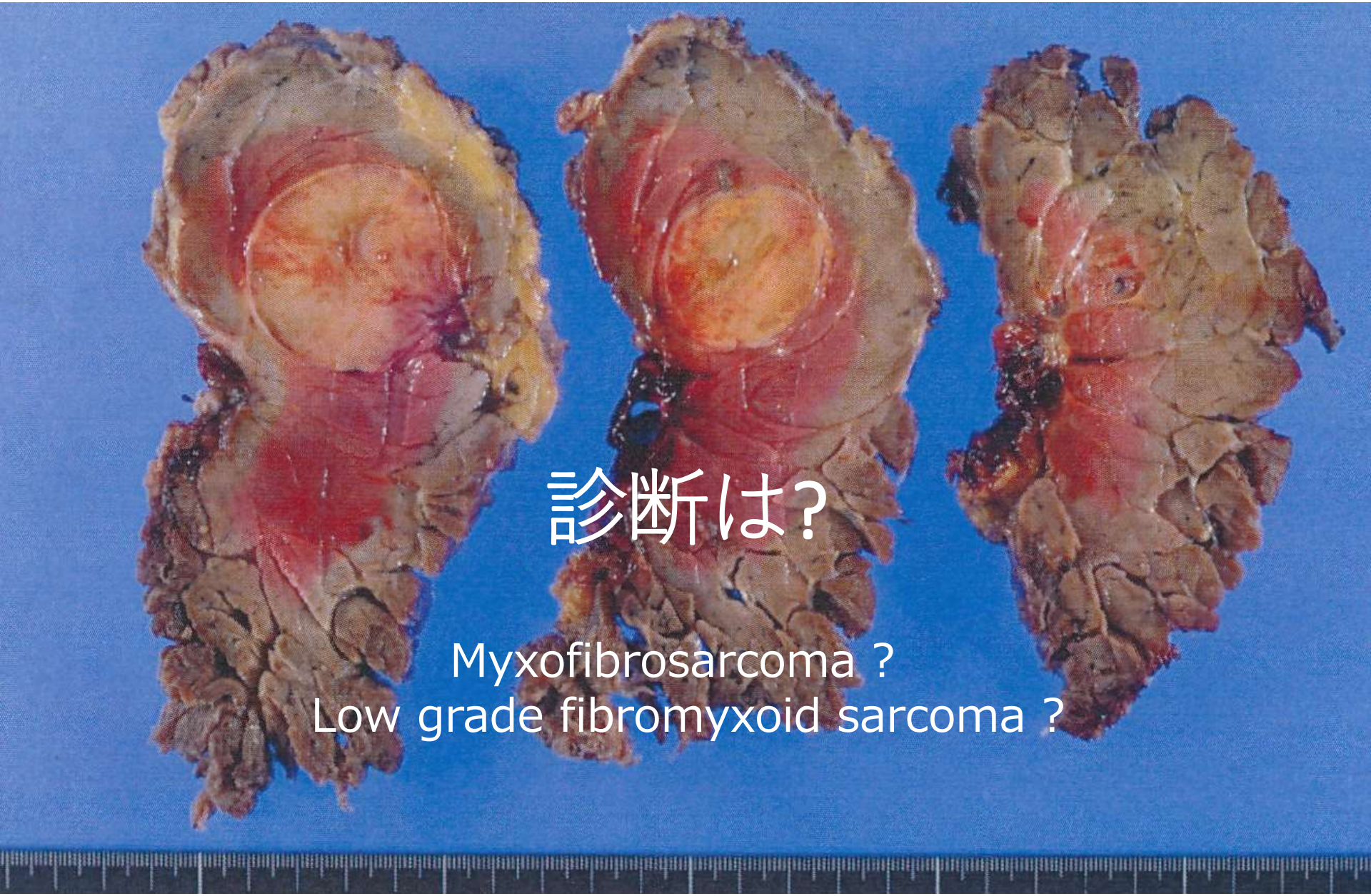








Case 1: 45M, buttock

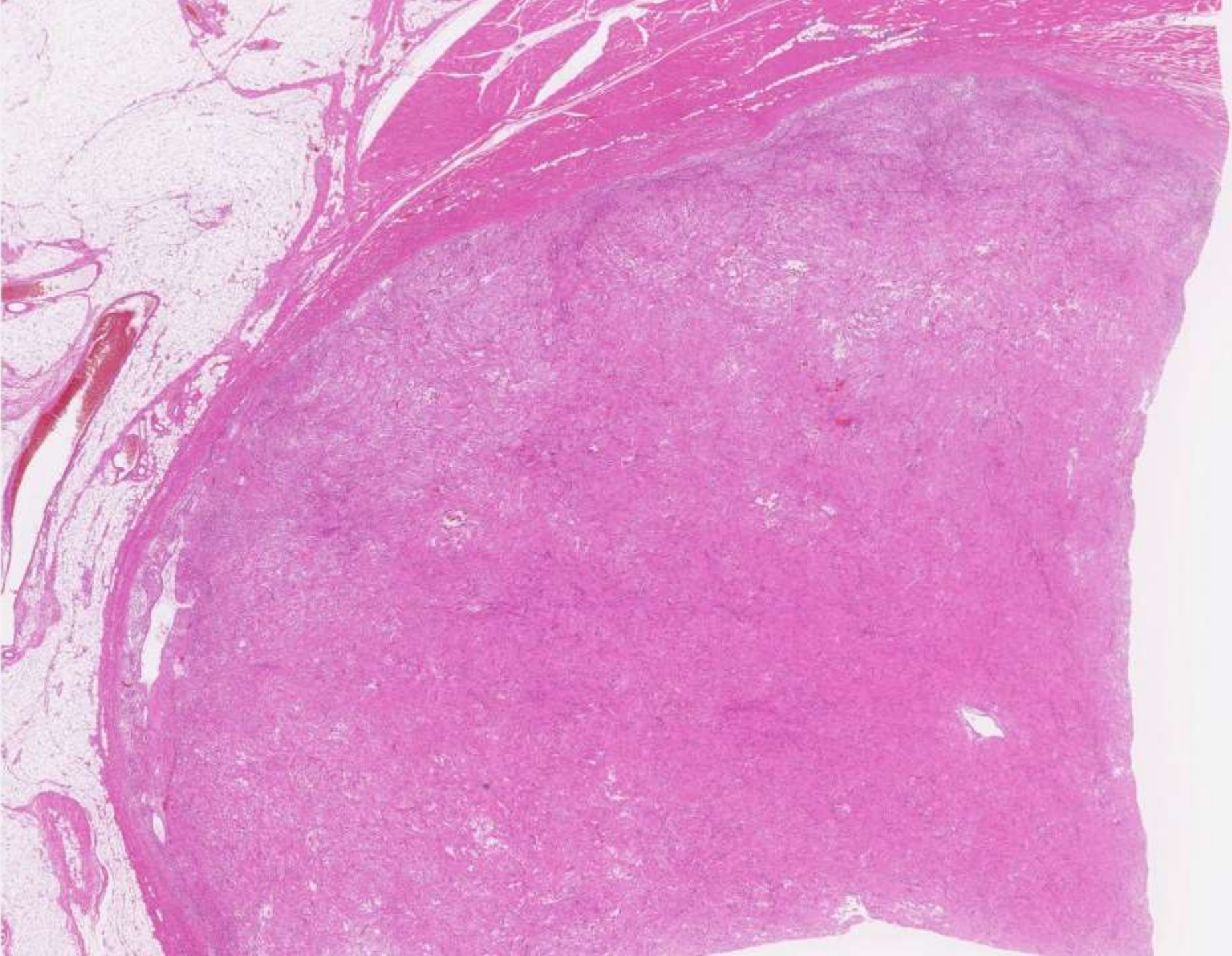


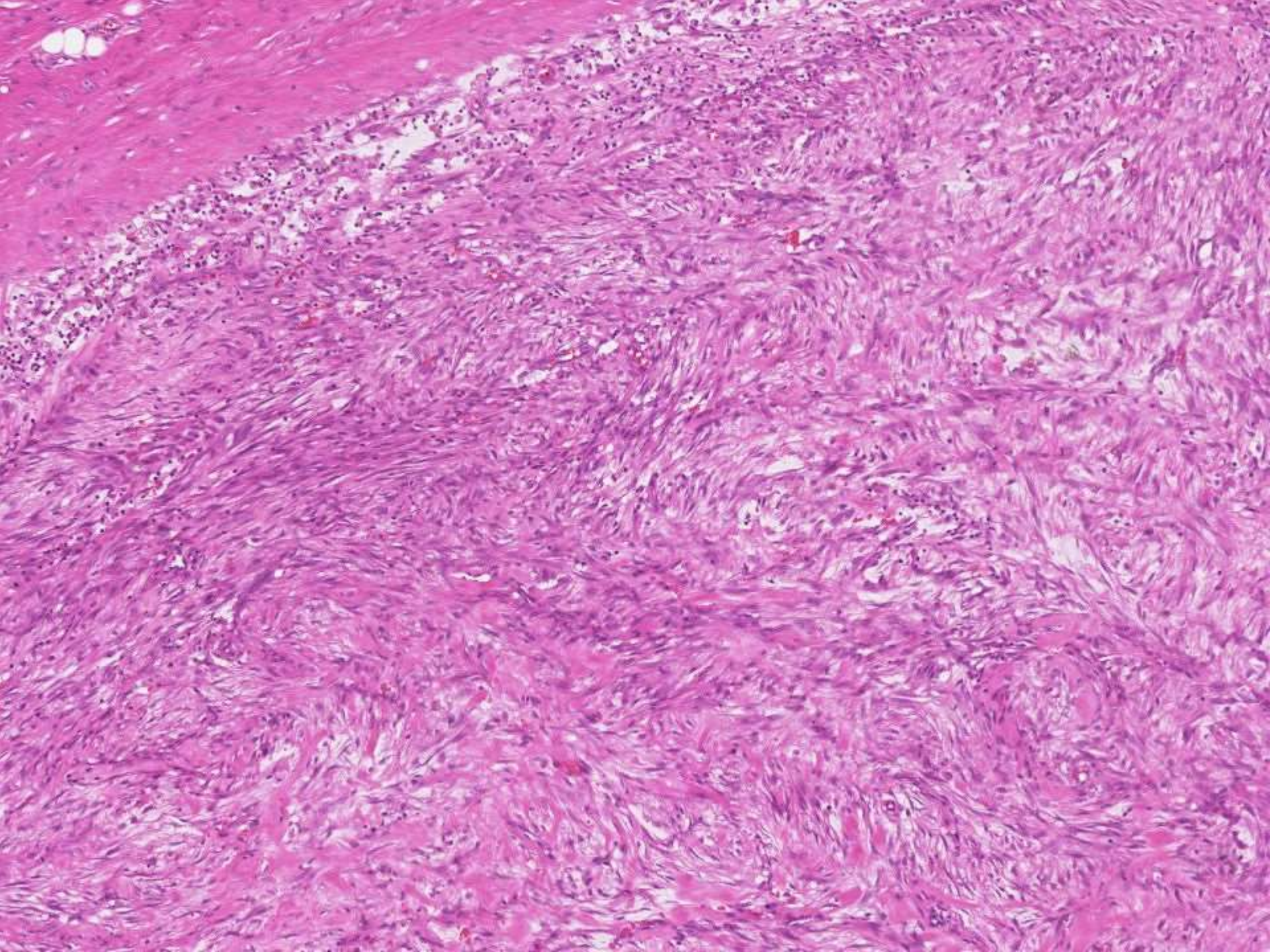
Diagnosis

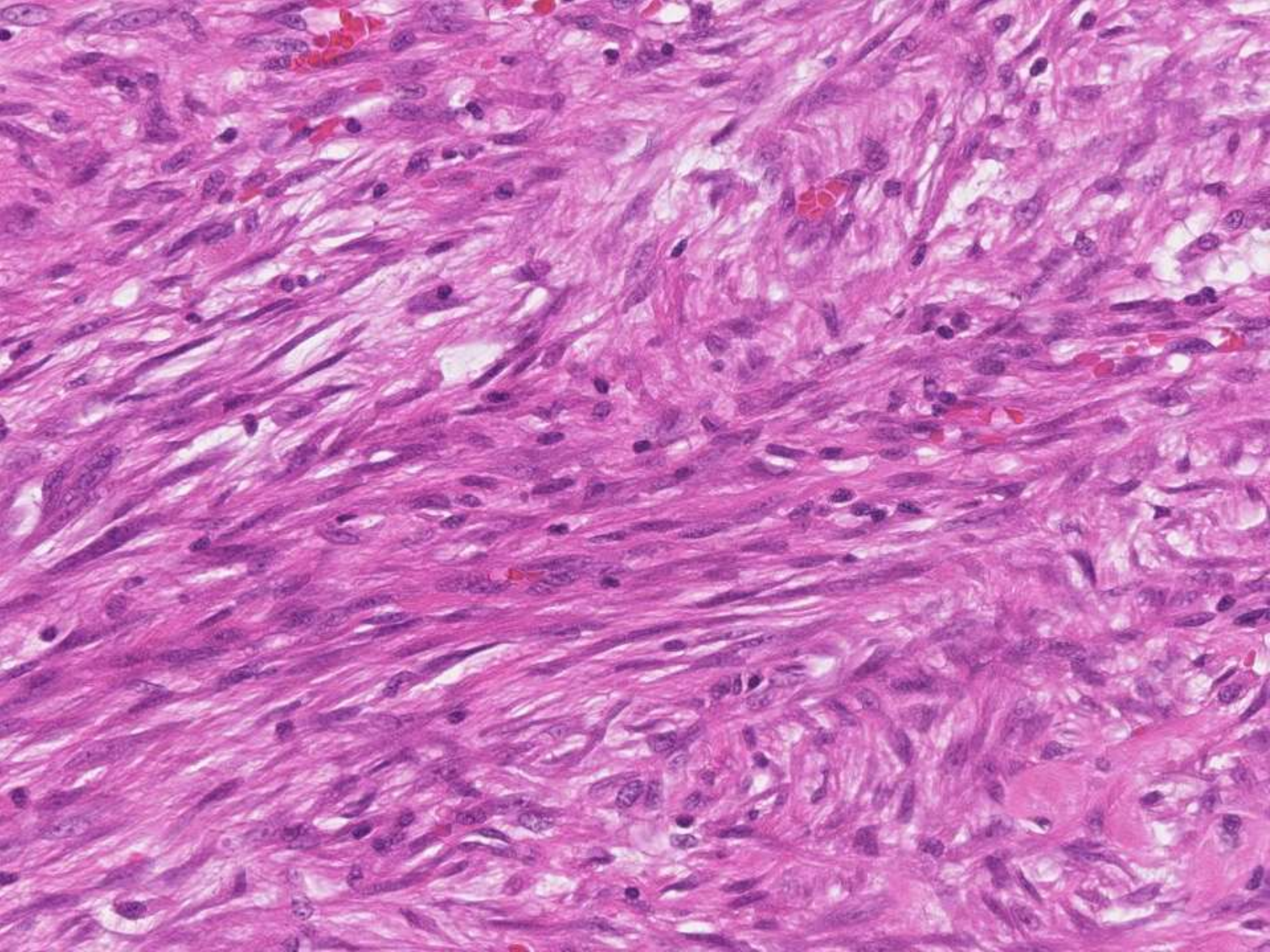
Nodular fasciitis

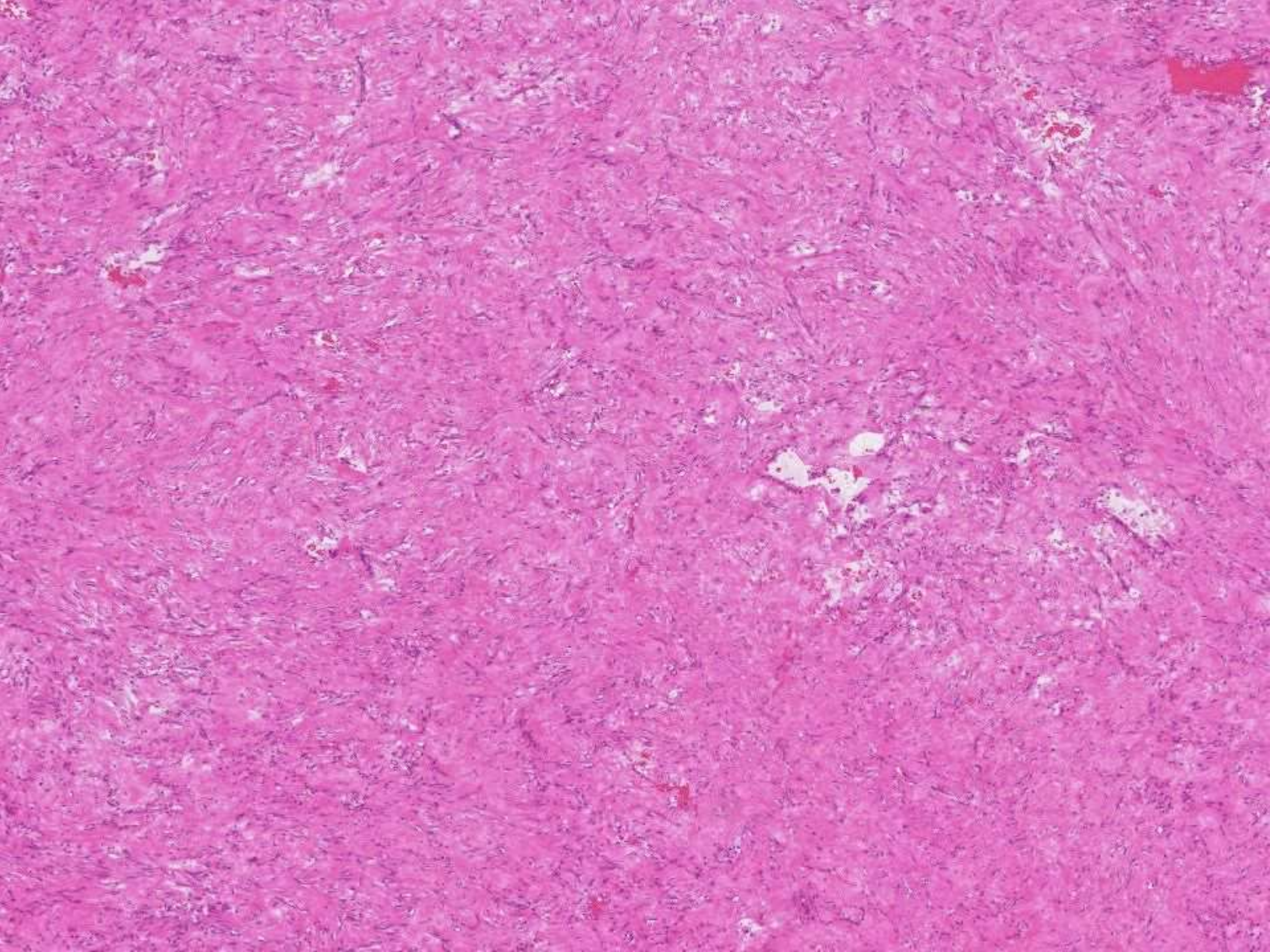
Nodular fasciitis

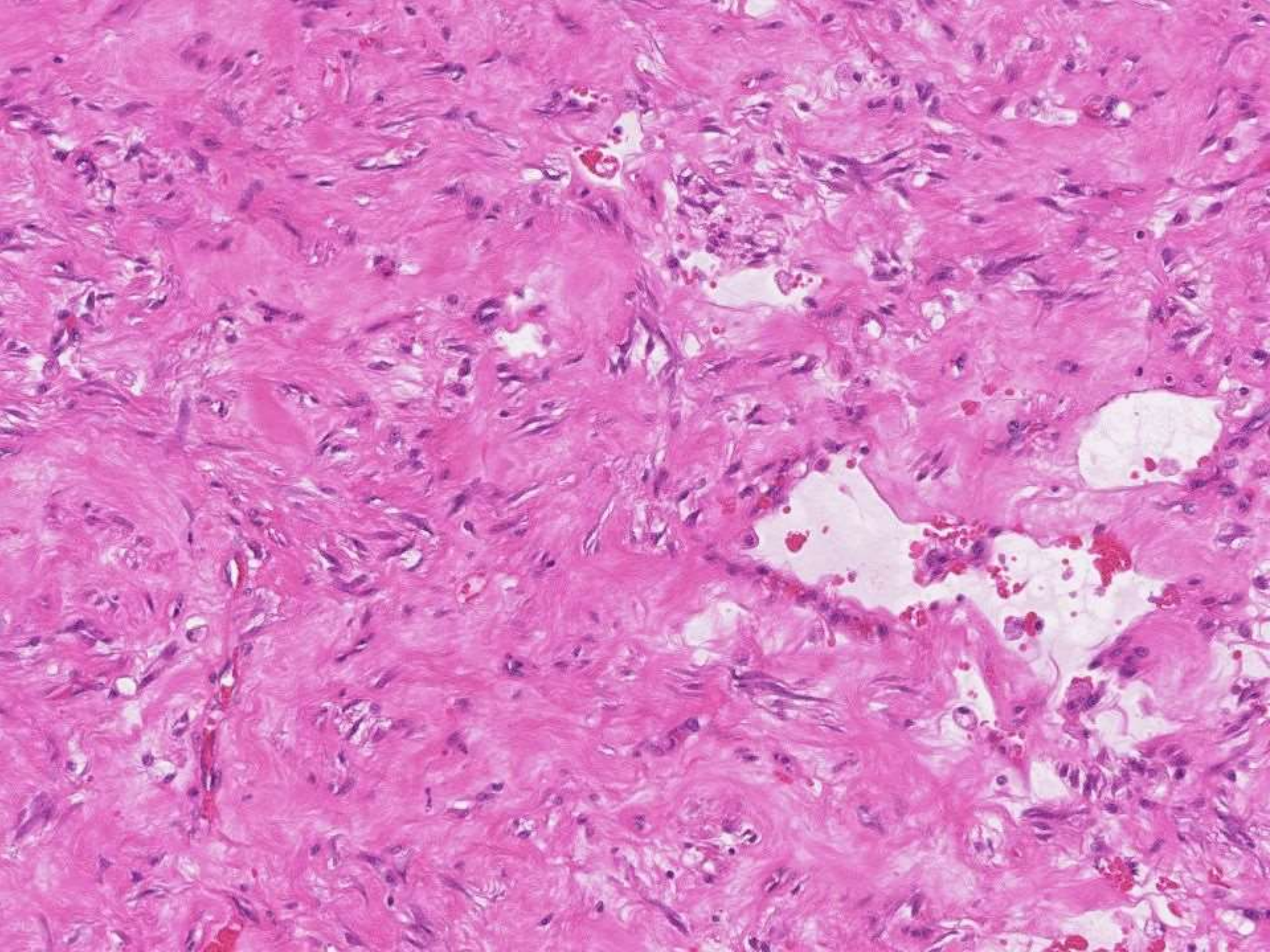
- 若年者＞中高年
- 上肢、体幹、頭頸部に多い
- 皮下に多い
- 急速に増大するものが多い
- 2 cm以下のものが多い
- 外傷歴(+/-)、自然軽快(+/-)

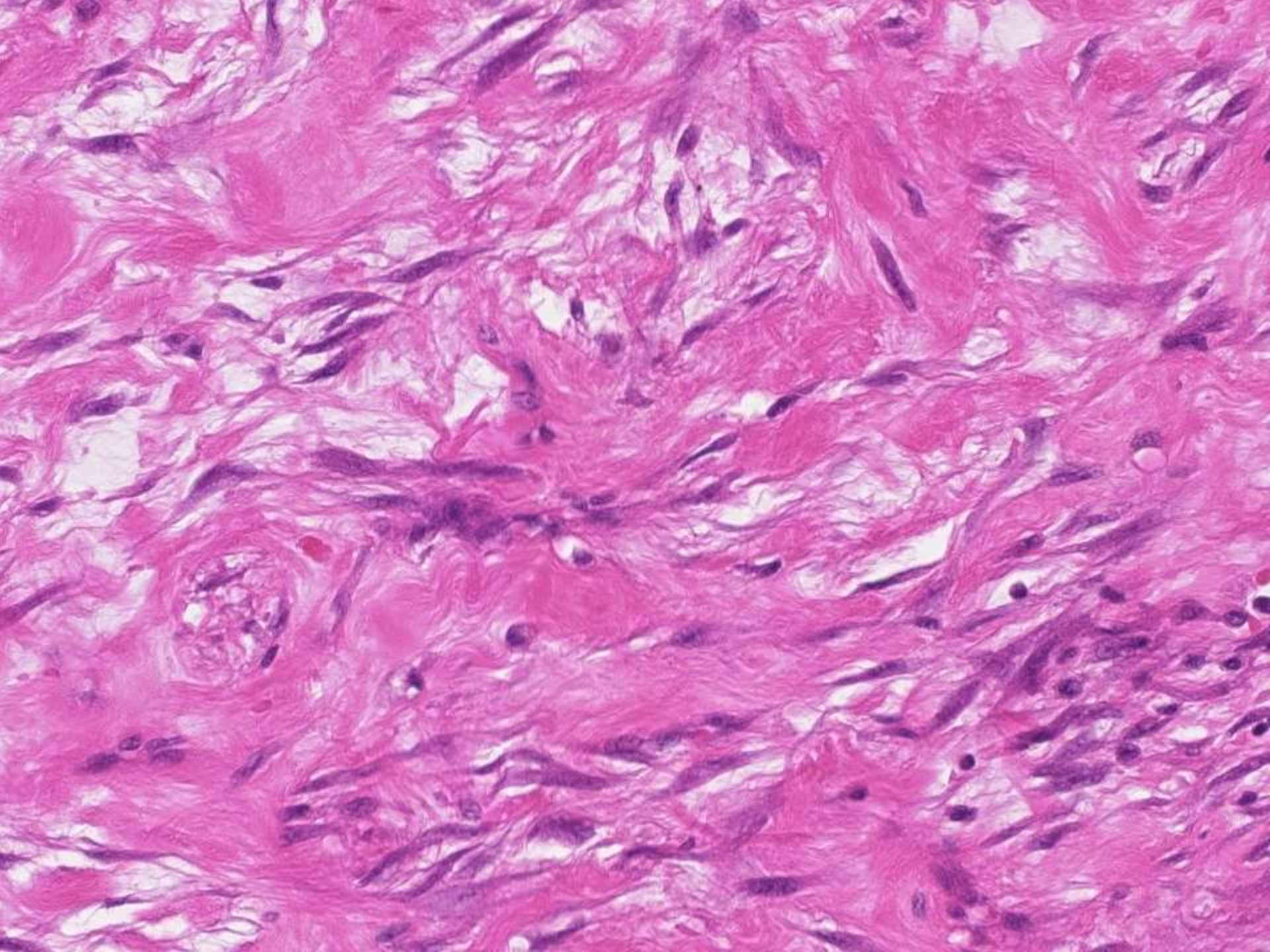


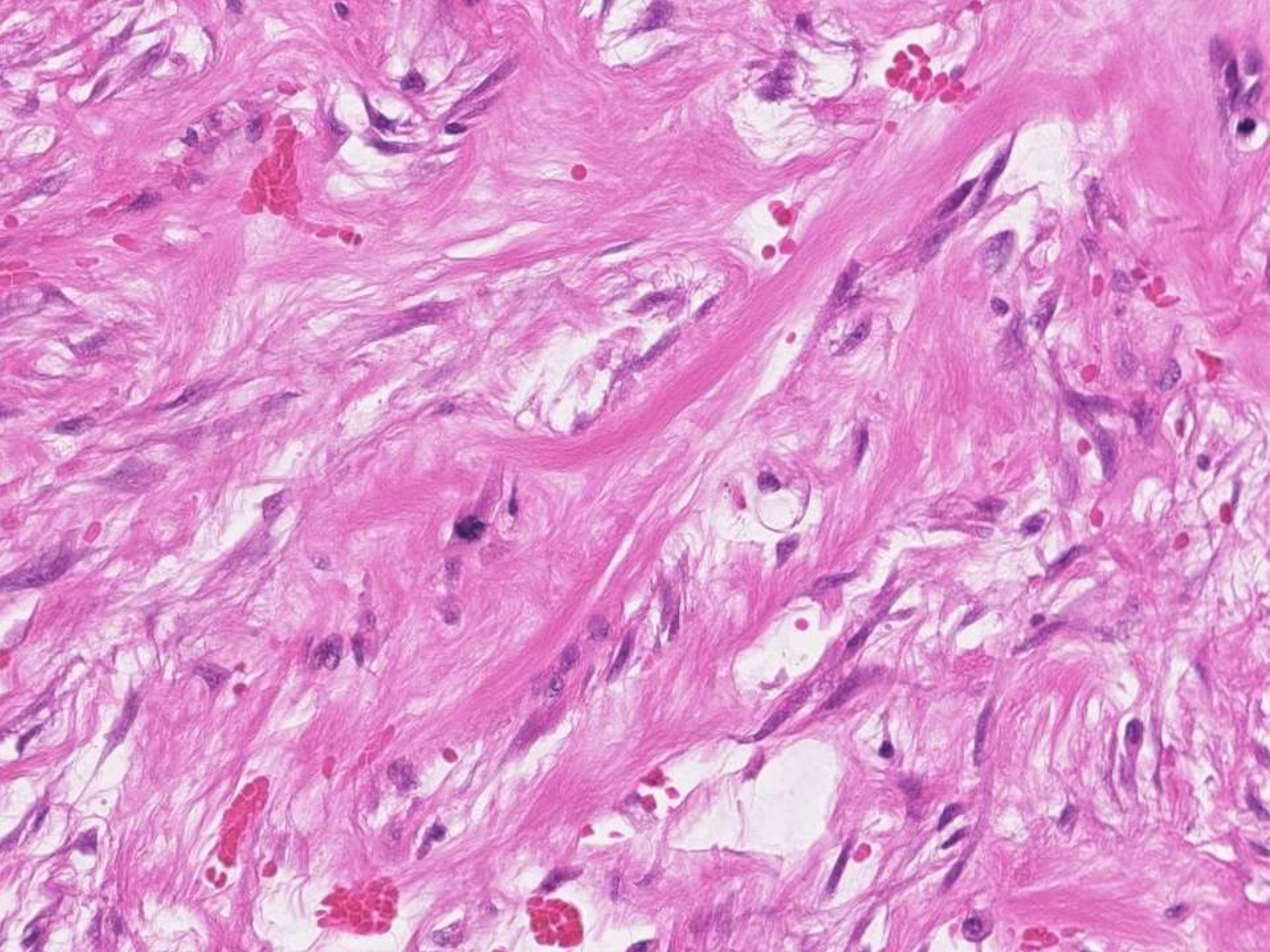












Macroscopic appearance:-

Macroscopically, nodular fasciitis may appear circumscribed or infiltrative, but it is not encapsulated. The cut surface varies from myxoid to fibrous, and occasionally there is central cystic change. Intravascular fasciitis ranges from nodular to plexiform, the latter contour resulting when there is extensive intravascular growth. Cranial fasciitis is typically circumscribed and rubbery to firm, and it may be focally myxoid or cystic in its centre.

Histopathology:-

Nodular fasciitis is composed of plump spindle-shaped cells lacking nuclear hyperchromasia or pleomorphism. Mitotic figures may be plentiful, but atypical forms are not observed. The lesion may be highly cellular, but typically it is partly discohesive and myxoid, with a torn, feathery, or tissue culture–like character. In more-cellular areas, there is often growth in S-shaped or C-shaped fascicles, or sometimes in a storiform pattern. There is normally little collagen, but collagen may be increased focally, and keloidal collagen bundles may be present and occasionally prominent. Microcystic stromal changes are also typical. Extravasated erythrocytes, lymphocytes, and osteoclast-like giant cells are frequently identified. The lesional border is typically infiltrative (at least focally), although it may be well delineated; peripheral extension is often seen between fat cells in the subcutis and between muscle cells in intramuscular locations. Small vessels are numerous, which may occasionally result in a resemblance to granulation tissue.

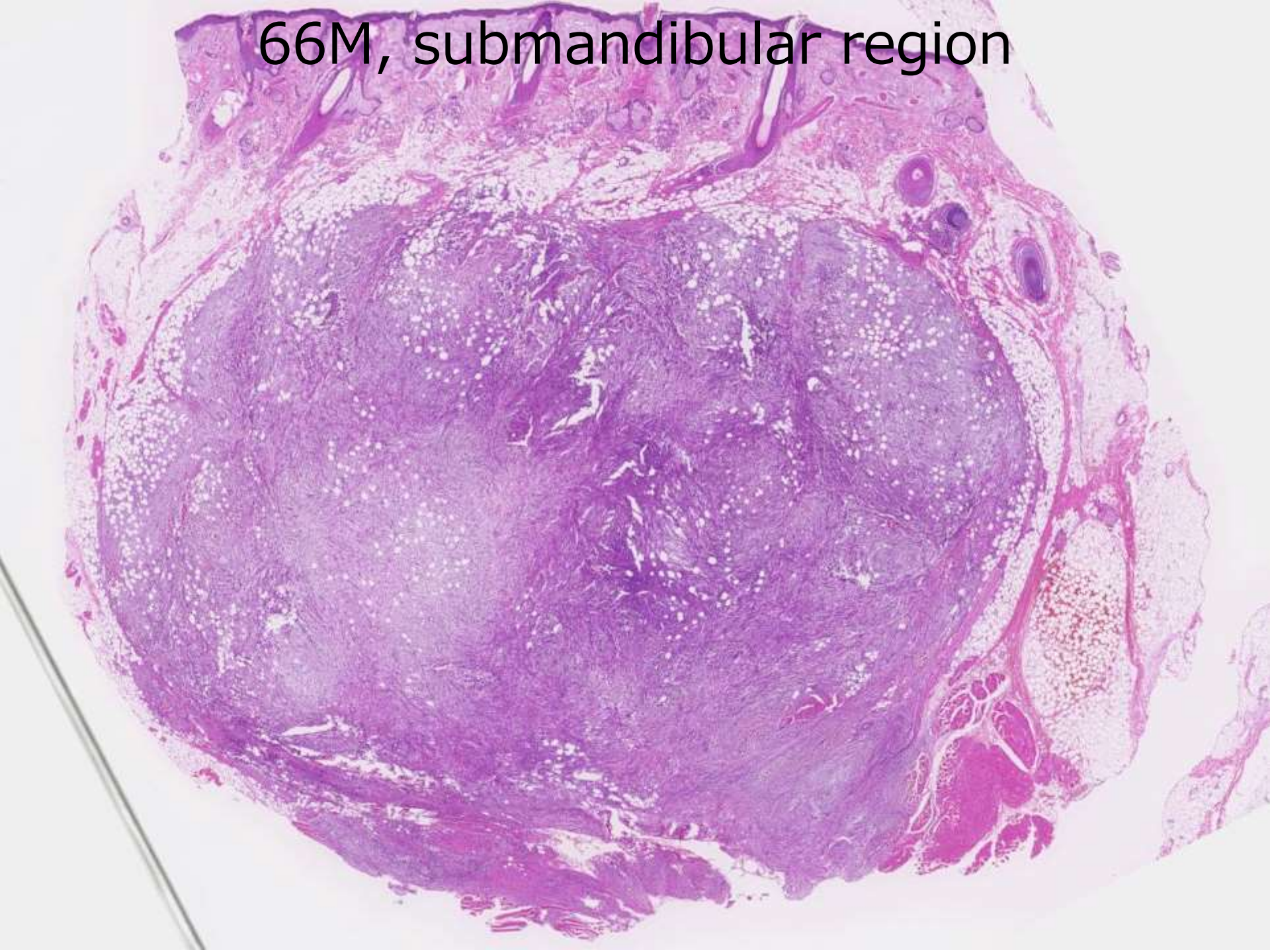
Intravascular fasciitis and cranial fasciitis are similar to nodular fasciitis histologically, although intravascular fasciitis often displays a greater number of osteoclast-like giant cells. Intravascular fasciitis ranges from predominantly extravascular, with only a minor intravascular component, to predominantly intravascular. Osseous metaplasia is occasionally seen in nodular fasciitis (fasciitis ossificans) and cranial fasciitis { 6814399 ; 4974865 }.

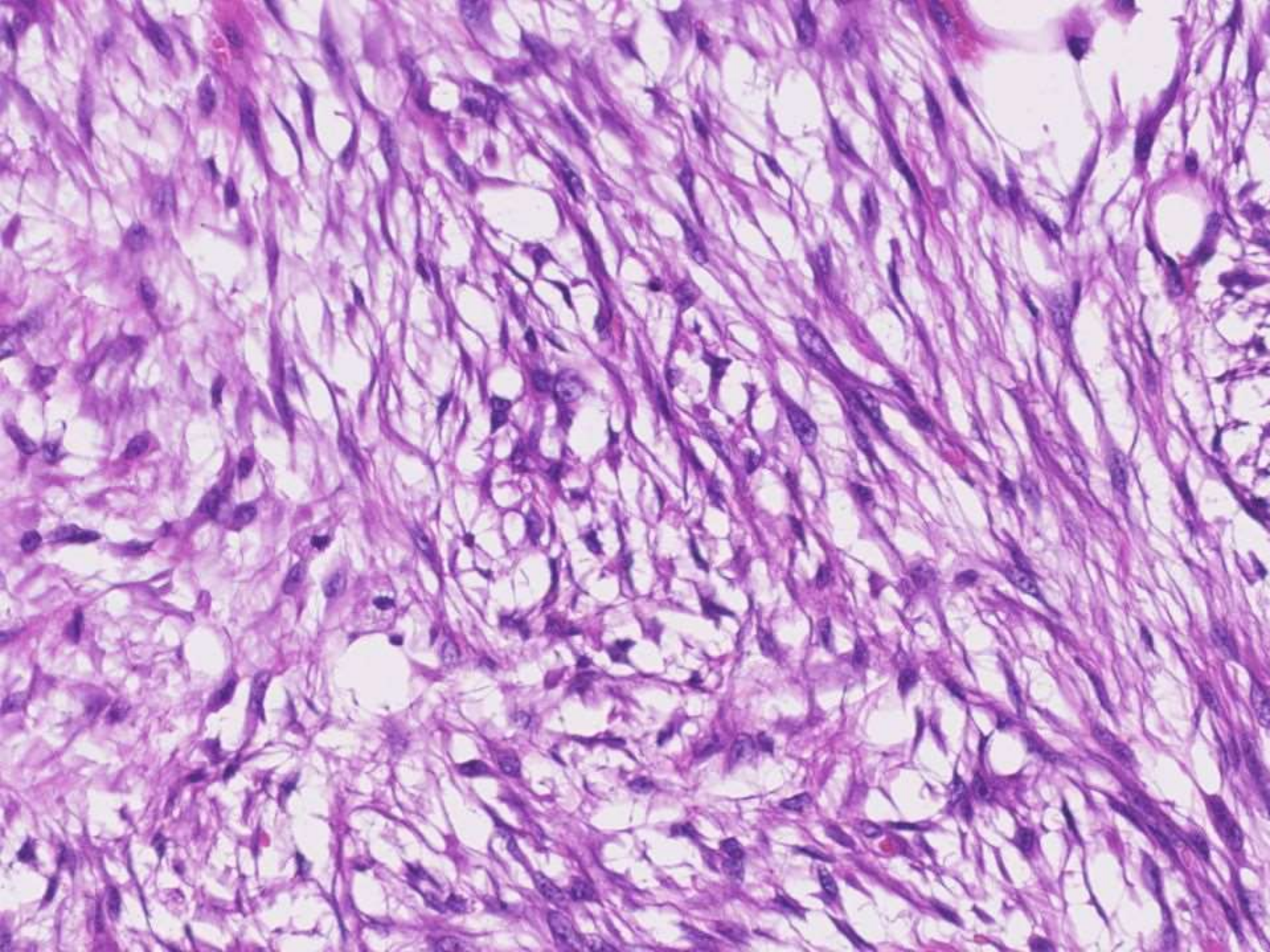
By immunohistochemistry, the neoplastic cells express SMA and MSA in a typical myofibroblastic (tram-track) pattern; desmin

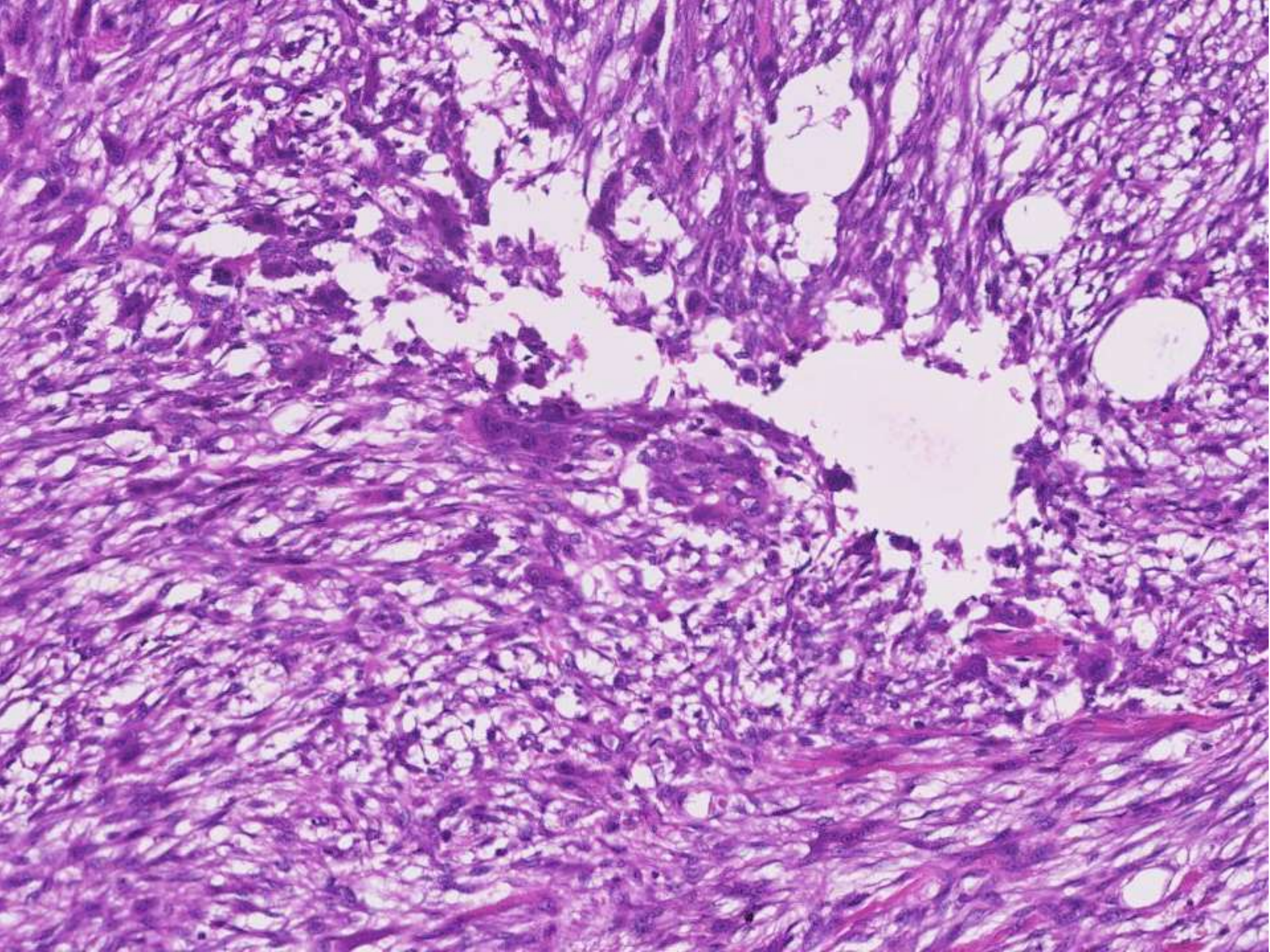
Tips 1

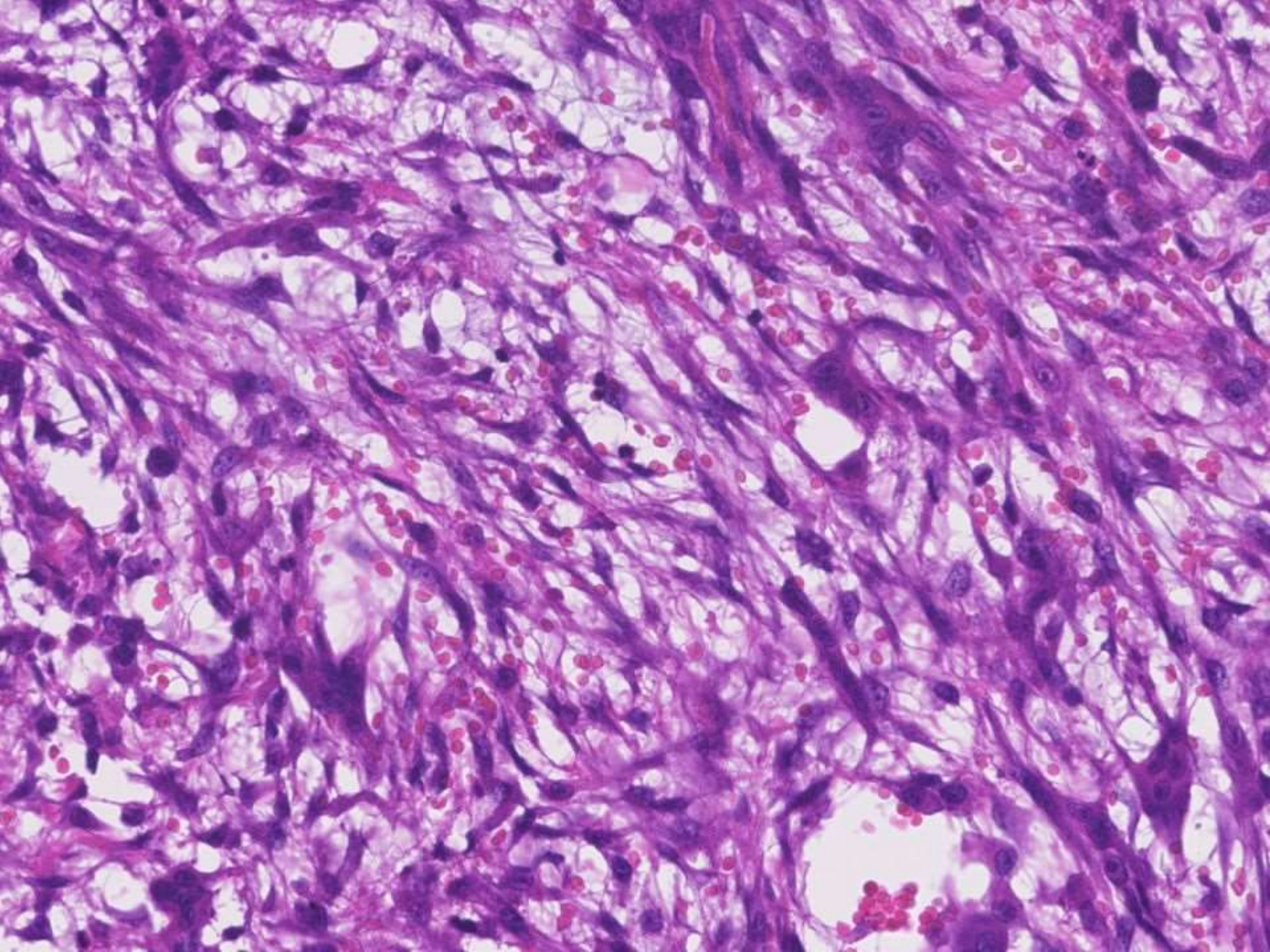
異型がはっきり分からない紡錘形細胞から成る軟部腫瘍を見たら、**まず**結節性筋膜炎の可能性を検討

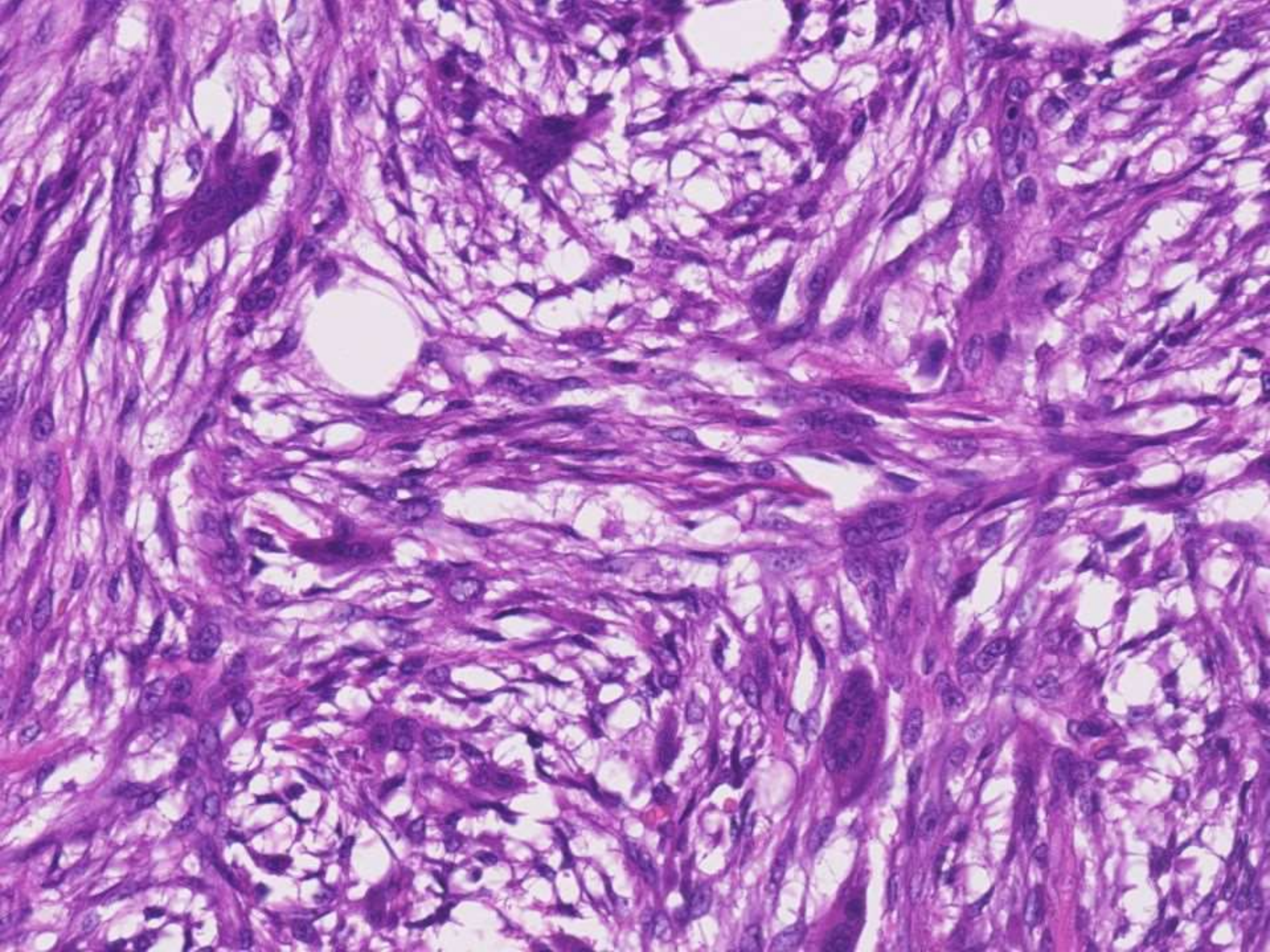
- 年齢、大きさ、発生部位、境界
- **myxoid (+ microcystic), inflammatory cells, extravasation**, giant cells

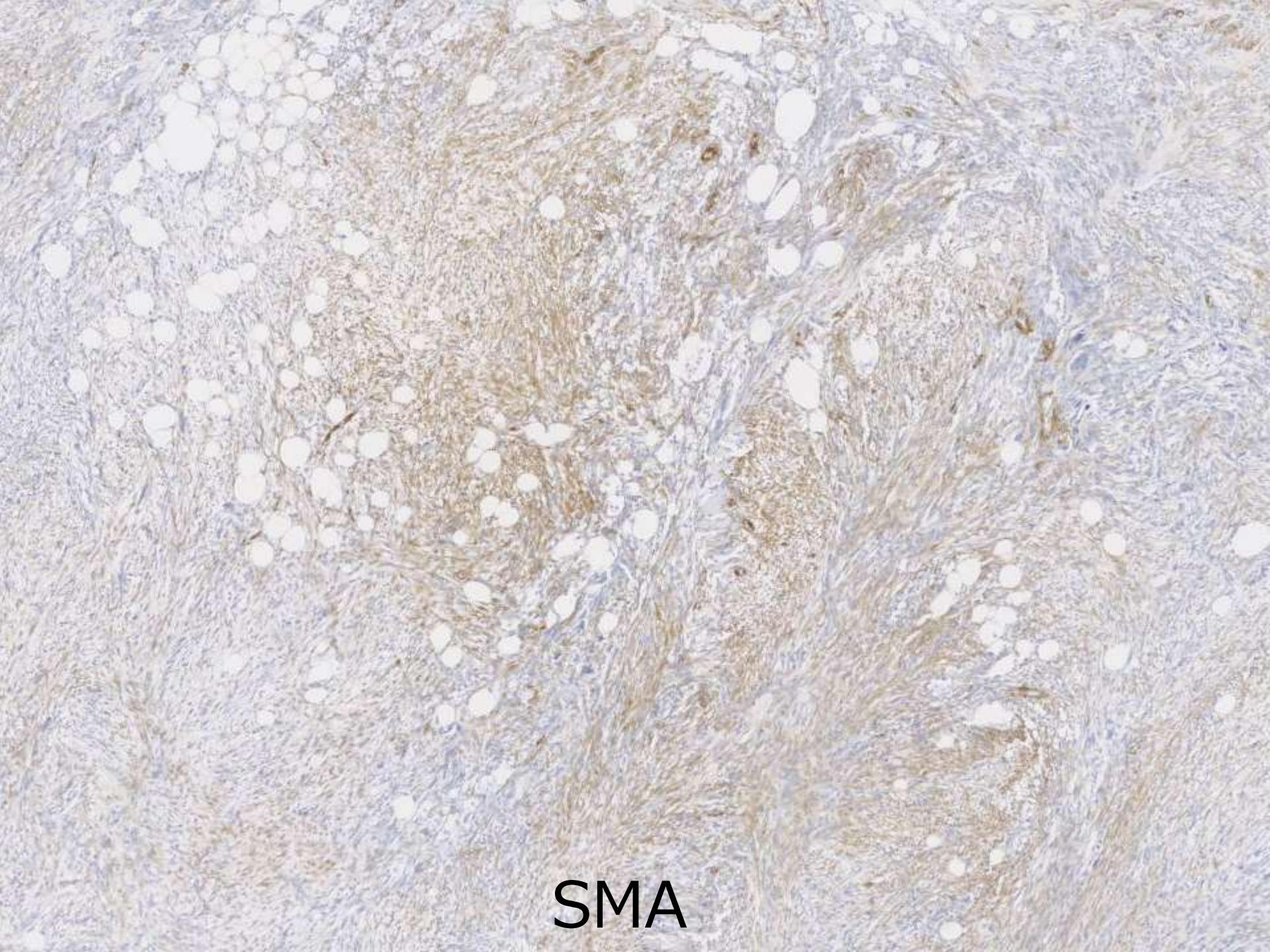

66M, submandibular region

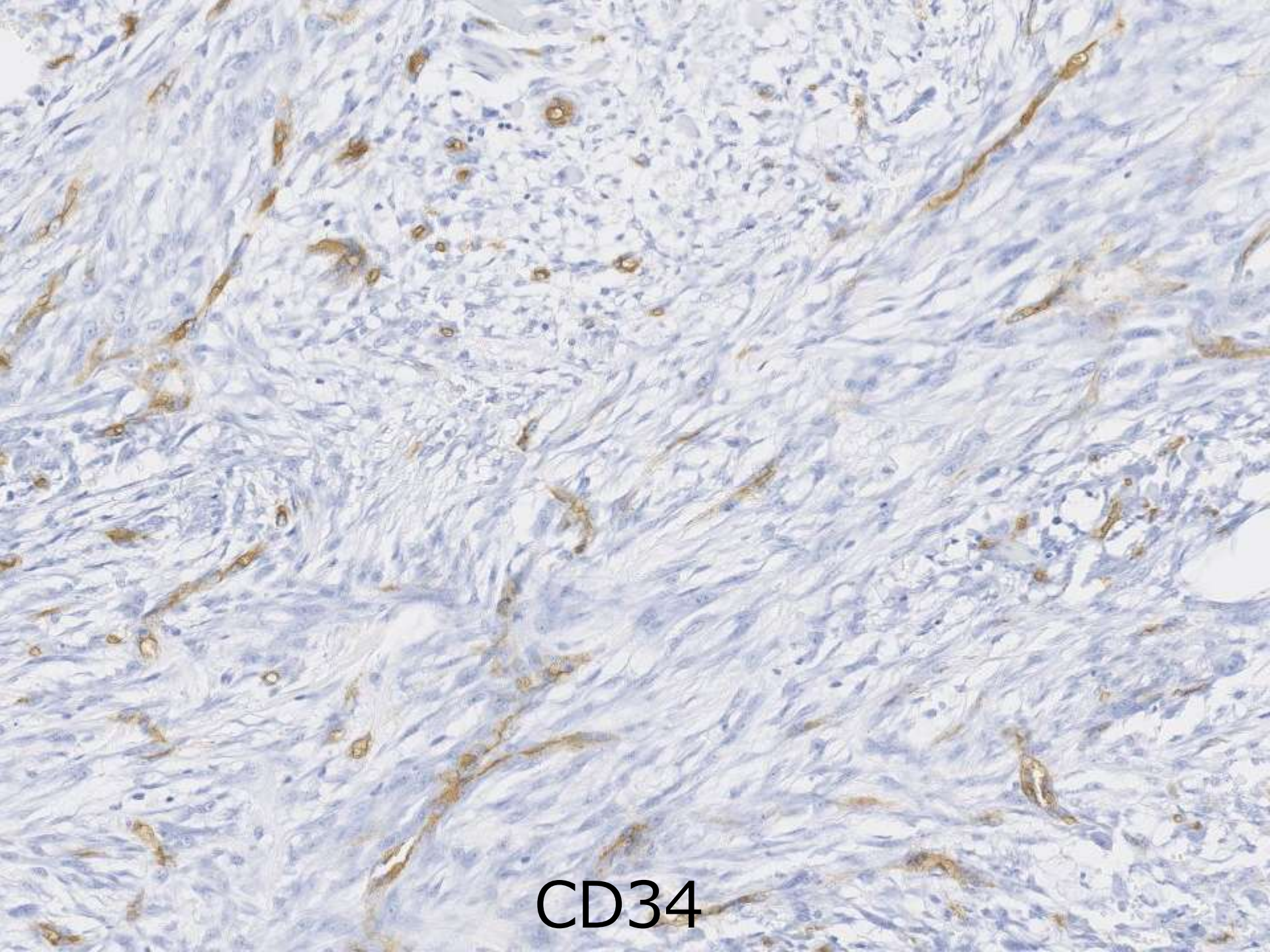




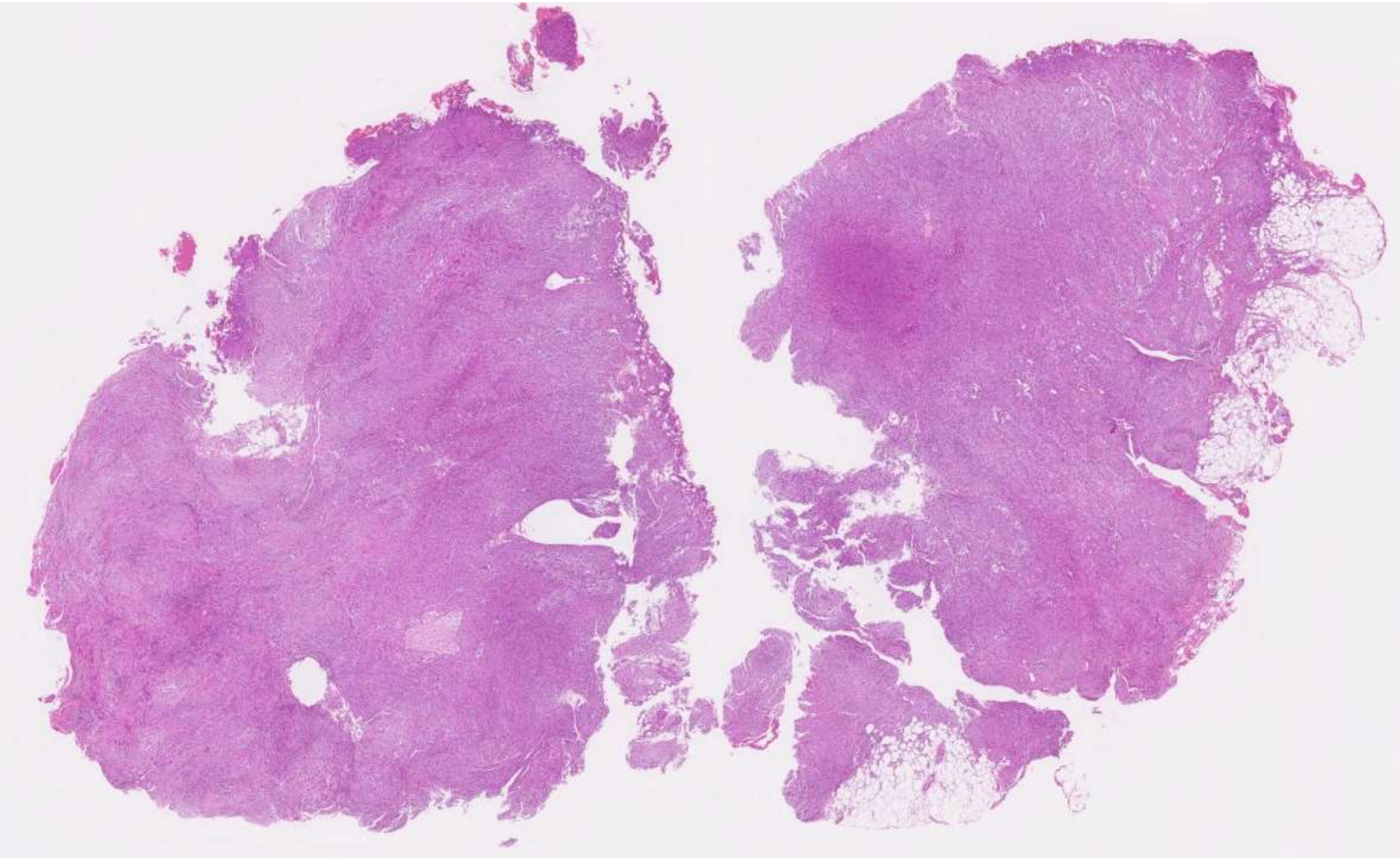


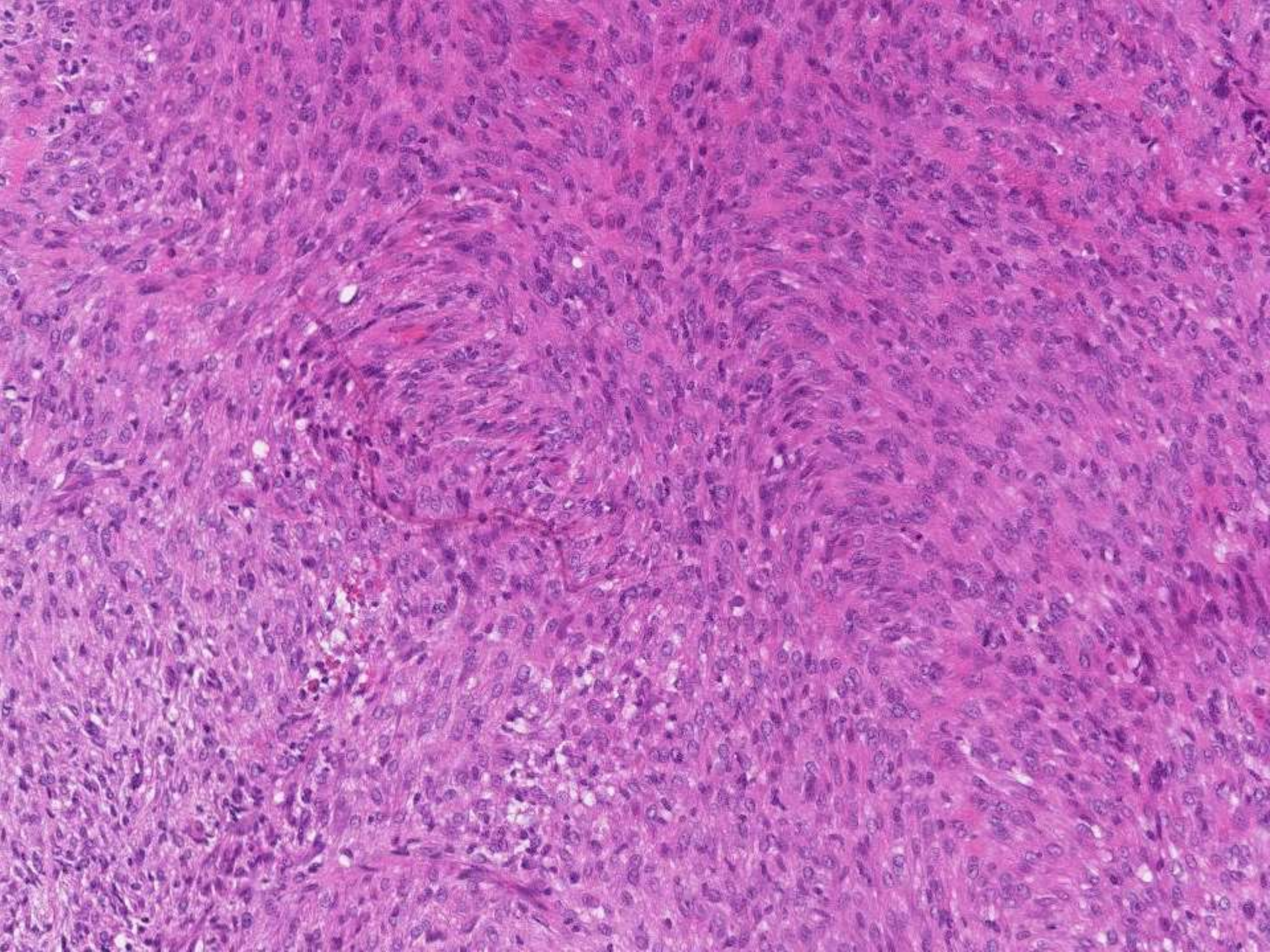


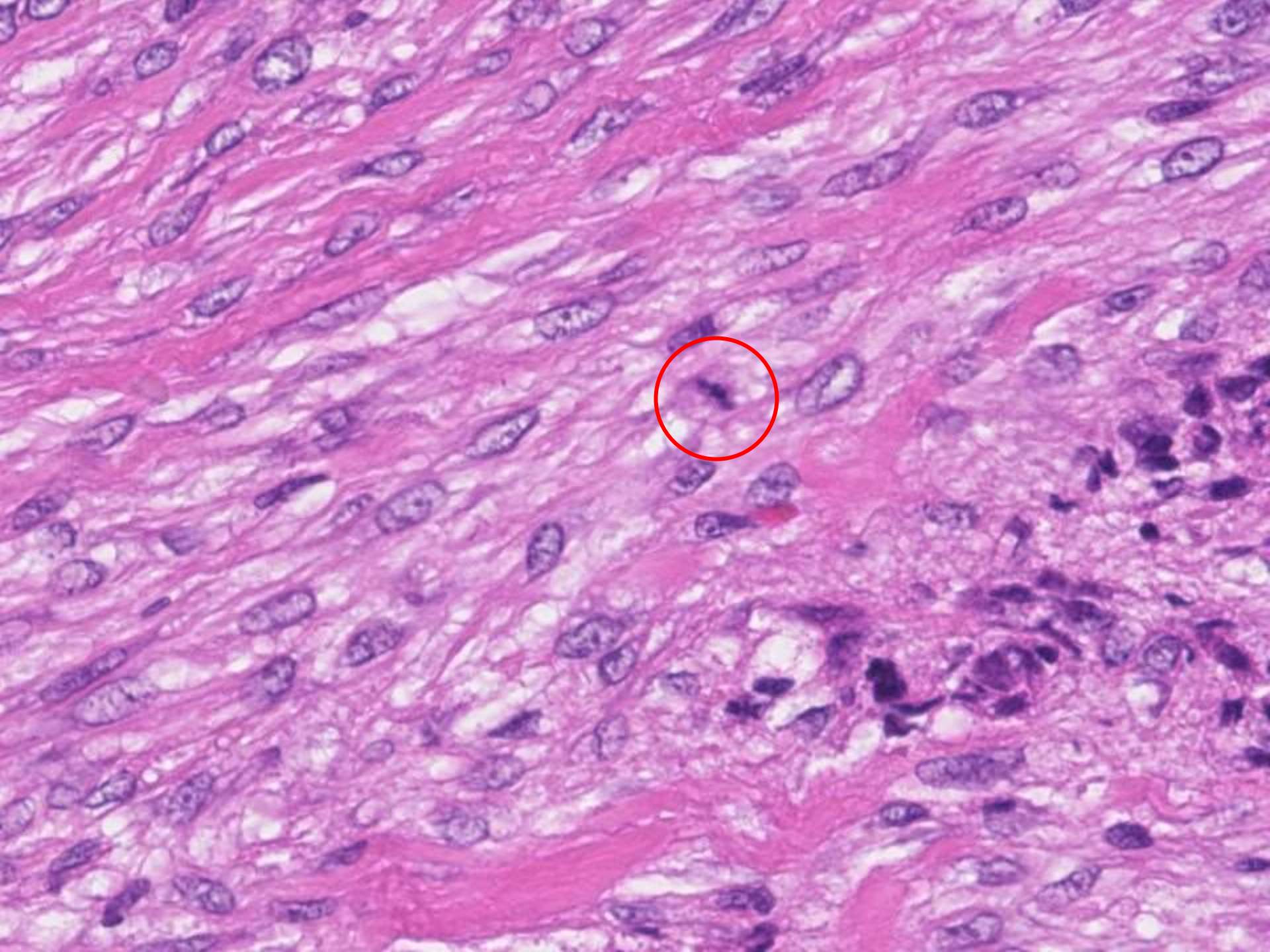

SMA

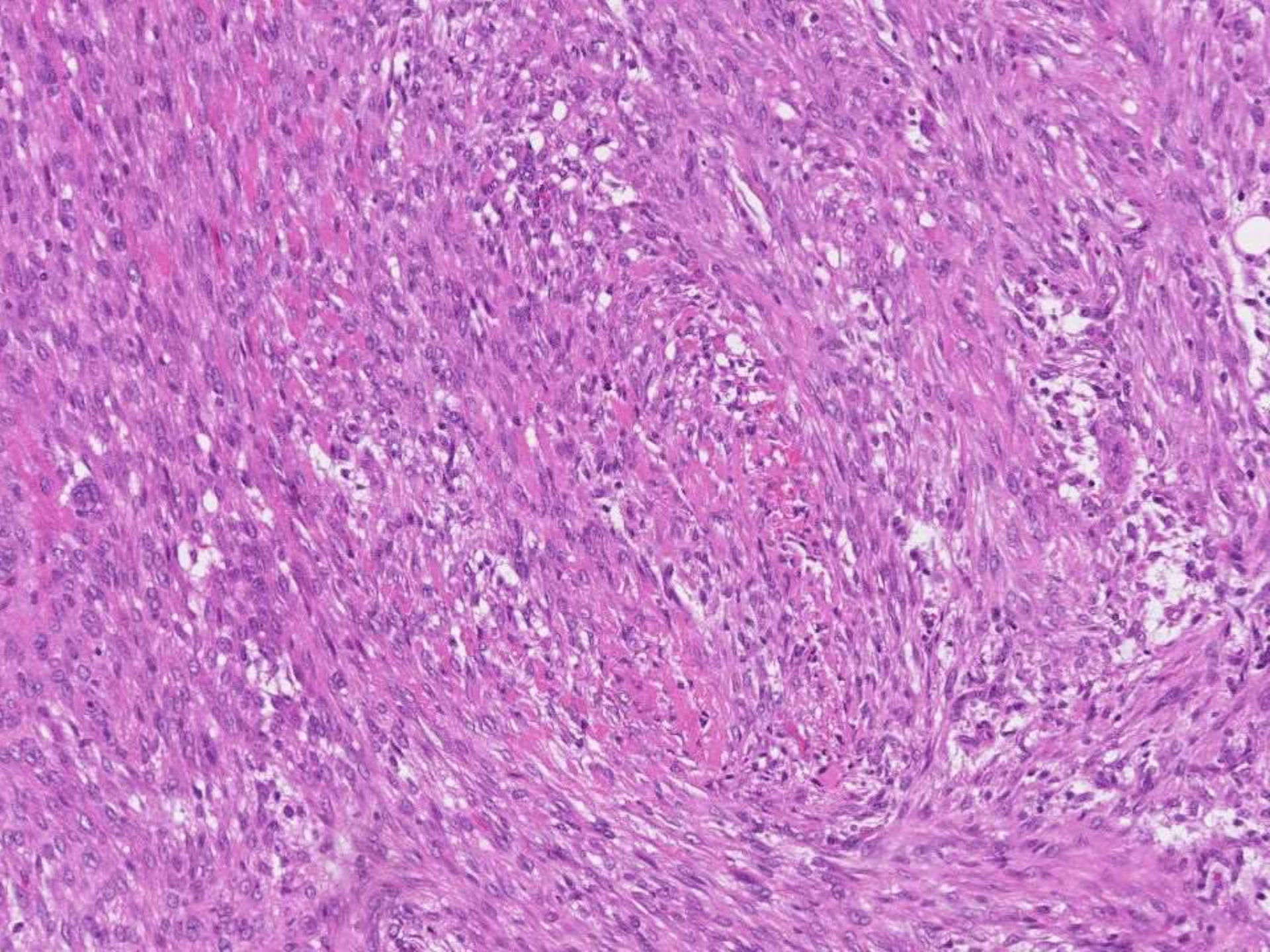

CD34

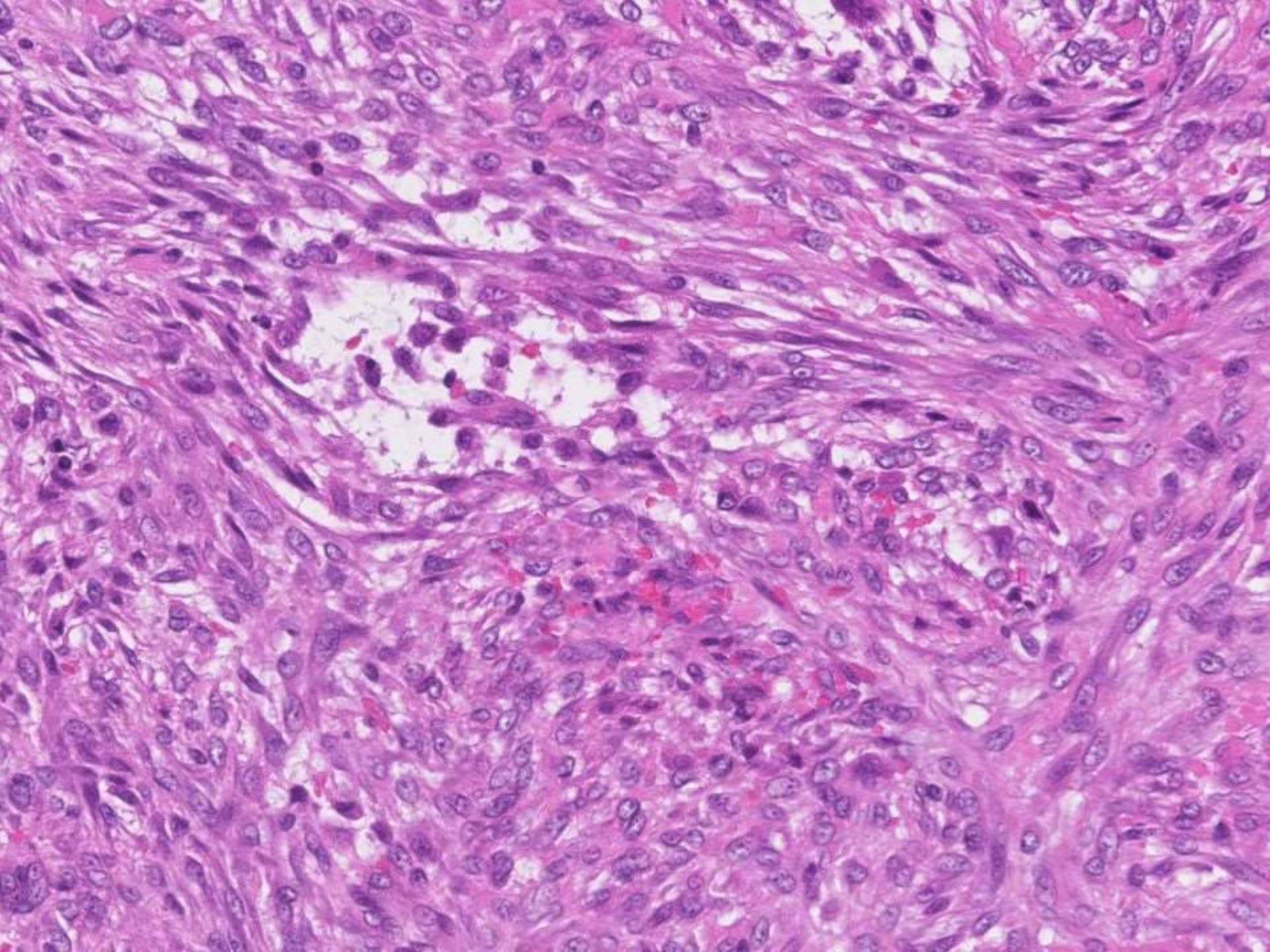
11F, shoulder



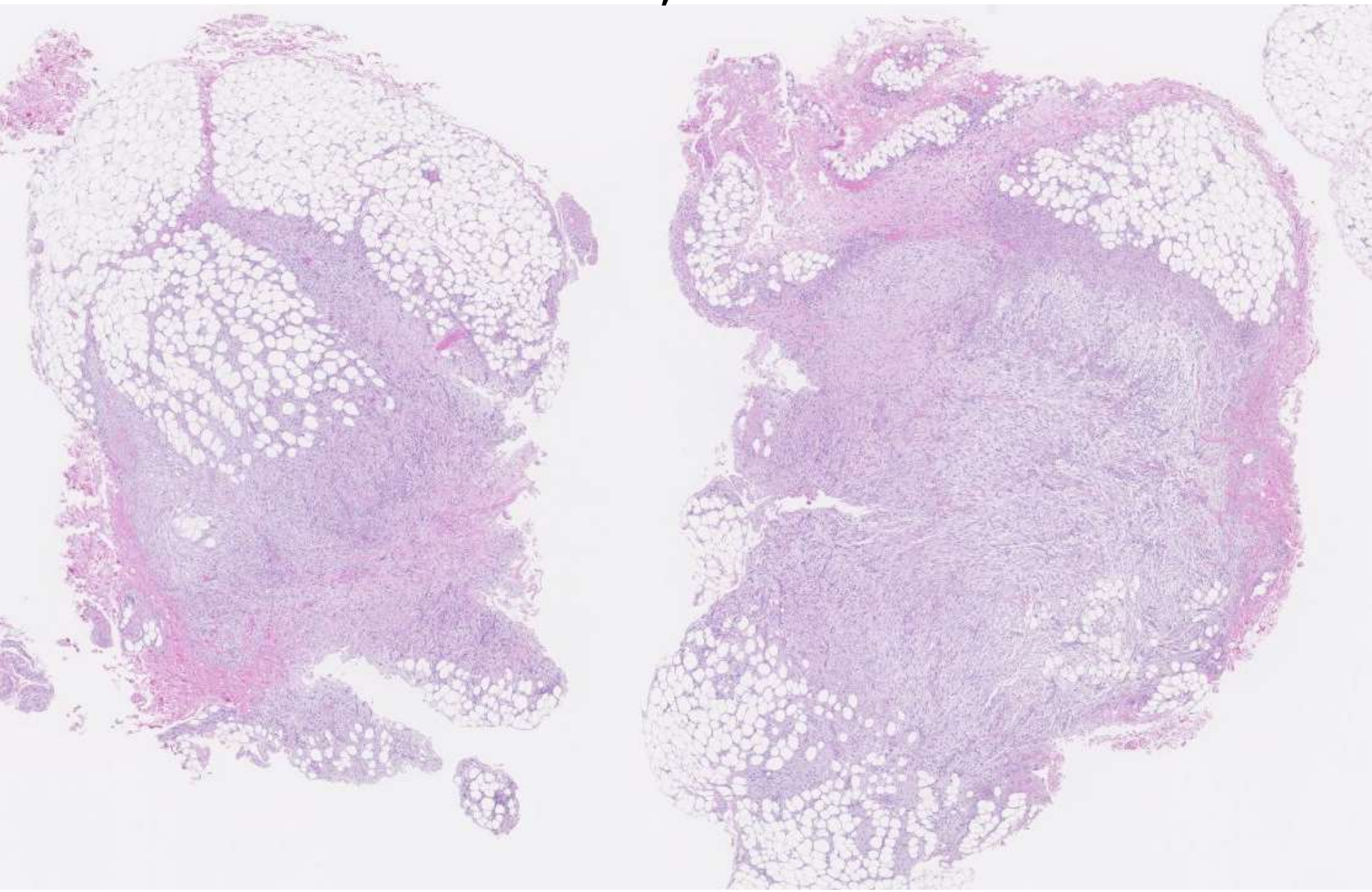


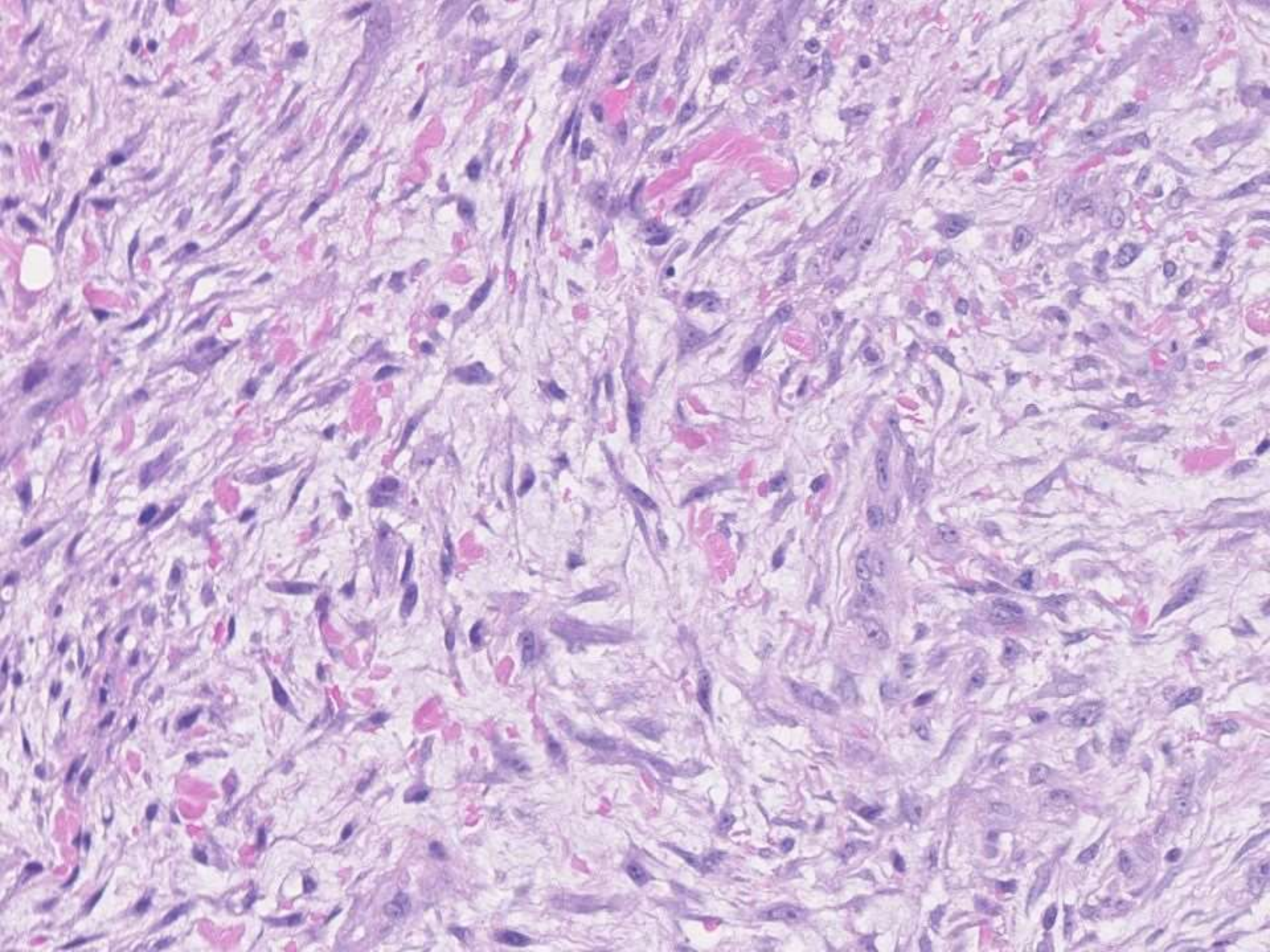


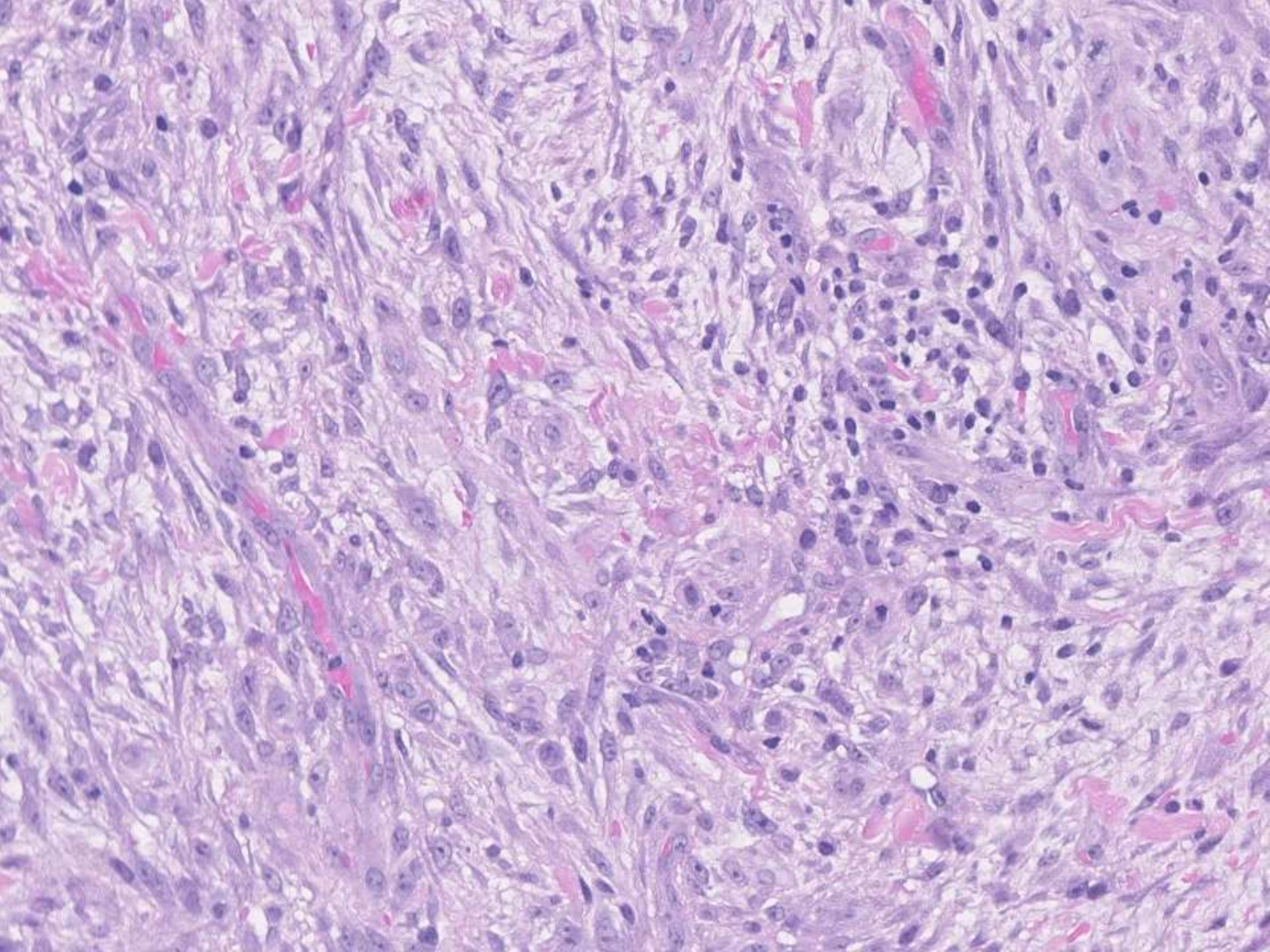


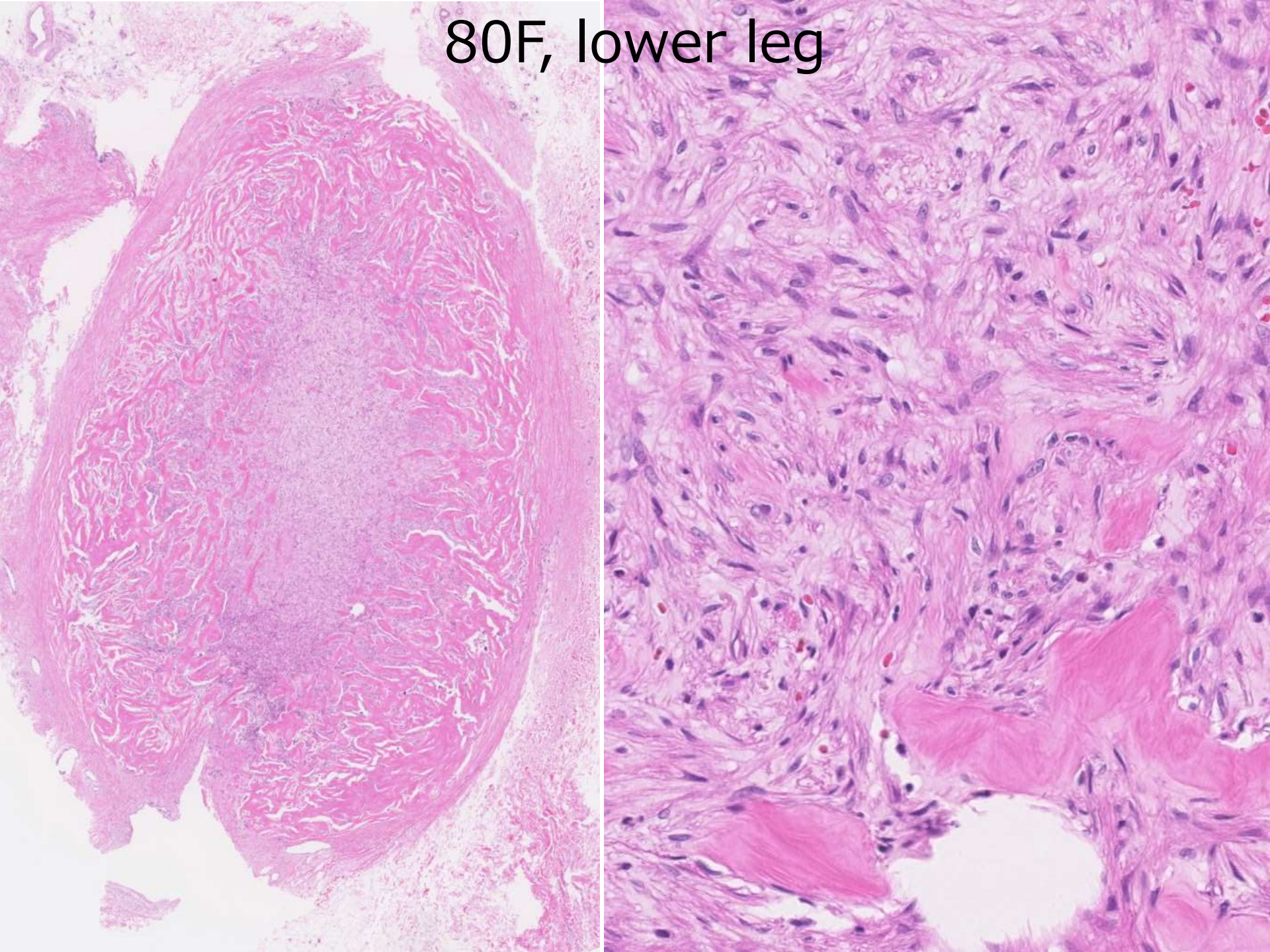


76M, knee





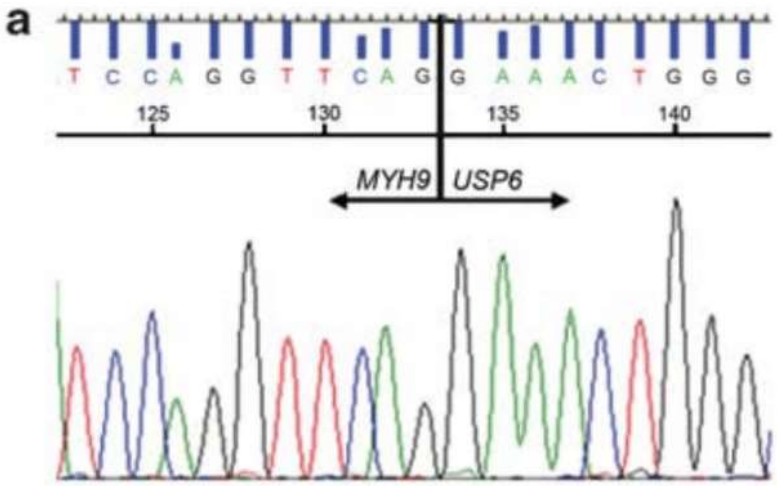



80F, lower leg

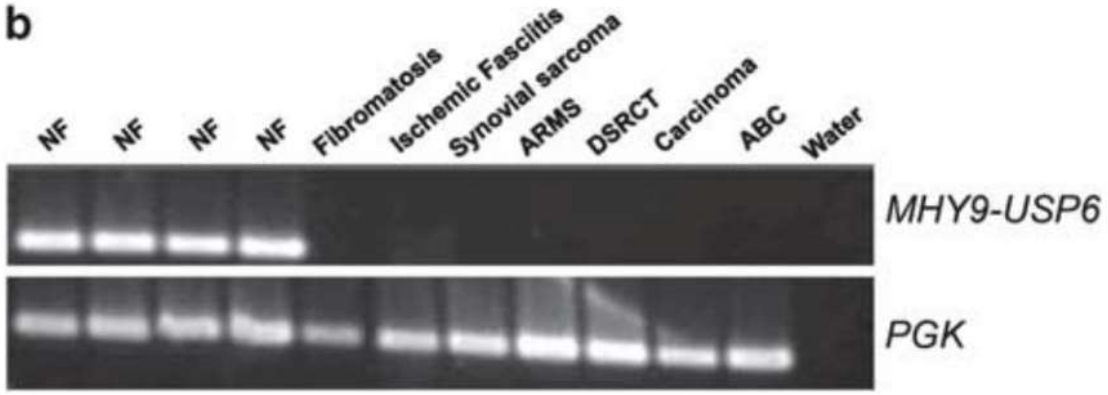
Laboratory Investigation (2011) 91, 1427–1433

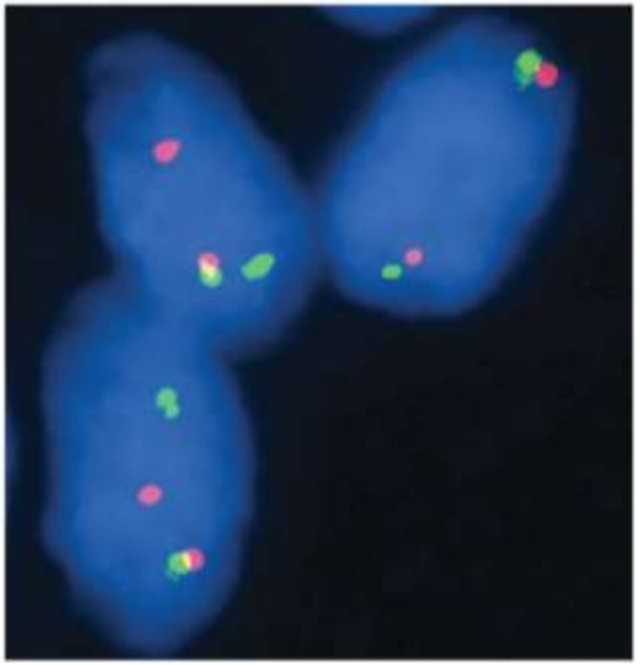
Nodular fasciitis: a novel model of transient neoplasia induced by *MYH9-USP6* gene fusion

Michele R Erickson-Johnson[1,*], Margaret M Chou[2,*], Barbara R Evers[1], Christopher W Roth[1], Amber R Seys[1], Long Jin[1], Ying Ye[2], Alan W Lau[2], Xiaoke Wang[1] and Andre M Oliveira[1]



USP6 rearrangement in 44/48 NFs
MYH9-USP6 fusion in 12/48 NFs

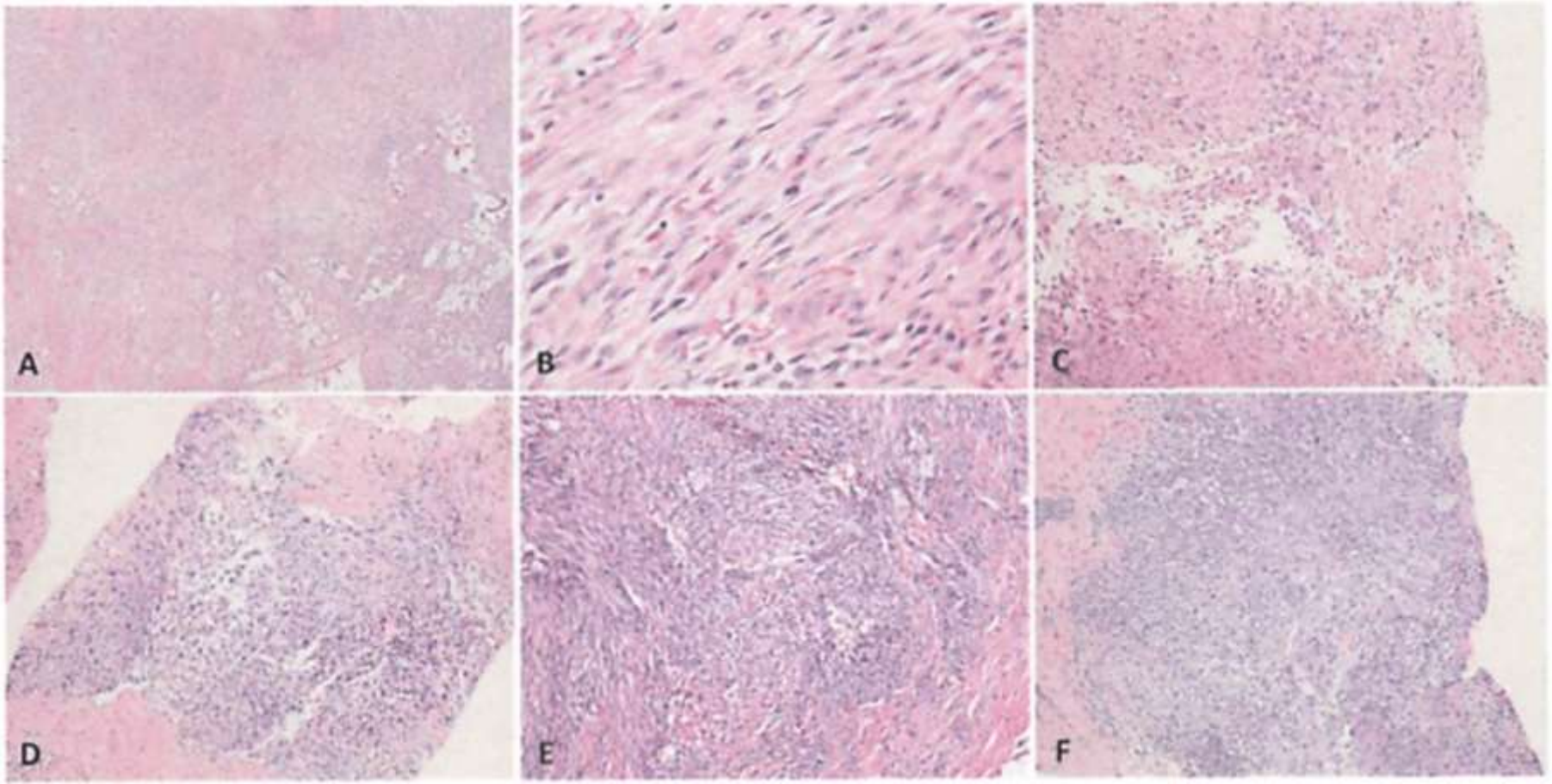




GENES, CHROMOSOMES & CANCER 55:640–649 (2016)

PPP6R3-USP6 Amplification: Novel Oncogenic Mechanism in Malignant Nodular Fasciitis

Ruifeng Guo,[1,2†] Xiaoke Wang,[1†] Margaret M Chou,[5] Yan Asmann,[4] Doris E. Wenger,[3] Alyaa Al-Ibraheemi,[1] Diana W Molavi,[6] Albert Aboulafia,[7] Long Jin,[1] Karen Fritchie,[1] Jennifer L. Oliveira,[1] Robert B. Jenkins,[1] Jennifer J. Westendorf,[1] Jie Dong,[1] and Andre M. Oliveira[1*]



MODERN PATHOLOGY (2016) 29, 865–869

USP6 genetic rearrangements in cellular fibroma of tendon sheath

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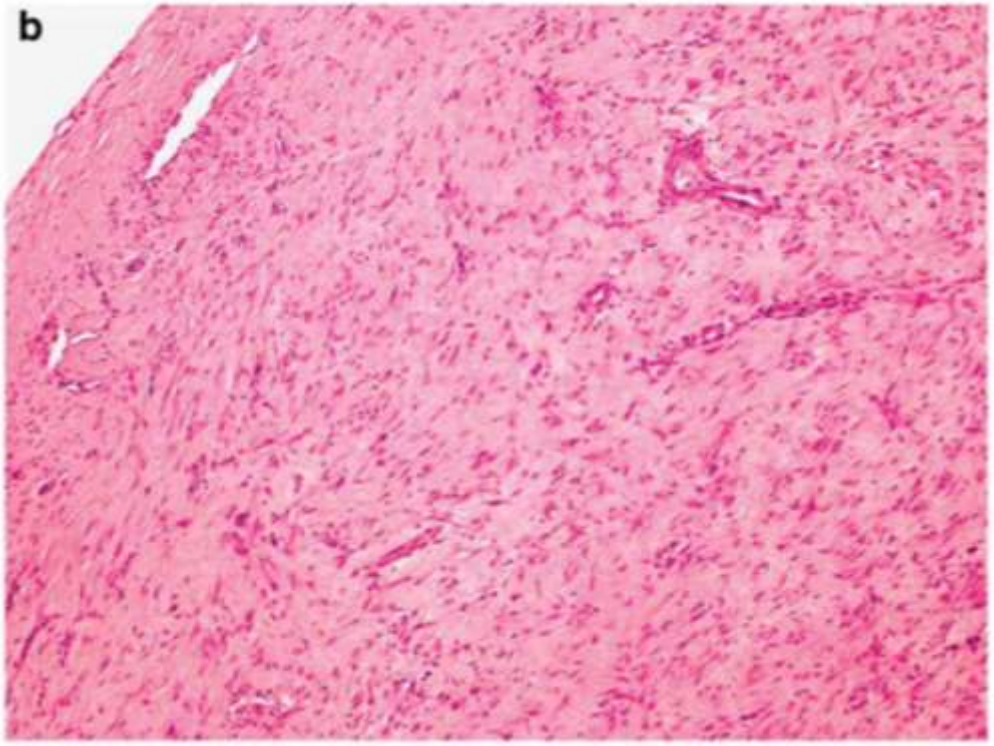
Table 1 Clinicopathological features and *USP6* and *MYH9* rearrangement status

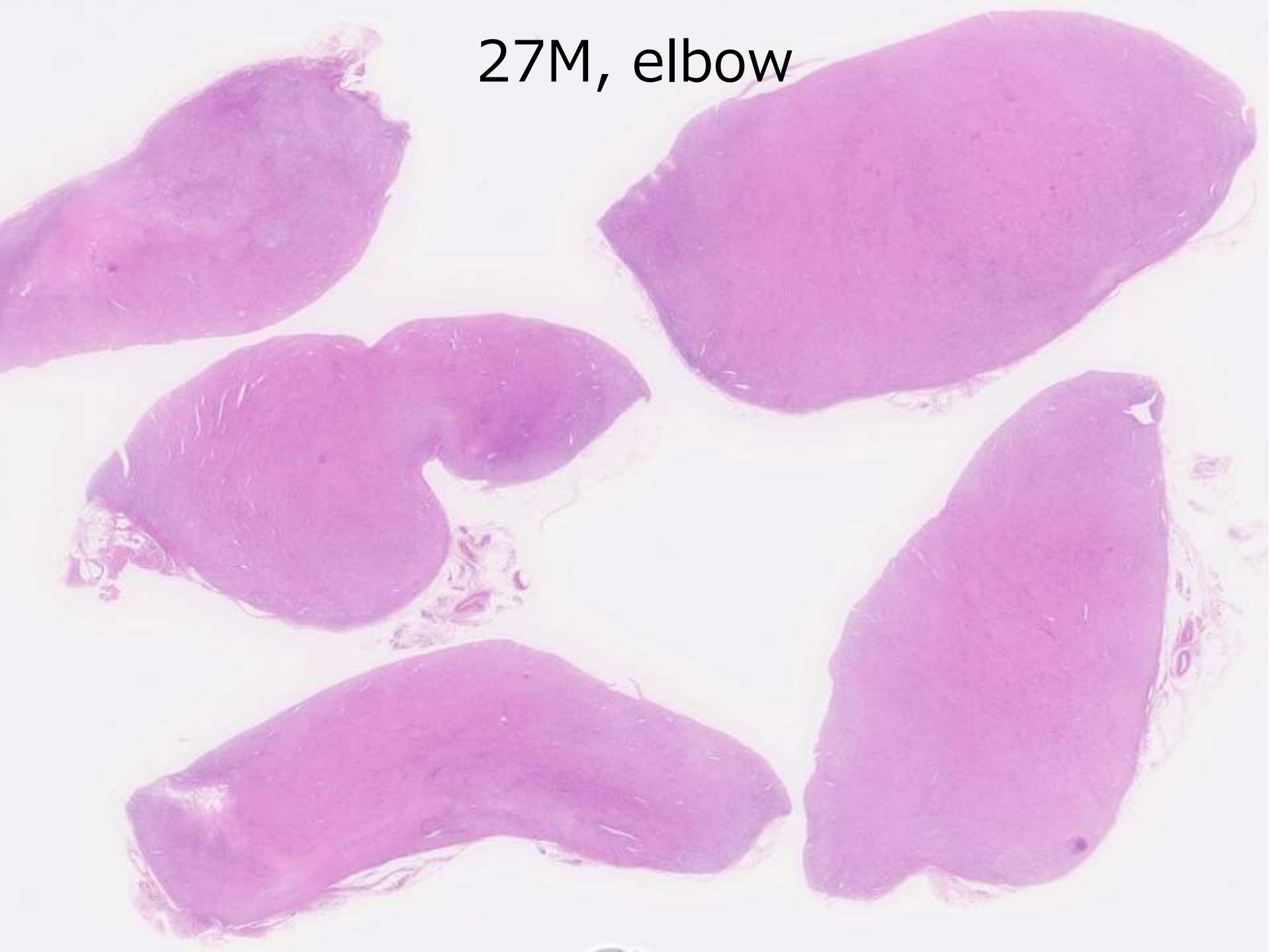
Variant of fibroma of tendon sheath	*Age/sex*	*Site*	*Size (cm)*	*USP6*	*MYH9*
Cellular	24/M	Wrist	1.6	+	–
Cellular	32/F	Hand	NA	+	–
Cellular	33/F	Finger	NA	+	–
Cellular	42/F	Finger	NA	+	–
Cellular	12/M	Finger	NA	+	NA
Cellular	46/M	Hand	NA	+	–
Cellular	26/F	Finger	0.5	–	NA
Cellular	31/M	Thumb	NA	–	NA
Cellular	38/M	Hand	1.1	–	NA
Classic	23/F	Finger	NA	–	NA
Classic	43/F	Finger	0.7	–	NA
Classic	52/F	Forearm	NA	–	–
Classic	53/M	Foot	NA	–	–
Classic	65/F	Hand	1.2	–	NA
Classic	69/M	Hand	1.2	–	NA
Classic	70/M	Wrist	3.6	–	NA
Classic	71/F	Foot	2	–	NA
Classic	71/M	Finger	1	–	NA
Classic	77/M	Hand	6.5	–	NA

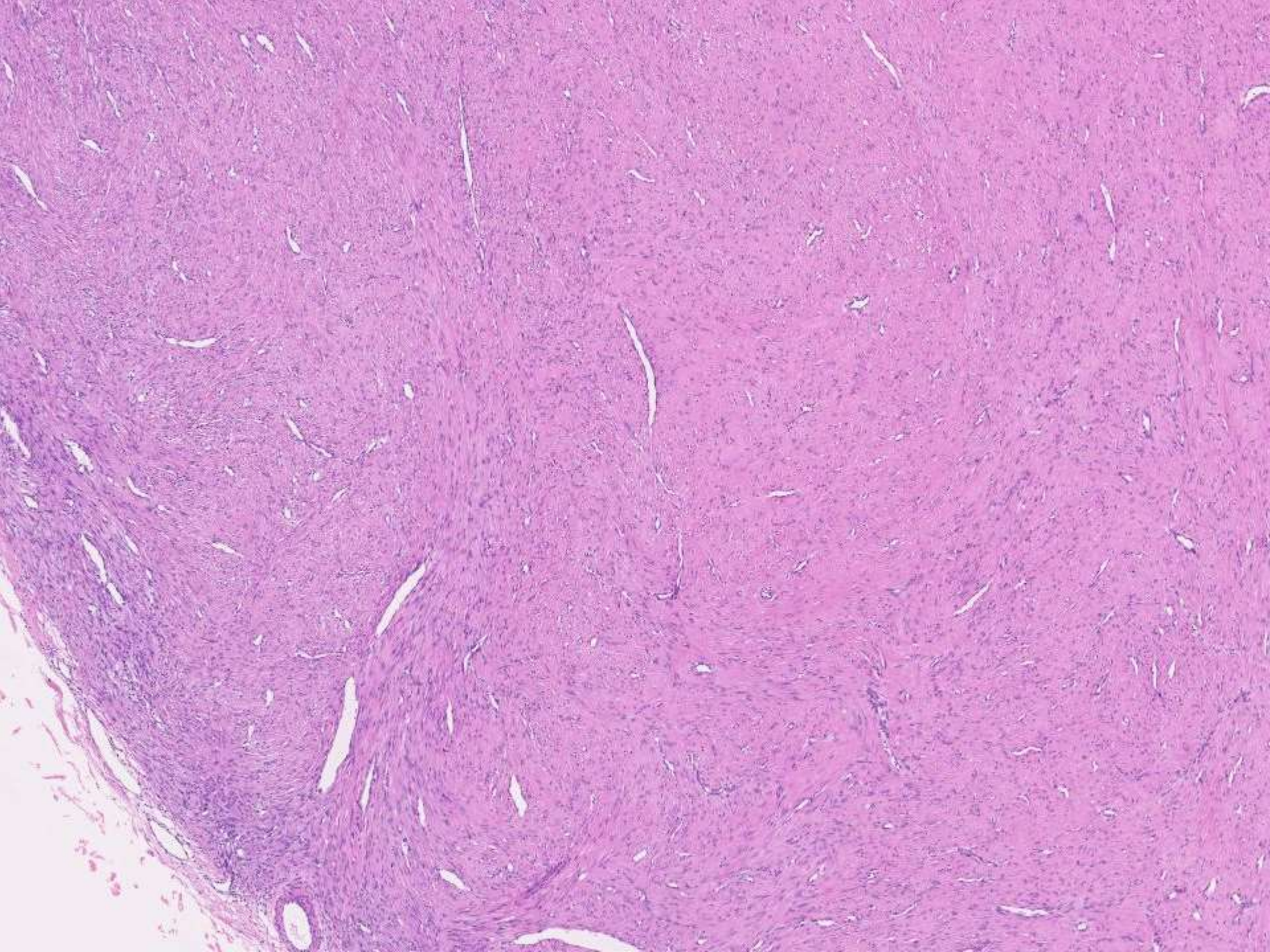


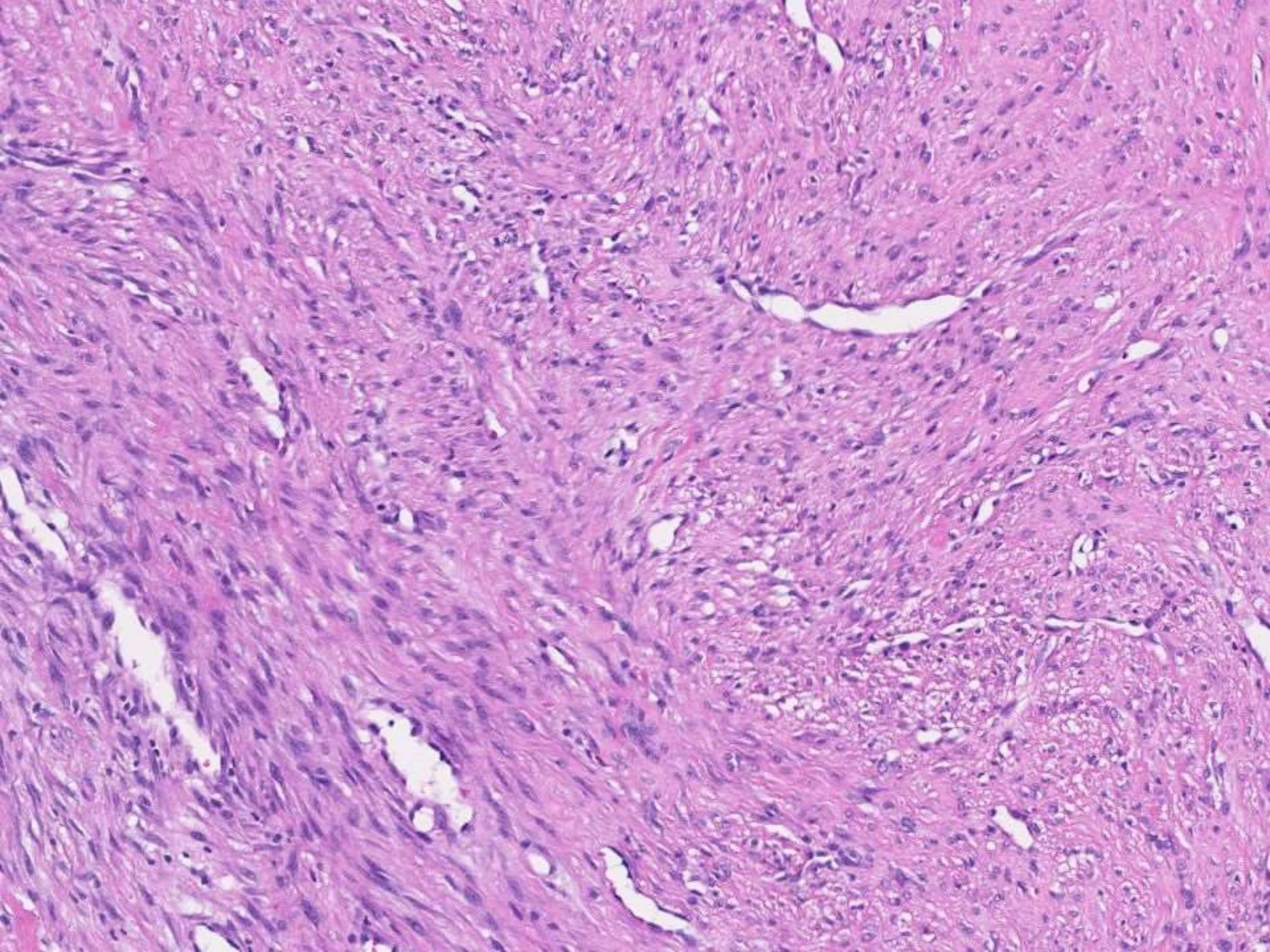
Fibroma of tendon sheath

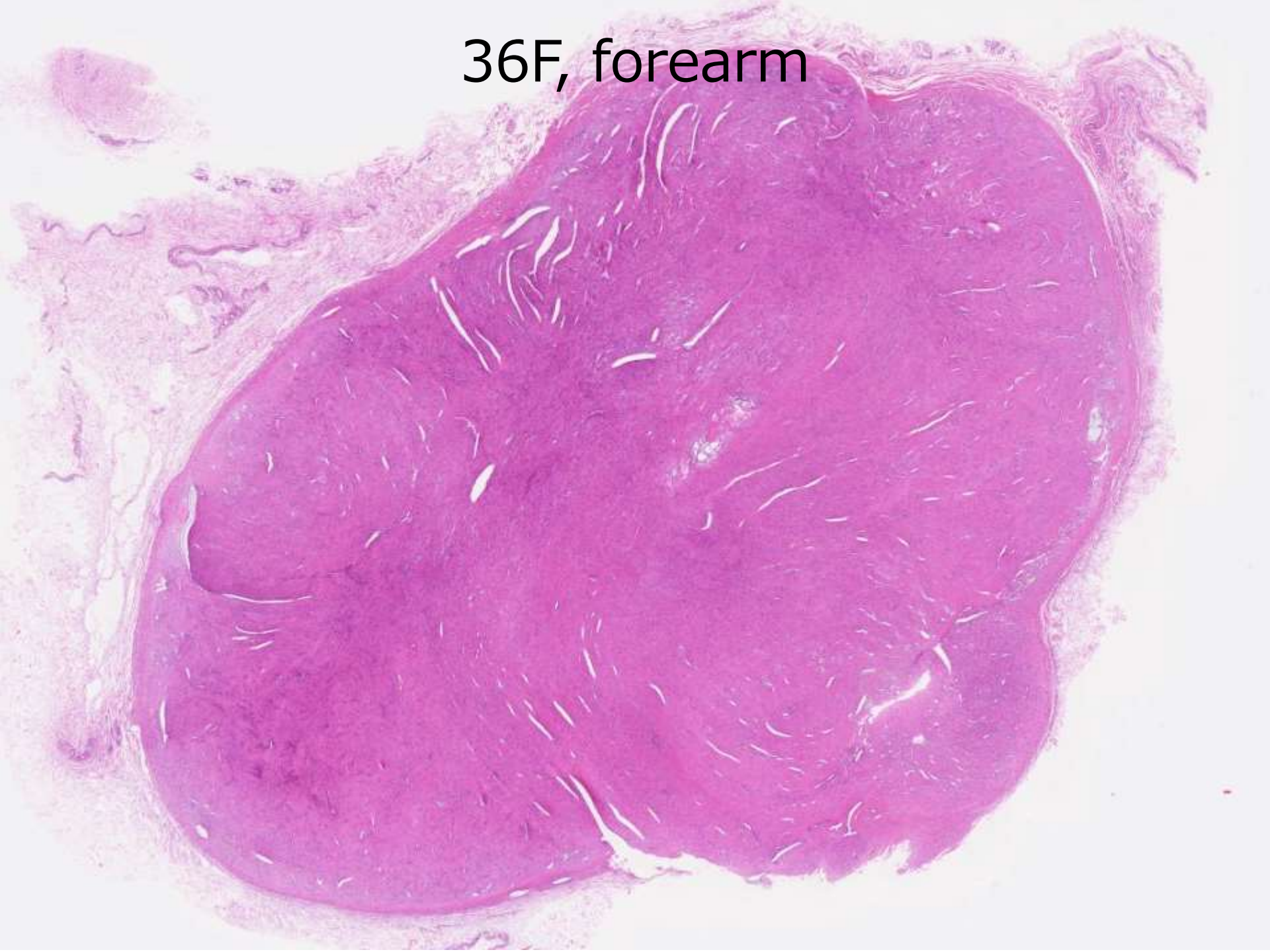
- 若年成人～中年
- 手末梢 58%、足 27%、他 15%
- 2 cm以下
- 境界明瞭

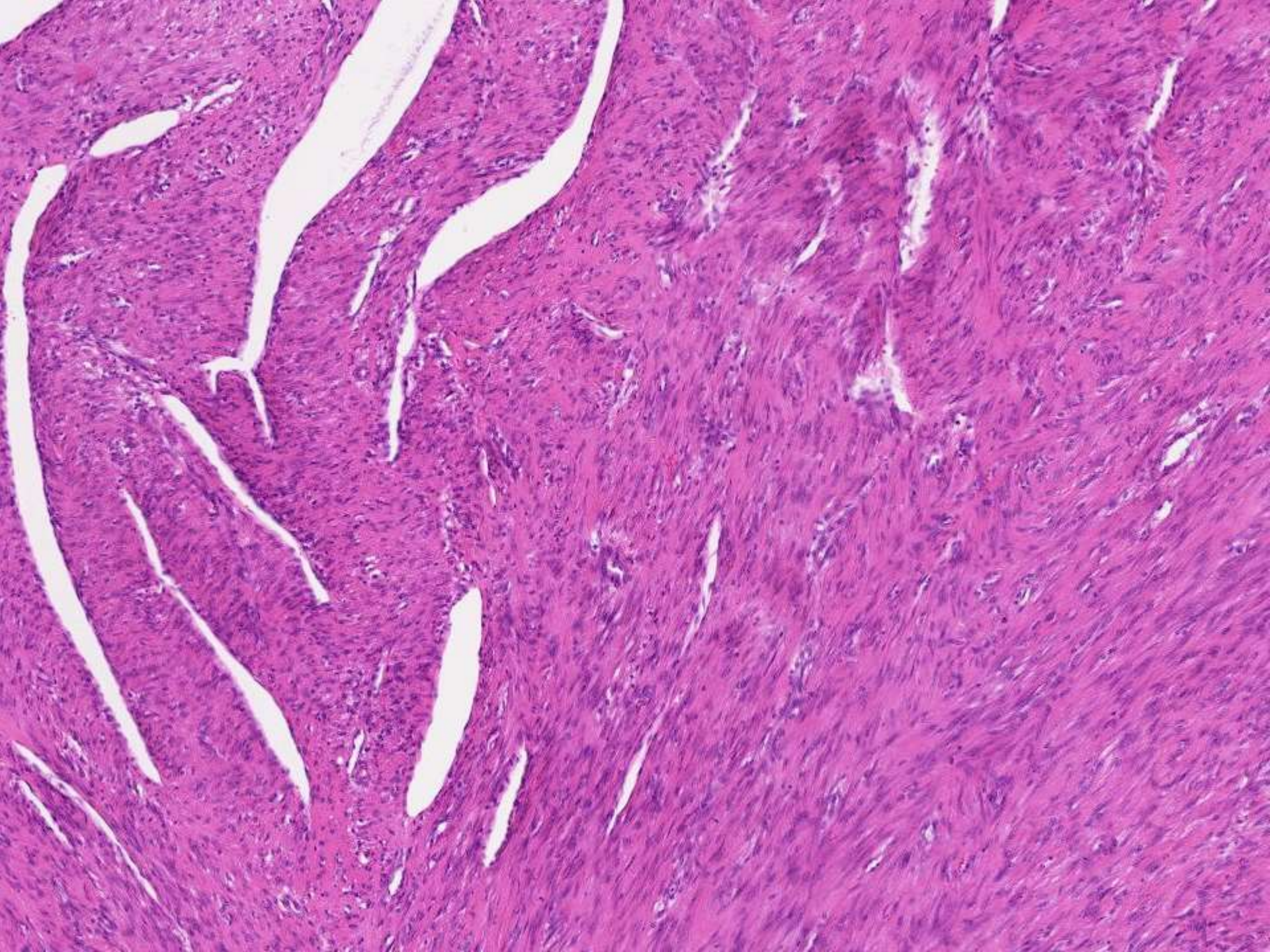
27M, elbow

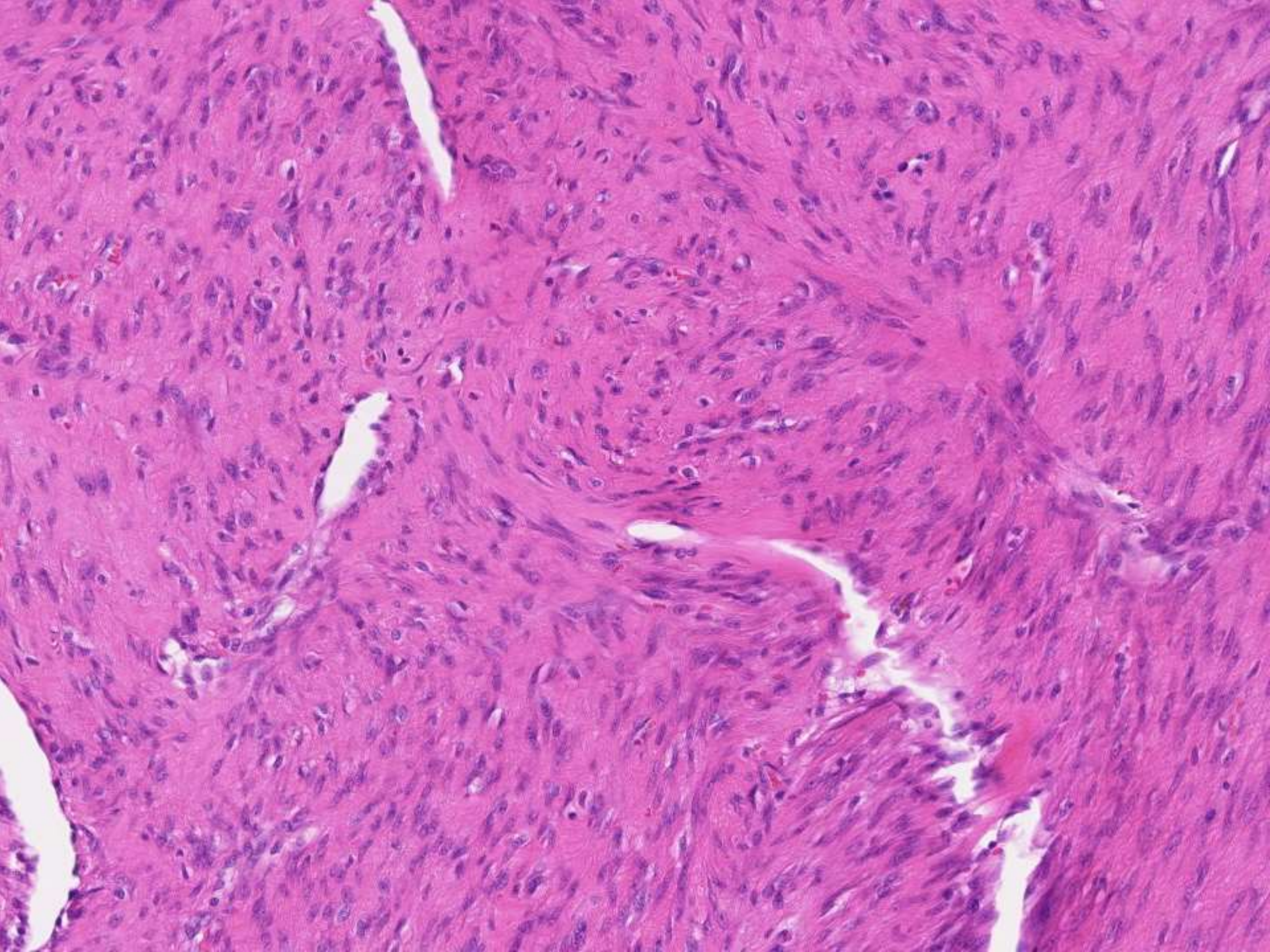


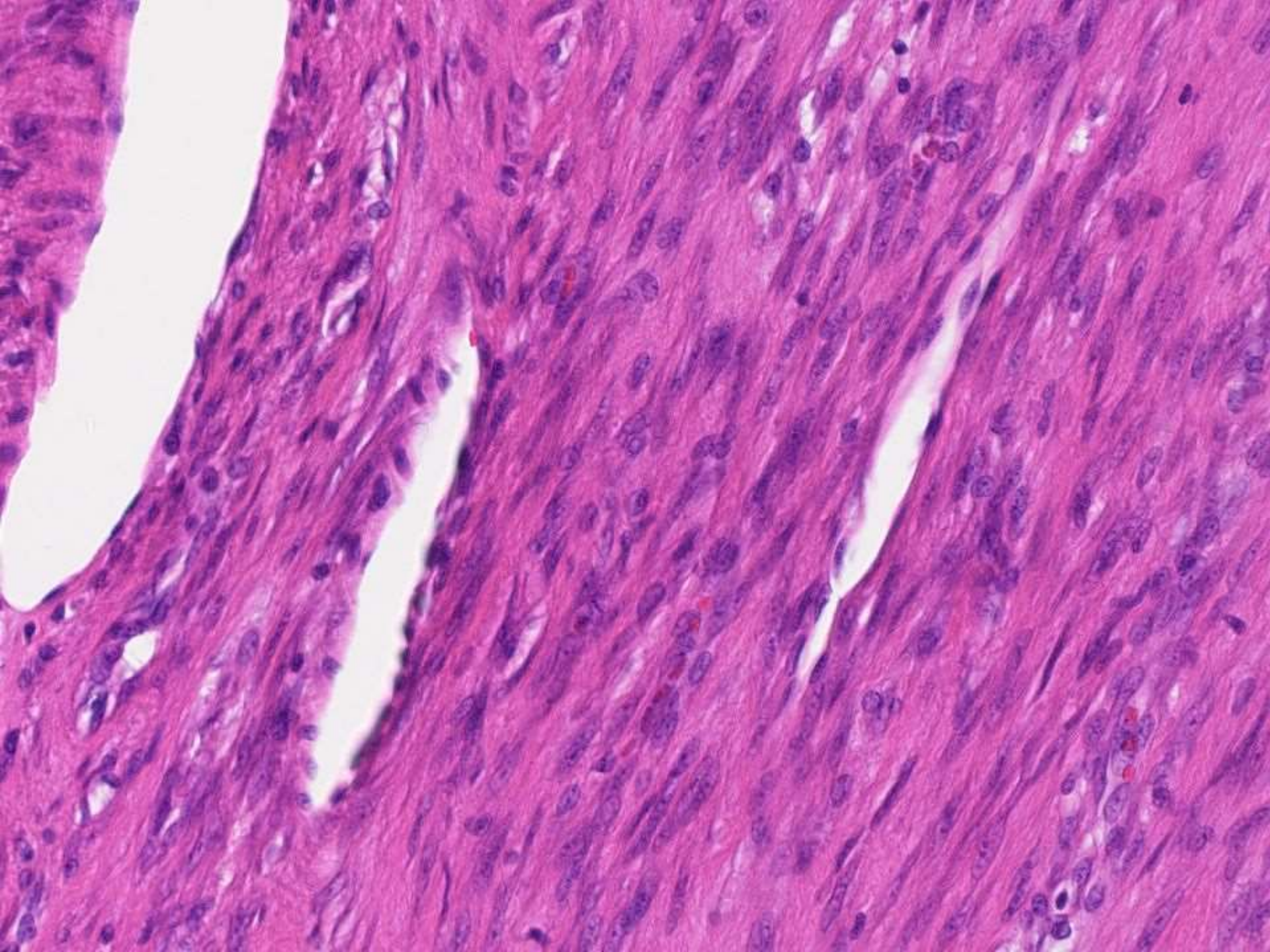




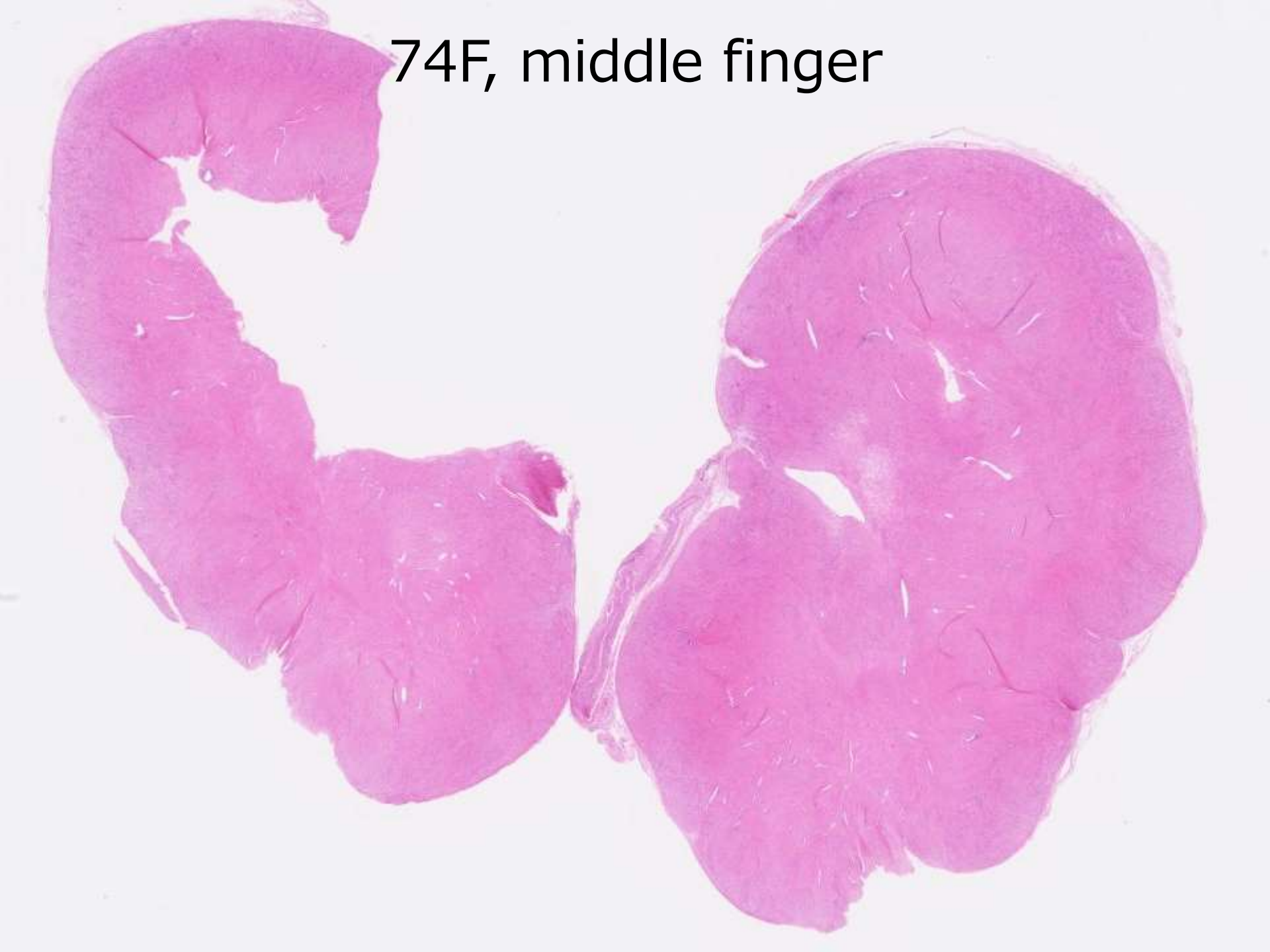

36F, forearm

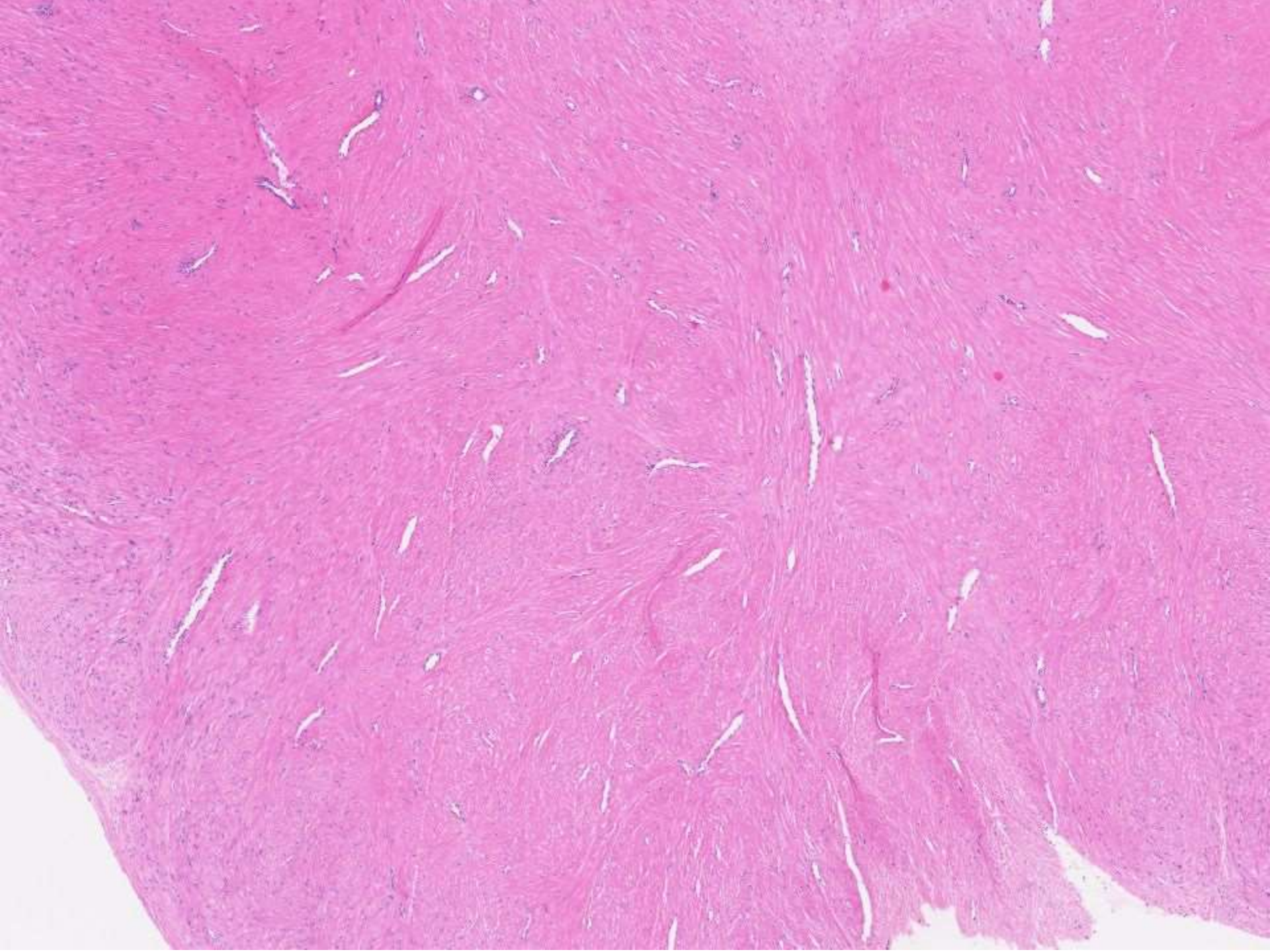


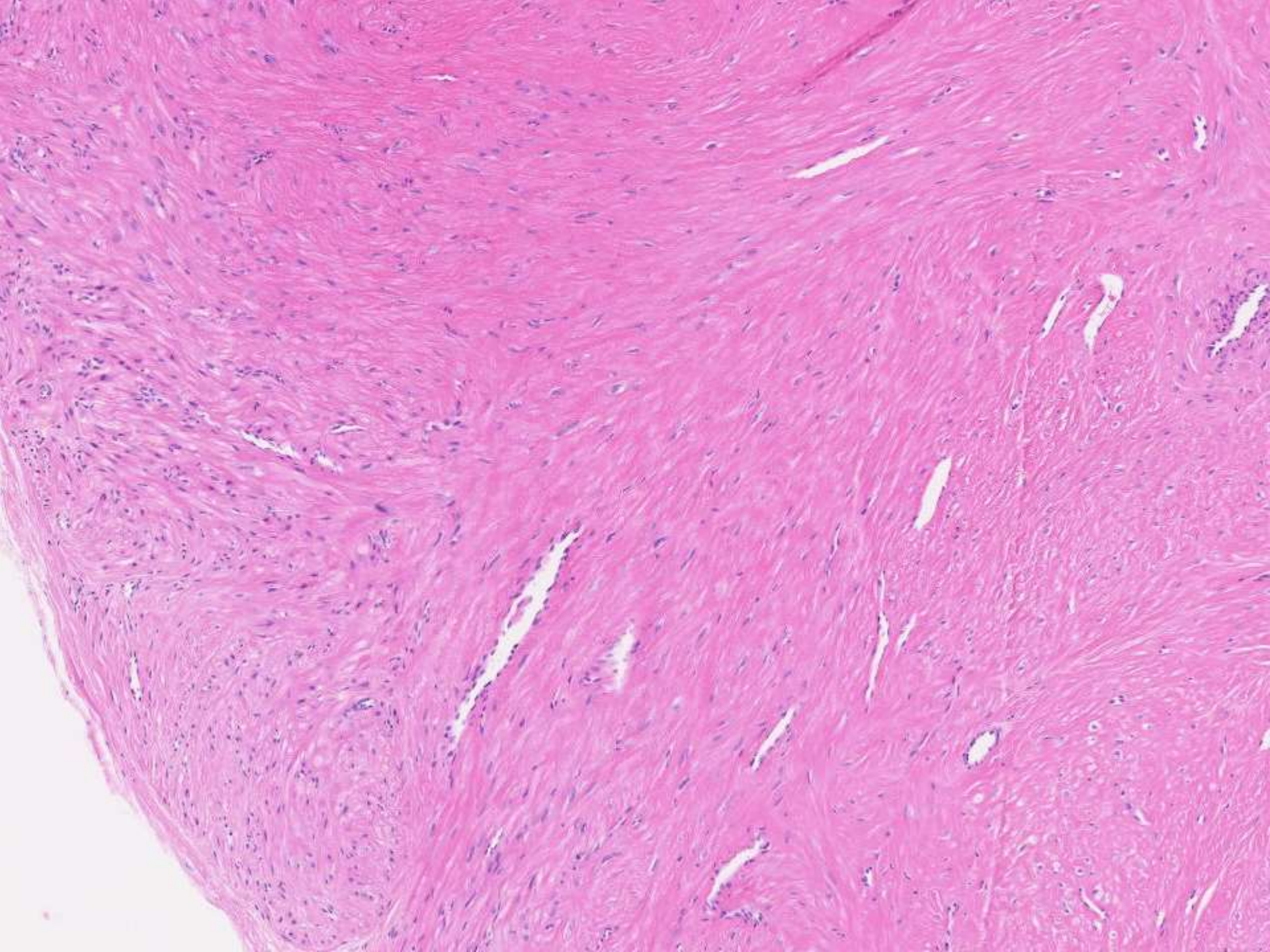


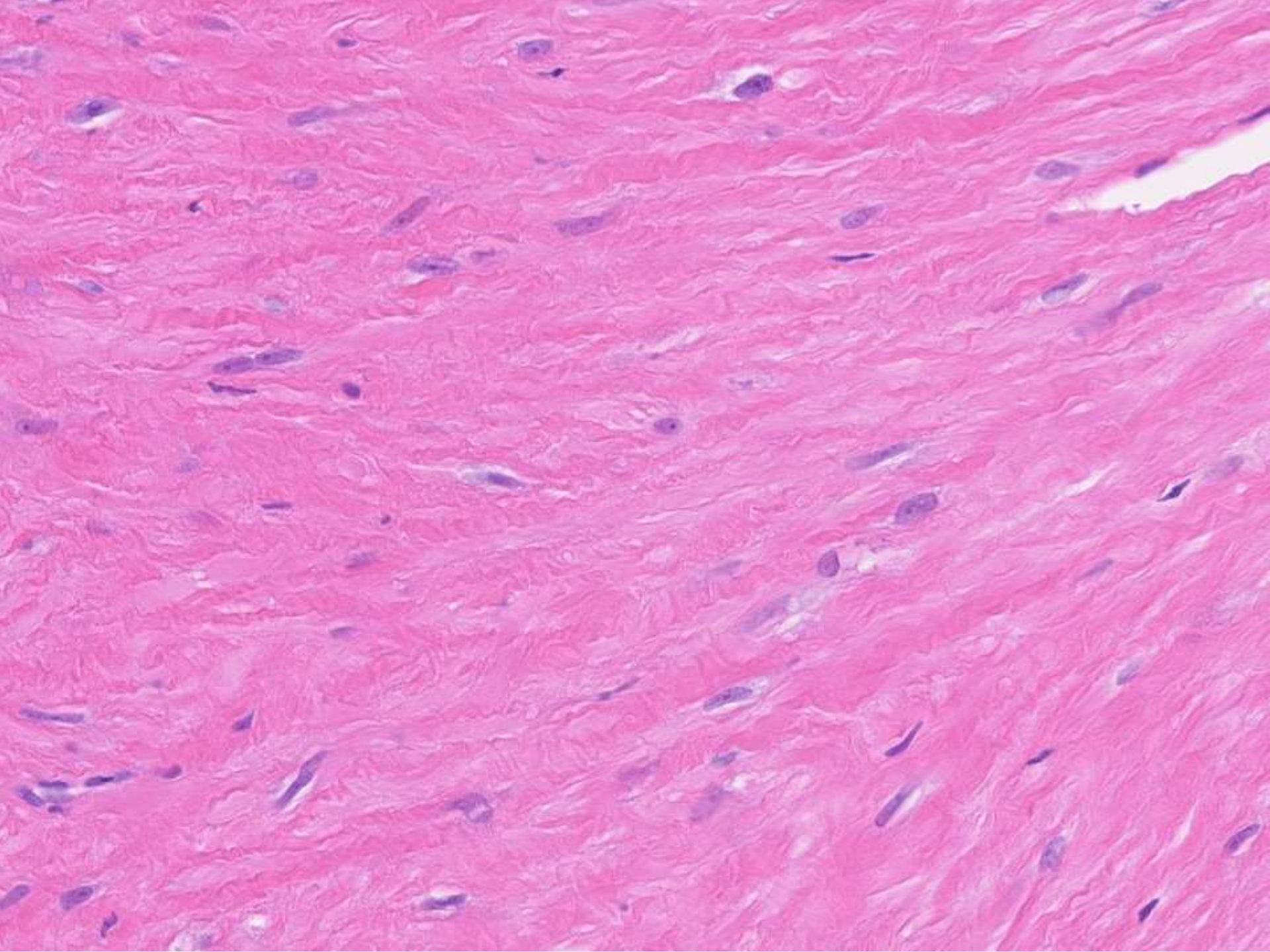


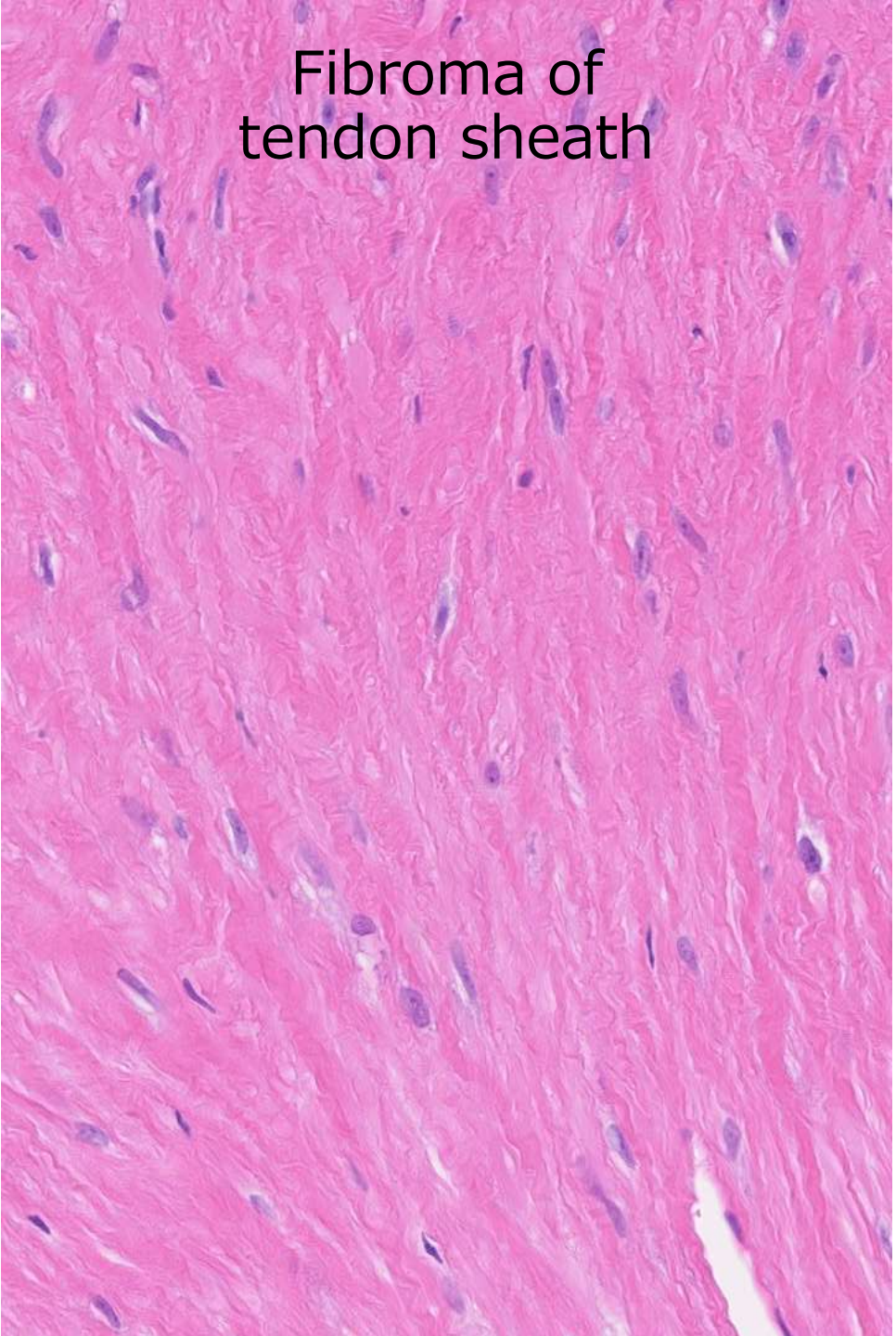
74F, middle finger

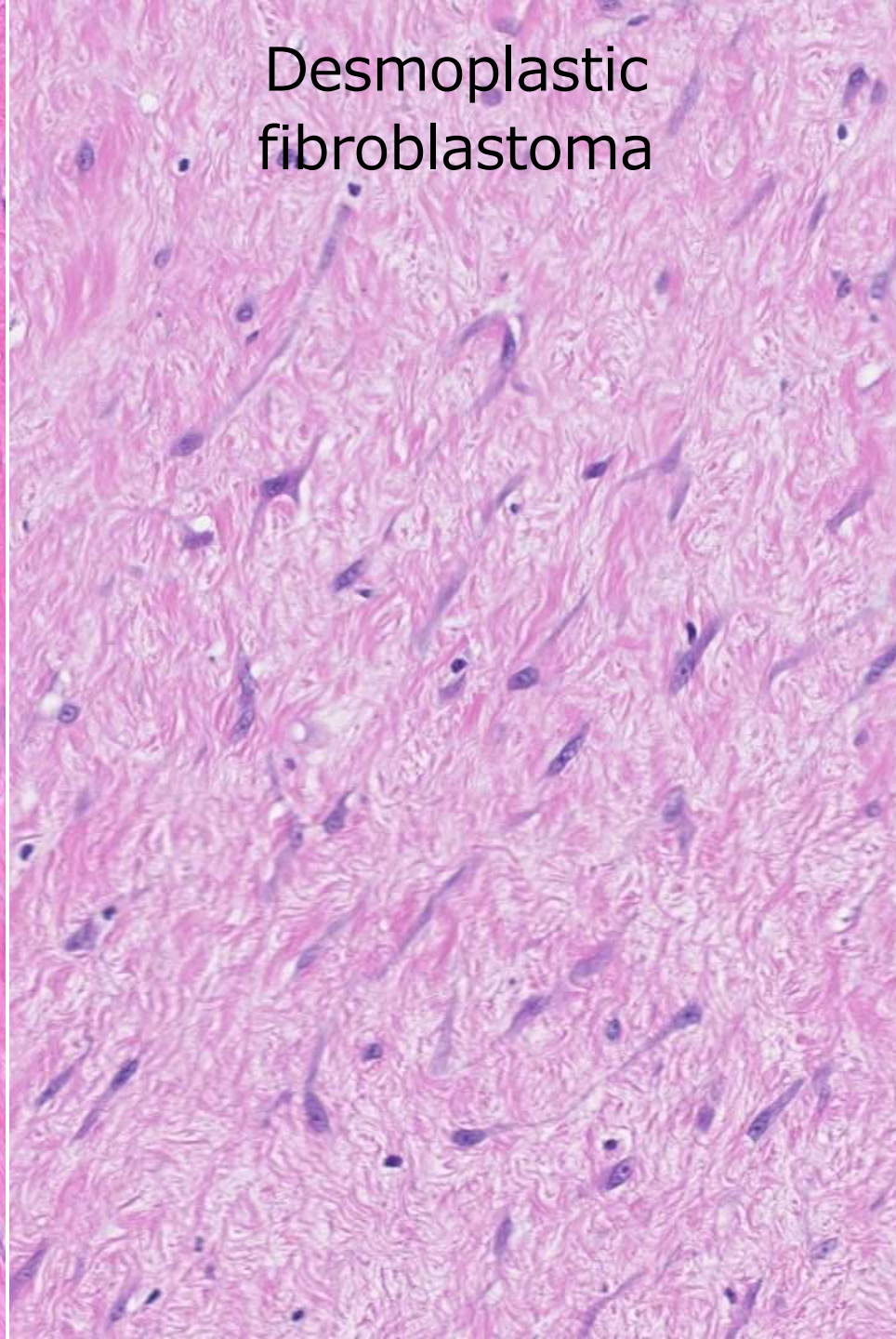









Fibroma of
tendon sheath


Desmoplastic
fibroblastoma

Histopathology

Histopathology 2016, **69**, 1012–1020. DOI: 10.1111/his.13042

FOSL1 immunohistochemistry clarifies the distinction between desmoplastic fibroblastoma and fibroma of tendon sheath

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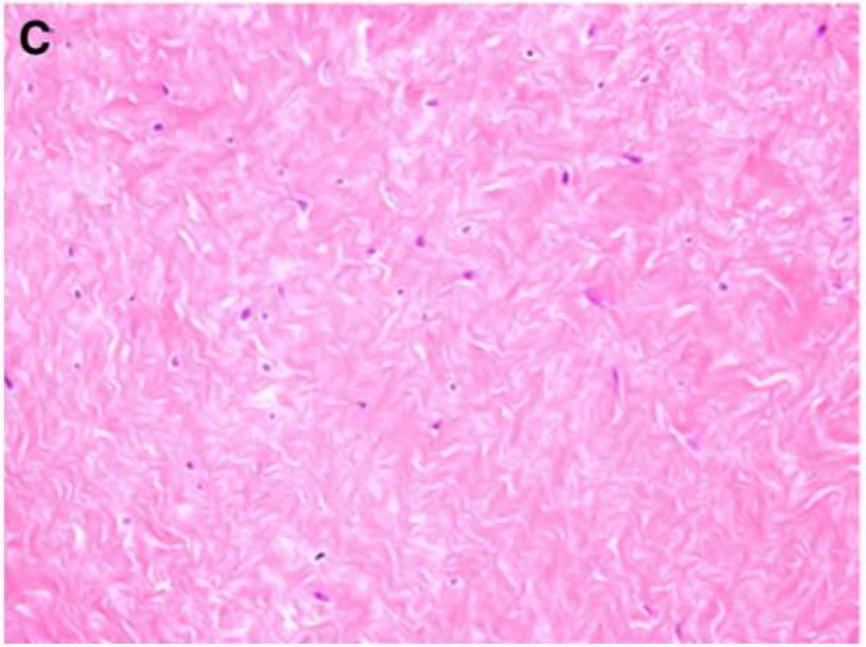
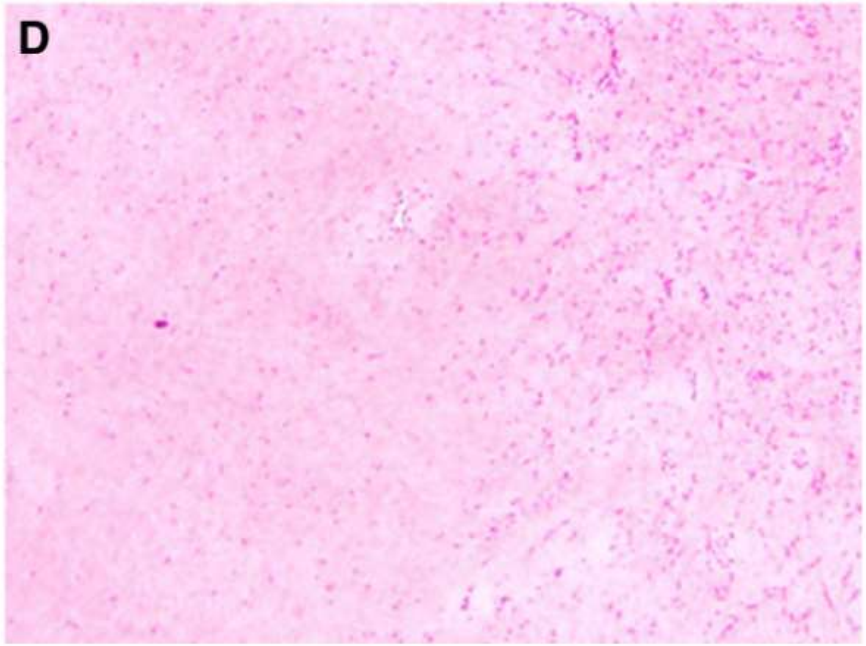
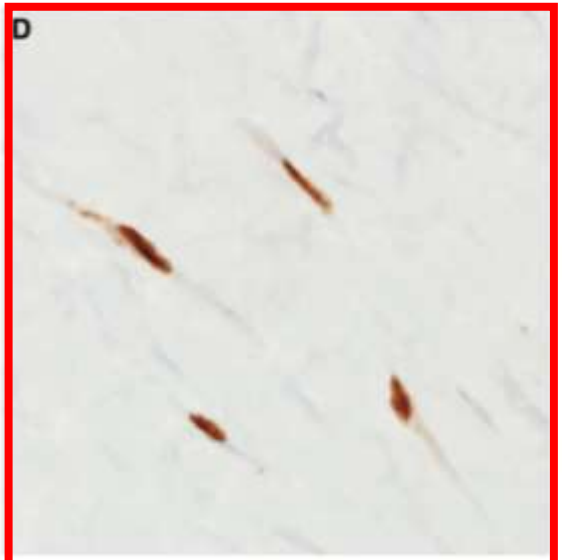


Desmoplastic fibroblastoma

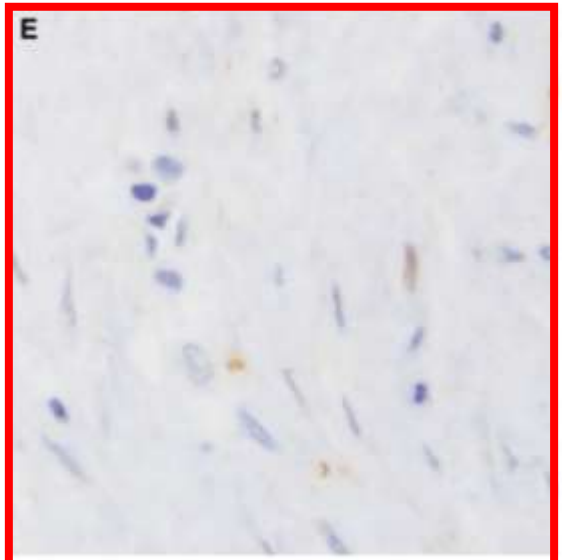


Fibroma of tendon sheath

IHC for FOSL1

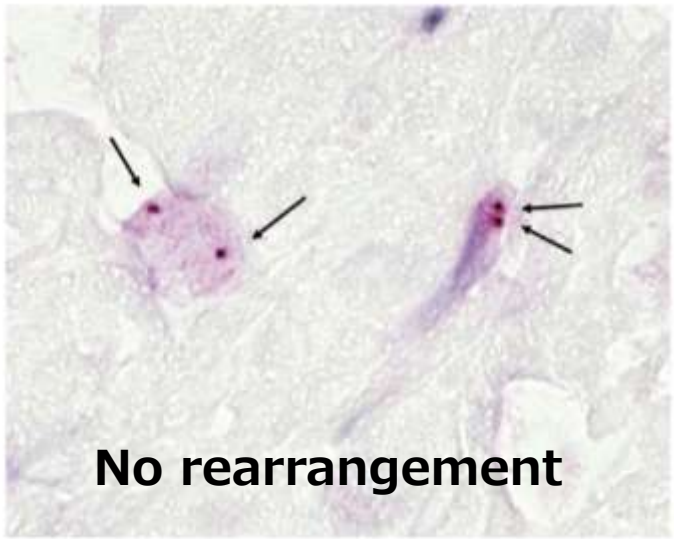


Desmoplastic fibroblastoma



Fibroma of tendon sheath

CISH of FOSL1



Desmoplastic fibroblastoma

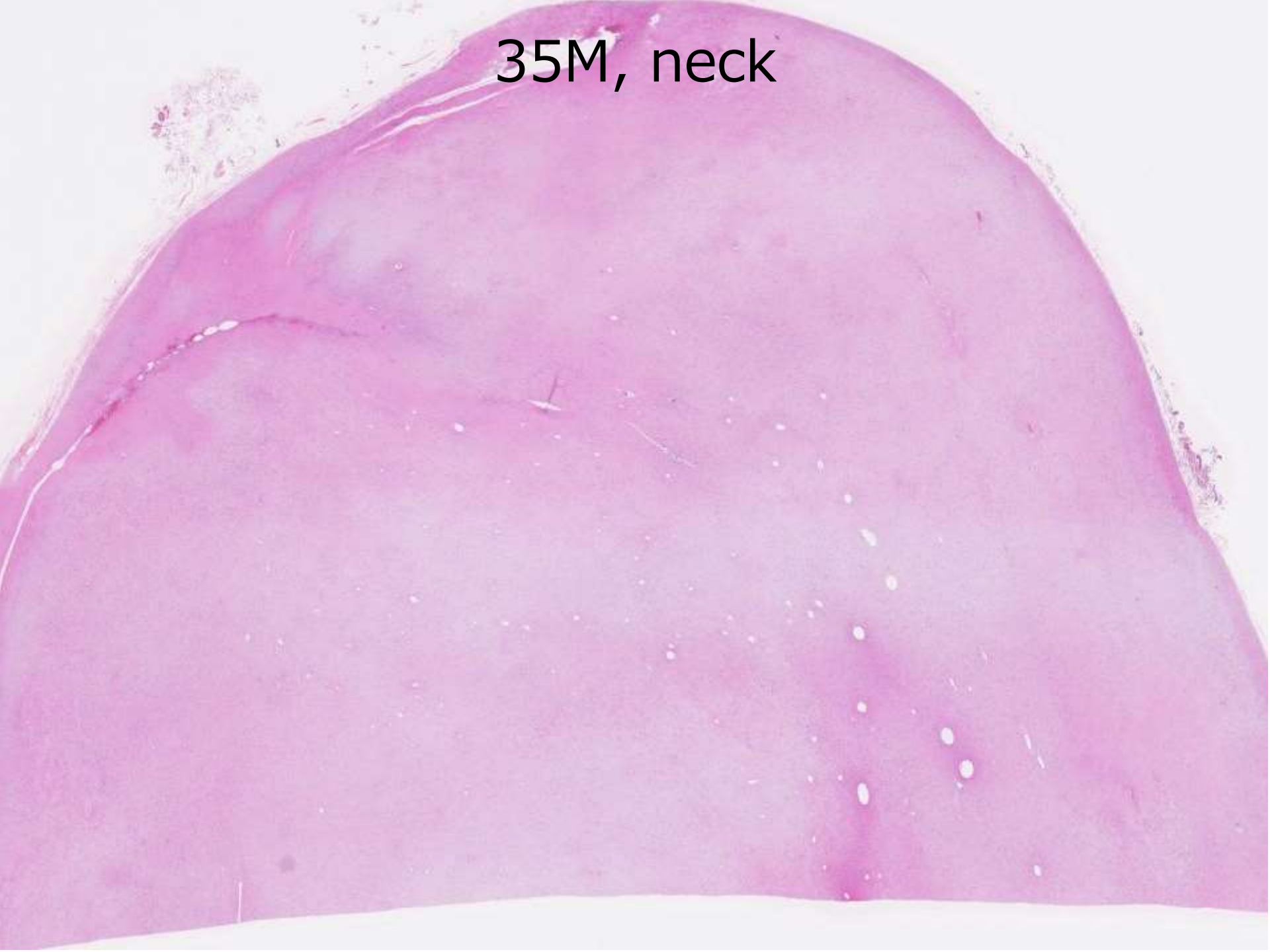
Table 3. Summary of FOSL1 immunohistochemical nuclear staining

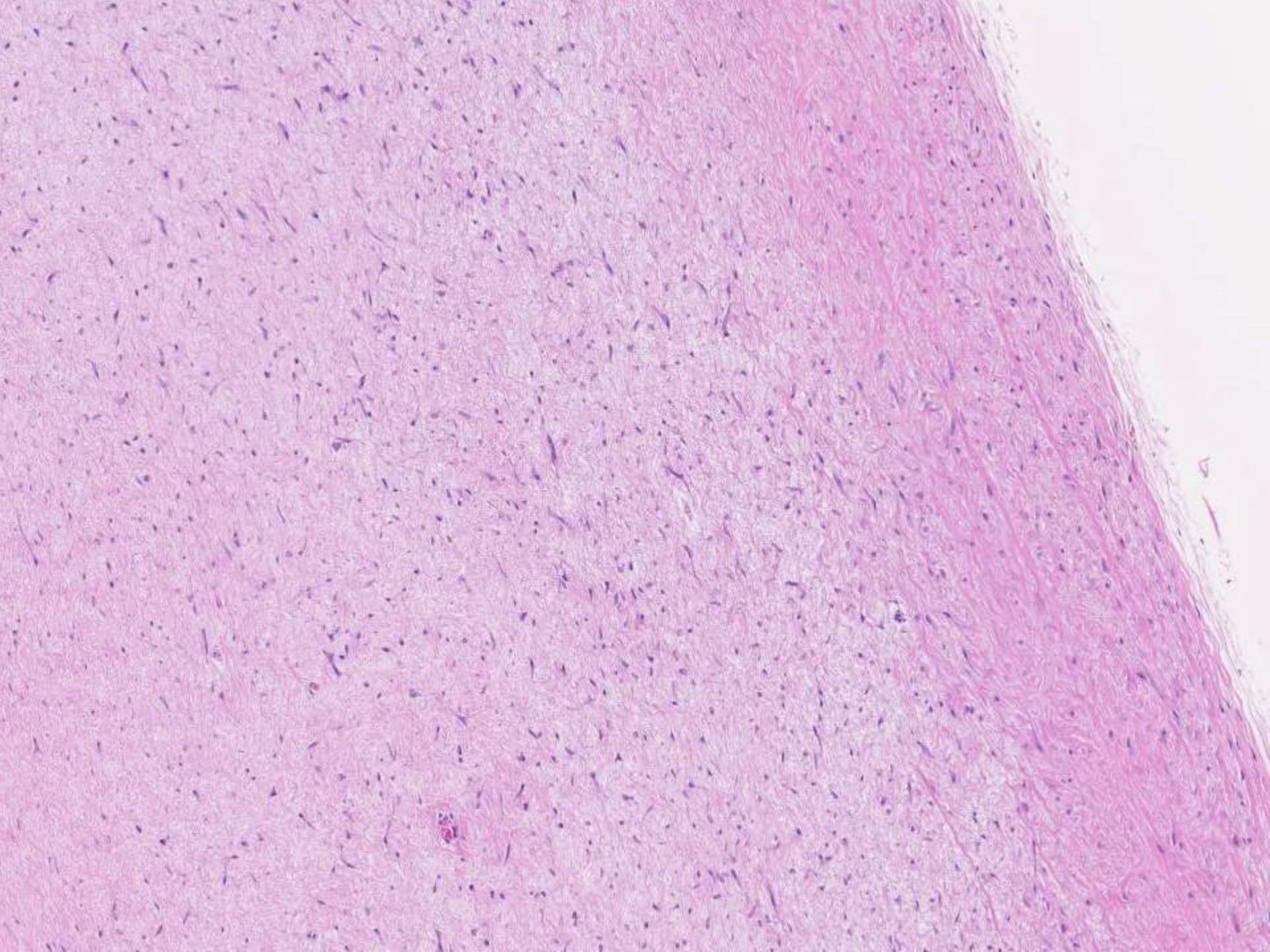
Tumour type	*n*	4+ (%)	3+ (%)	2+ (%)	1+ (%)	0 (%)
DFB	25	25 (100)	0	0	0	0
FTS	16	0	0	2 (13)	6 (37)	8 (50)
Other spindle cell tumours	42					

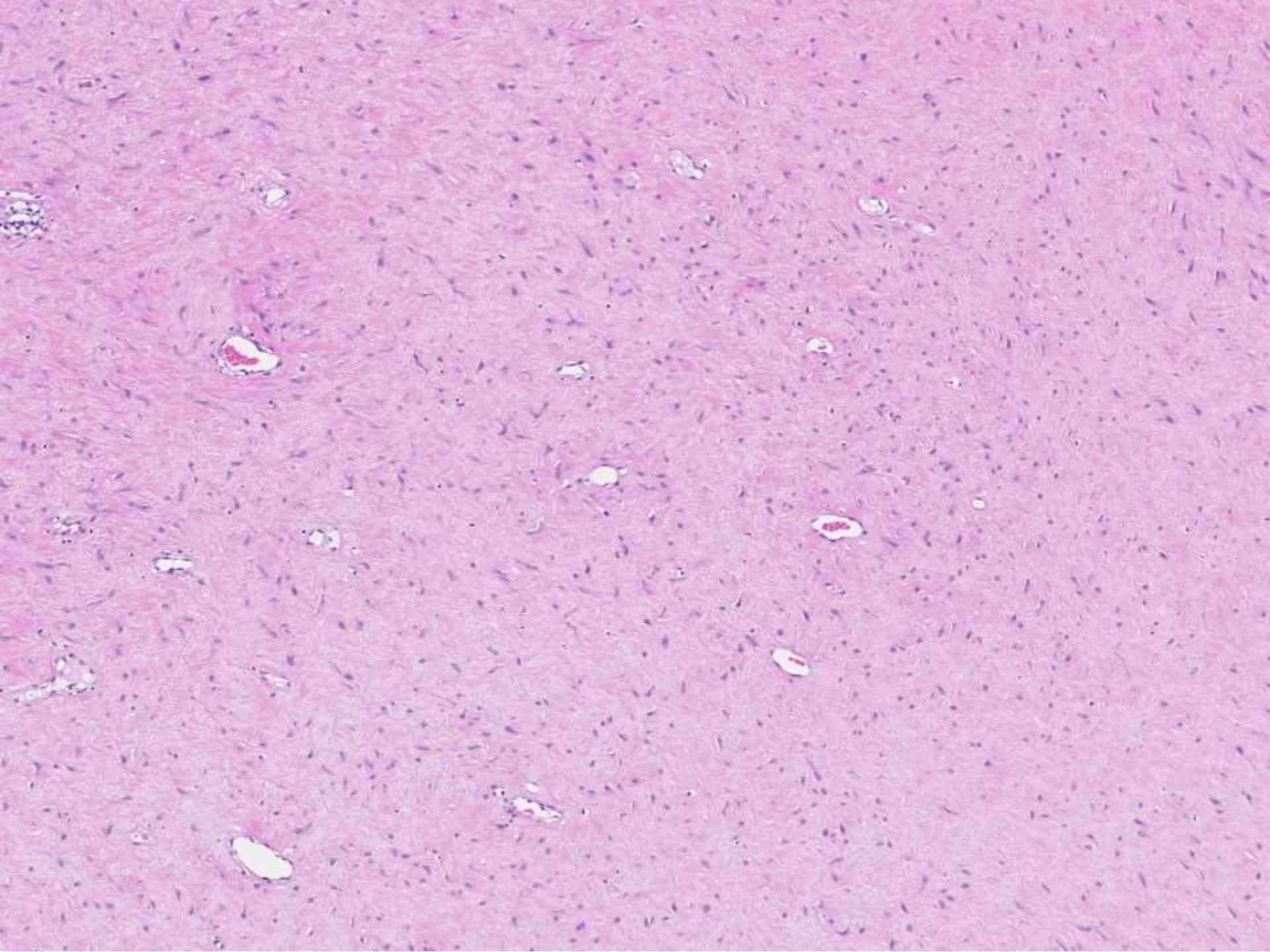
Kato I, et al. FOSL1 immunohistochemistry clarifies the distinction between desmoplastic fibroblastoma and fibroma of tendon sheath. Histopathology 69: 1012-1020, 2016.

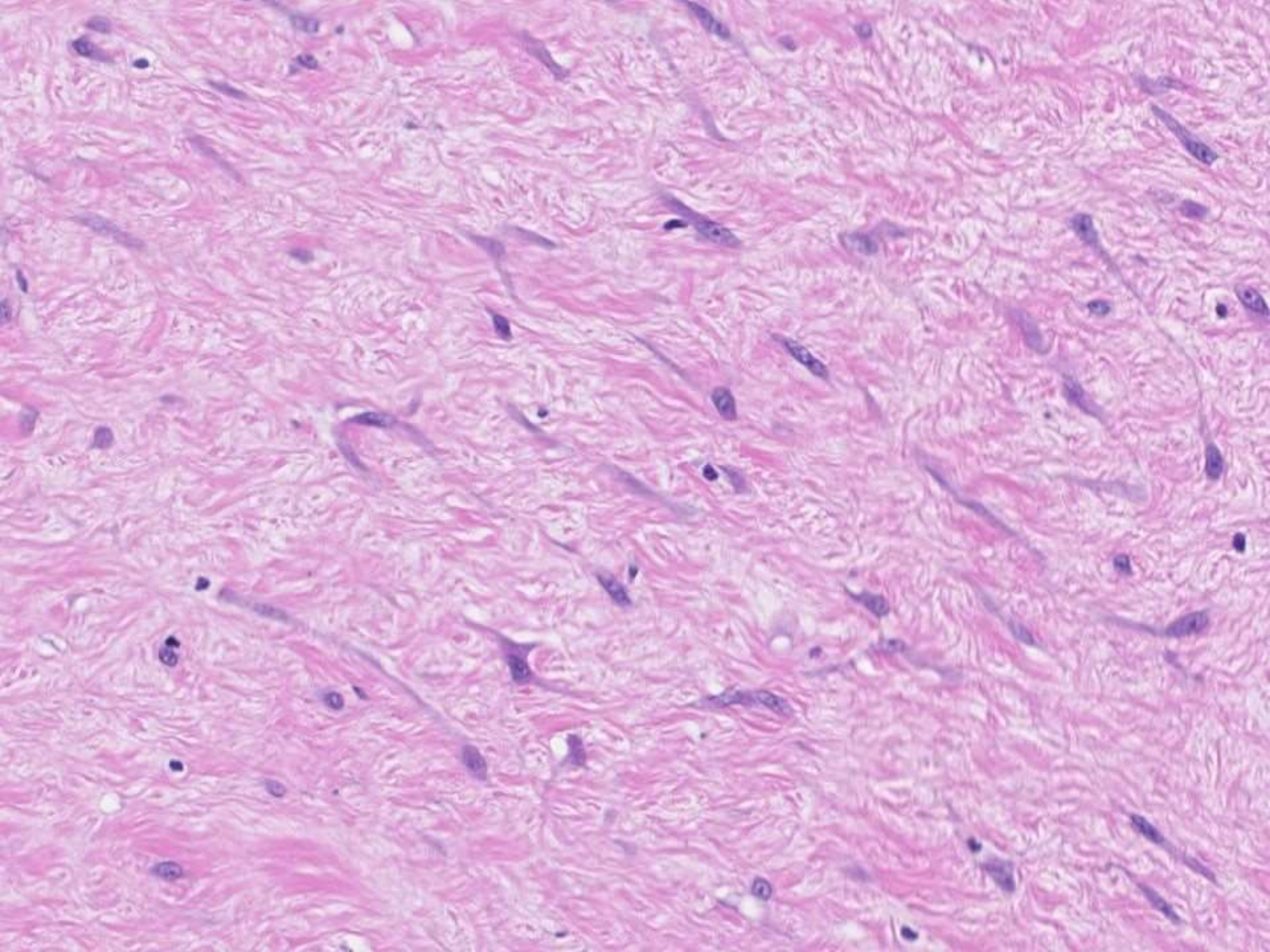
Desmoplastic fibroblastoma

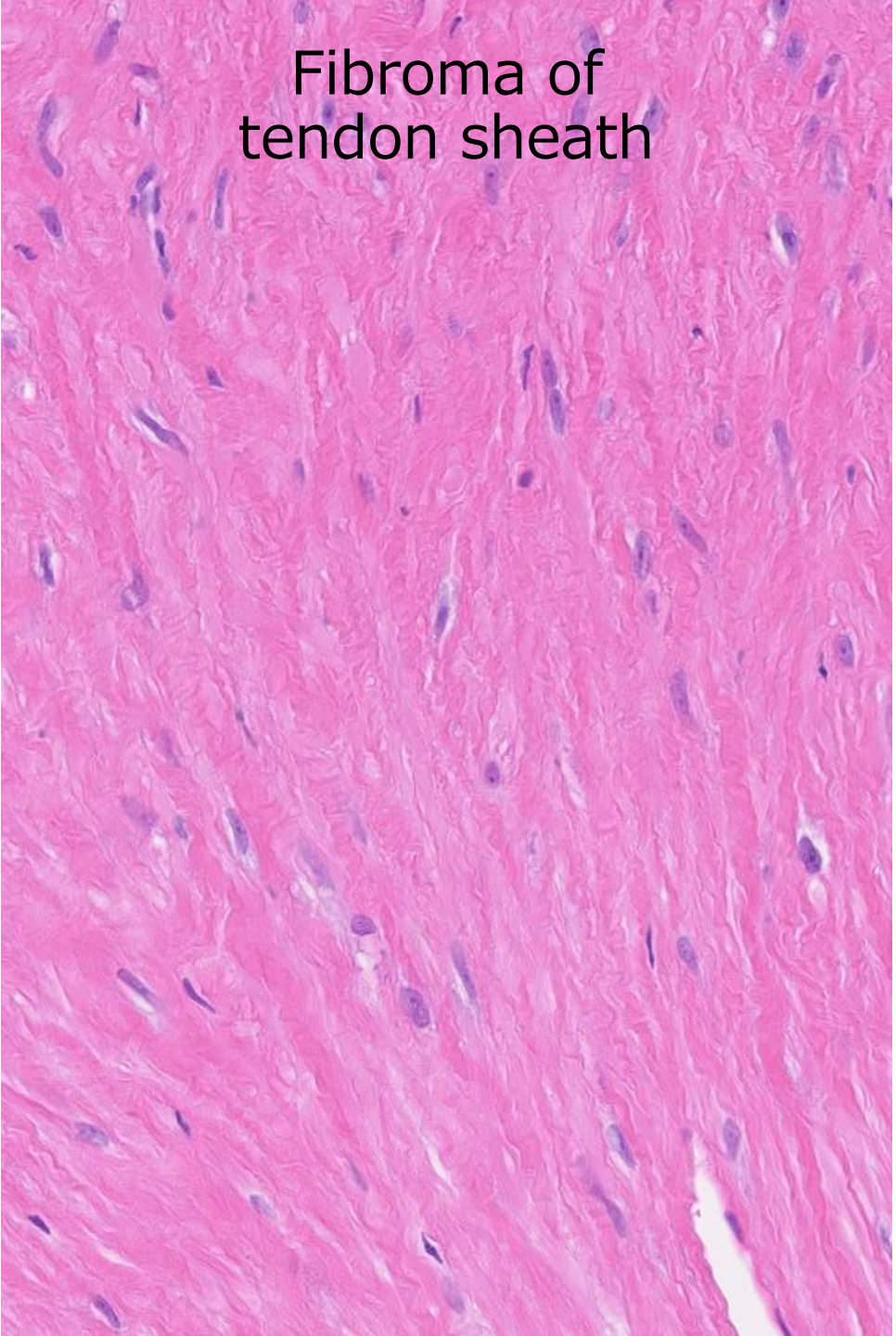
- 中高年に多い
- 上下肢、体幹
- 皮下＞筋内
- ～20 cm （多くは～5 cm)
- t(2;11)(q31;q12)
- 免役染色でFOSL1(+)だが、FOSL1(11q13)の遺伝子再構成はない
- 細胞密度が低い

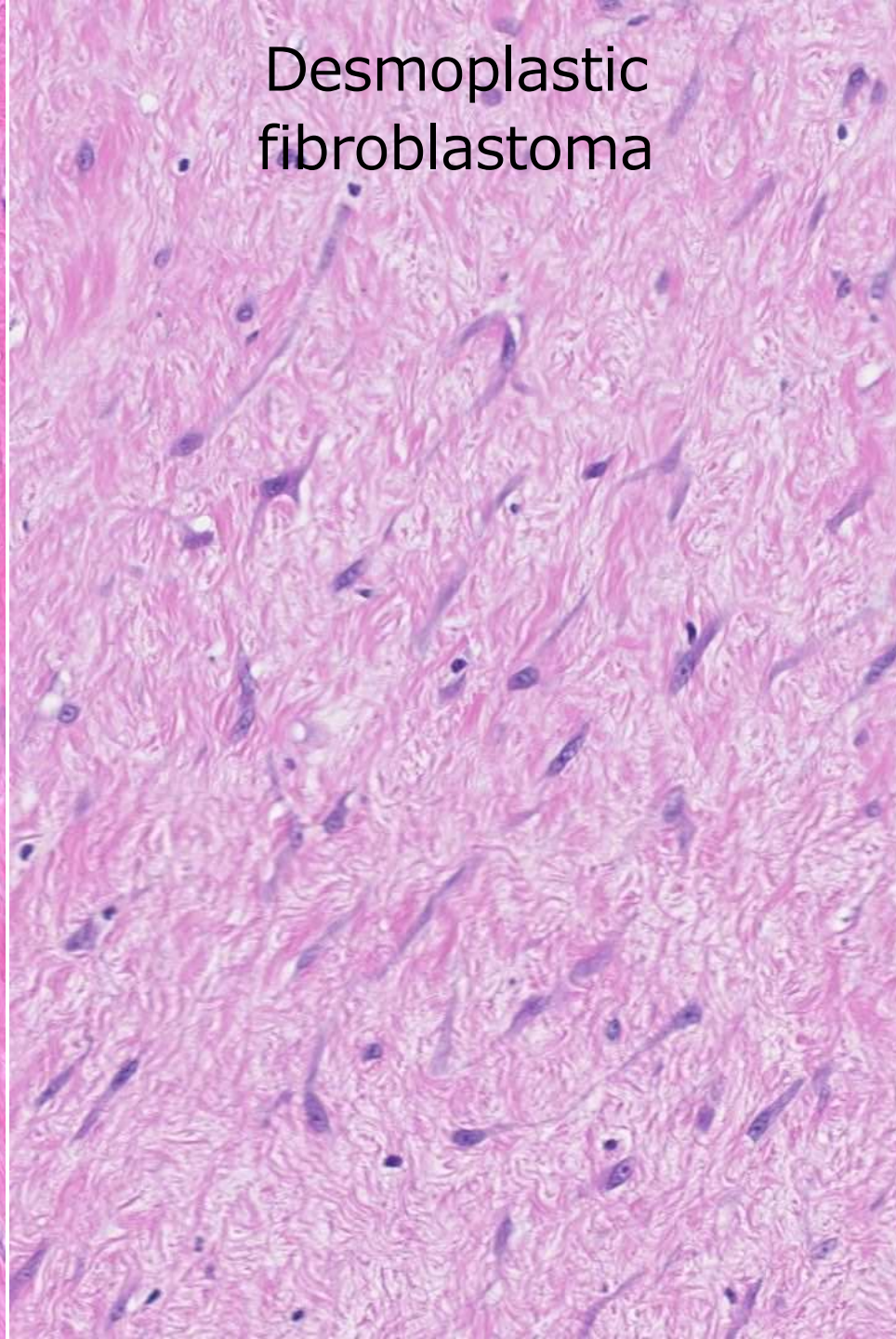

35M, neck

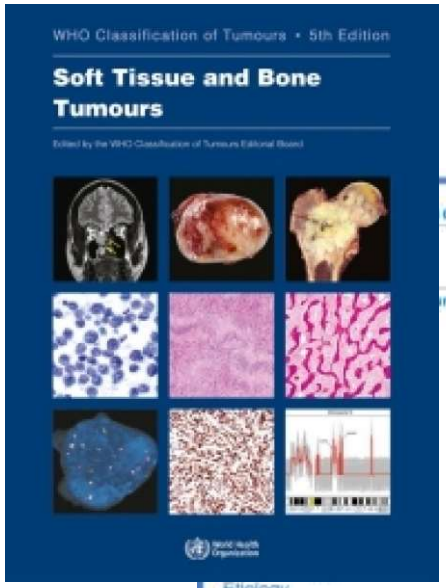







Fibroma of
tendon sheath


Desmoplastic
fibroblastoma



Macroscopic appearance:-

Fibroma of tendon sheath has a lobular fibrous appearance, reminiscent of a localized tenosynovial giant cell tumour, except for the pigment, which is absent in fibroma of tendon sheath.

Histopathology:-

The lesion is well circumscribed and contains bland spindle cells in a collagenous background. The cellularity is usually low but can be variable and is often higher at the tumour edge. There are characteristic slit-like thin-walled vessels. Degenerative features such as myxoid/cystic changes, chondroid or osseous metaplasia, and bizarre pleomorphic cells can be seen. The morphological features of the cellular subtype are identical to those of nodular fasciitis.

Macroscopic appearance:-

Desmoplastic fibroblastomas are usually relatively small, often measuring 1–4 cm in greatest dimension, but examples > 10 cm and as large as 20 cm have occurred. Grossly, the lesions appear well circumscribed and form oval, fusiform, or discoid masses. Some examples have an externally lobulated, cobblestone-like surface. The tumours have a firm, cartilage-like consistency, and on cut section they have a homogeneous pearl-grey colour.

Histopathology:-

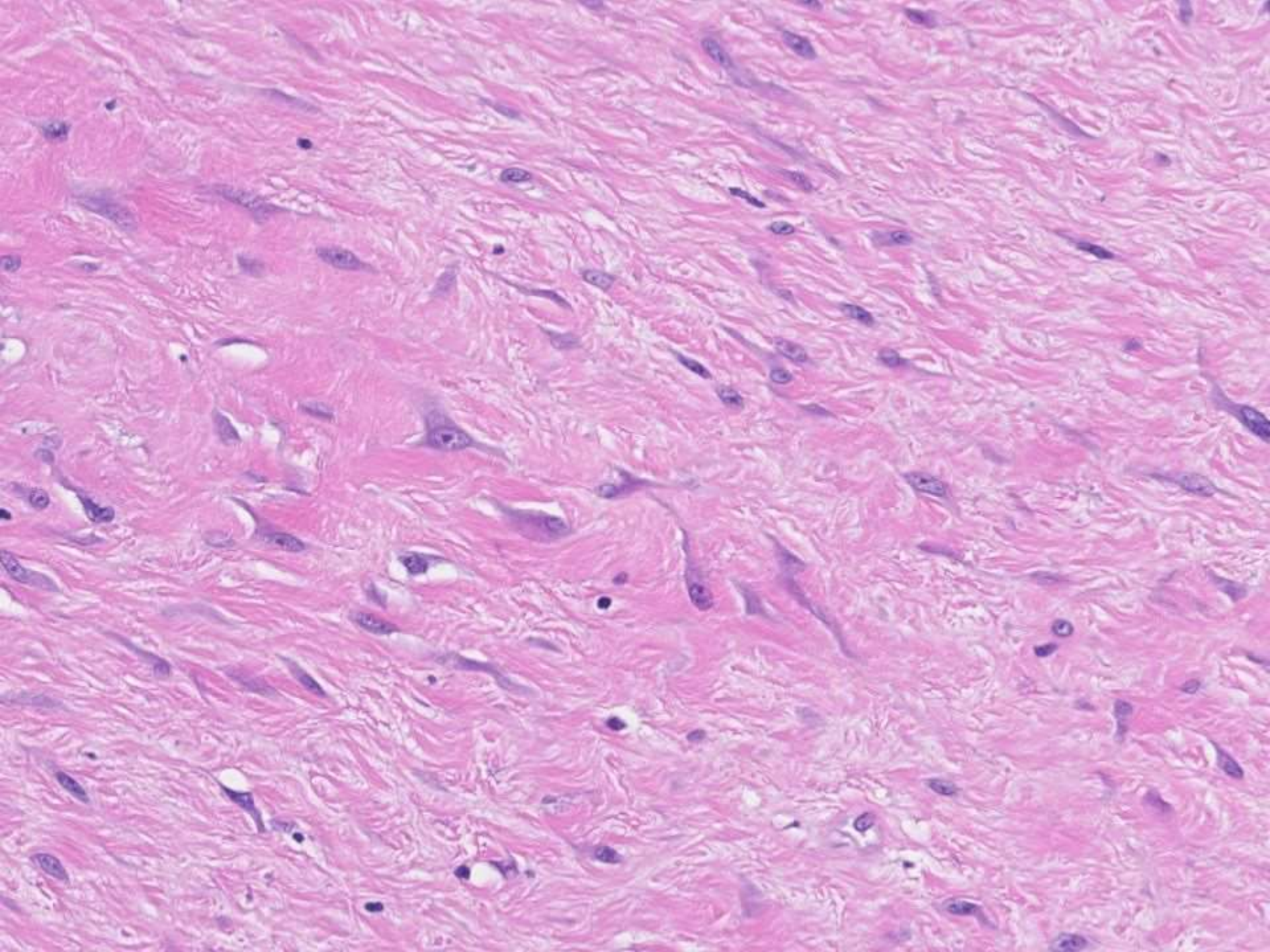
Although often well demarcated grossly, most tumours microscopically infiltrate into subcutaneous fat, and approximately 25% extend into skeletal muscle { 7661281 ; 8832562 ; 9670823 }. Rare examples are purely intramuscular. The lesions have abundant collagenous or myxocollagenous matrix with low vascularity. Cellularity ranges from low to moderate, and the neoplastic cells tend to be uniformly distributed within the extracellular matrix. The lesional cells are stellate-shaped, bipolar, and spindled, and they have uniform, bland nuclei with distinct small nucleoli. Mitotic figures are uncommon. Rare examples have focal intravascular growth. The tumour cells may be focally positive for SMA.

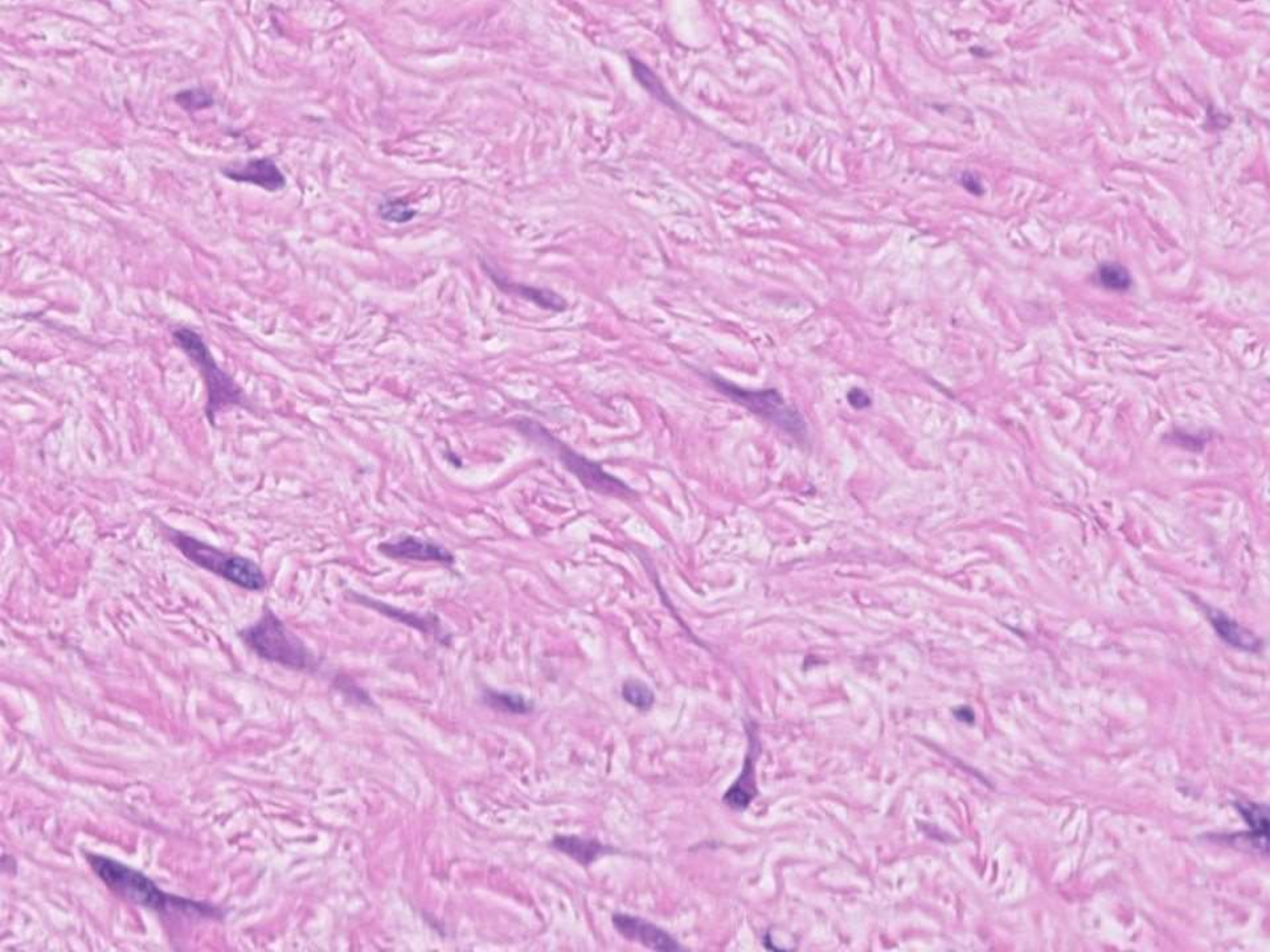
Cytology:-

Not clinically relevant

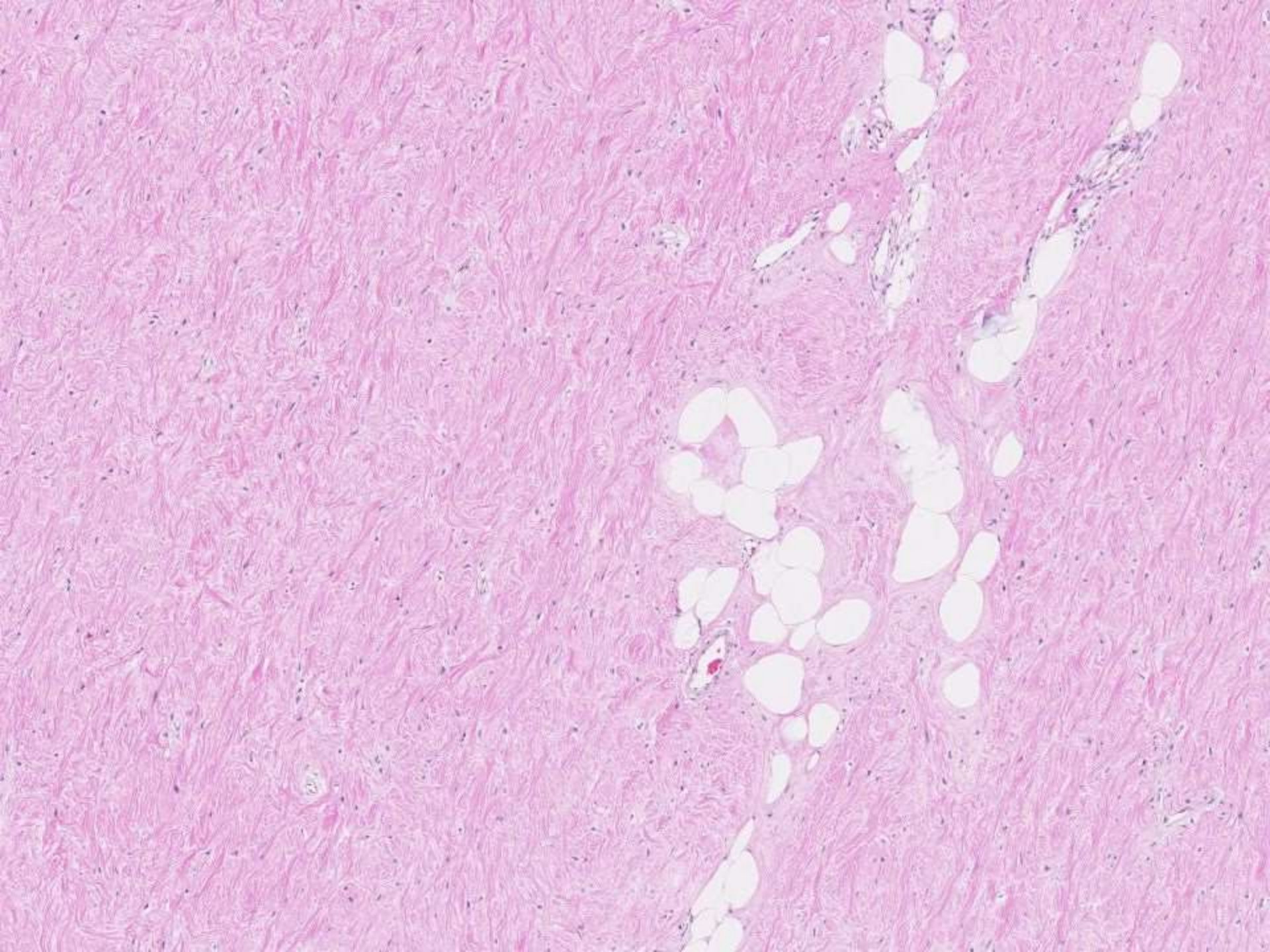
Diagnostic molecular pathology:-

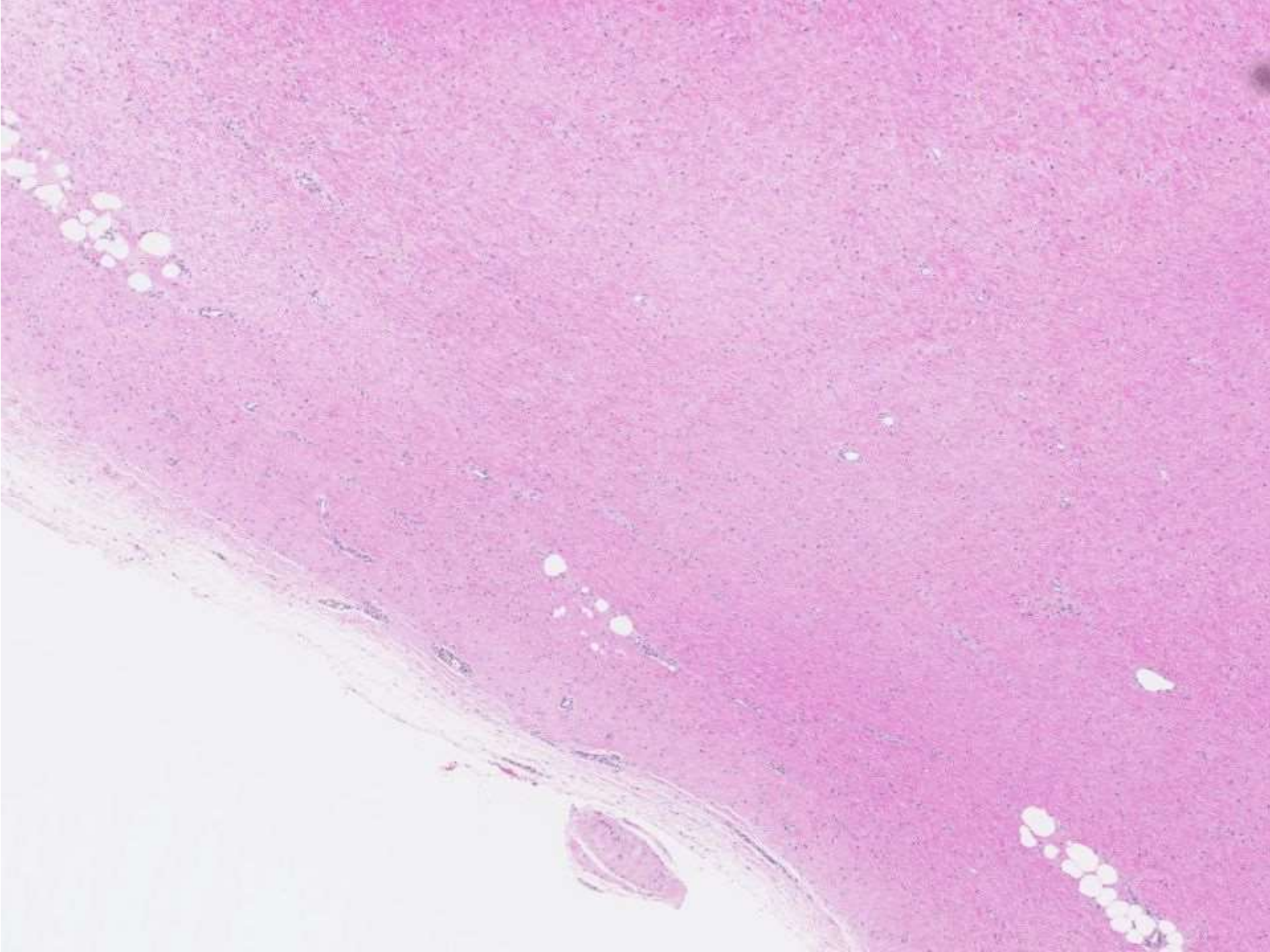
Not clinically relevant

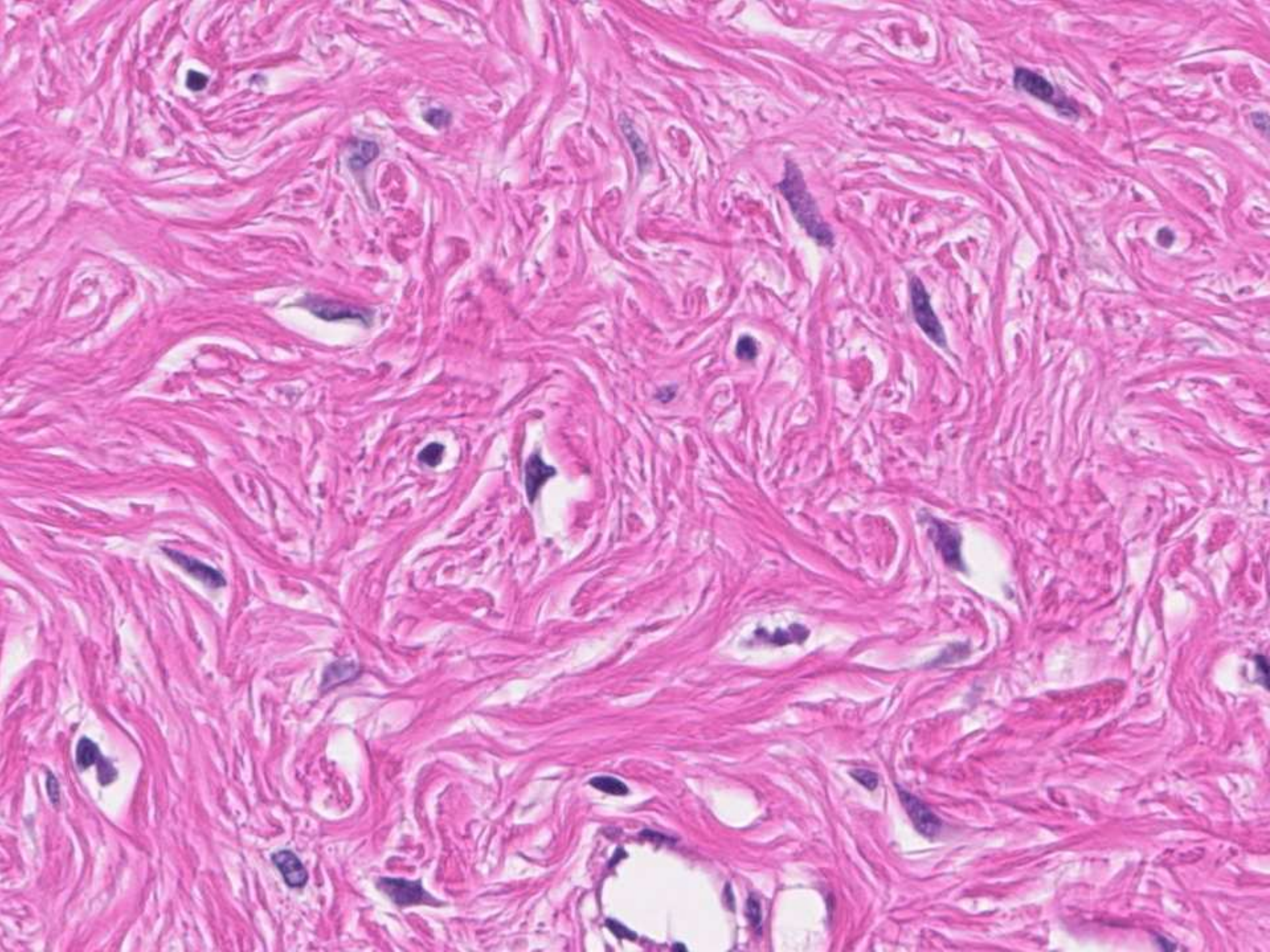


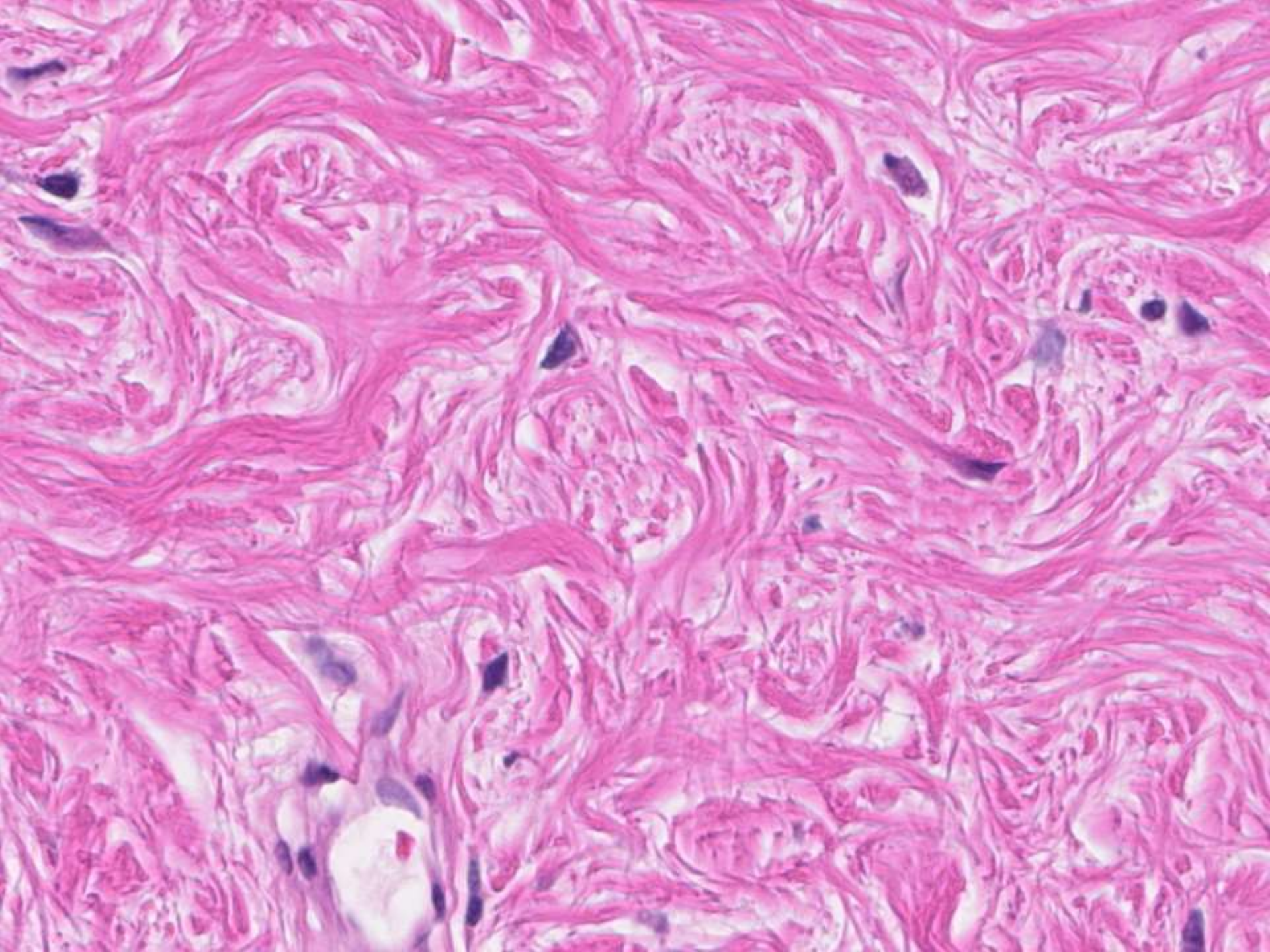



46F, thigh

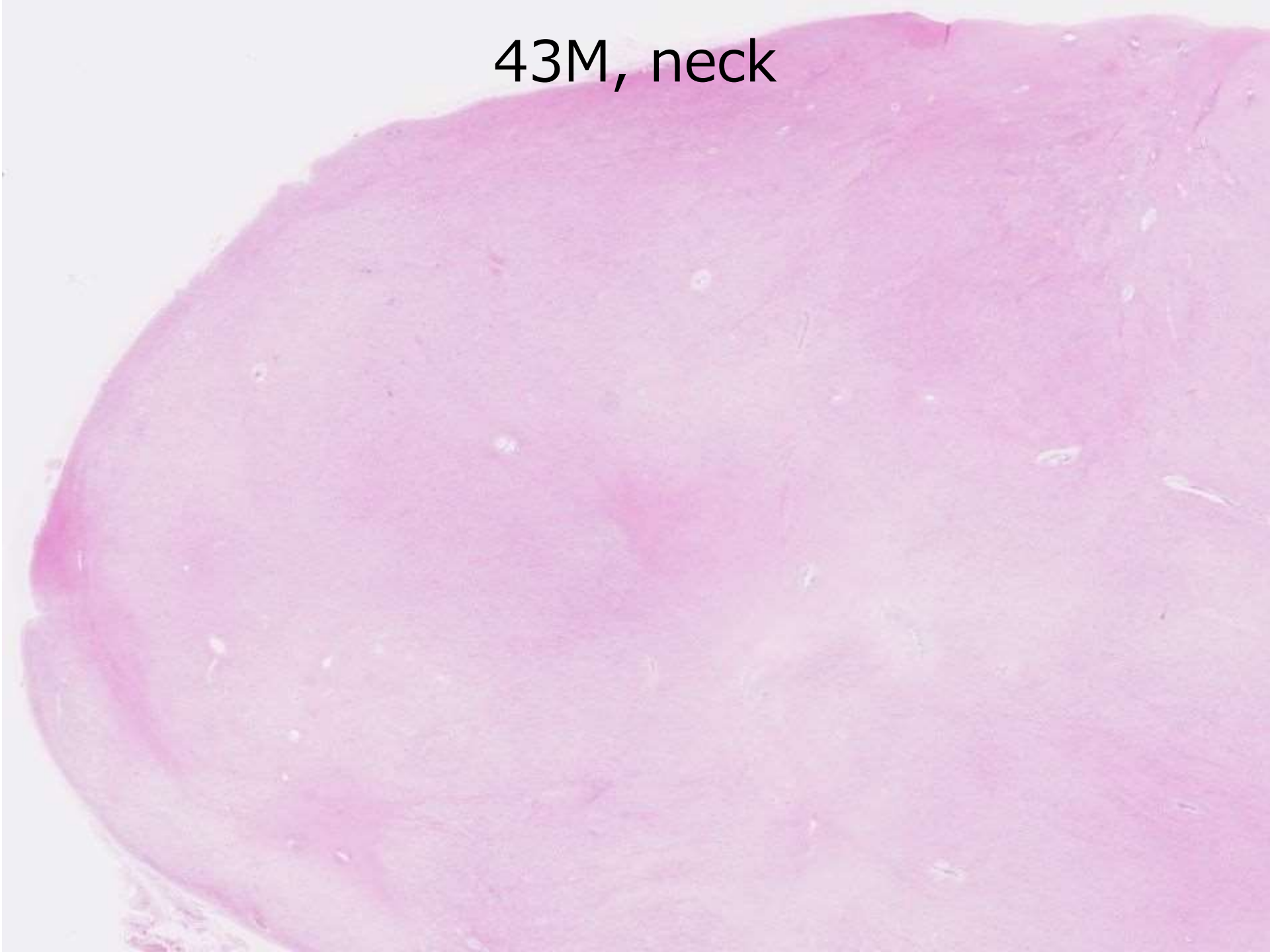


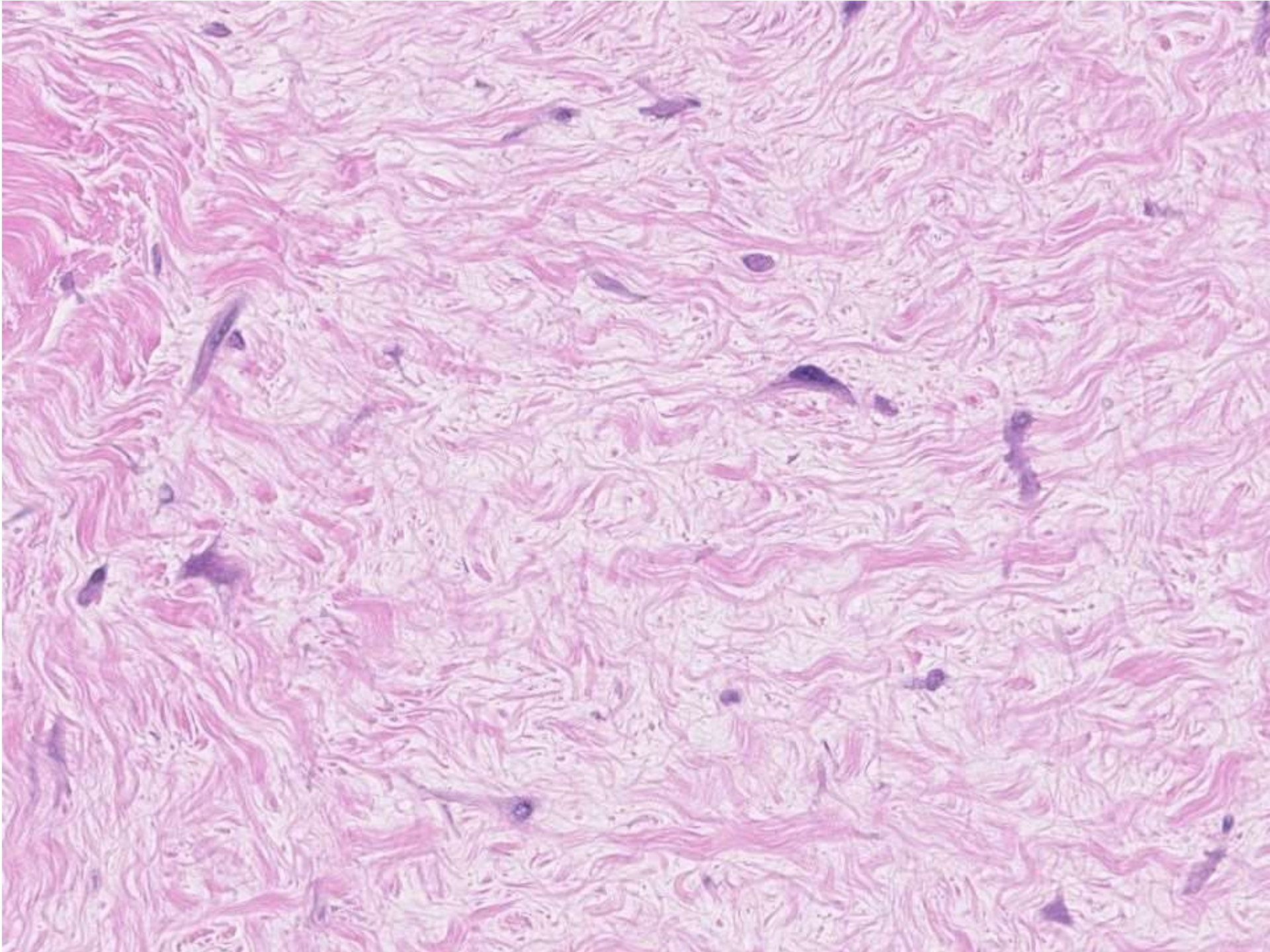


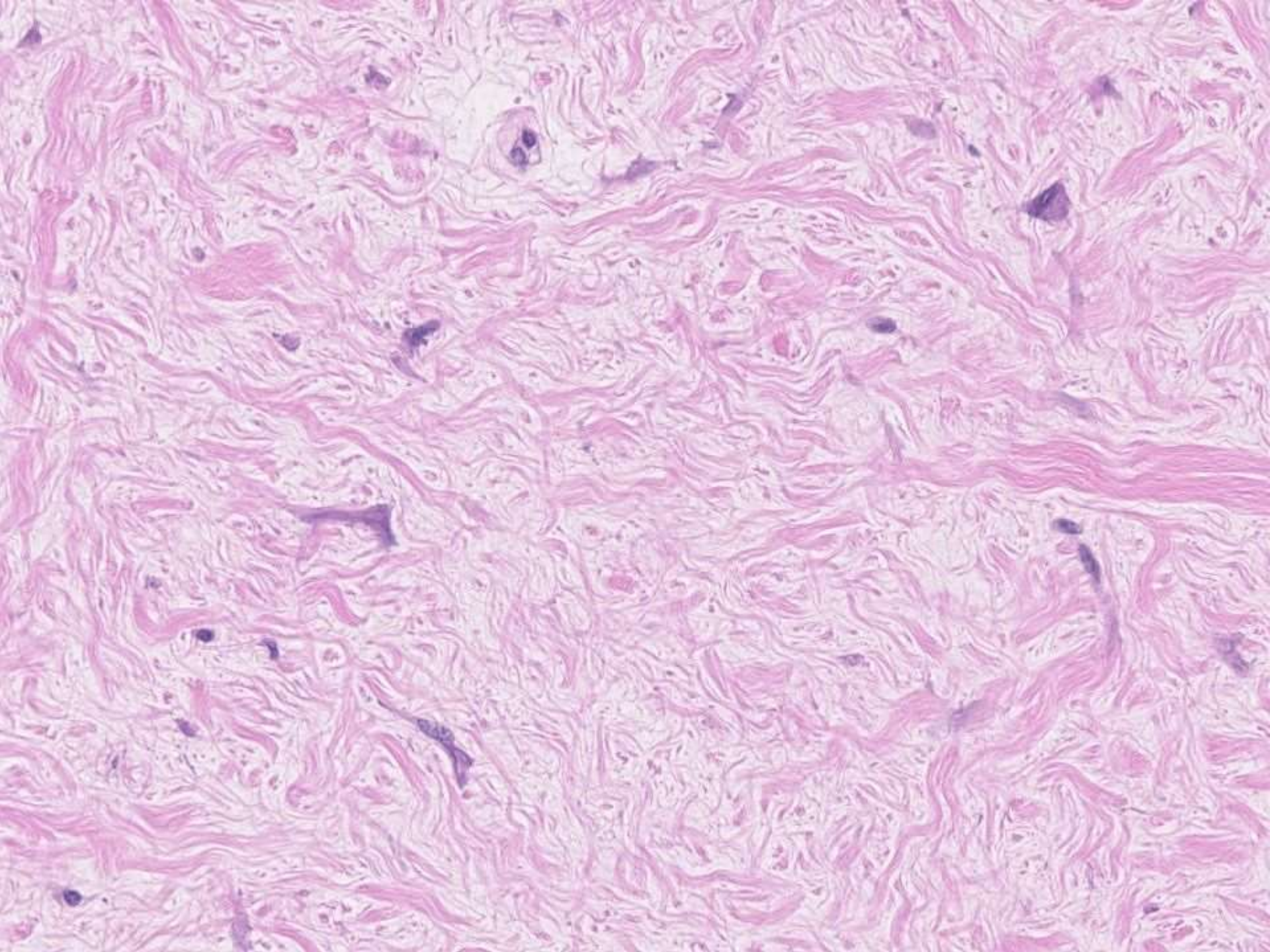


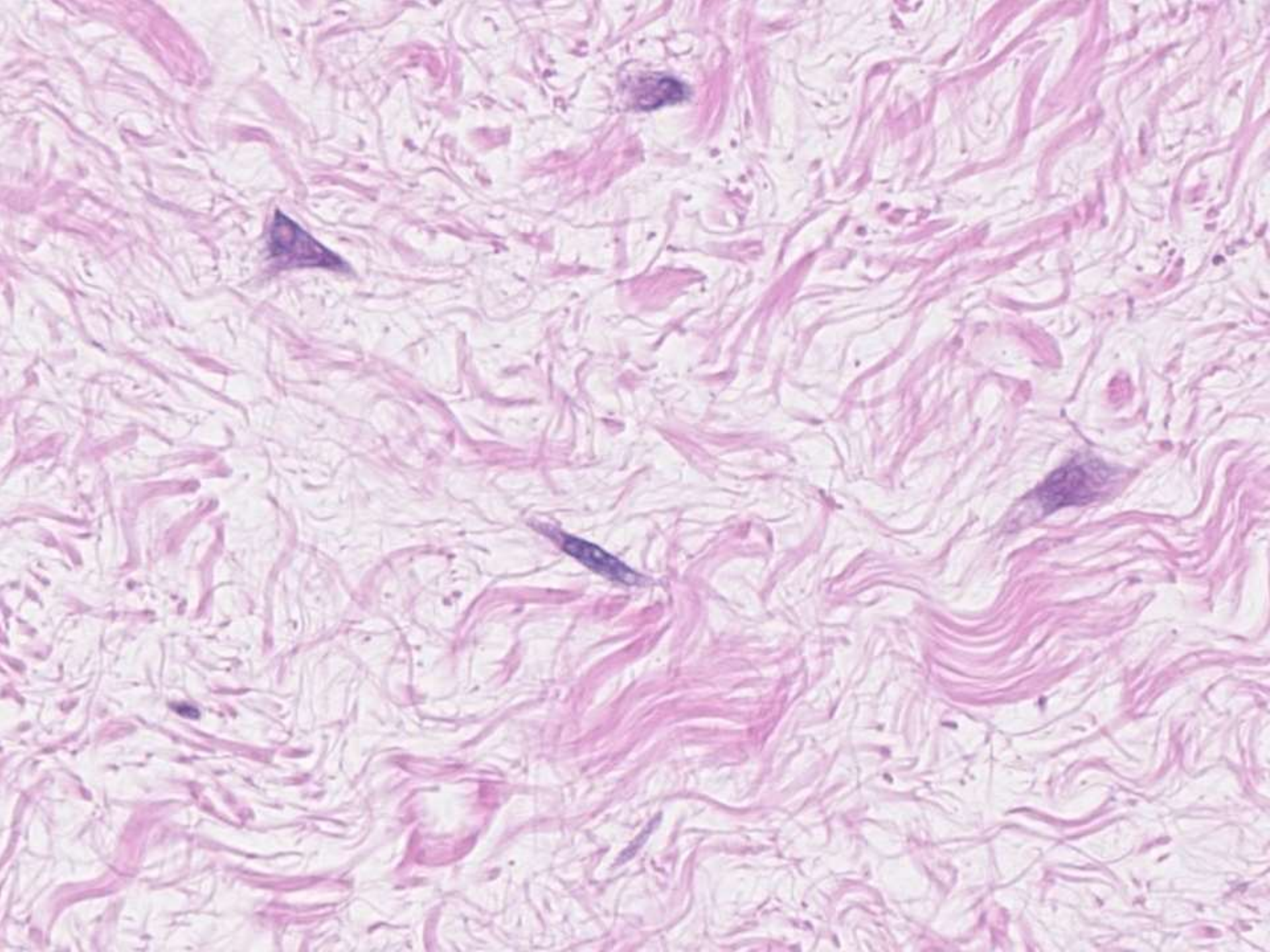


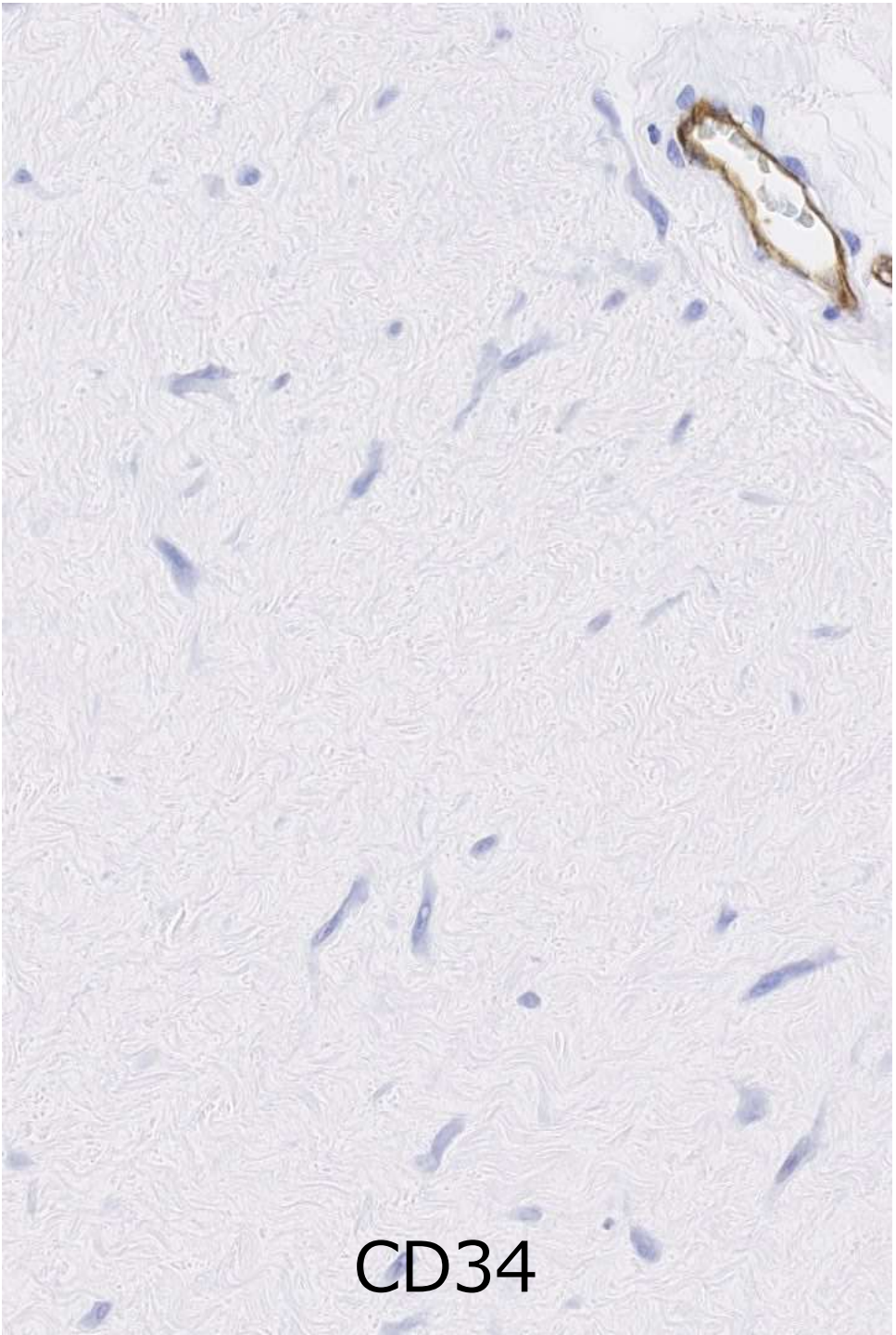
43M, neck

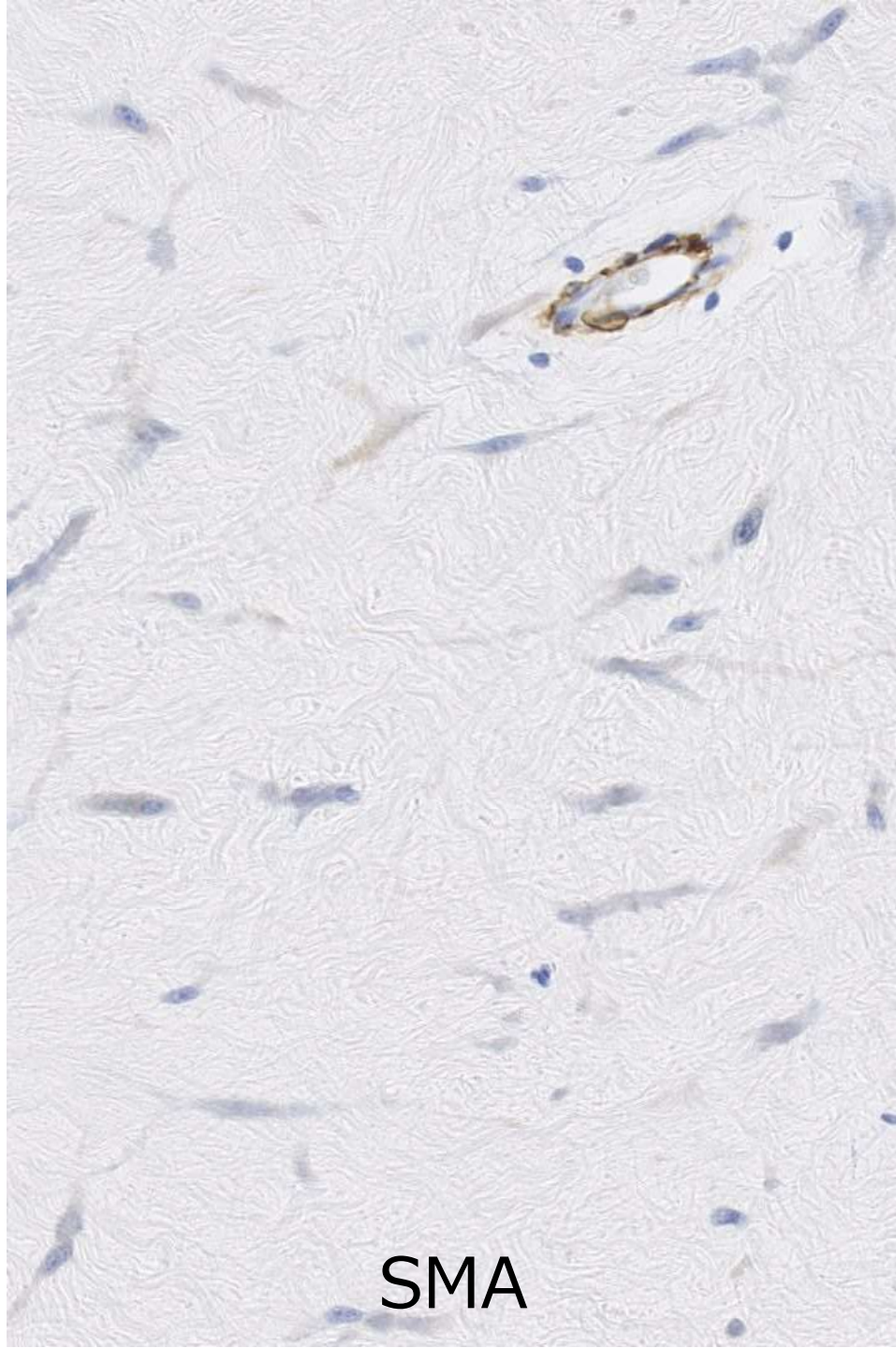









CD34


SMA

Nodular fasciitis

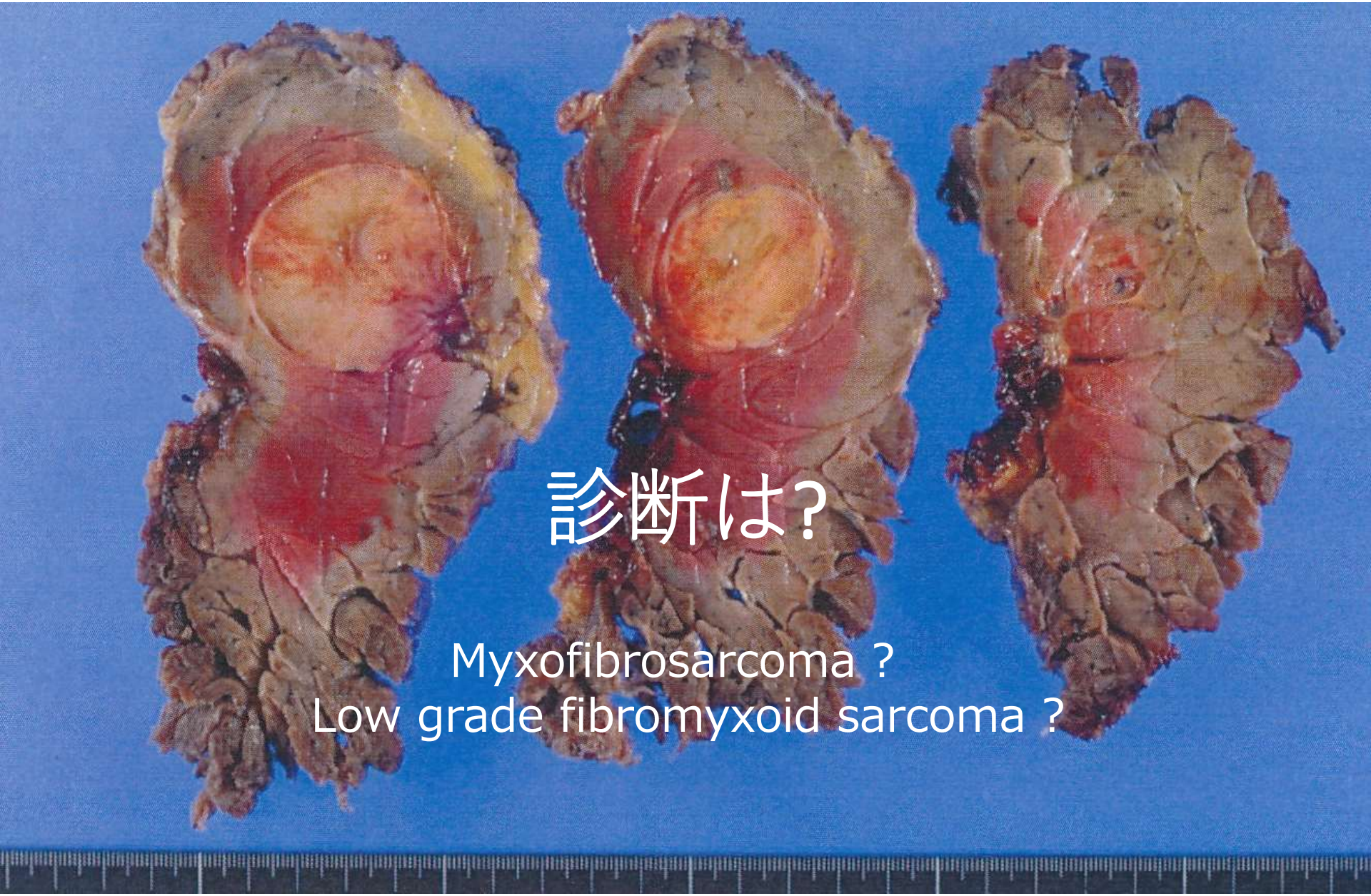
USP6 rearrangements

Fibroma of tendon sheath

Desmoplastic fibroblastoma

t(2;11)(q31;q12) FOSL (+)

Case 1: 45M, buttock



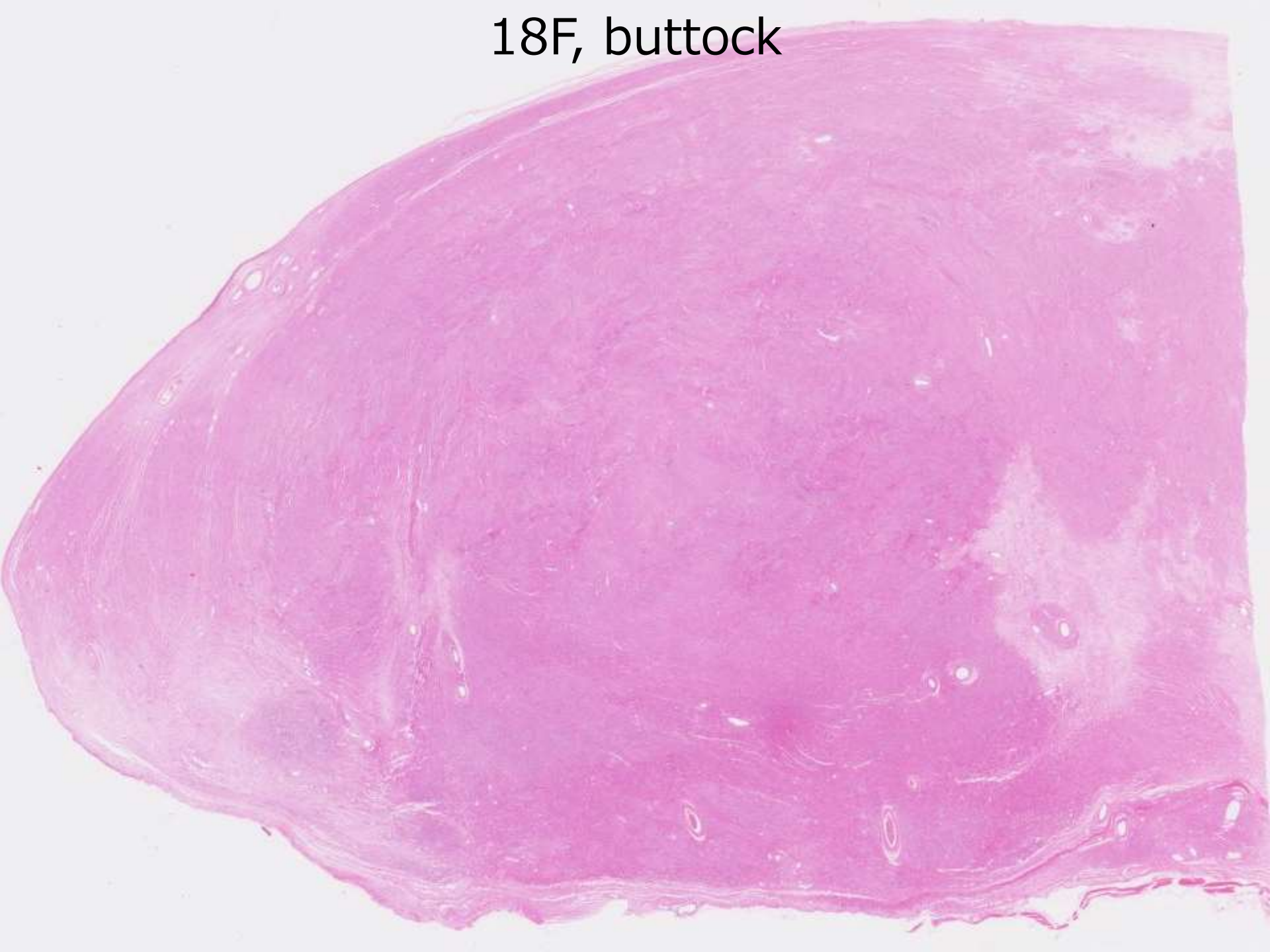
異型に乏しい紡錘形細胞腫瘍でしばしば鑑別に挙げられる肉腫

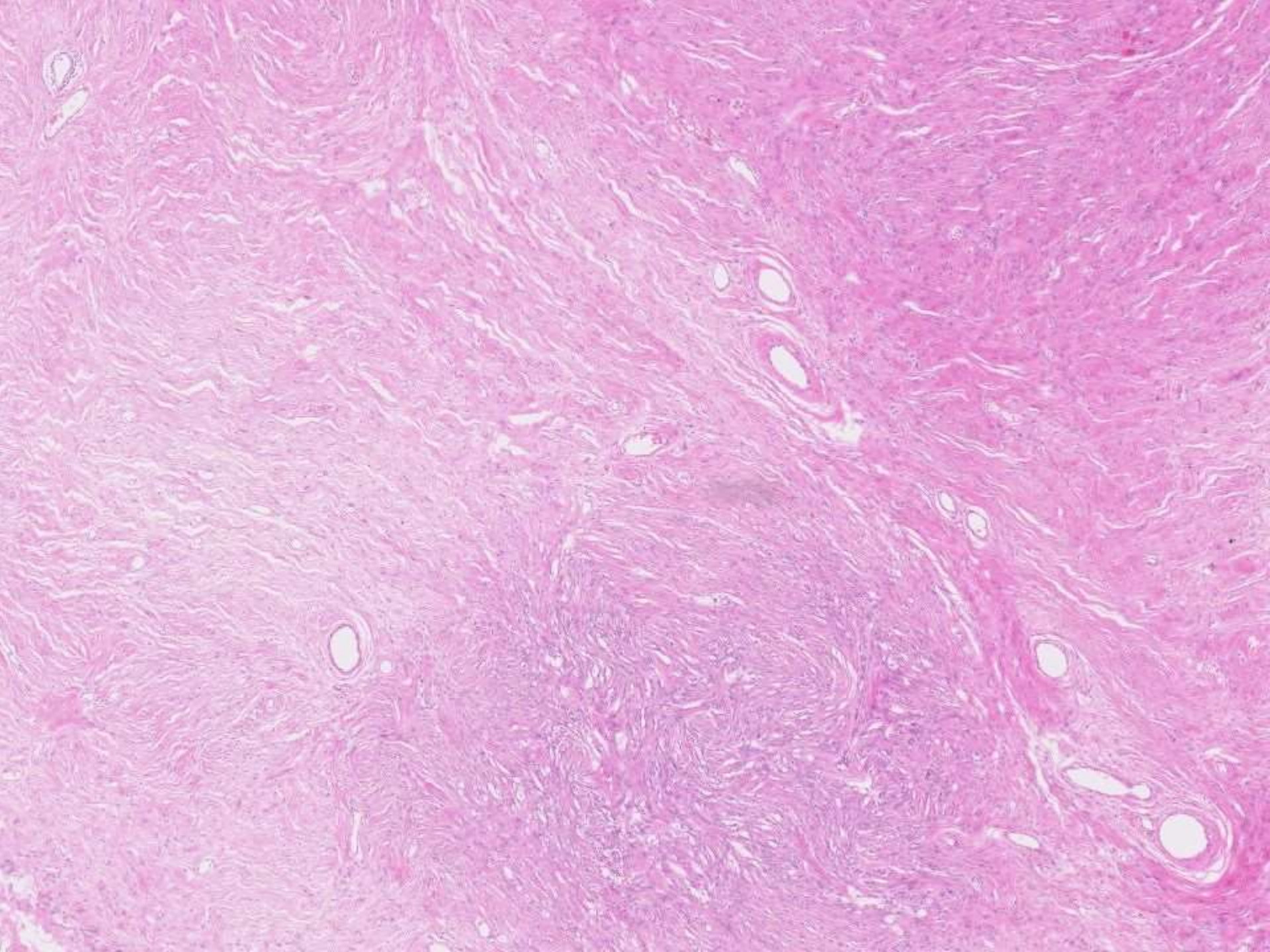
1. Low grade fibromyxoid sarcoma
2. (low grade) myxofibrosarcoma
3. Low grade myofibroblastic sarcoma

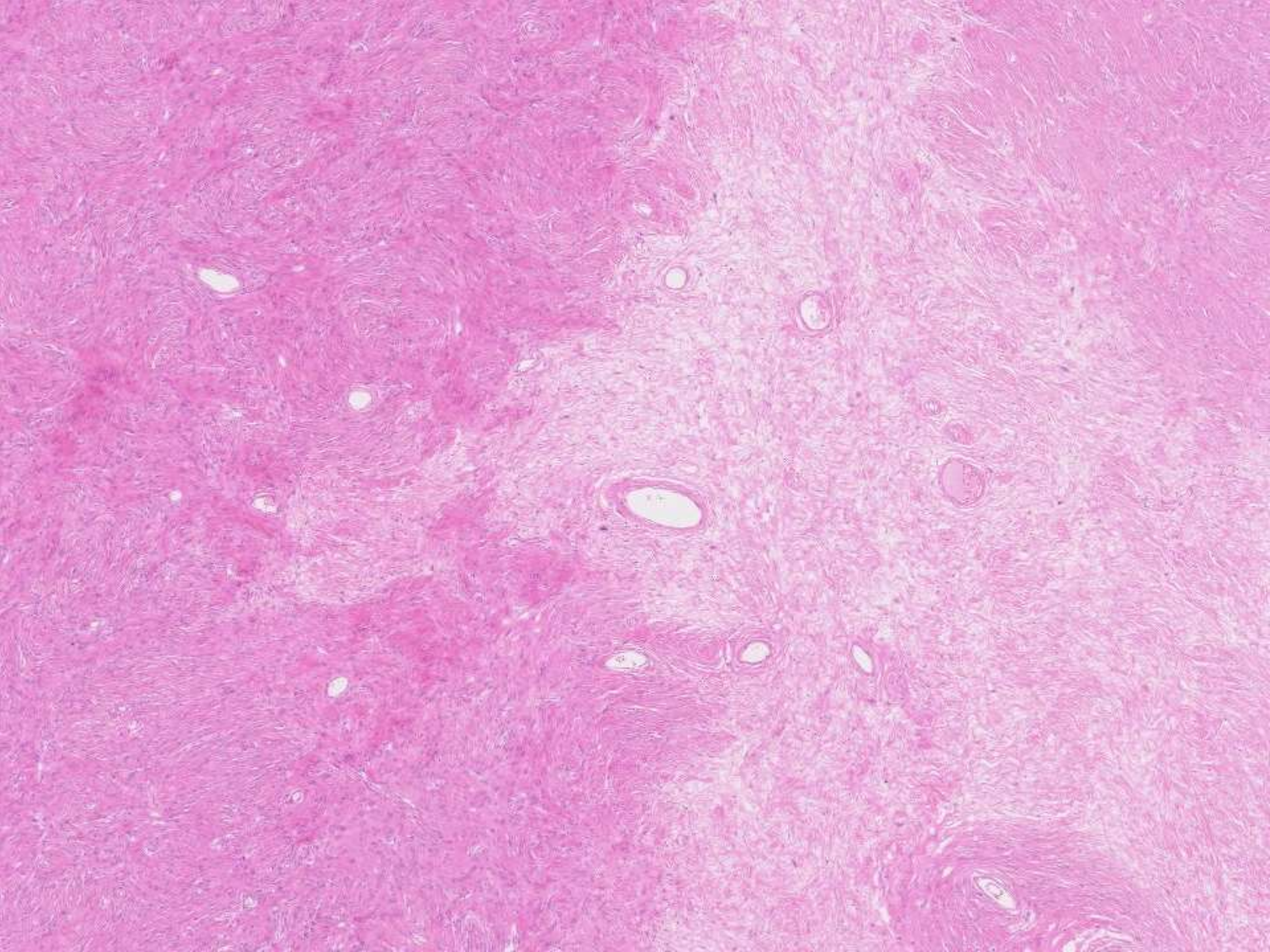
異型に乏しい紡錘形細胞腫瘍でしばしば鑑別に挙げられる肉腫

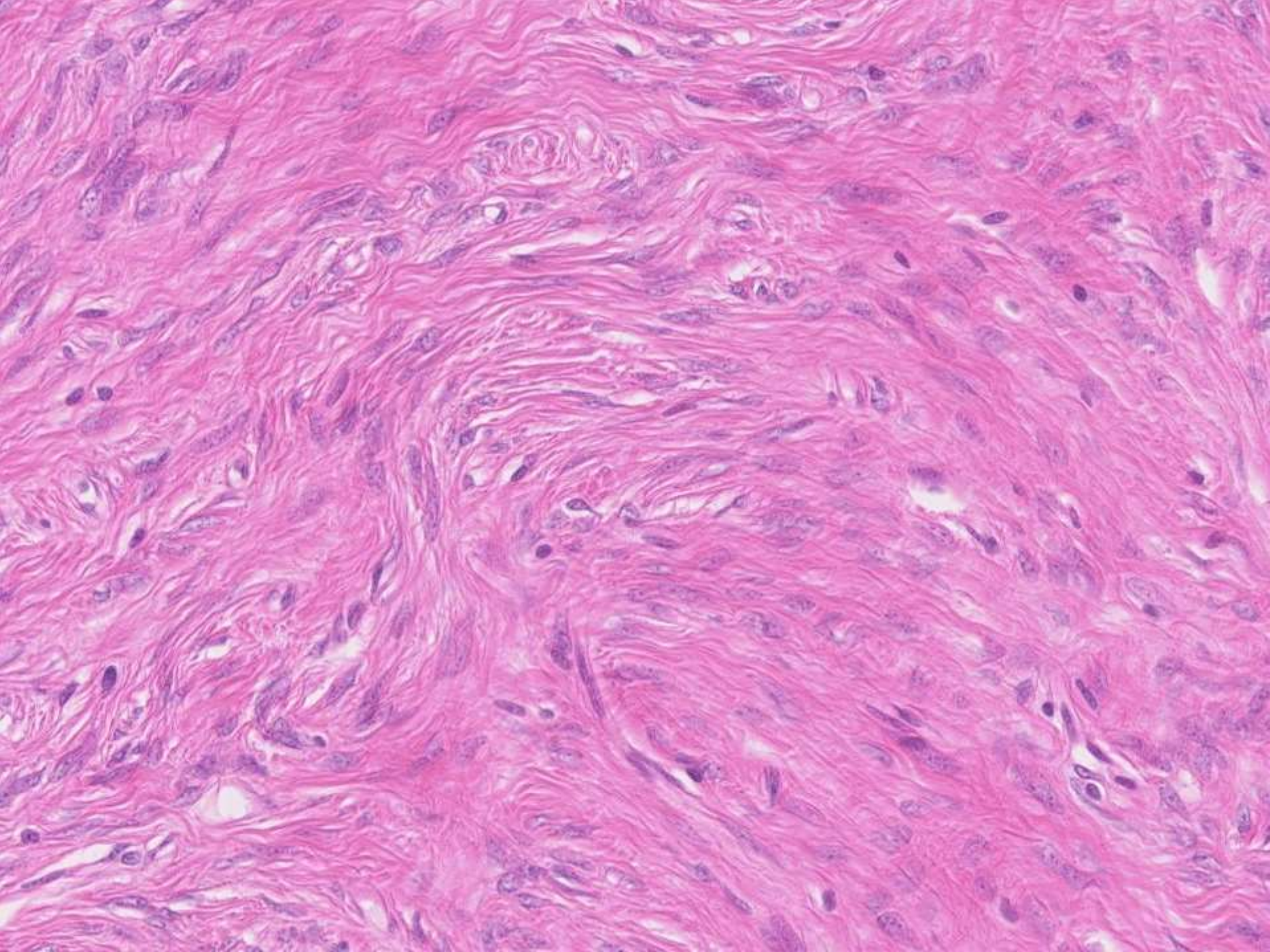
1. Low grade fibromyxoid sarcoma
2. (low grade) myxofibrosarcoma
3. Low grade myofibroblastic sarcoma

18F, buttock









Brief Scientific Reports

Low-Grade Fibromyxoid Sarcoma

A Report of Two Metastasizing Neoplasms Having a Deceptively Benign Appearance

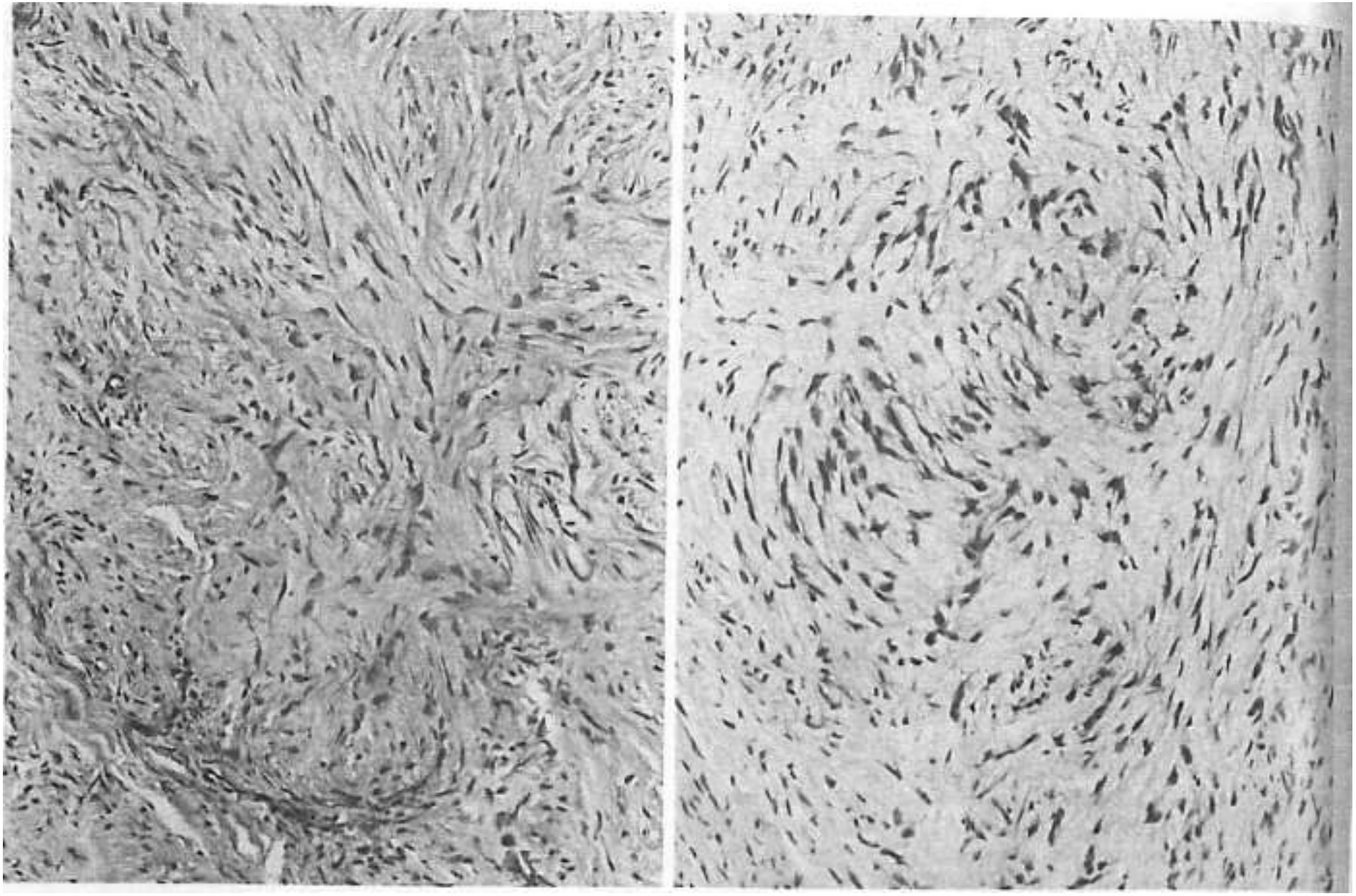
HARRY L. EVANS., M.D.

Department of Pathology, University of Texas M.D. Anderson Hospital, Houston, Texas

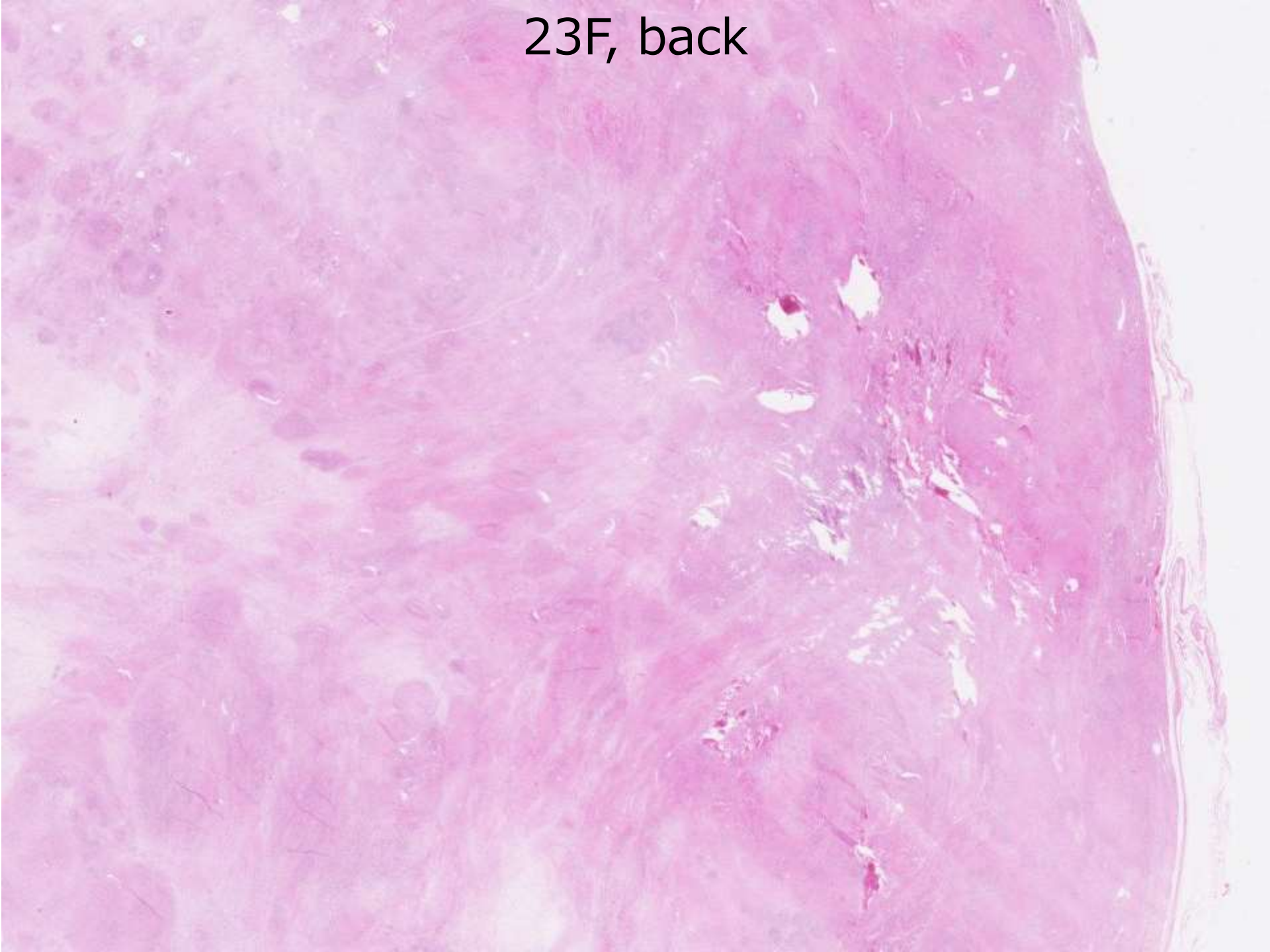
Two deceptively benign-appearing, unclassifiable but very similar fibromyxoid sarcomas characterized histologically by bland, innocuous-appearing fibroblastic cells and a swirling, whorled growth pattern are presented. The tumors both occurred in women in their late twenties and were located in the soft tissues of the scapular area and the axillary–chest wall area, respectively. Lung metastases developed in both cases; one patient died 94 months after excision of the primary neoplasm, whereas the other was alive at 82 months. The designation "low-grade fibromyxoid sarcoma" is suggested for these tumors. (Key words: Low-grade fibromyxoid sarcoma; Soft-tissue sarcoma) Am J Clin Pathol 1987; 88: 615–619

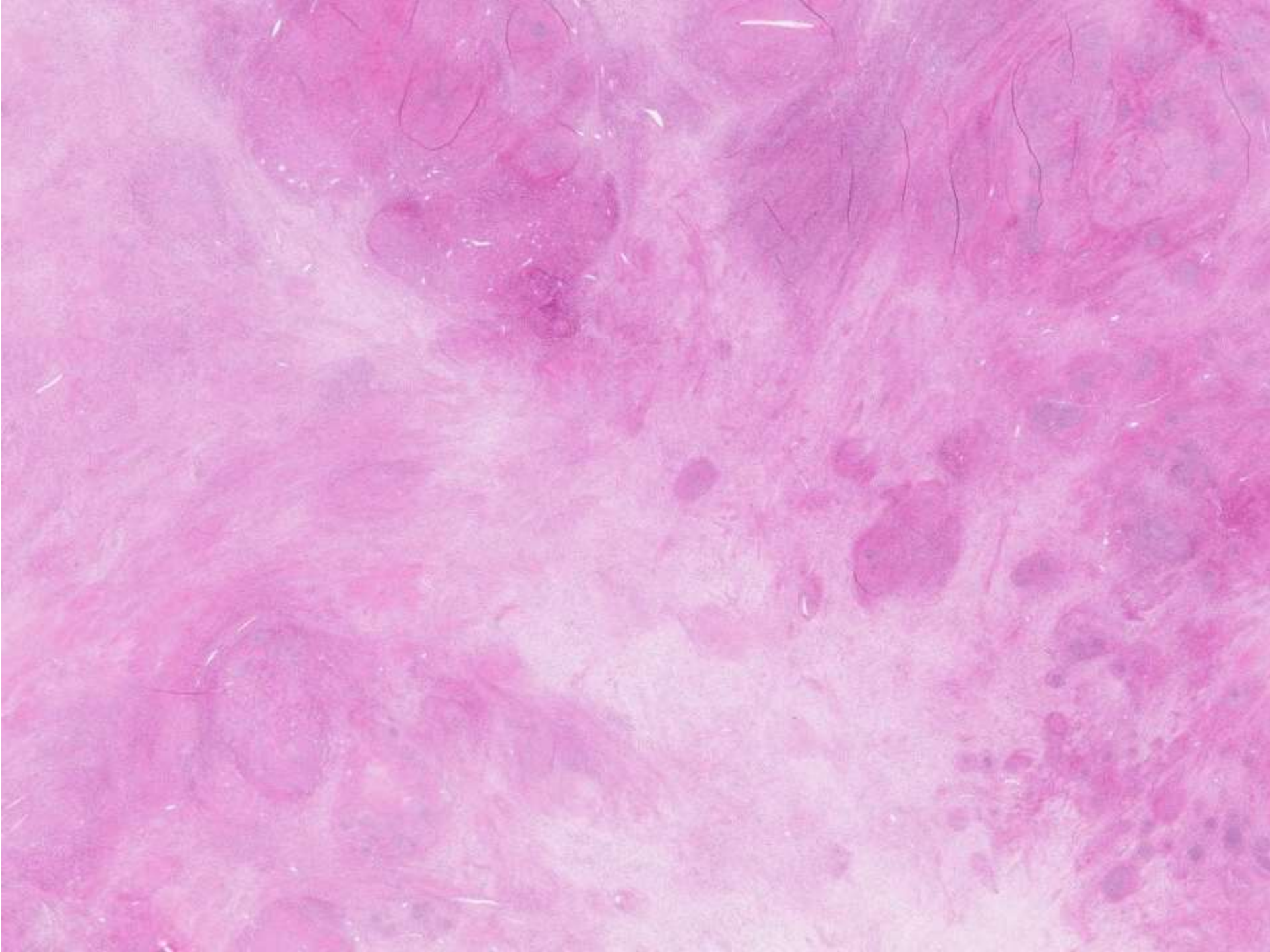
excision of the recurrence, the patient had an attack of "bronchitis," and chest x-ray revealed multiple, bilateral lung nodules. An open biopsy of the left upper lobe demonstrated metastatic sarcoma, and the patient was treated with a Cytoxan–Adriamycin–DTIC regimen for four months, during which time the nodules increased slightly in size. Radiotherapy in a dose of 4,000 rads and hyperthermia were then administered to the largest metastasis (in the left lower lobe), and, two

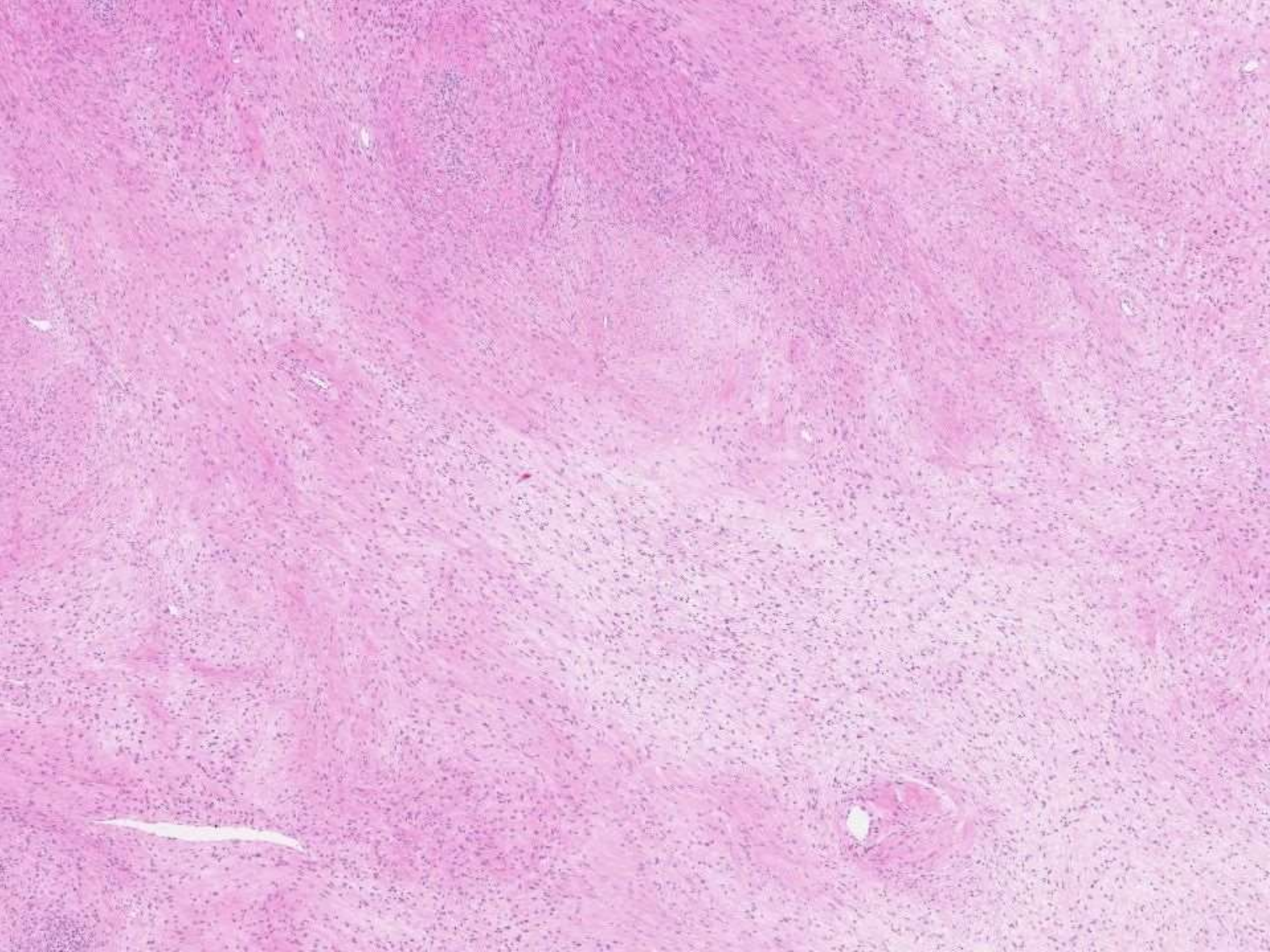
Fibroma, fibromatosis (desmoid)と診断されていた2例

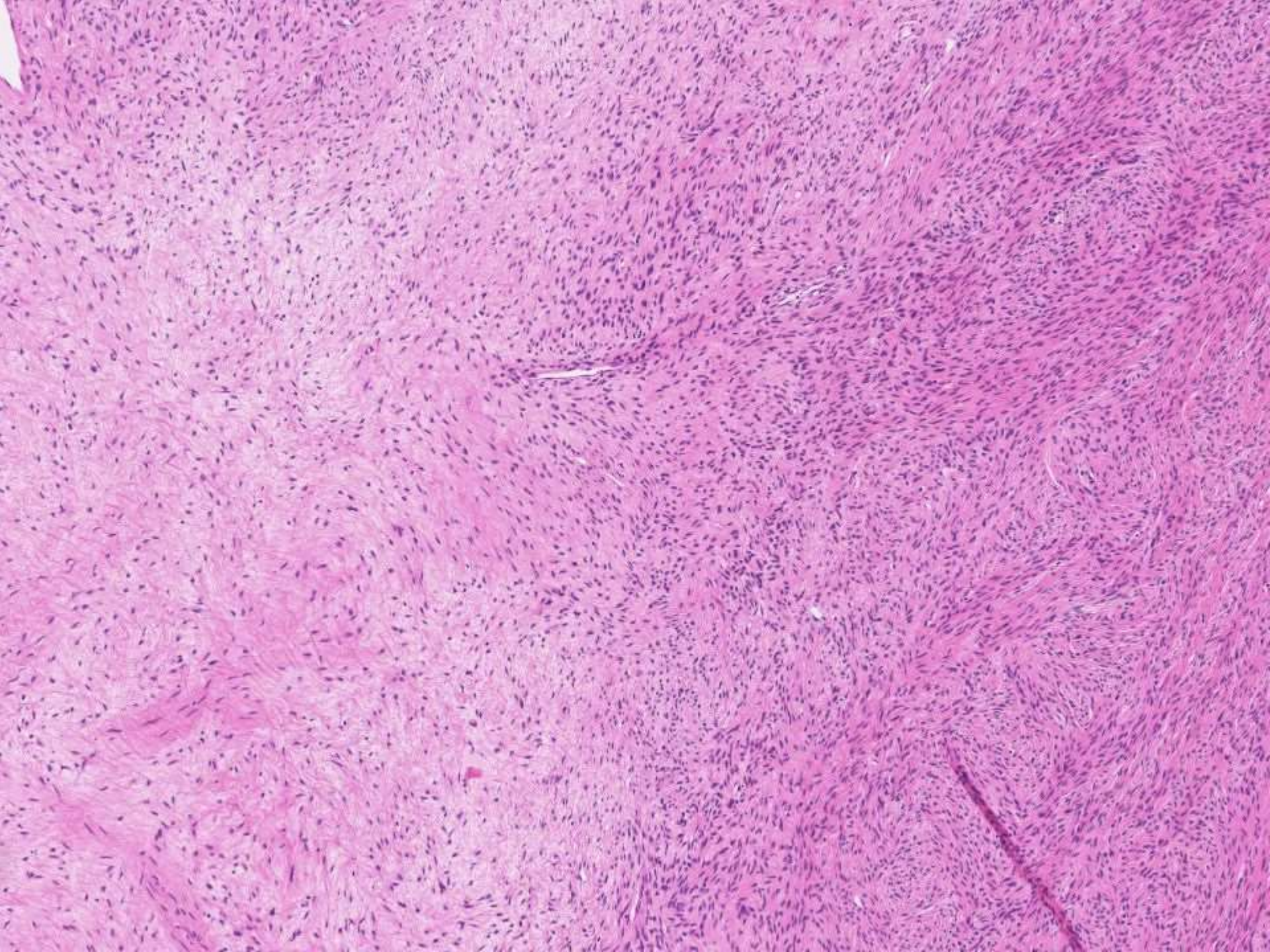


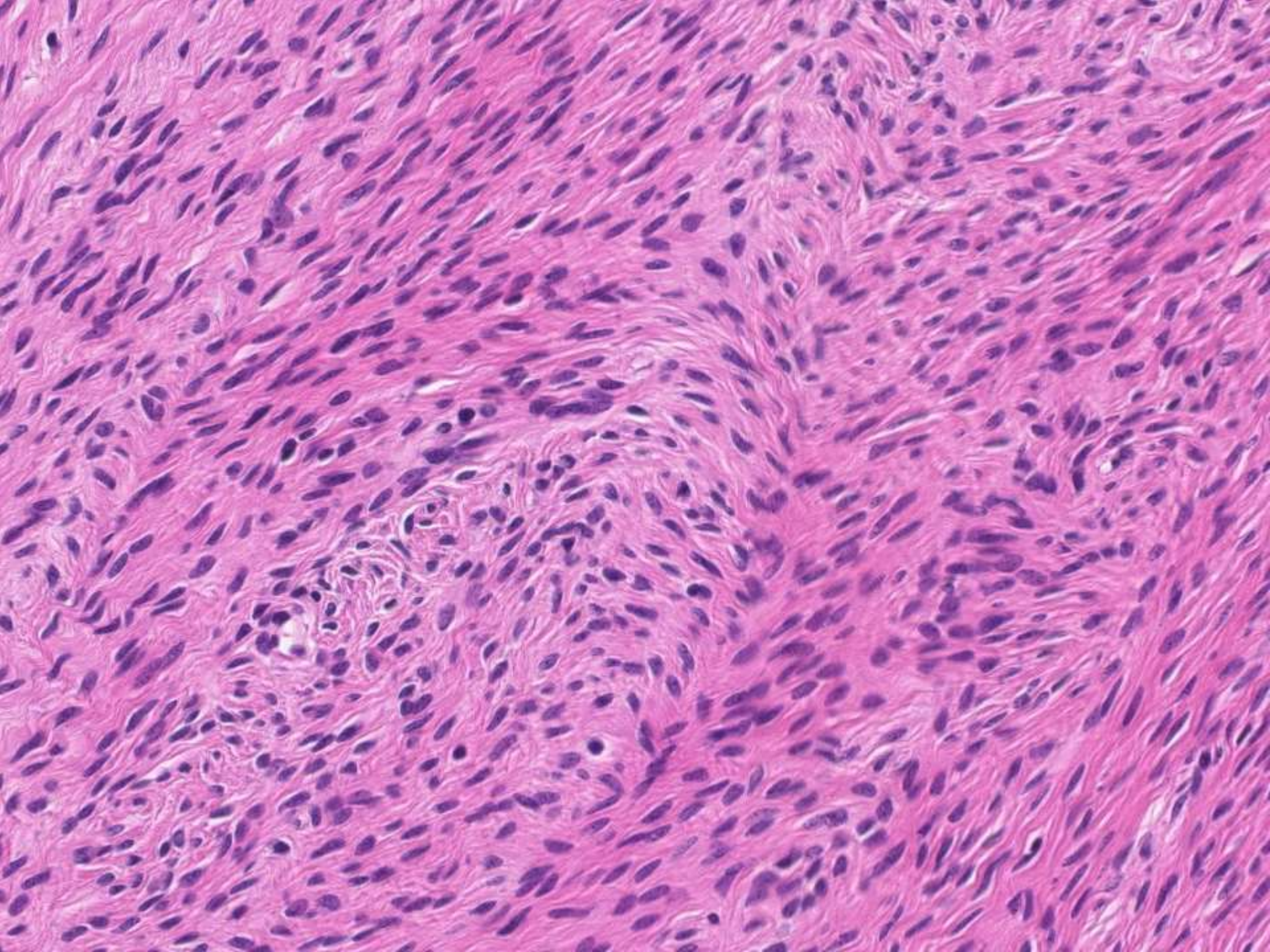
Evans HL. Low-grade fibromyxoid sarcoma. A report of two metastasizing neoplasms having a deceptively benign appearance. Am J Clin Pathol 88: 615-619, 1987.

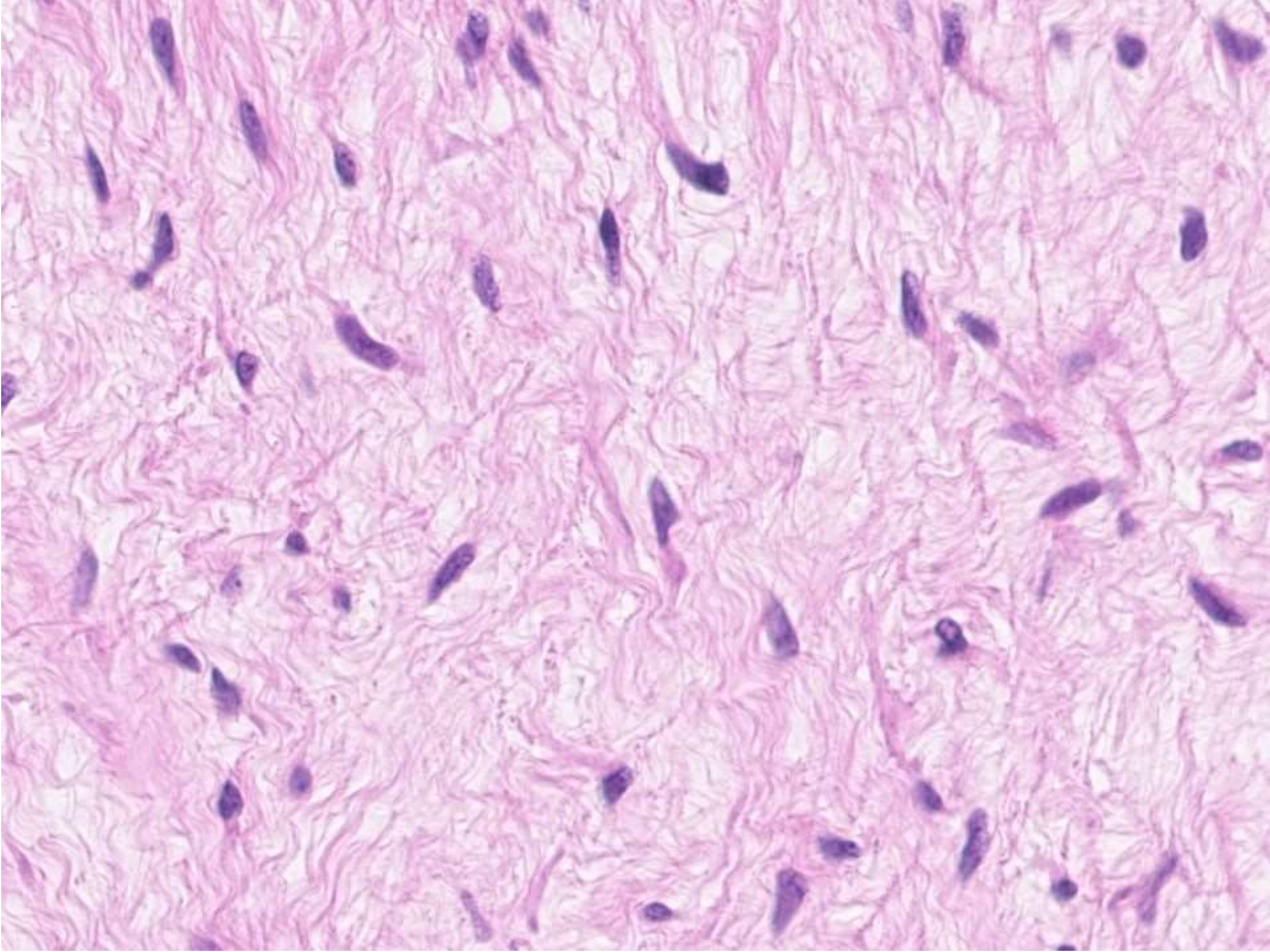

23F, back

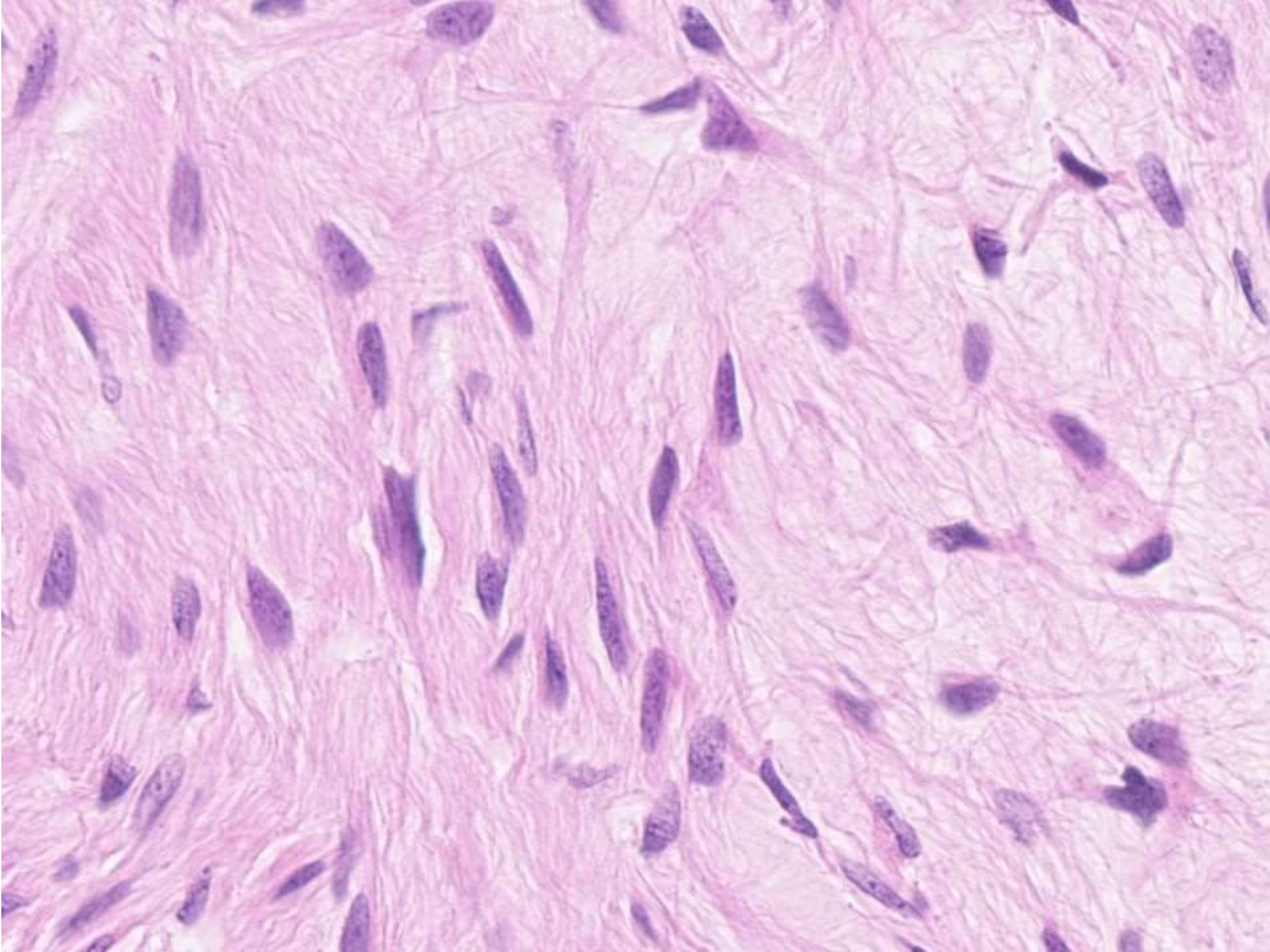


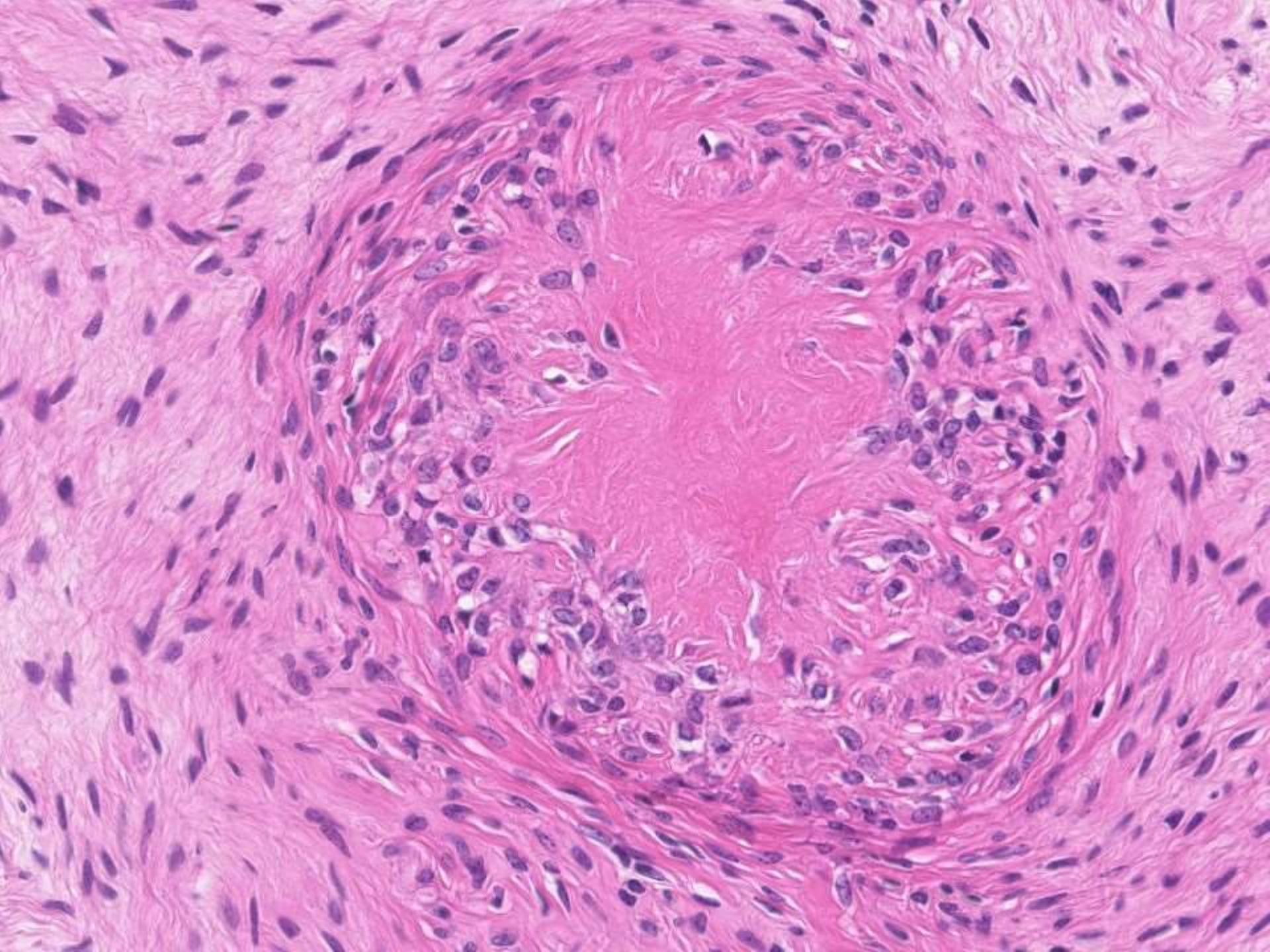


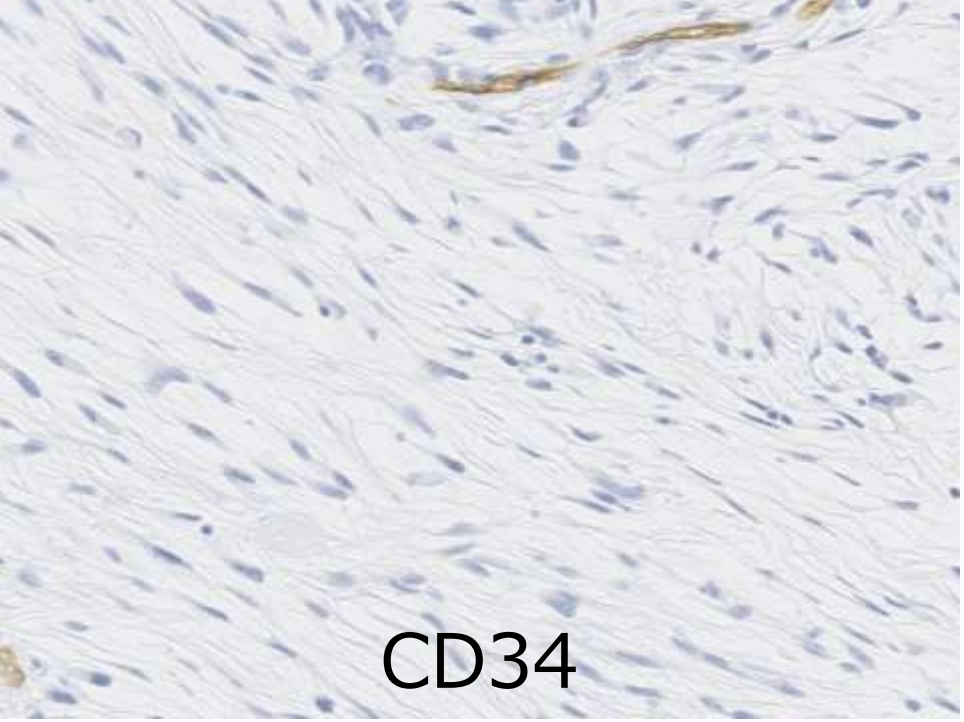


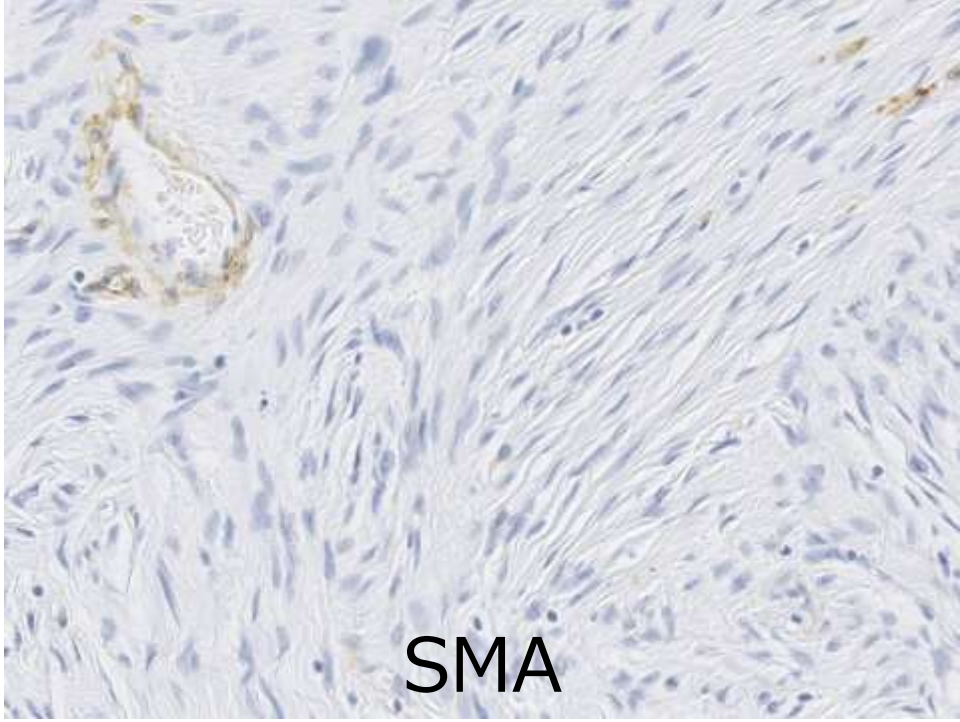


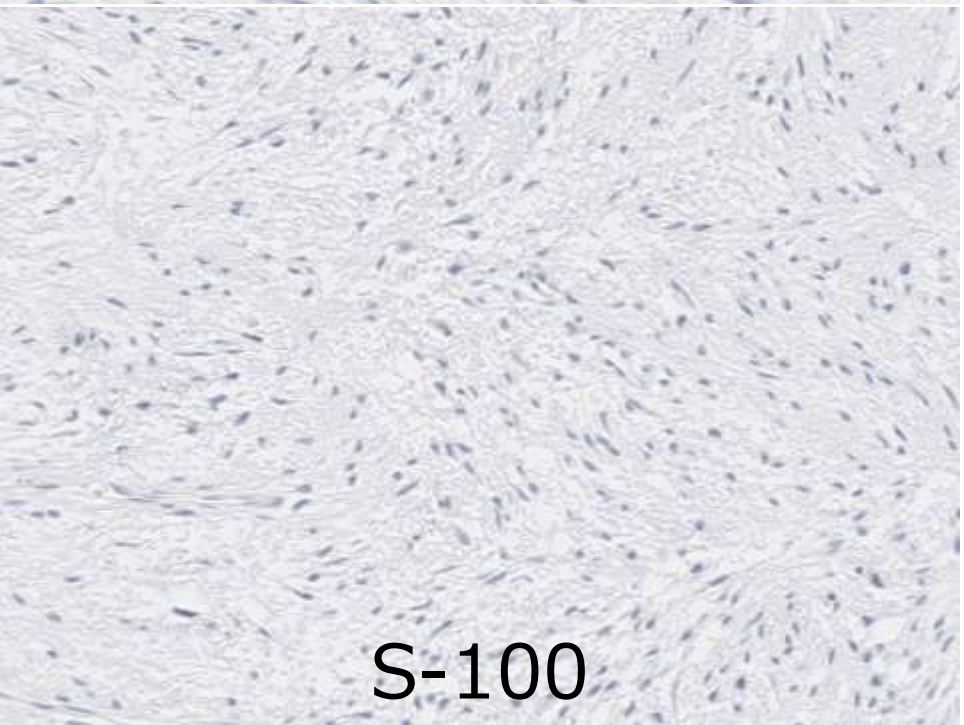


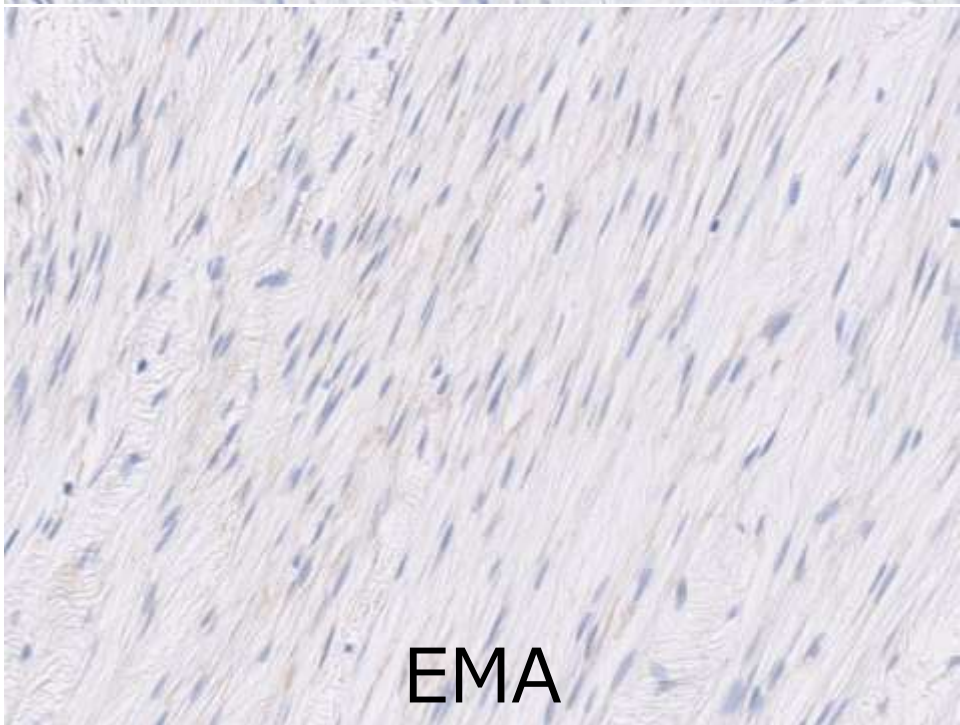




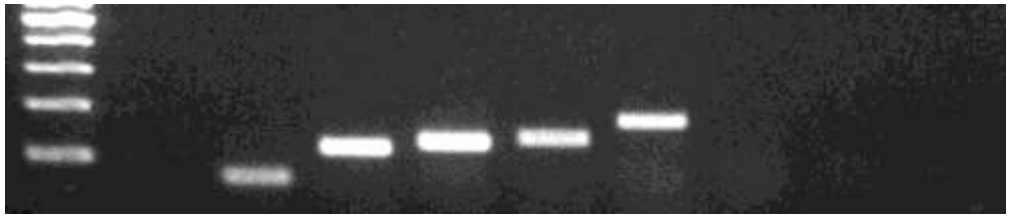

CD34

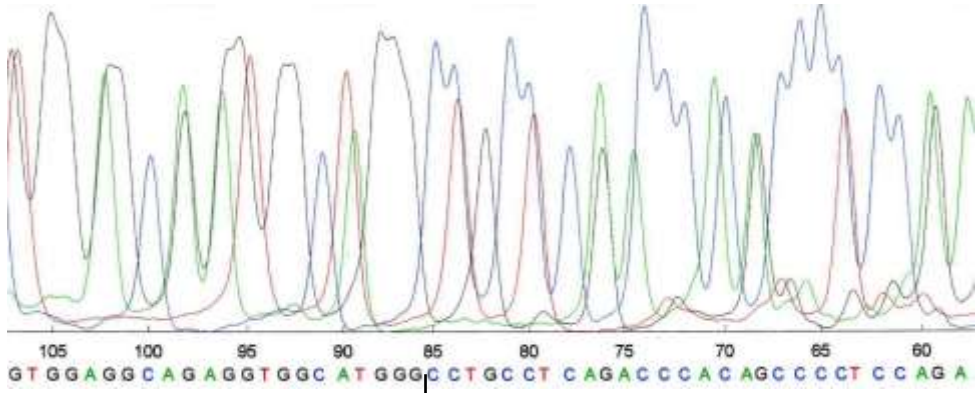

SMA


S-100

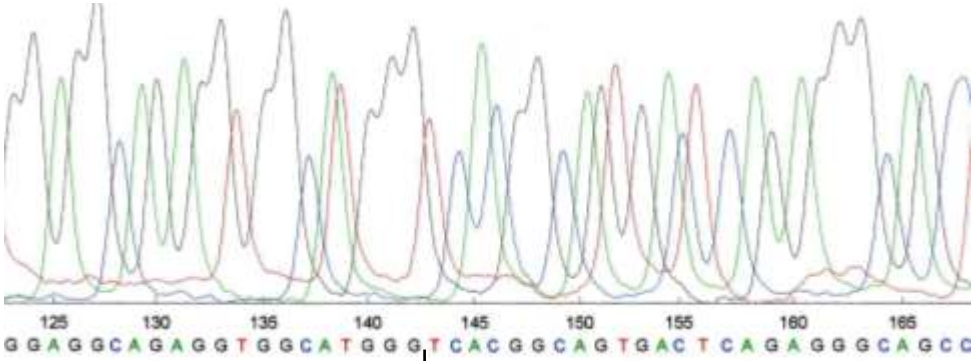

EMA

RT-PCR for FUS-CREB3L2 in LGFMS





FUS exon 7 ↔ CREB3L2 exon 5



FUS exon 6 ↔ CREB3L2 exon 5

Brief Scientific Reports

Low-Grade Fibromyxoid Sarcoma

A Report of Two Metastasizing Neoplasms Having a Deceptively Benign Appearance

HARRY L. EVANS., M.D.

Department of Pathology, University of Texas M.D. Anderson Hospital, Houston, Texas

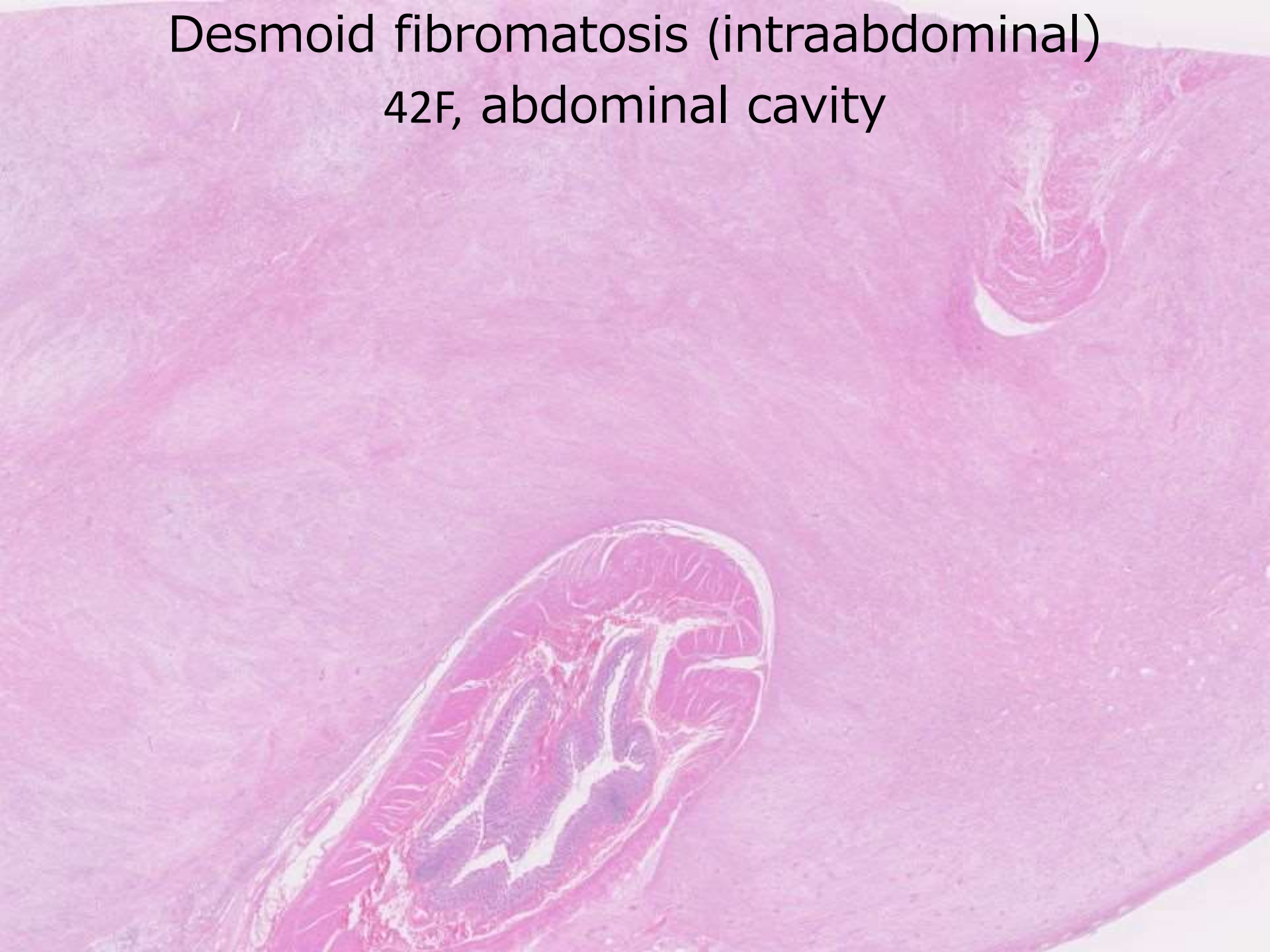
Two deceptively benign-appearing, unclassifiable but very similar fibromyxoid sarcomas characterized histologically by bland, innocuous-appearing fibroblastic cells and a swirling, whorled growth pattern are presented. The tumors both occurred in women in their late twenties and were located in the soft tissues of the scapular area and the axillary–chest wall area, respectively. Lung metastases developed in both cases; one patient died 94 months after excision of the primary neoplasm, whereas the other was alive at 82 months. The designation "low-grade fibromyxoid sarcoma" is suggested for these tumors. (Key words: Low-grade fibromyxoid sarcoma; Soft-tissue sarcoma) Am J Clin Pathol 1987; 88: 615–619

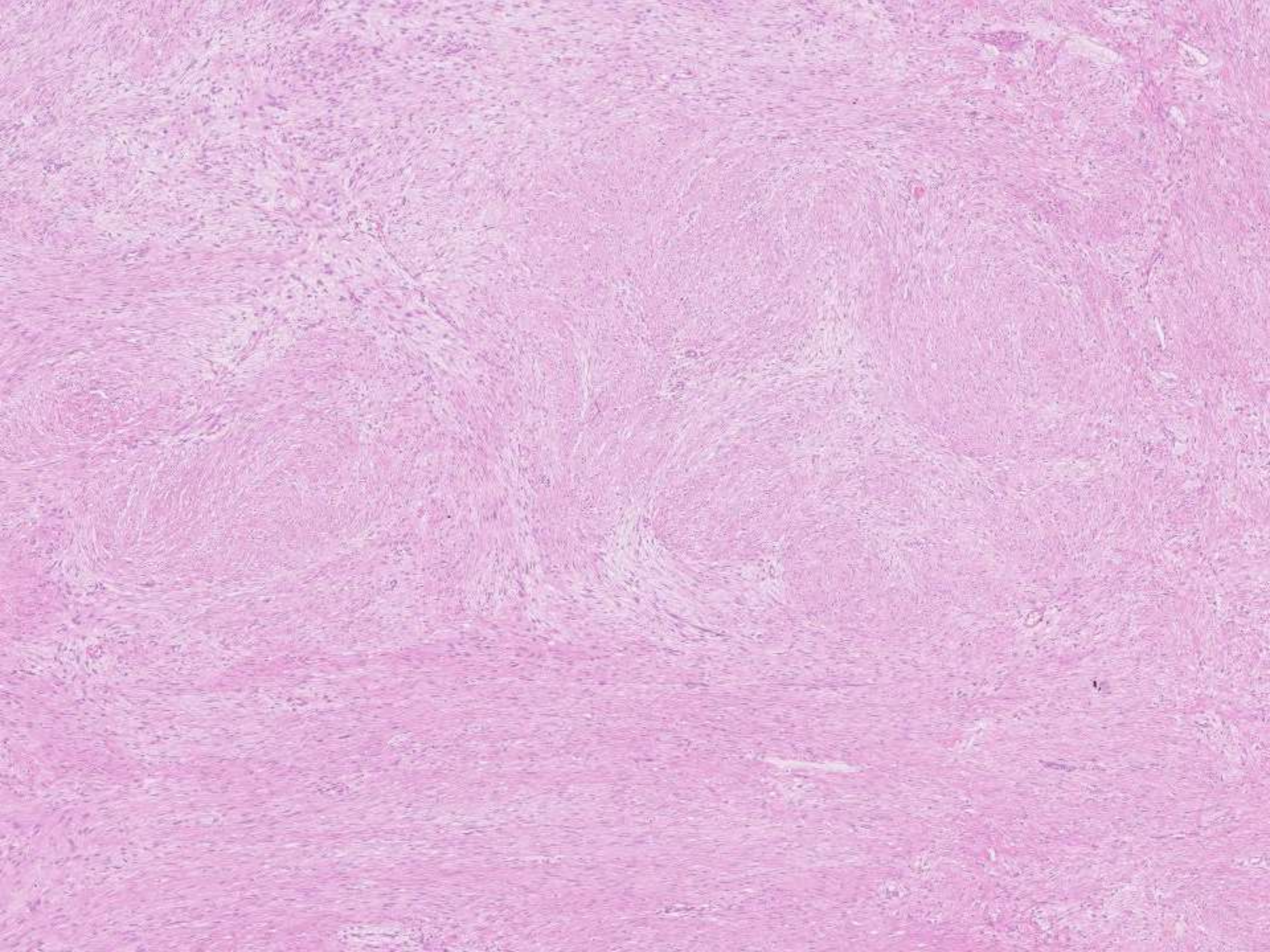
excision of the recurrence, the patient had an attack of "bronchitis," and chest x-ray revealed multiple, bilateral lung nodules. An open biopsy of the left upper lobe demonstrated metastatic sarcoma, and the patient was treated with a Cytoxan–Adriamycin–DTIC regimen for four months, during which time the nodules increased slightly in size. Radiotherapy in a dose of 4,000 rads and hyperthermia were then administered to the largest metastasis (in the left lower lobe), and, two

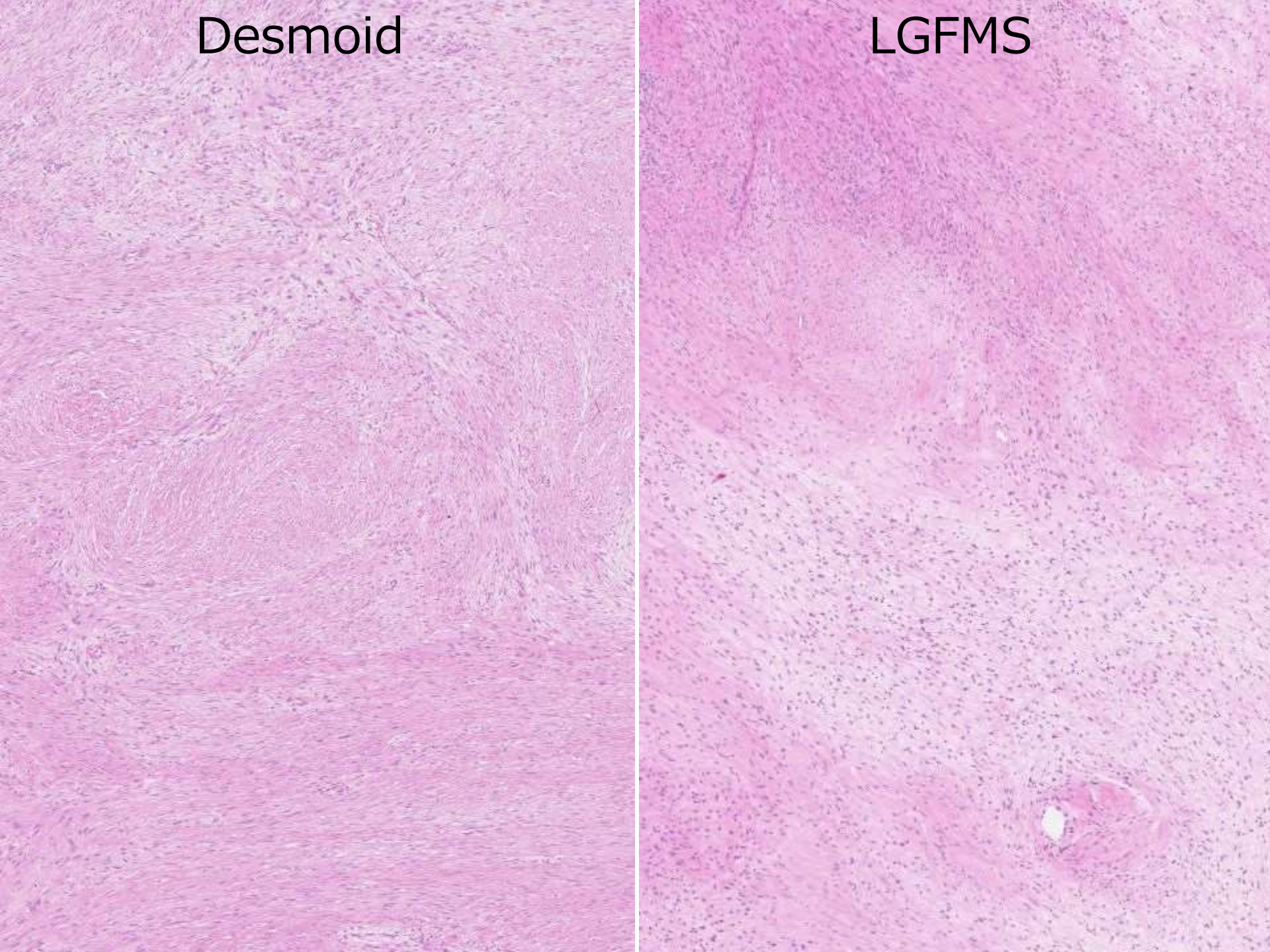
Fibroma, fibromatosis (desmoid)と診断されていた2例

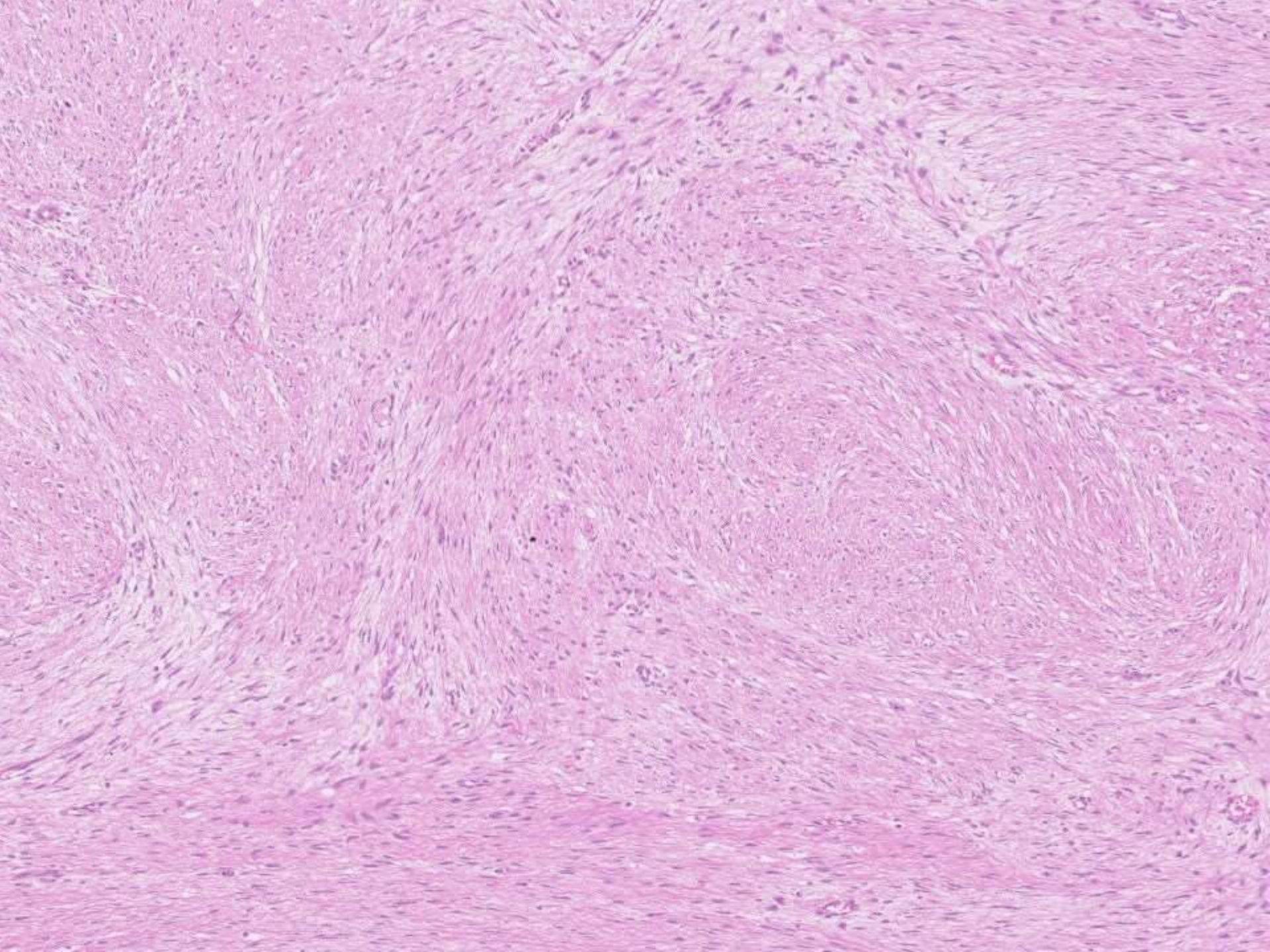
Desmoid fibromatosis (intraabdominal)

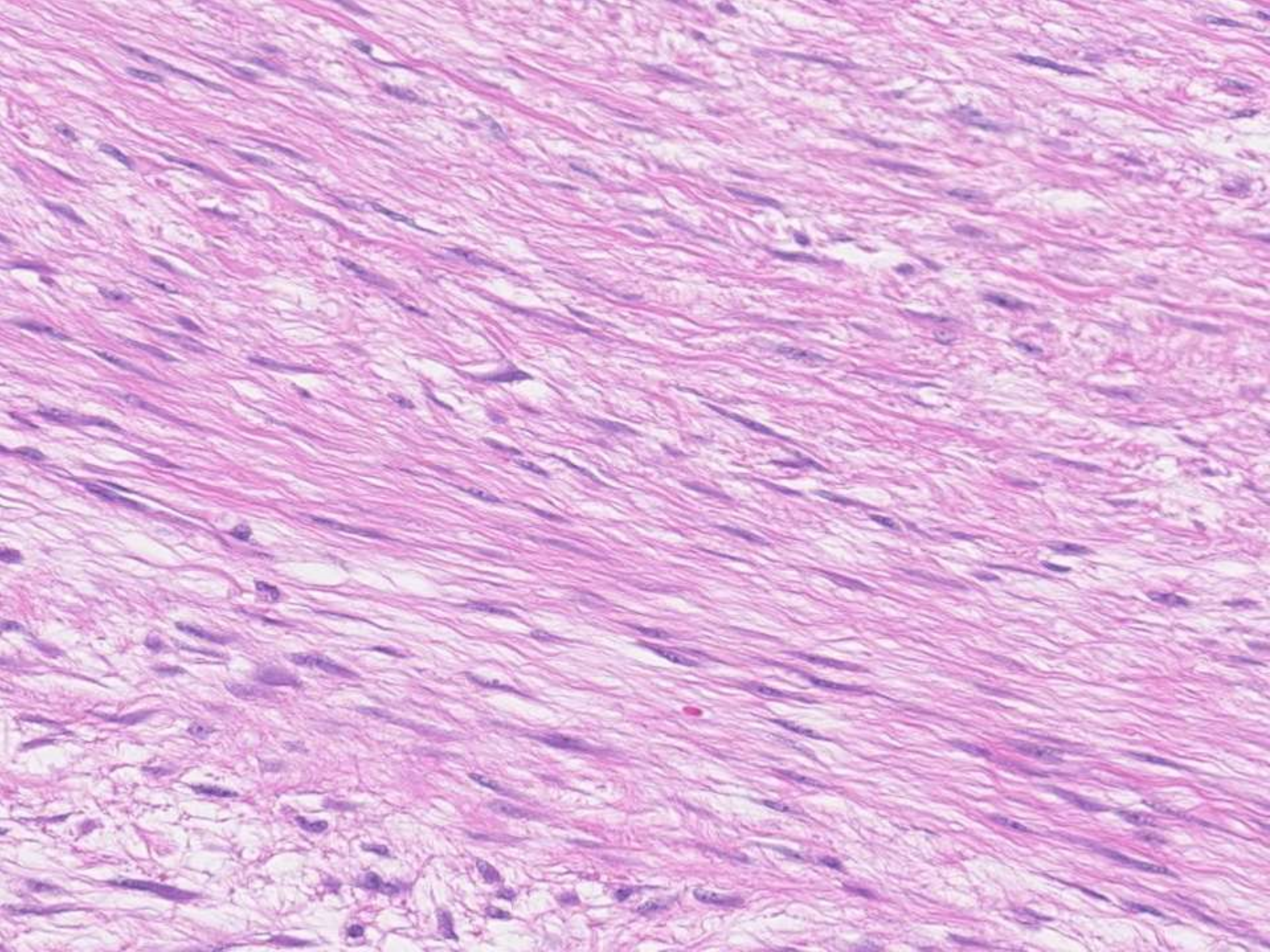
42F, abdominal cavity

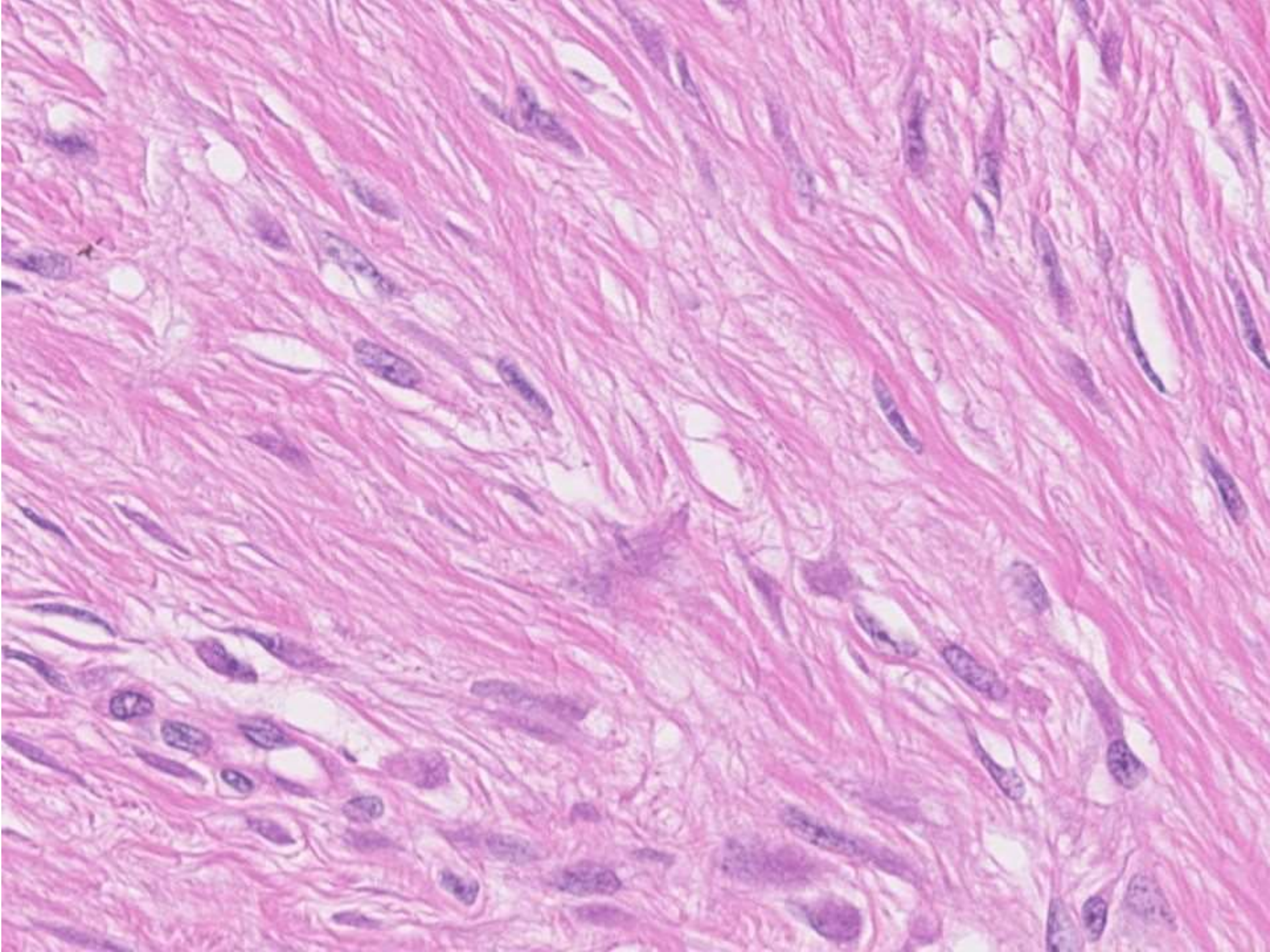


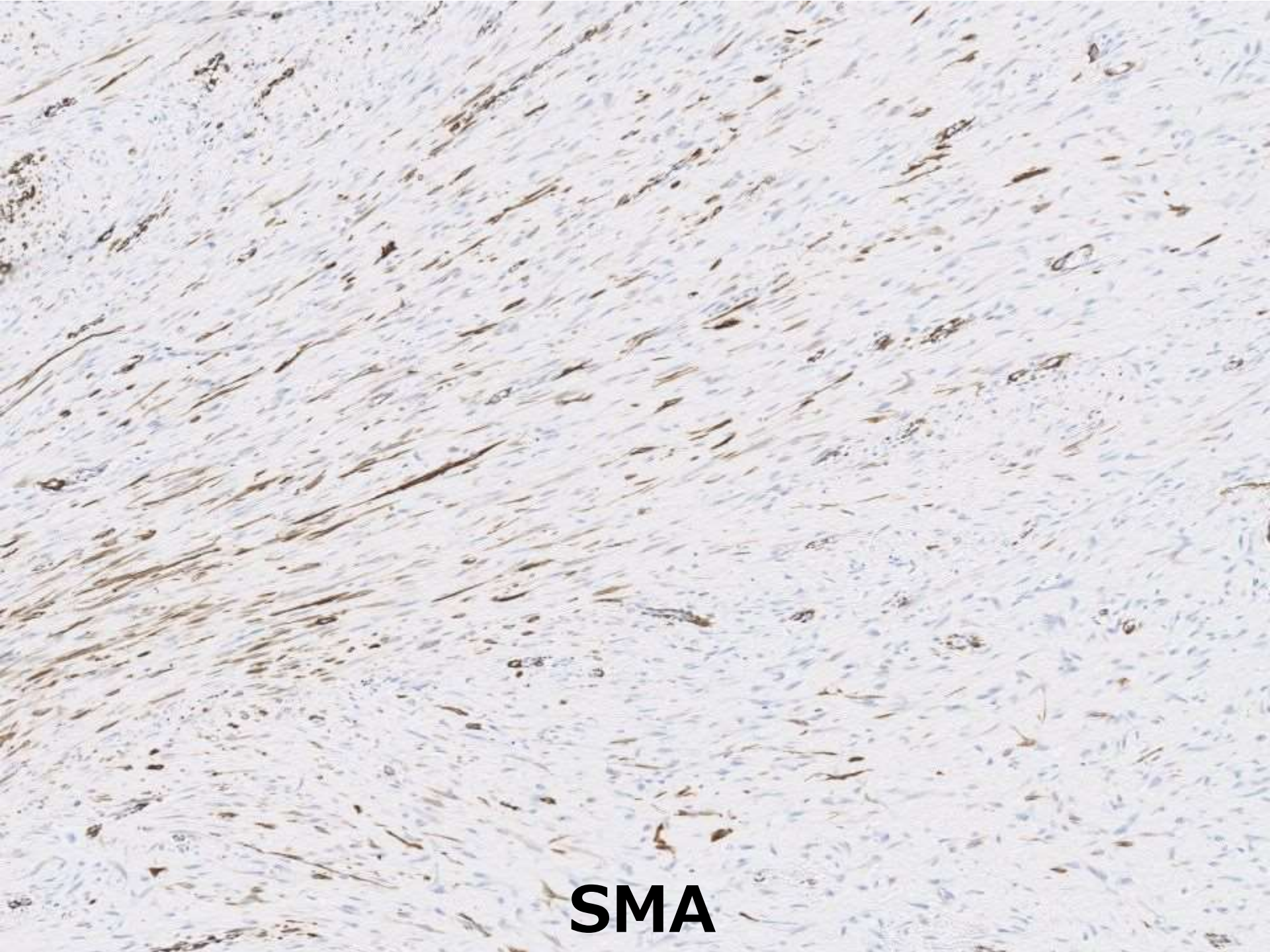


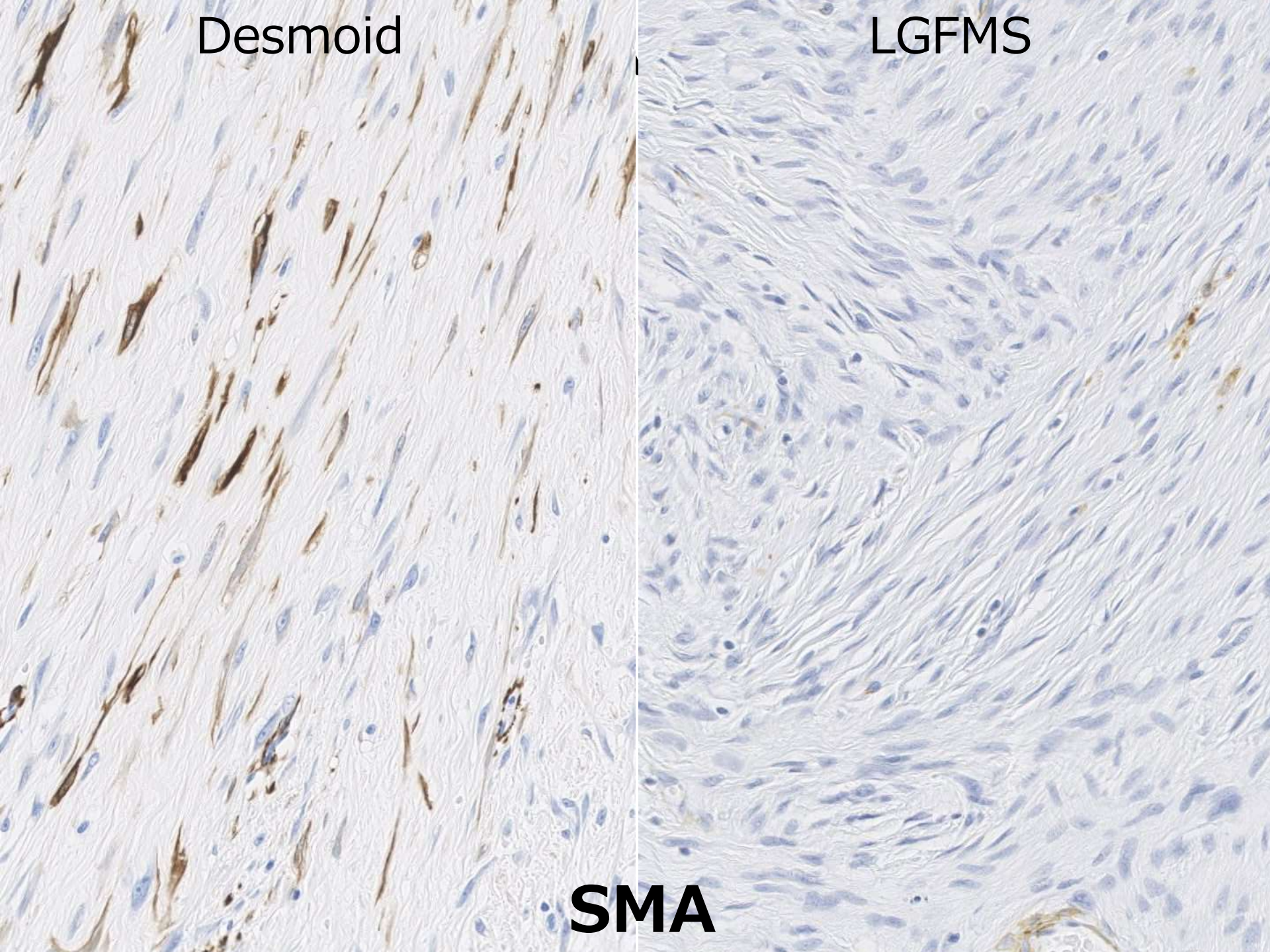

Desmoid
LGFMS

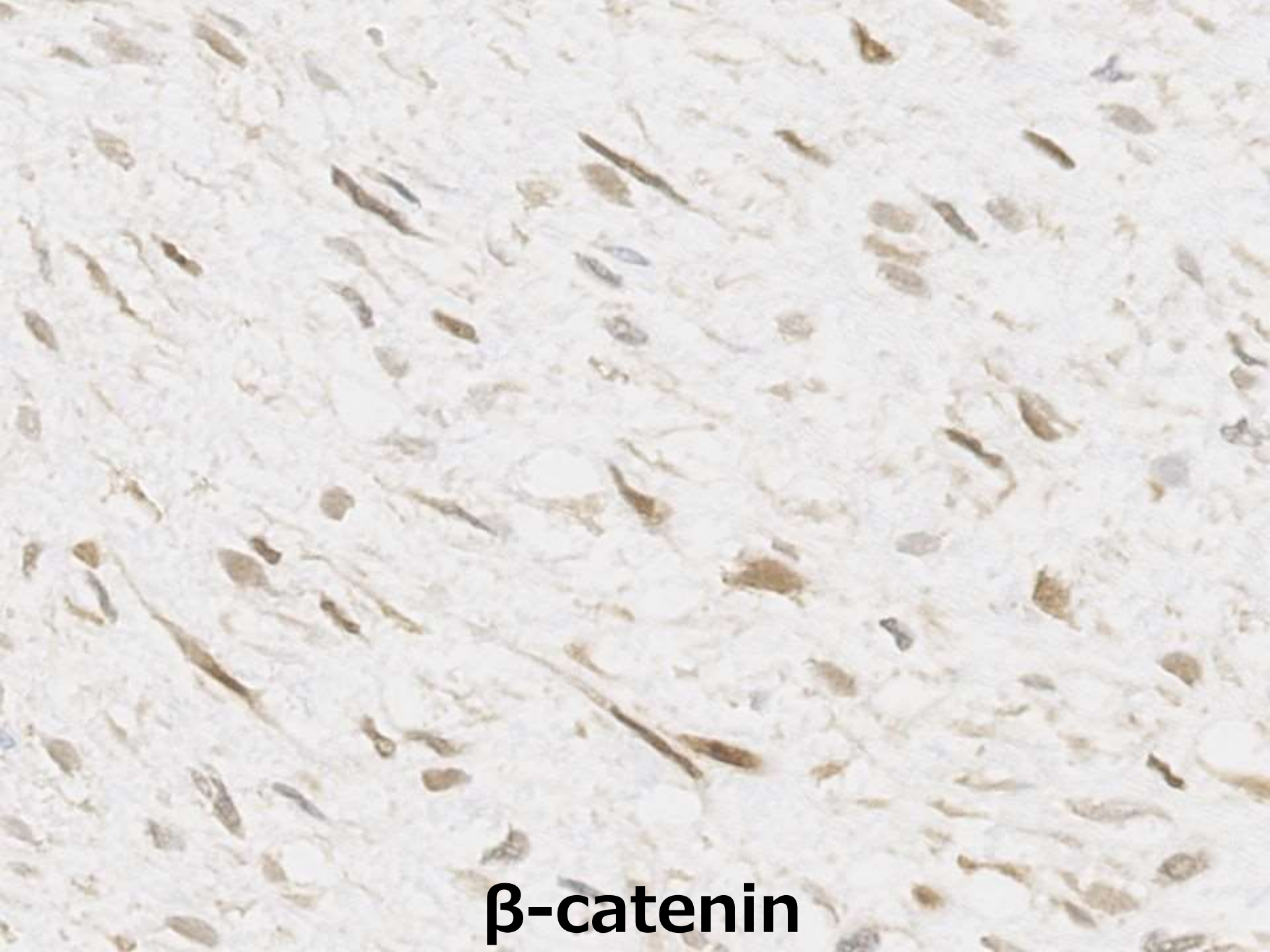







SMA


Desmoid
LGFMS
SMA

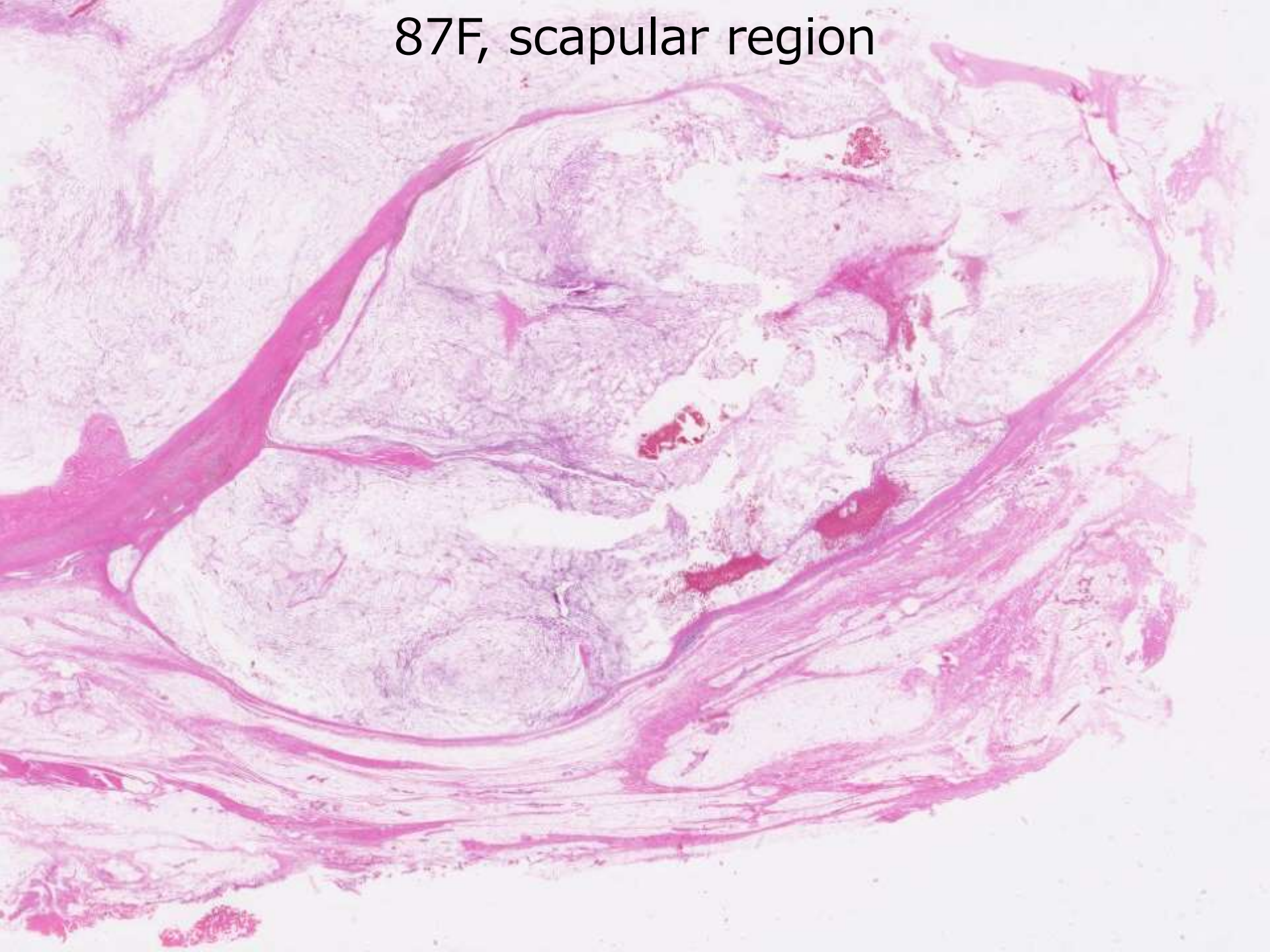

β-catenin

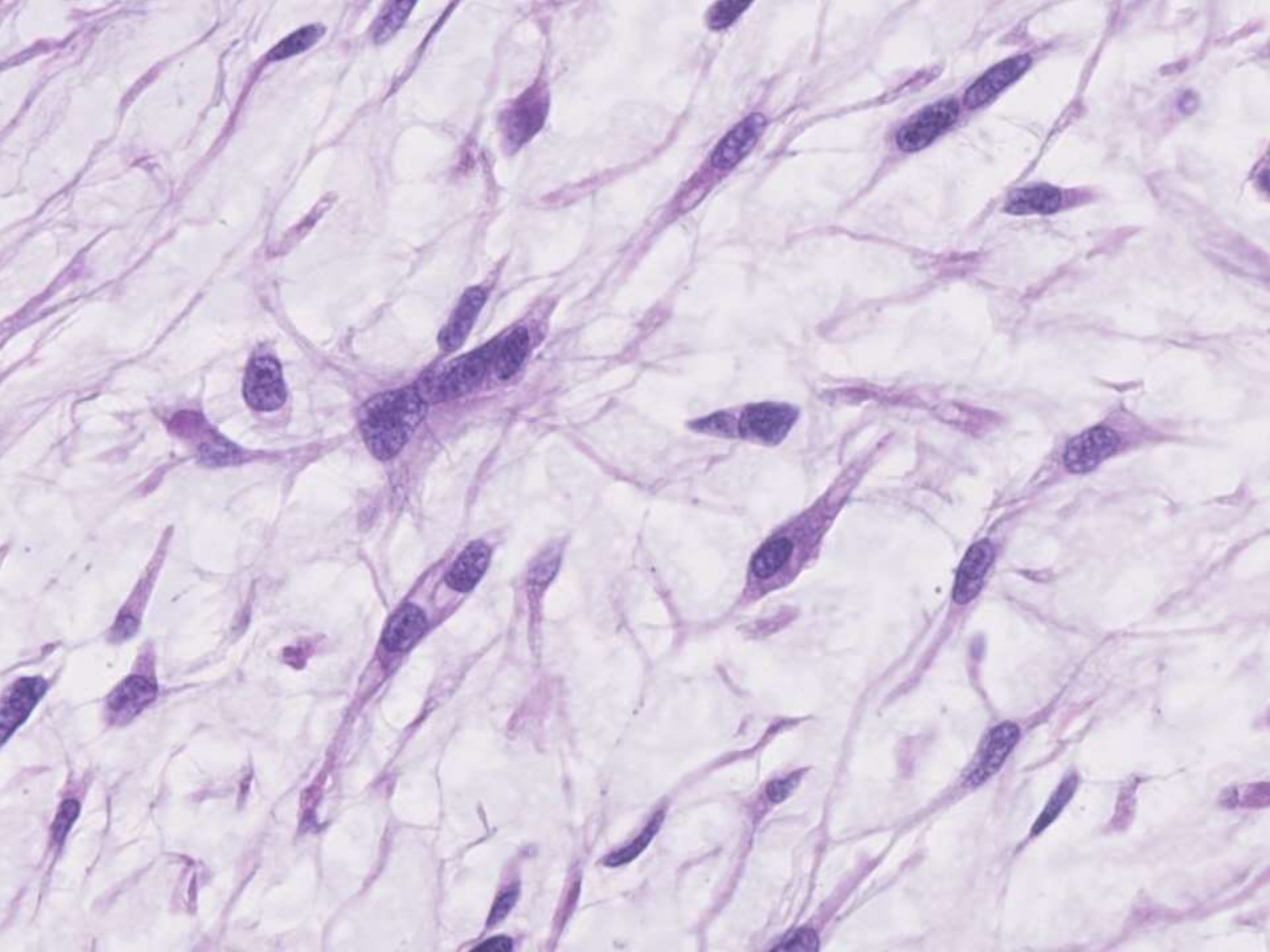
Tips 2

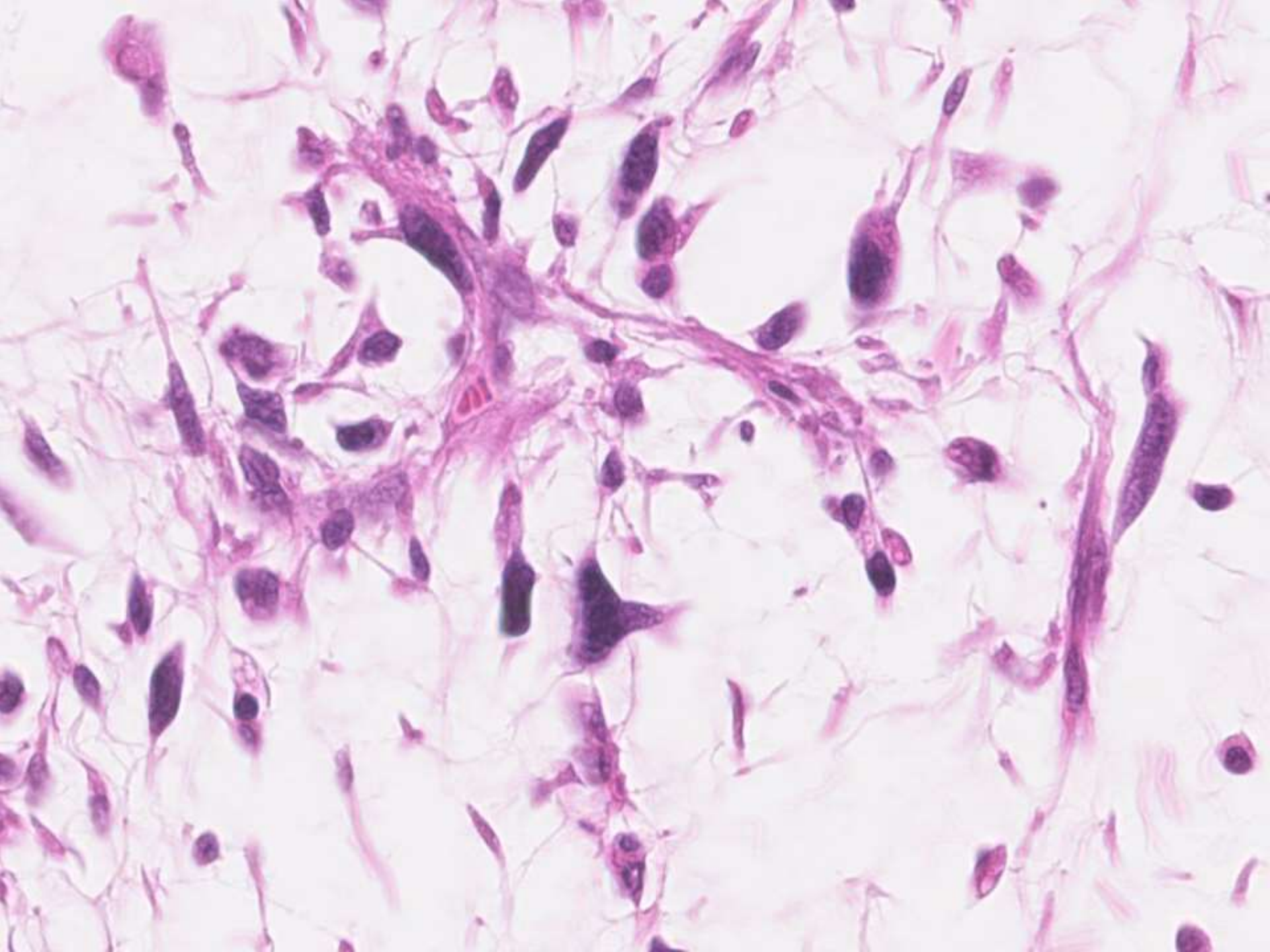
- LGFMSの粘液腫状間質はあまり高度ではない（ズブズブにはならない）
- LGFMSに一般的な核異型はない
- LGFMSはSMA(-)、desmin(-)、CD34(-)

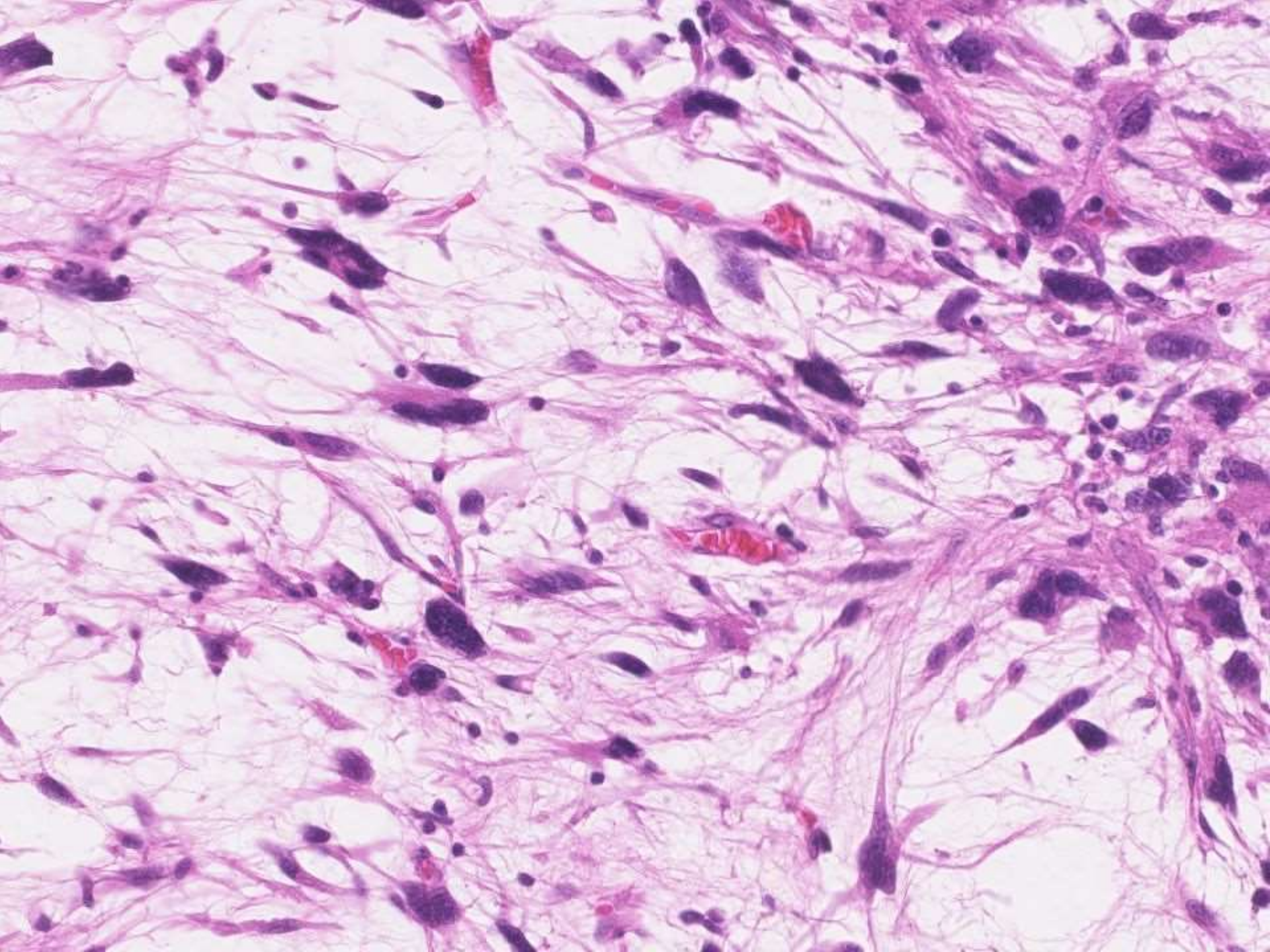
異型に乏しい紡錘形細胞腫瘍でしばしば鑑別に挙げられる肉腫

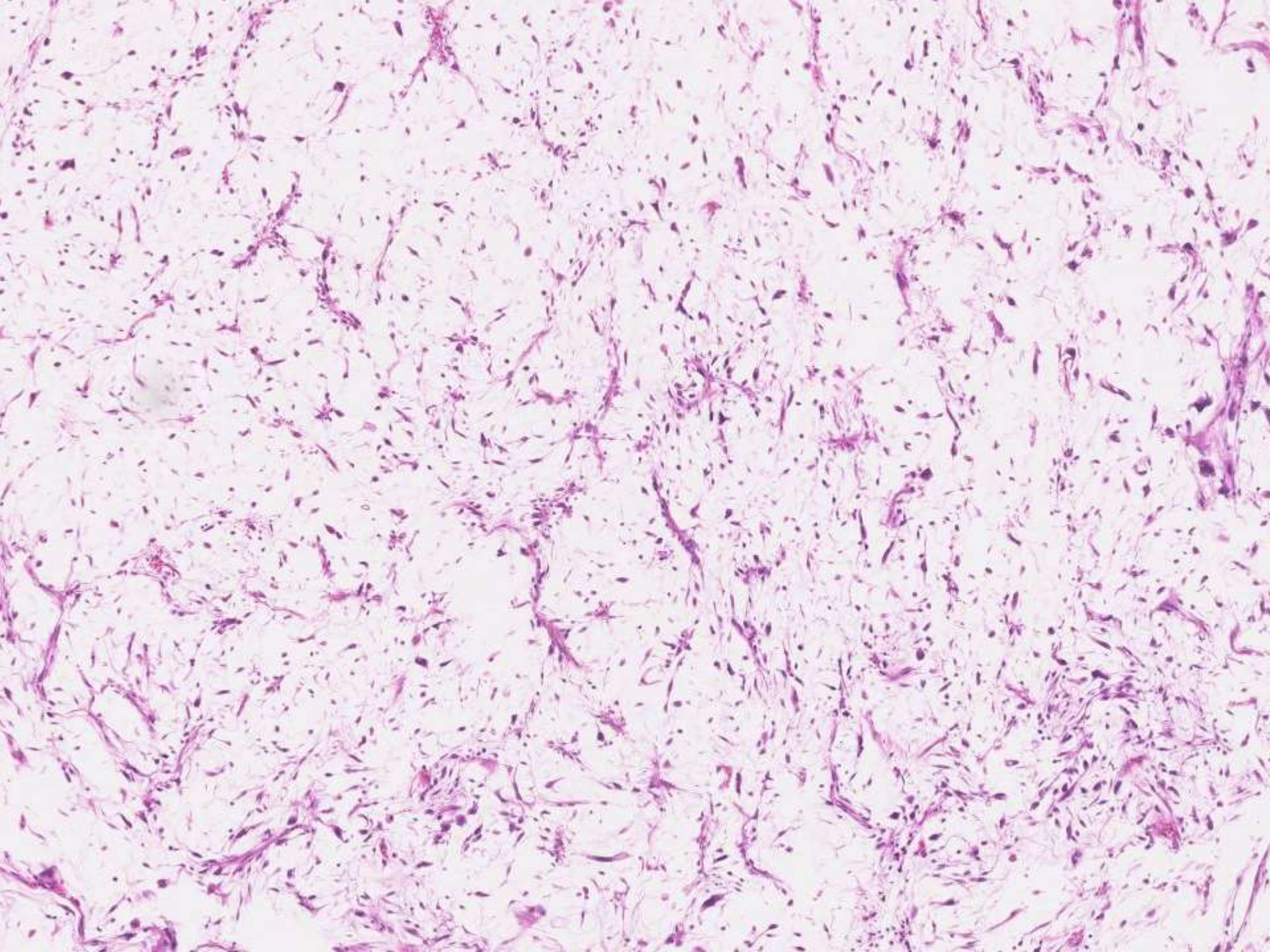
1. Low grade fibromyxoid sarcoma
2. (low grade) myxofibrosarcoma
3. Low grade myofibroblastic sarcoma


87F, scapular region

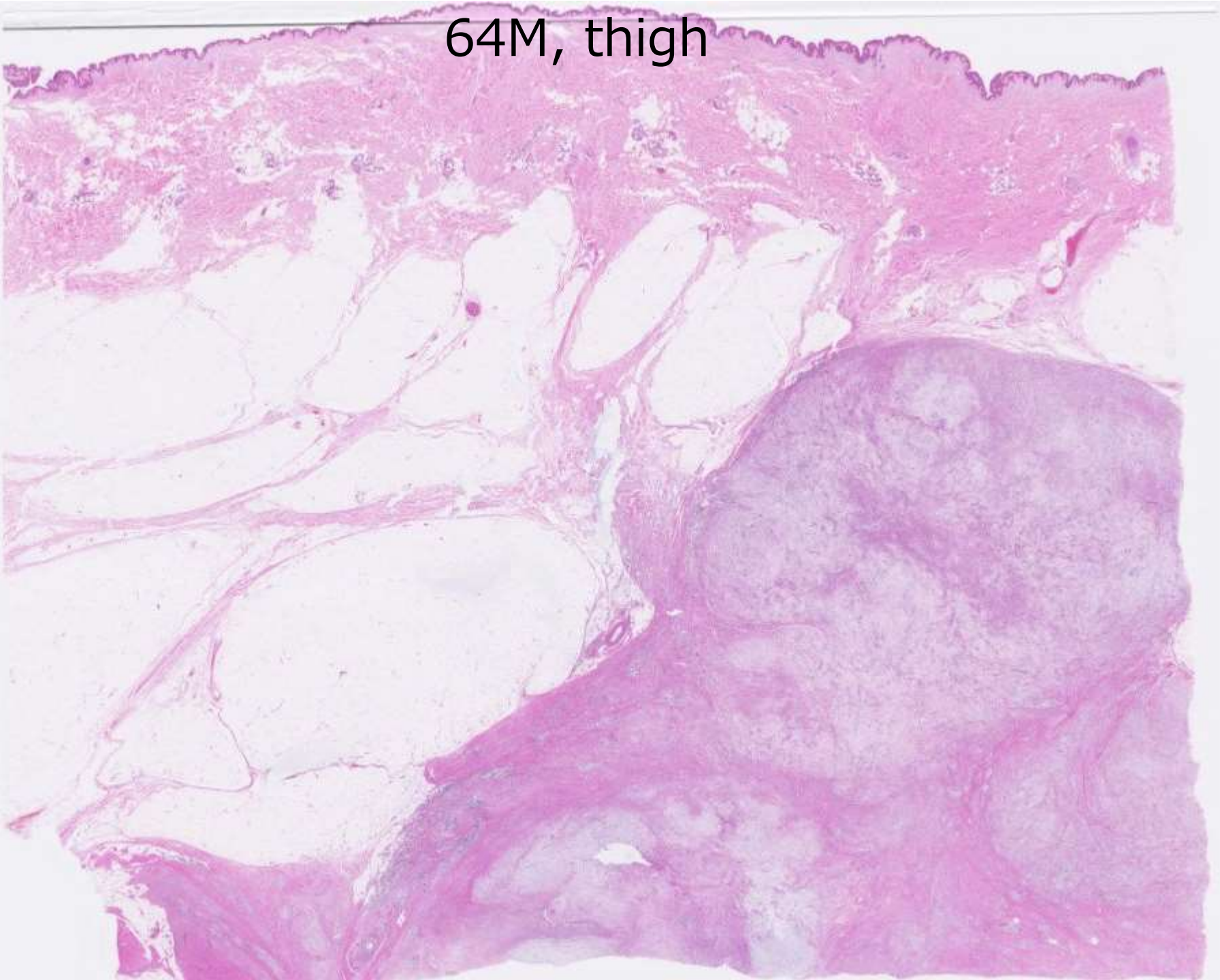


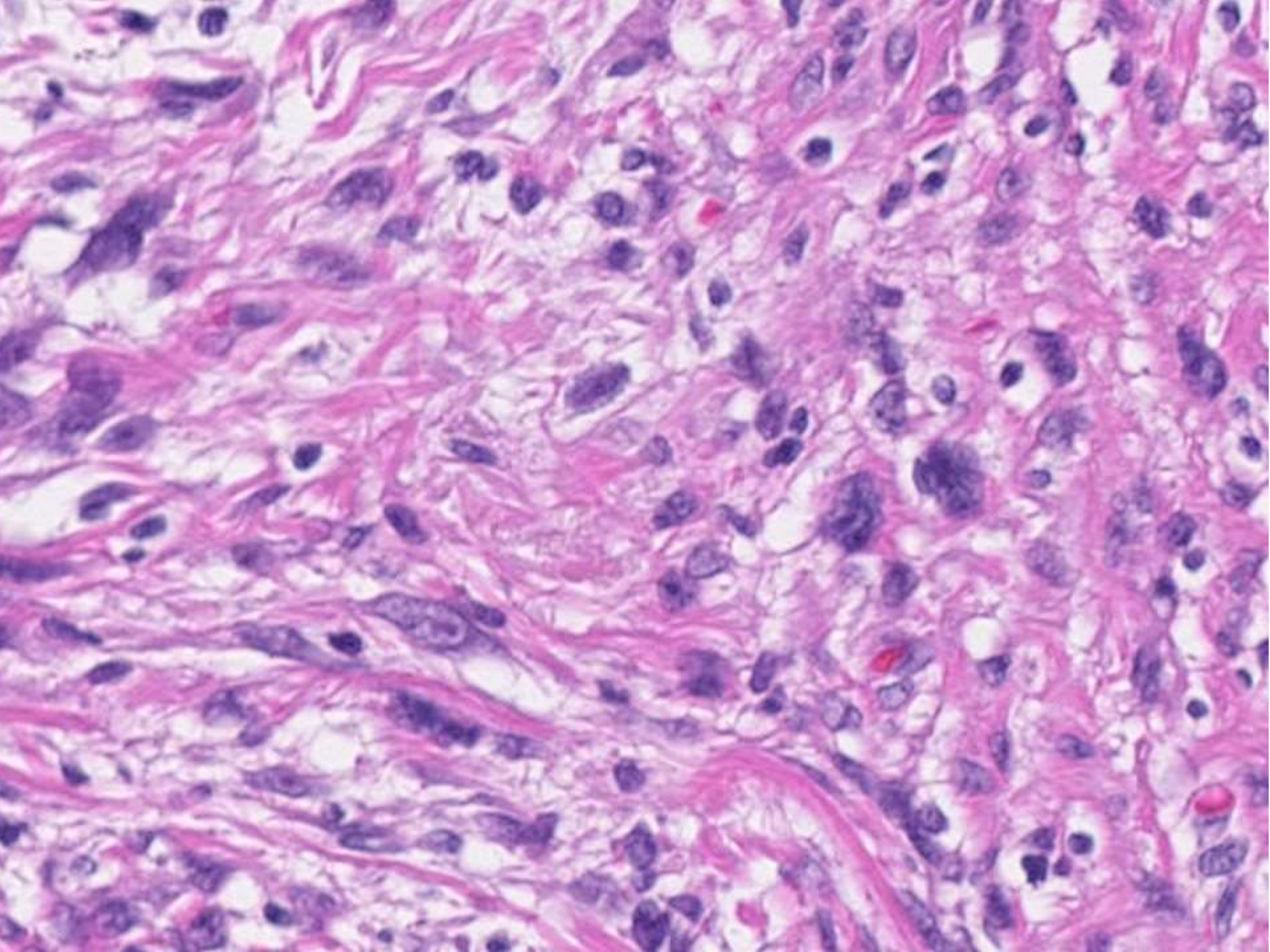


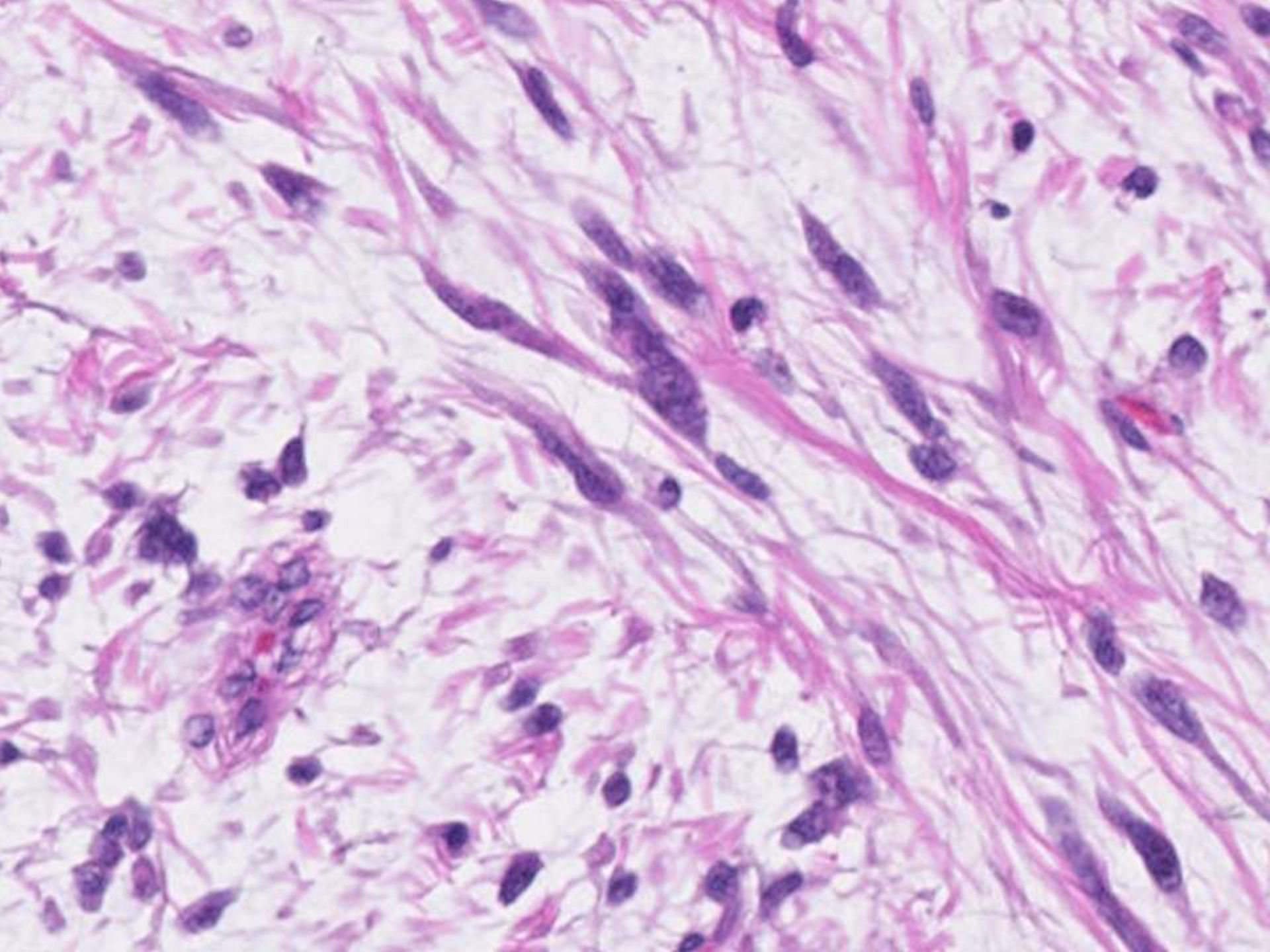


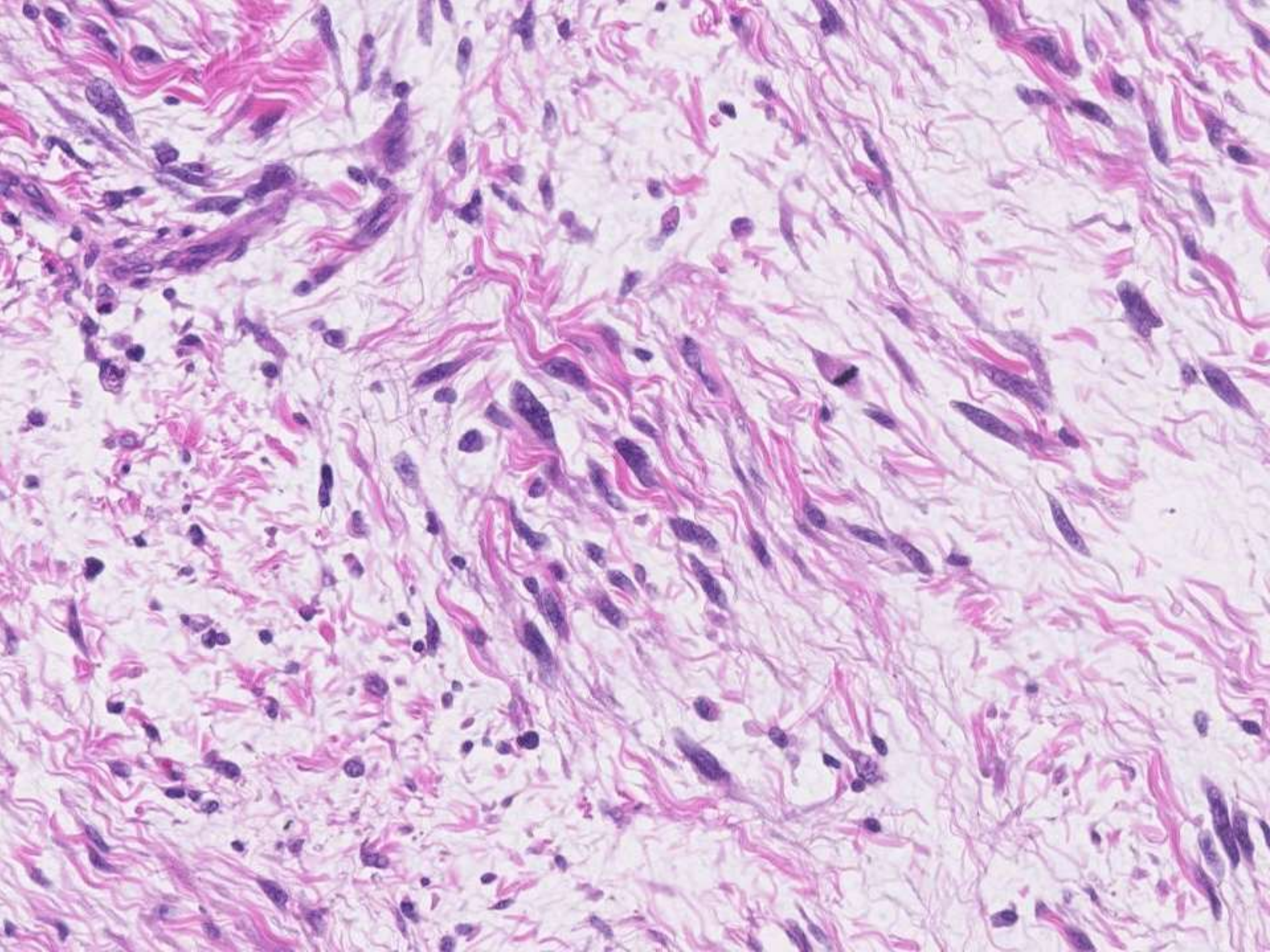


64M, thigh

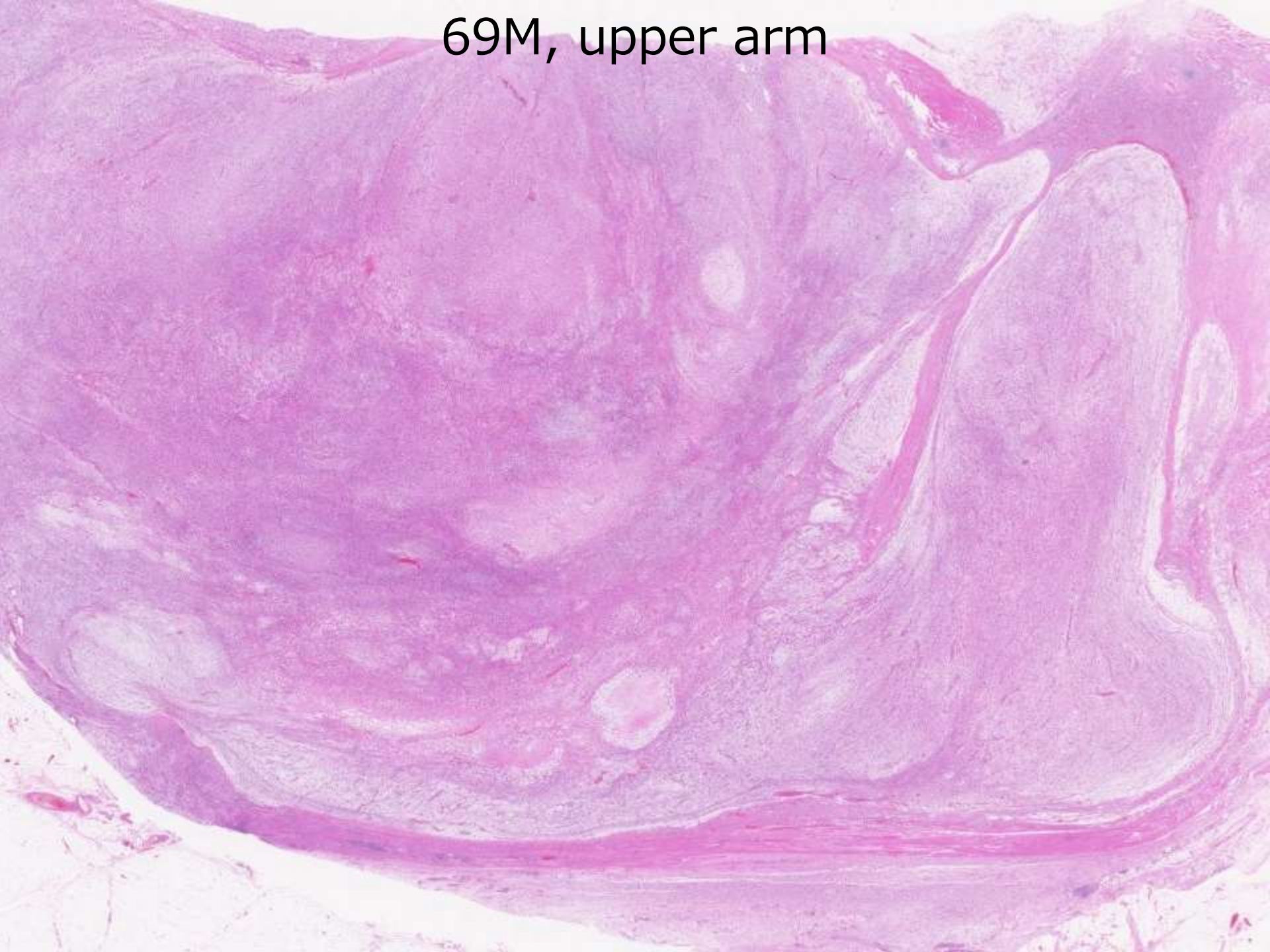


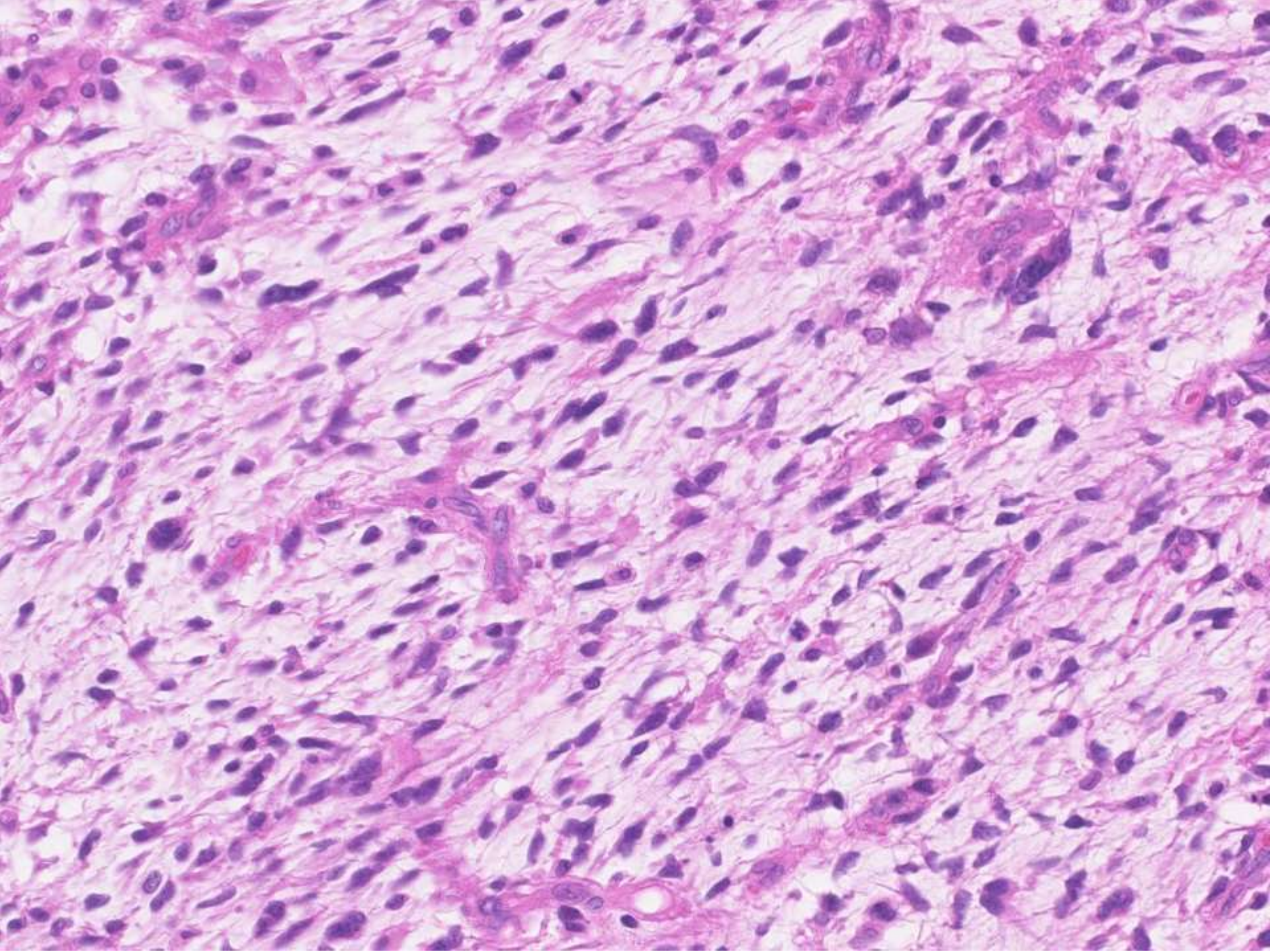


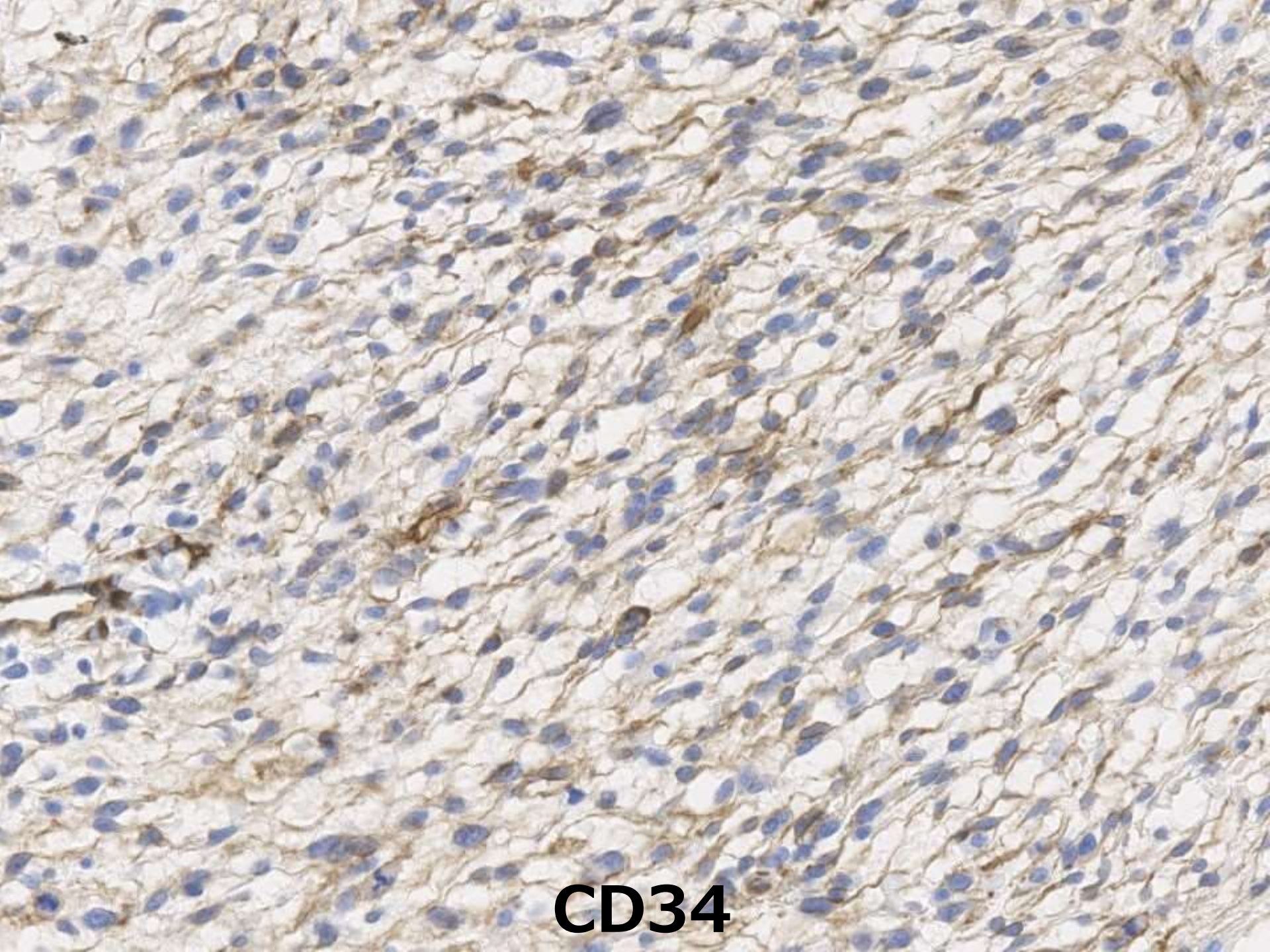




69M, upper arm






CD34

Tips 3

- Myxofibrosarcomaの粘液腫状間質は部分的には高度なものが多い（ズブズブになる）
- Myxofibrosarcomaは多形性が重要で、多核の異型細胞は指標になる
- 免役染色は他の腫瘍の否定

異型に乏しい紡錘形細胞腫瘍でしばしば鑑別に挙げられる肉腫

1. Low grade fibromyxoid sarcoma
2. (low grade) myxofibrosarcoma
3. Low grade myofibroblastic sarcoma

Macroscopic appearance:-

Grossly, the tumour is usually a firm mass with pale, fibrous cut surfaces and ill-defined margins { 9777985 }; a minority are well circumscribed with pushing margins { 11176071 }.

Histopathology:-

Histologically, low-grade myofibroblastic sarcomas are characterized by a diffusely infiltrative growth pattern, and (in deeply located neoplasms) tumour cells often grow between individual skeletal muscle fibres. Most cases are composed of spindle-shaped tumour cells arranged in cellular fascicles or show a storiform growth pattern. Neoplastic cells have ill-defined palely eosinophilic cytoplasm and fusiform nuclei that are either elongated and wavy with evenly distributed chromatin or plumper, more rounded, and vesicular with small nucleoli. Rarely, hypocellular neoplasms with a more prominent collagenous (sometimes hyalinized) stroma have been described. Importantly, neoplastic cells show, at least focally, moderate nuclear atypia with enlarged, hyperchromatic, and irregular nuclei and slightly increased proliferative activity. These neoplasms may contain numerous thin-walled capillaries. The tumours may progress to morphologically higher-grade myofibroblastic sarcomas { 11176071 }. By immunohistochemistry, neoplastic cells in low-grade myofibroblastic sarcoma show variable positivity for SMA and/or desmin. A subset show nuclear β-catenin staining { 17711447 }.

Cytology:-

Not clinical

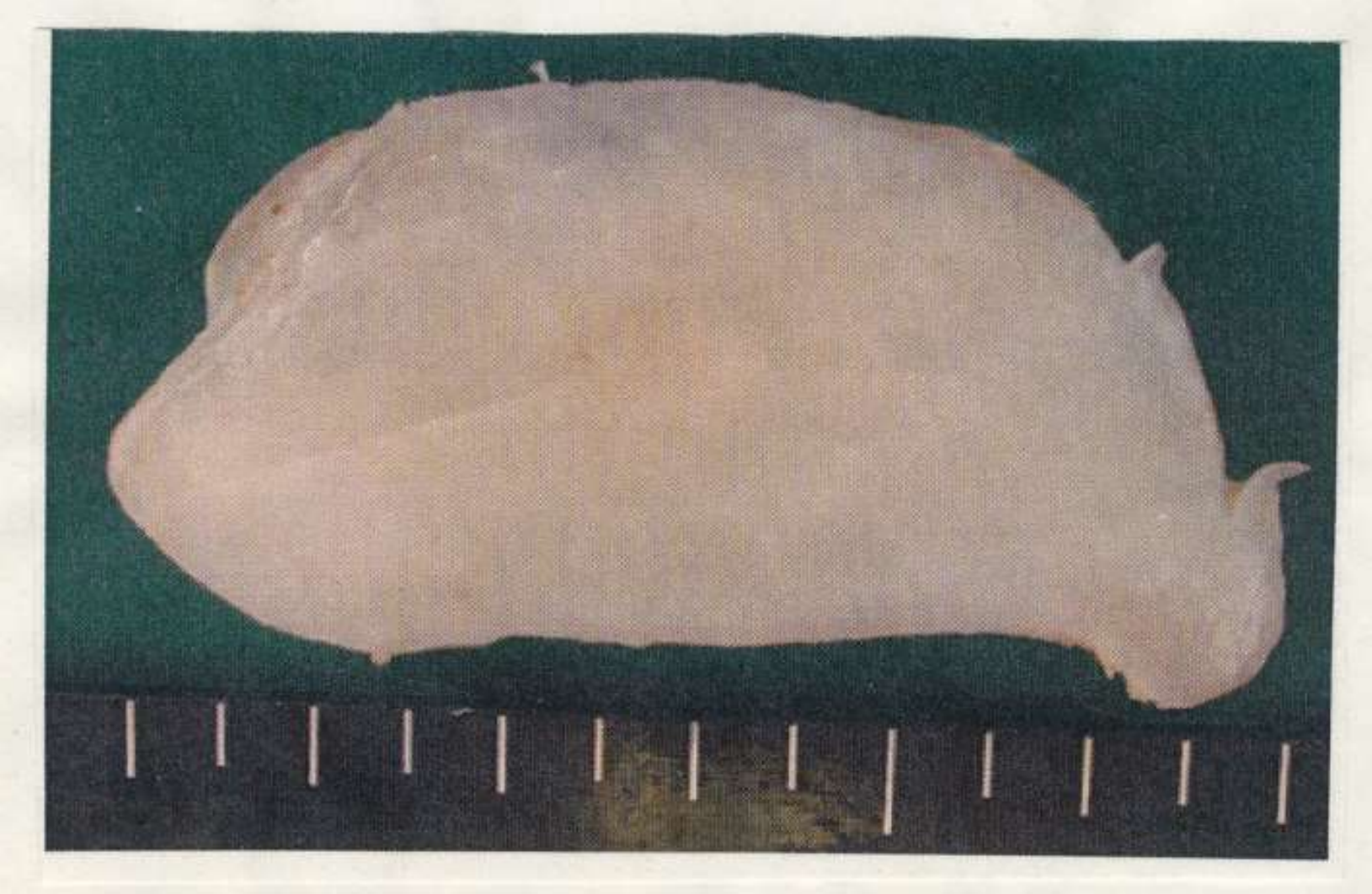
Diagnosti

Not clinical

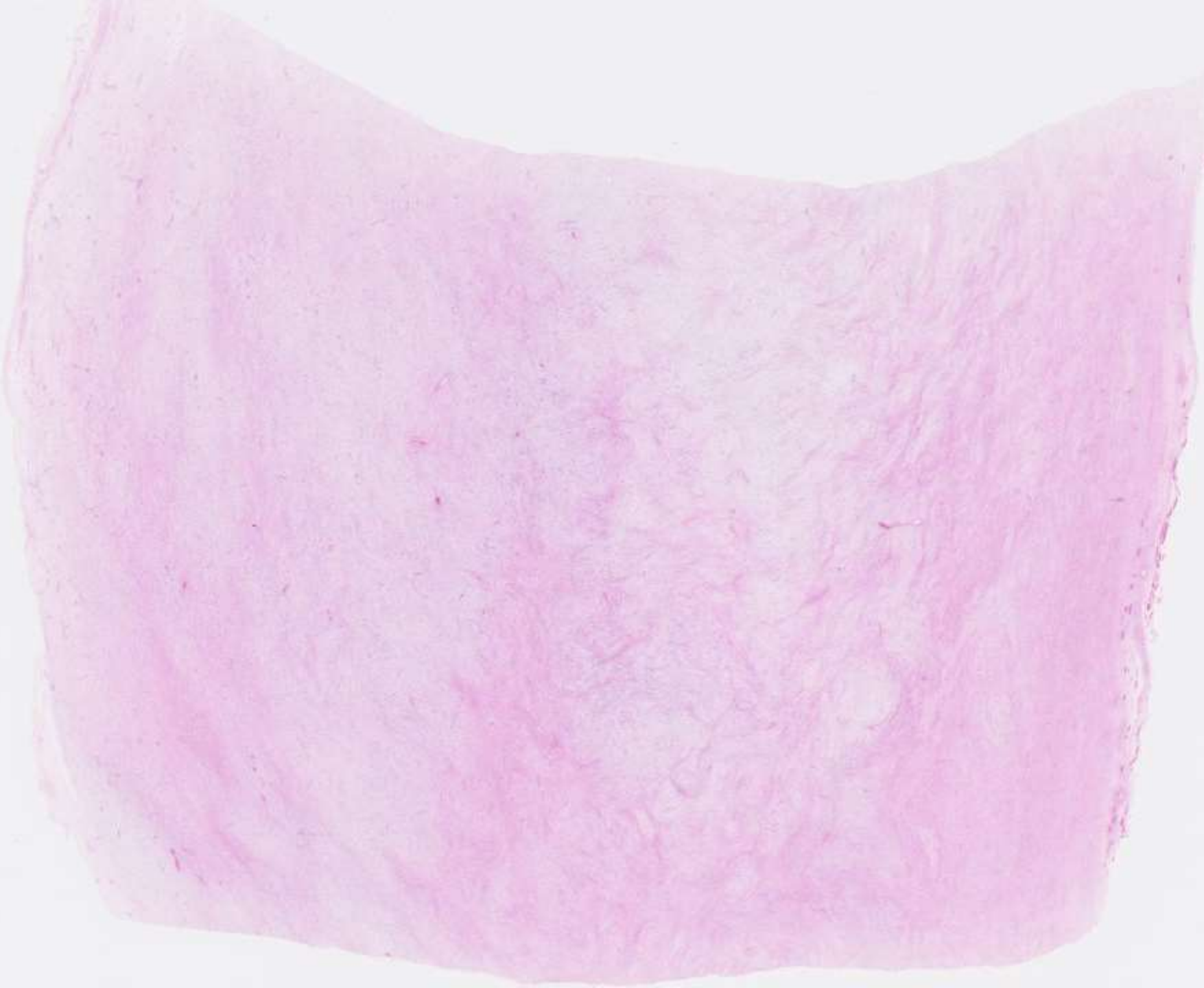
- 典型像はない
- 腫瘍細胞のほとんどがSMA陽性
- 他の疾患概念に当てはまらない筋線維芽細胞分化を示す肉腫

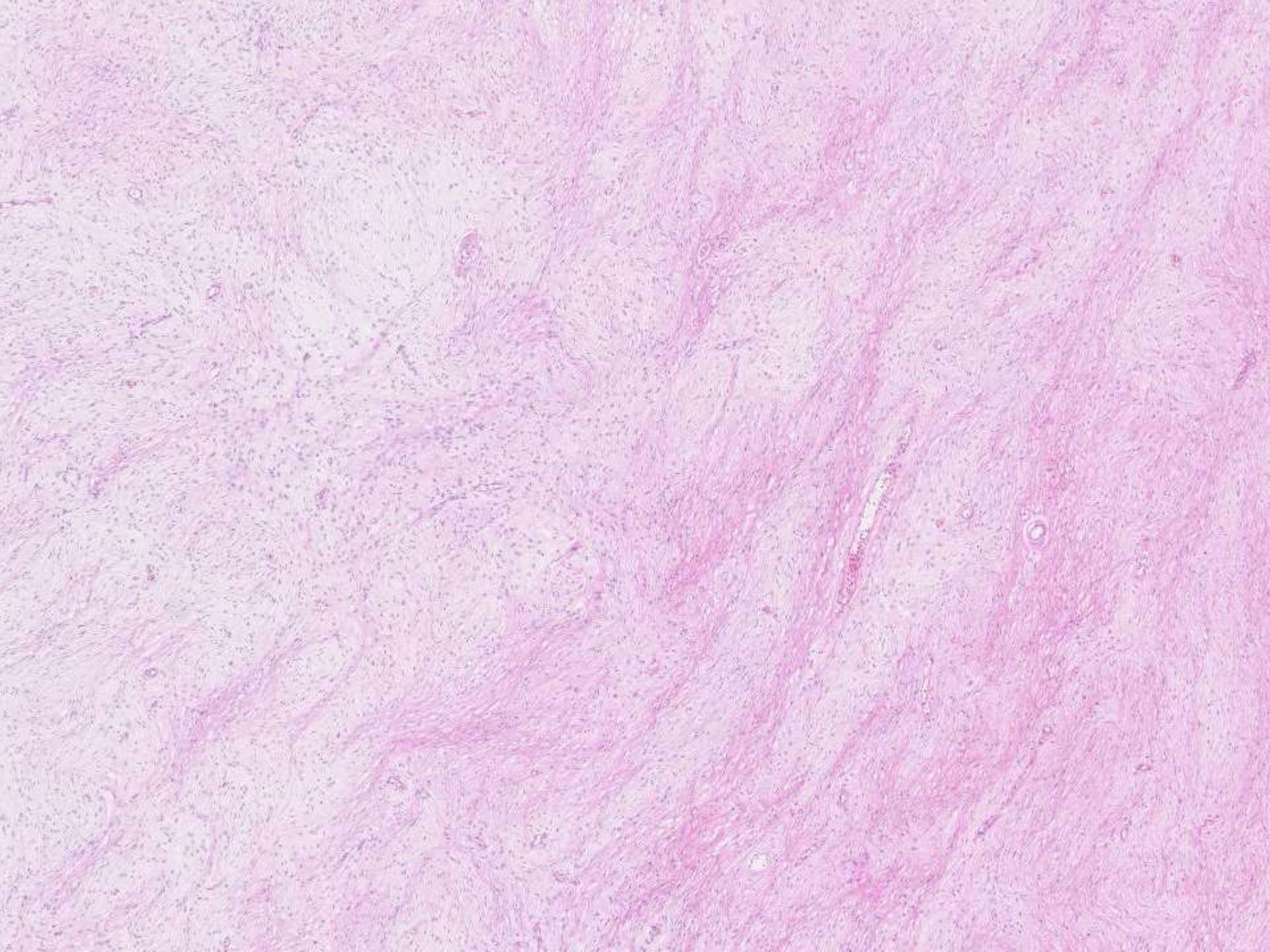
Essential and desirable diagnostic criteria:-

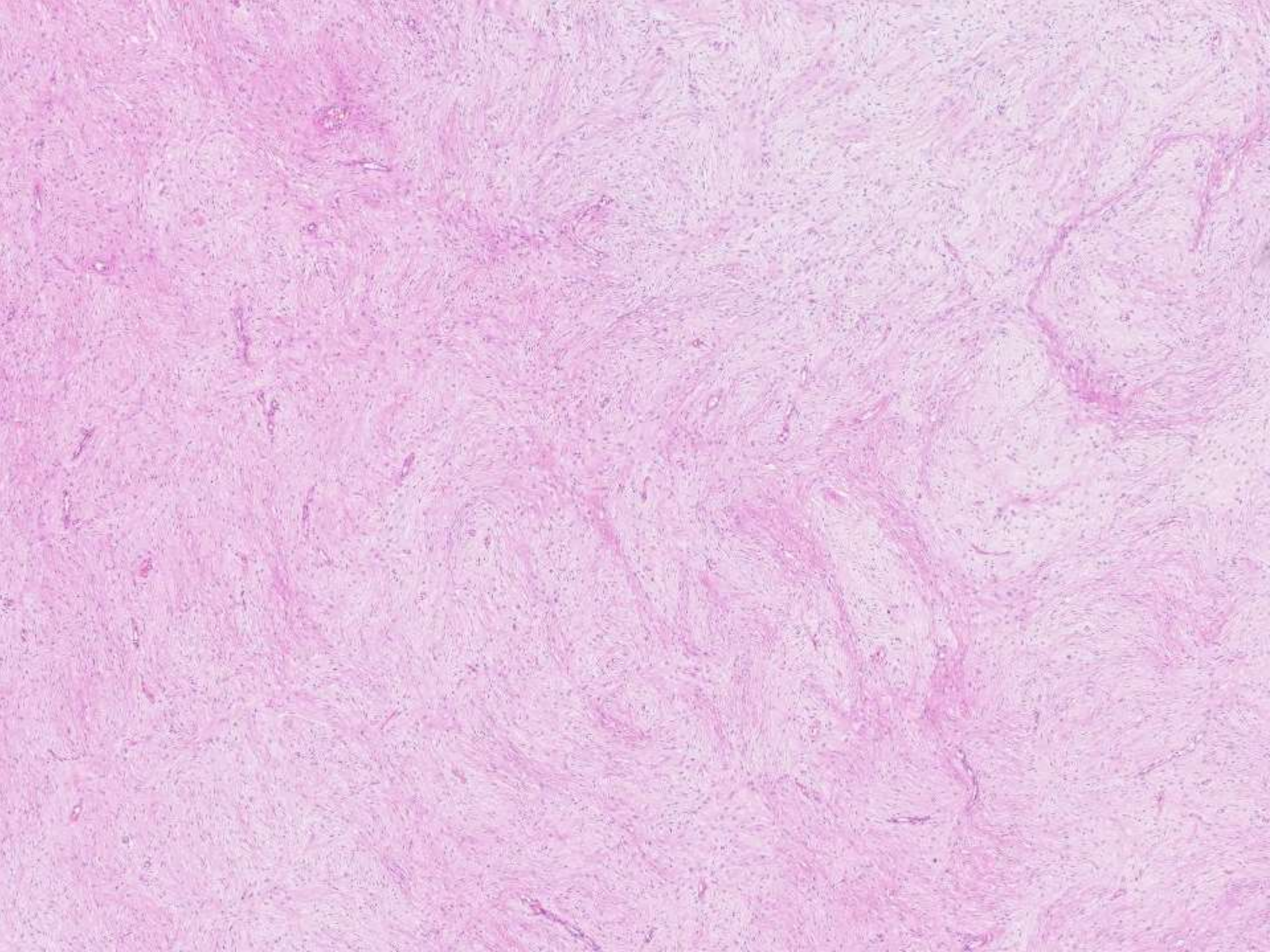
Case 2: 30F, back

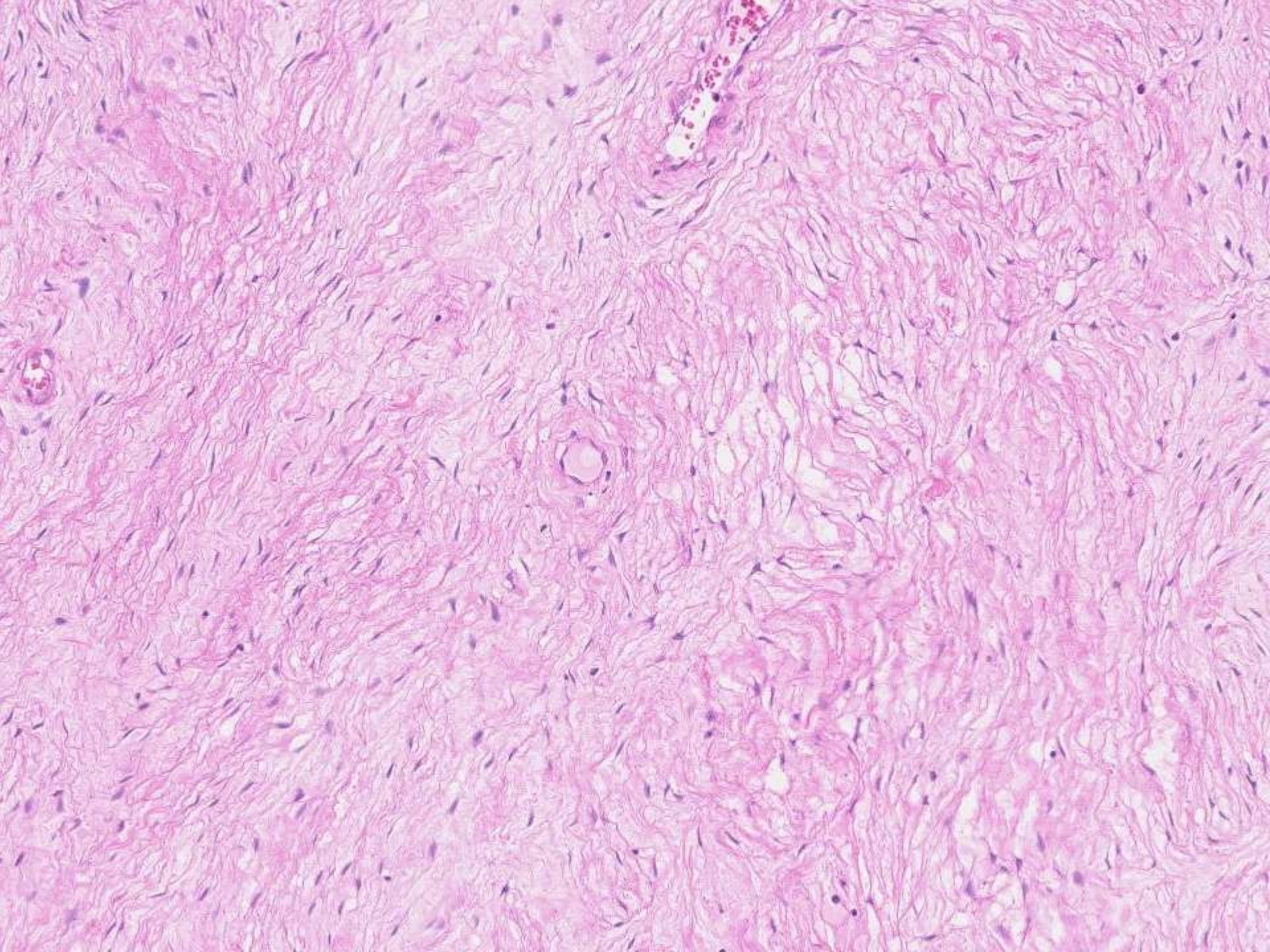


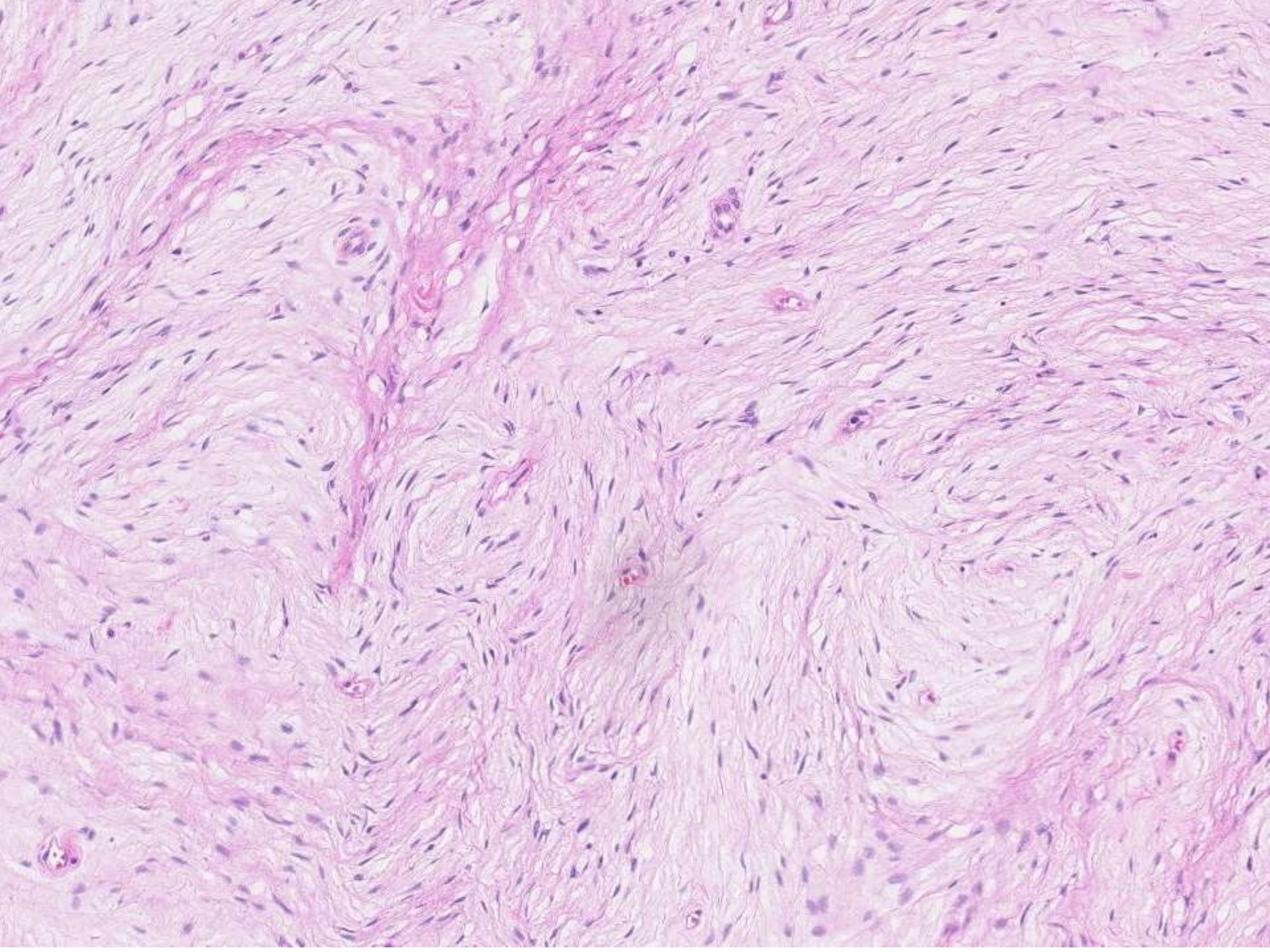
3年前に気づく。徐々に増大。広背筋内の7 cmの腫瘍

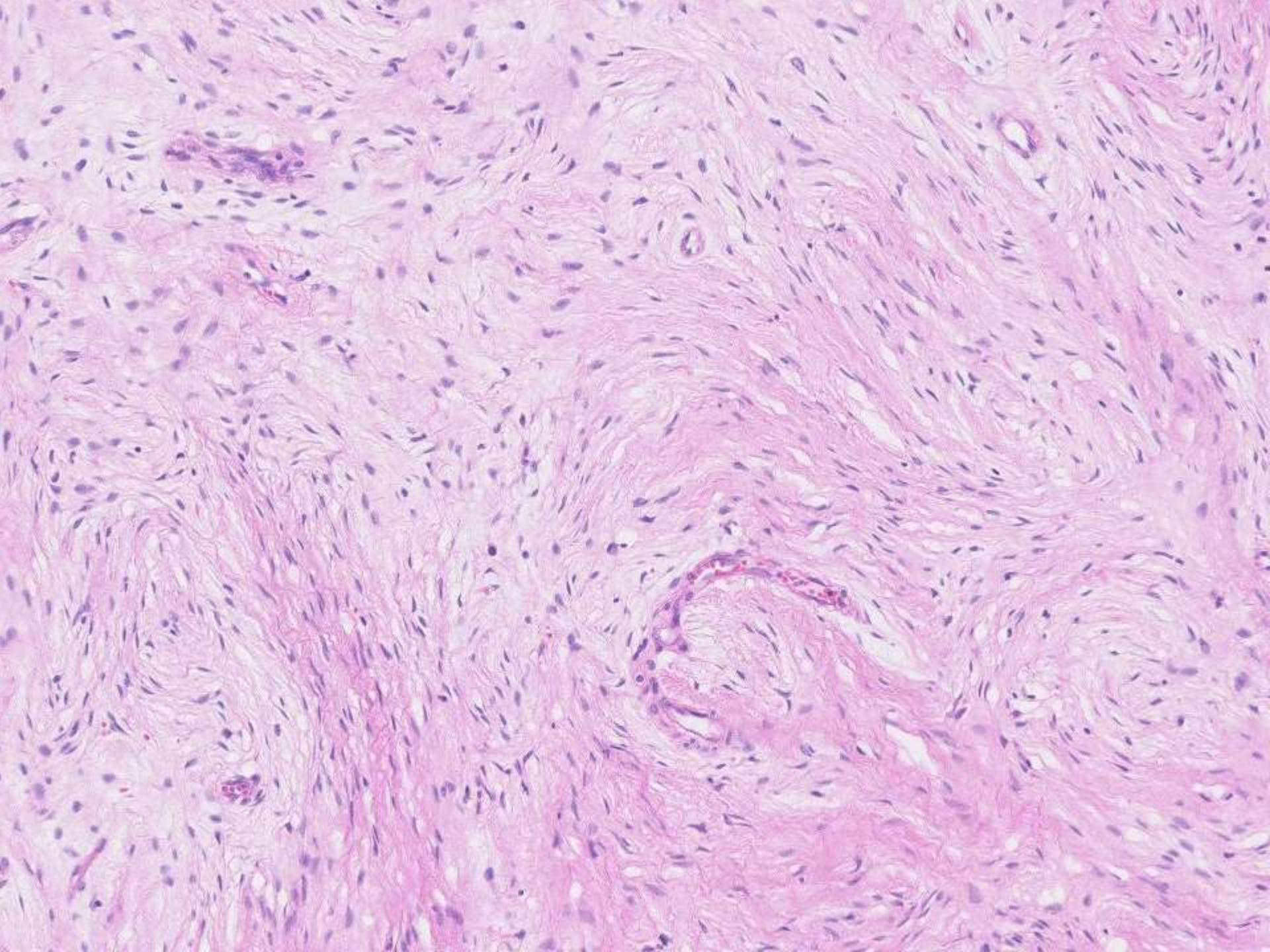


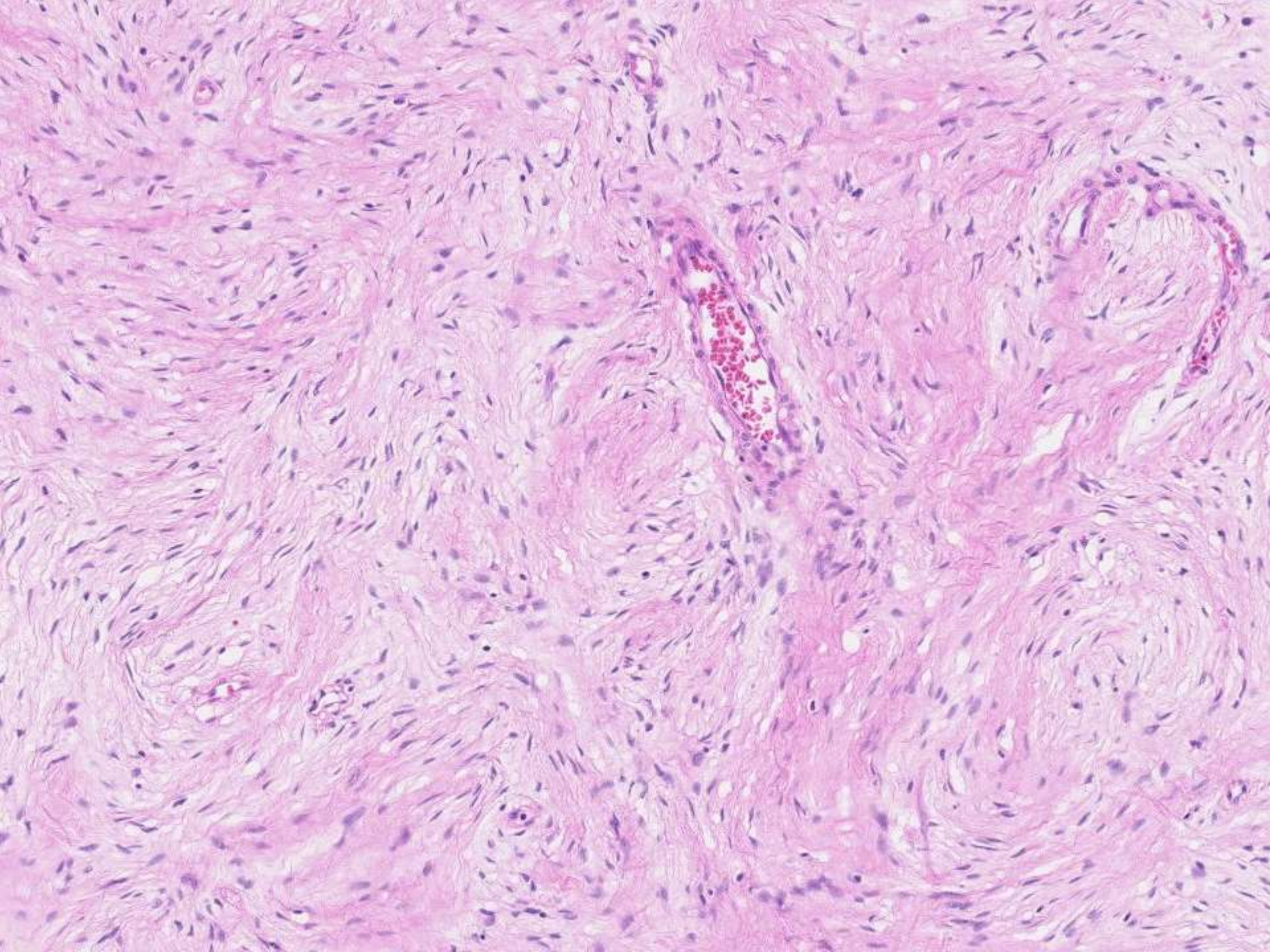


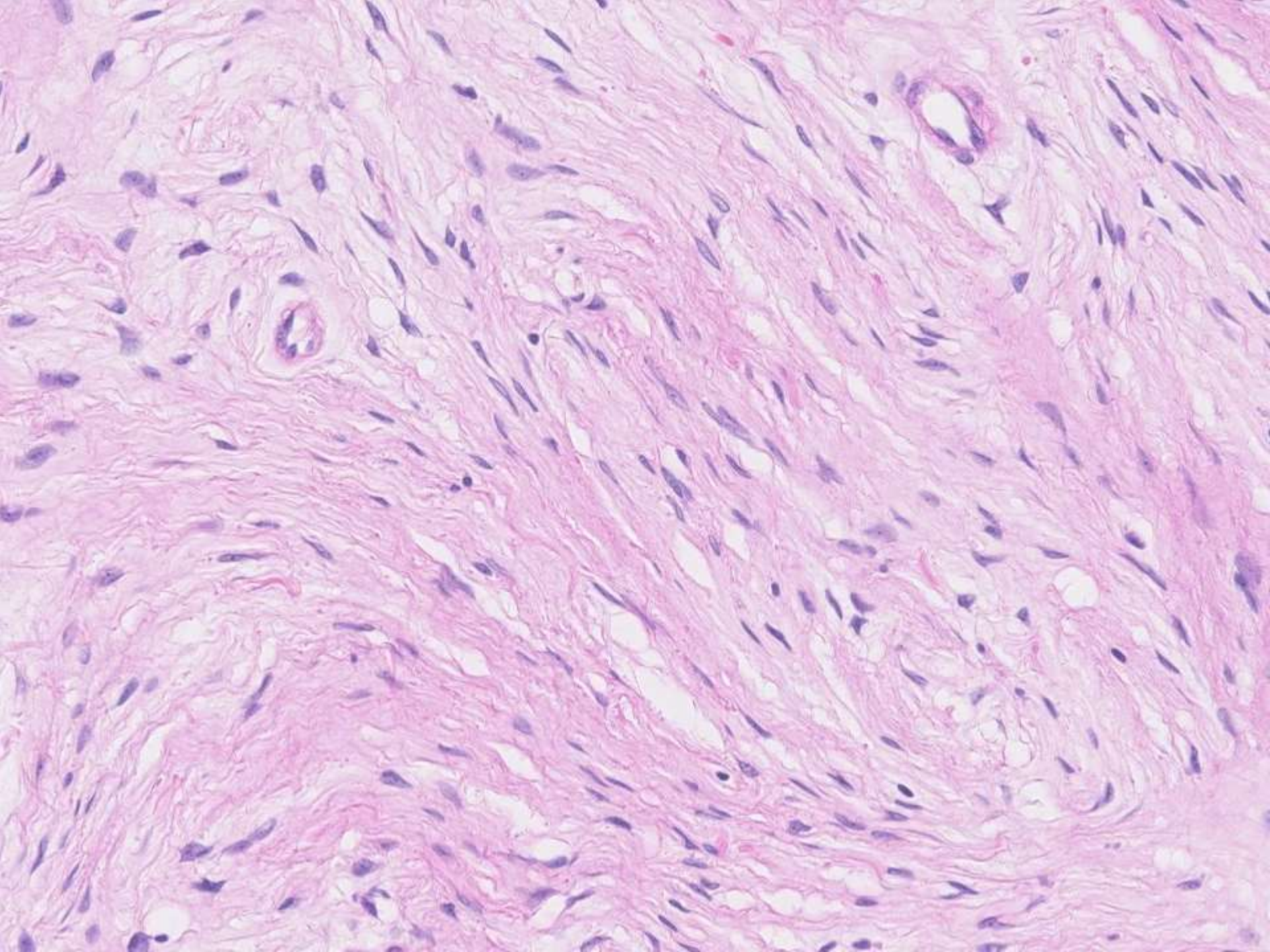


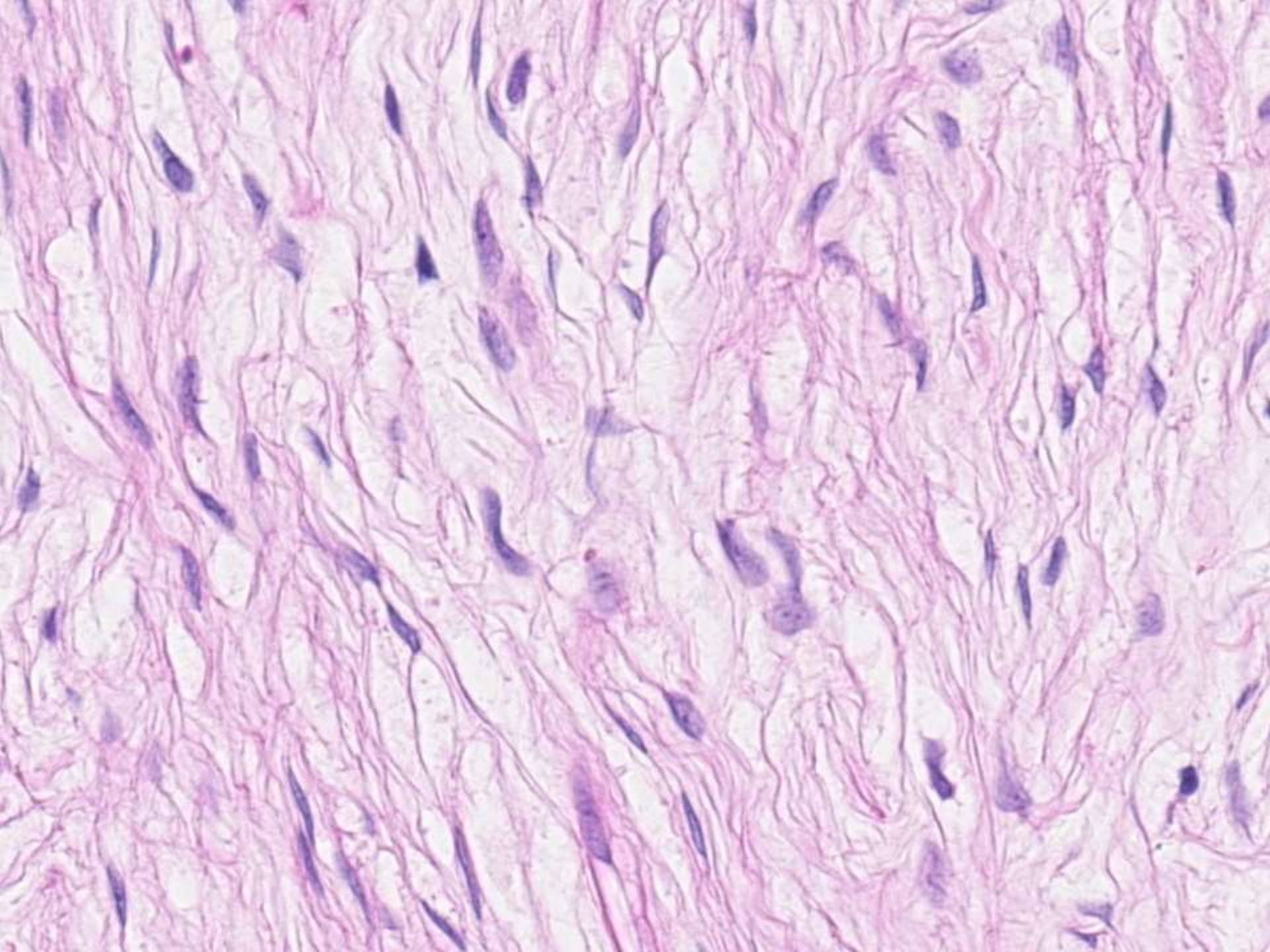




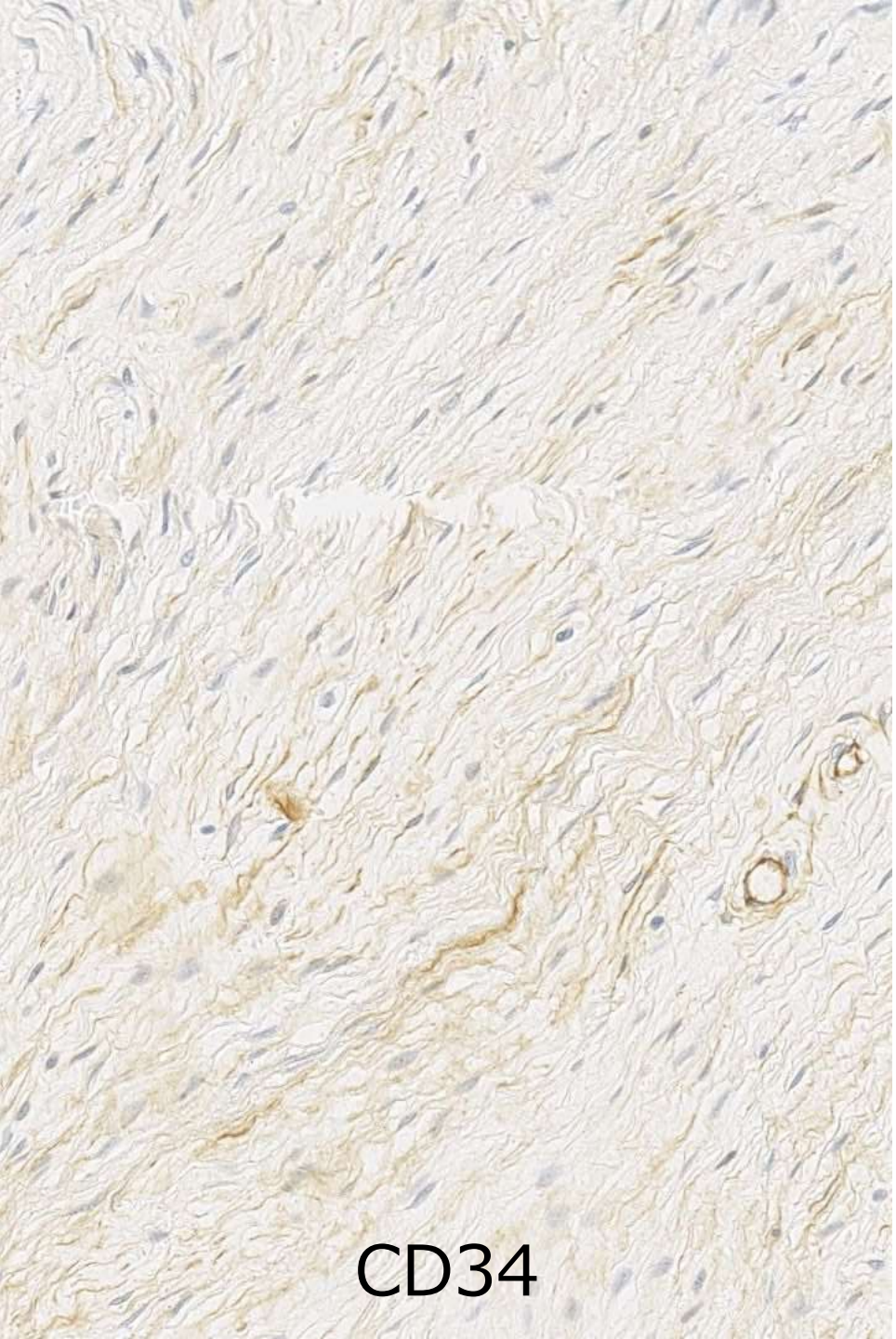


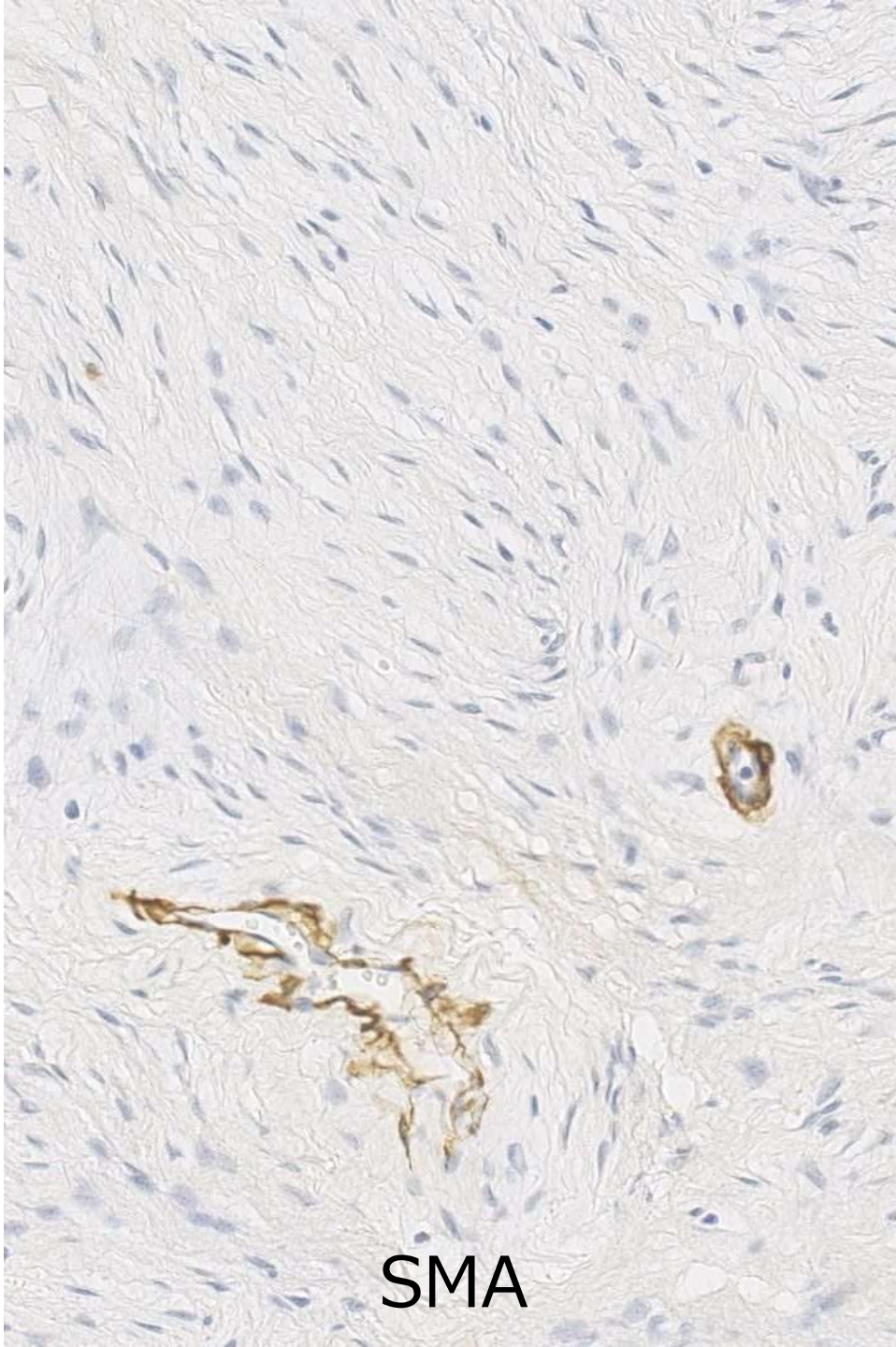


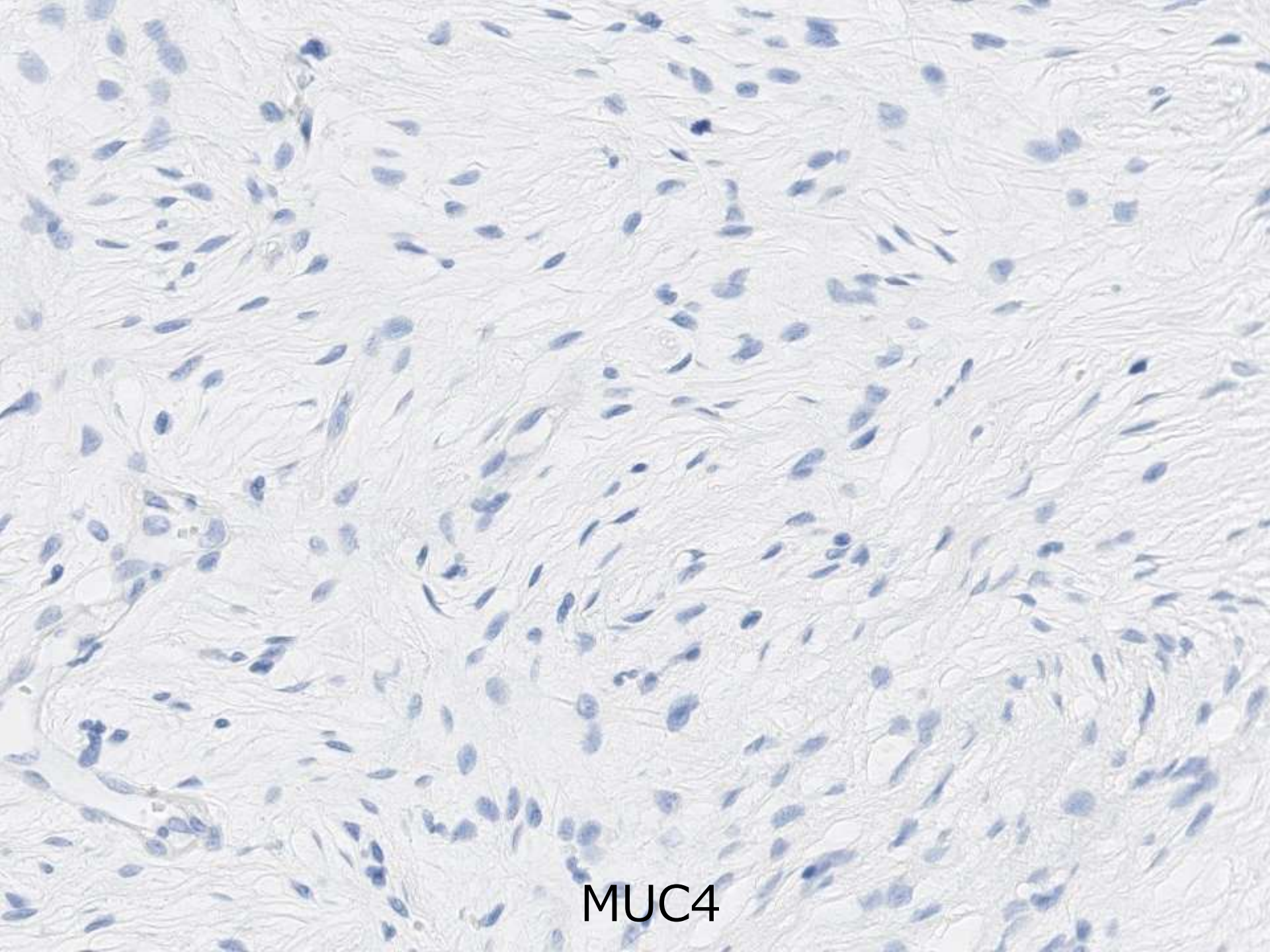




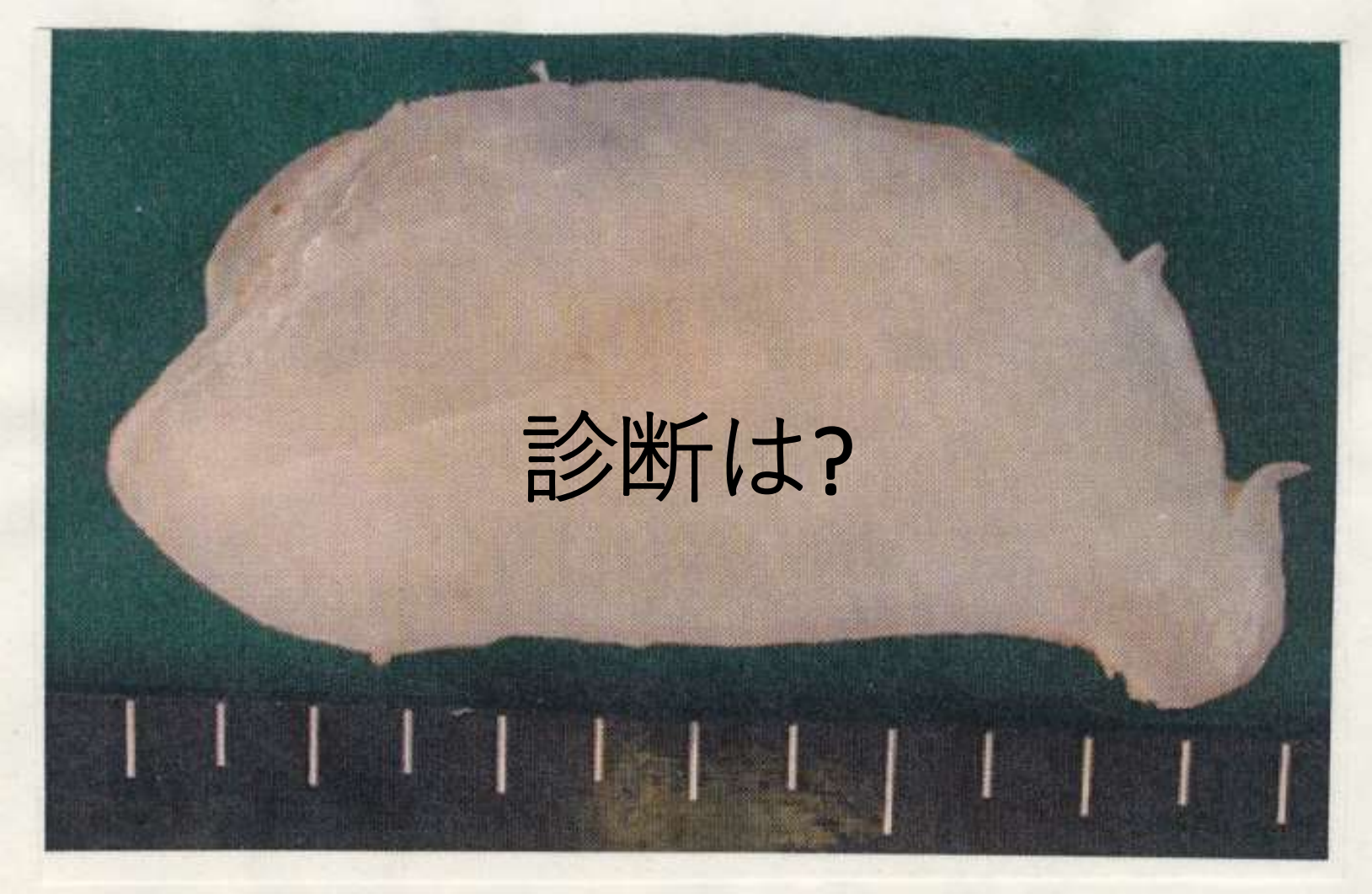

EMA


CD34


SMA


MUC4

Case 2: 30F, back

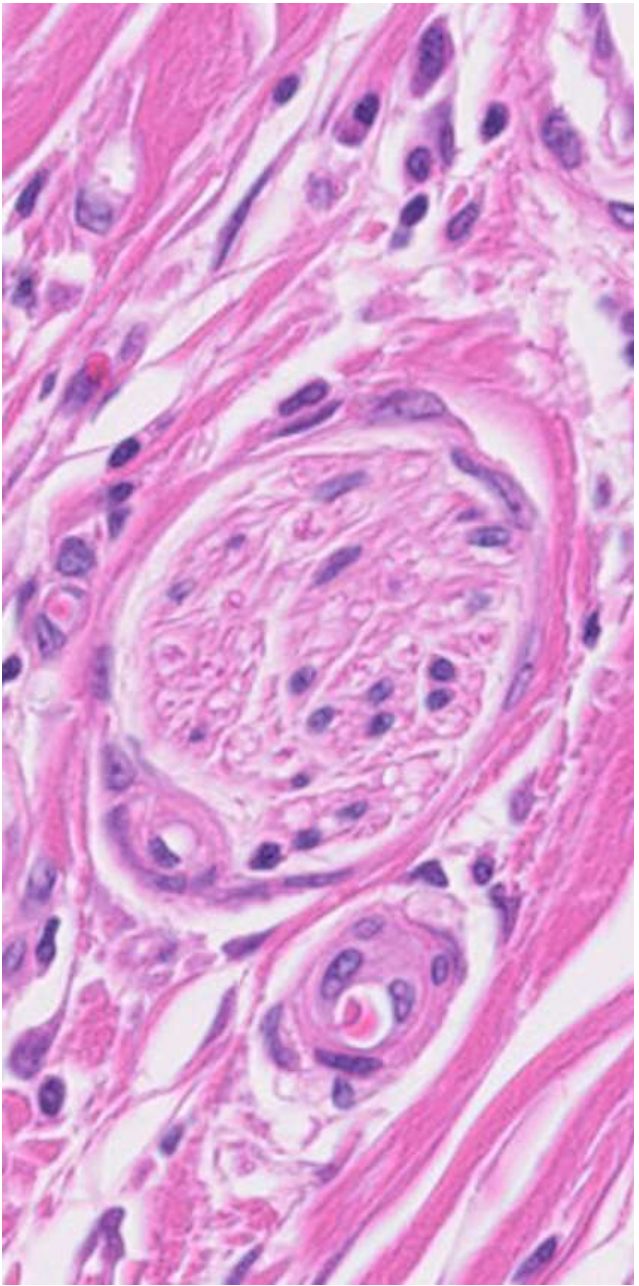


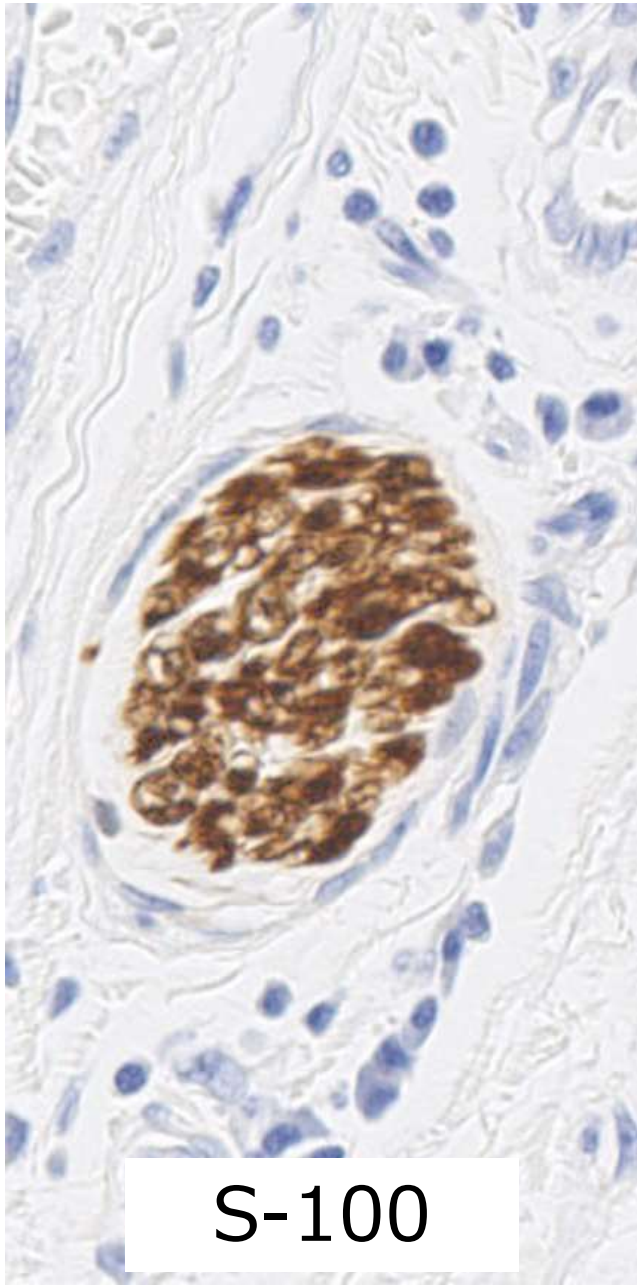
3年前に気づく。徐々に増大。広背筋内の7 cmの腫瘍

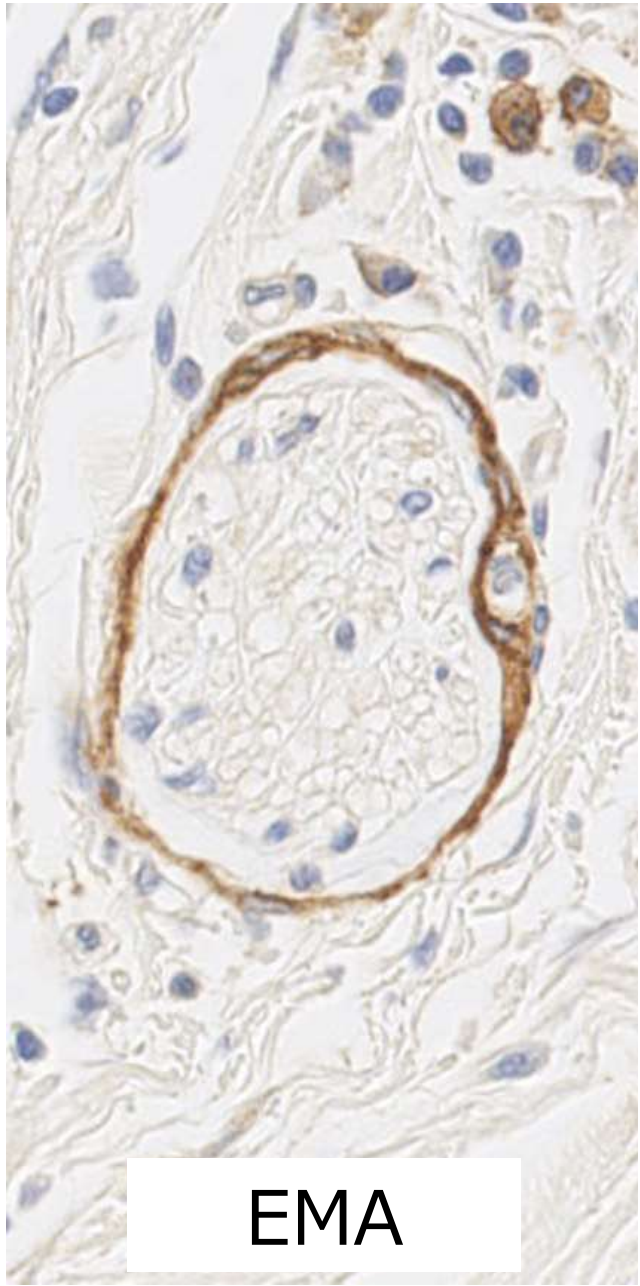
Diagnosis

Perineurioma of soft tissue

神経周膜細胞 Perineurial cell

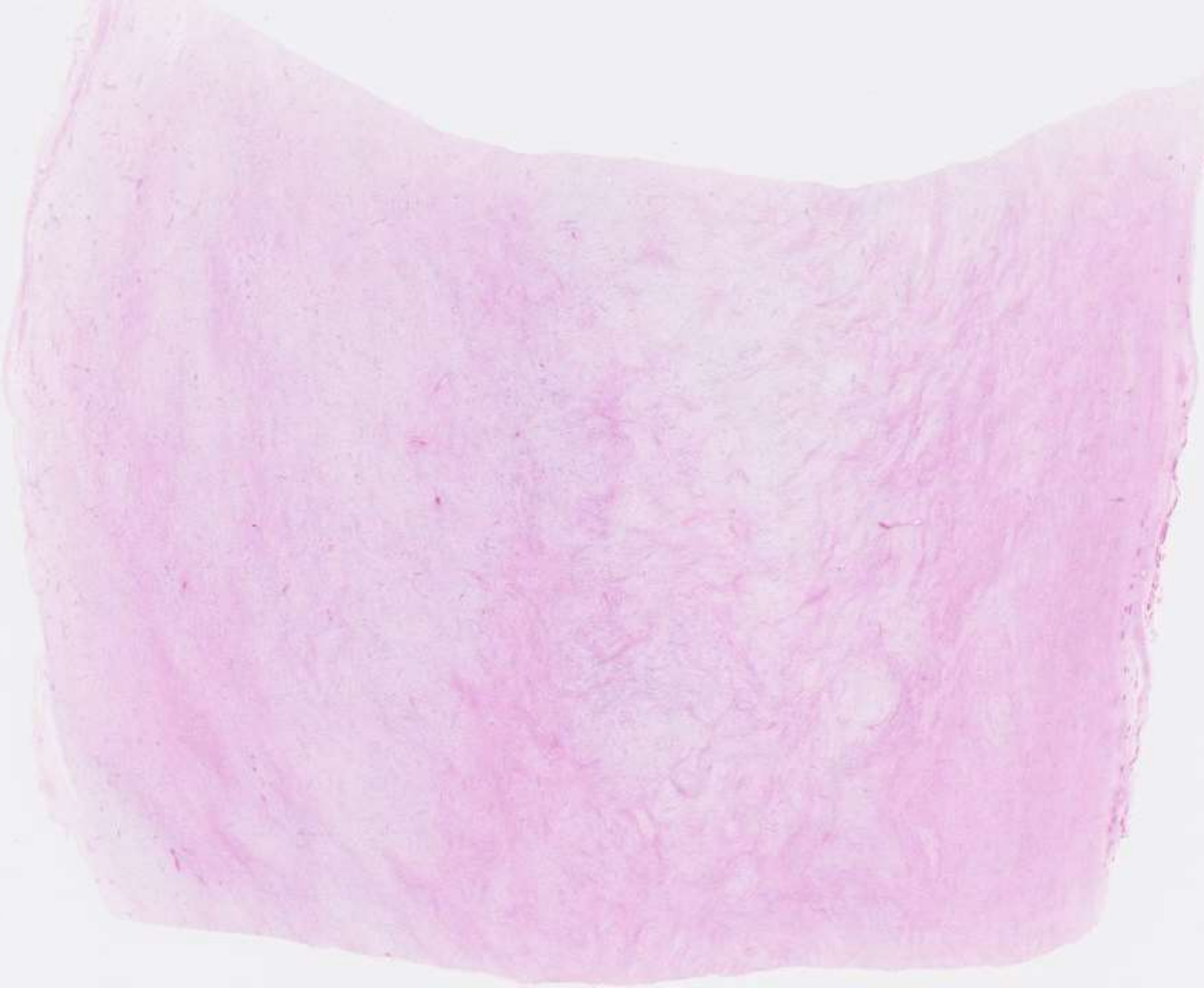


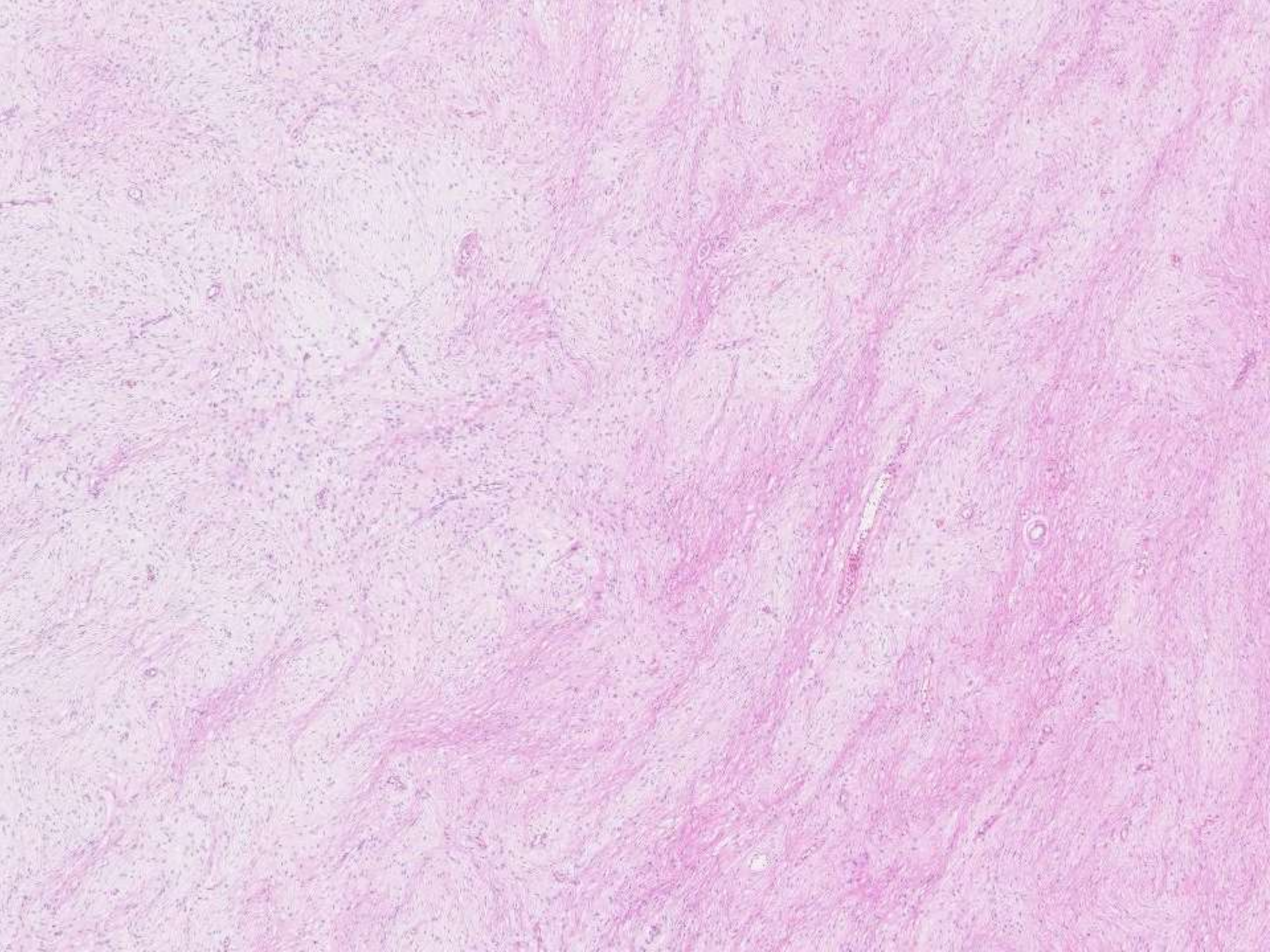


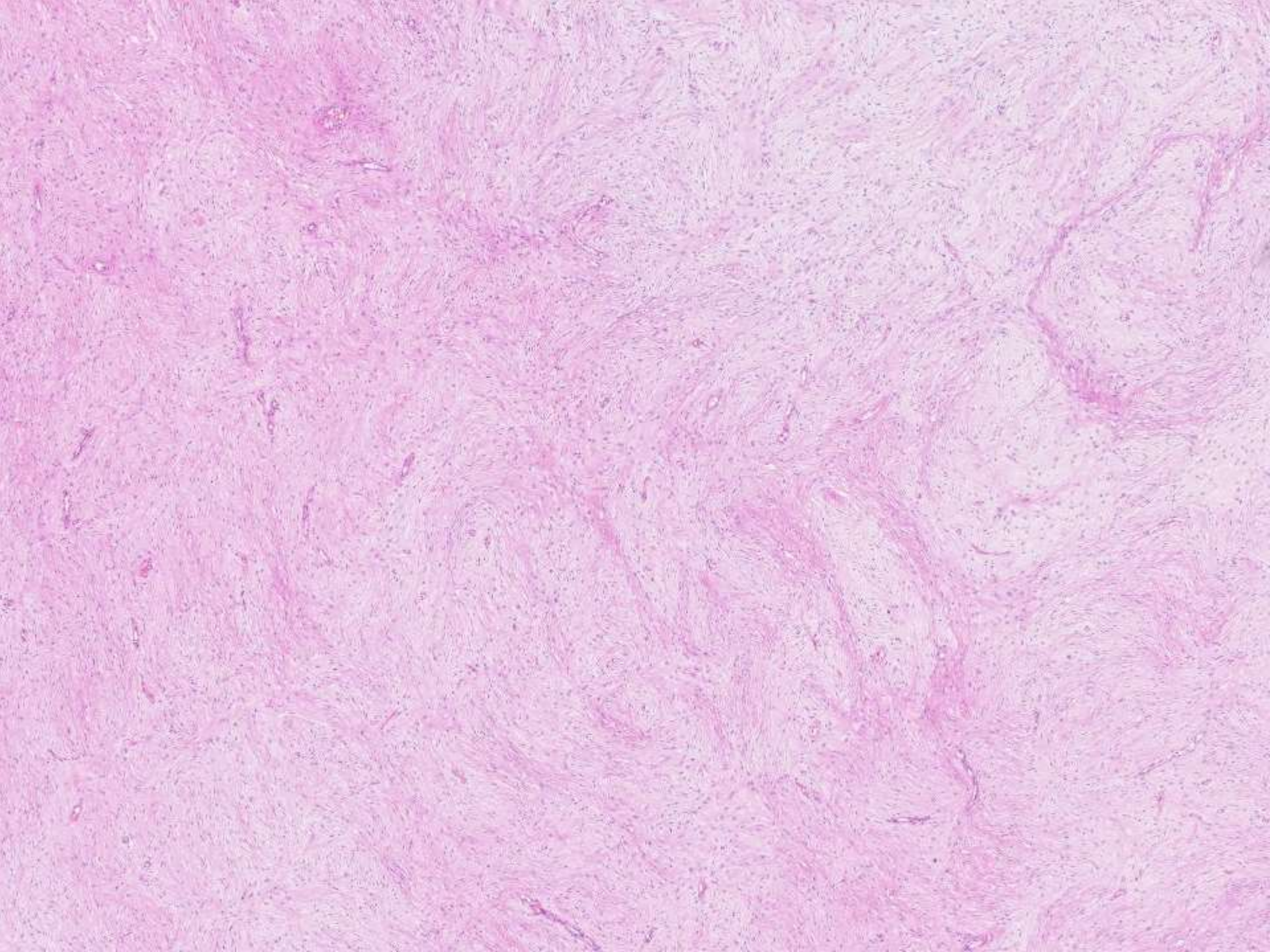


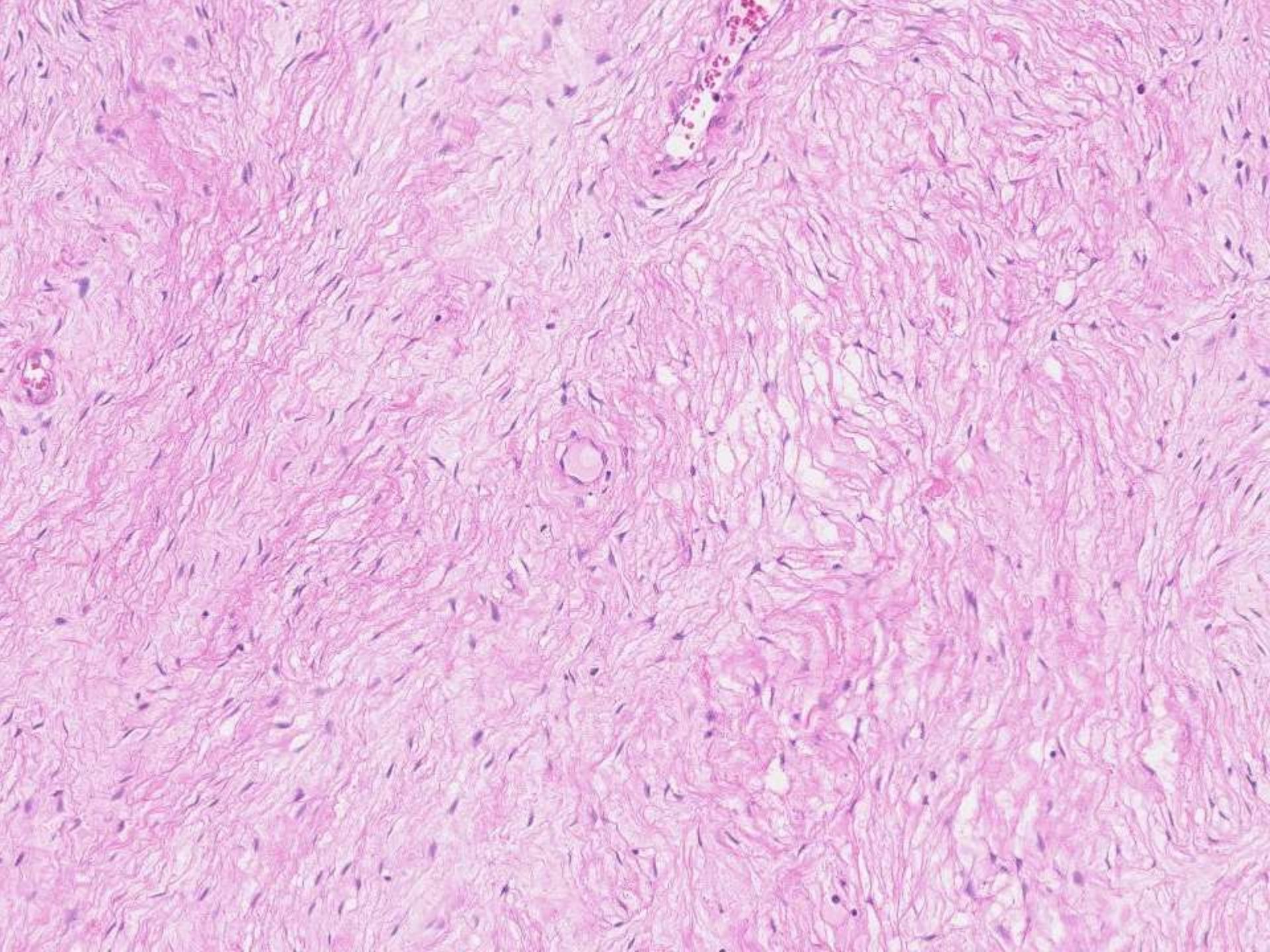
Perineurioma

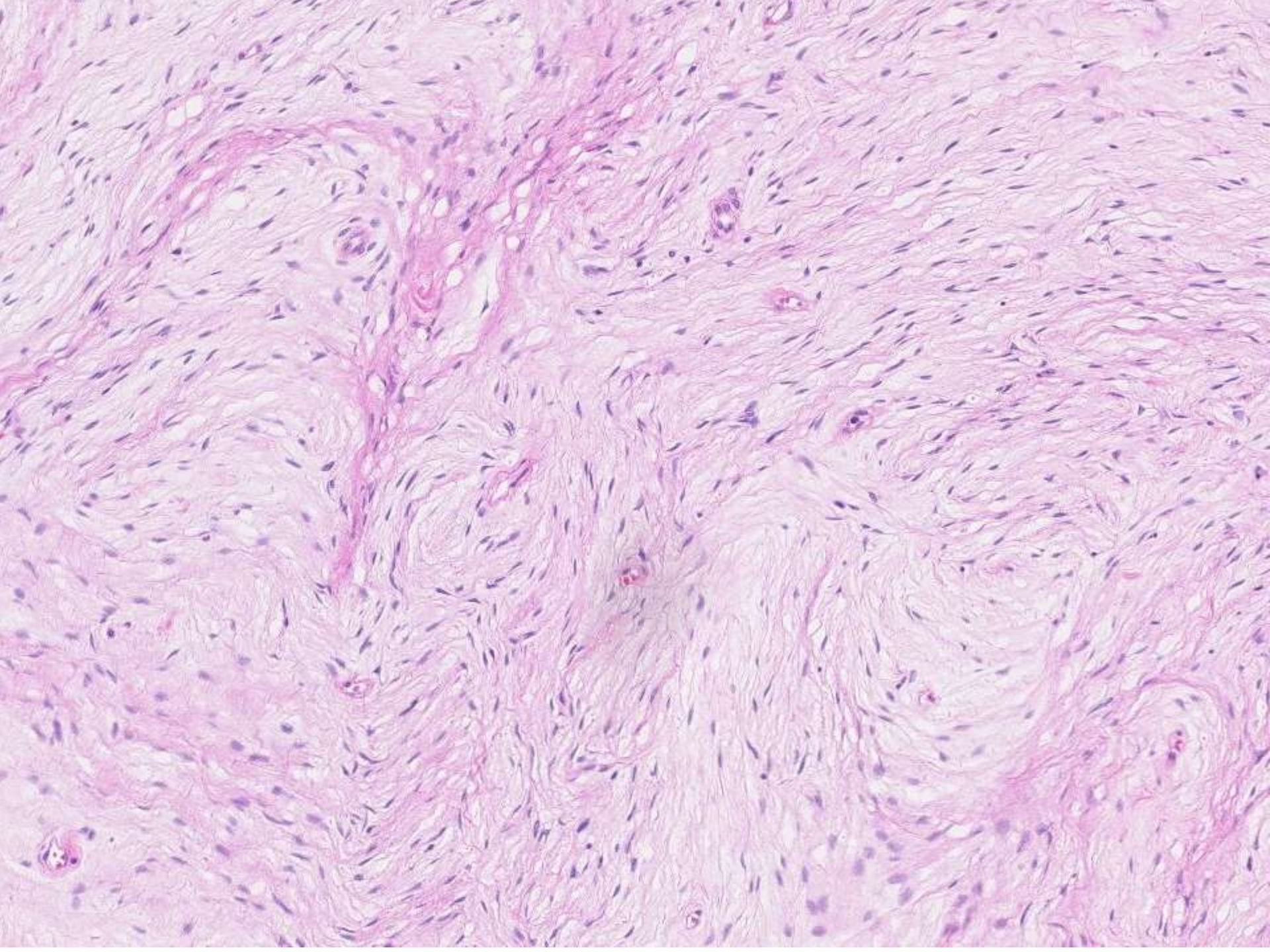
- Intraneural perineurioma
- Soft tissue (extraneural) perineurioma
  - (Storiform perineurial fibroma)
  - Reticular perineurioma
  - Sclerosing perineurioma

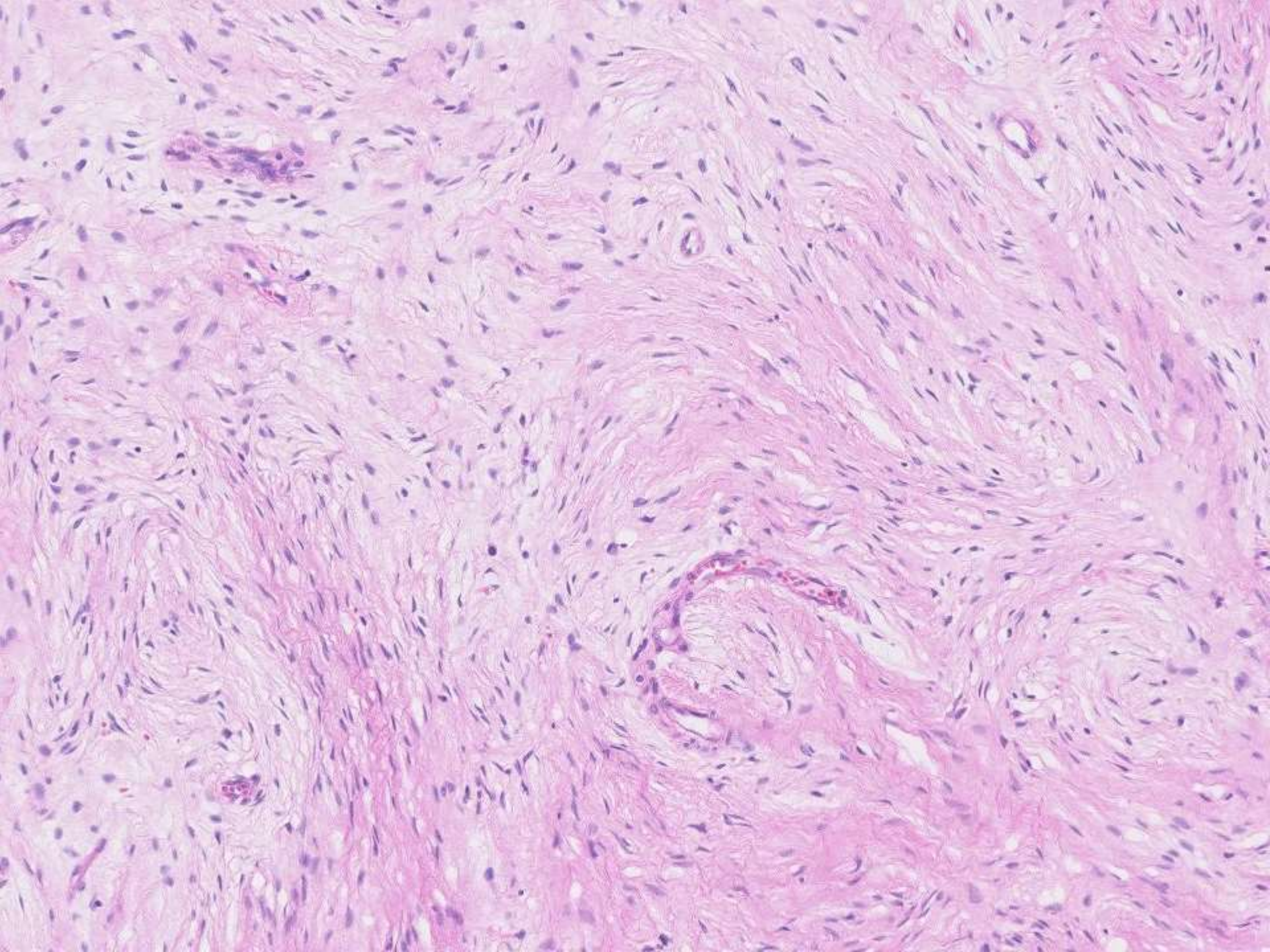


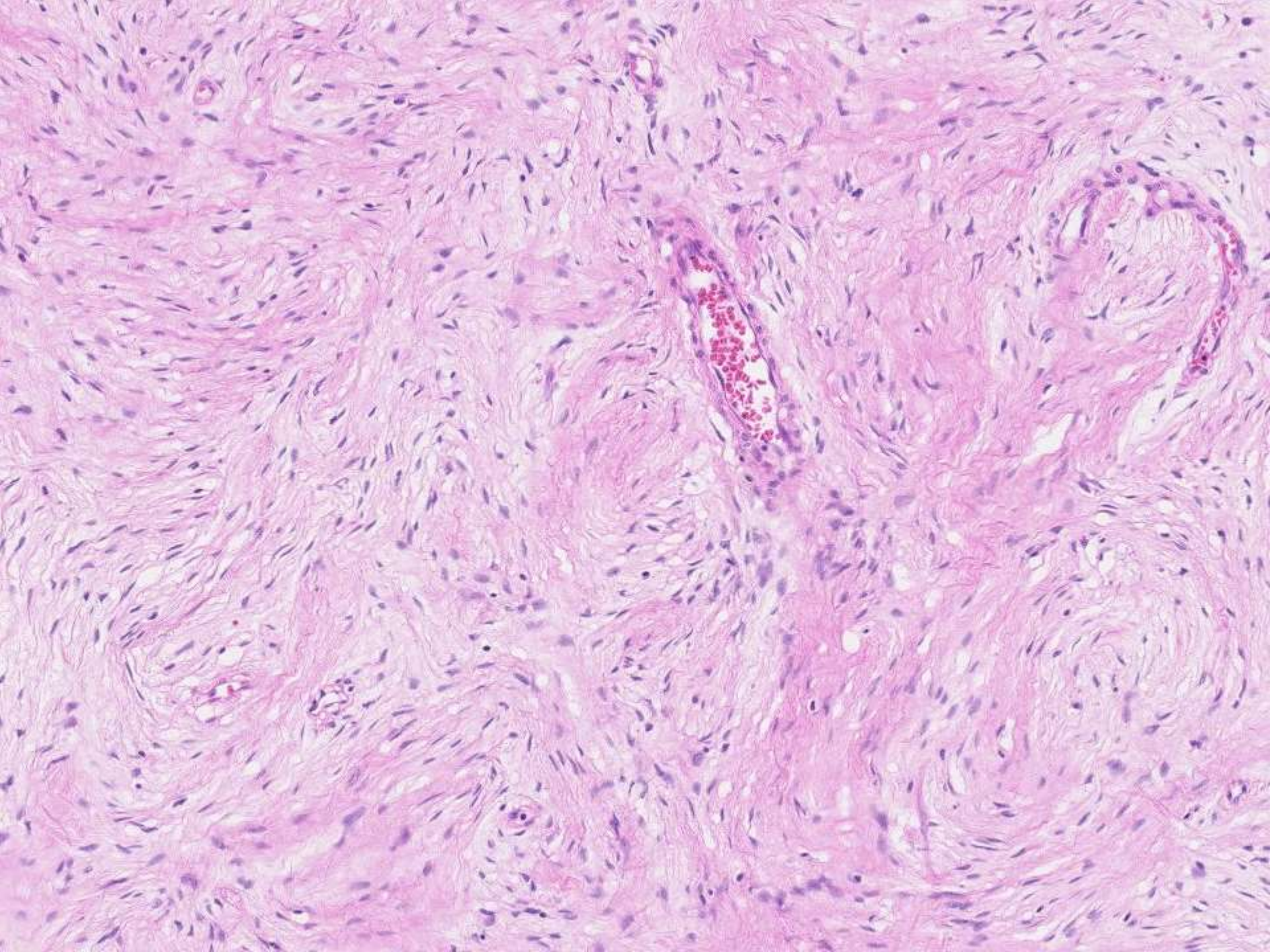


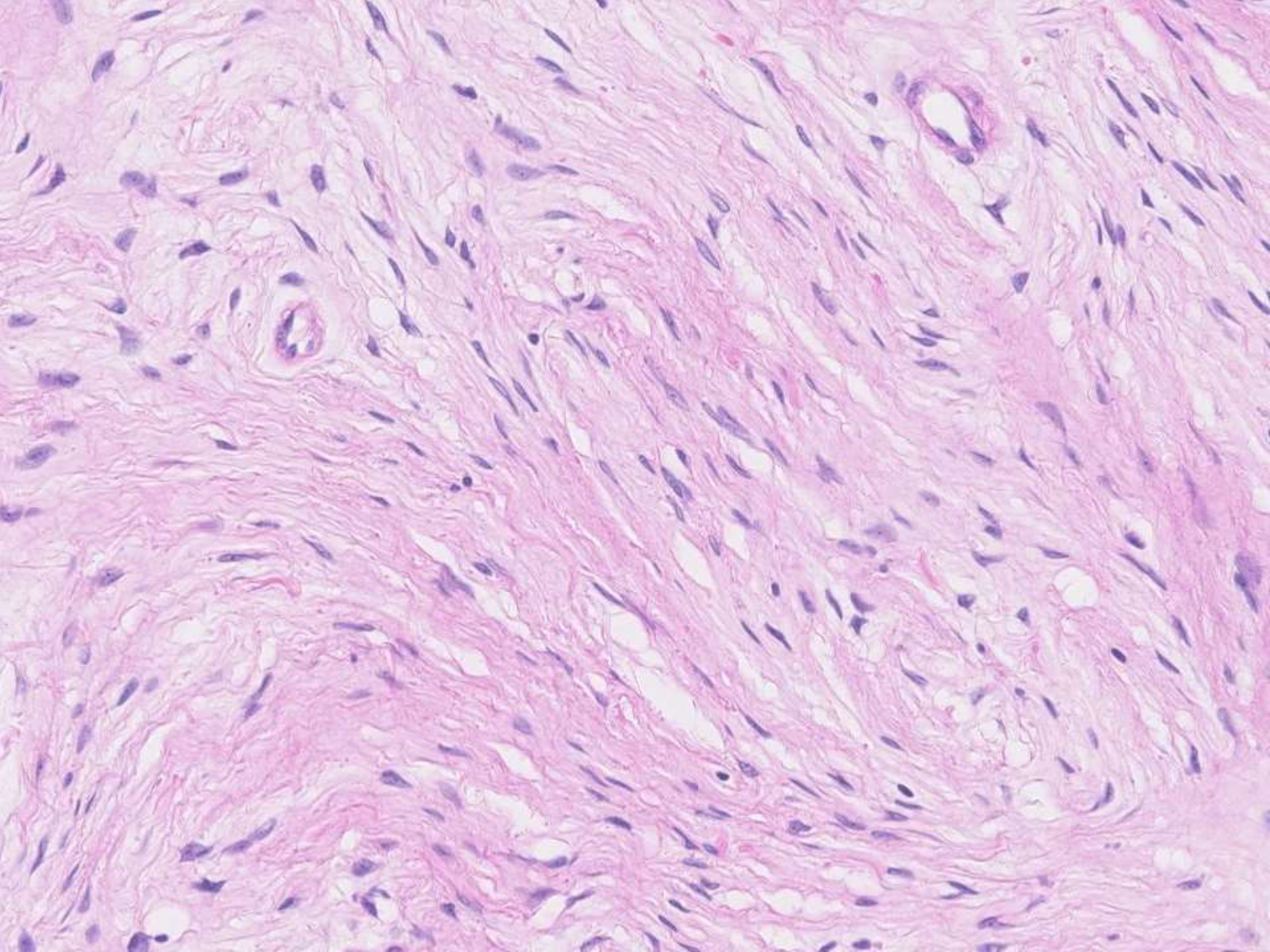


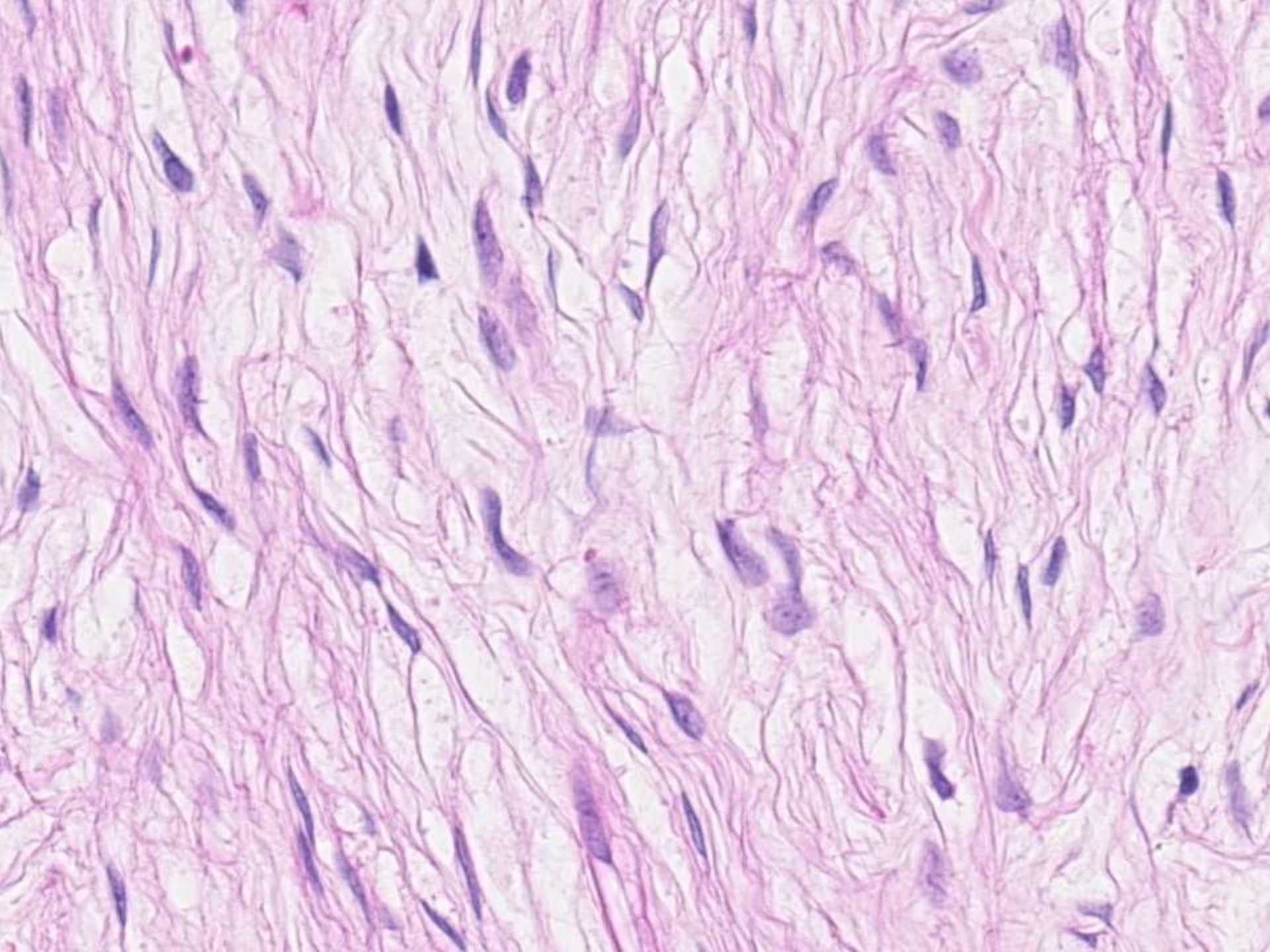


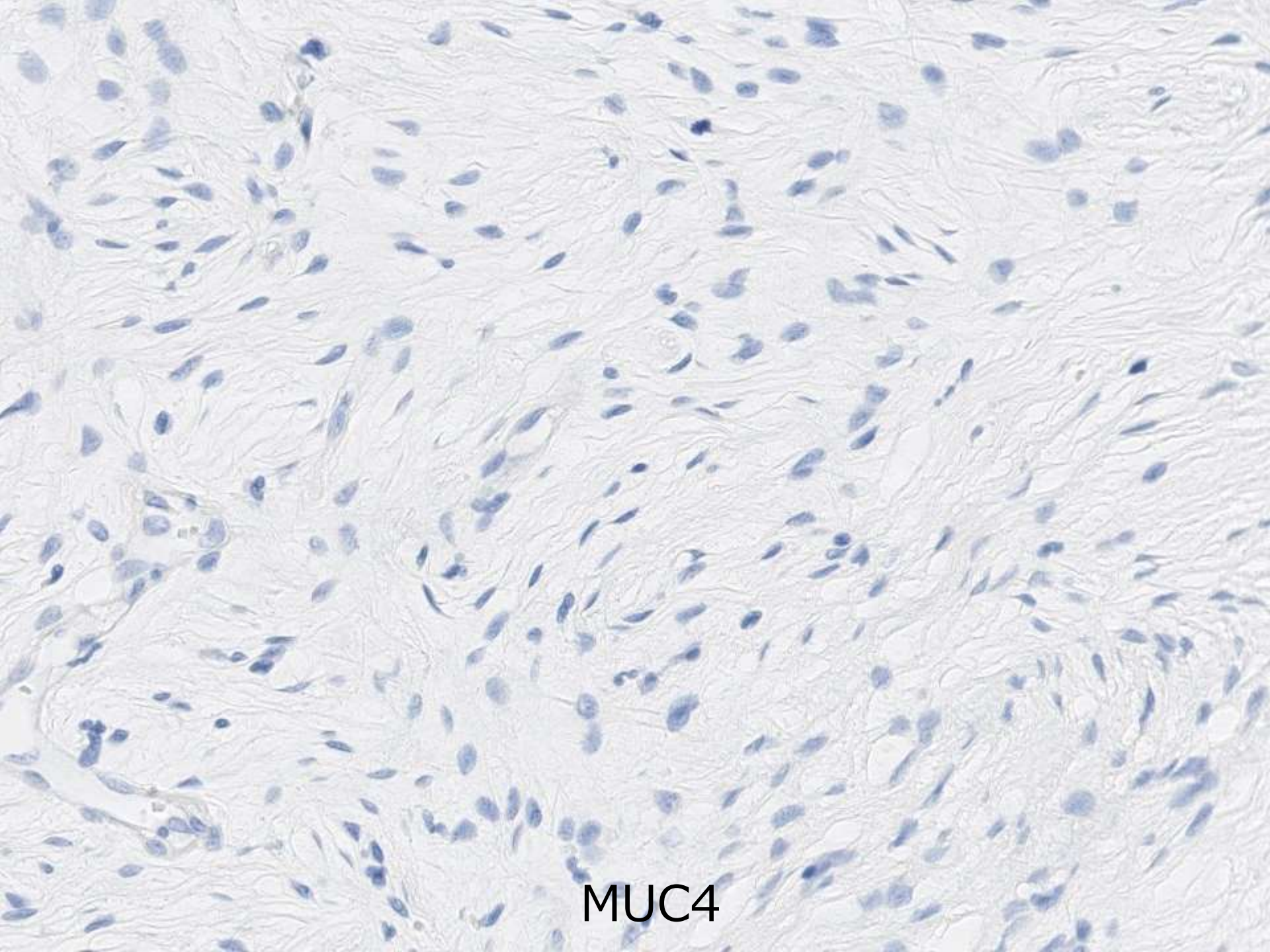


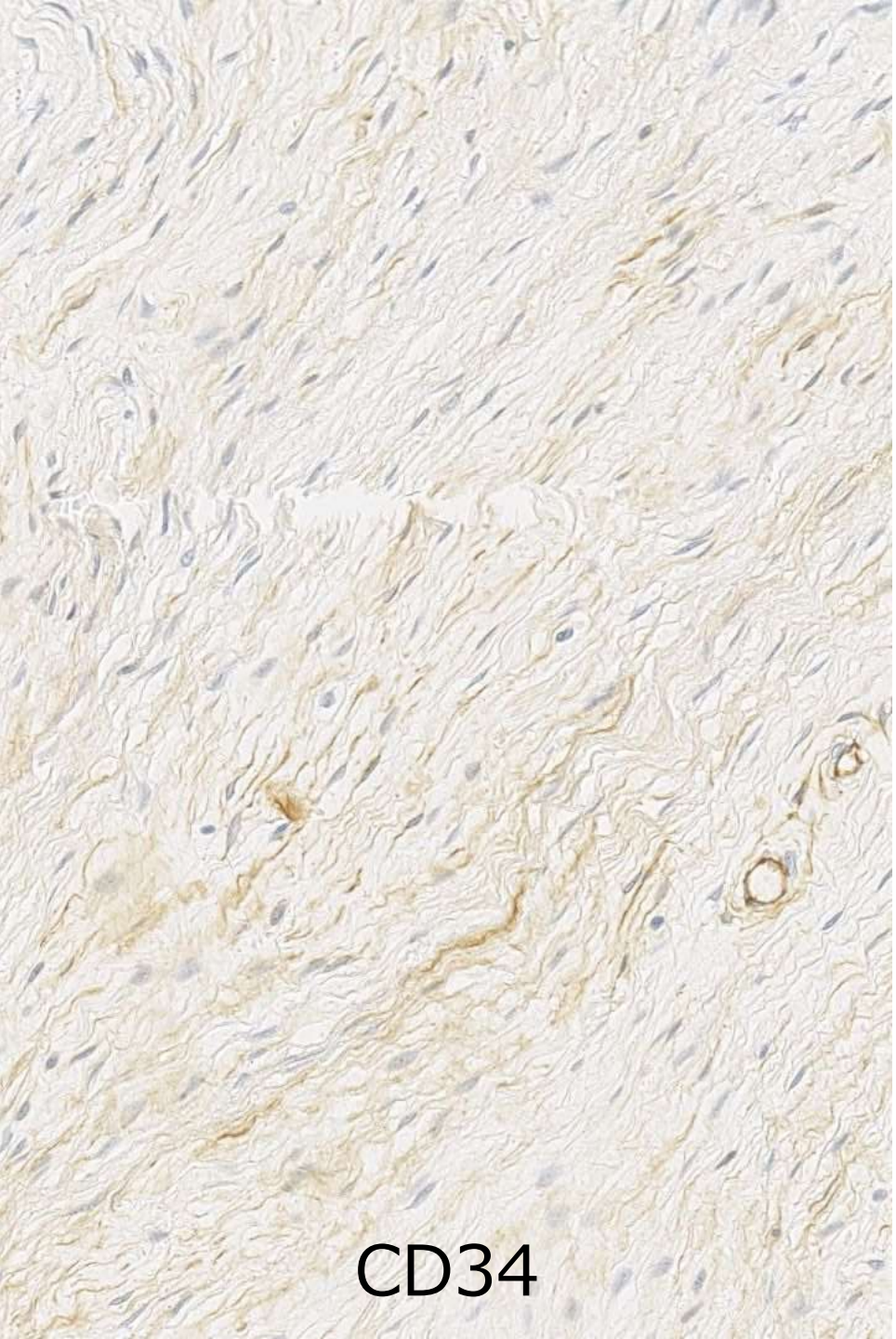


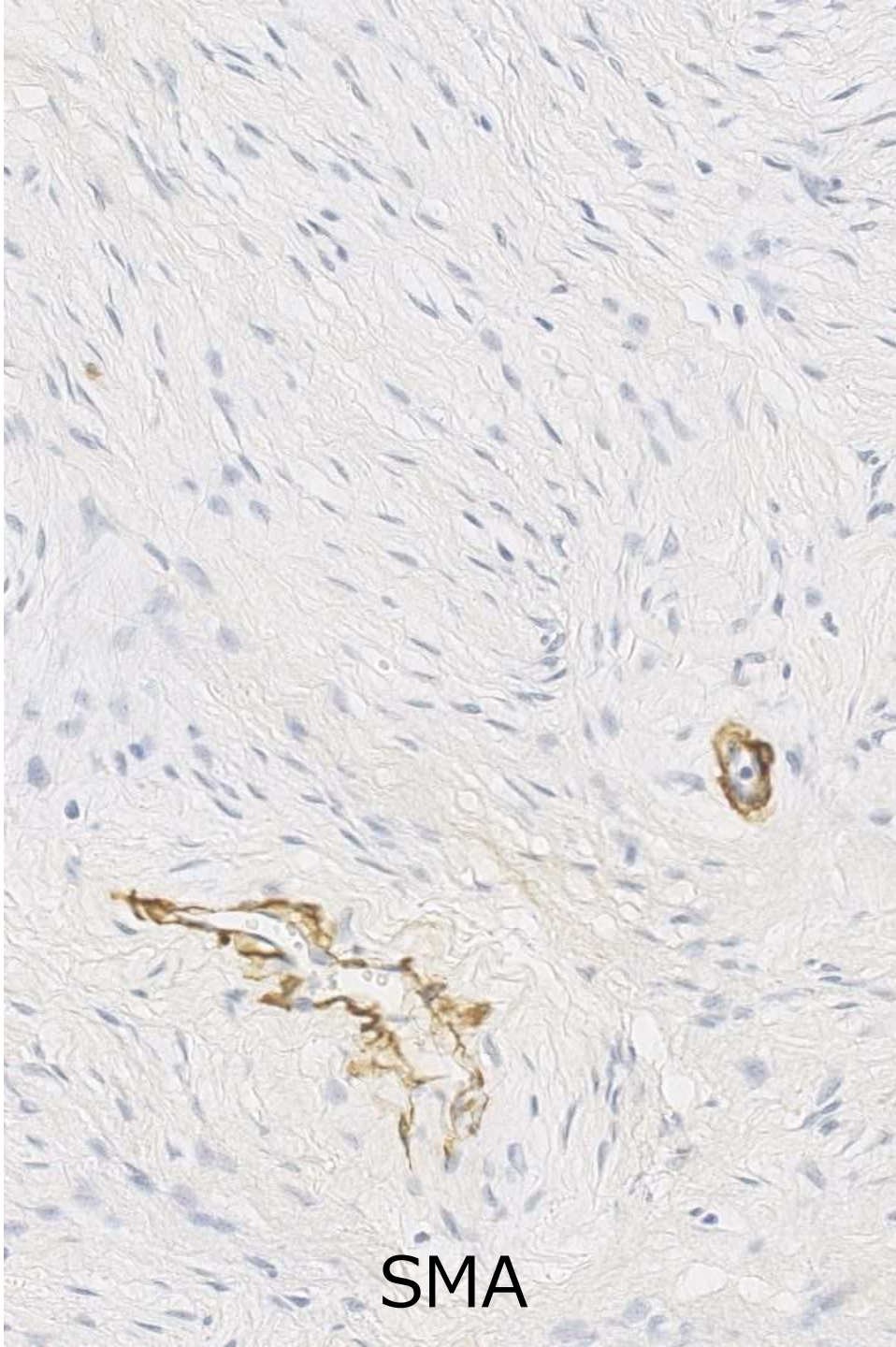






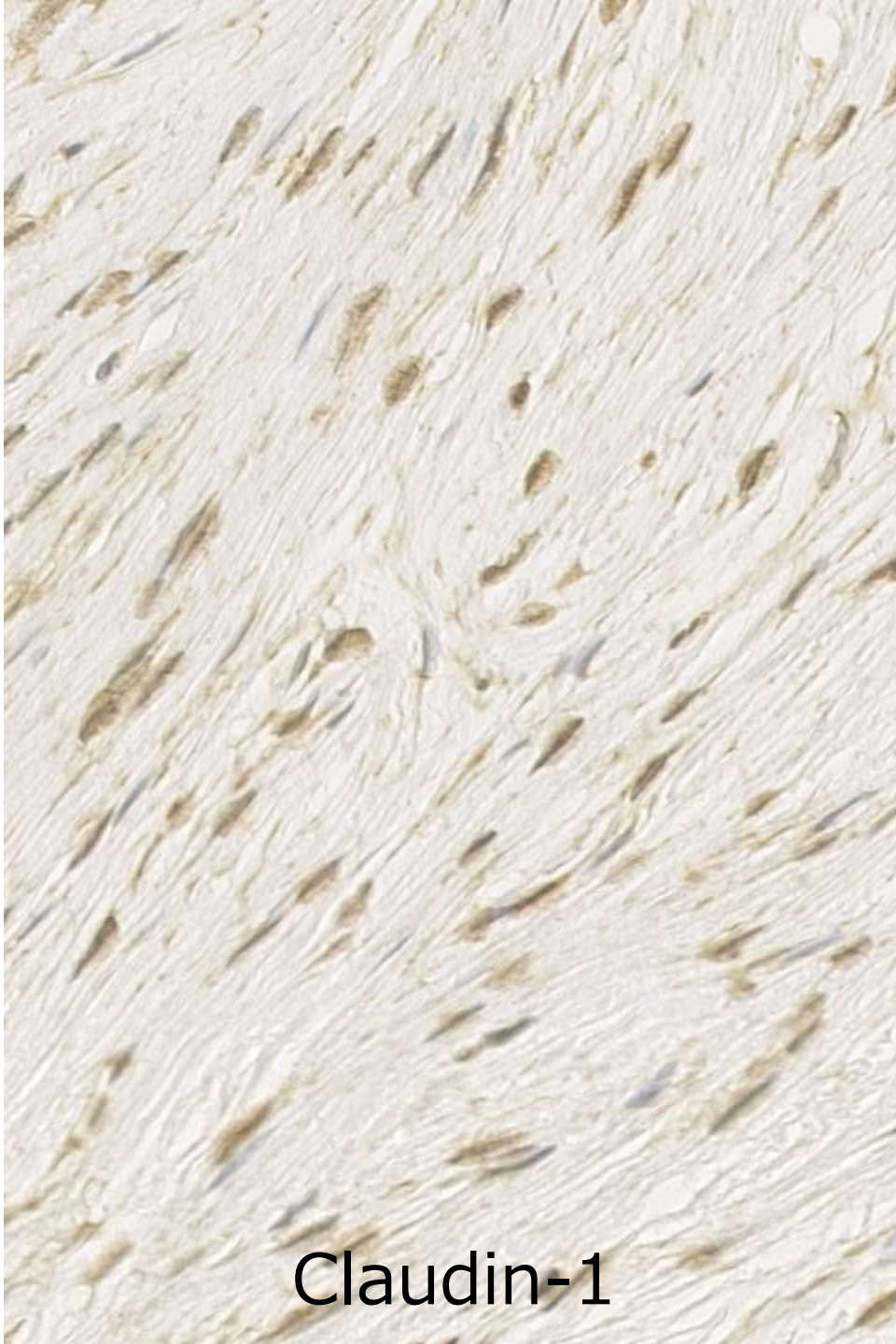

MUC4

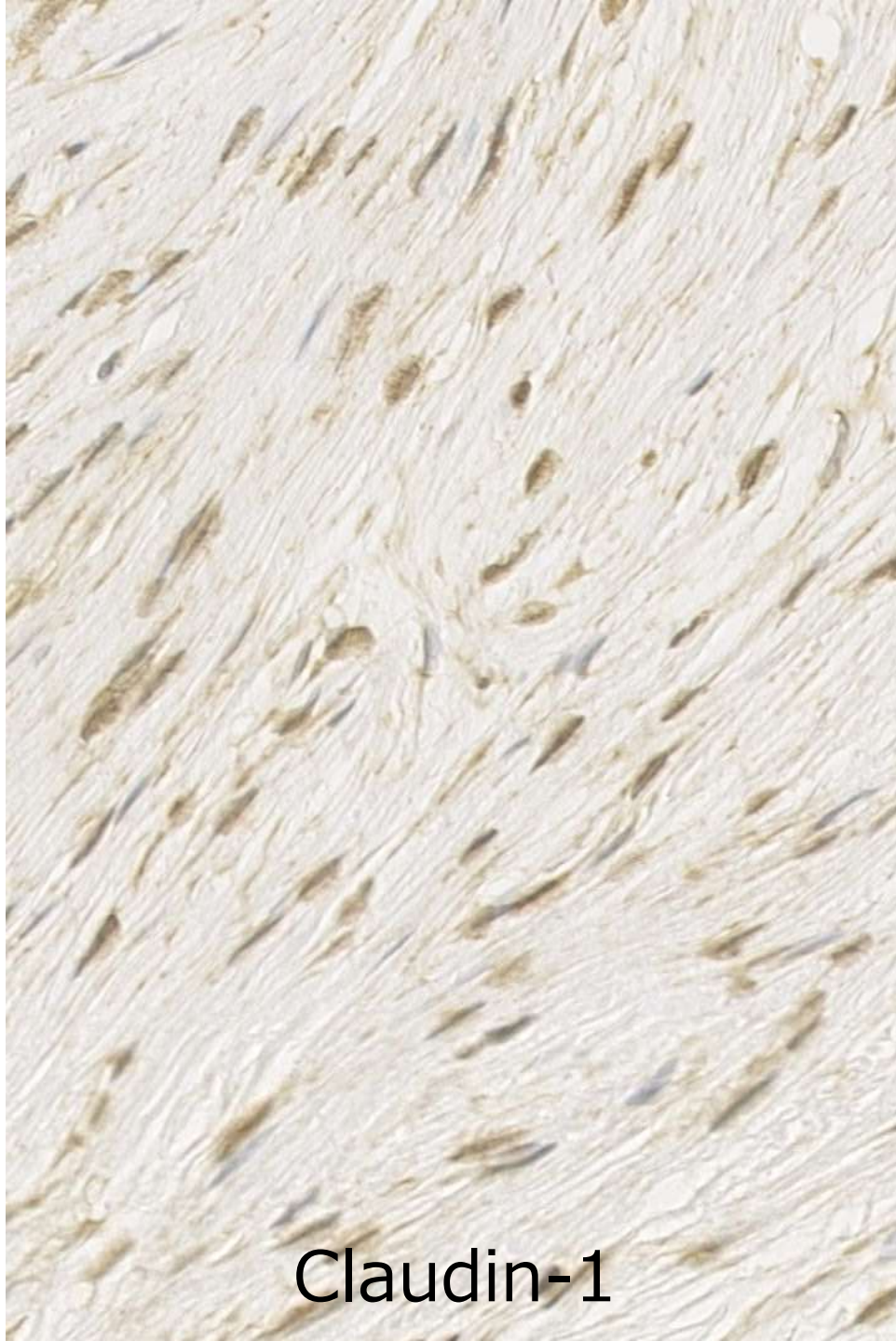

CD34


SMA

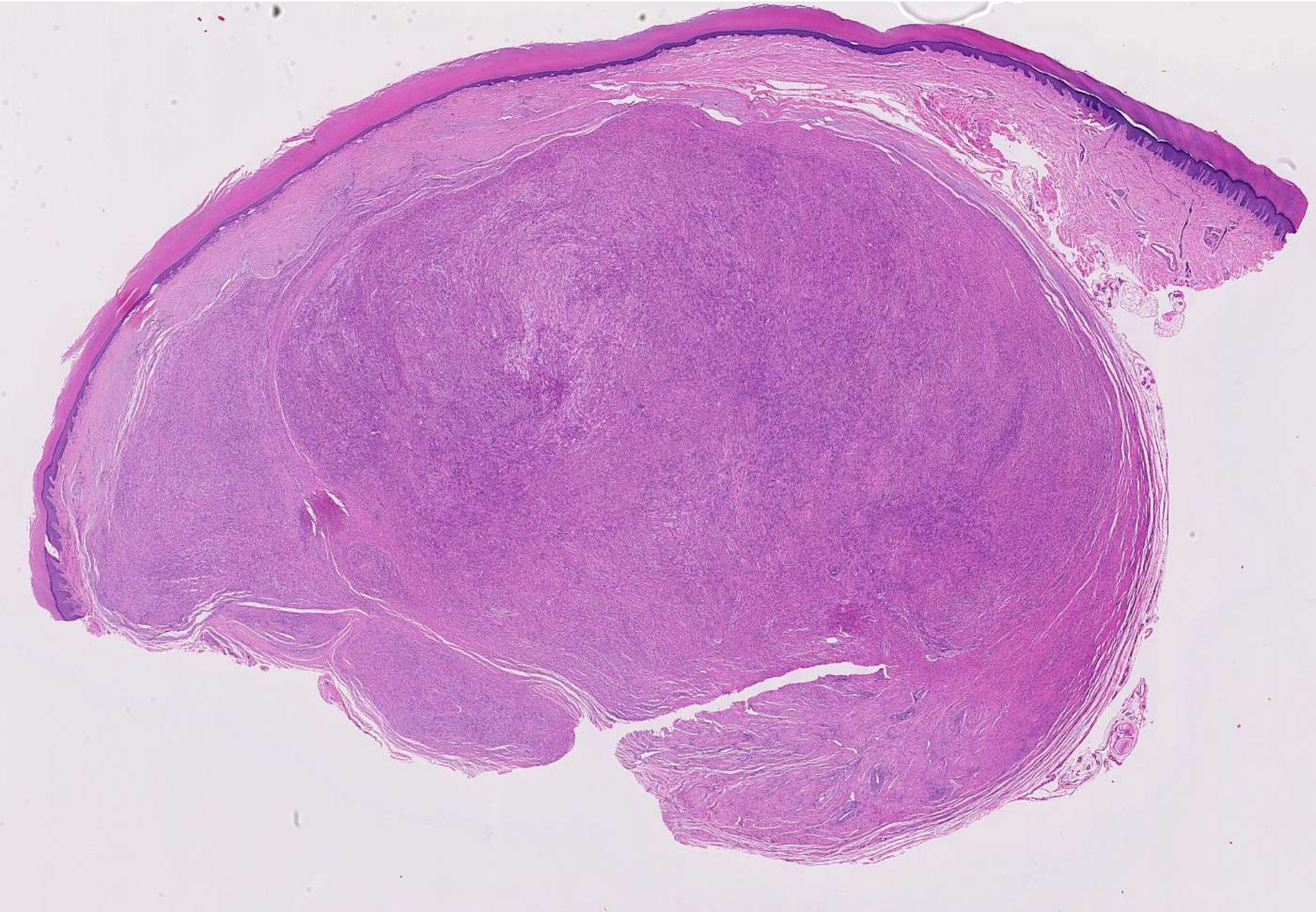

EMA

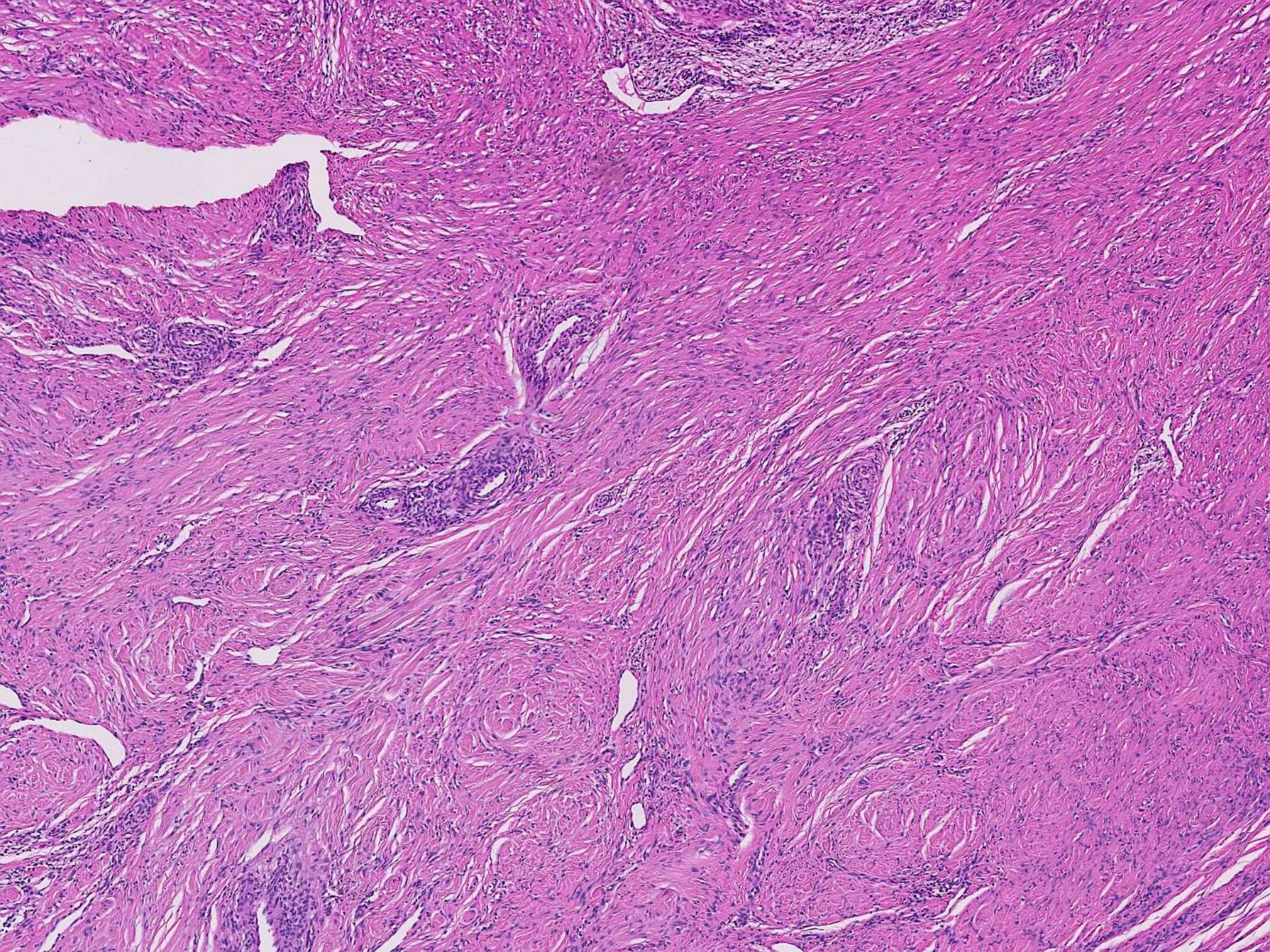

Glut-1

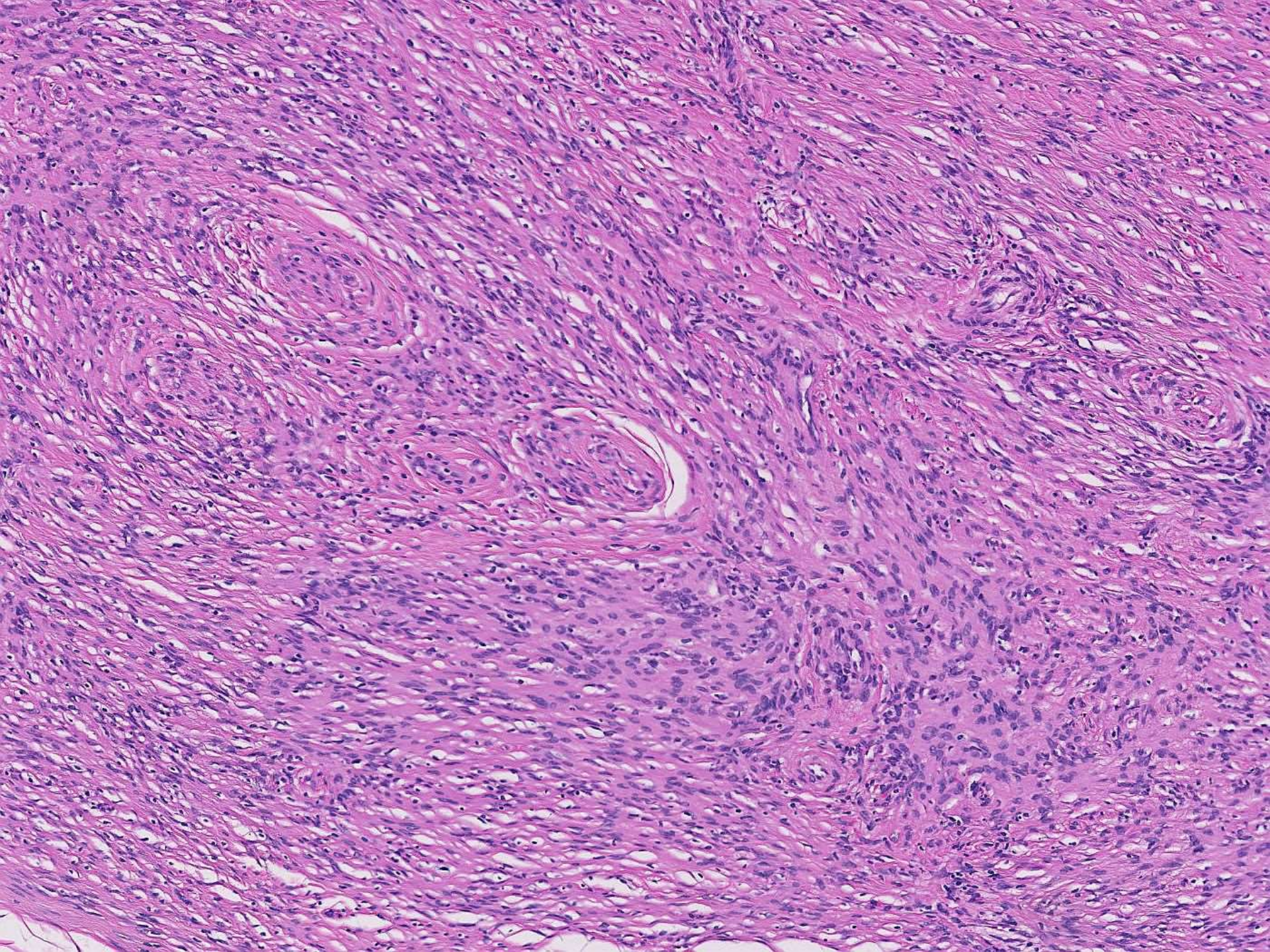

Claudin-1

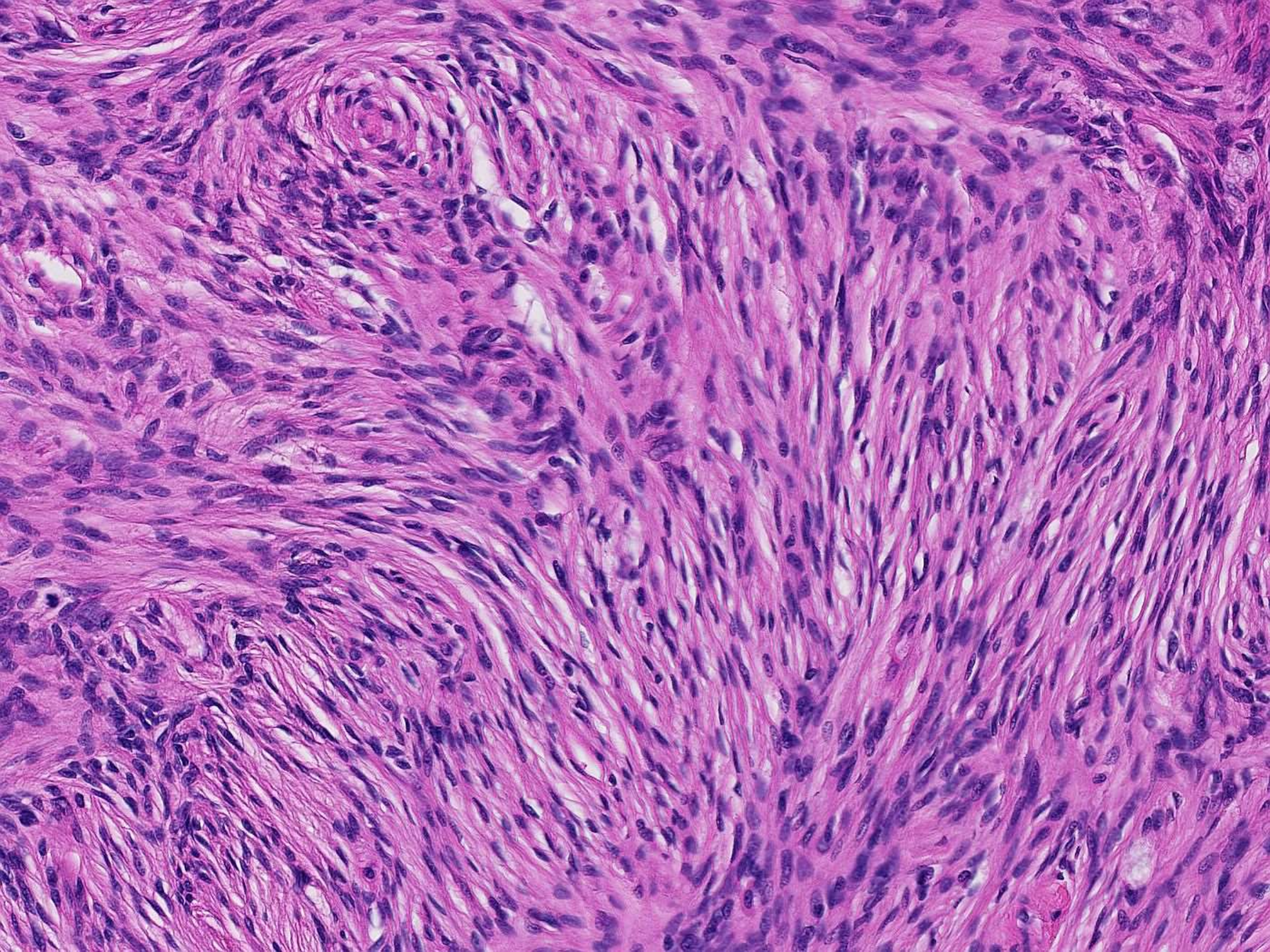

Claudin-1

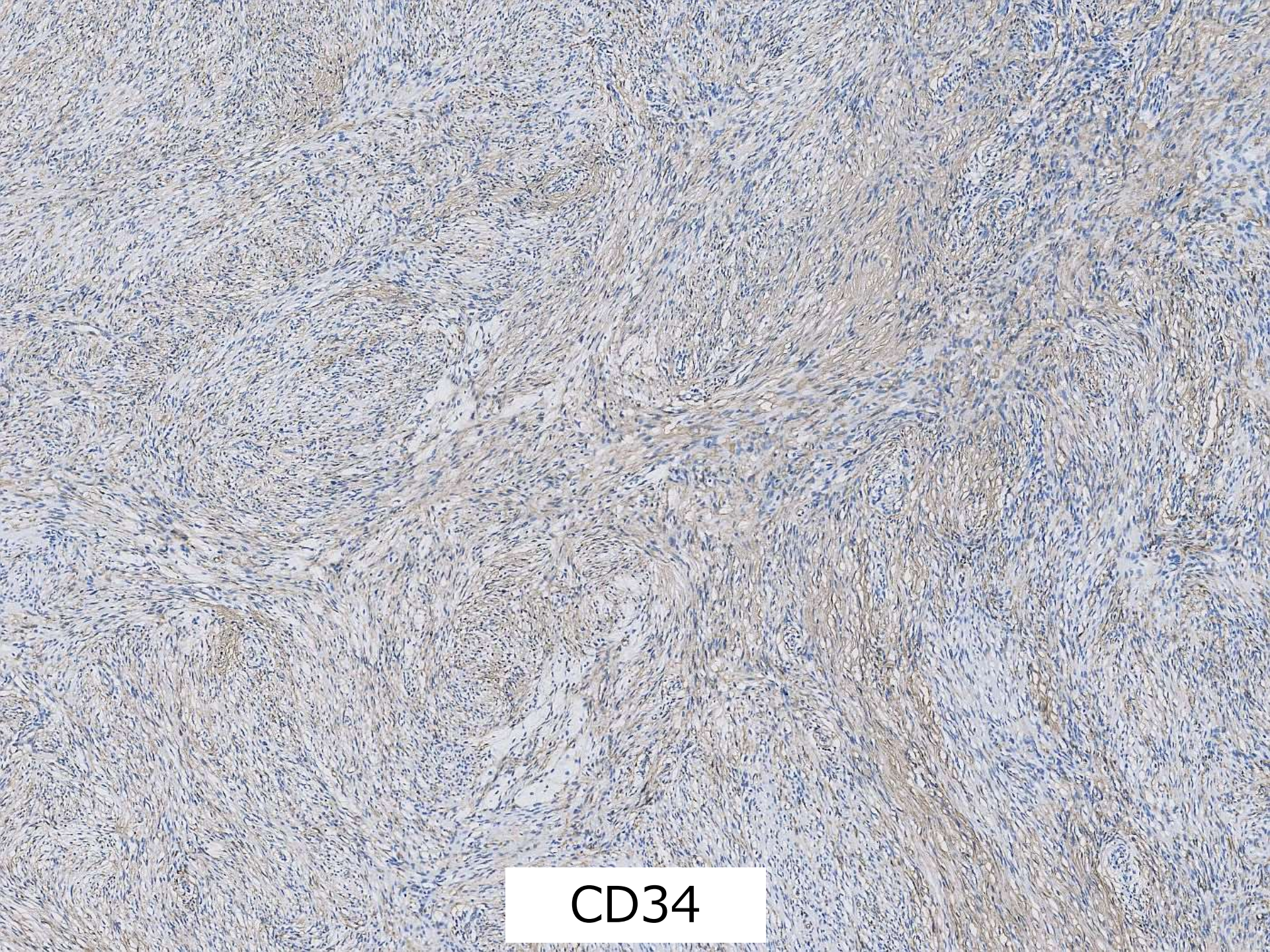
47F, thumb







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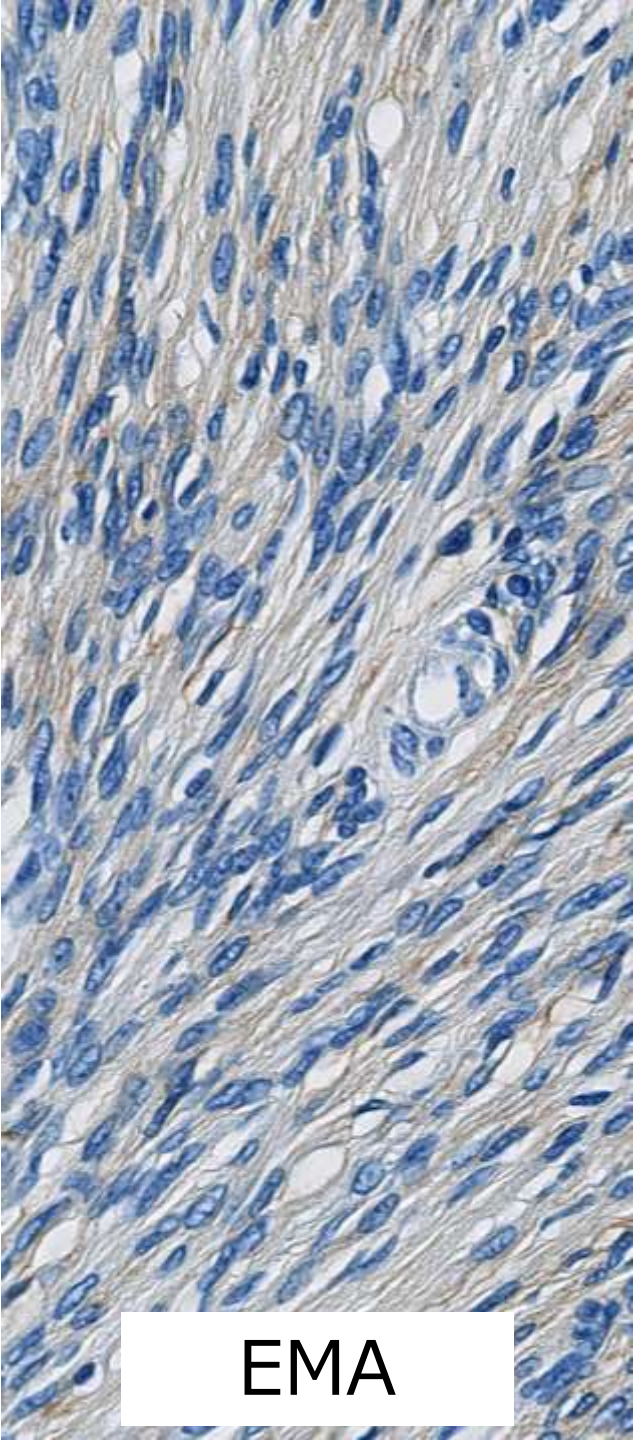
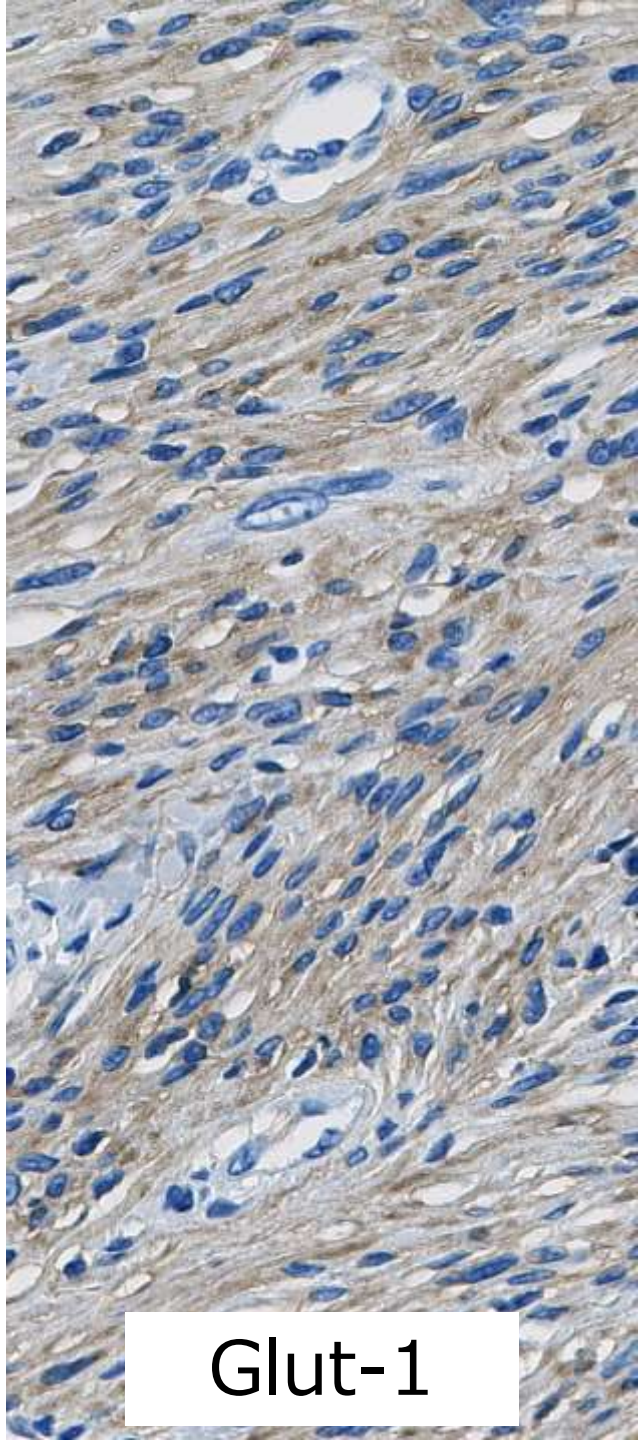
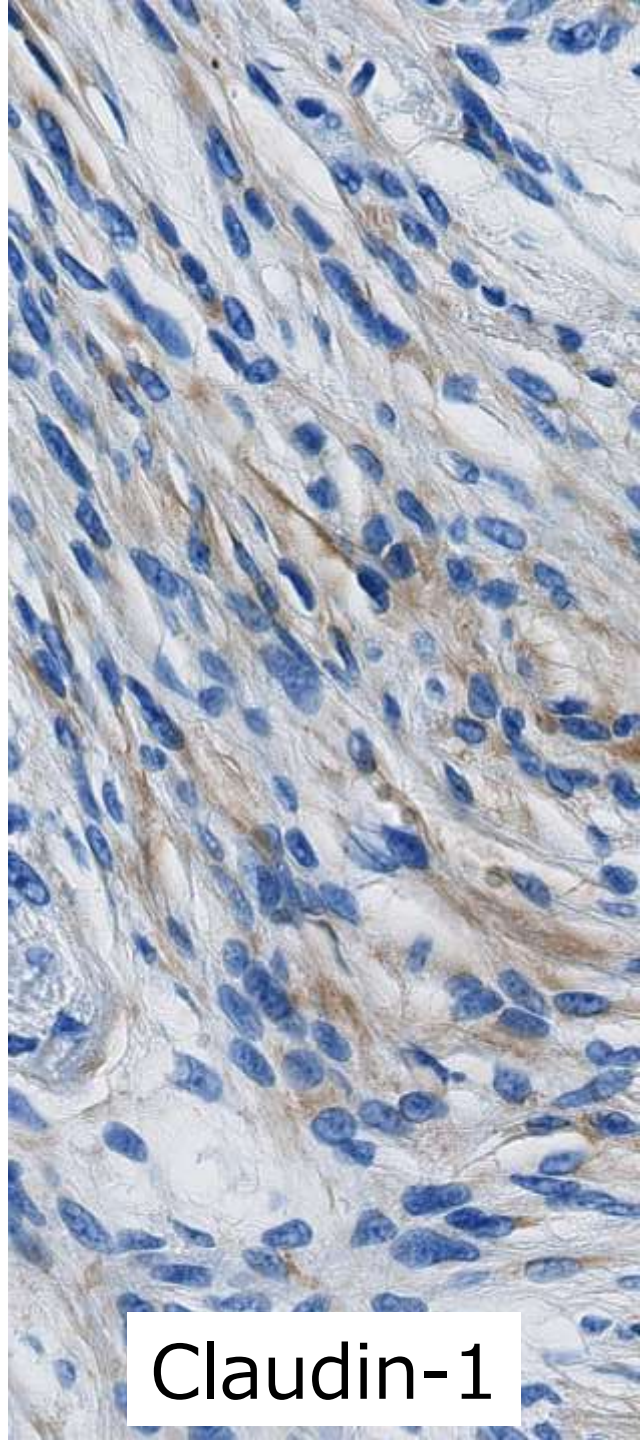
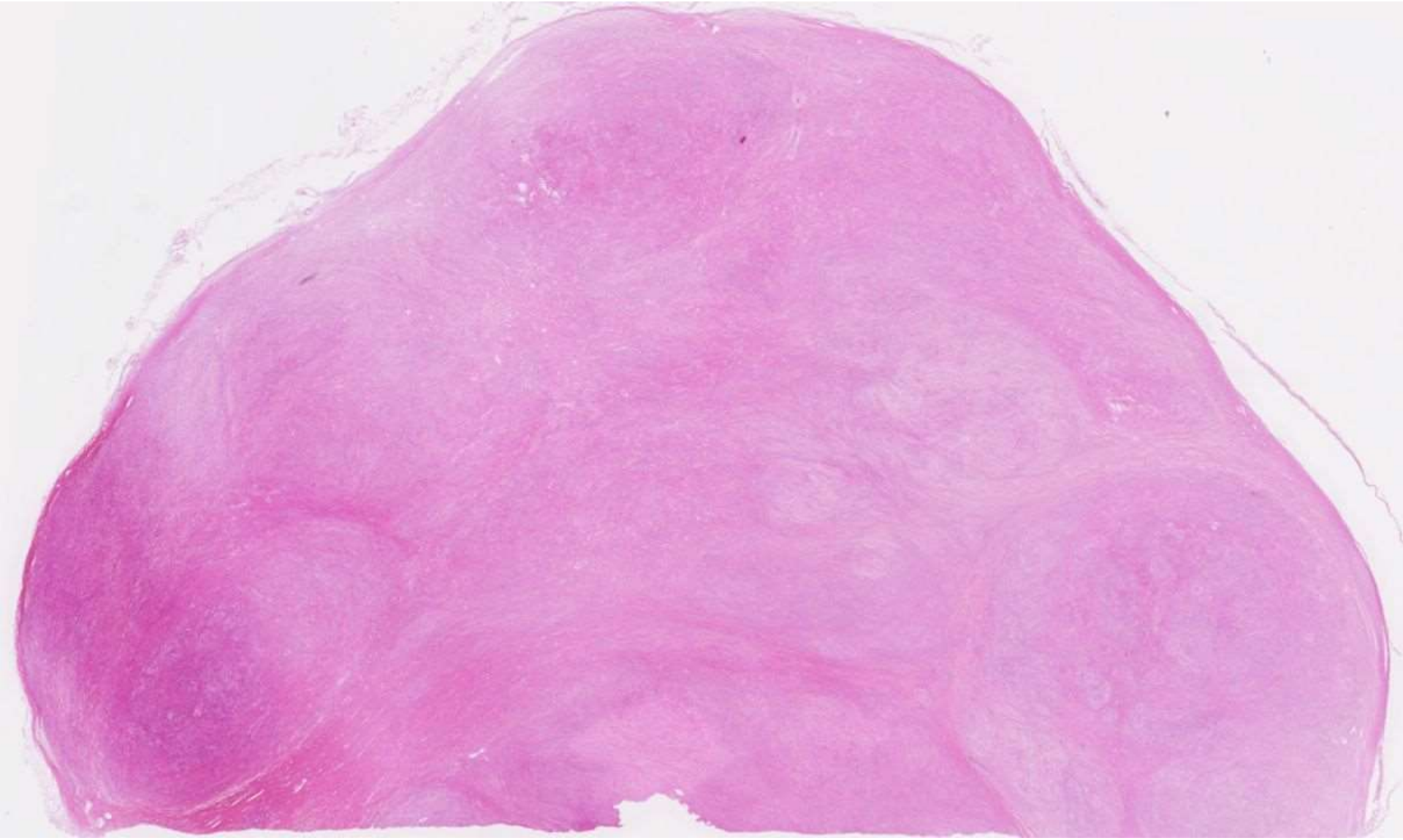
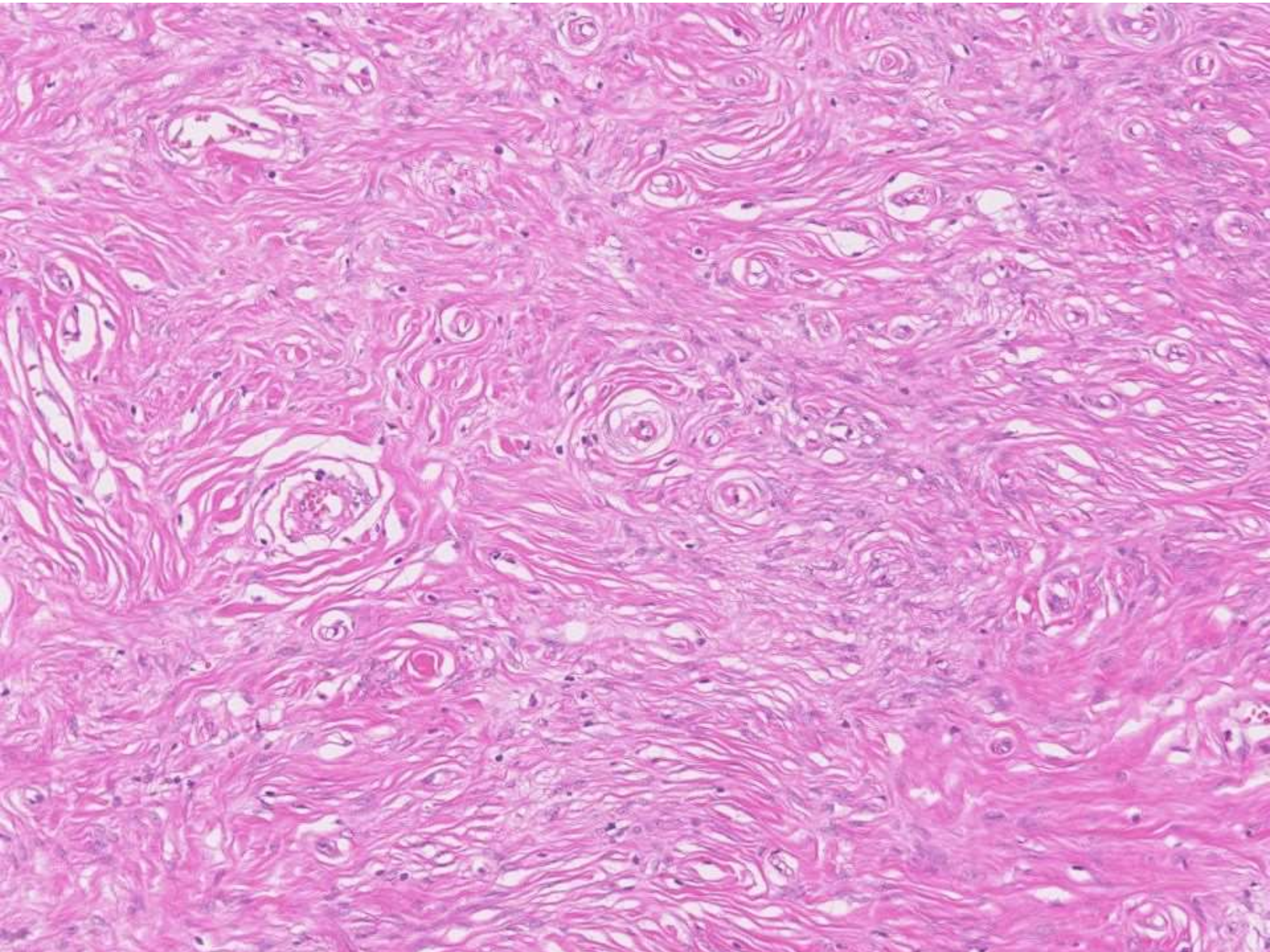

EMA

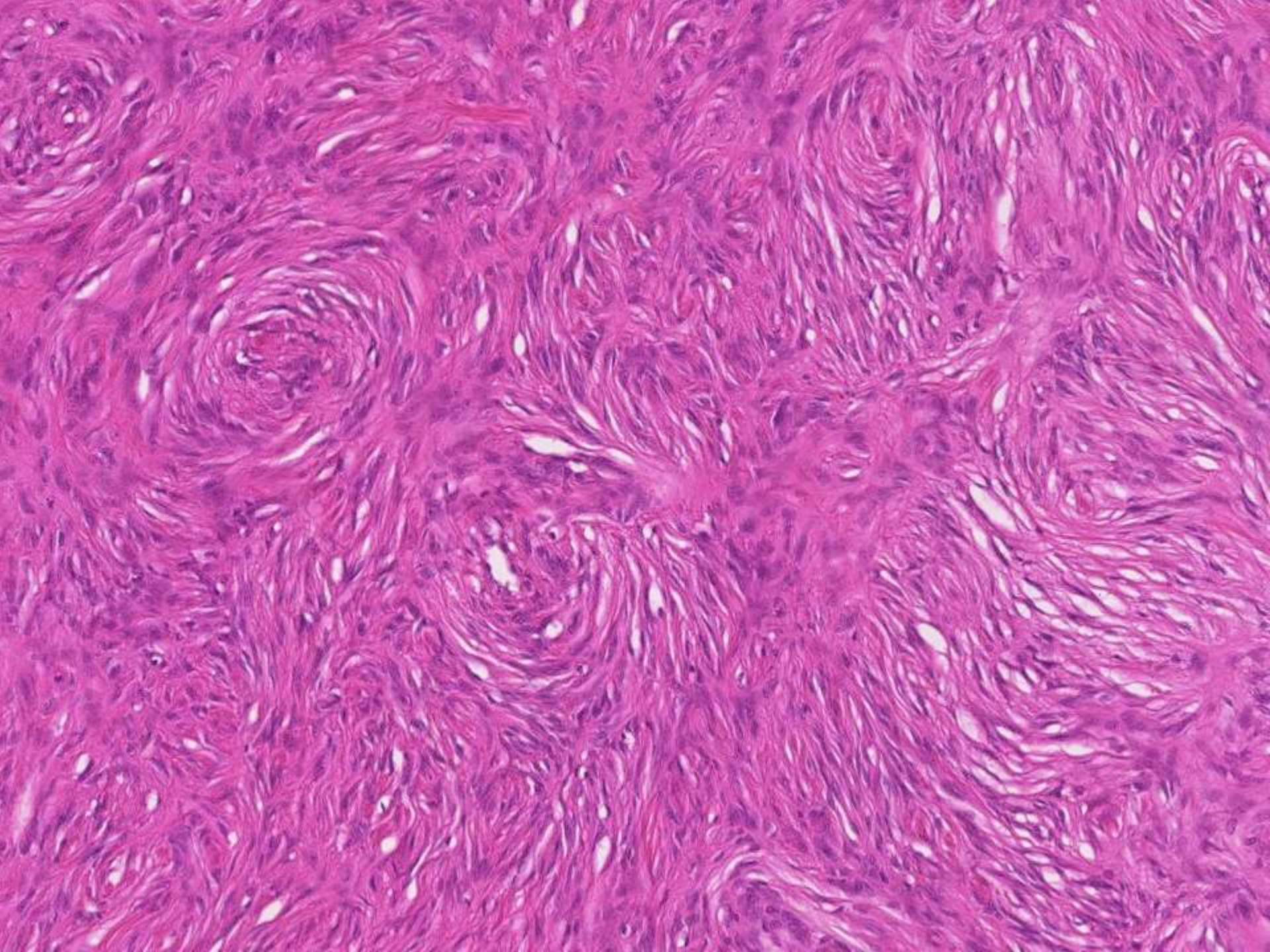

Glut-1

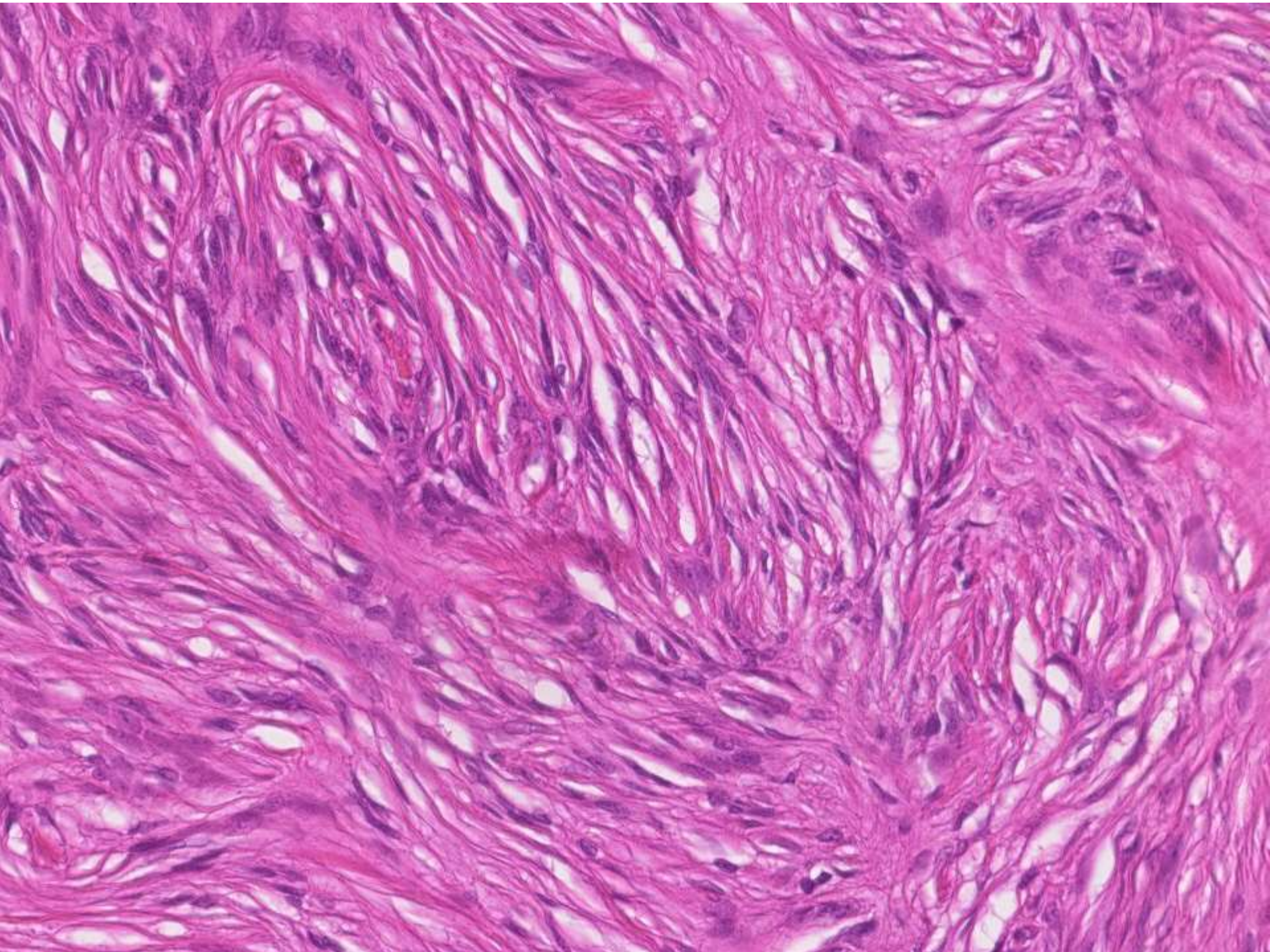

Claudin-1

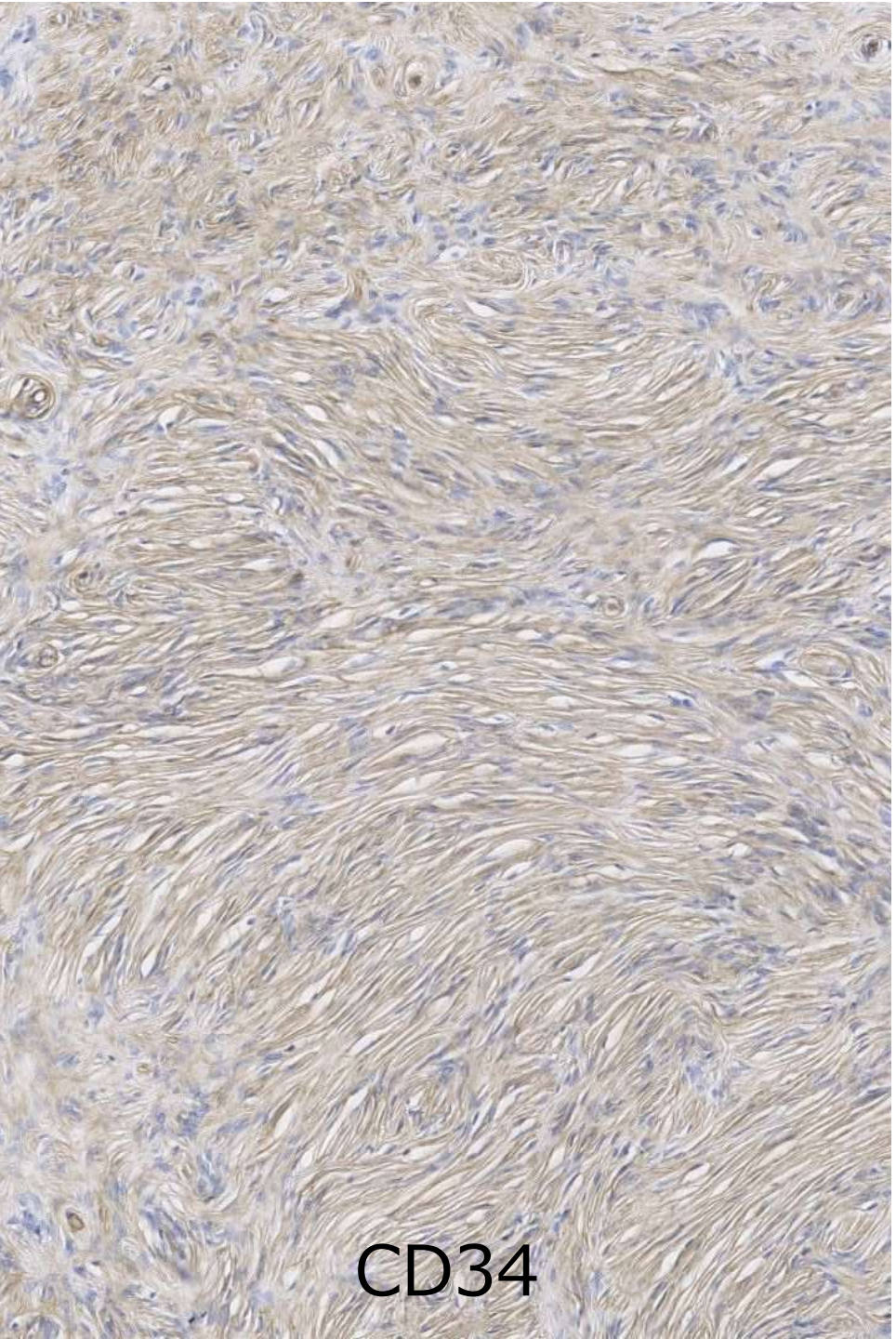
19M, upper arm







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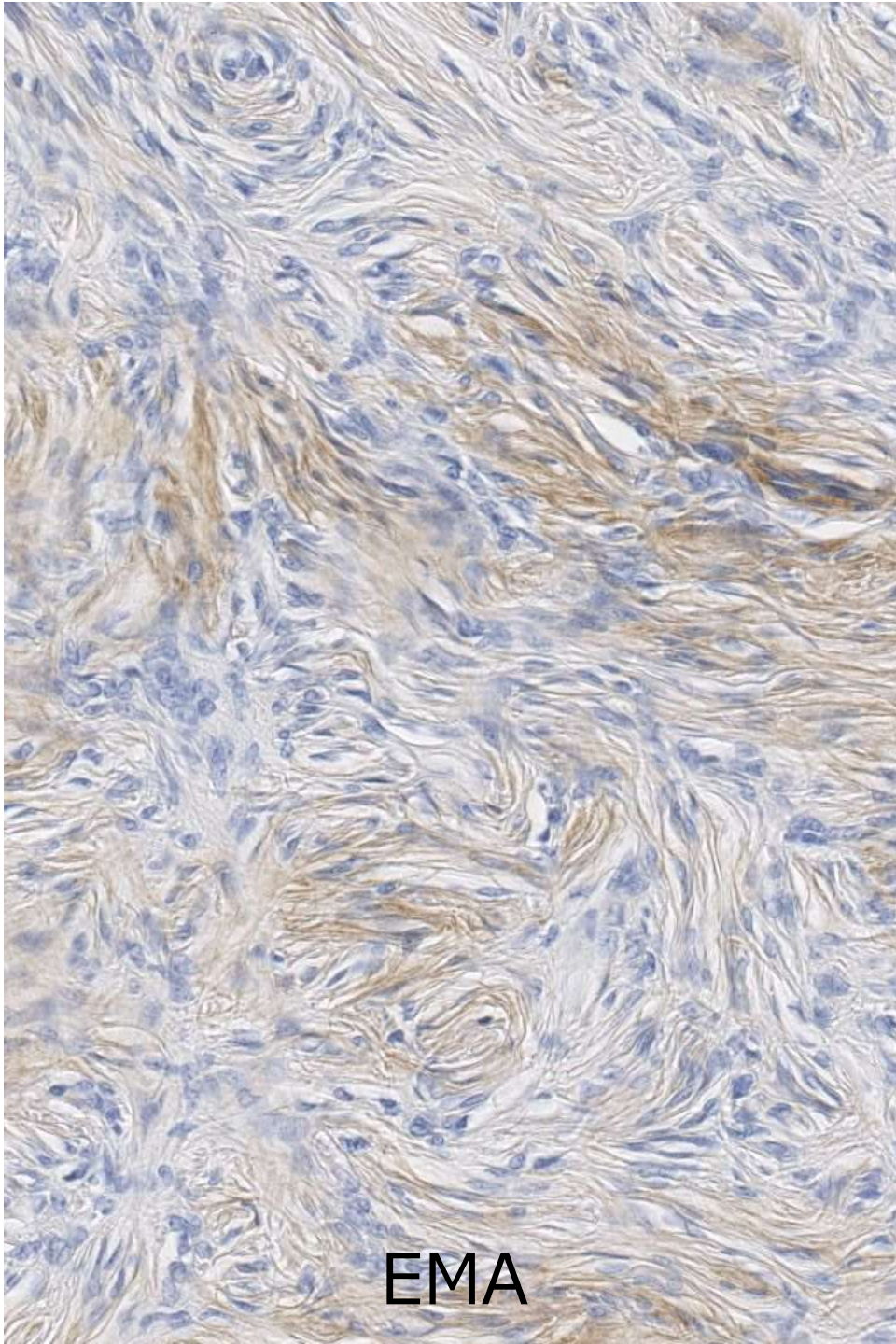

EMA

Tips 4

Desmoid fibromatosis
vs.
Low grade fibromyxoid sarcoma
vs.
Soft tissue perineurioma

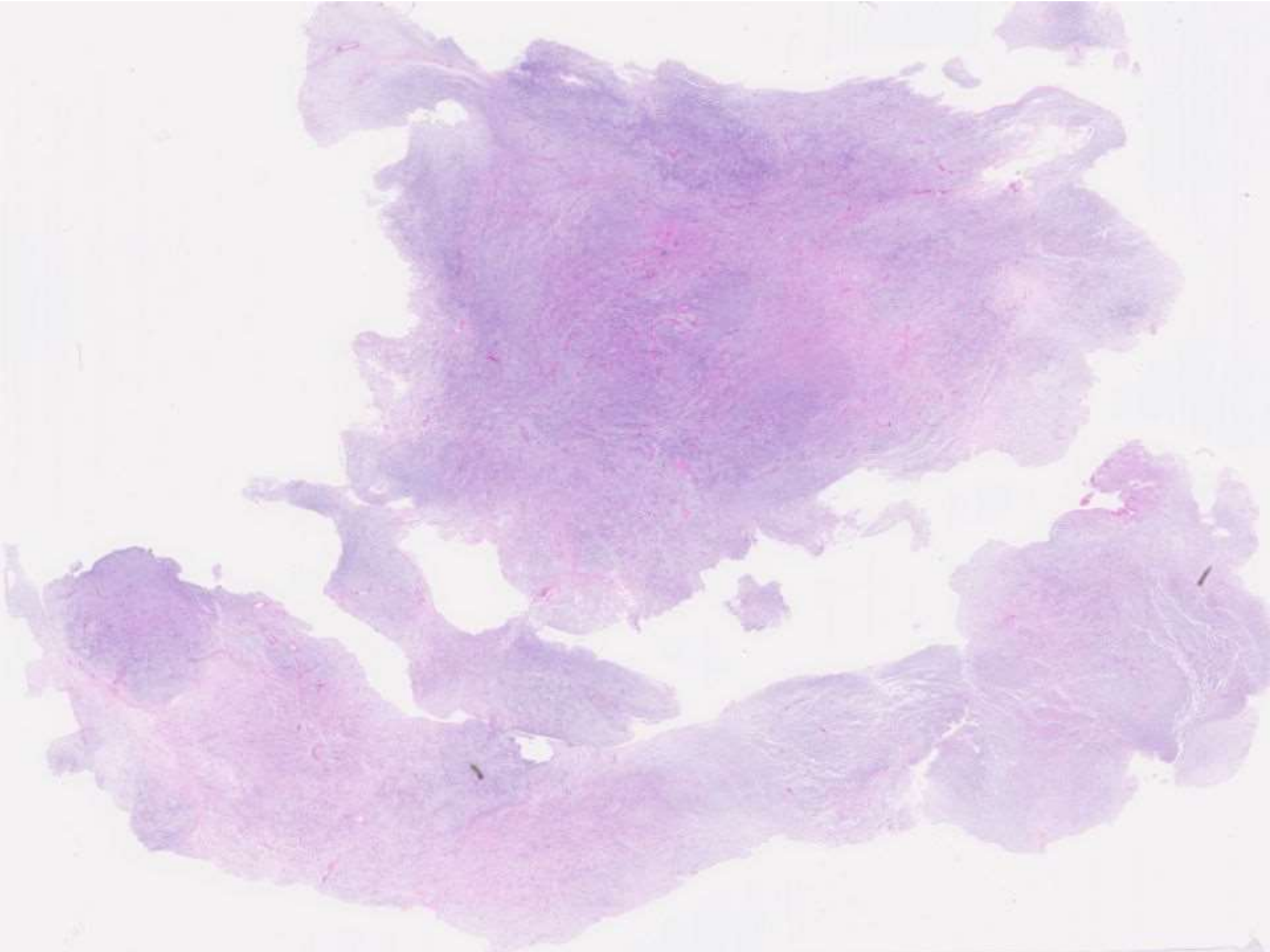
Desmoid : 直線的な細胞束、筋線維芽細胞の特徴（形態・IHC）
LGFMS : 曲線的で、collagenous>myxoid
Perineurioma : 曲線的で、渦巻き状配列が通常めだつが、多彩

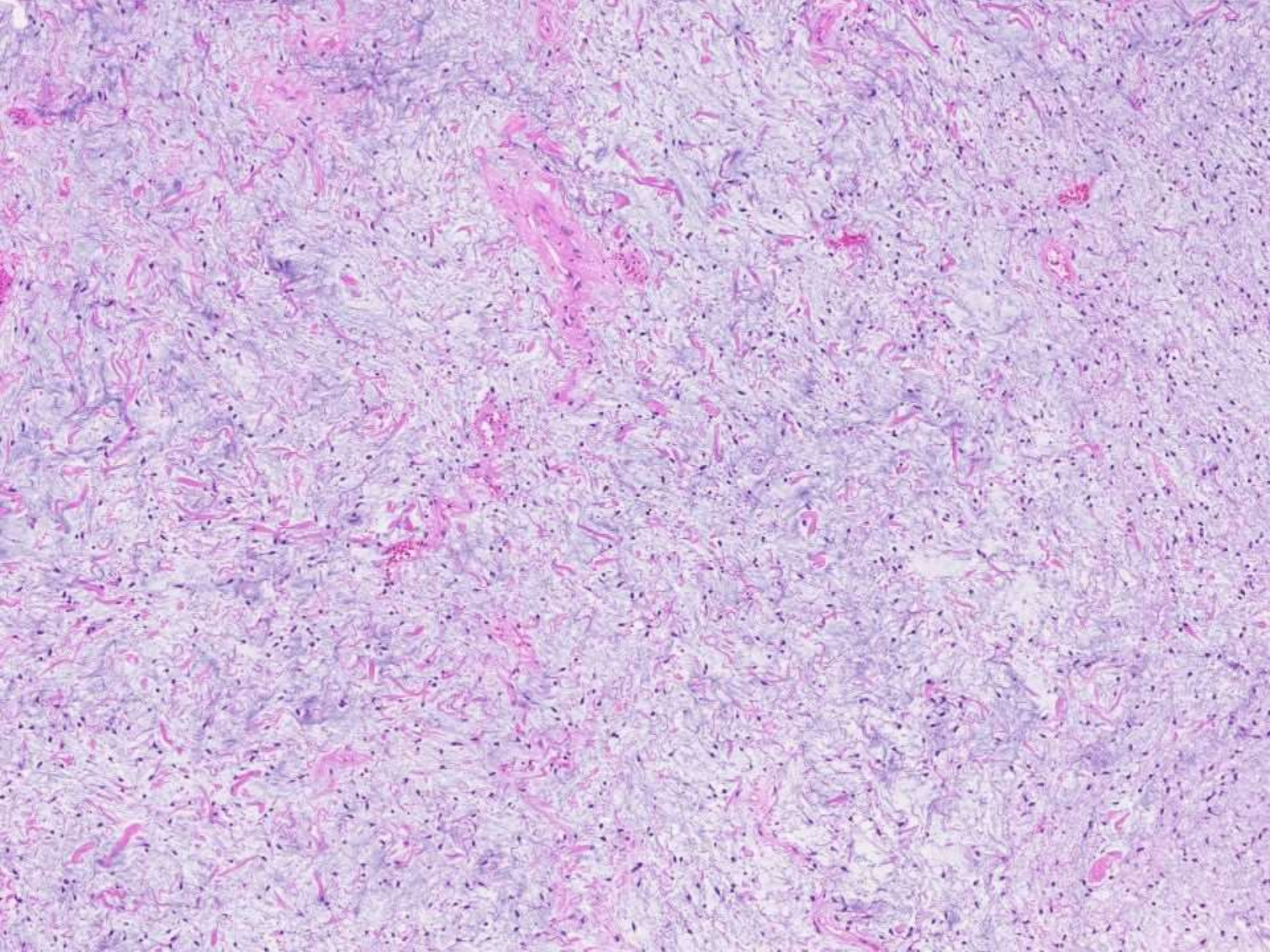
＊MyxofibrosarcomaはLGFMSの鑑別の対象となりにくい

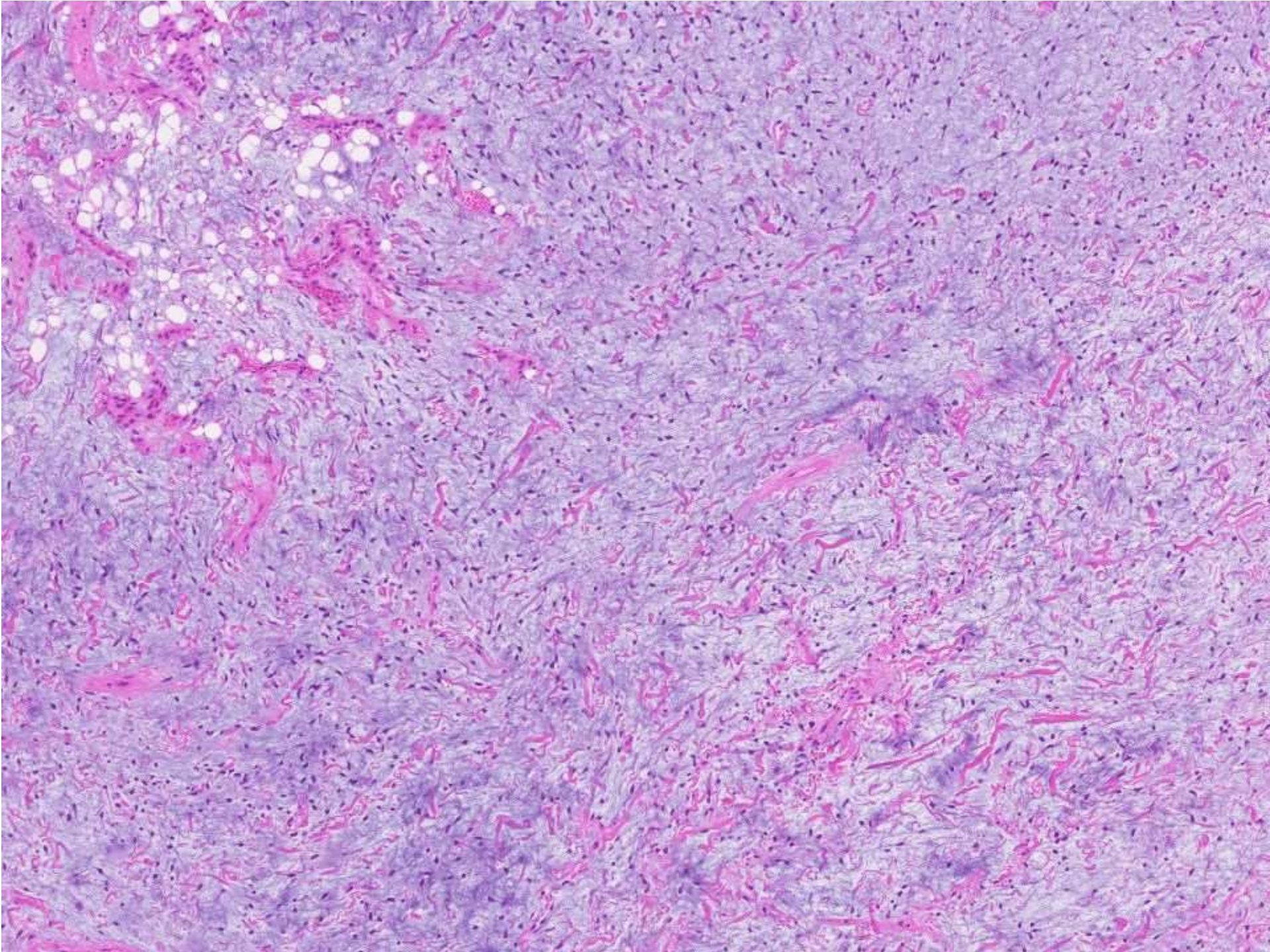
Case 3: 36M, thigh

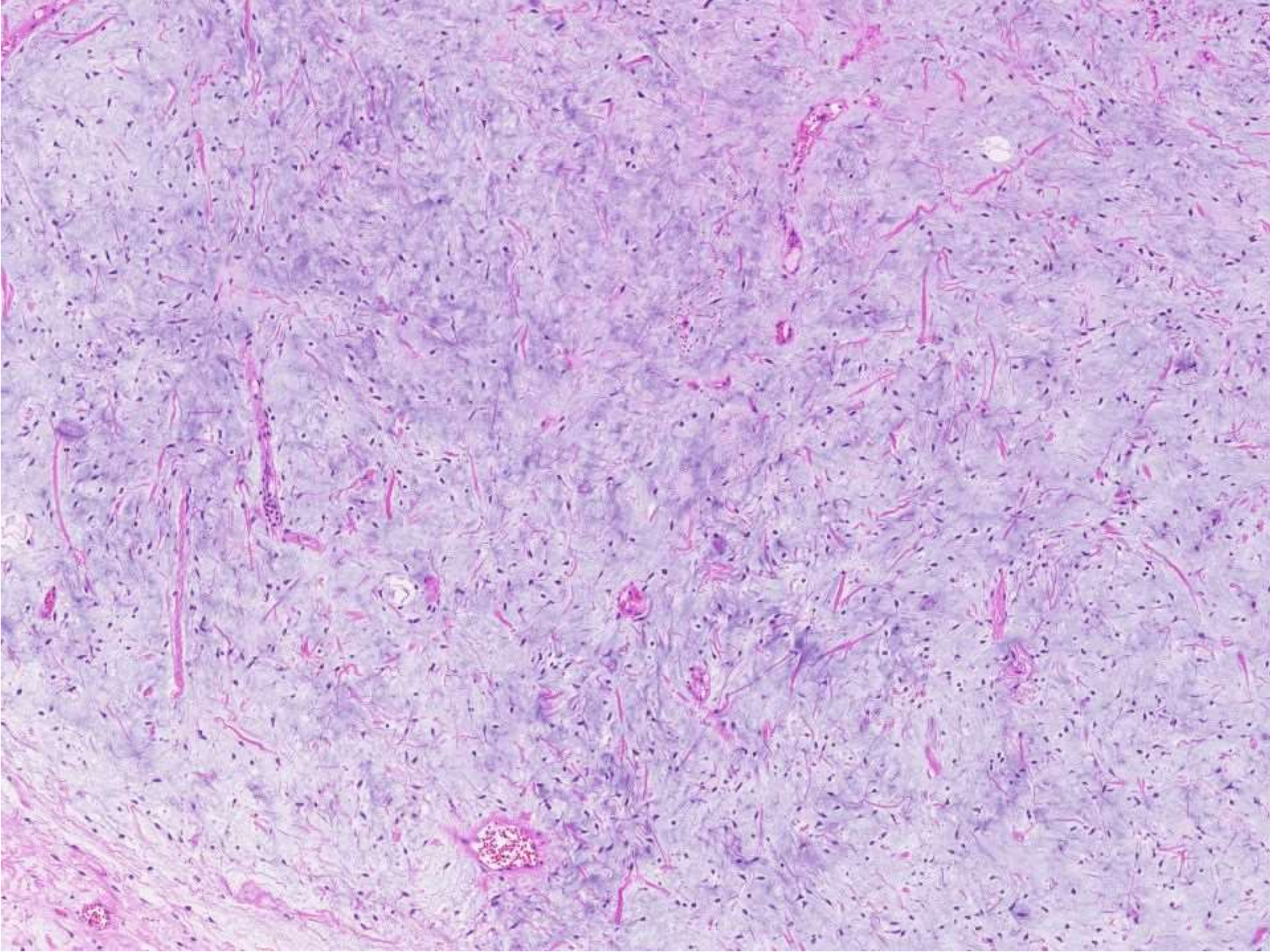
右大腿の筋間の3 cm大の腫瘍

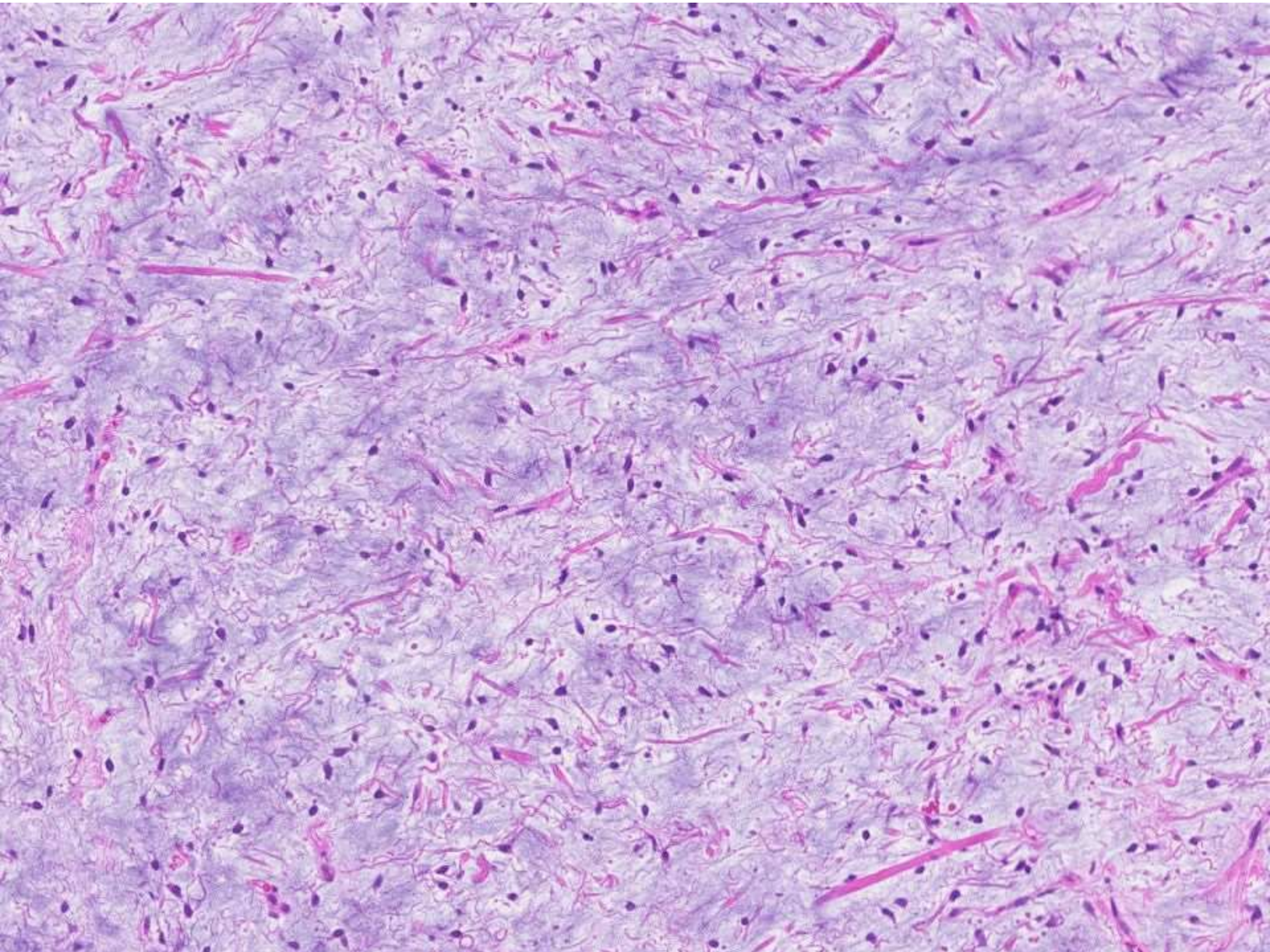
Low grade fibromyxoid sarcoma?
Myxoid liposarcoma?

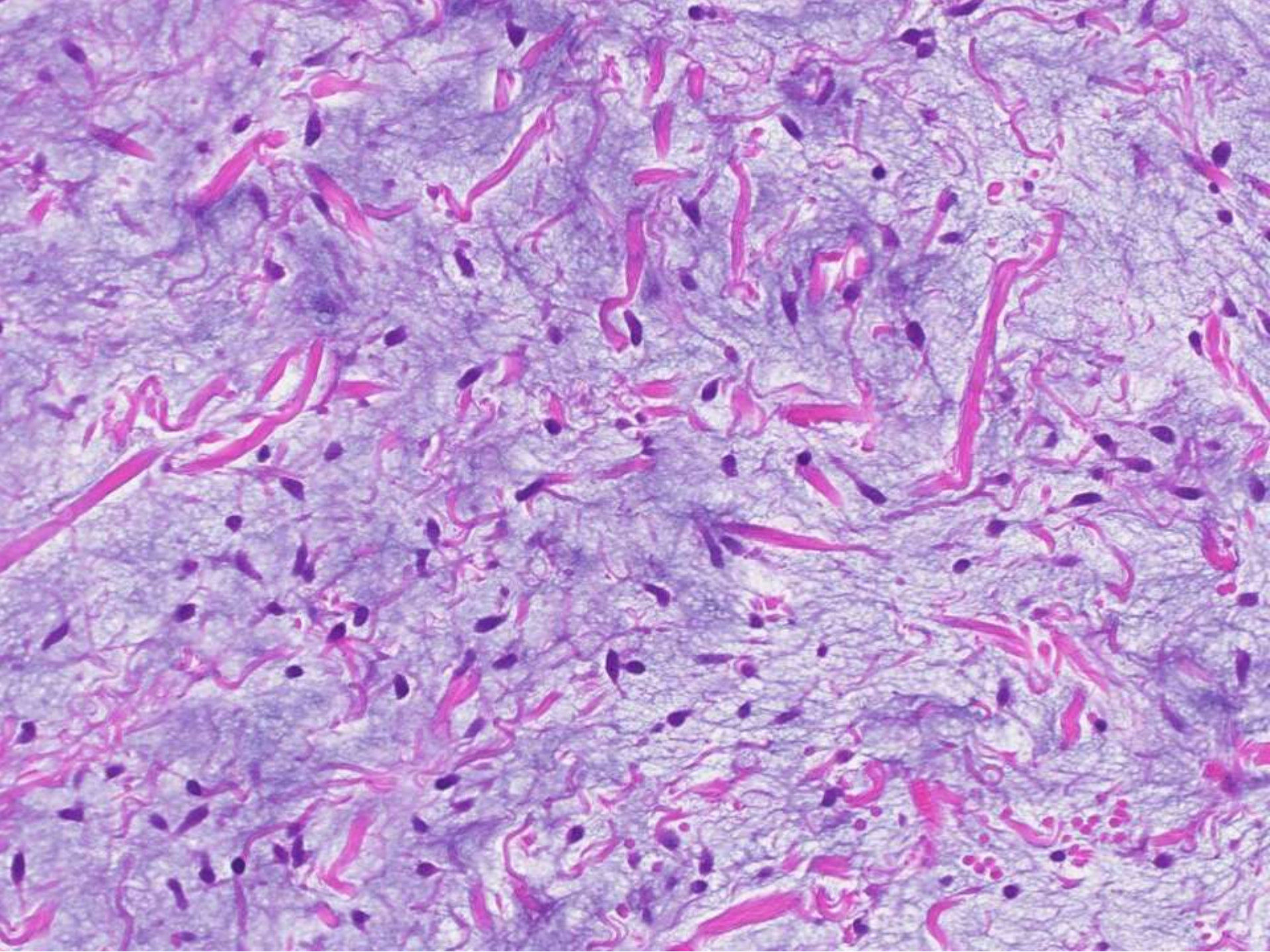












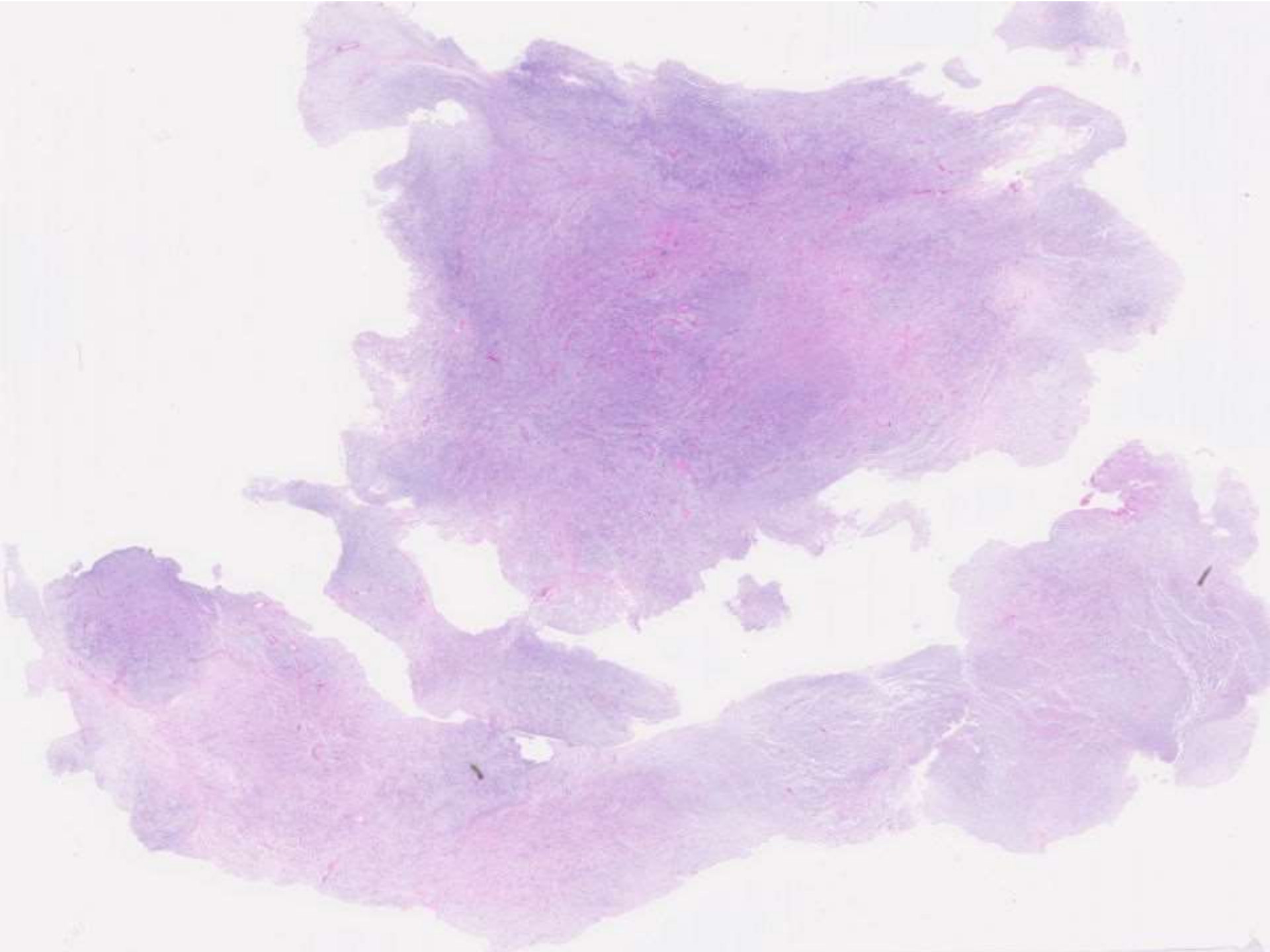
?

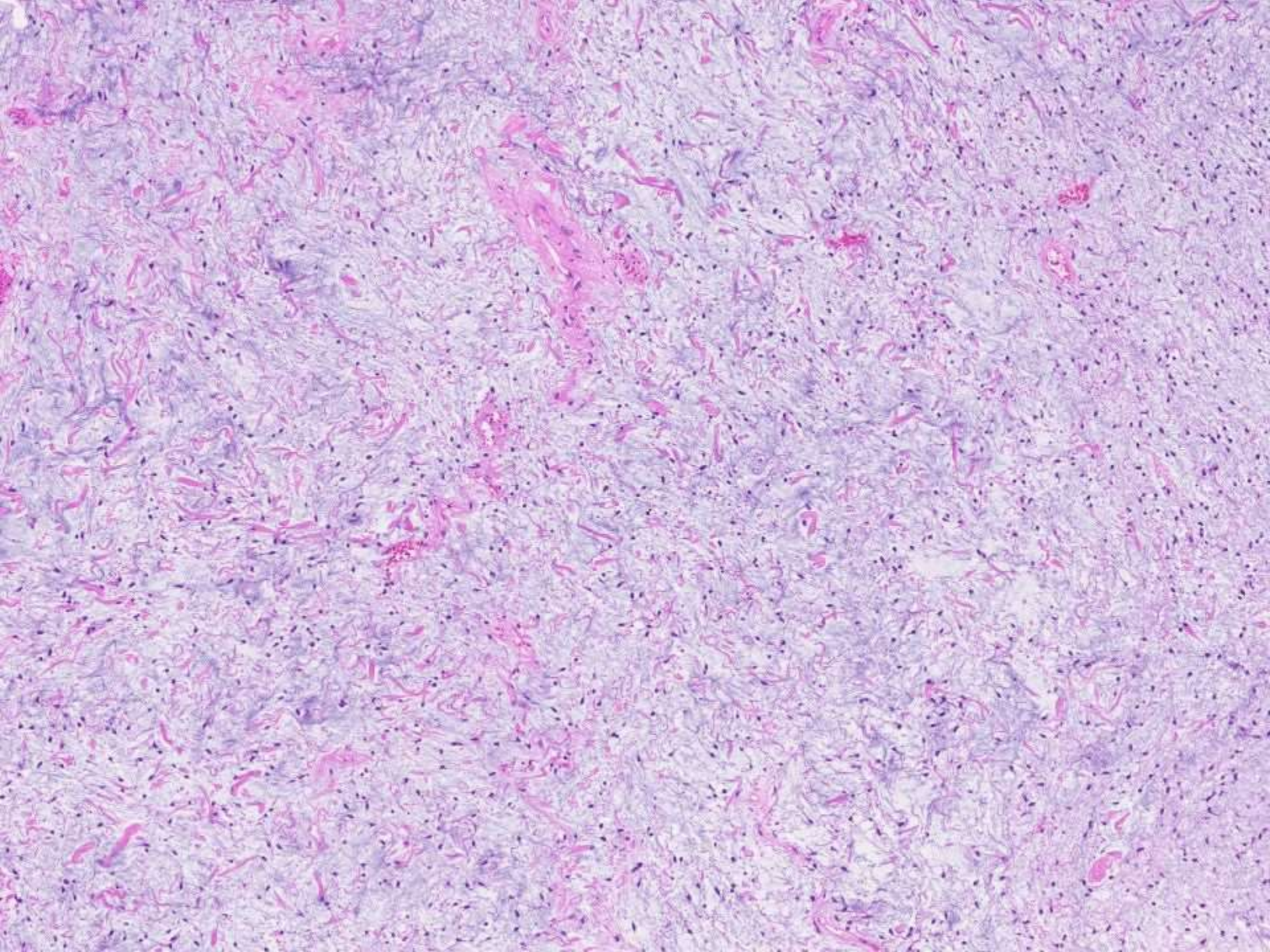
Diagnosis

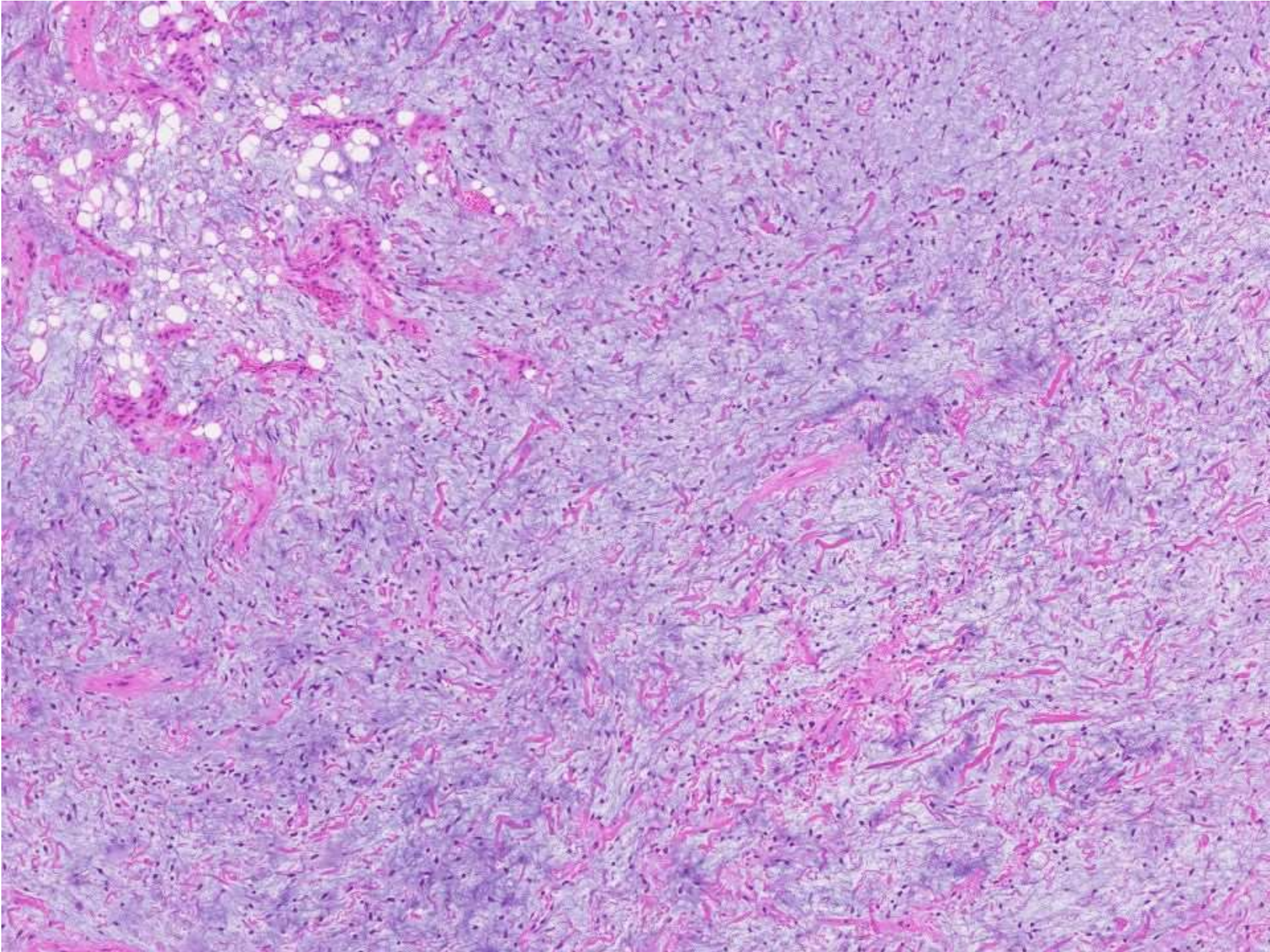
Spindle cell lipoma

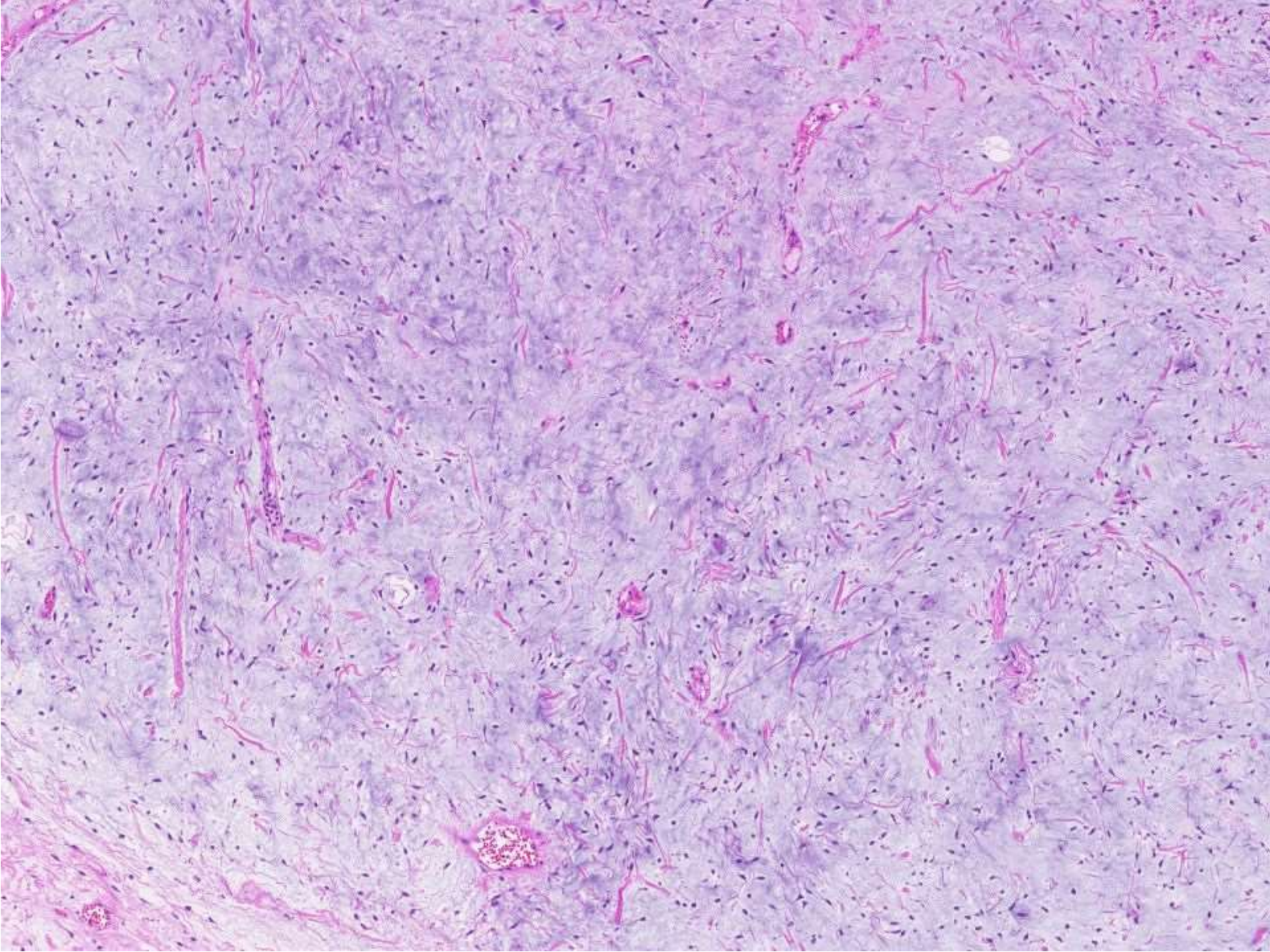
Spindle cell lipoma

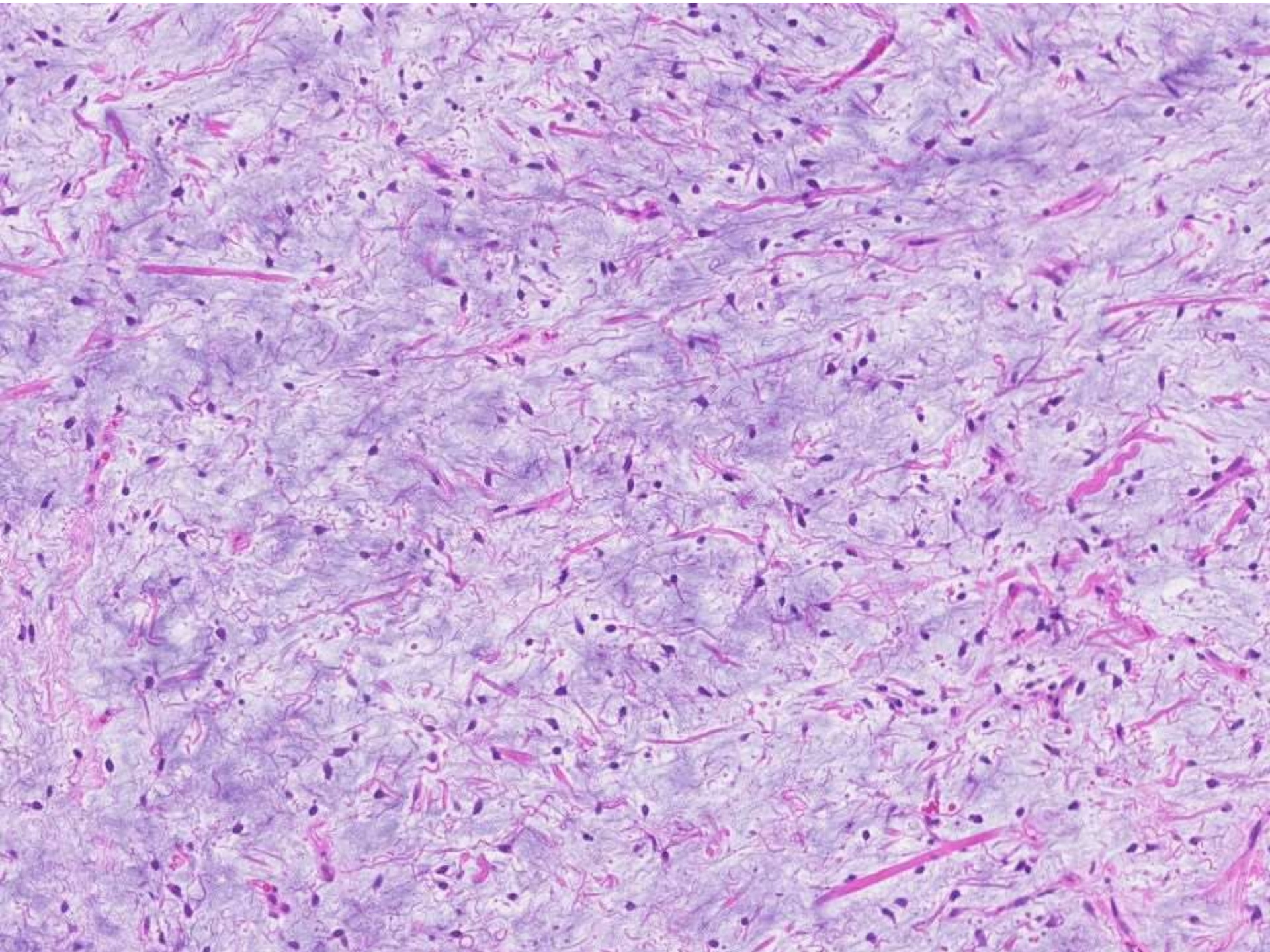
- M >> F
- 80%が後頸部・肩・背部
- 皮下発生が多い
- 多くは～5 cm
- 脂肪は多いことも少ないことも

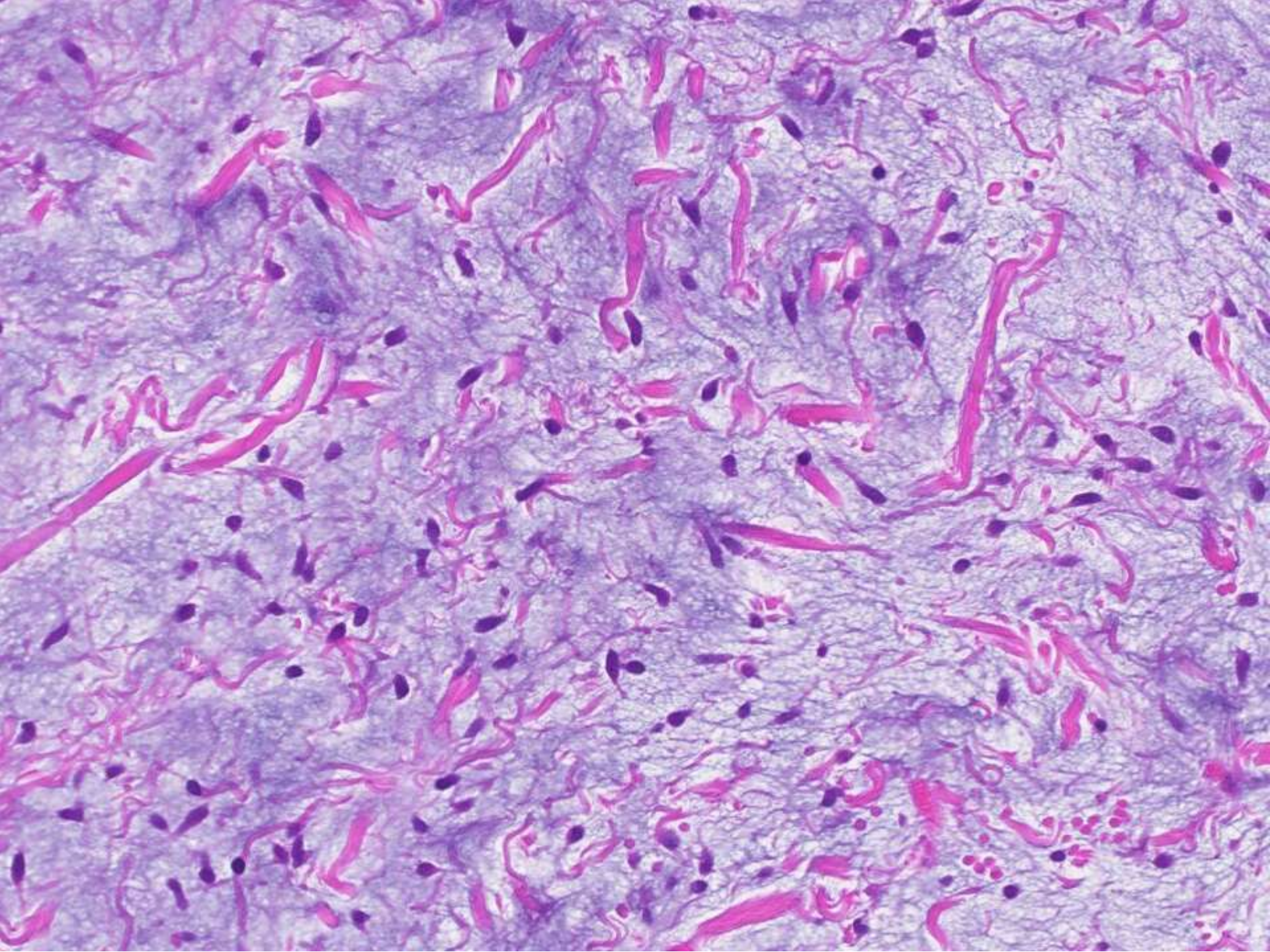




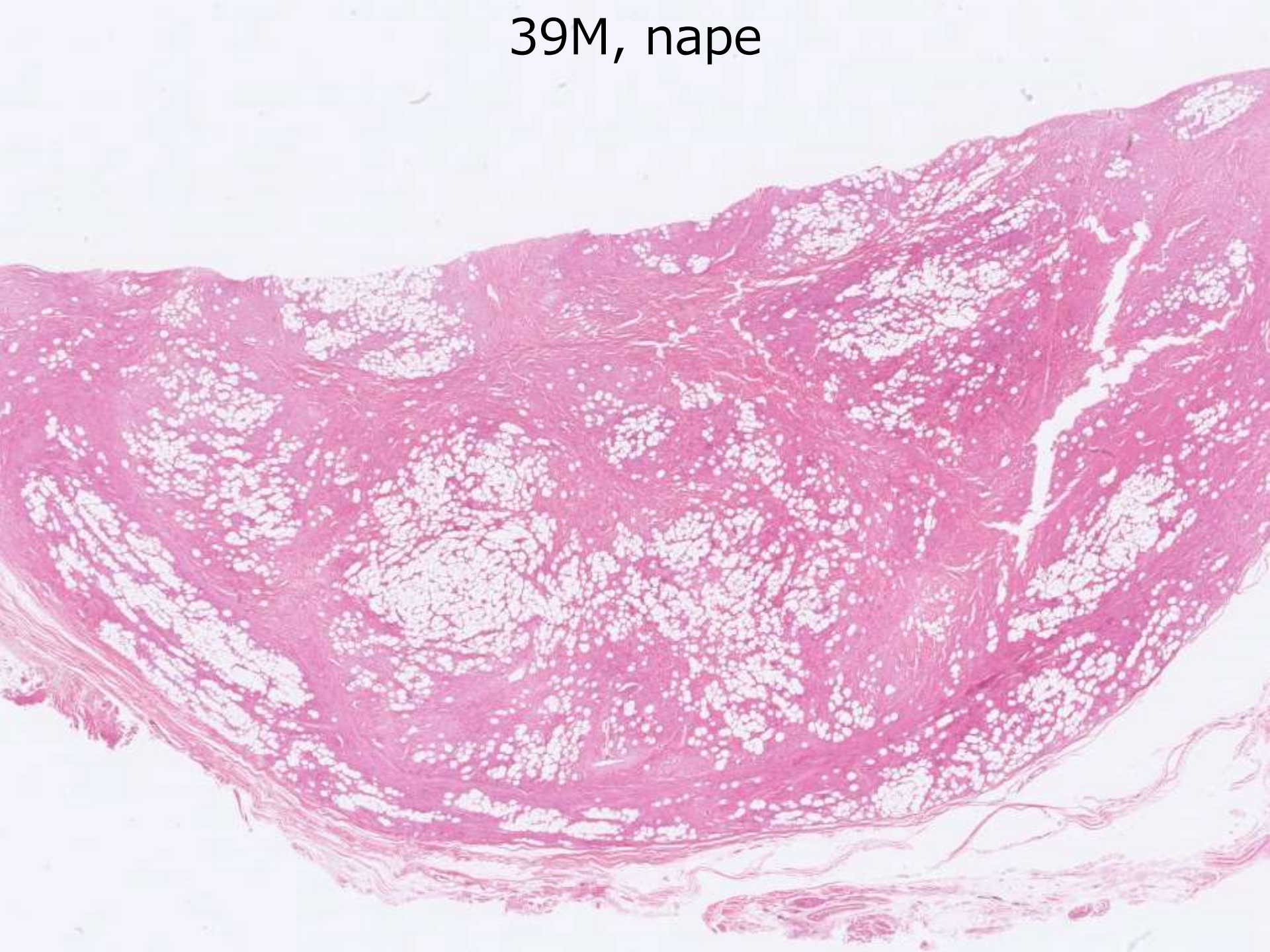


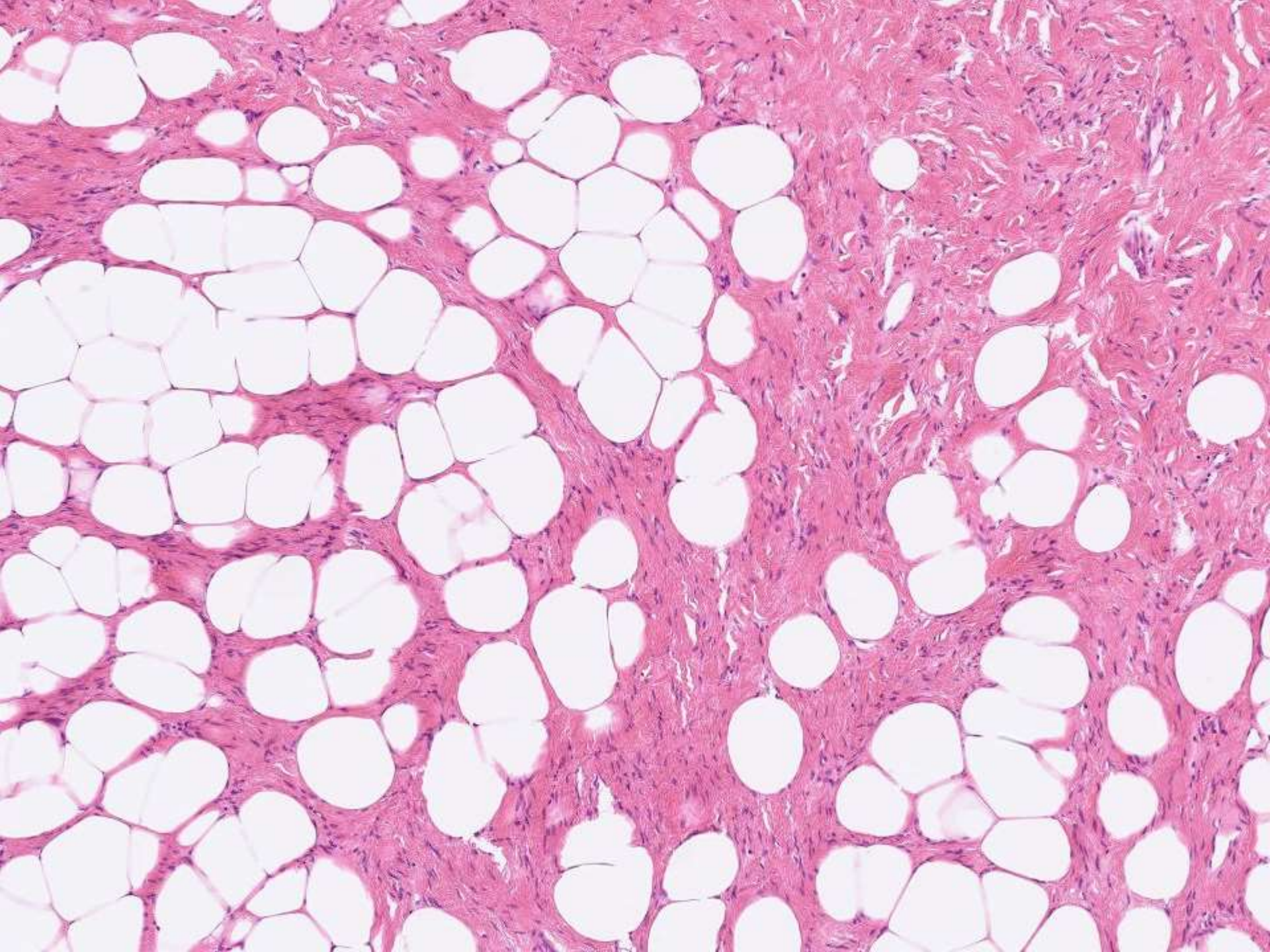


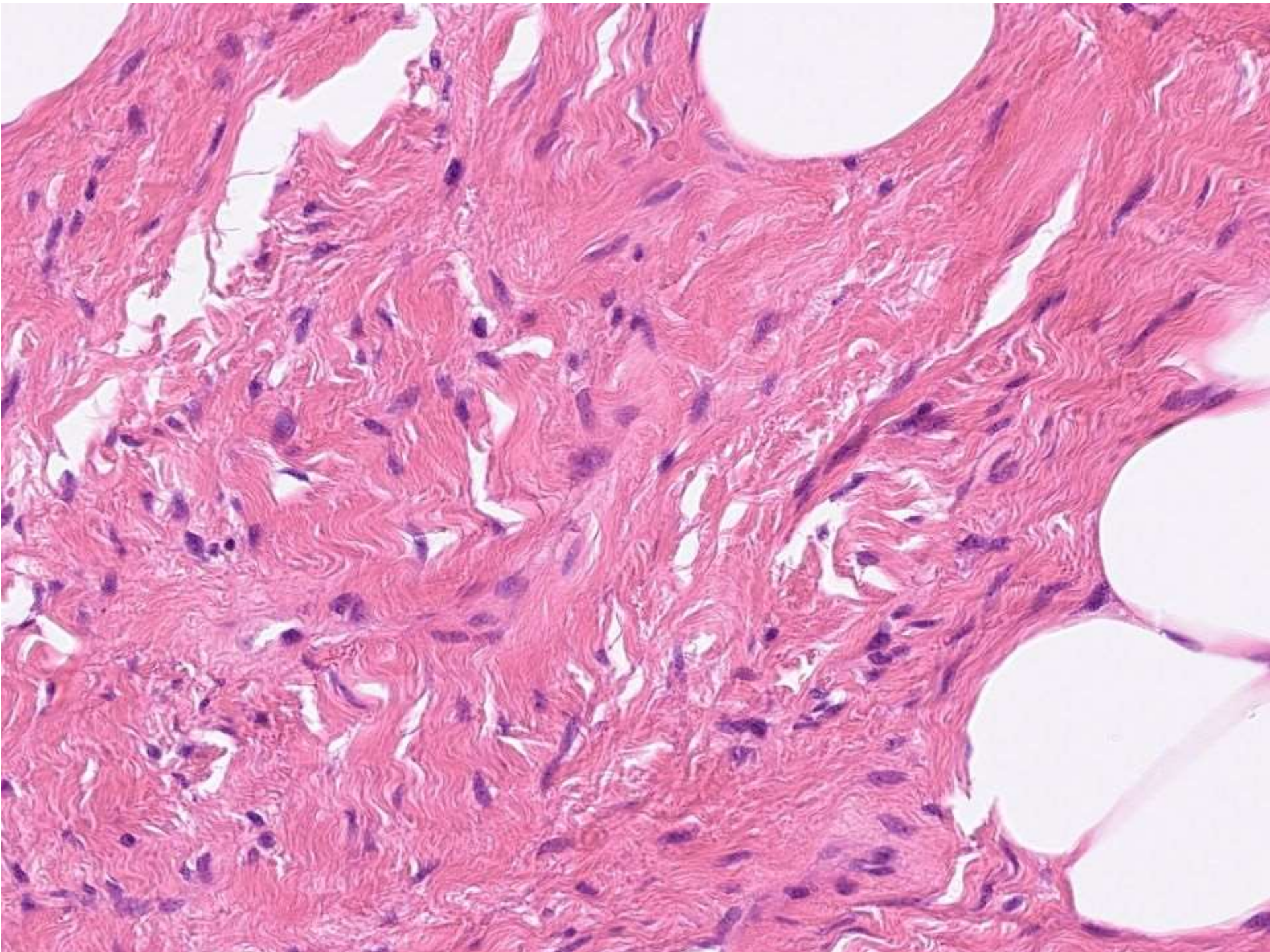


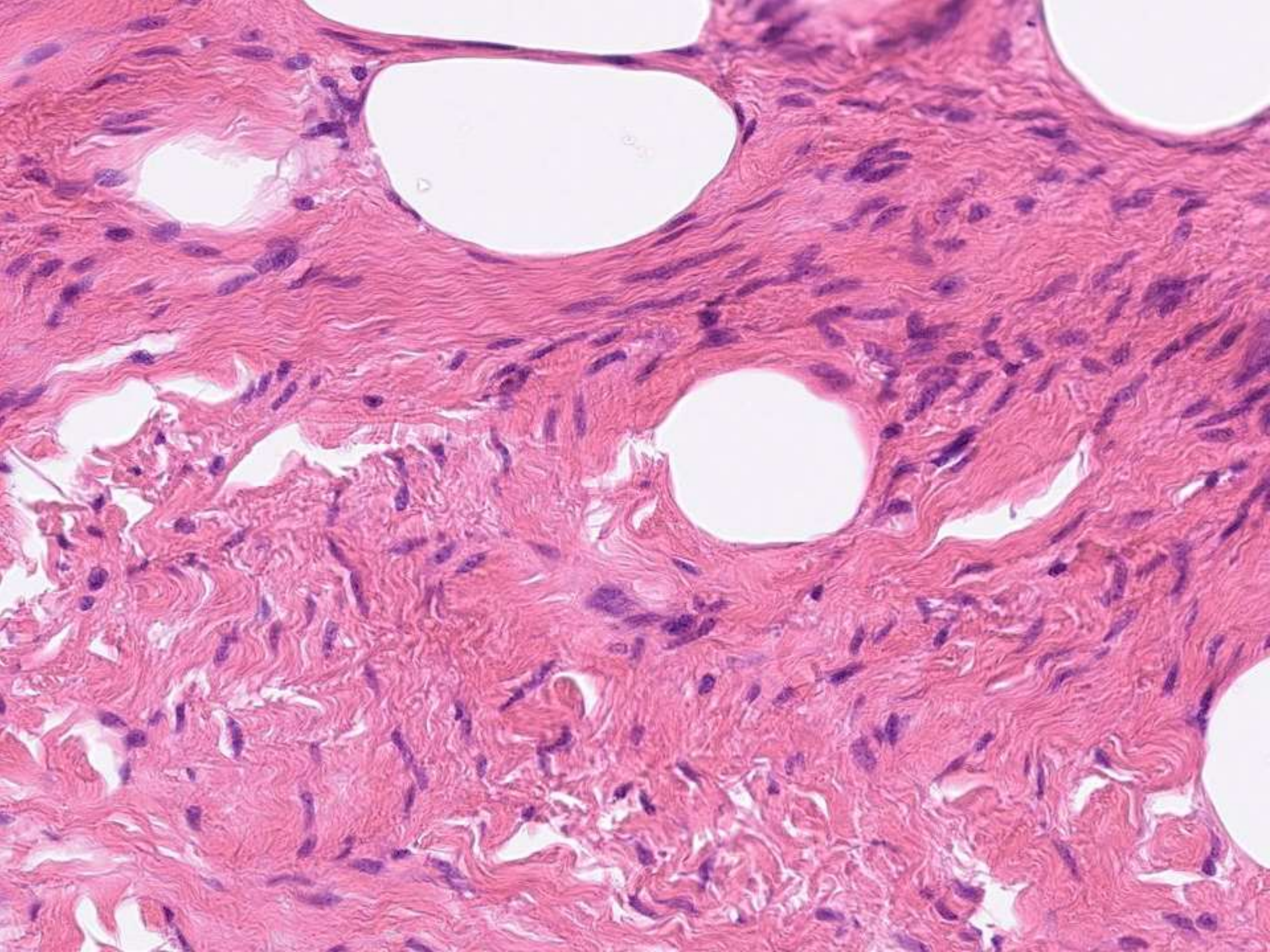


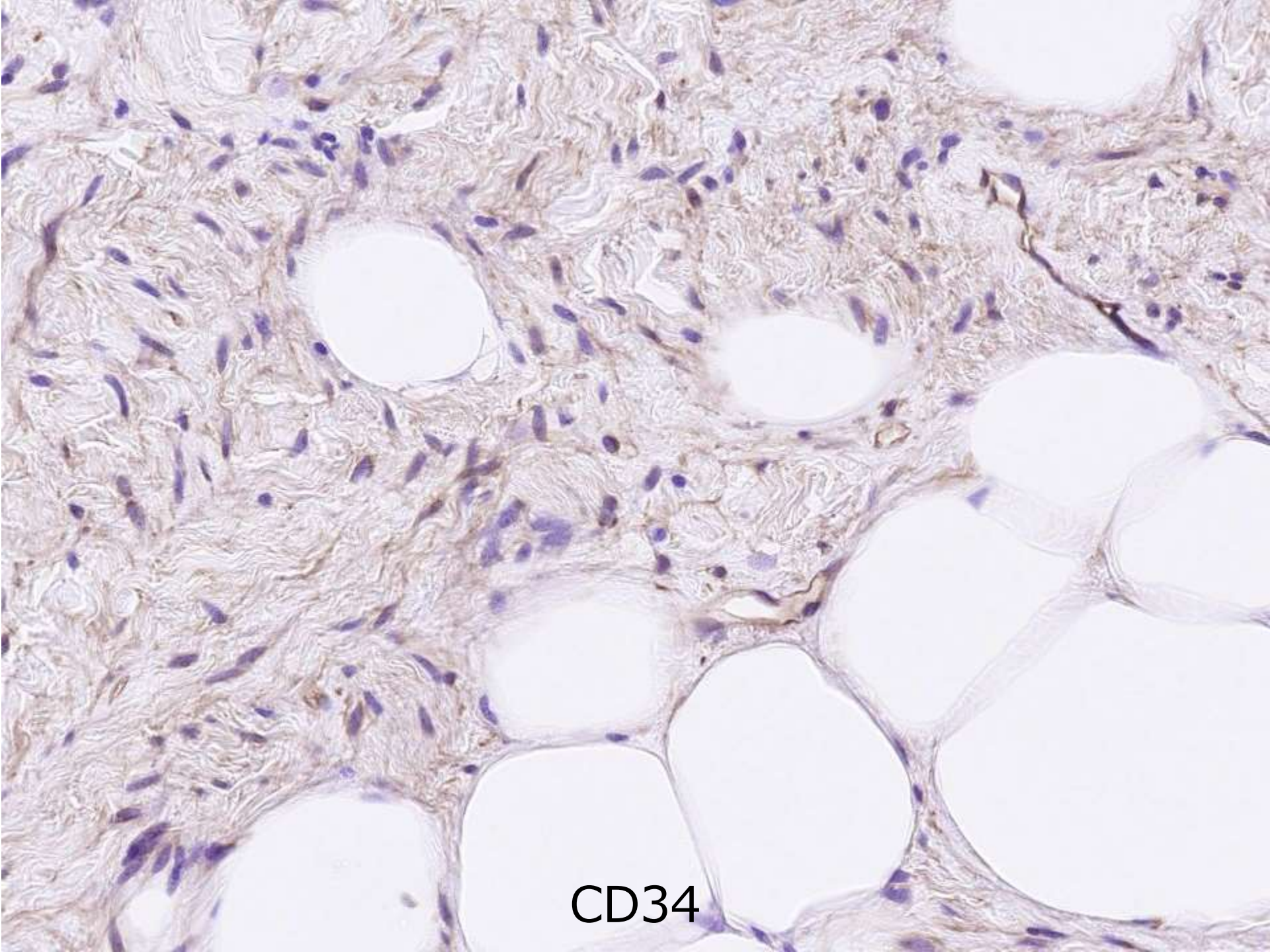
39M, nape

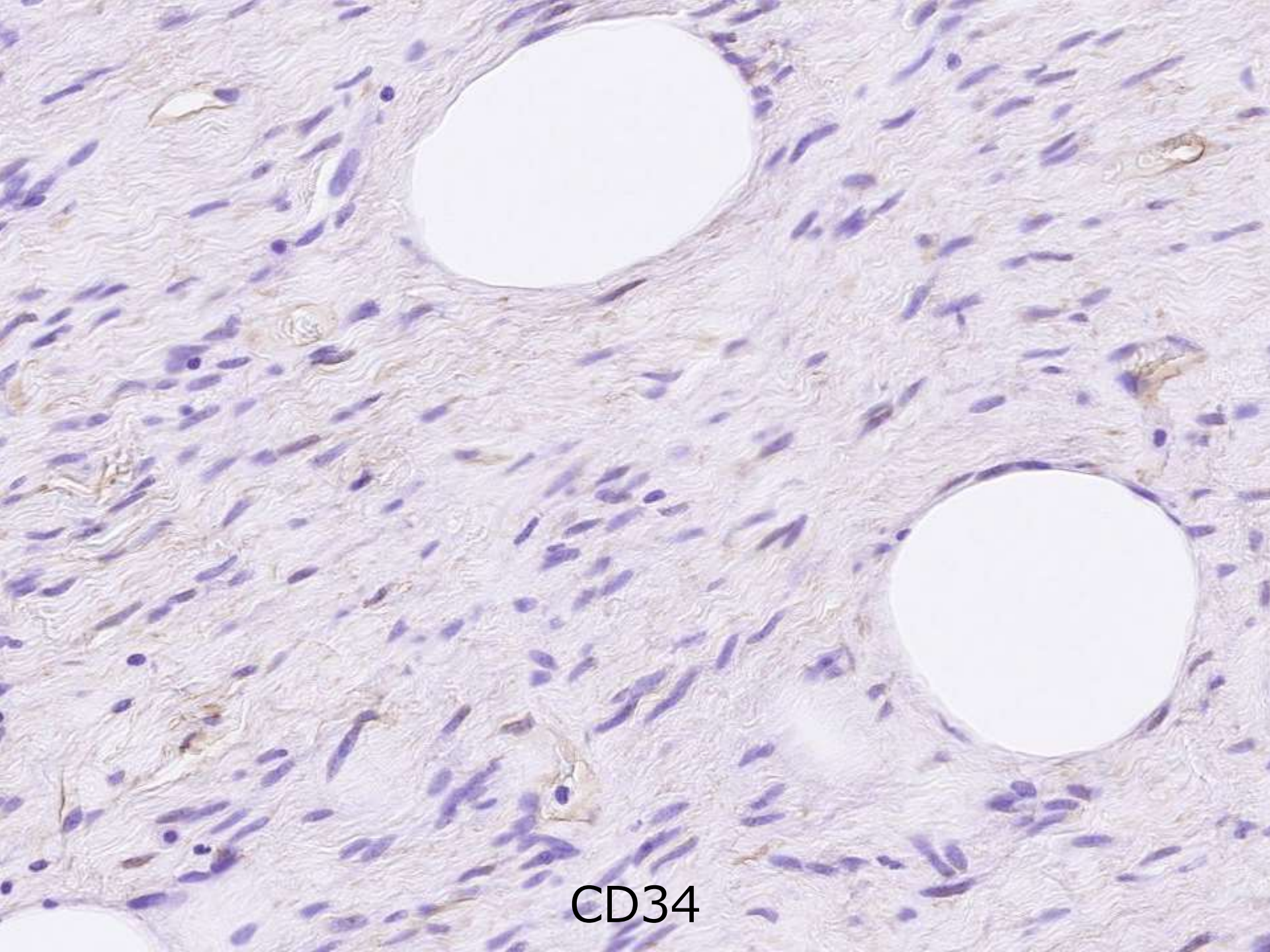









CD34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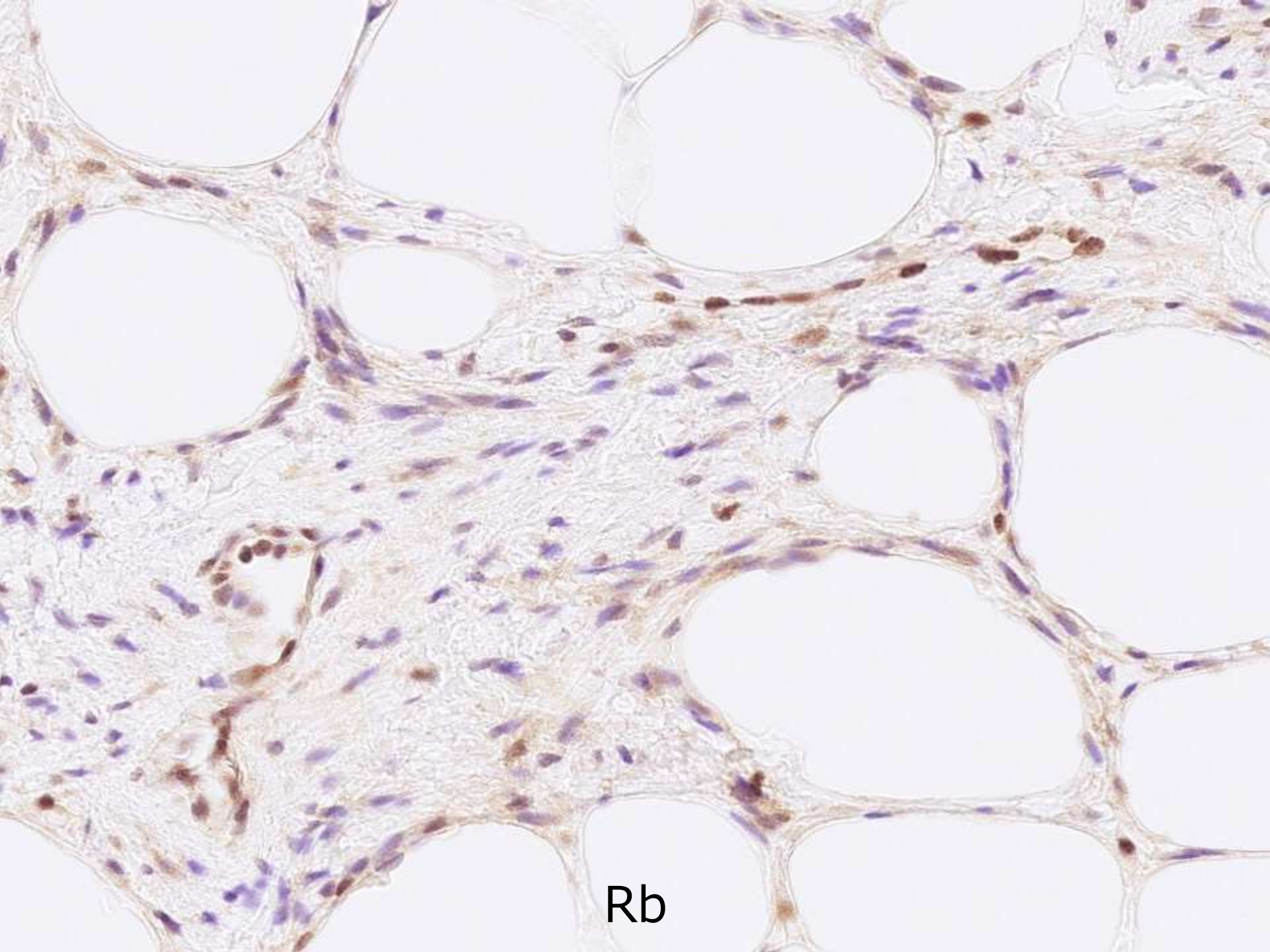
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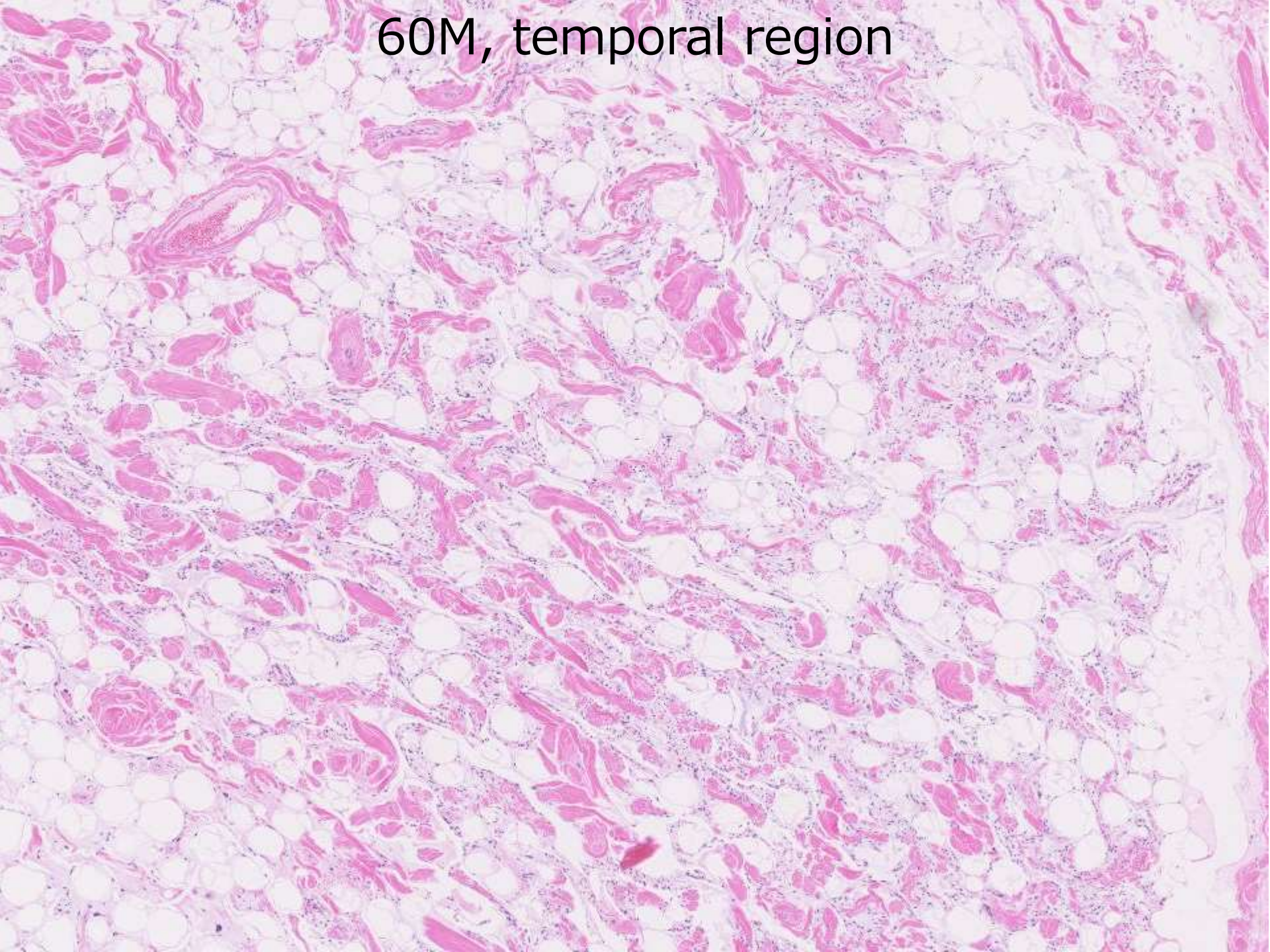
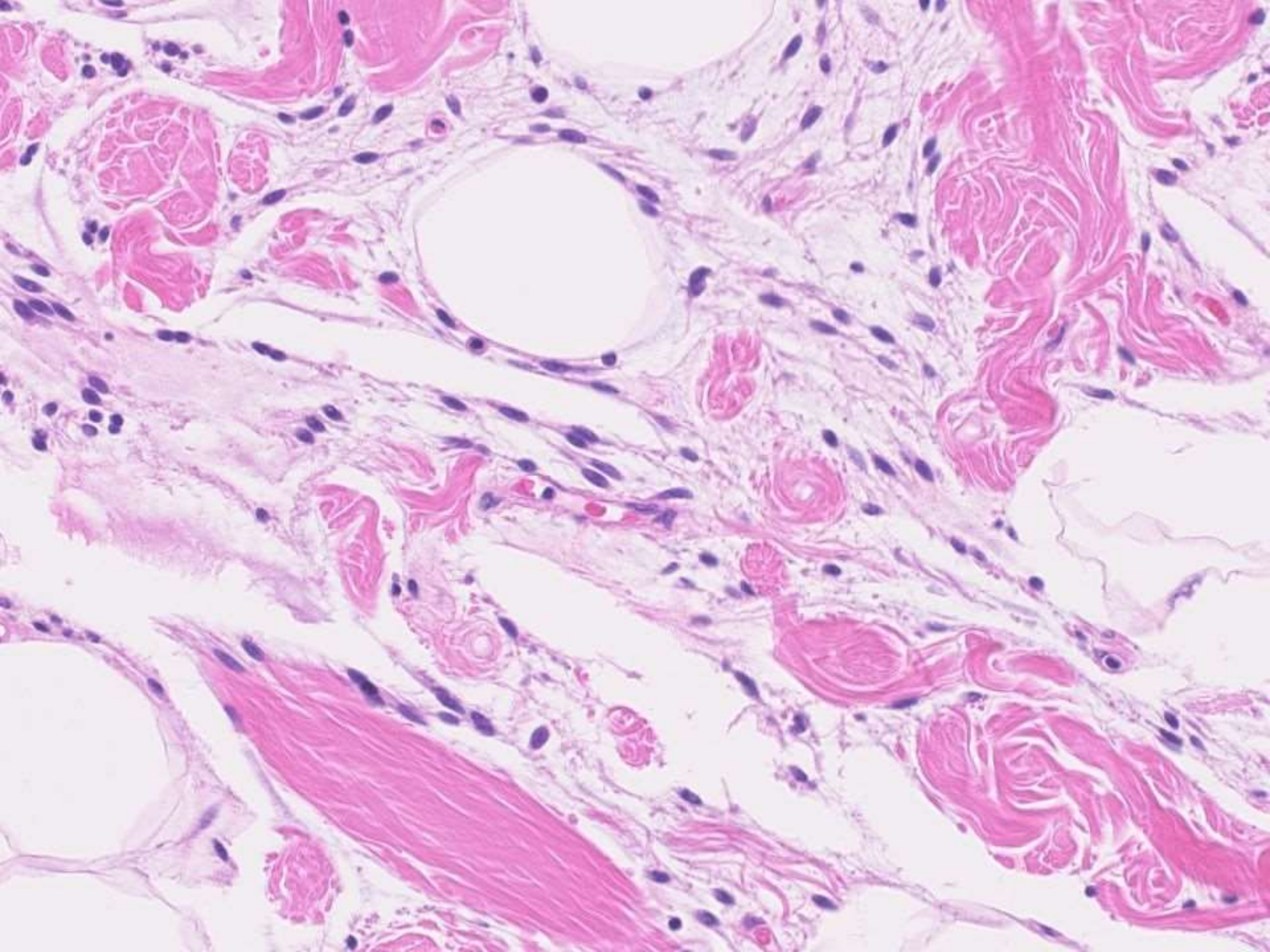
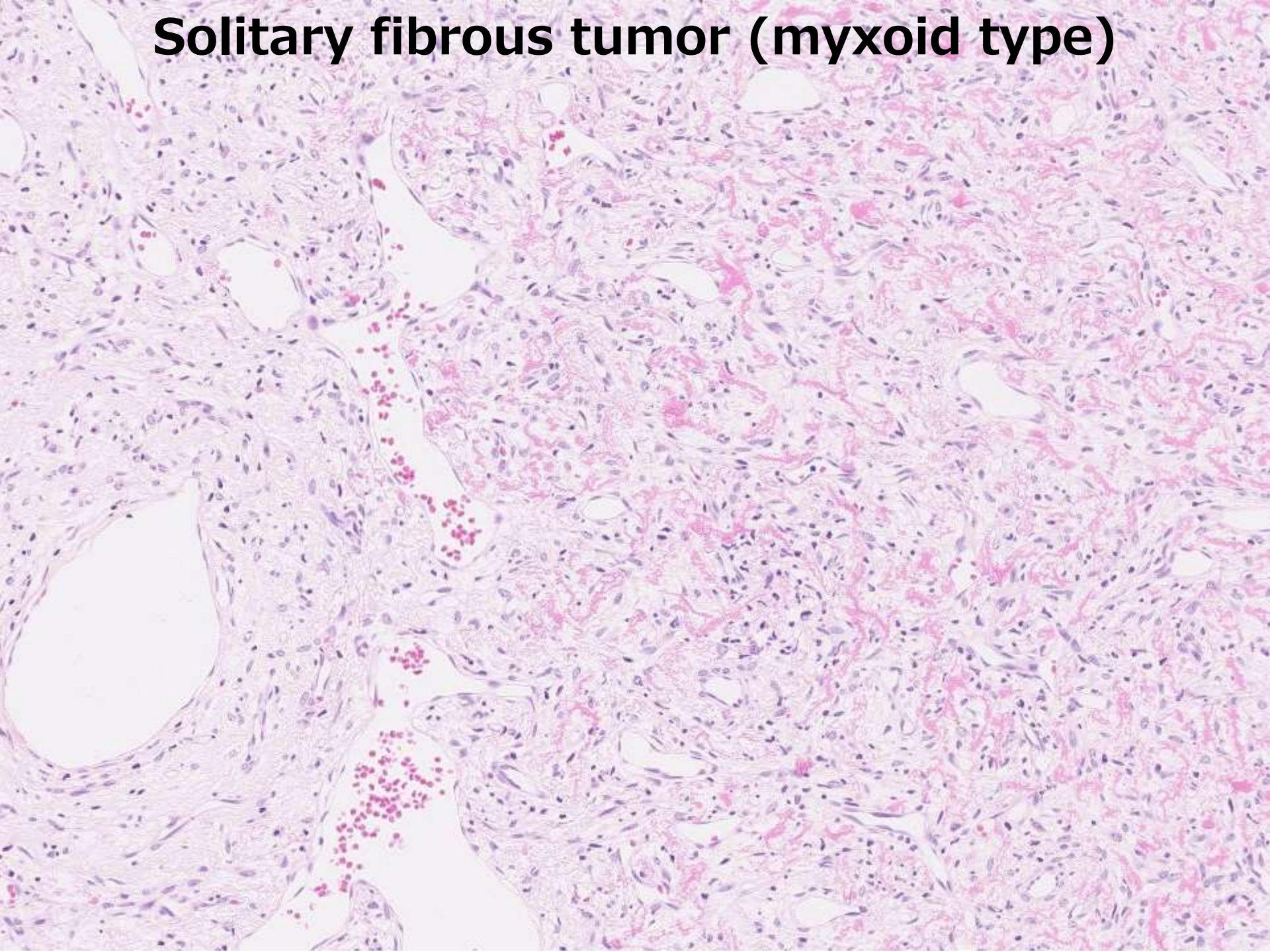
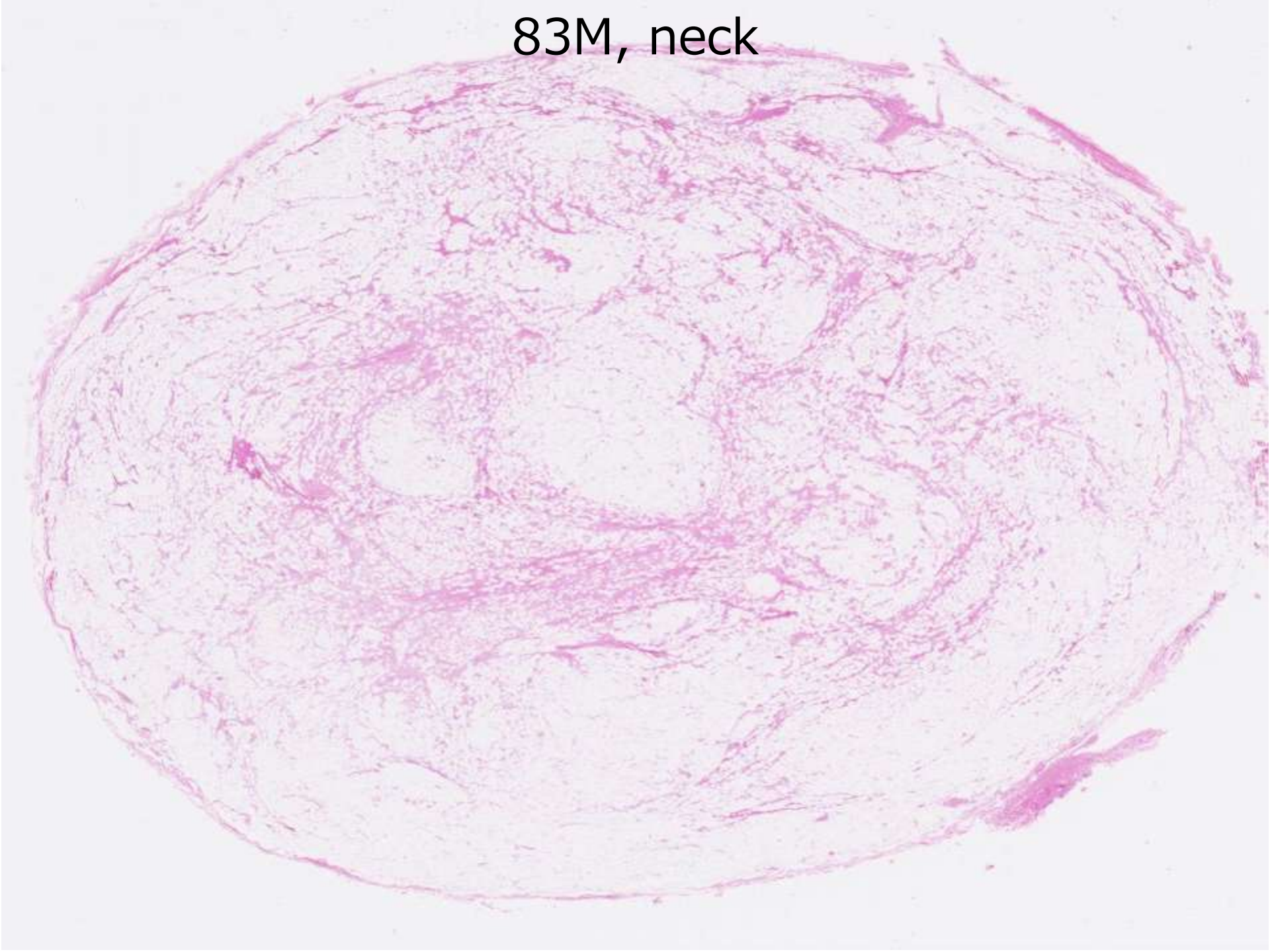

60M, temporal region

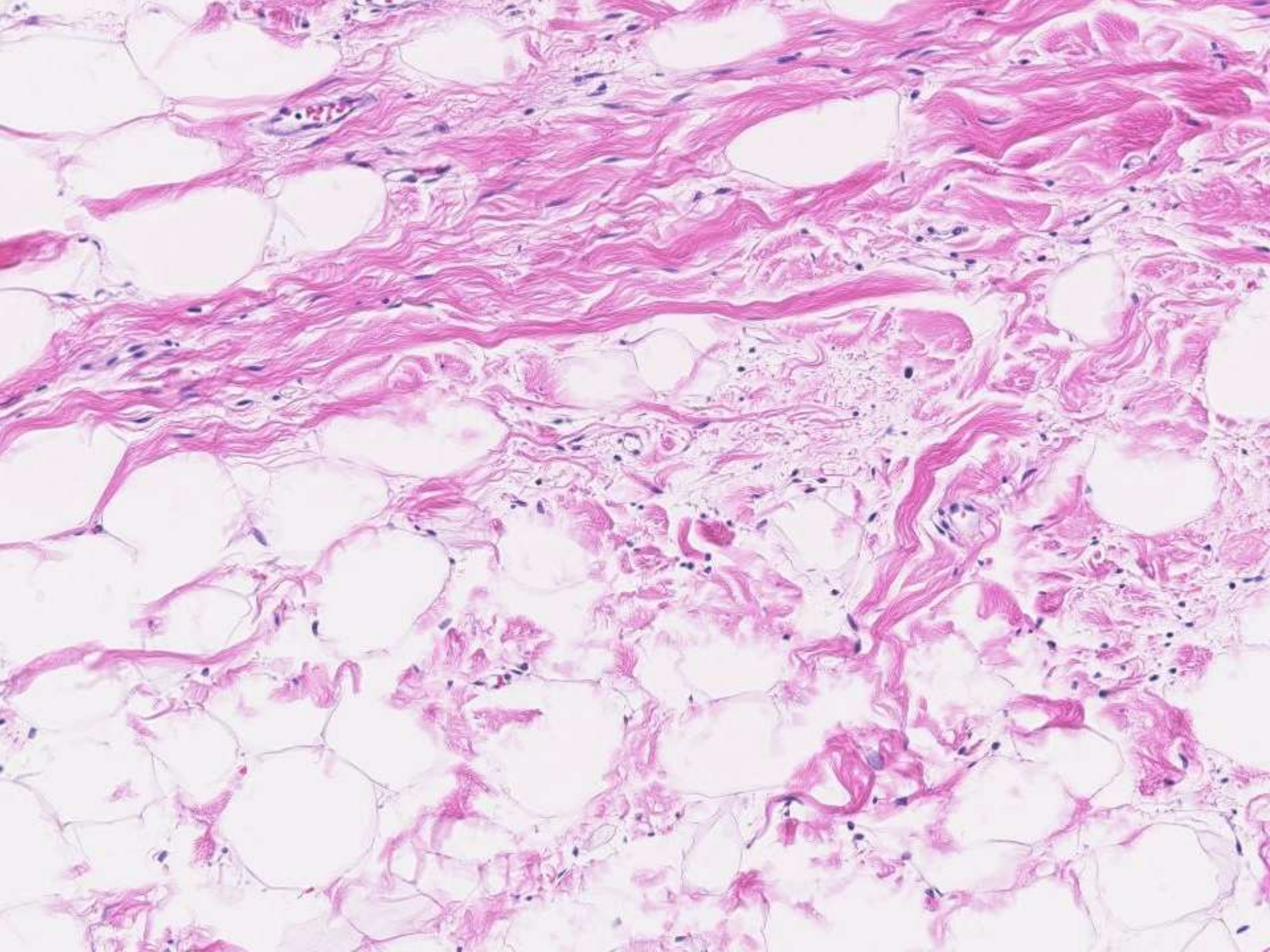


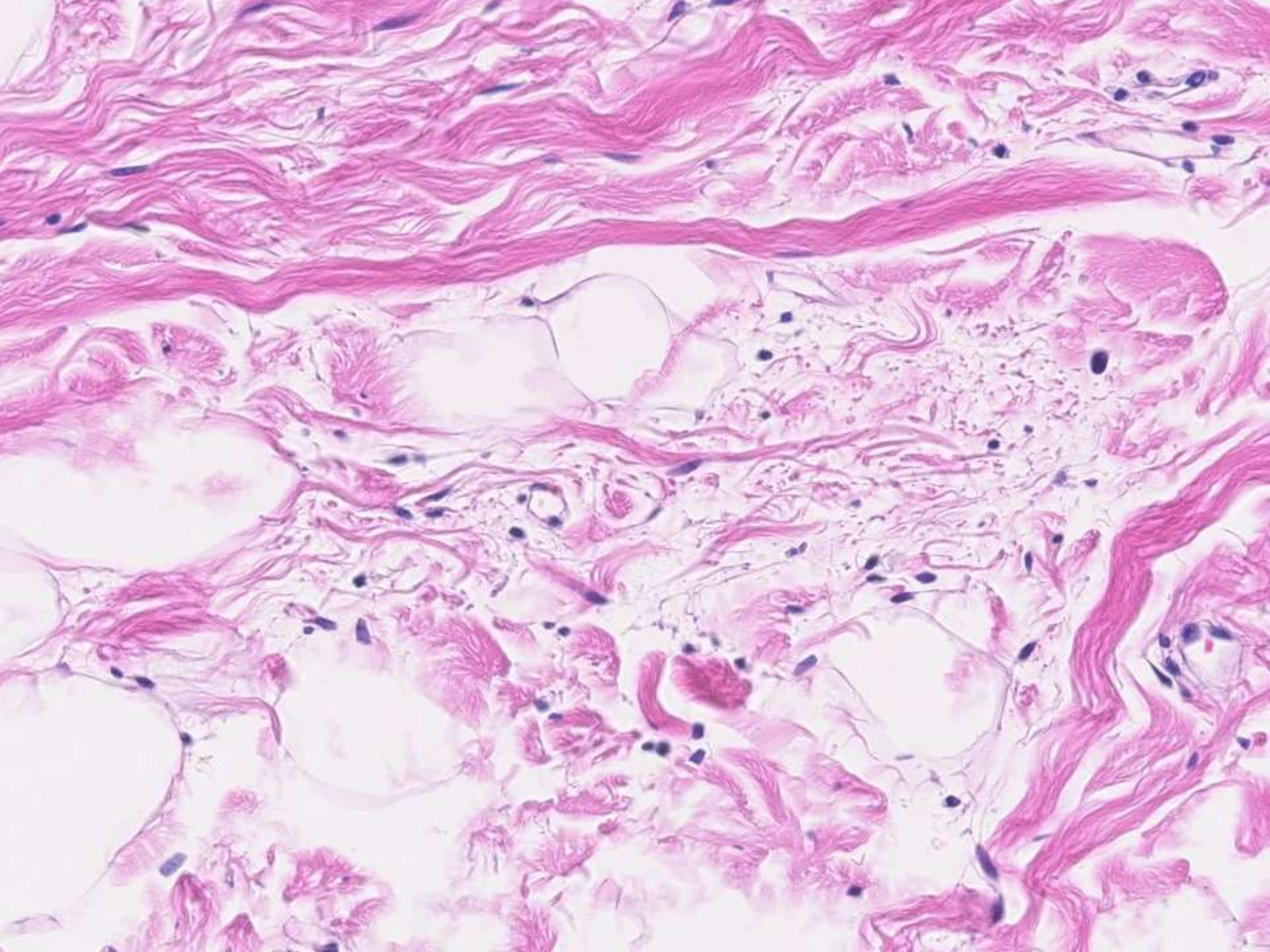
Solitary fibrous tumor (myxoid type)

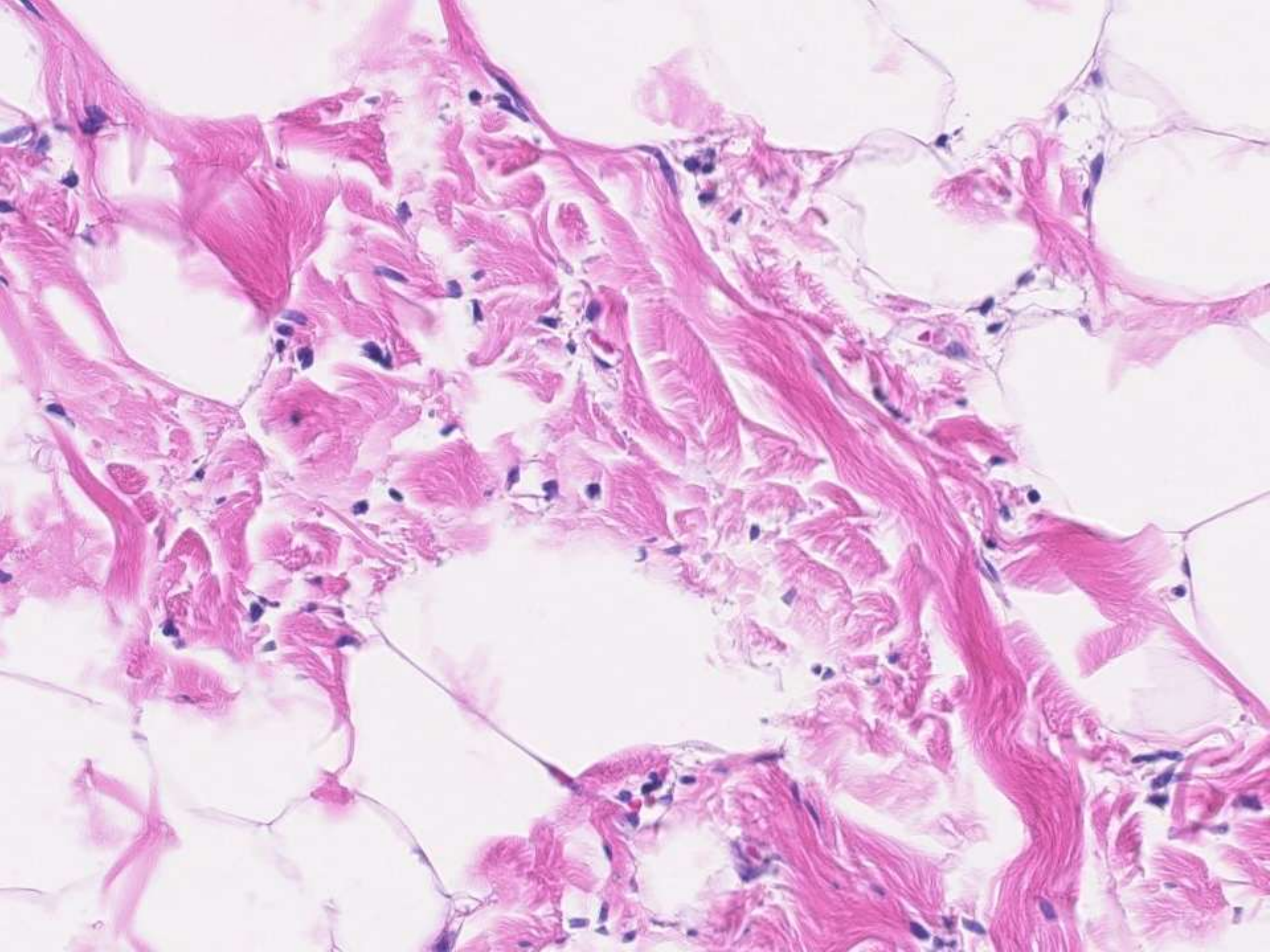


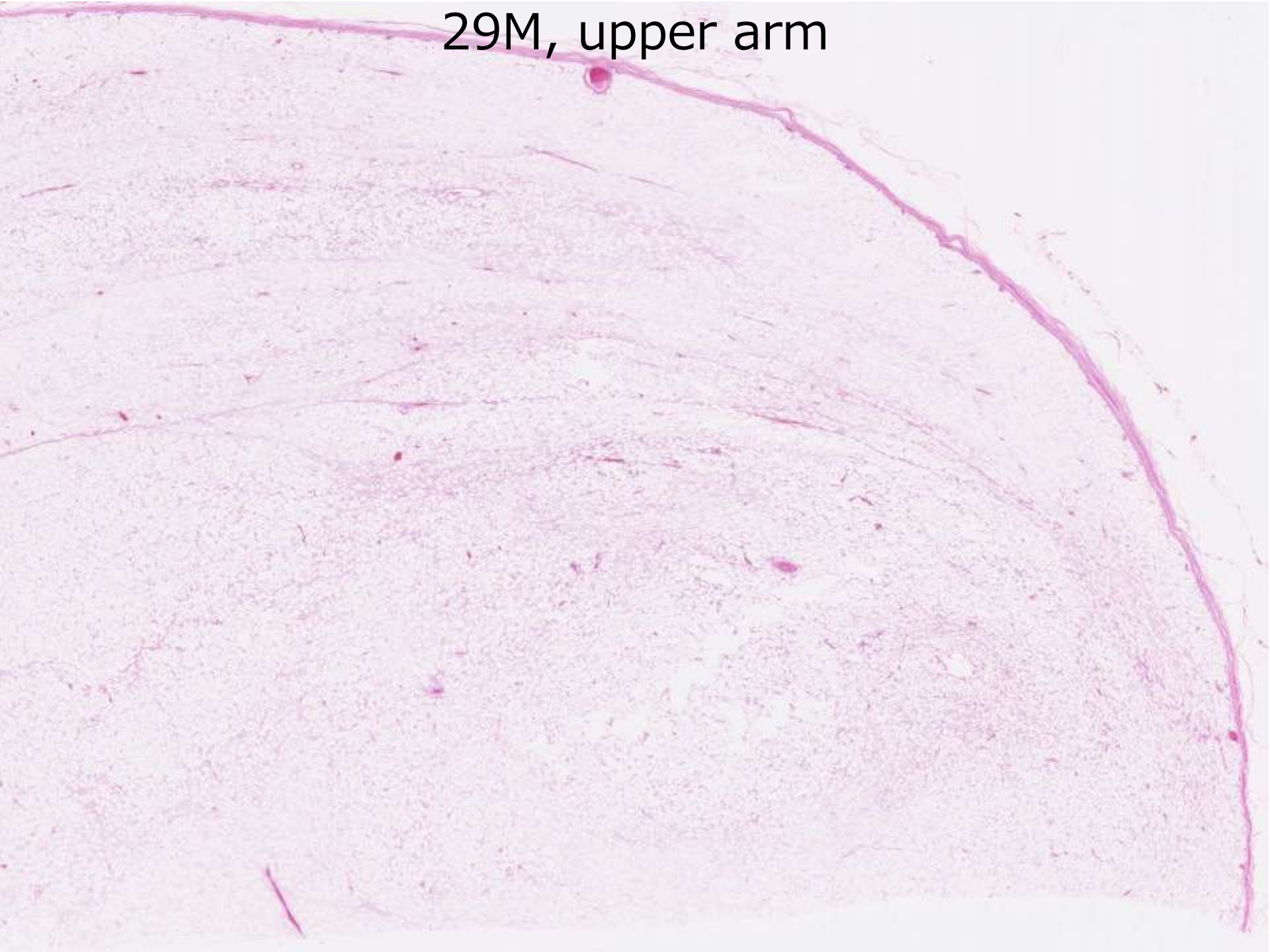
83M, neck

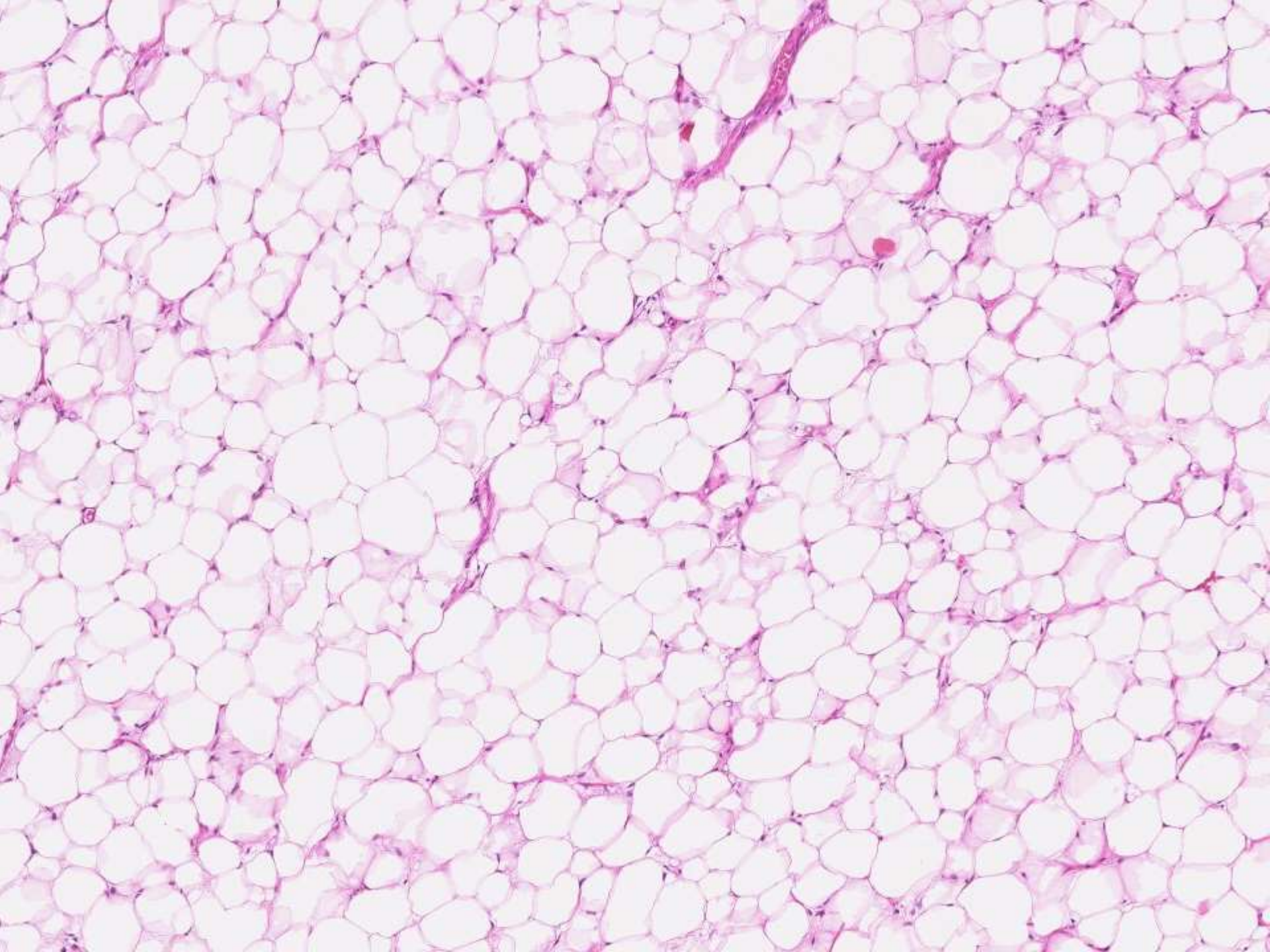


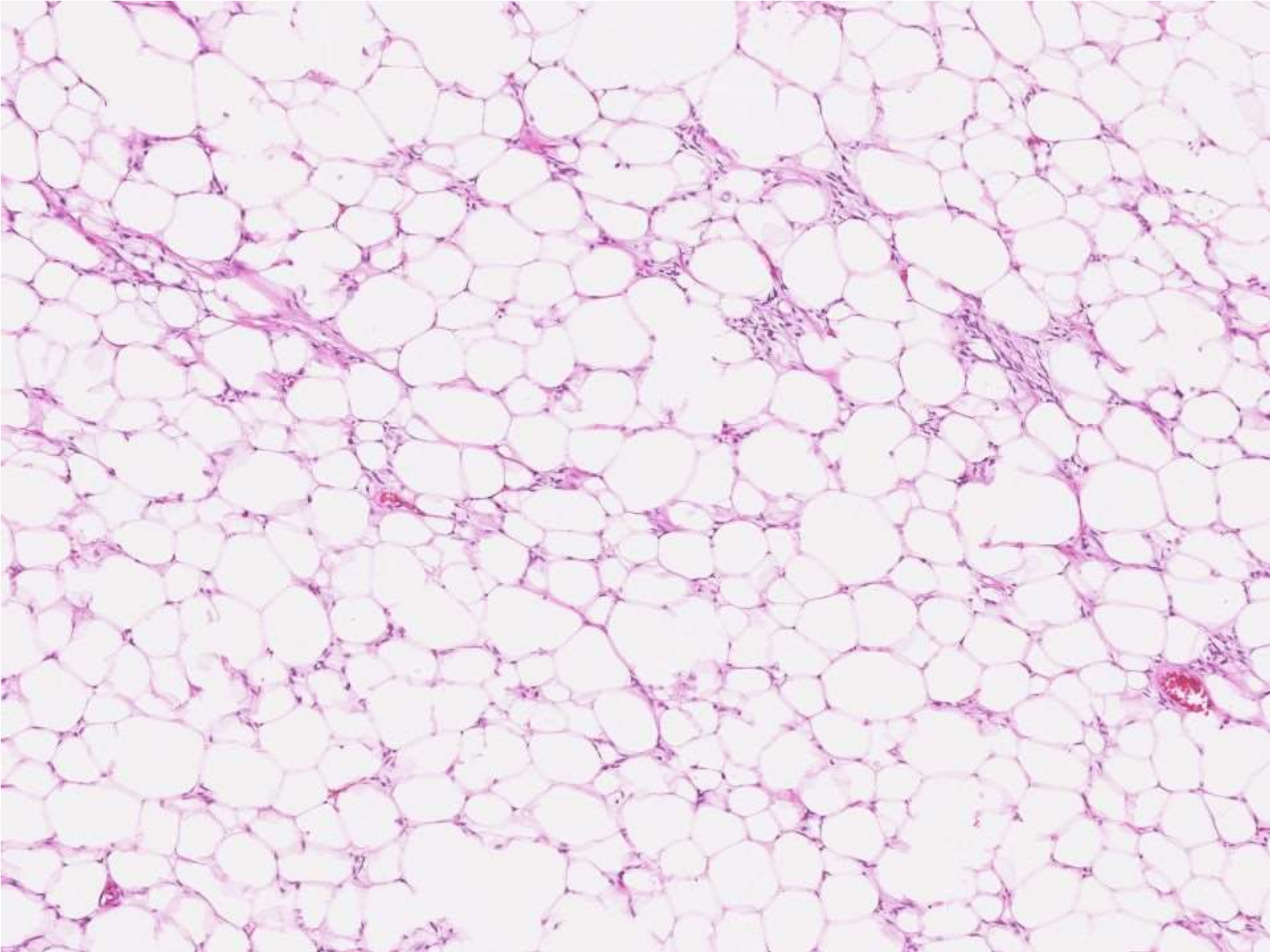


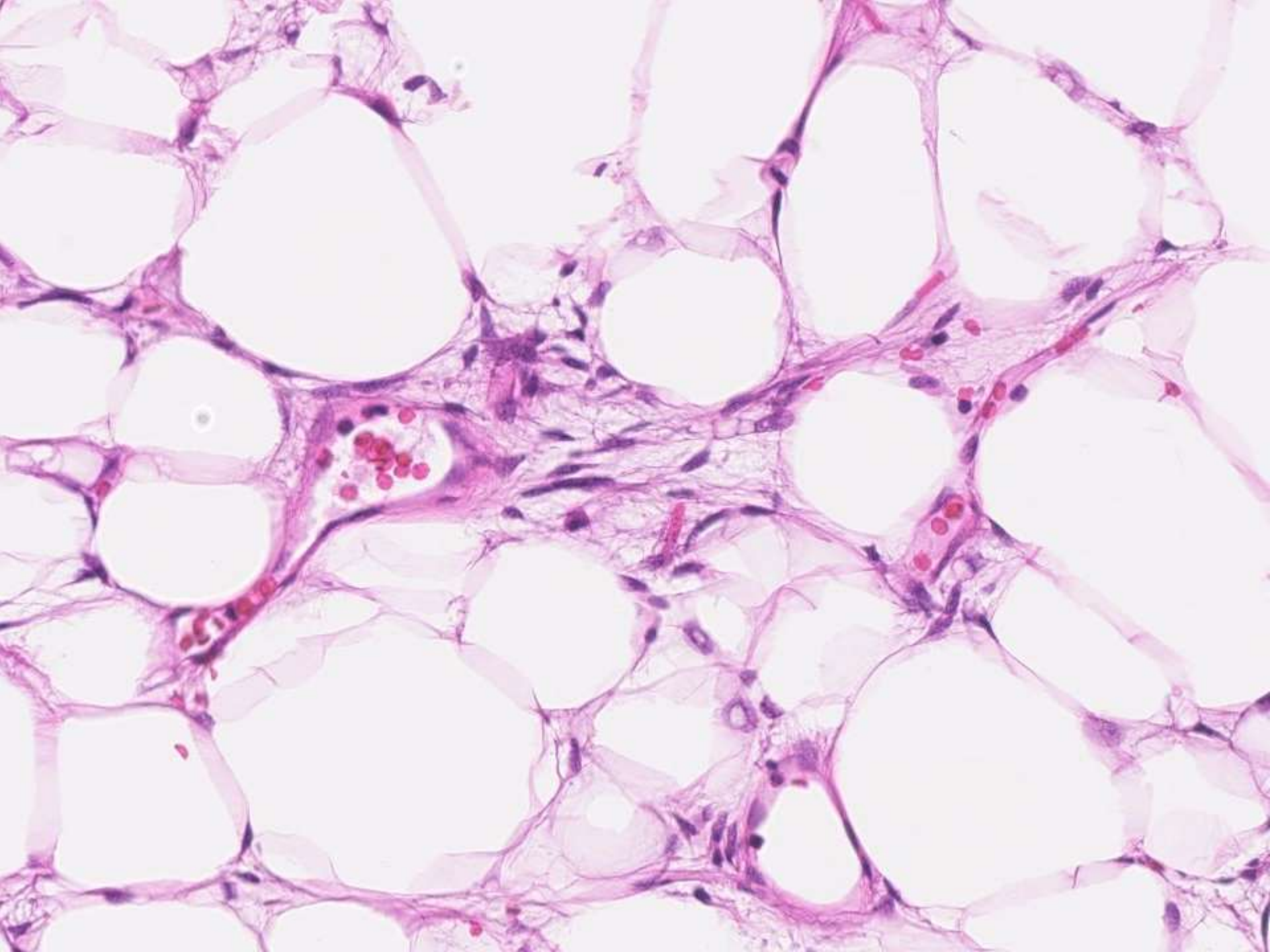


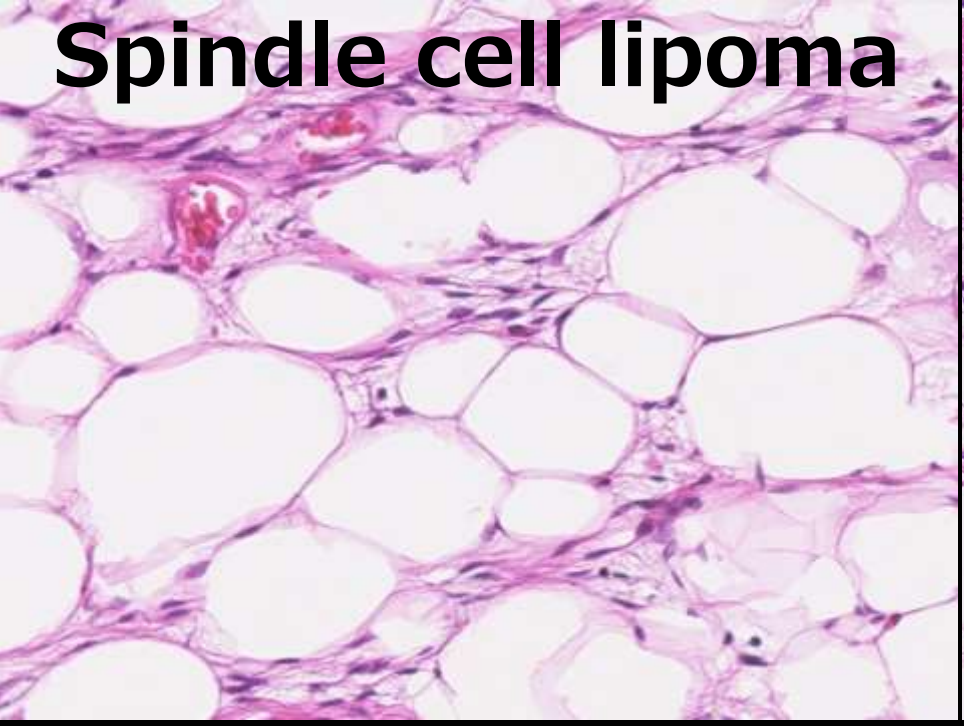


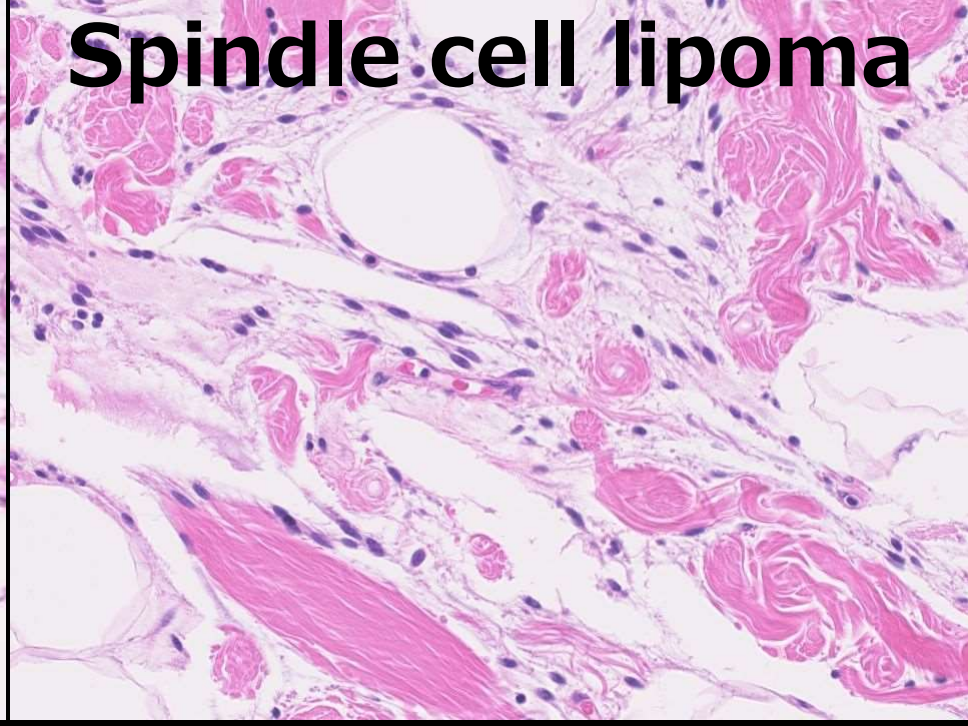

29M, upper arm

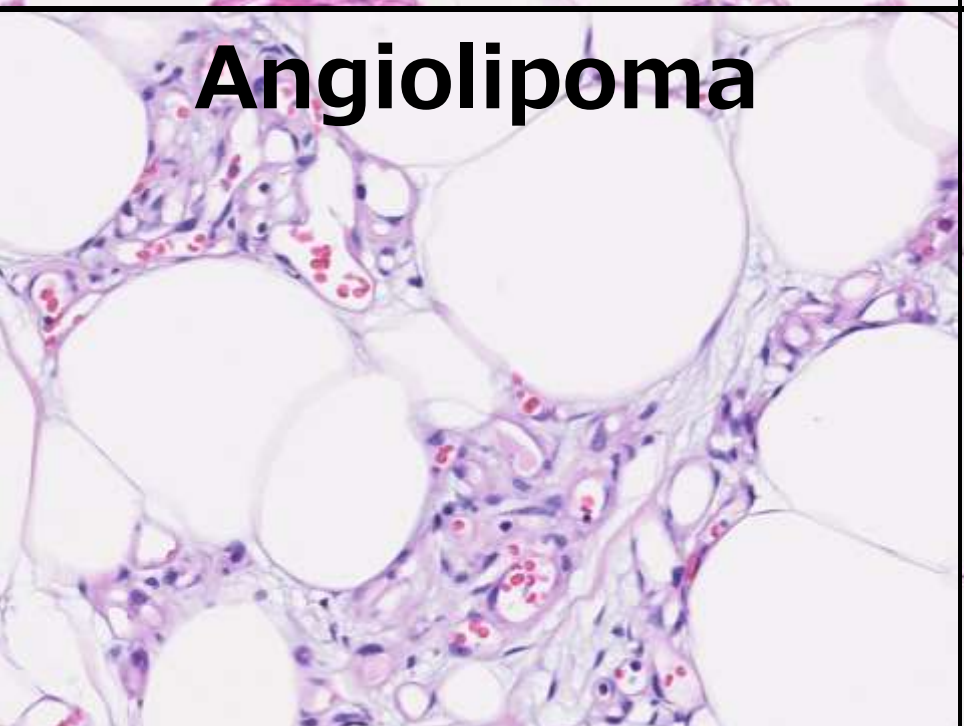


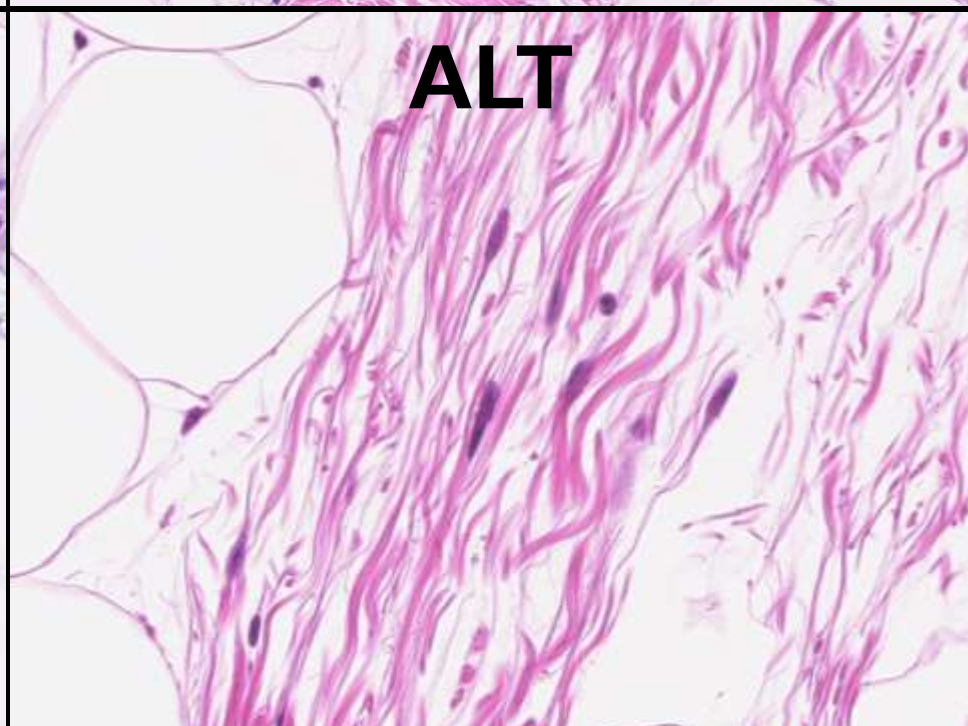




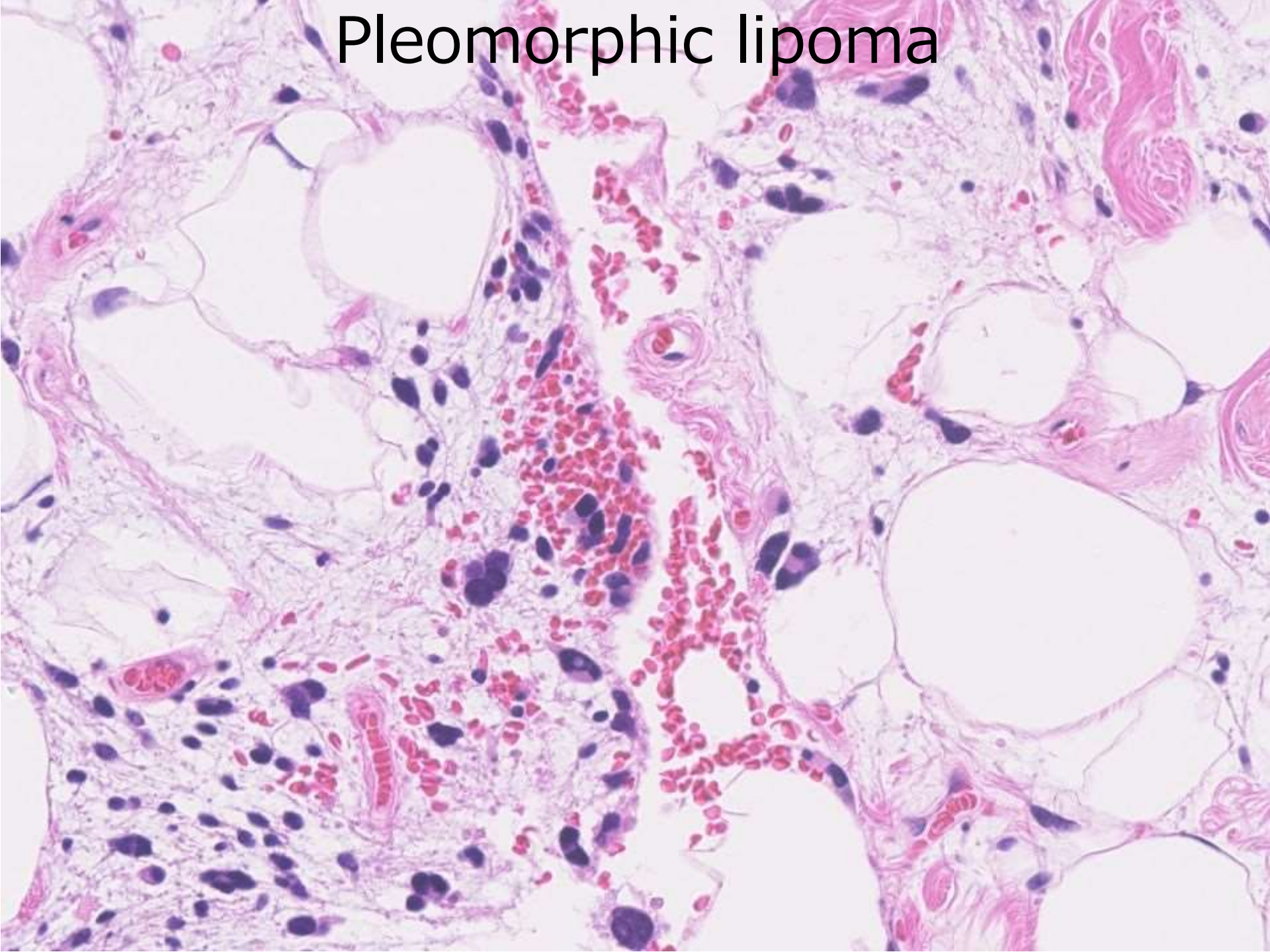

Spindle cell lipoma


Spindle cell lipoma

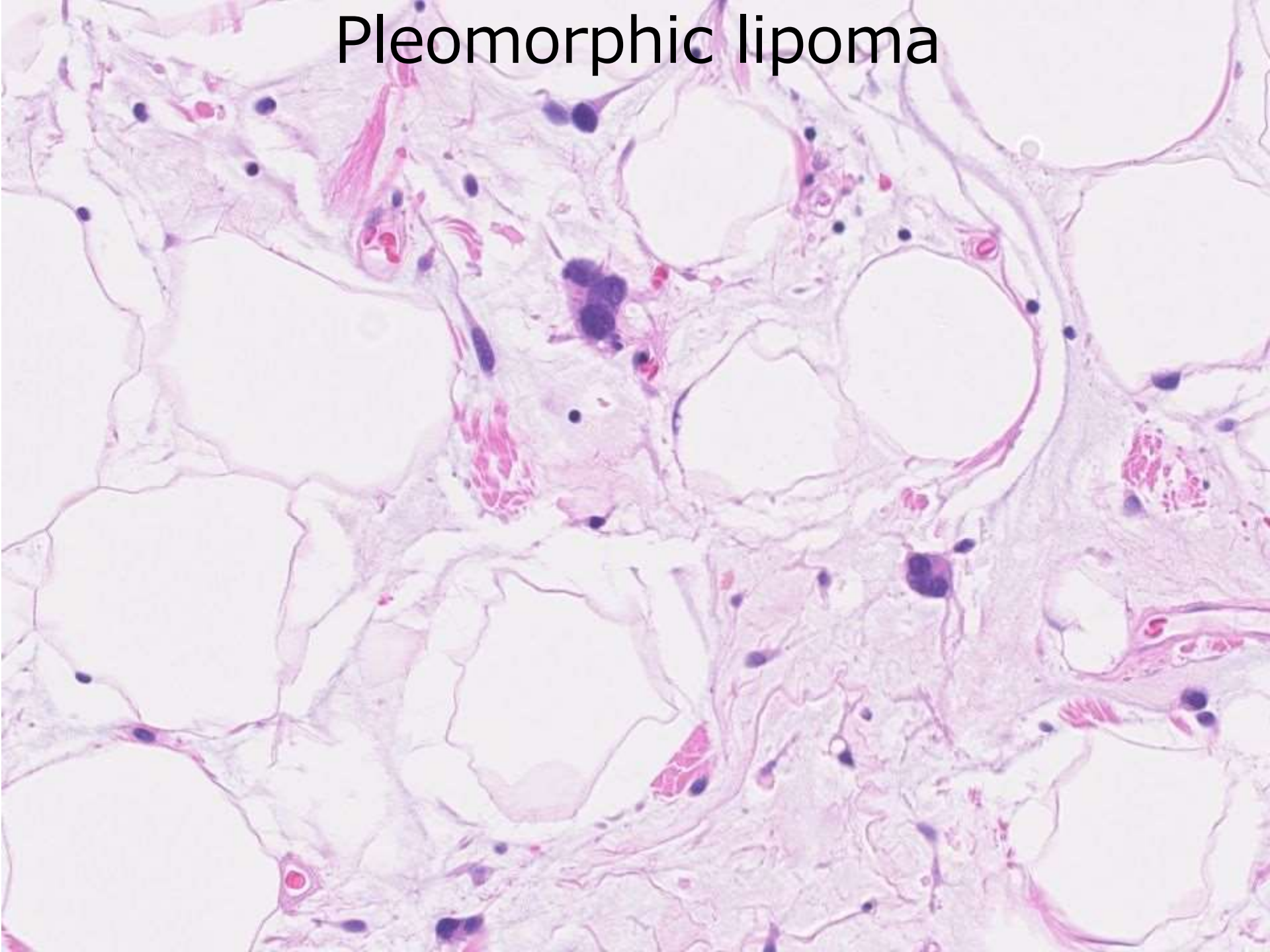

Angiolipoma


ALT

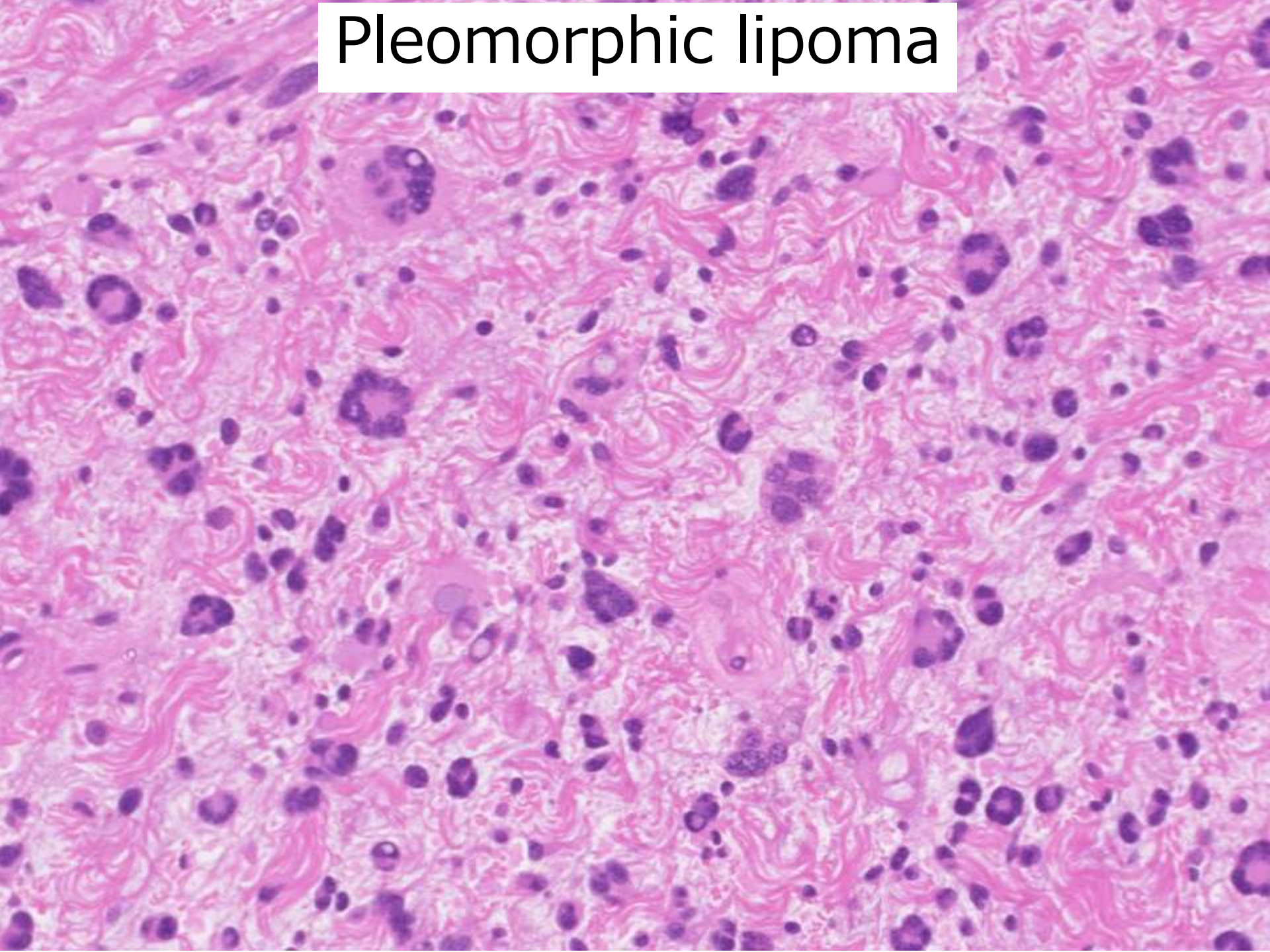
Pleomorphic lipoma



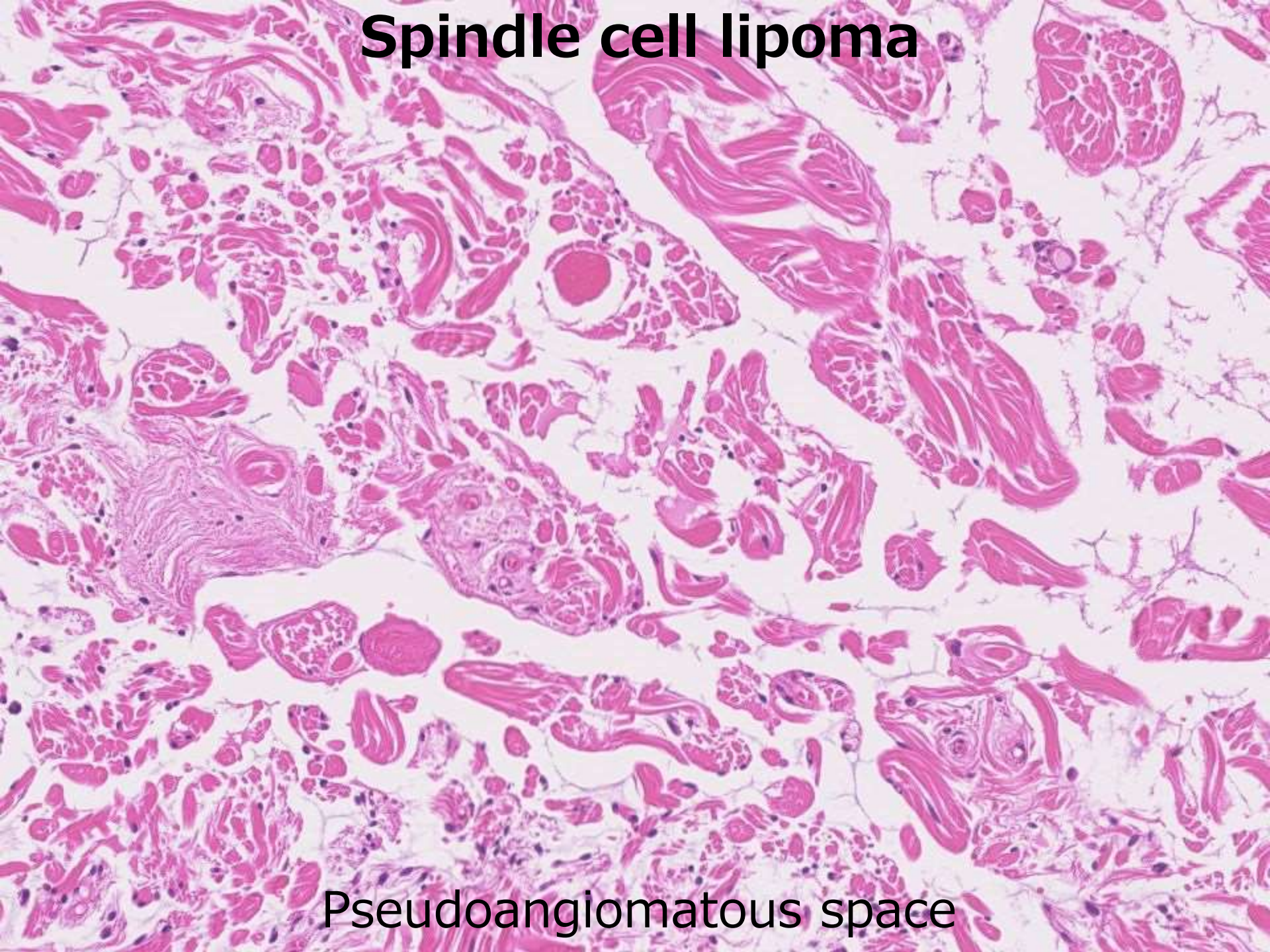
Pleomorphic lipoma



Pleomorphic lipoma

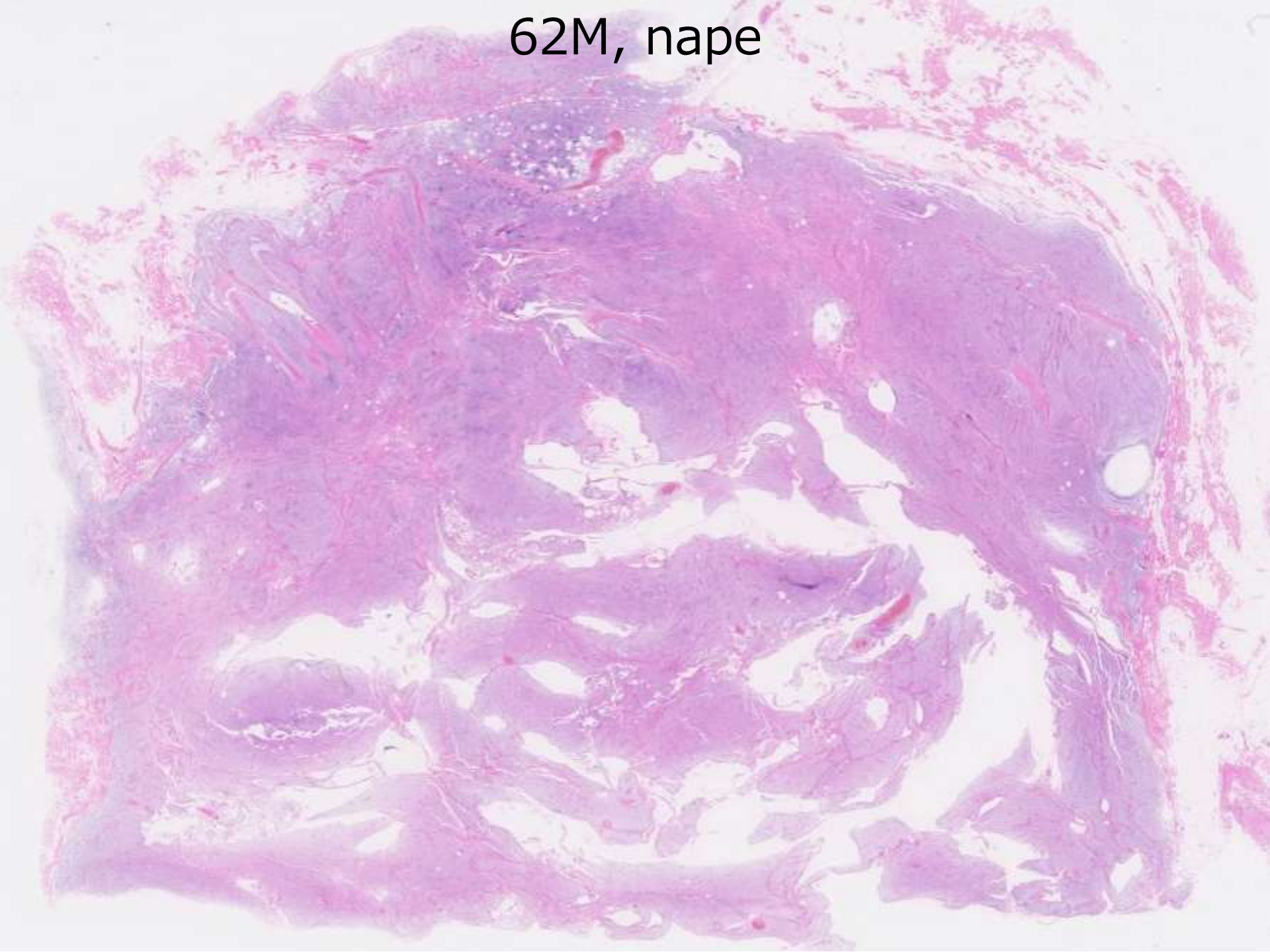


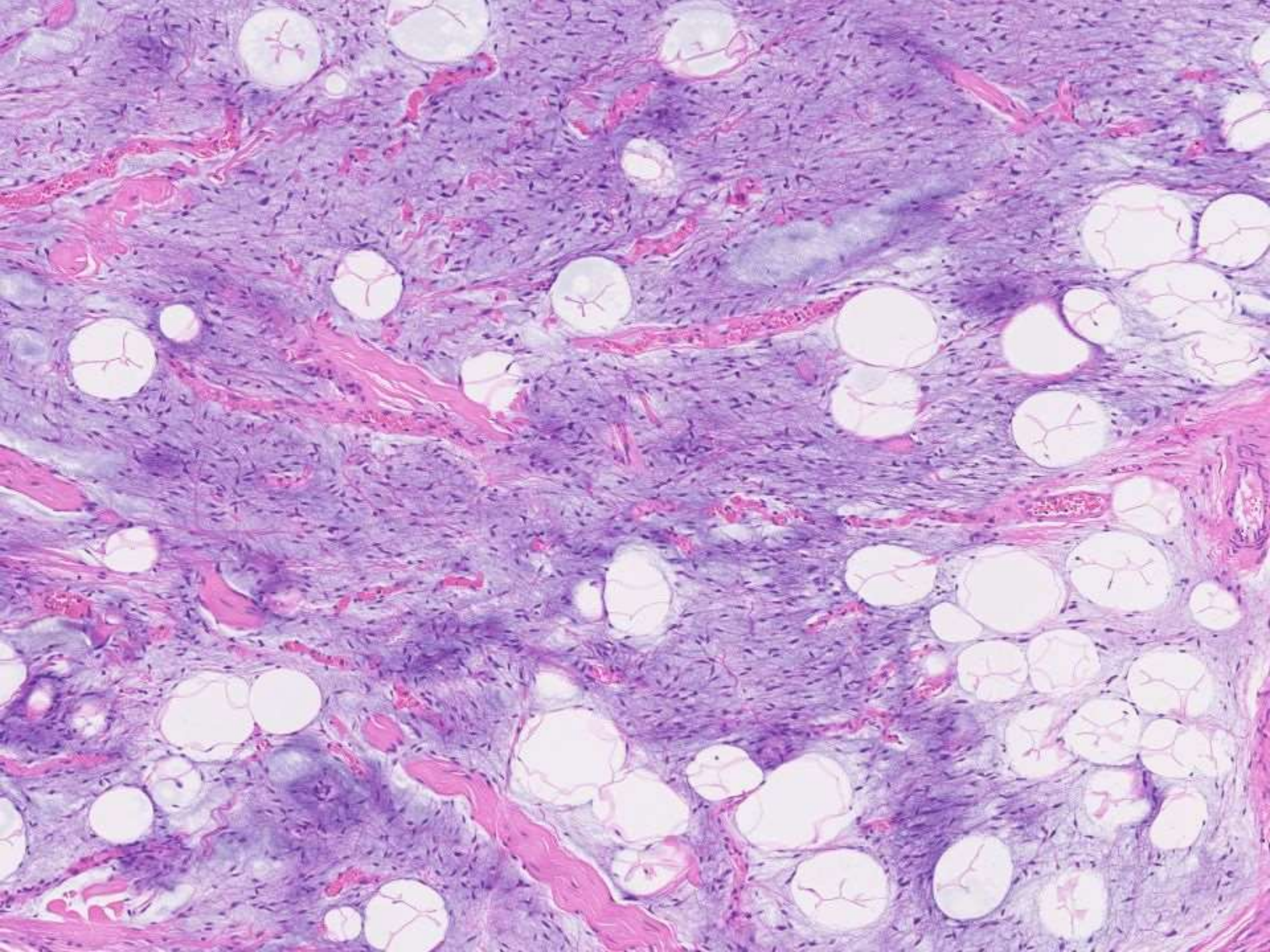
Spindle cell lipoma

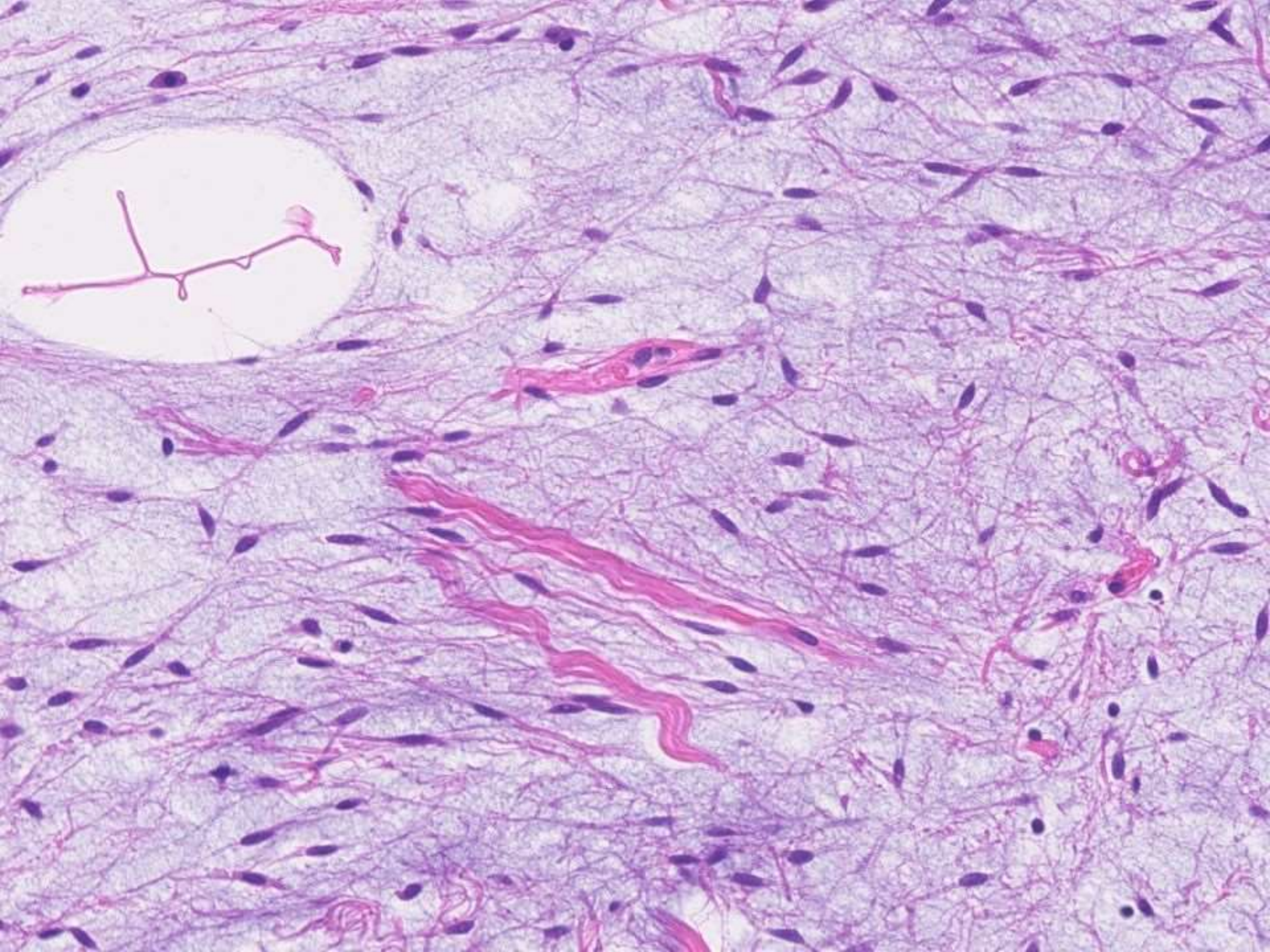


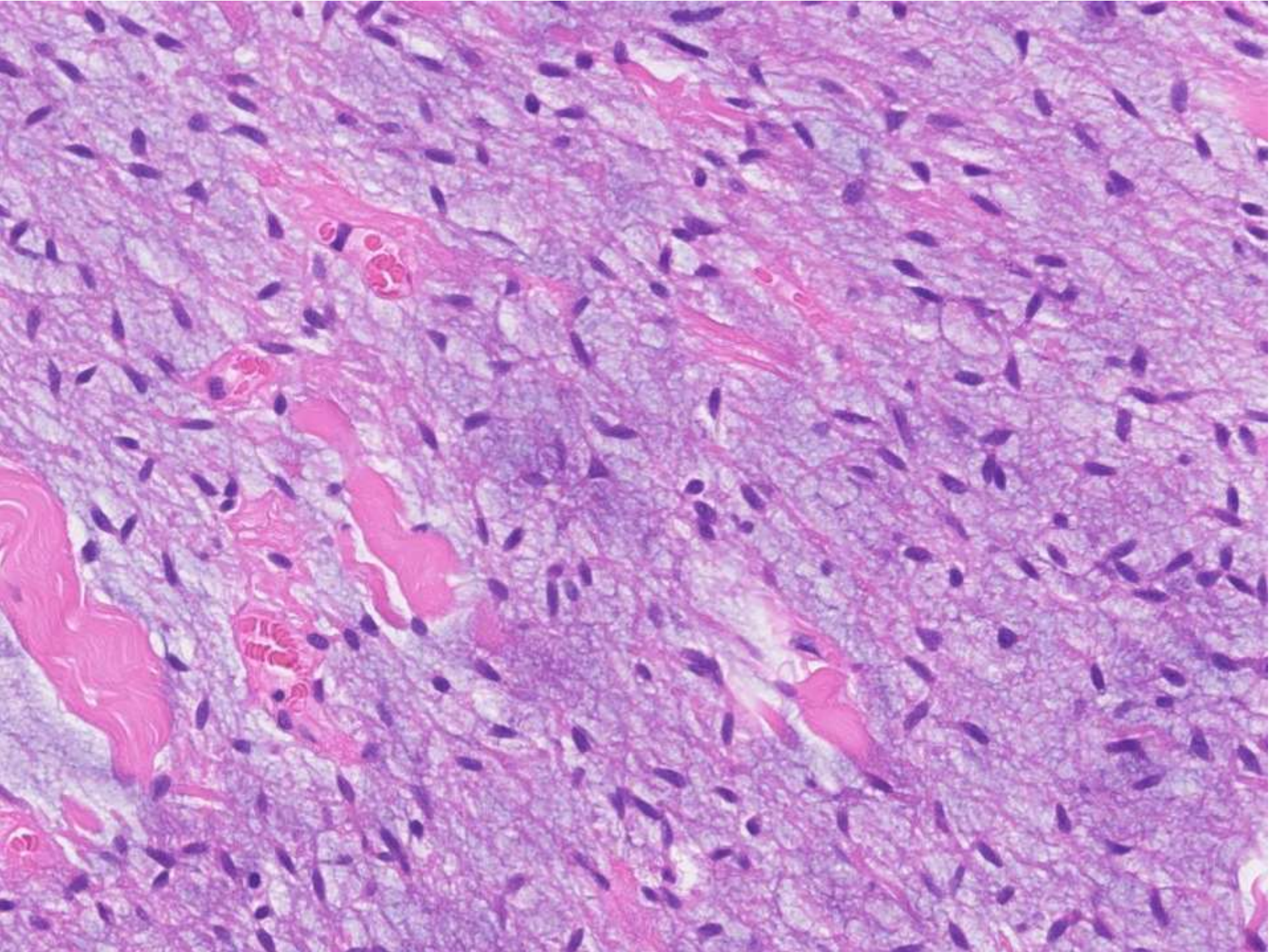
Pseudoangiomatous space

62M, nape



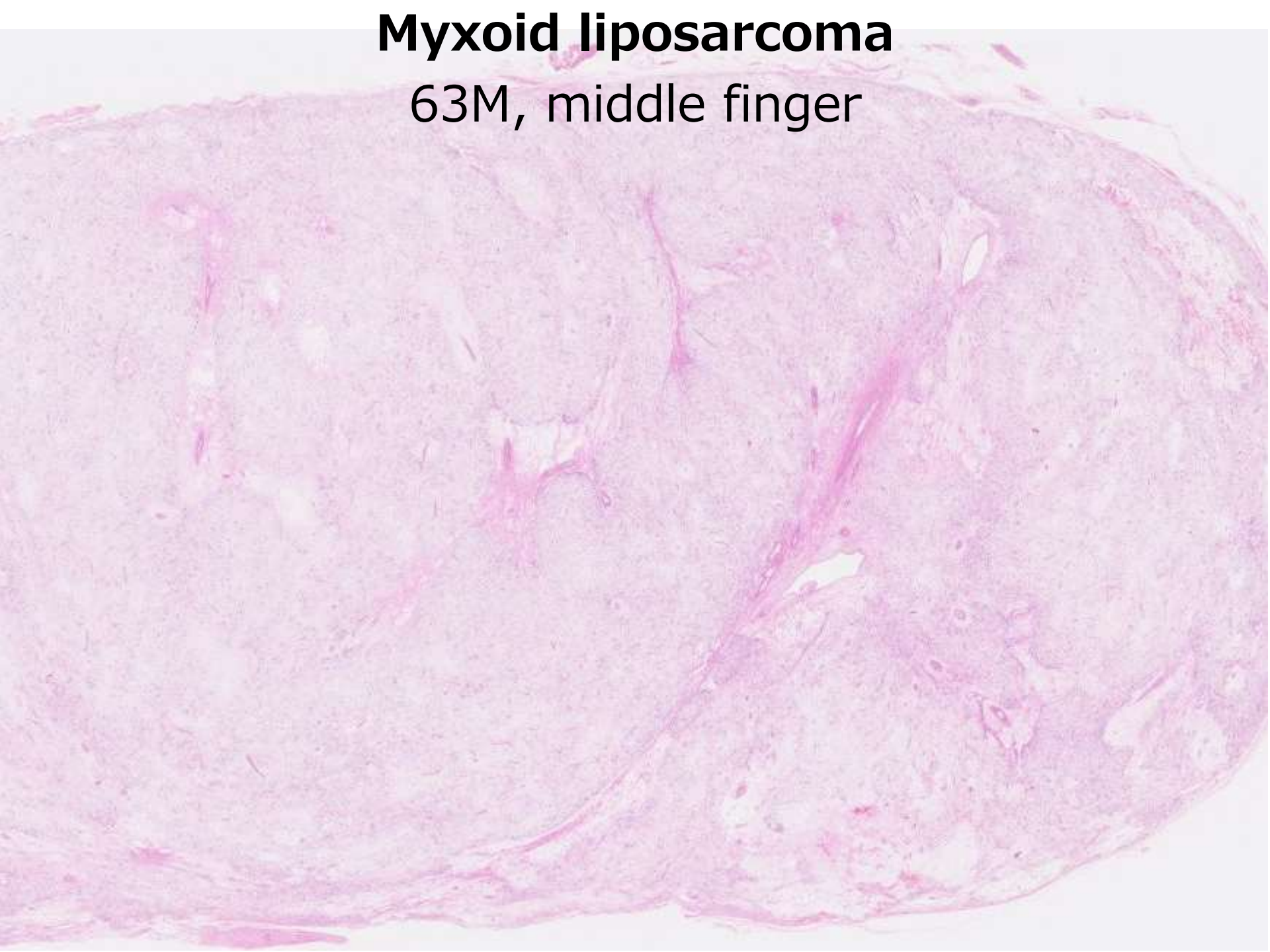


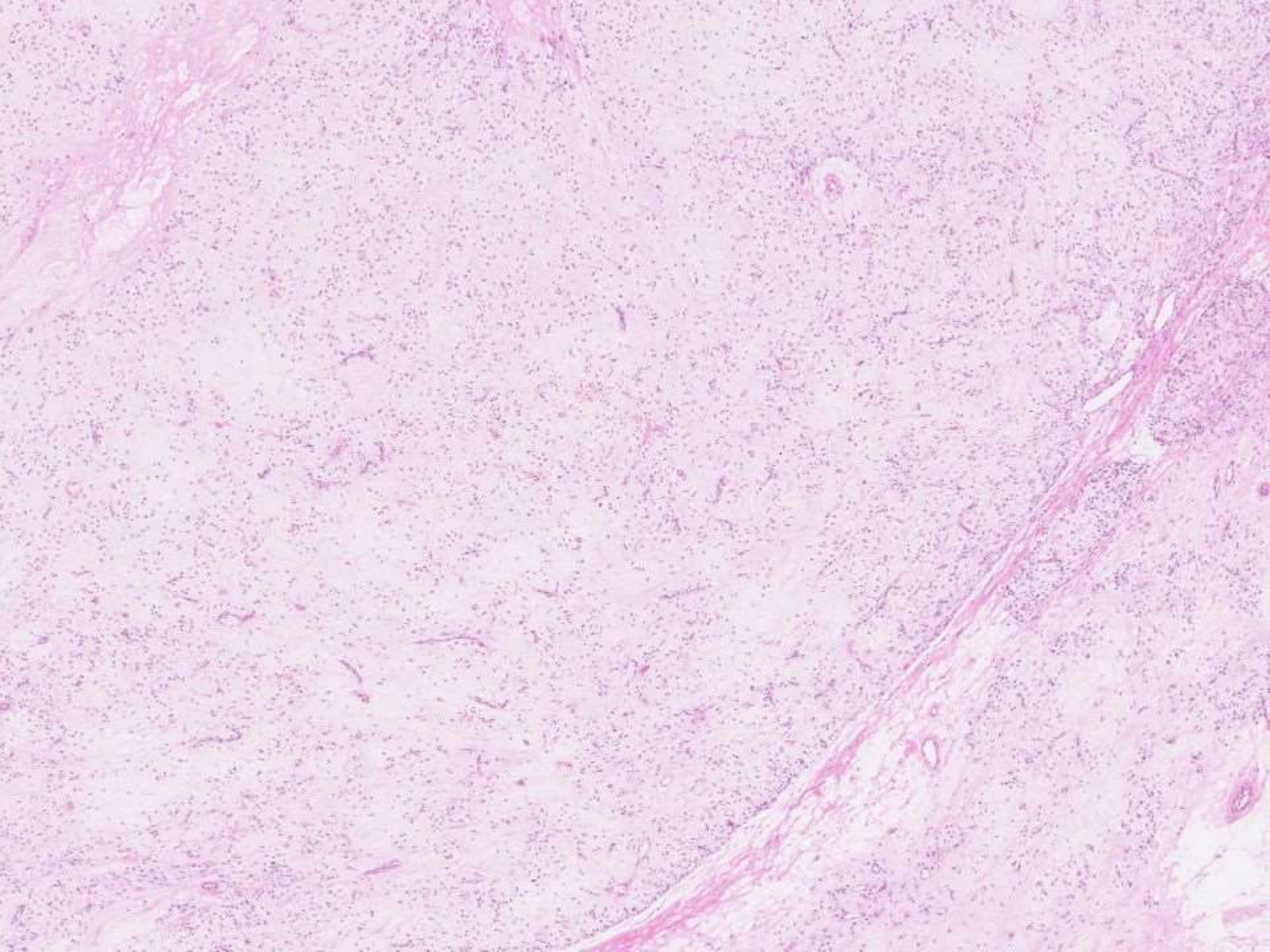


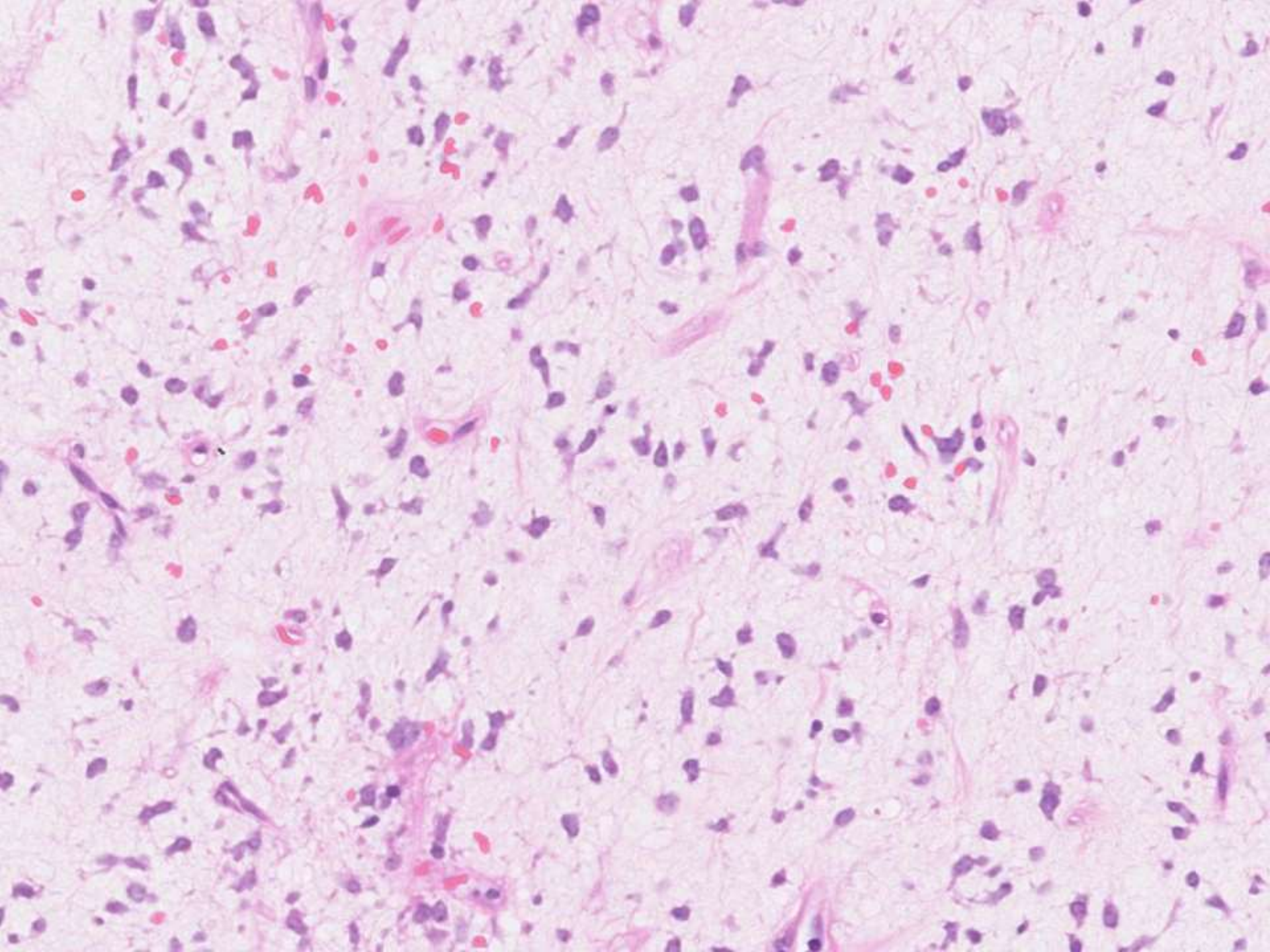


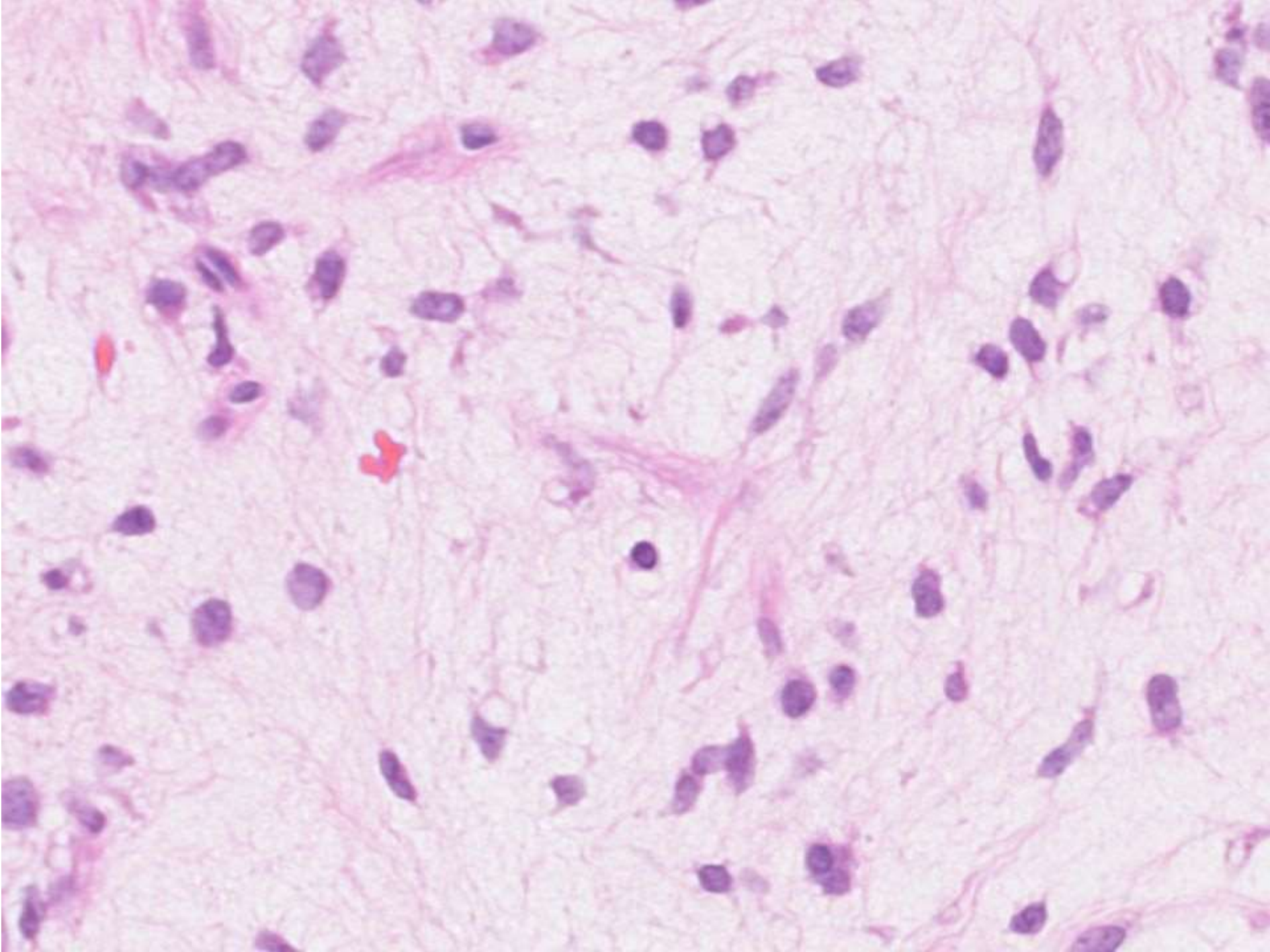
Myxoid liposarcoma

63M, middle finger

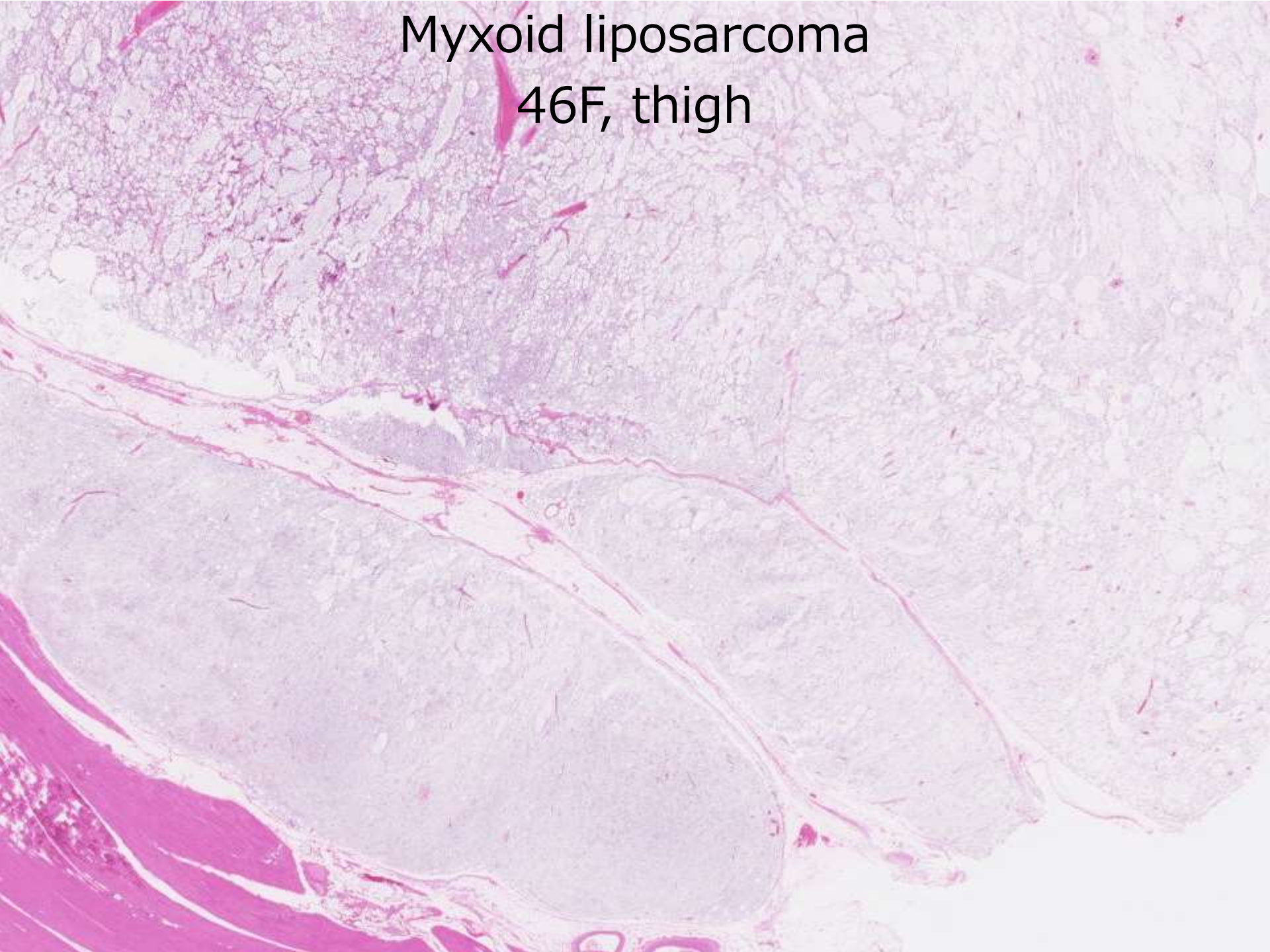


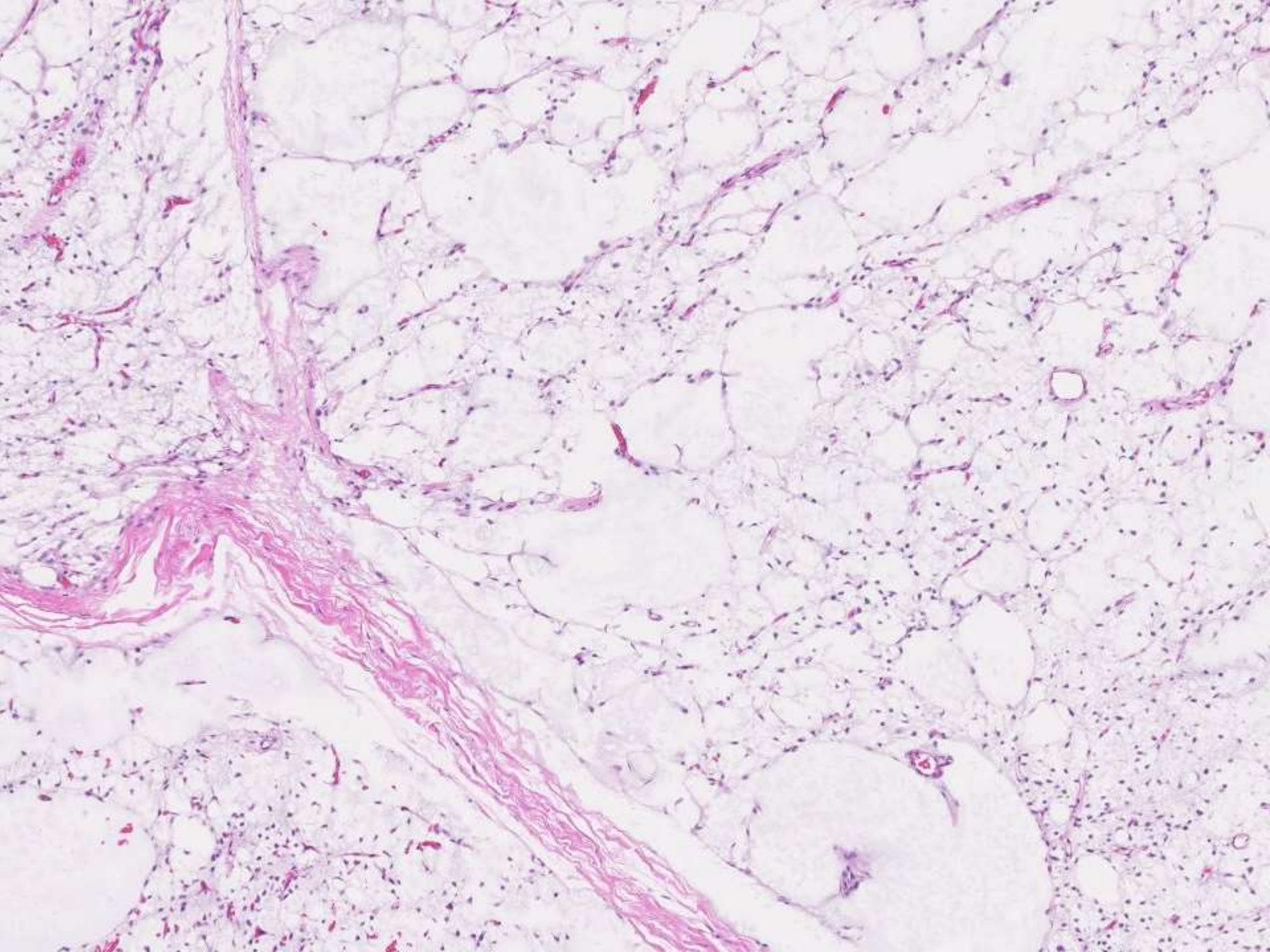


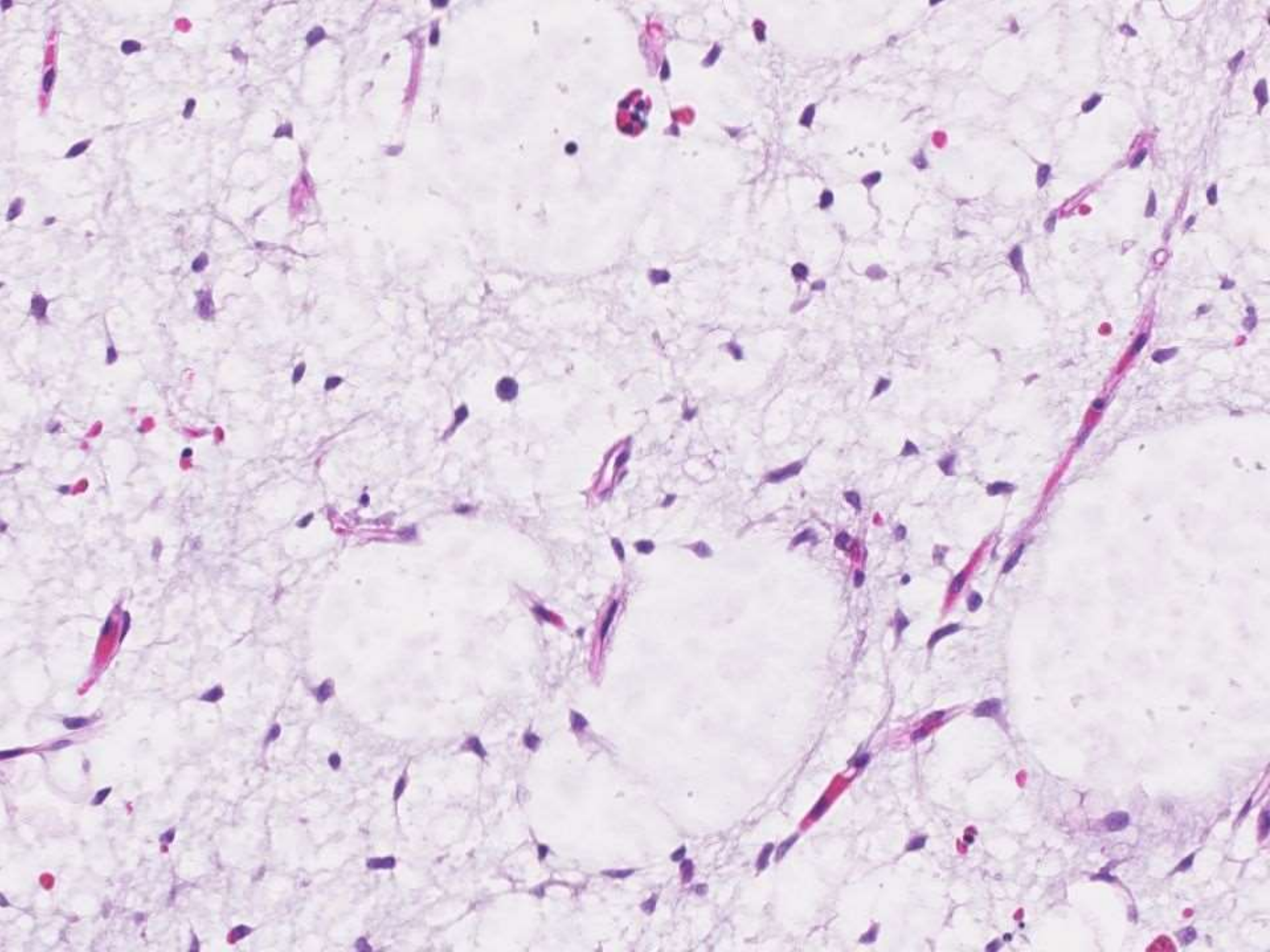


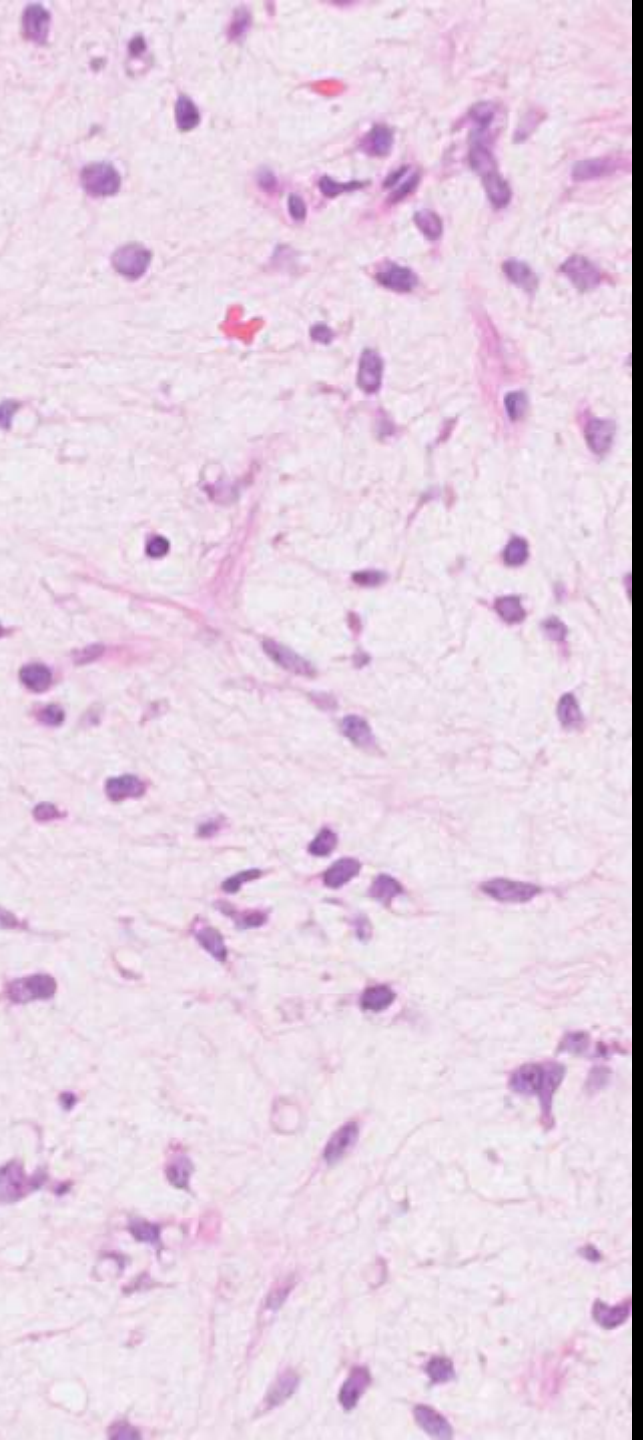
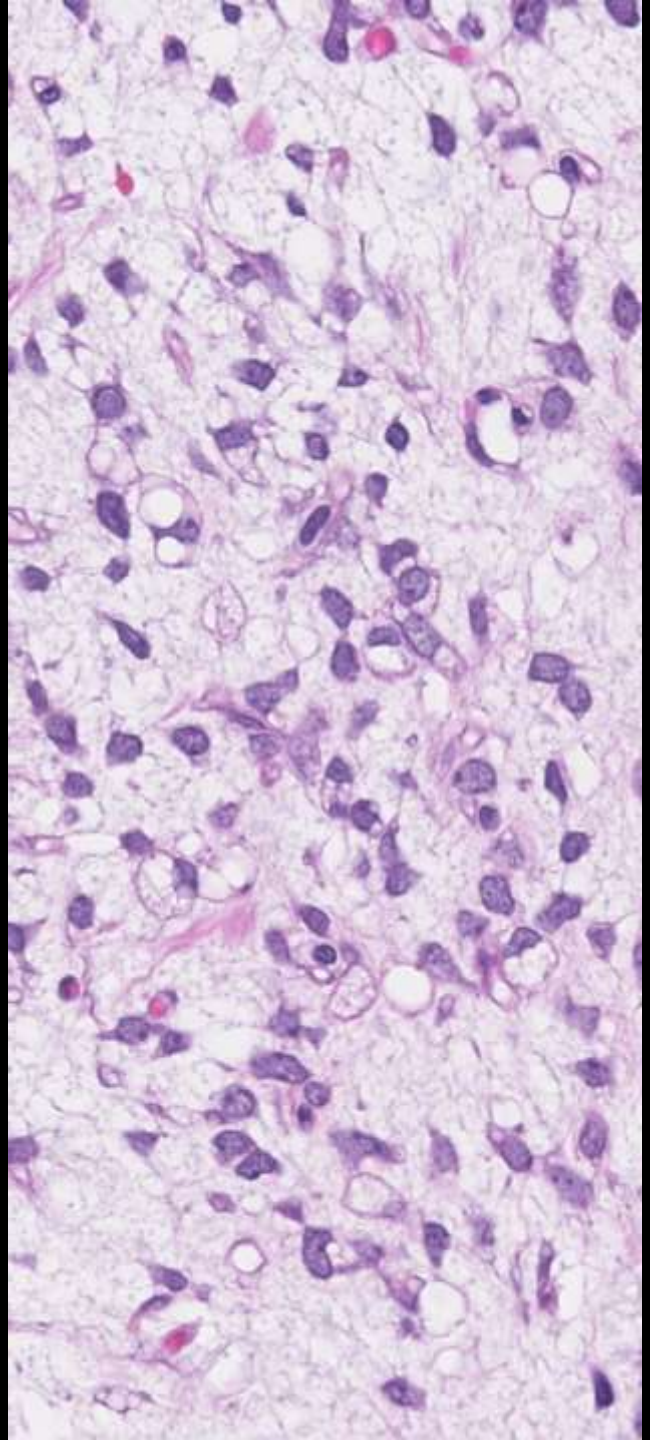
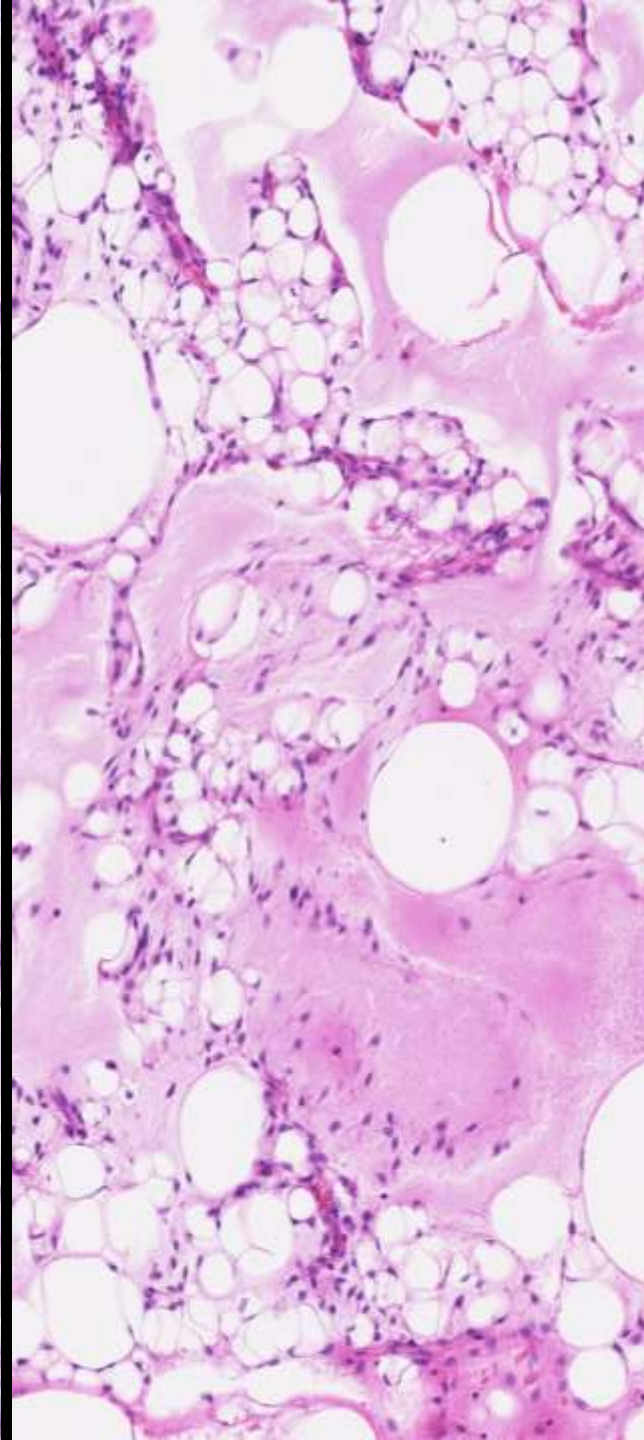


Myxoid liposarcoma
46F, thigh

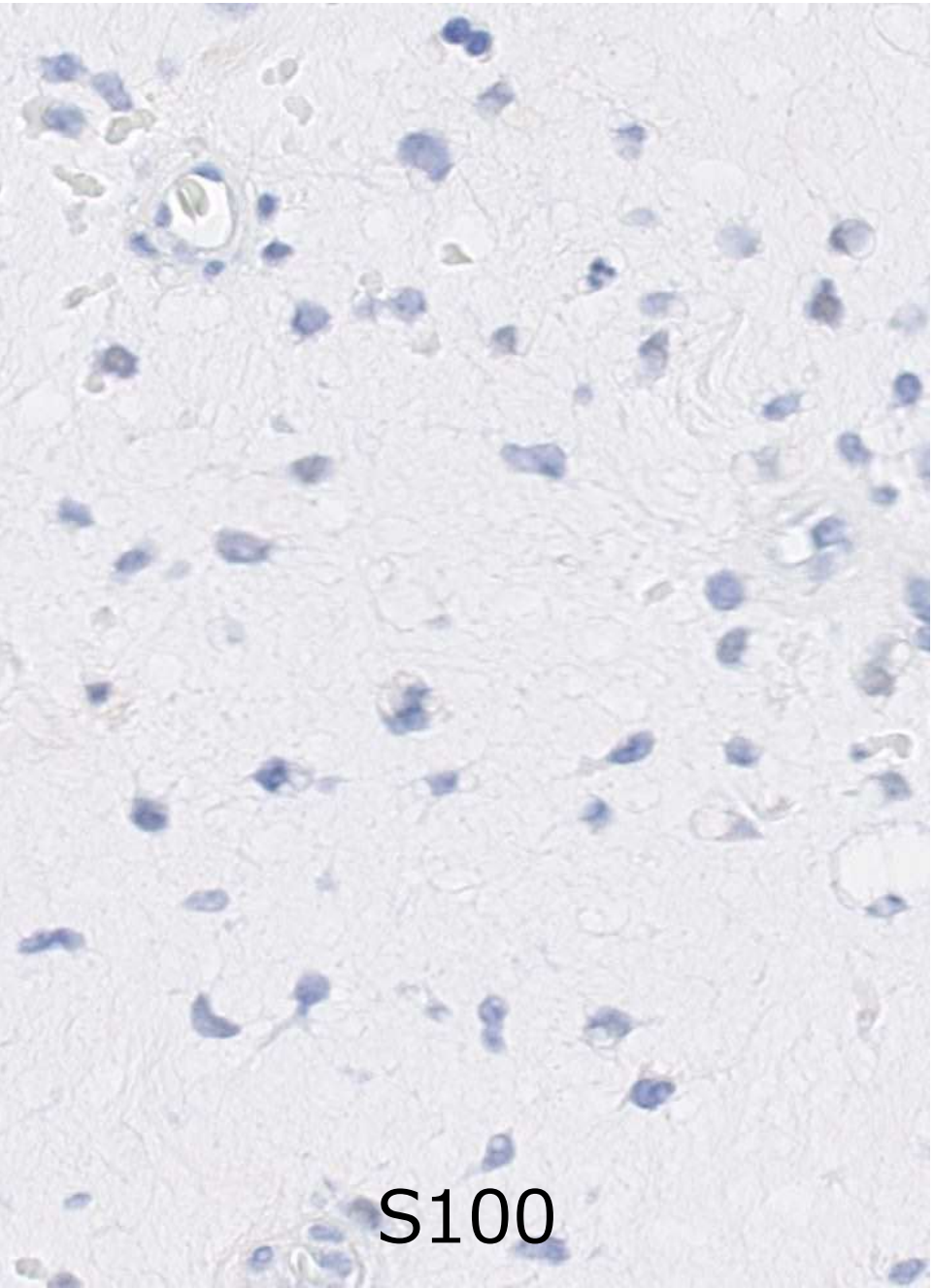


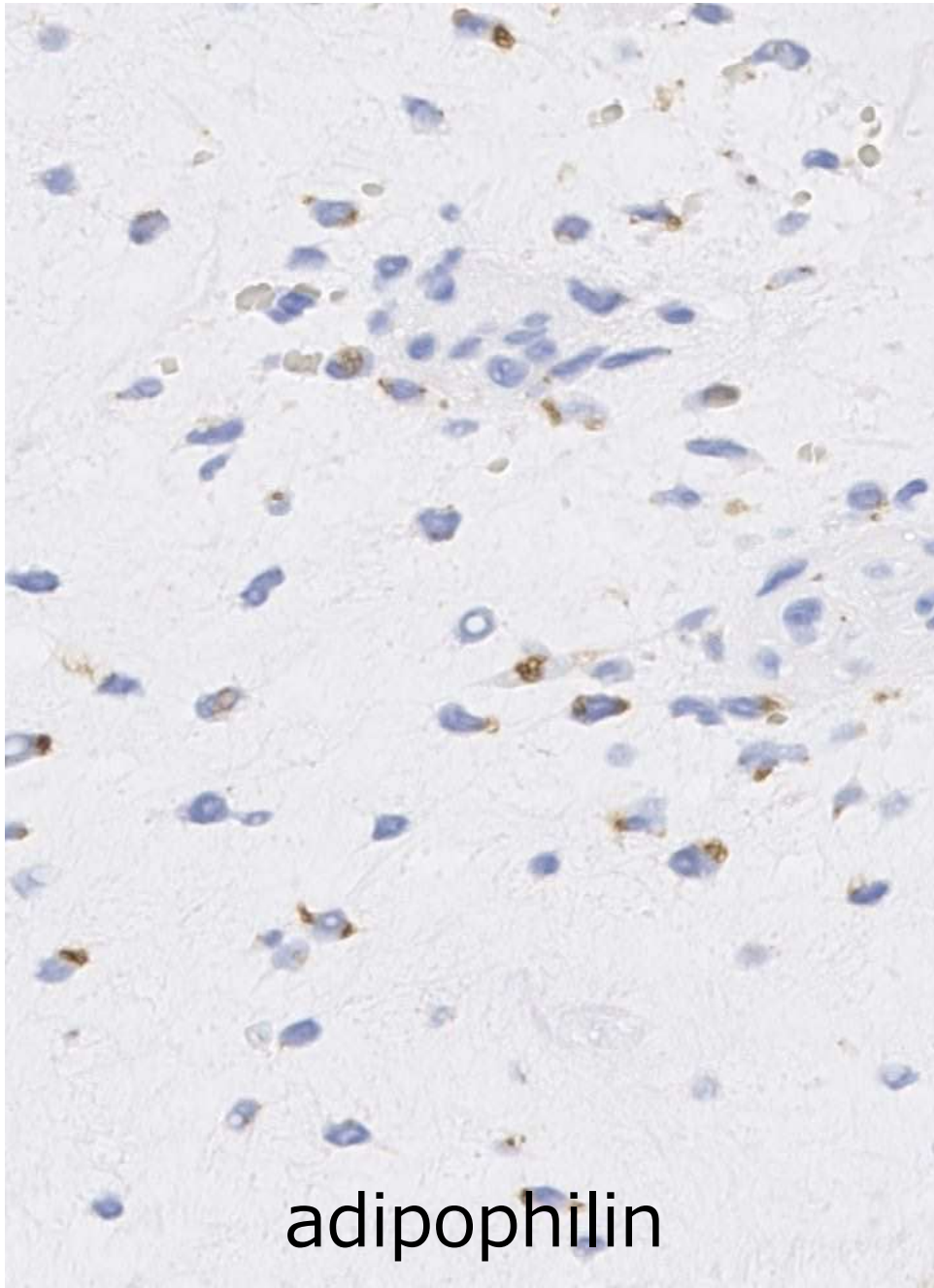




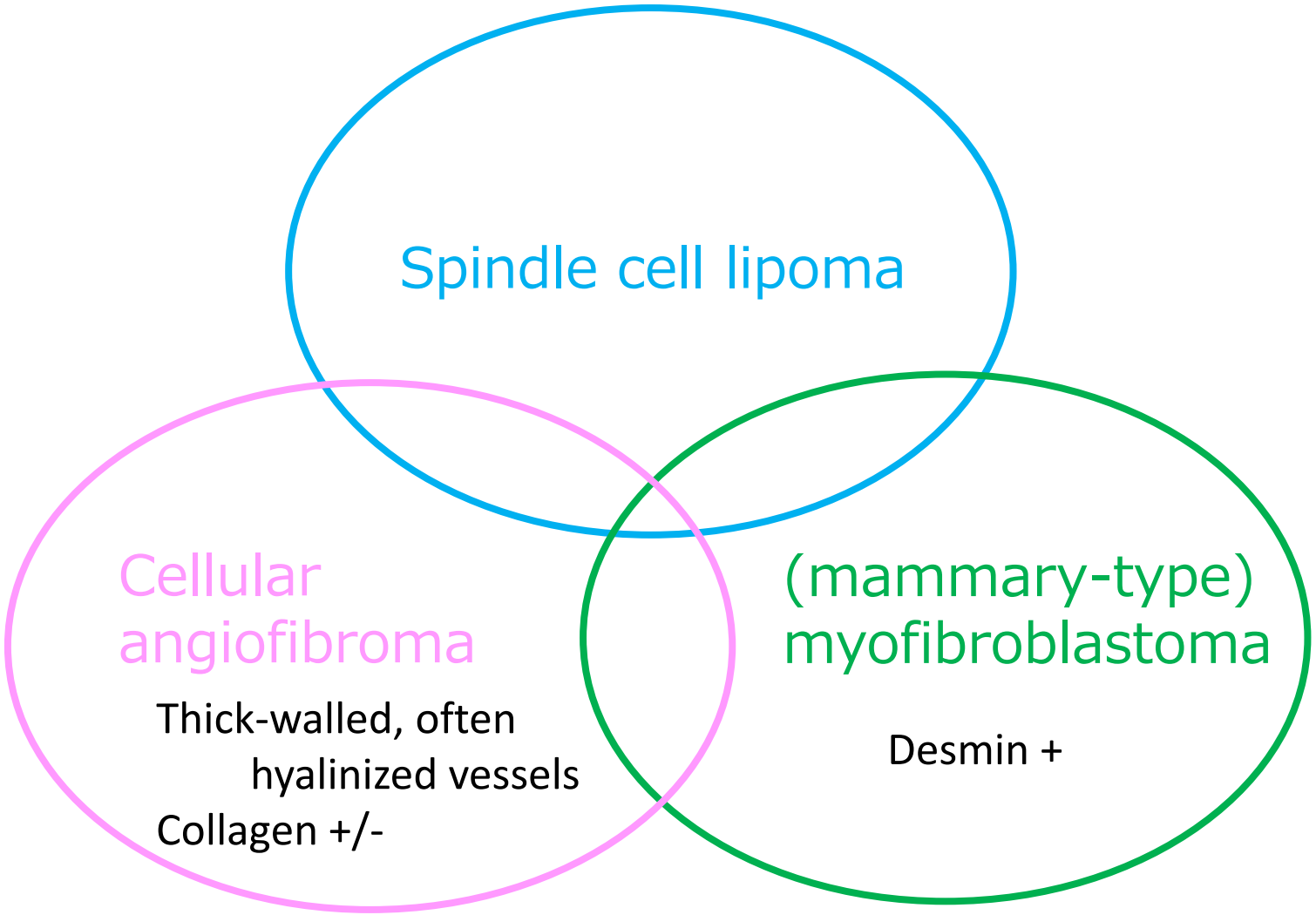
Myxoid liposarcoma

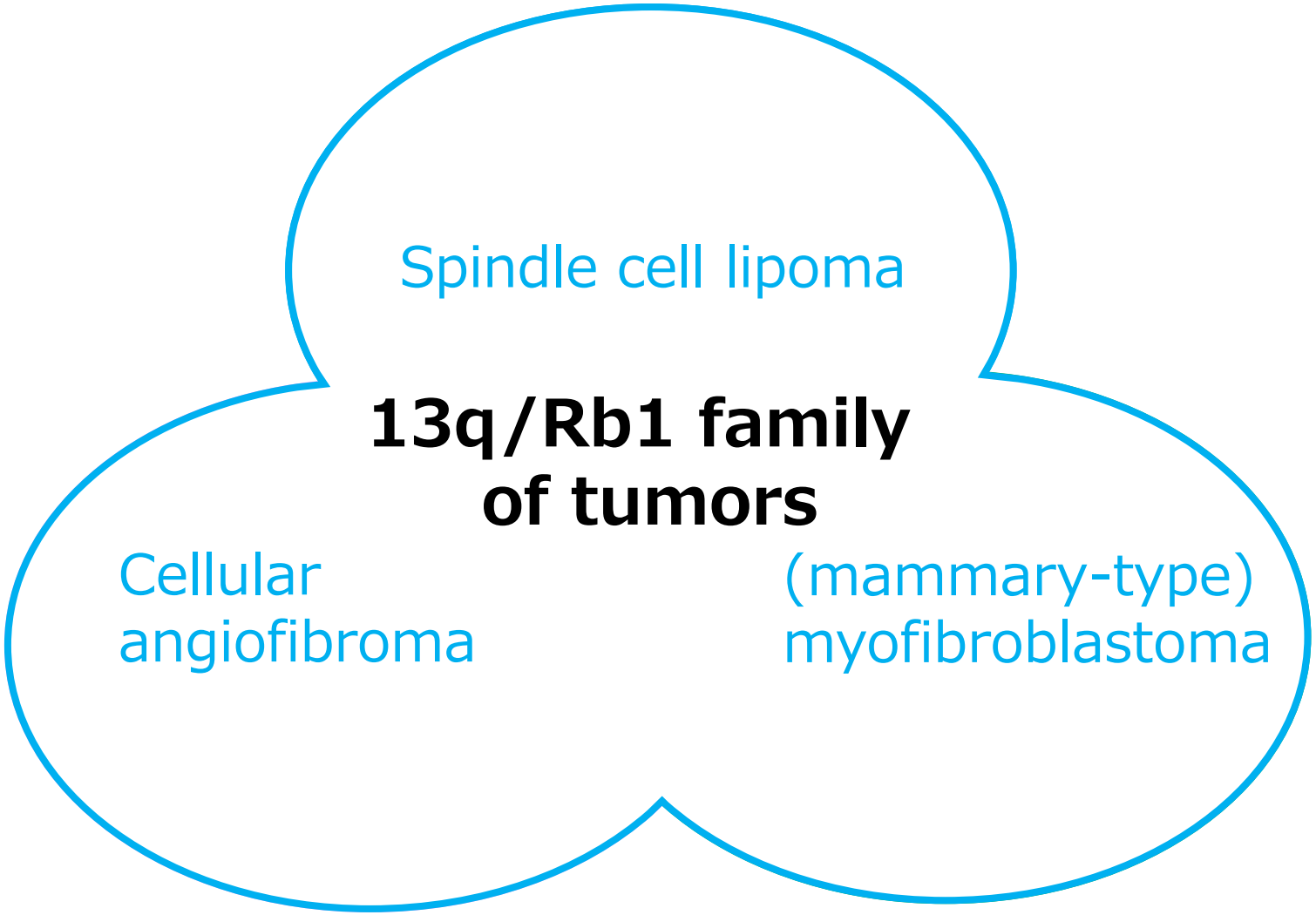




Tips 5

- spindle cell lipomaはしばしば高度の粘液腫状変化を示す
- 粘液腫状の場合、脂肪が少ない／ほとんどない
- 脂肪の間（隔壁ではない）に短紡錘形細胞とropey collagenがみられる

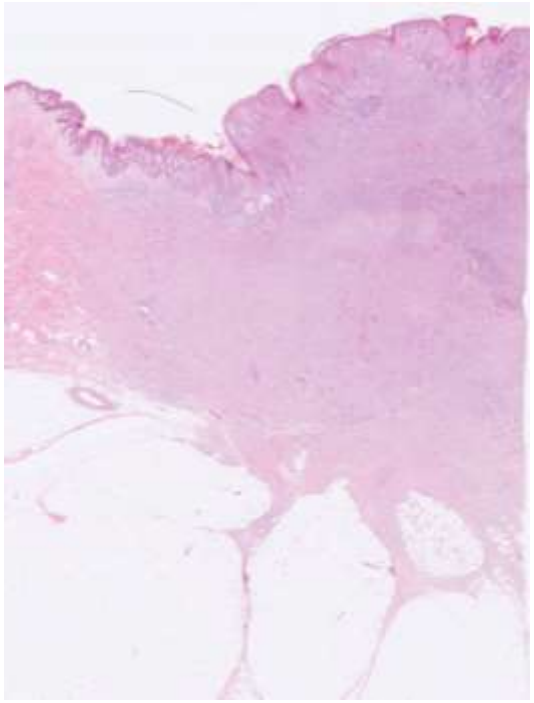

Spindle cell lipoma
Cellular angiofibroma
Thick-walled, often hyalinized vessels
Collagen +/-
(mammary-type) myofibroblastoma
Desmin +

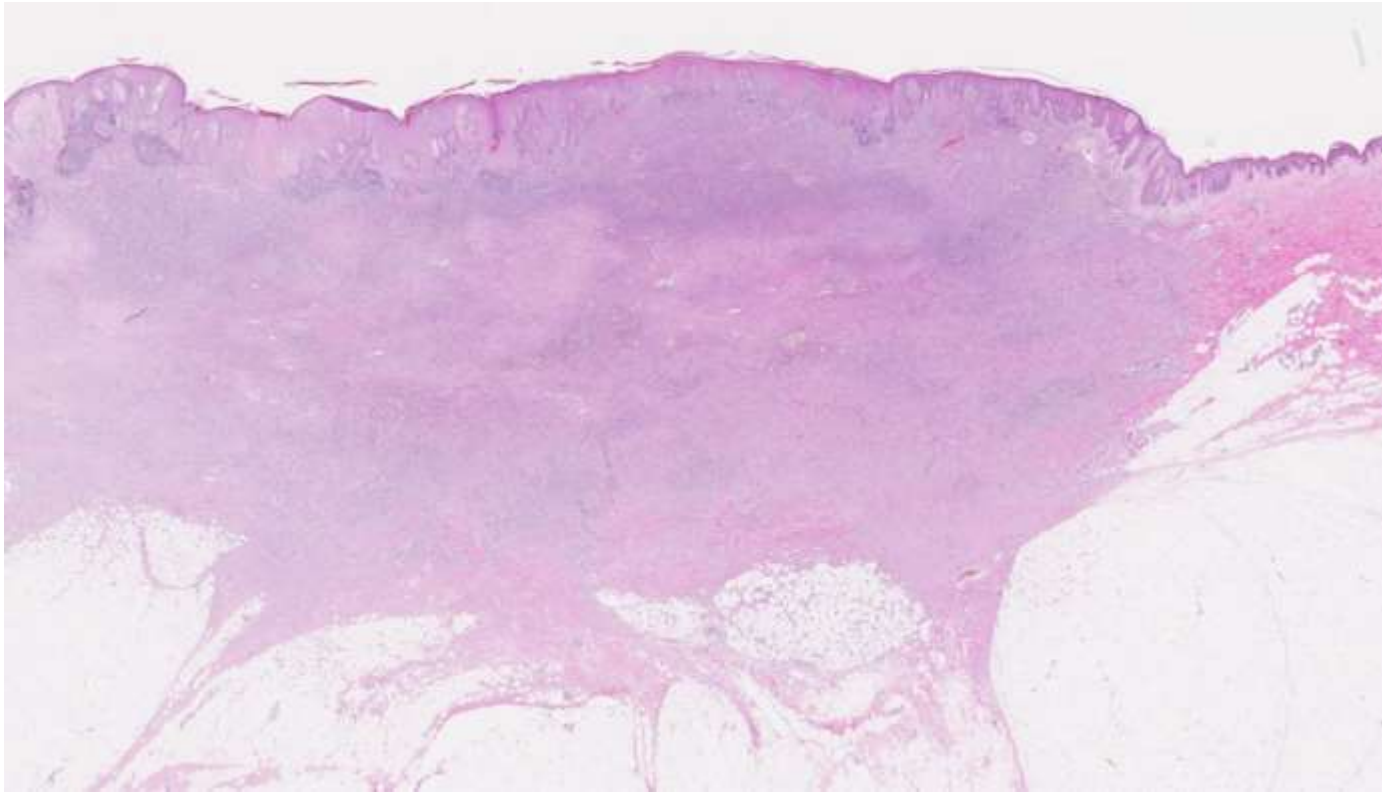

Spindle cell lipoma
13q/Rb1 family of tumors
Cellular angiofibroma
(mammary-type) myofibroblastoma

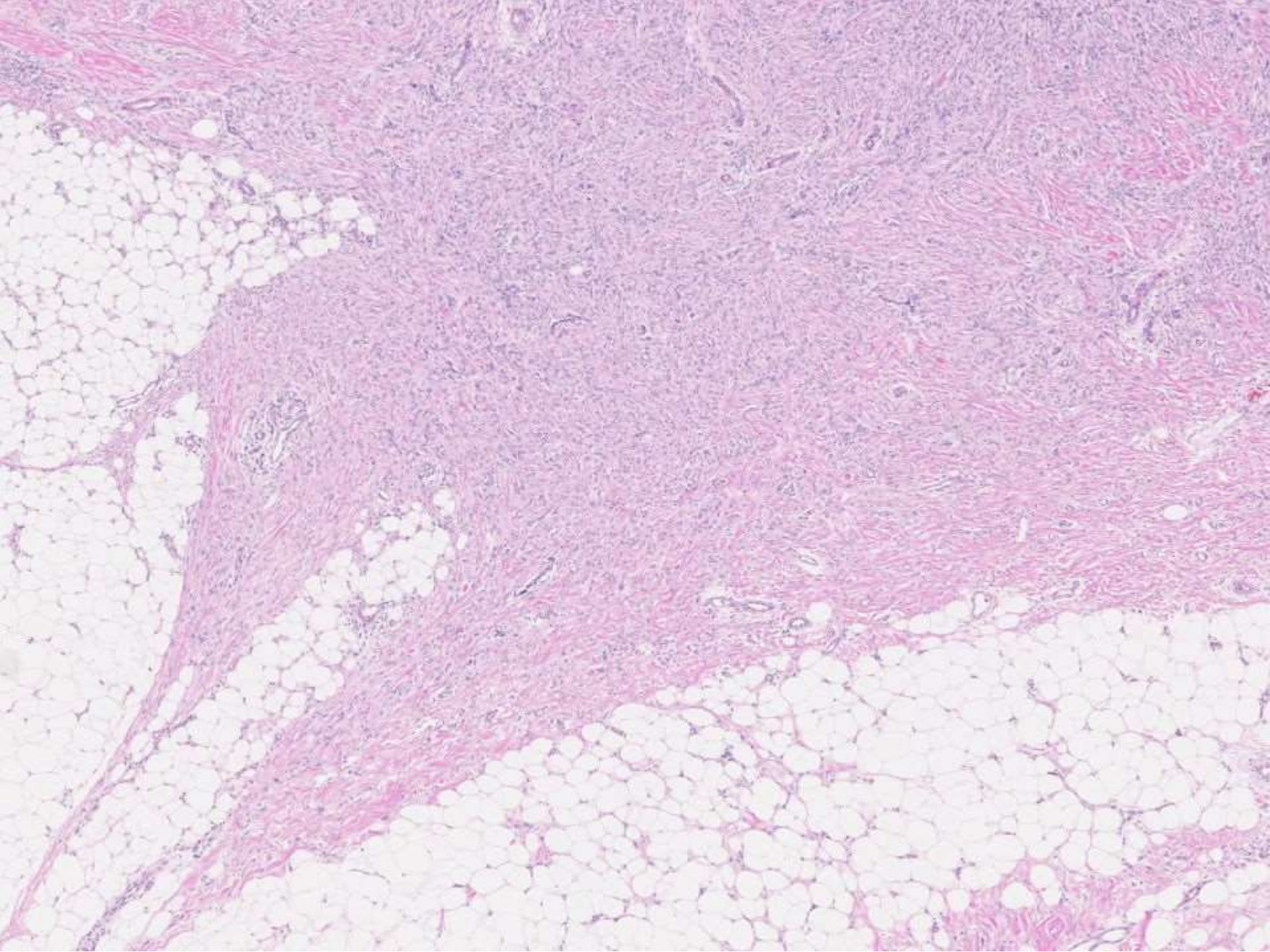
Case 4: 72F, chest wall

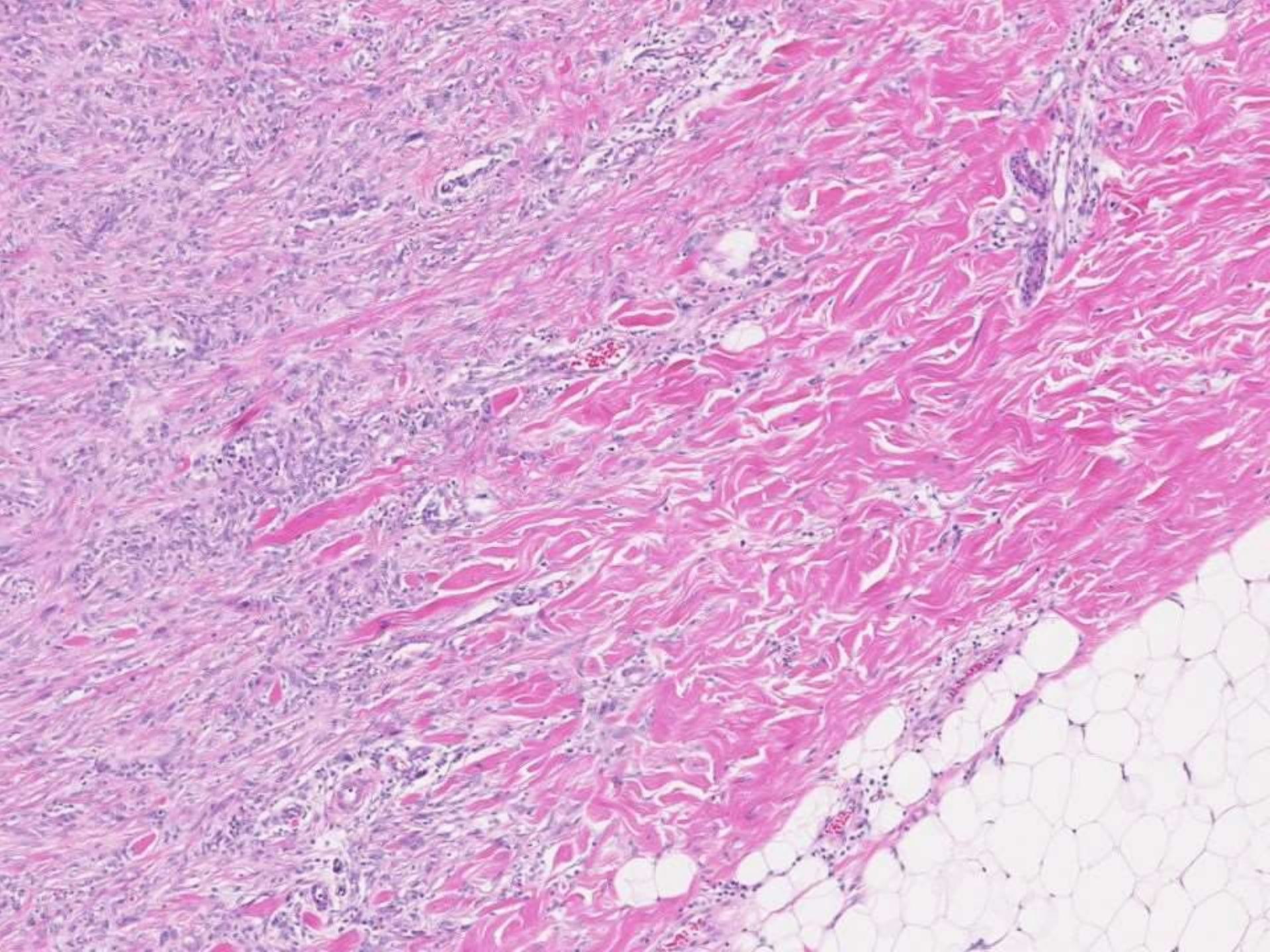
約2年前に2 cm大のケロイド様の腫瘤が出現し、様々な治療を行われたが改善しない

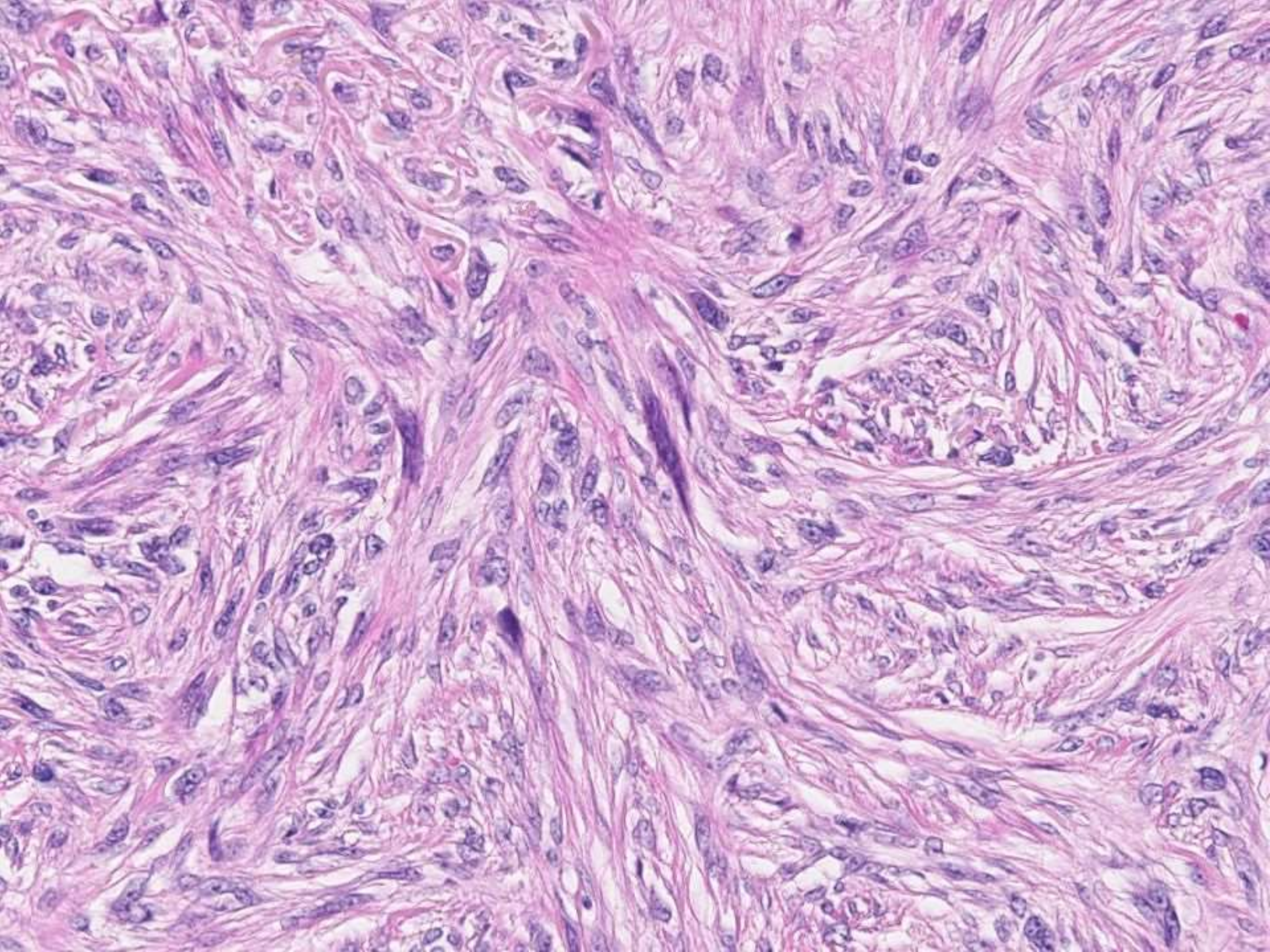
Dermatofibroma ?
Dermatofibrosarcoma protuberans ?

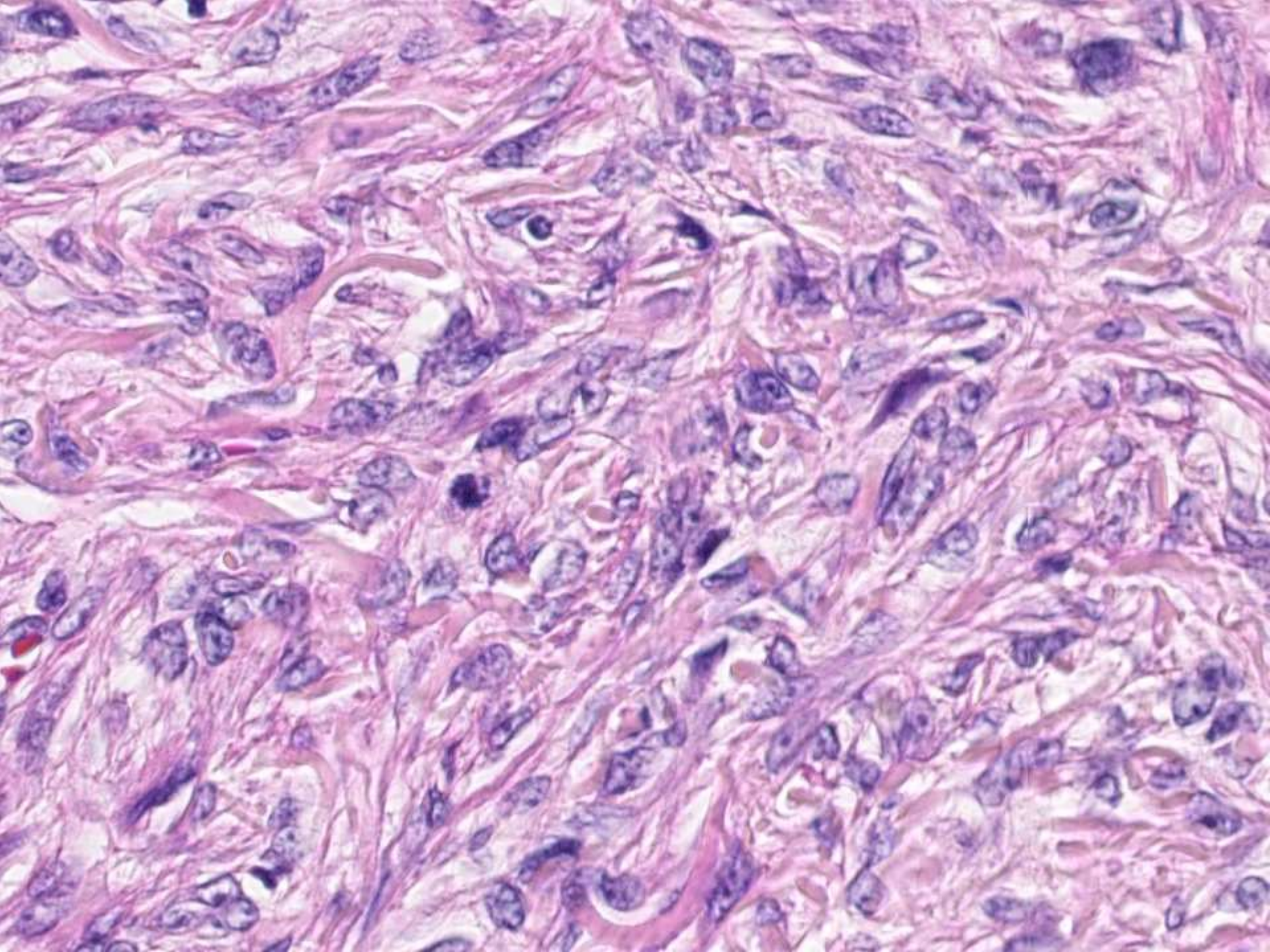


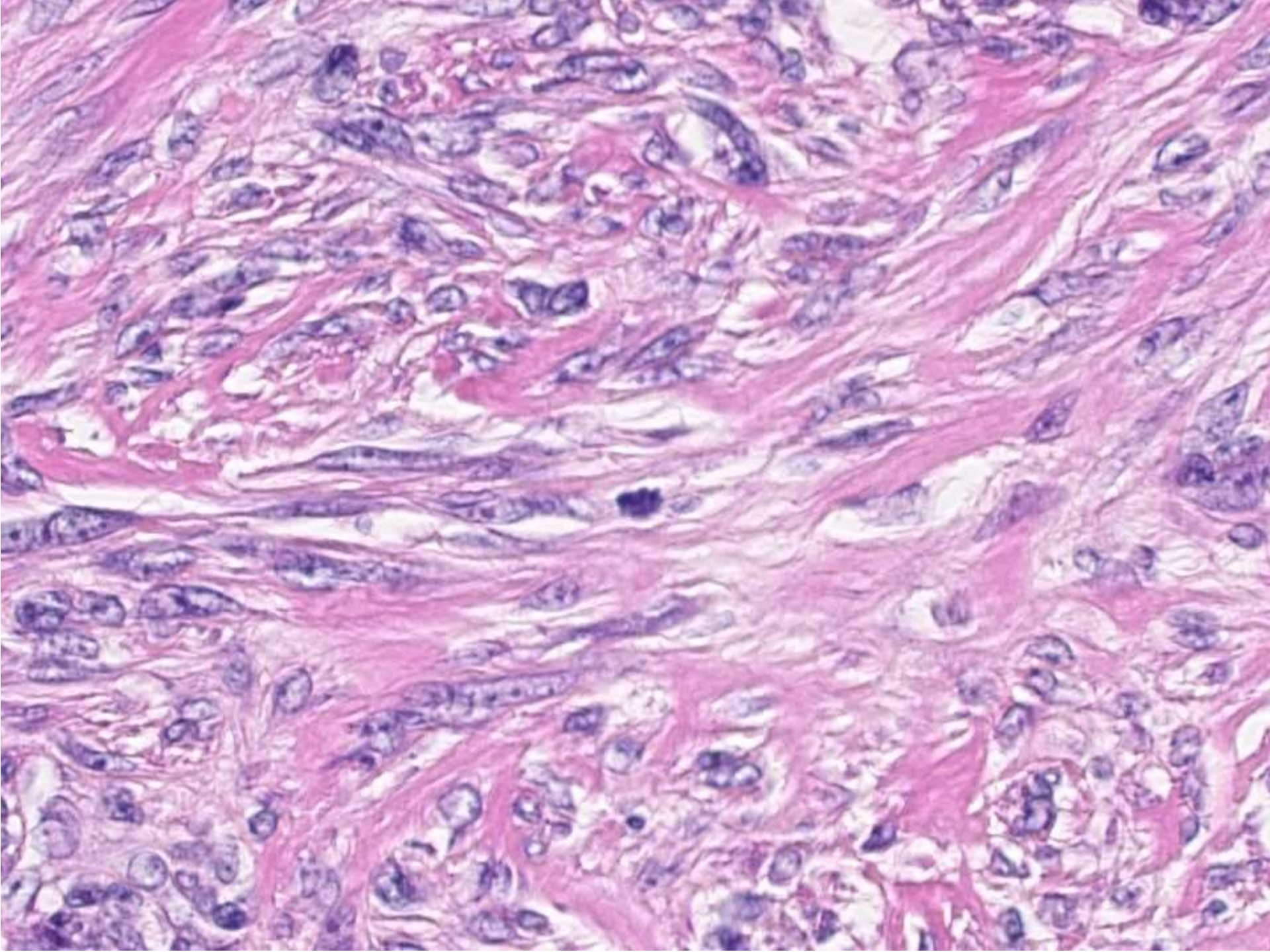


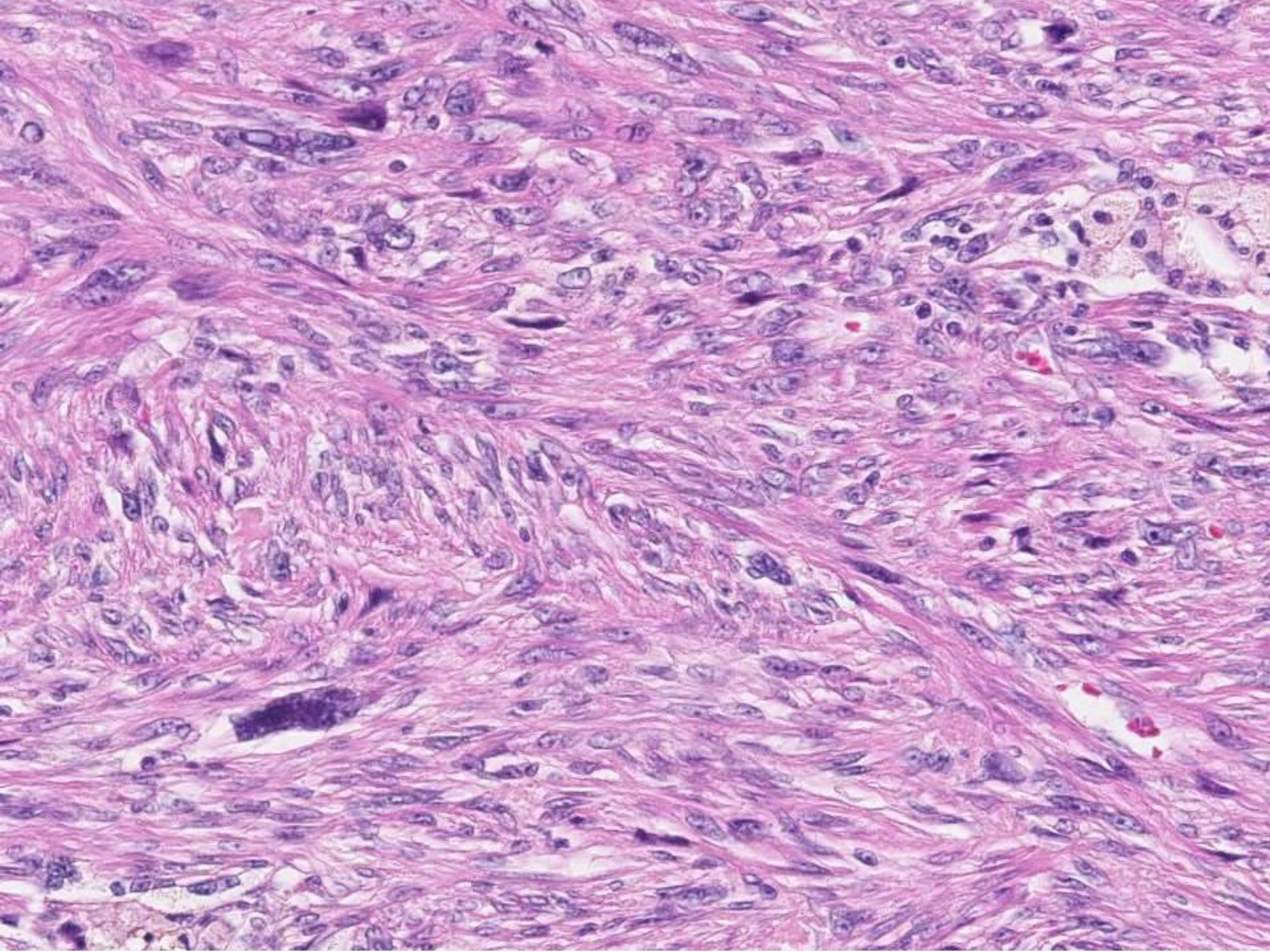








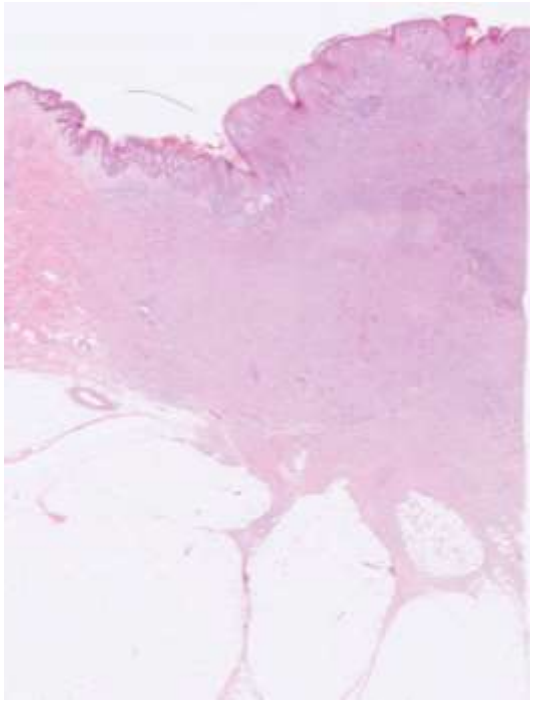


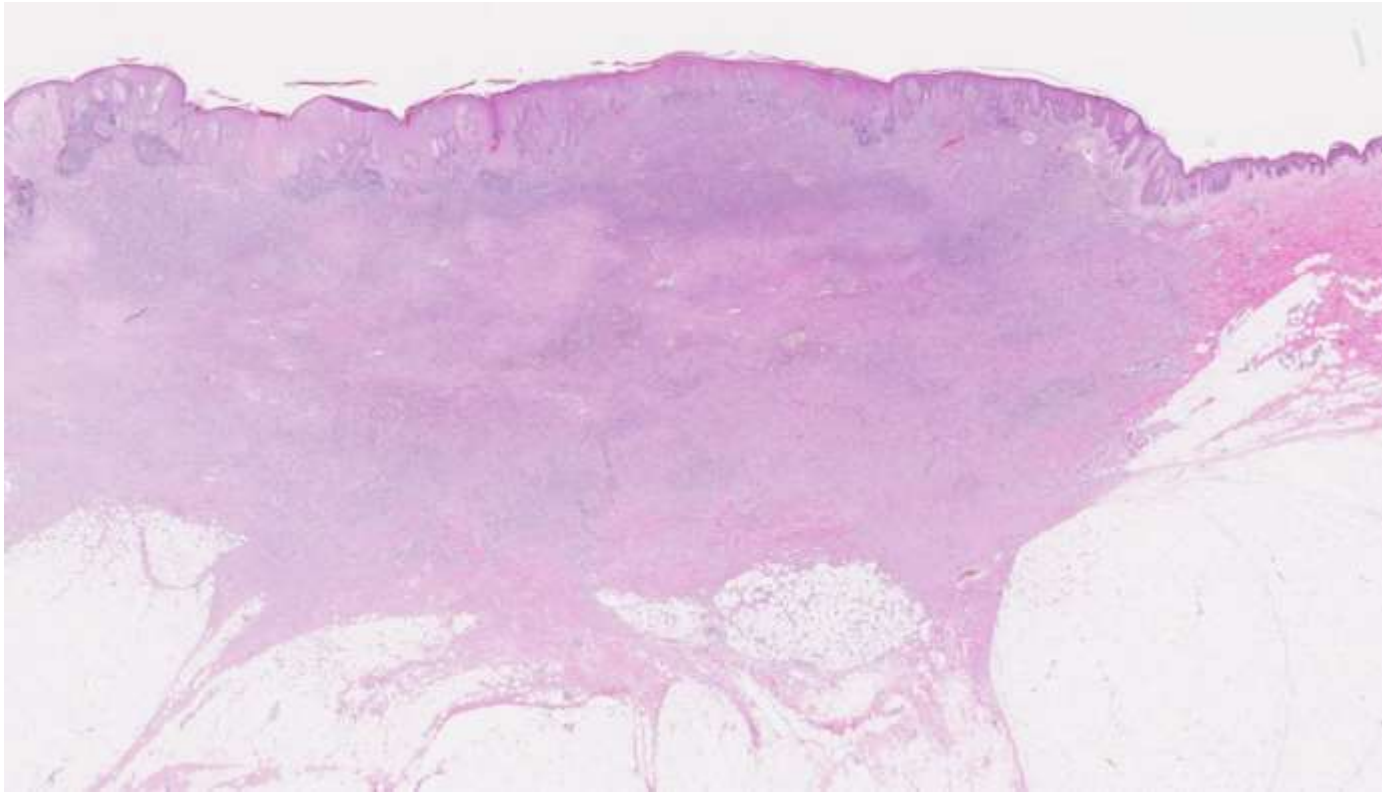


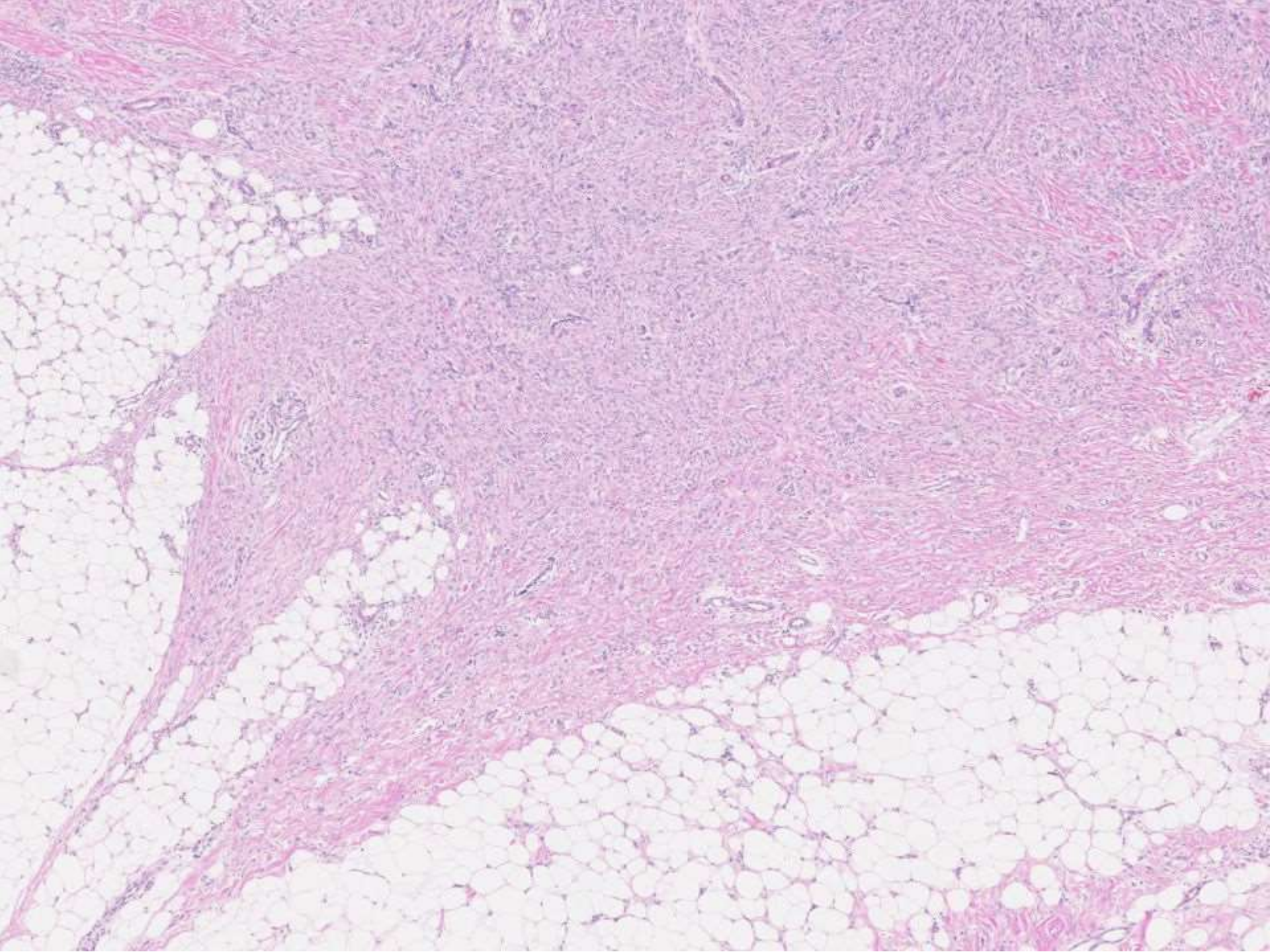
?

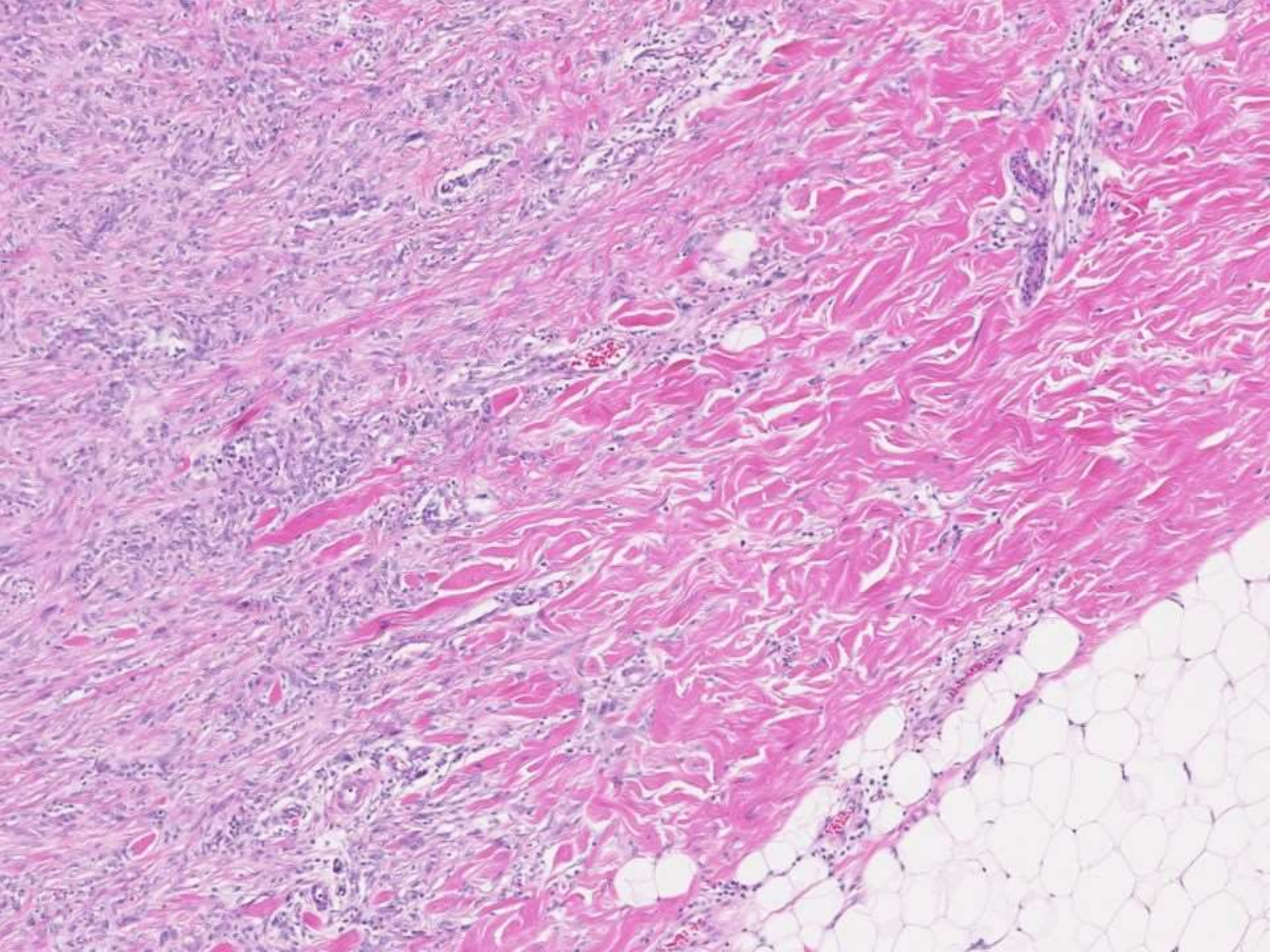
Diagnosis

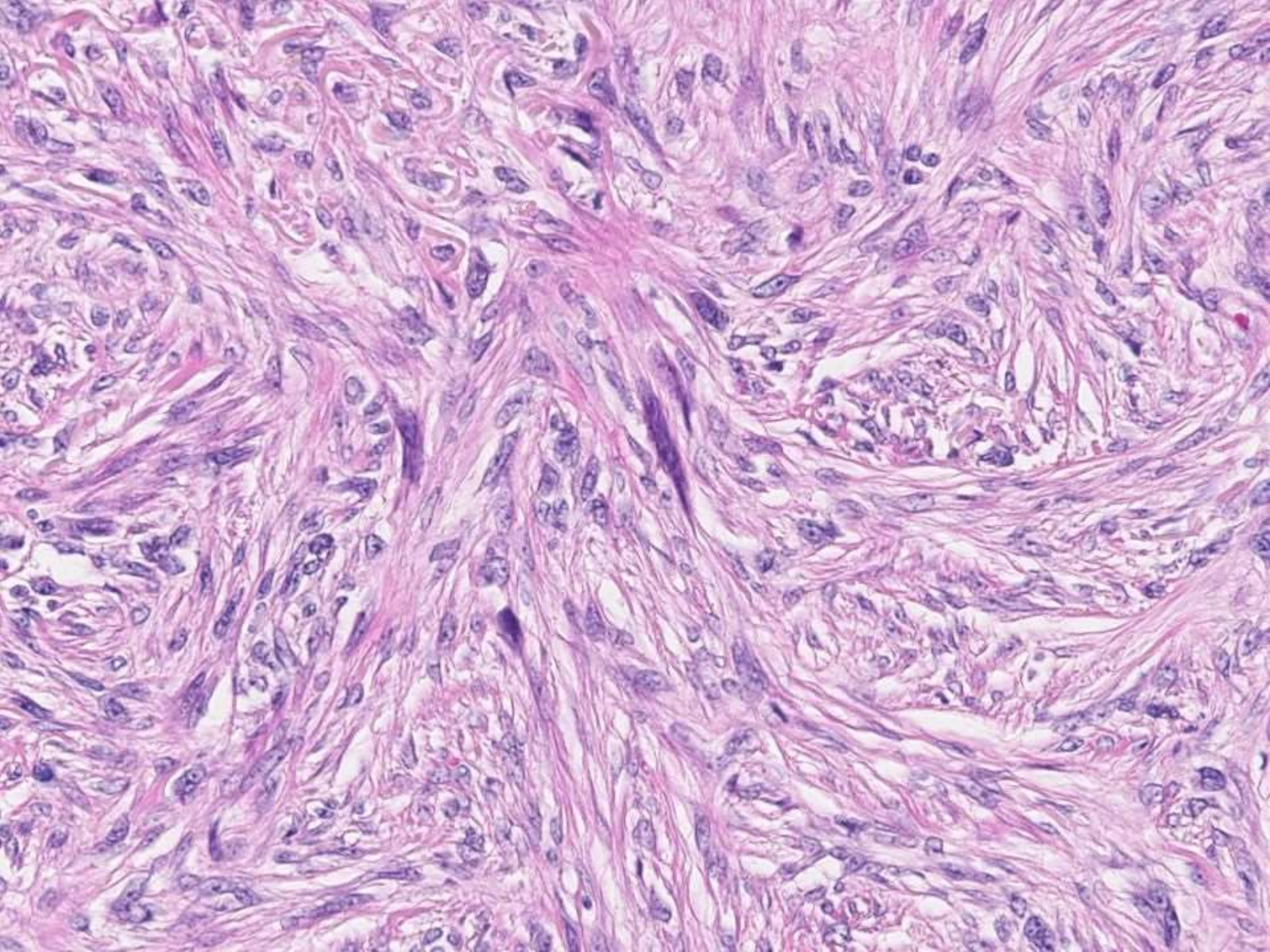
Atypical fibrous histiocytoma

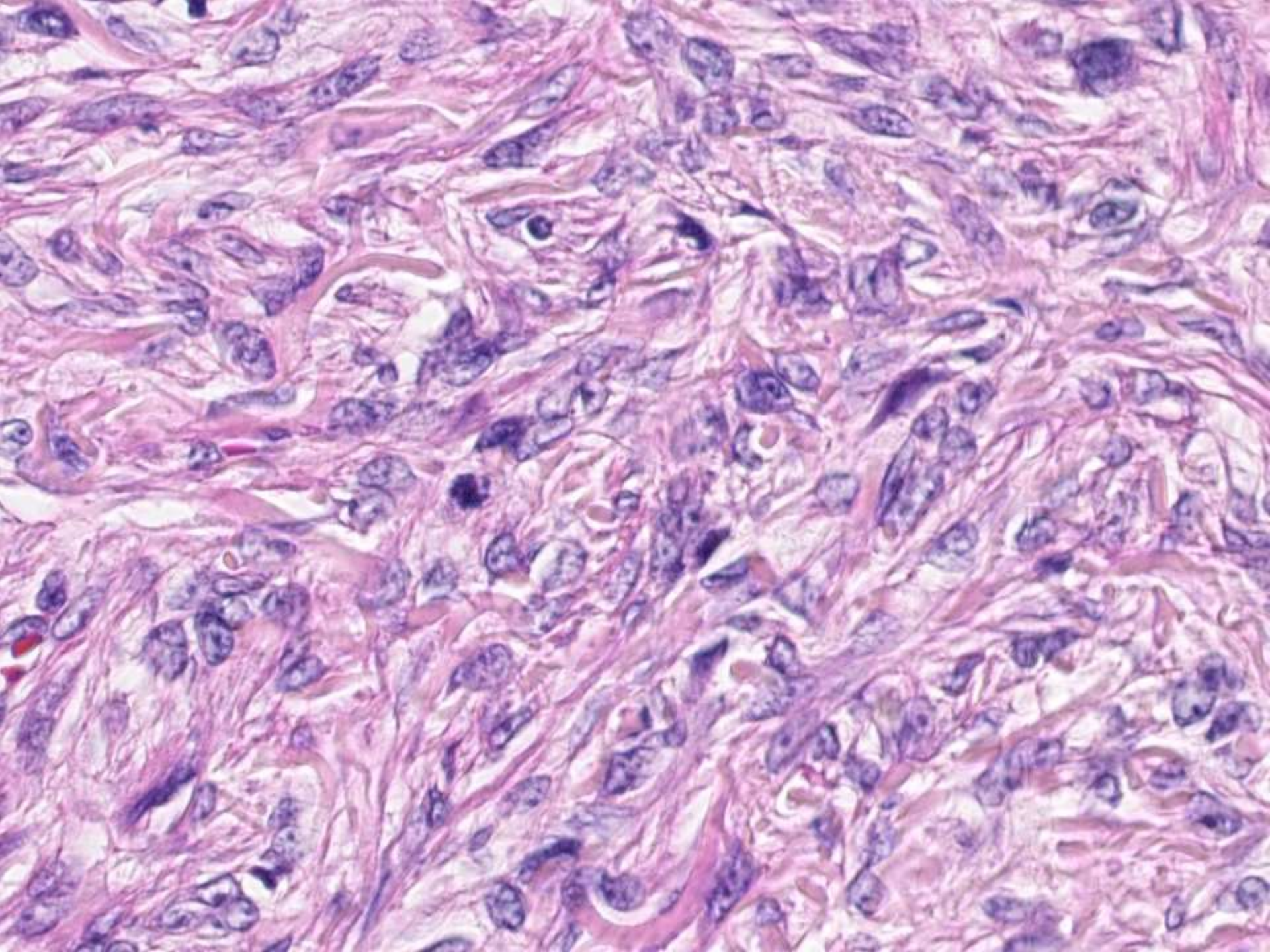


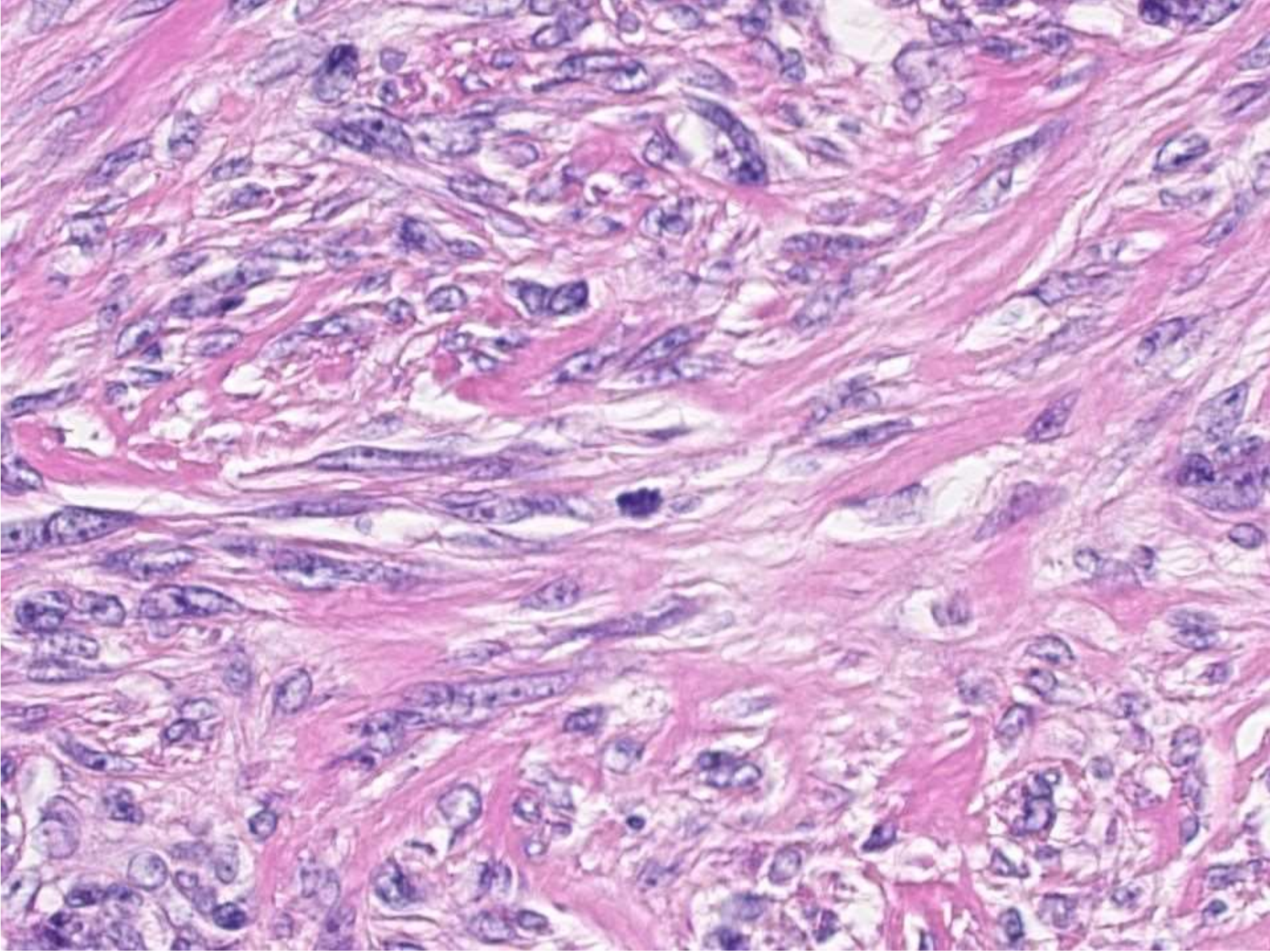


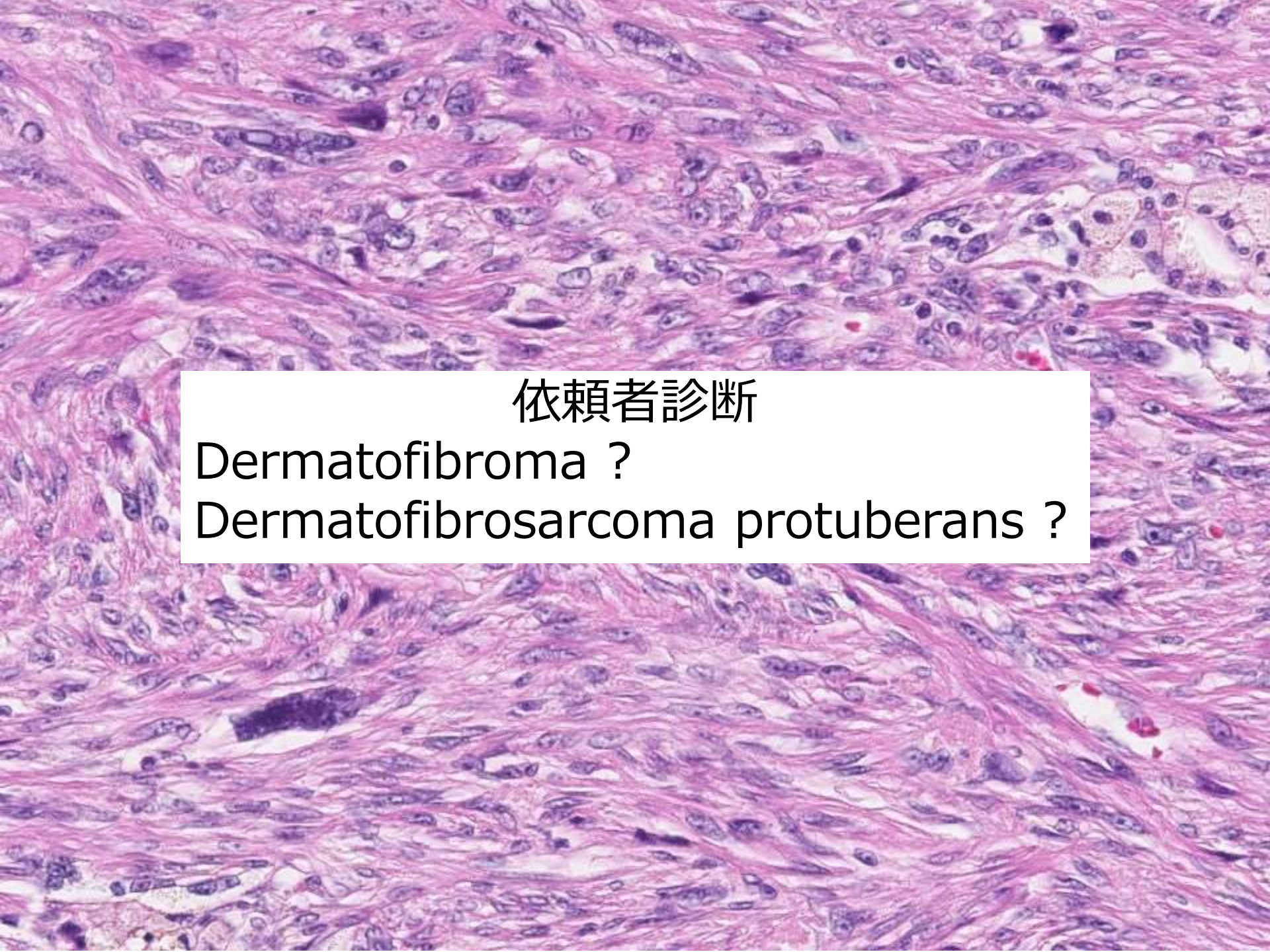






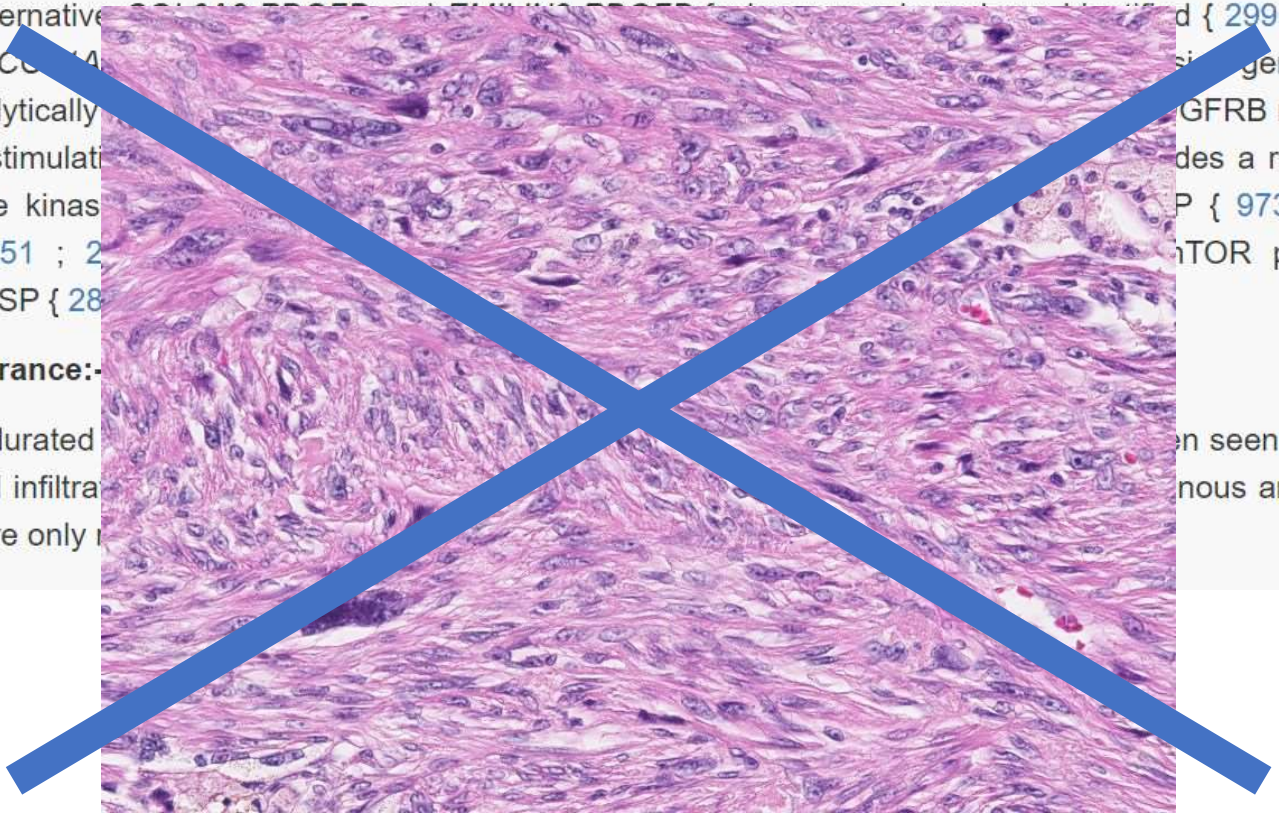





依頼者診断
Dermatofibroma ?
Dermatofibrosarcoma protuberans ?

Pathogenesis:-

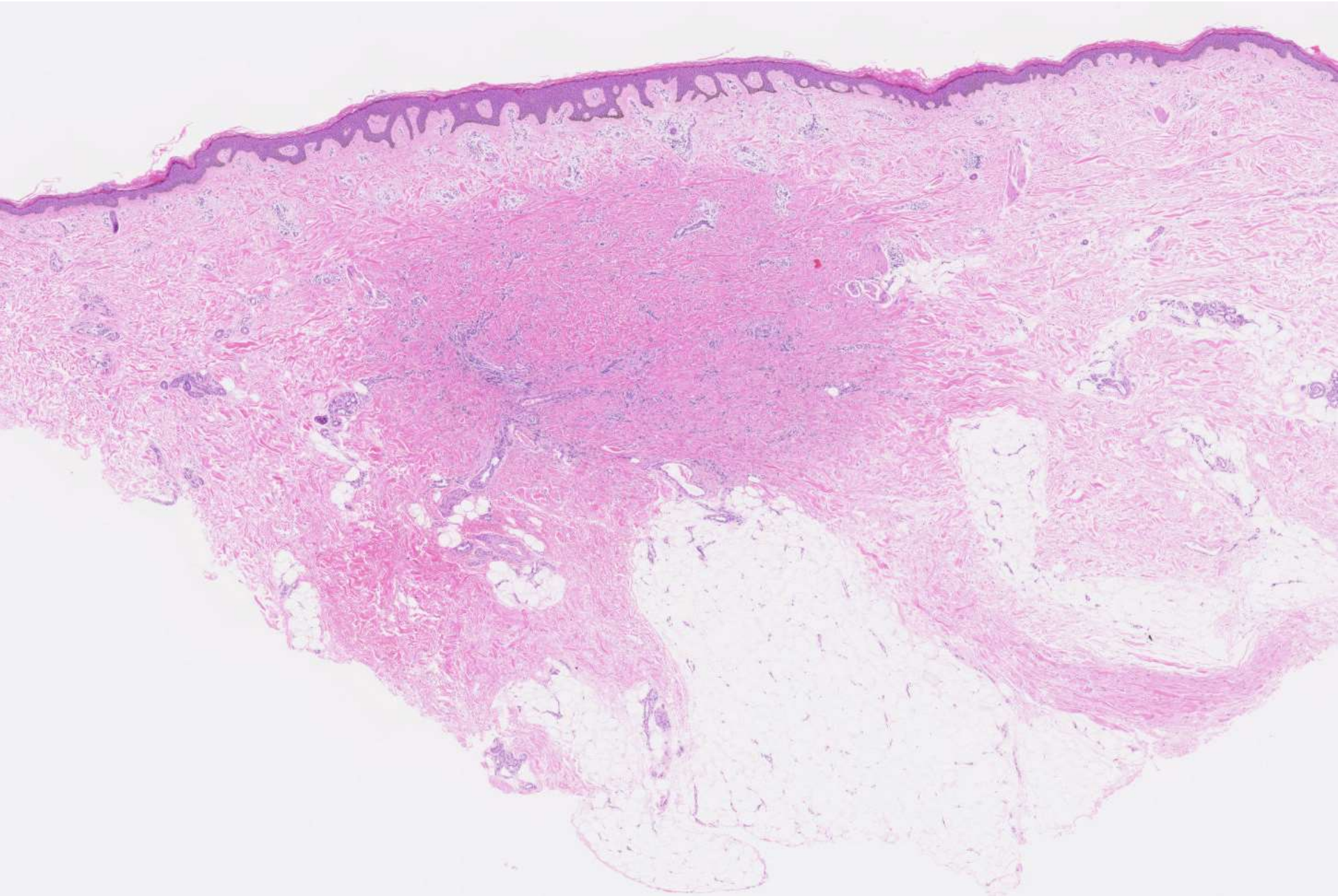
DFSP is characterized by the presence of supernumerary ring chromosomes { 12661001 ; 12550751 } that contain the centromere of chromosome 22 and comprise interspersed sequences from chromosomes 17 and 22 { 7757993 }. Additional aberrations, such as trisomy 5 and trisomy 8, are also observed { 12661001 ; 12550751 }. Unbalanced t(17;22)(q21.3;q13.1) translocations are present in most children and rarely in adults { 12661001 ; 12550751 }. Most DFSP cells harbour not only a structural rearrangement but also a gain of 17q21.3-17qter and 22q10-q31 sequences { 14633610 ; 17124411 }. Both ring and der(22)t(17;22) chromosomes contain a chimeric gene fusing *COL1A1* at 17q21.33 with *PDGFB* at 22q13.1 { 8988177 }. The breakpoint in *COL1A1* is variable: the chimeric gene is composed of at least the first 6 exons up to exon 49 of *COL1A1* and a consistent fragment retaining all but exon 1 of the *PDGFB* gene. Fewer than 5% of typical DFSP cases are negative for the *COL1A1-PDGFB* fusion gene by routine molecular testing; alternative d { 29955147 ; 30014607 }, and in some cases, C gene encodes a fusion protein that is proteolytically GFRB receptor on their cell surfaces, autocrine stimulati des a rationale for targeted therapy with tyrosine kinas P { 9739023 ; 19635106 ; 17950782 ; 20194851 ; 2 nTOR pathway is seen in fibrosarcomatous DFSP { 28

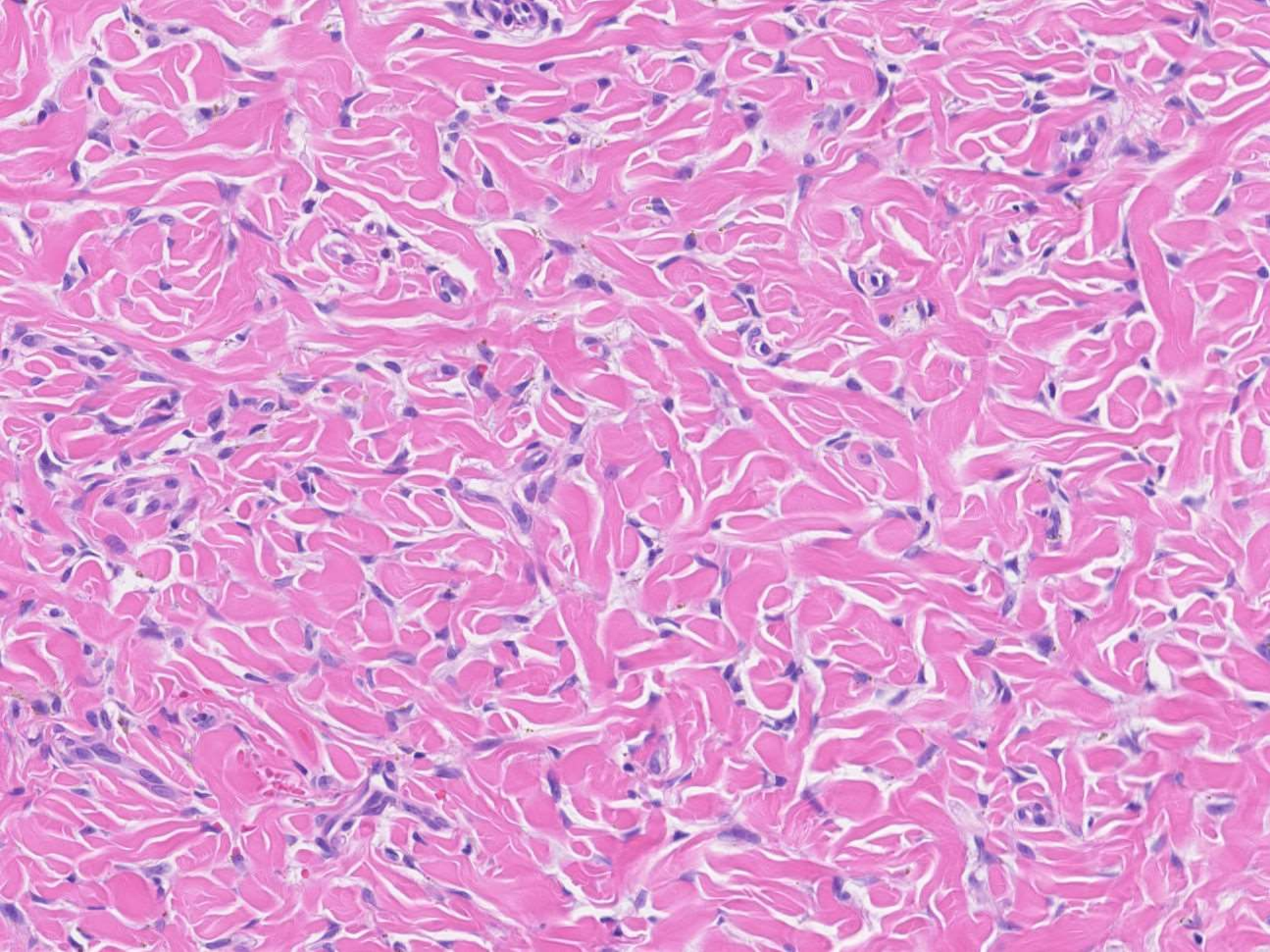


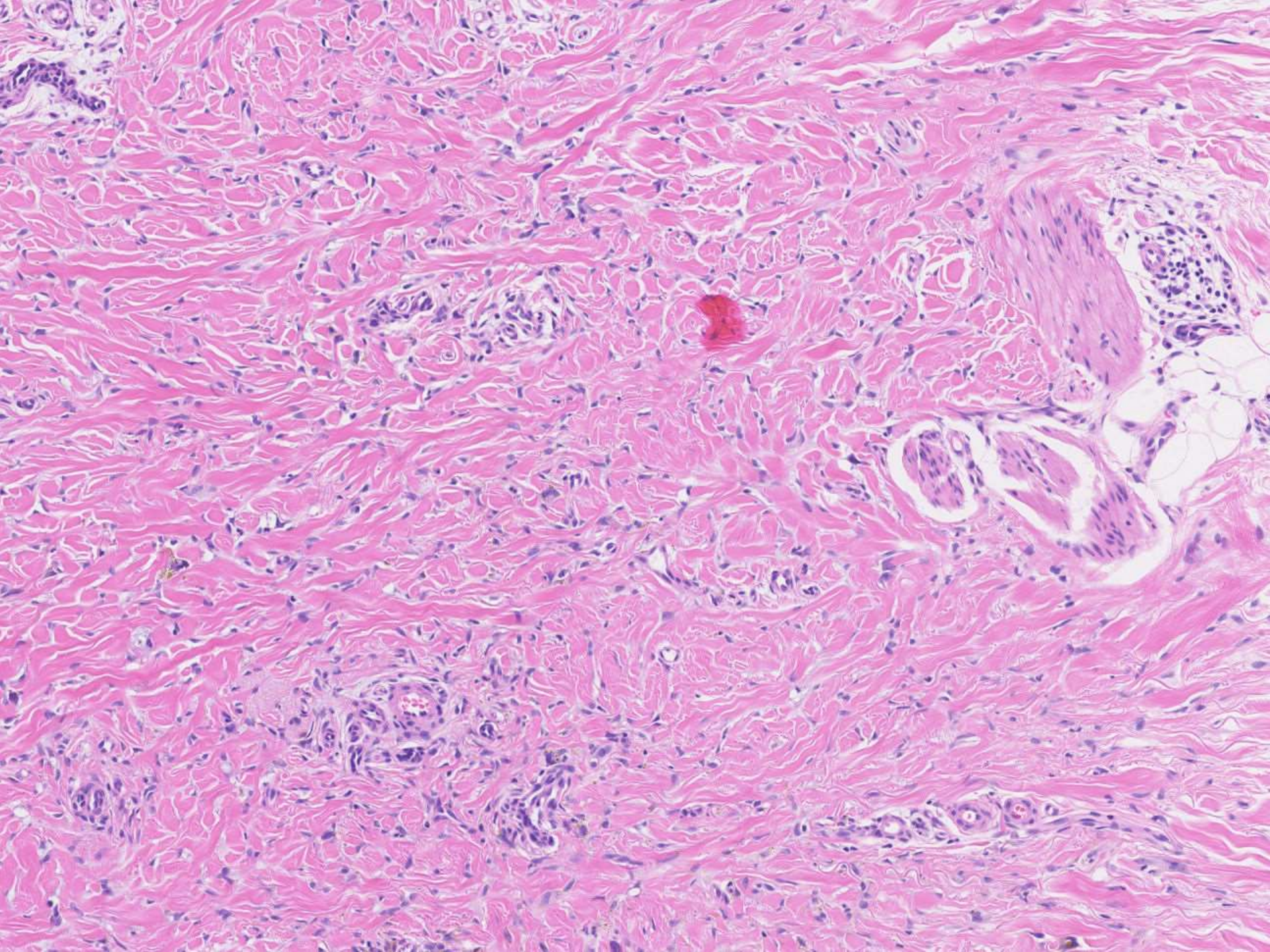
Macroscopic appearance:-

DFSP lesions are indurated n seen in recurrent lesions. These ill-defined and infiltra nous areas, whereas areas of tumour necrosis are only

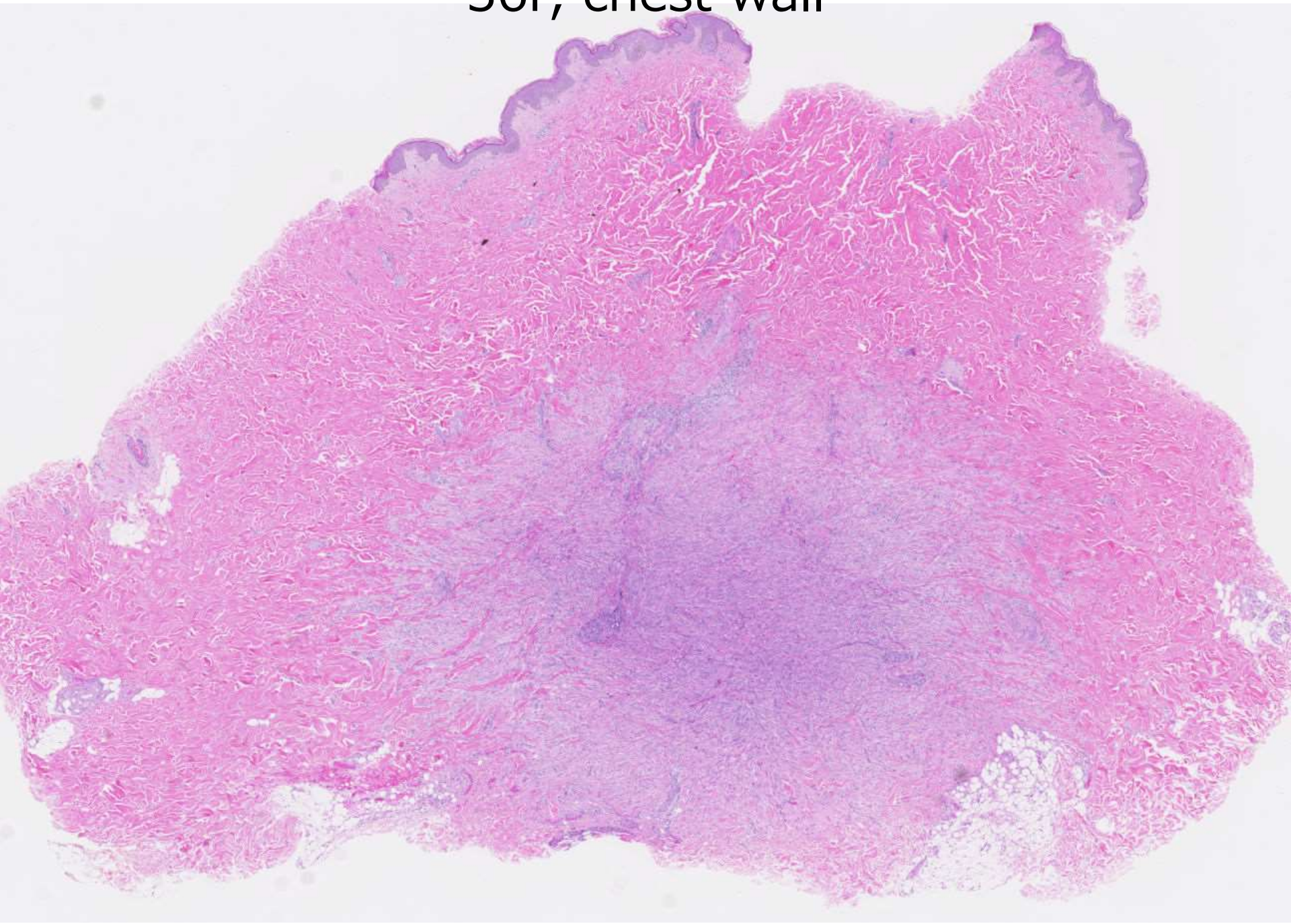
81M, lower leg

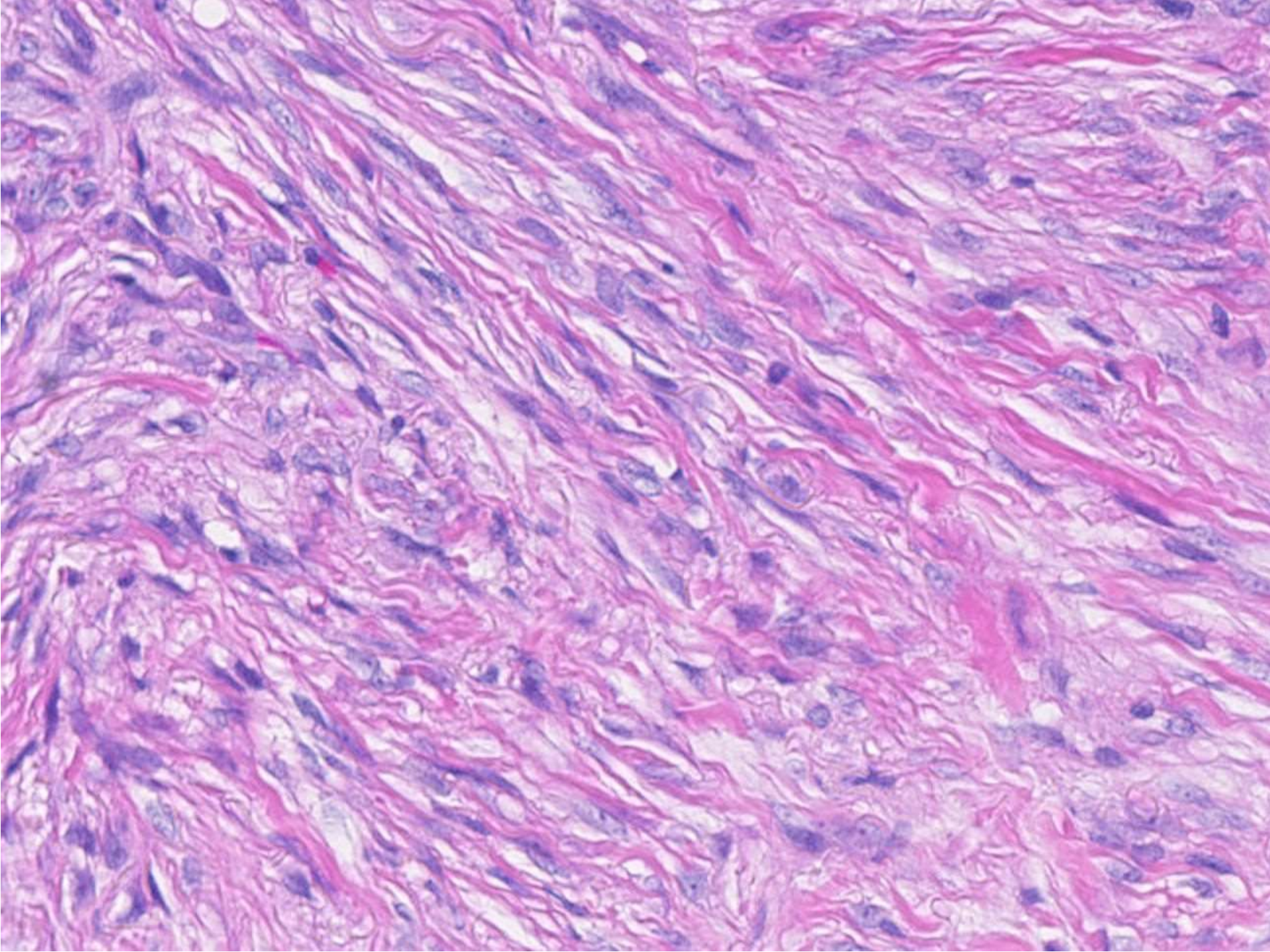


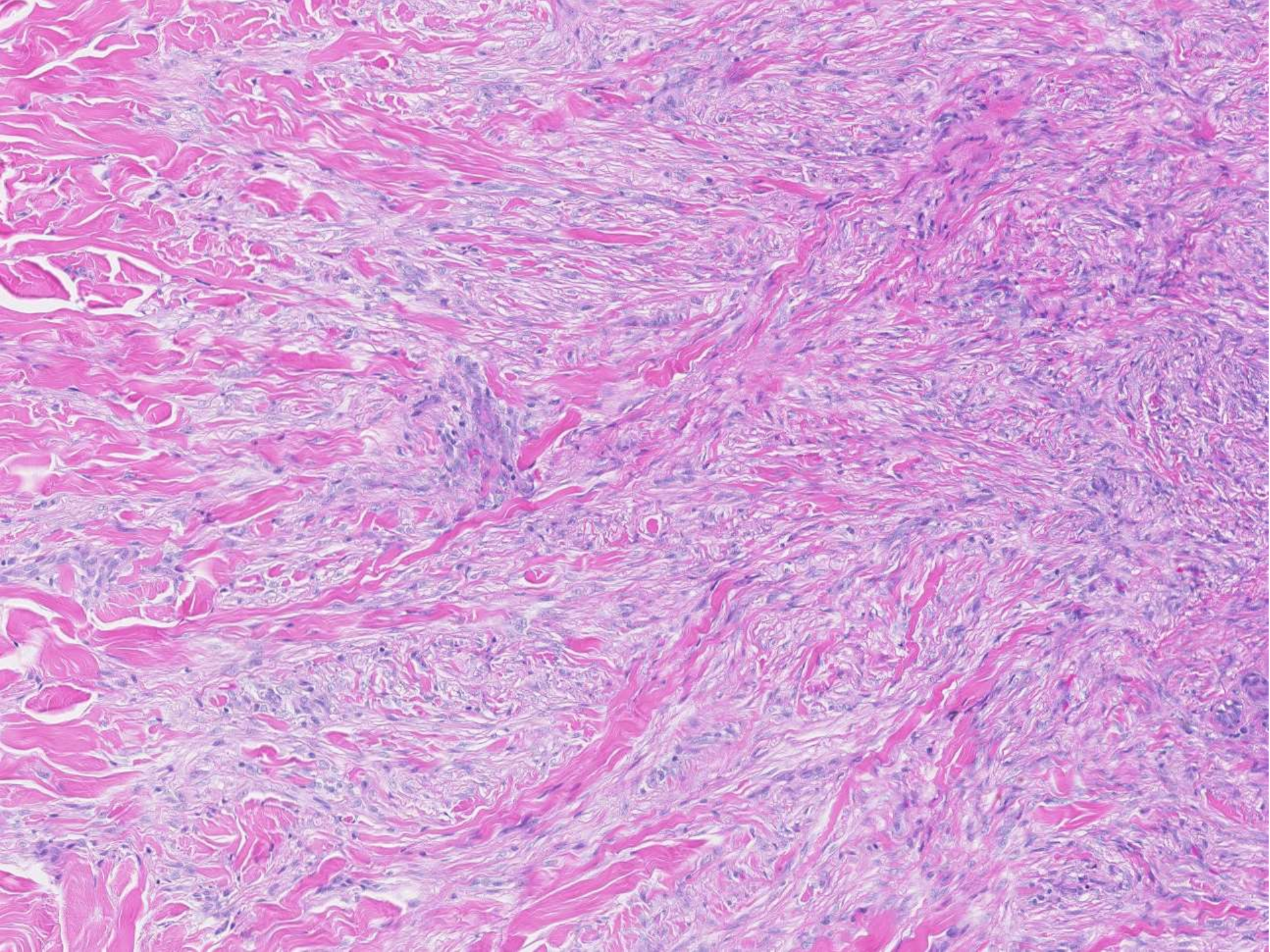




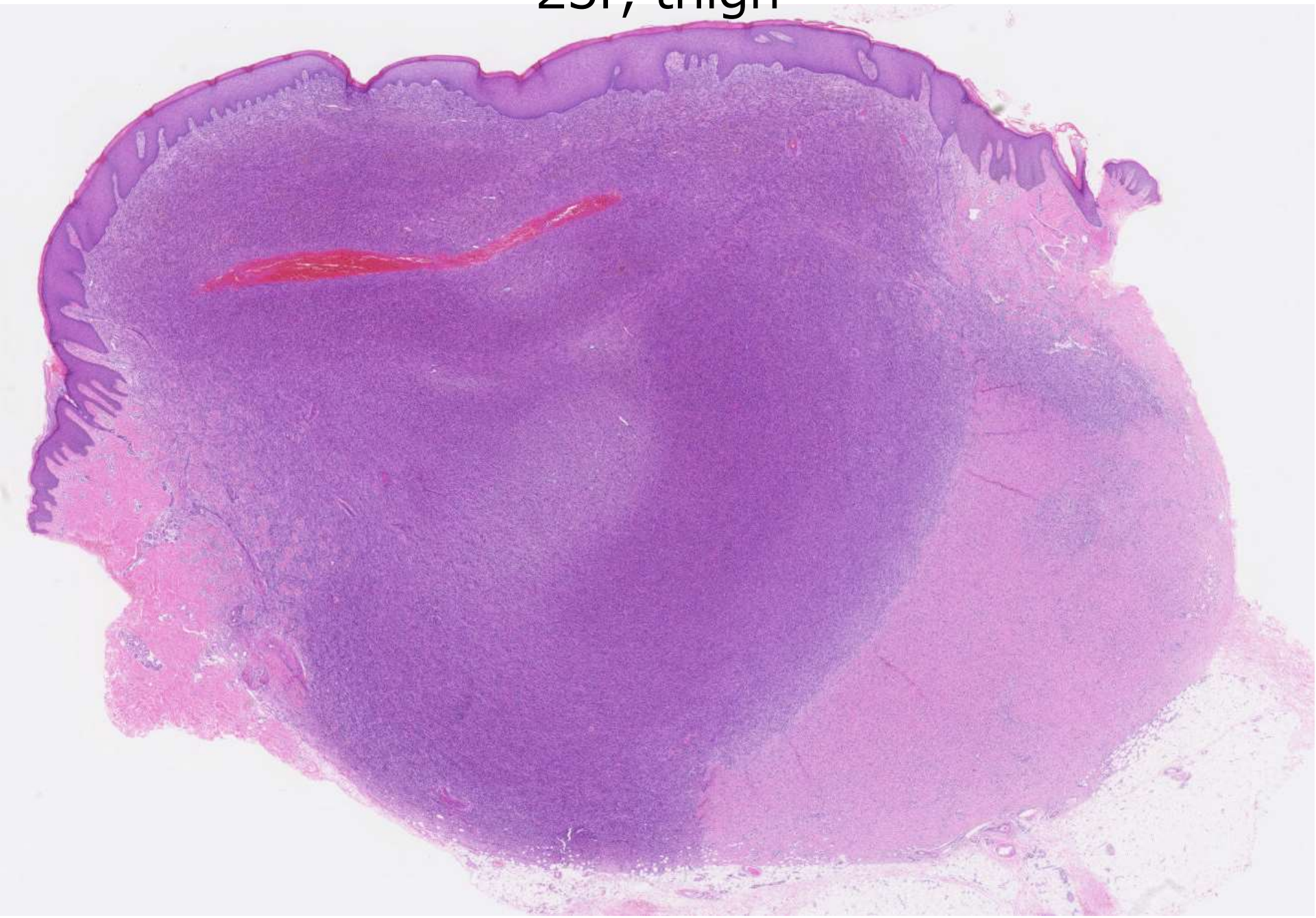
36F, chest wall

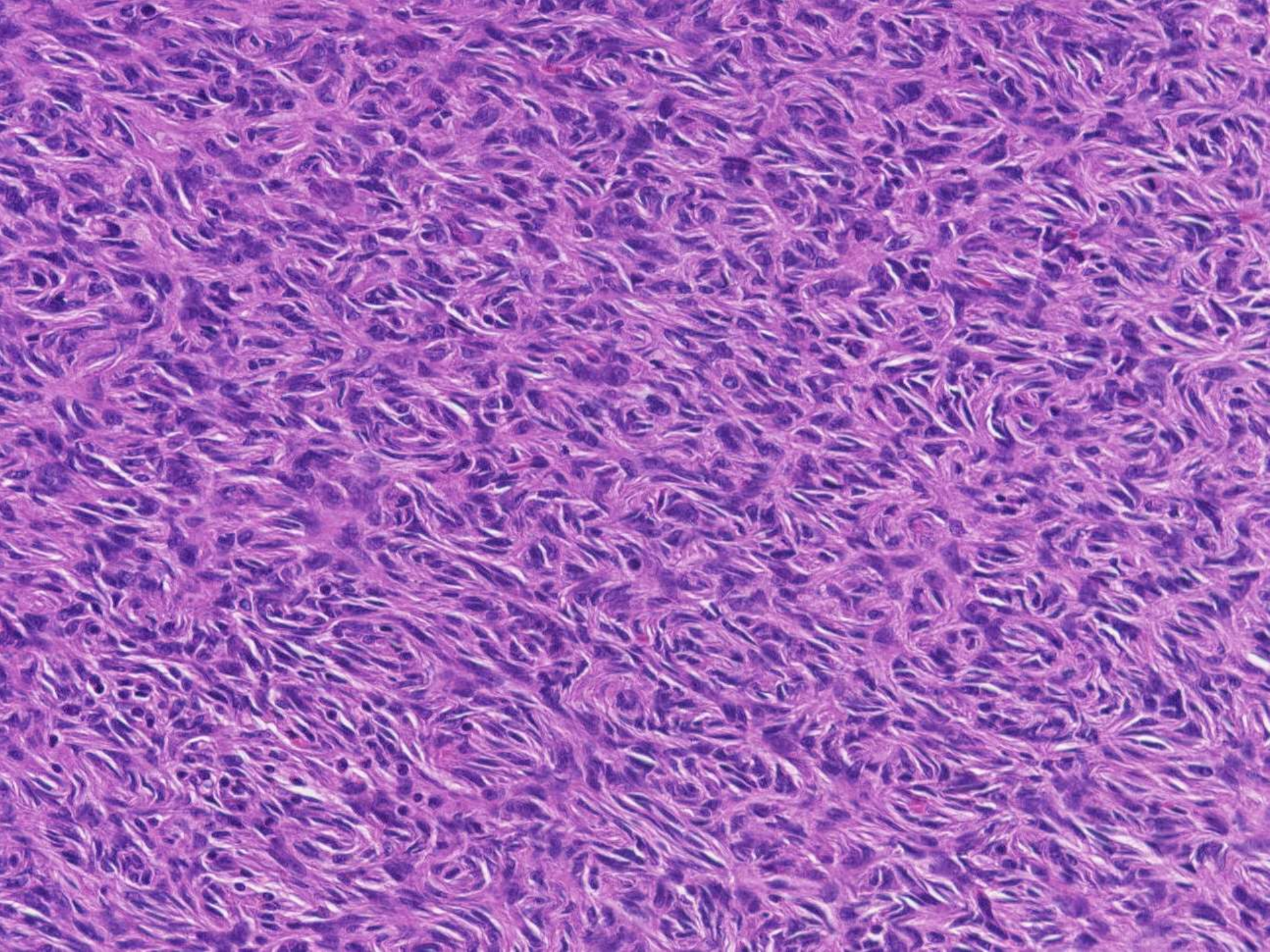


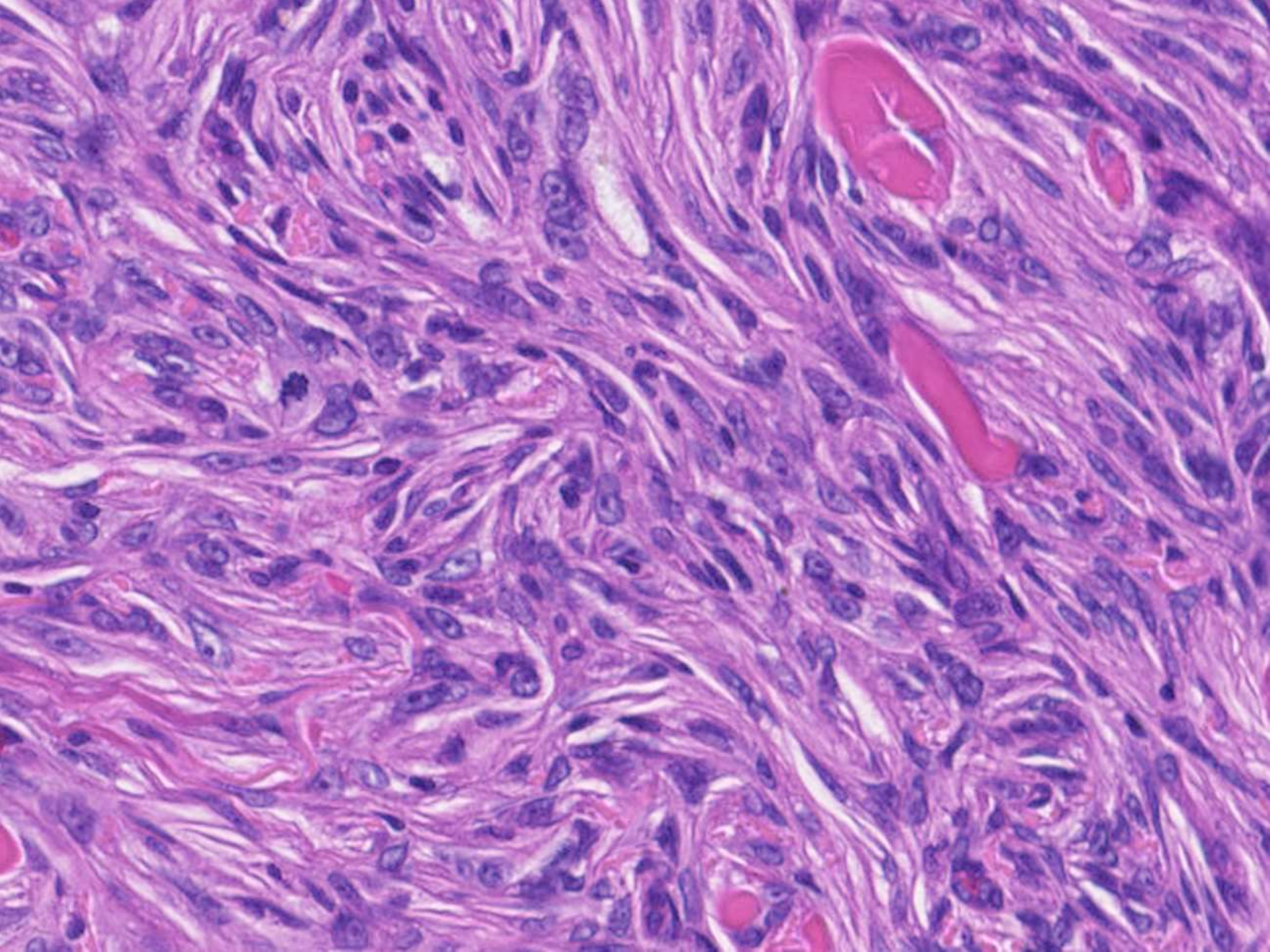


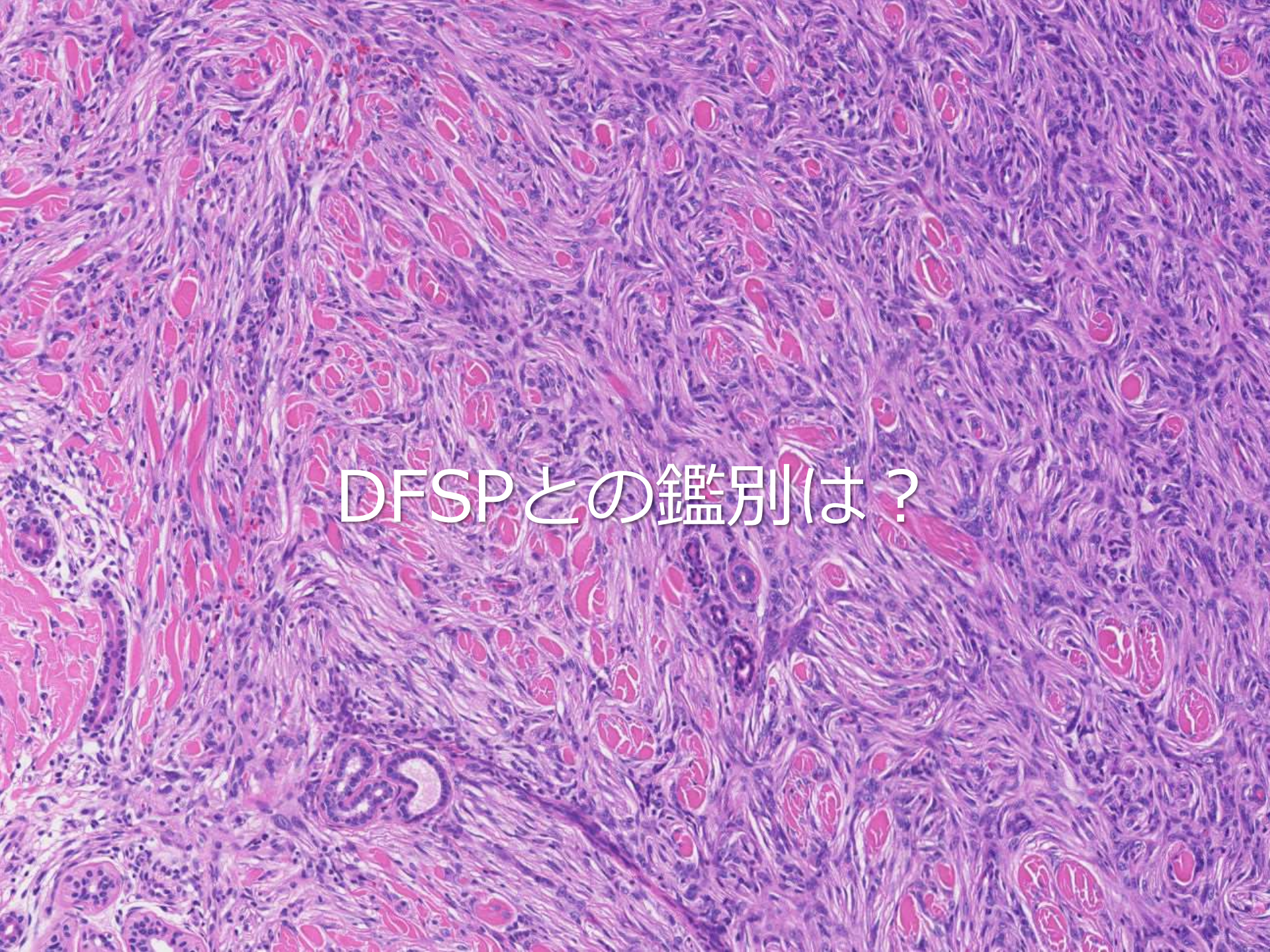


23F, thigh

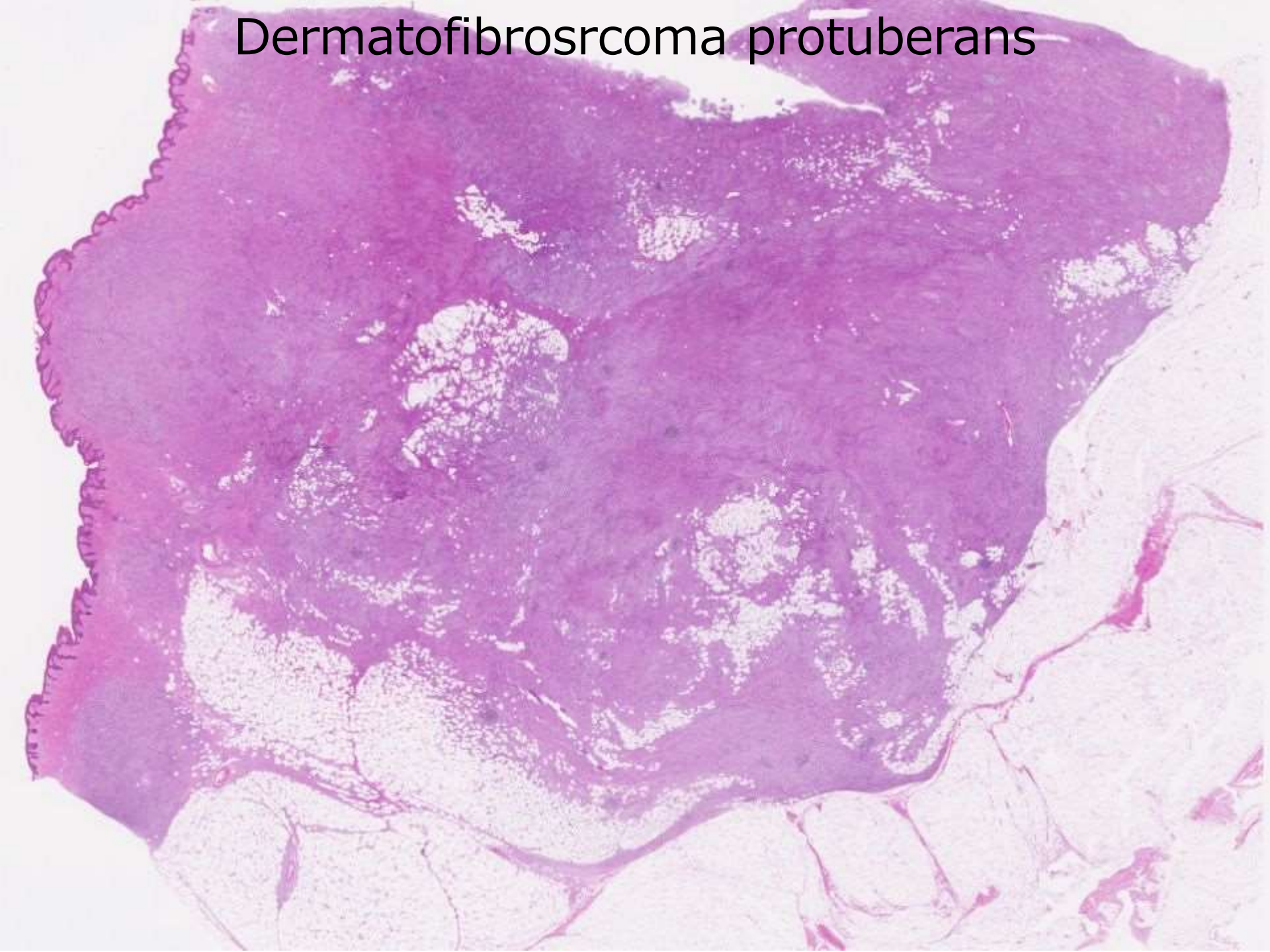


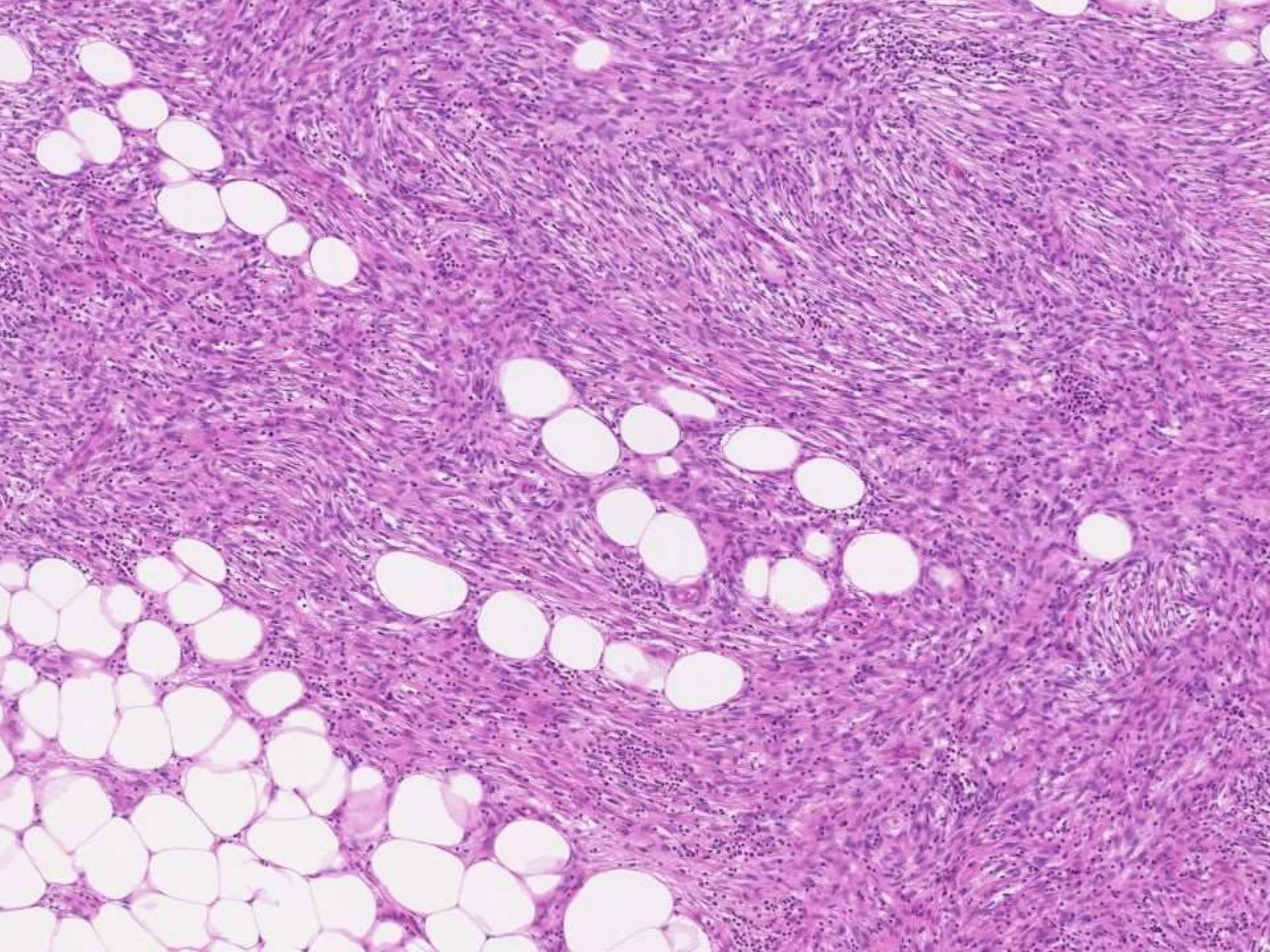


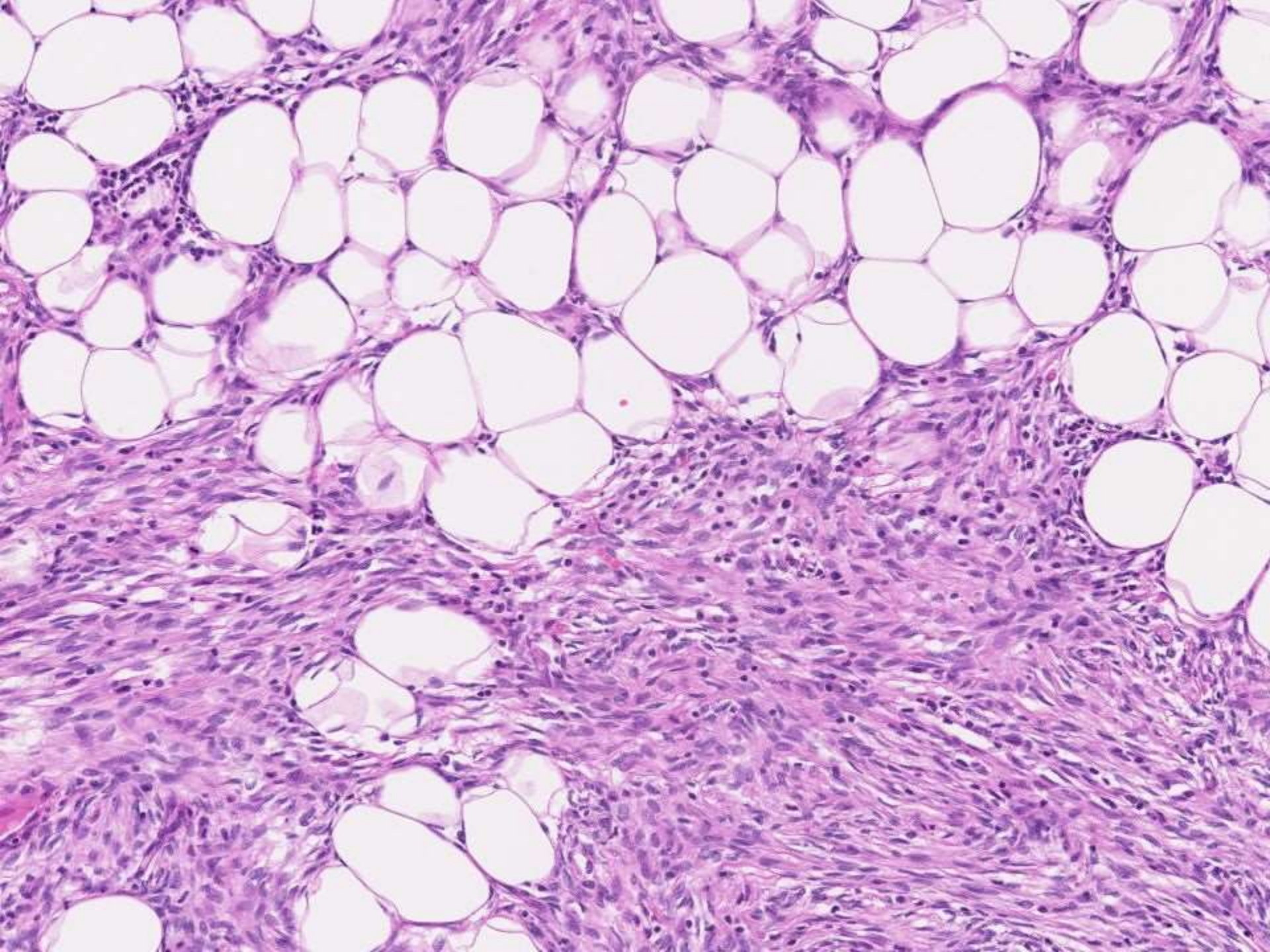


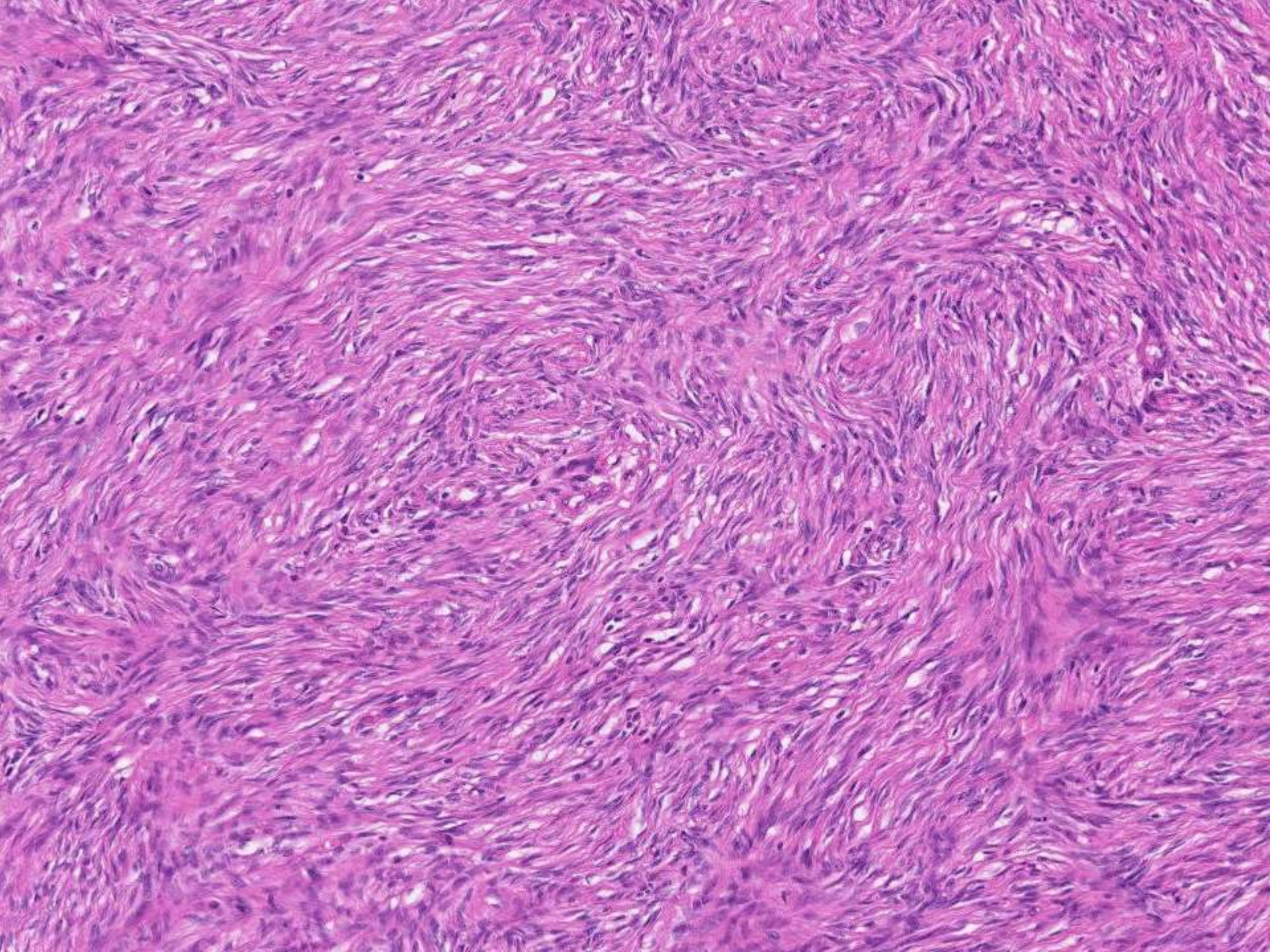

DFSPとの鑑別は？

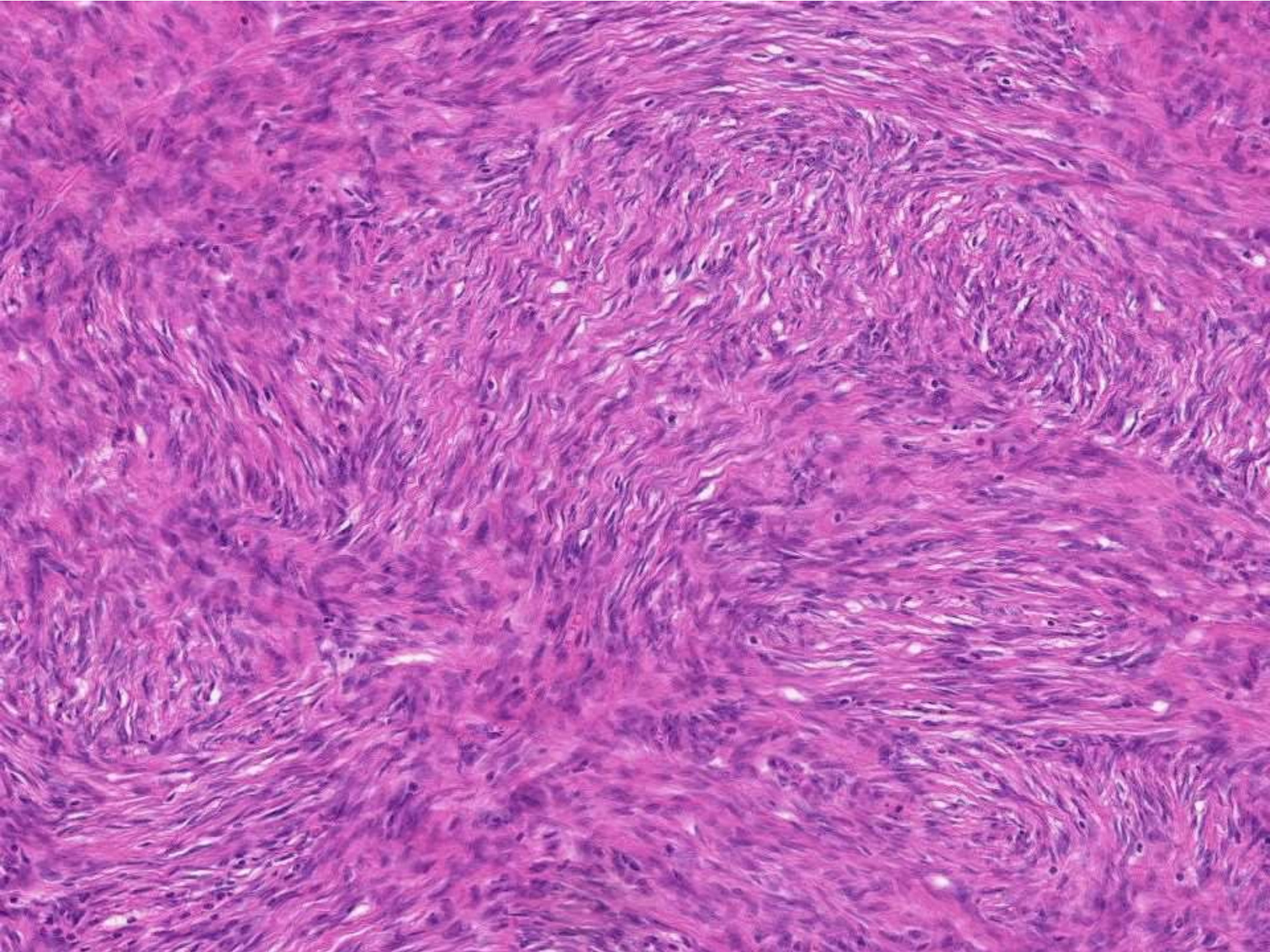
Dermatofibrosrcoma protuberans



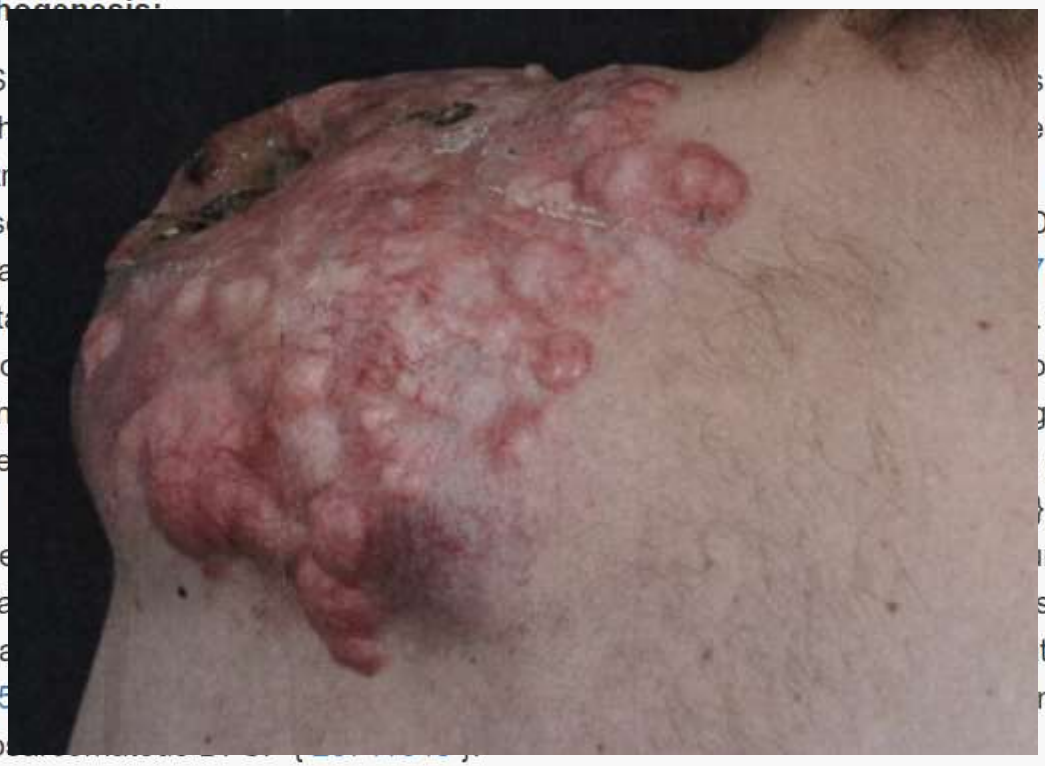


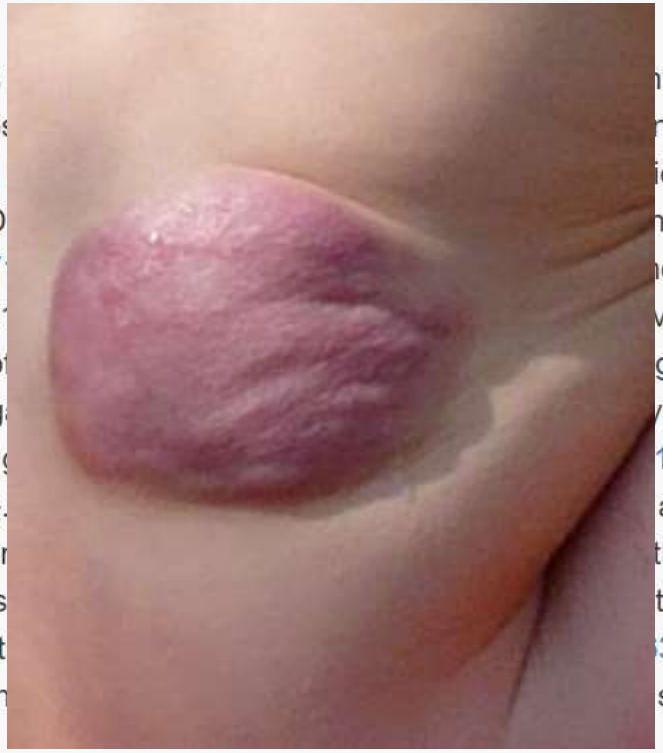






Pathogenesis:





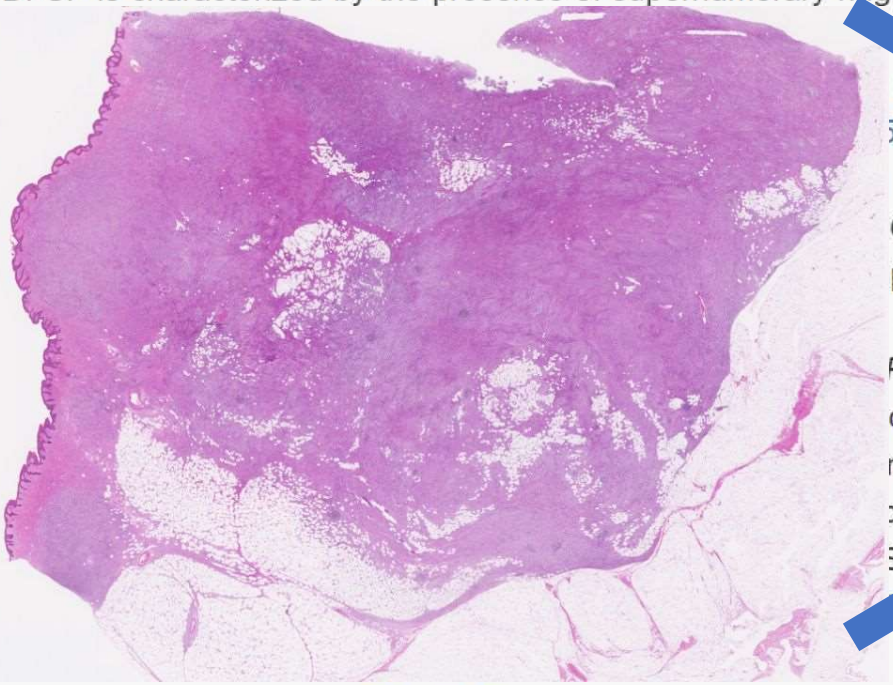
DFS ntromere of ch ns, such as tr ions are pres ngement but a osomes conta variable: the g all but exon routine mole 14607 }, and a fusion prote their cell surfa targeted thera 35106 ; 1795 seen in fibro

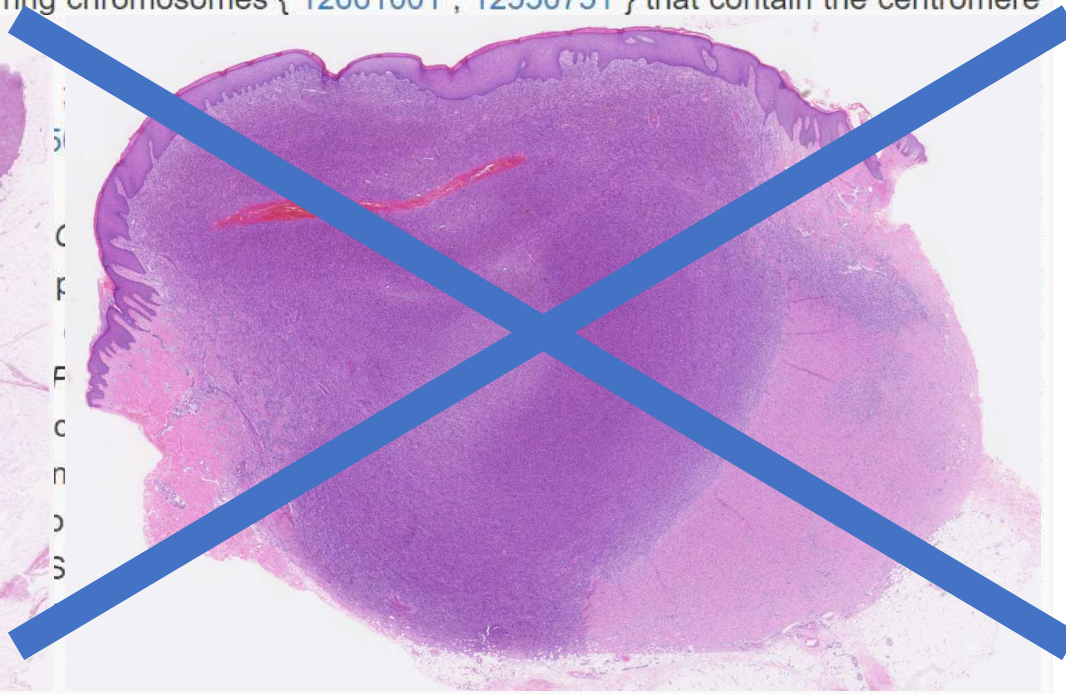
Macroscopic appearance:-

DFSP lesions are indurated plaques with one or multiple nodules. Multiple protuberant tumours are often seen in recurrent lesions. These ill-defined and infiltrative neoplasms have firm, greyish-white cut surfaces with occasional gelatinous areas, whereas areas of tumour necrosis are only rarely observed.

Pathogenesis:-

DFSP is characterized by the presence of supernumerary ring chromosomes { 12661001 ; 12550751 } that contain the centromere

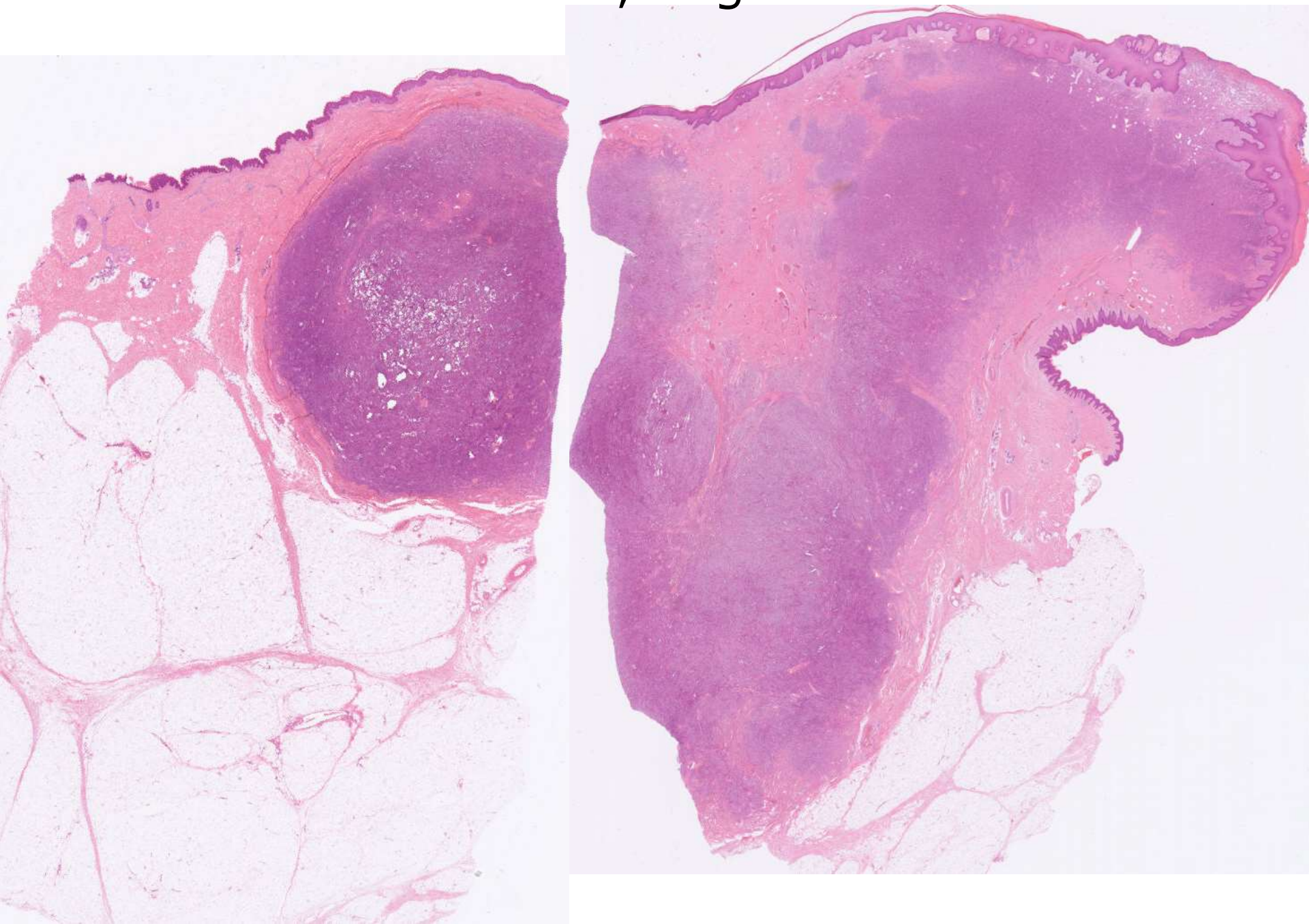


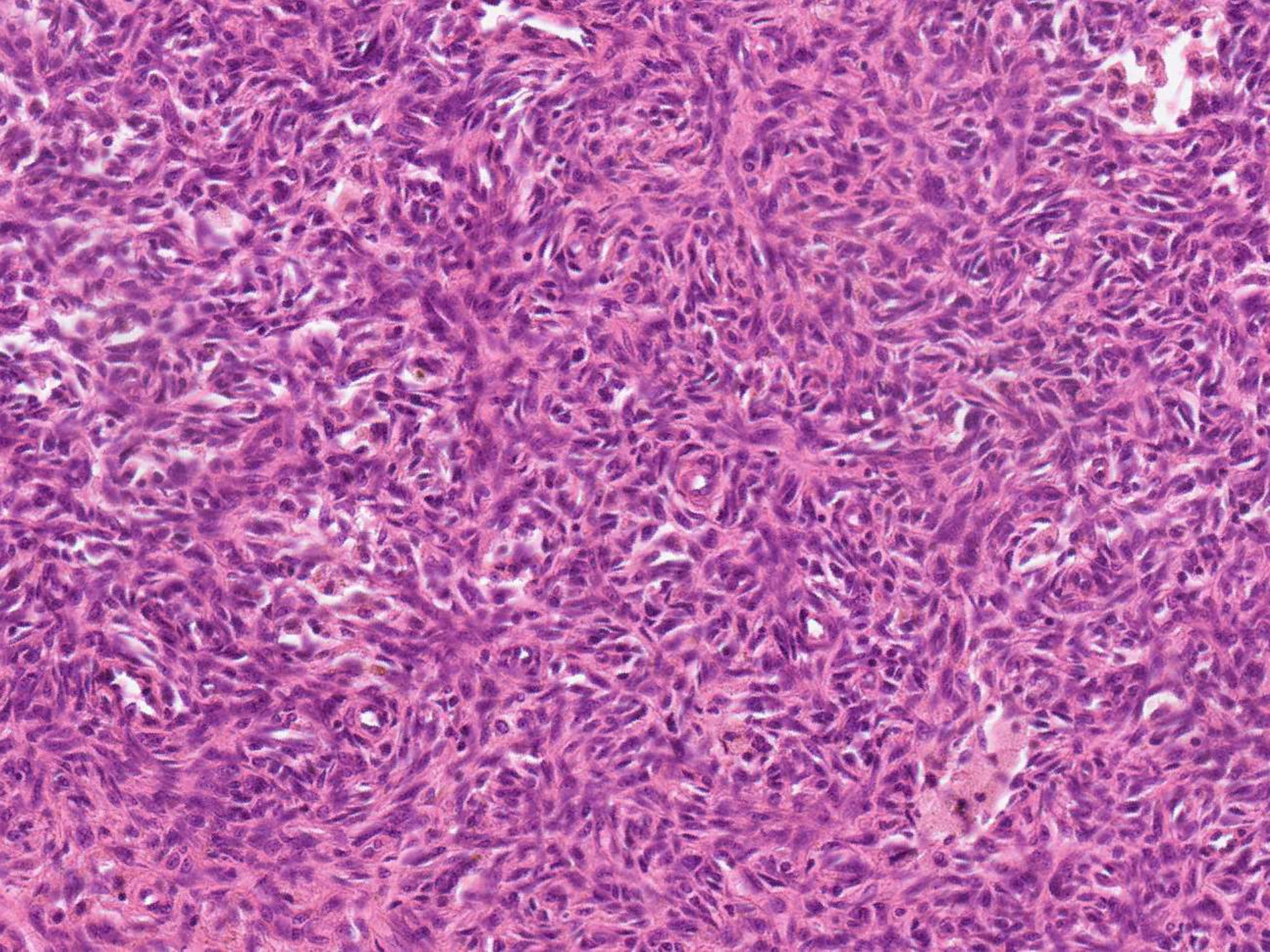


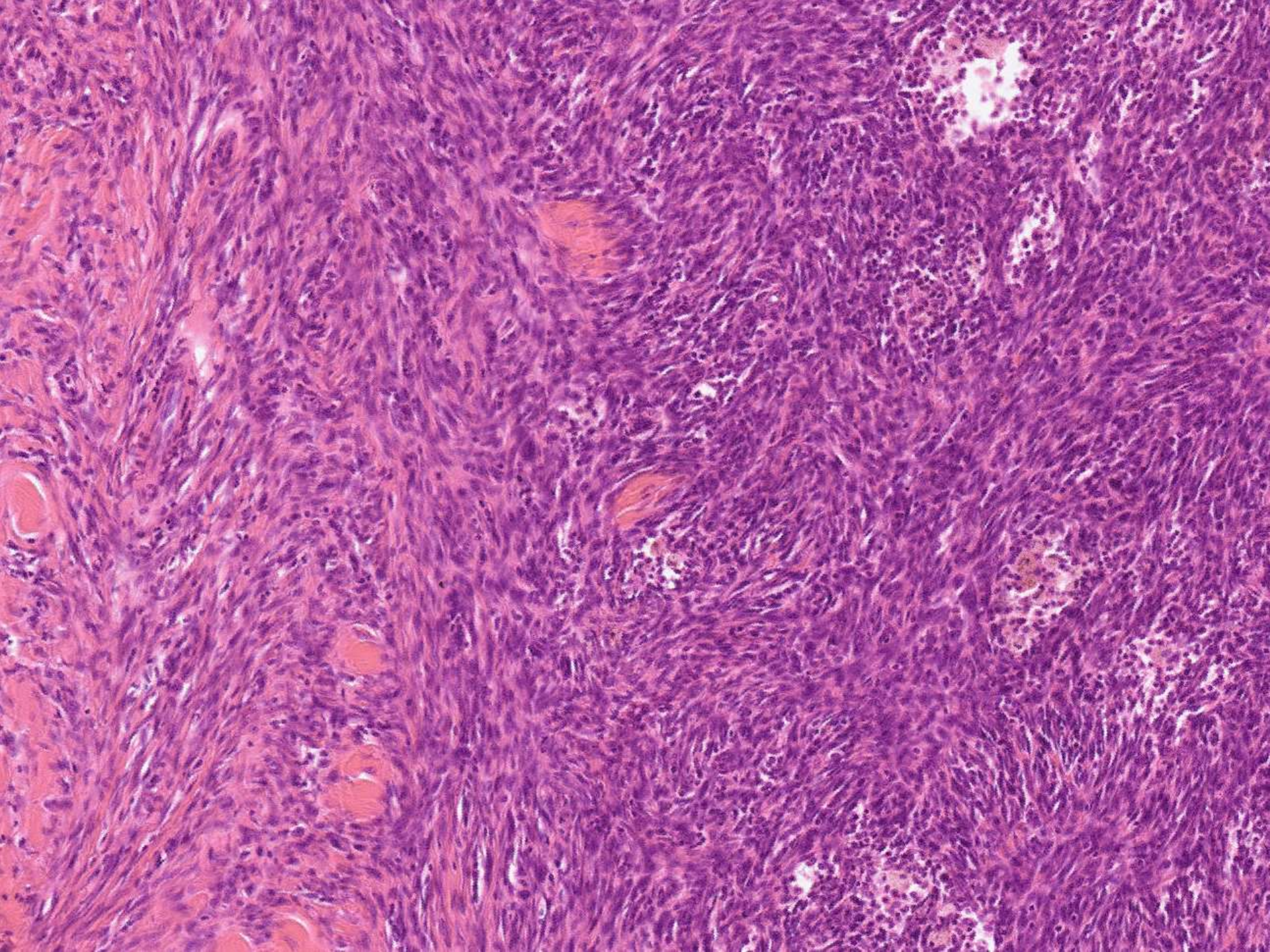
Macroscopic appearance:-

DFSP lesions are indurated plaques with one or multiple nodules. Multiple protuberant tumours are often seen in recurrent lesions. These ill-defined and infiltrative neoplasms have firm, greyish-white cut surfaces with occasional gelatinous areas, whereas areas of tumour necrosis are only rarely observed.

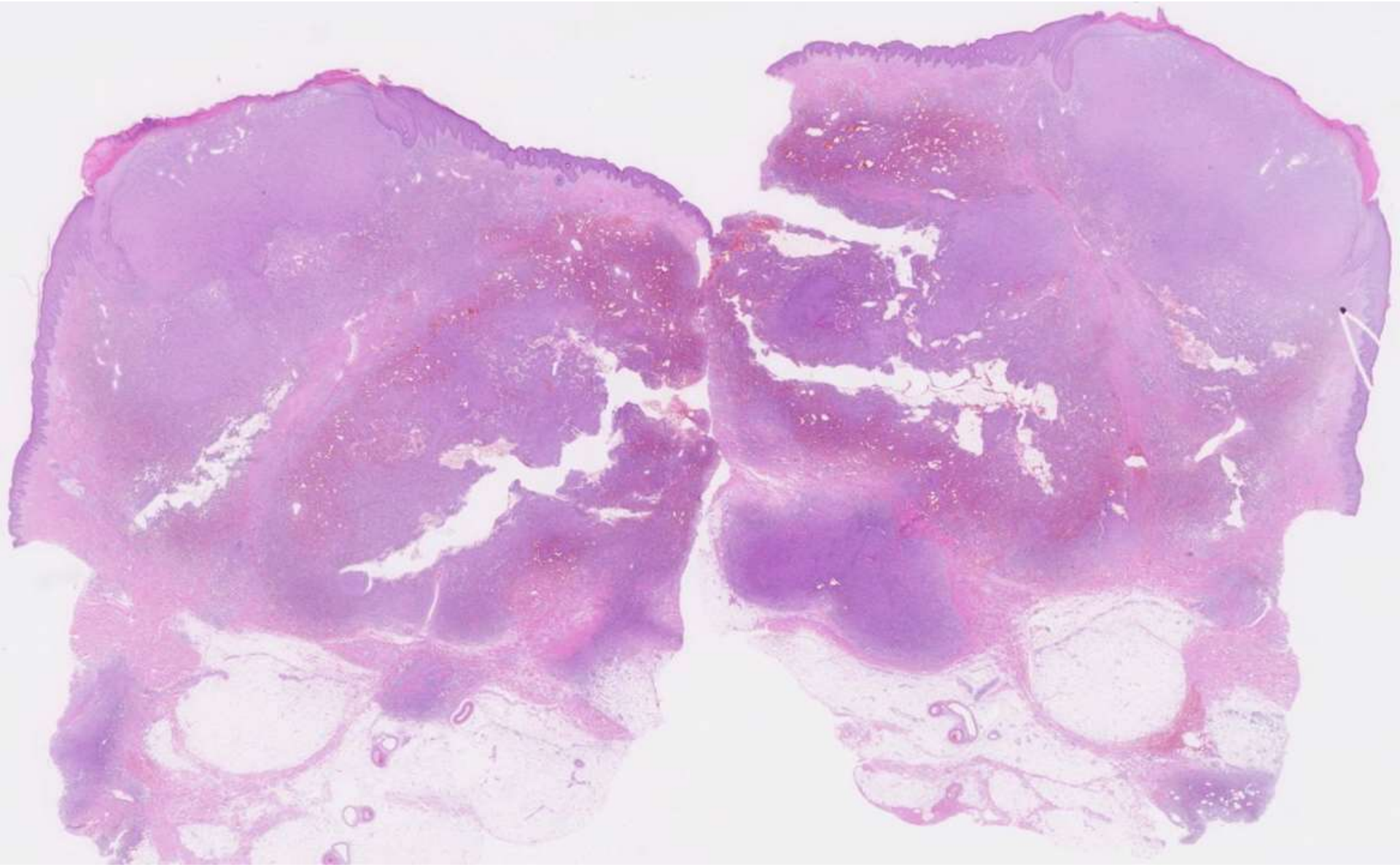
18F, thigh

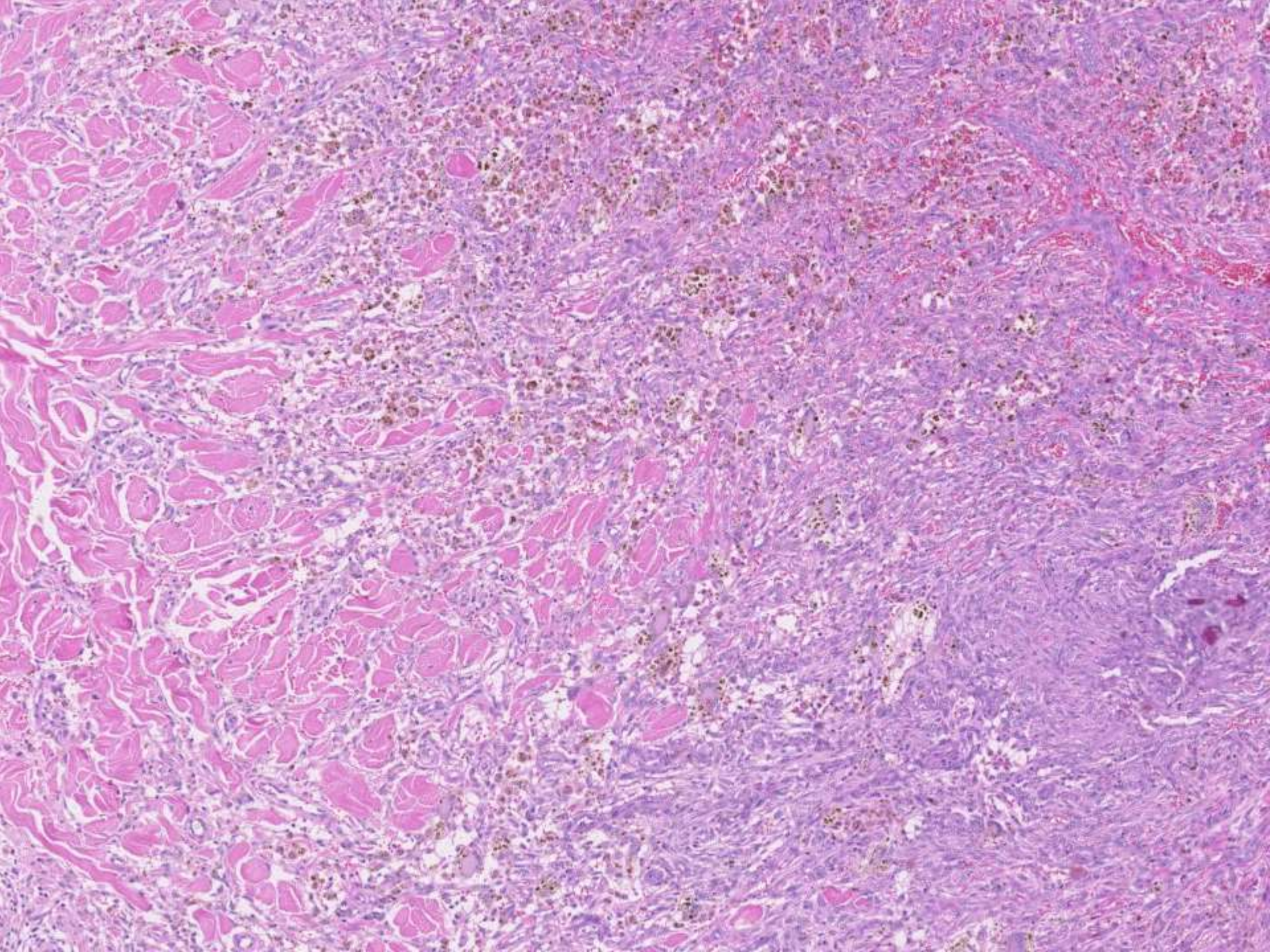


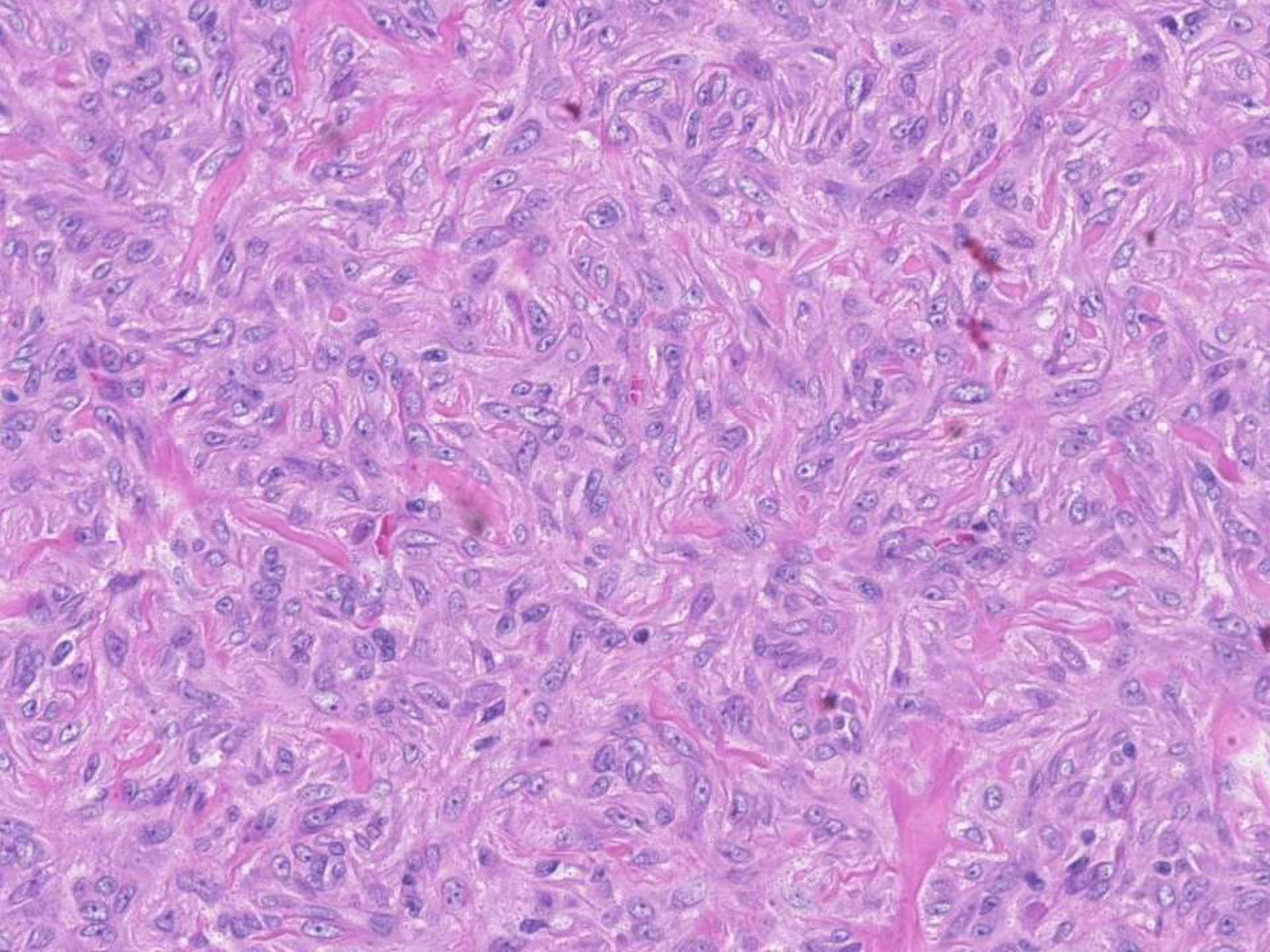


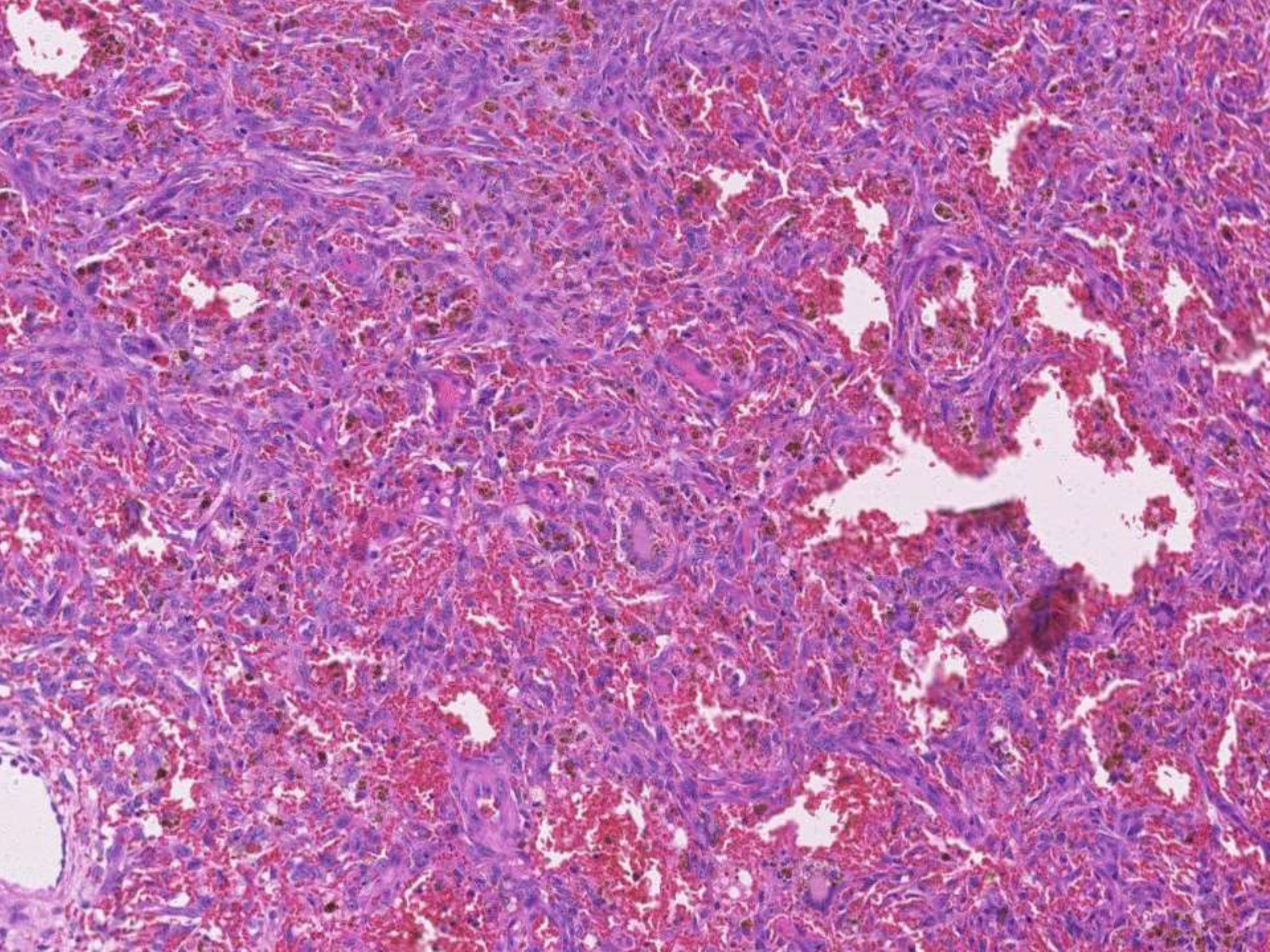


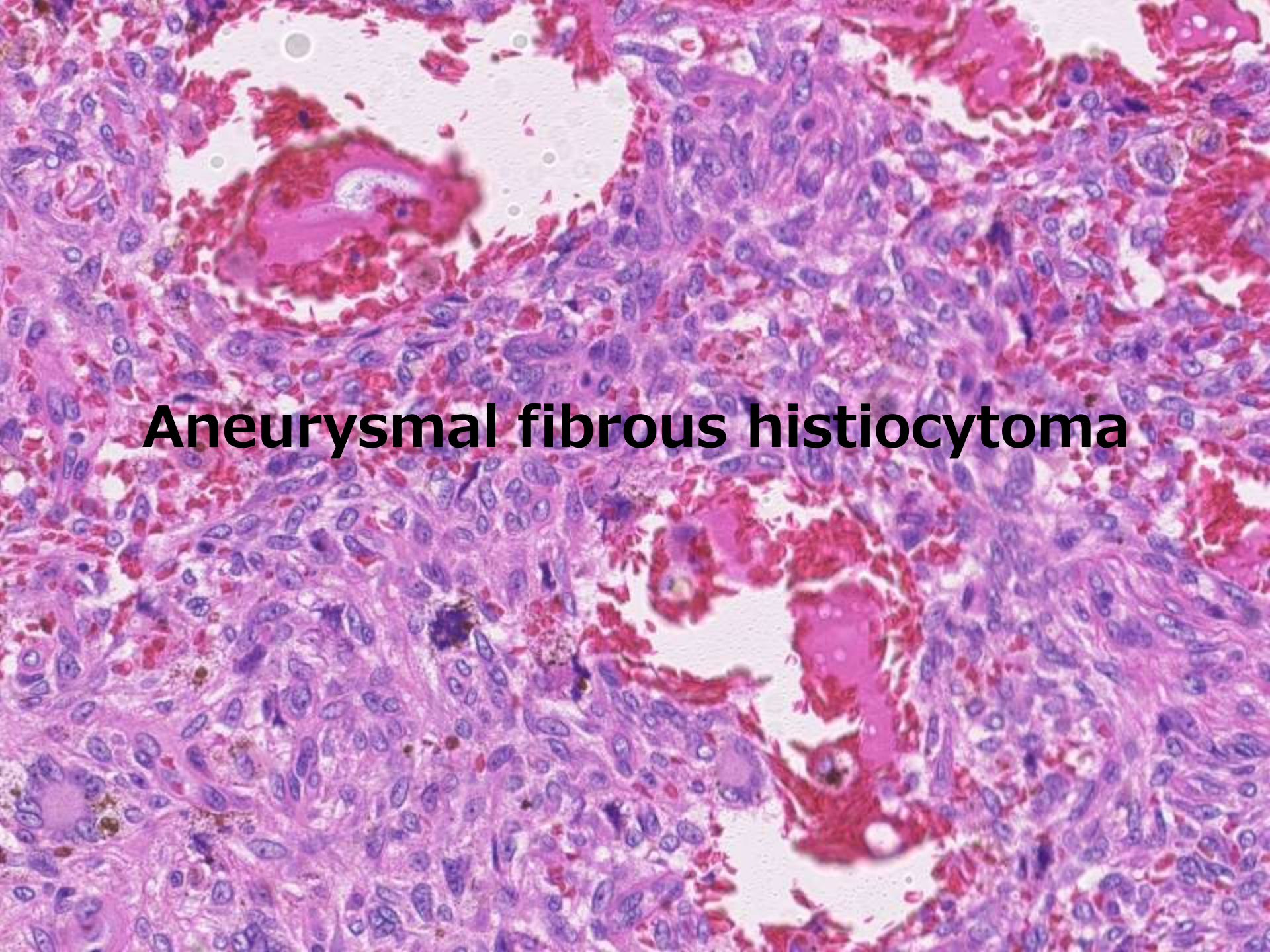
17M, loin










Aneurysmal fibrous histiocytoma

Aneurysmal fibrous histiocytoma

benign fibrous histiocytoma (dermatofibroma)の亜型だが、
稀に転移する

Angiomatoid fibrous histiocytoma

以前はangiomatoid malignant fibrous histiocytomaと呼ばれていた
Tumor of uncertain differentiationに含まれるintermediate tumor

Metastasizing dermatofibroma (fibrous histiocytoma)

- Atypical fibrous histiocytoma
- Aneurysmal fibrous histiocytoma
- (Cellular fibrous histiocytoma)

ELSEVIER

Cancer Genetics 208 (2015) 545–551

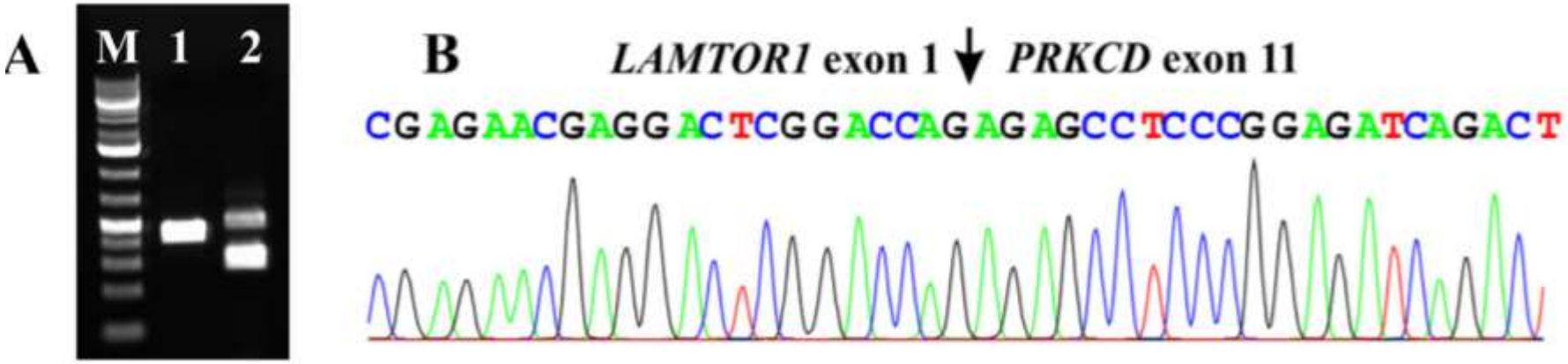
Cancer Genetics

ORIGINAL ARTICLE

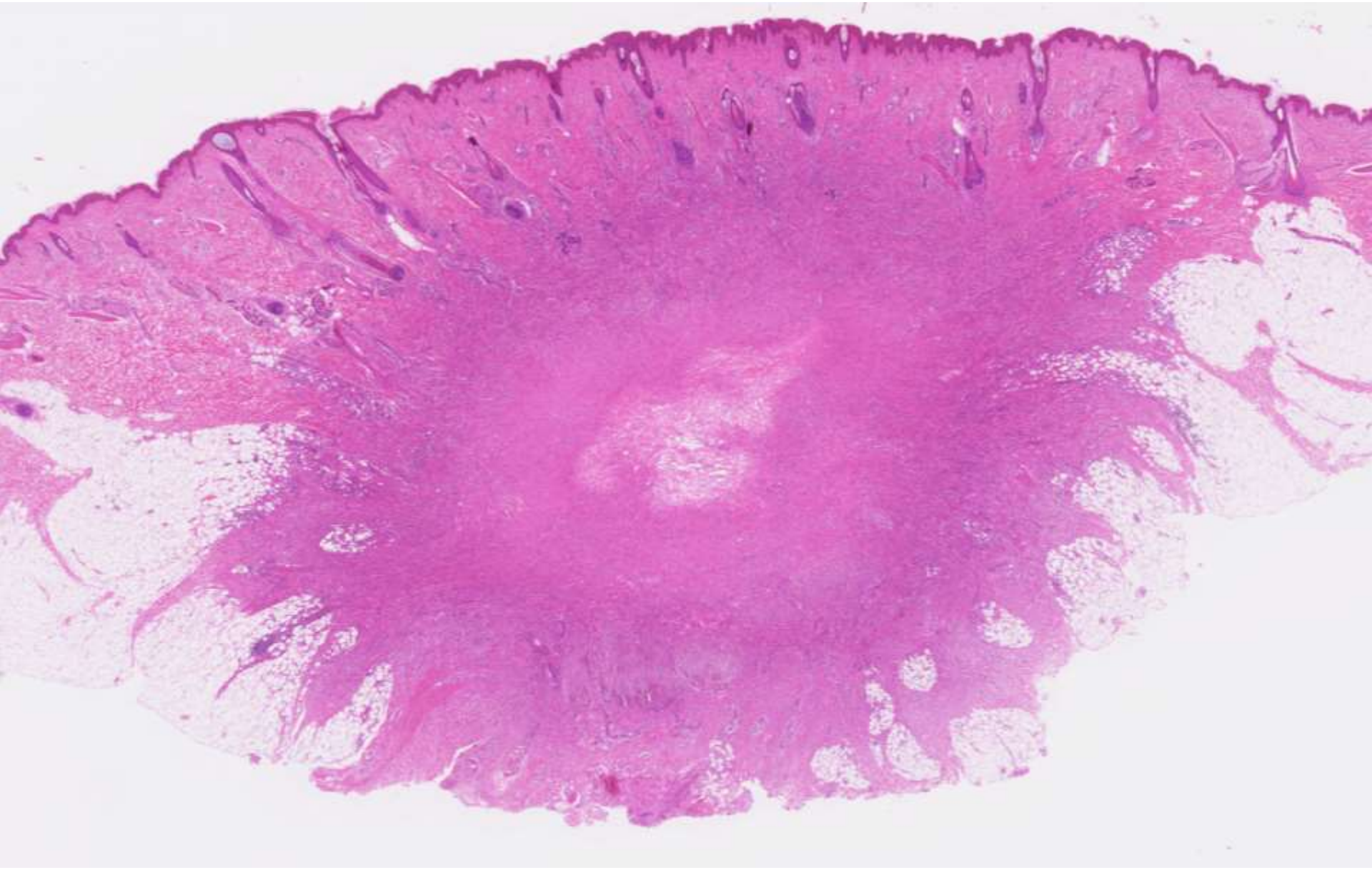
LAMTOR1-PRKCD and *NUMA1-SFMBT1* fusion genes identified by RNA sequencing in aneurysmal benign fibrous histiocytoma with t(3;11)(p21;q13)

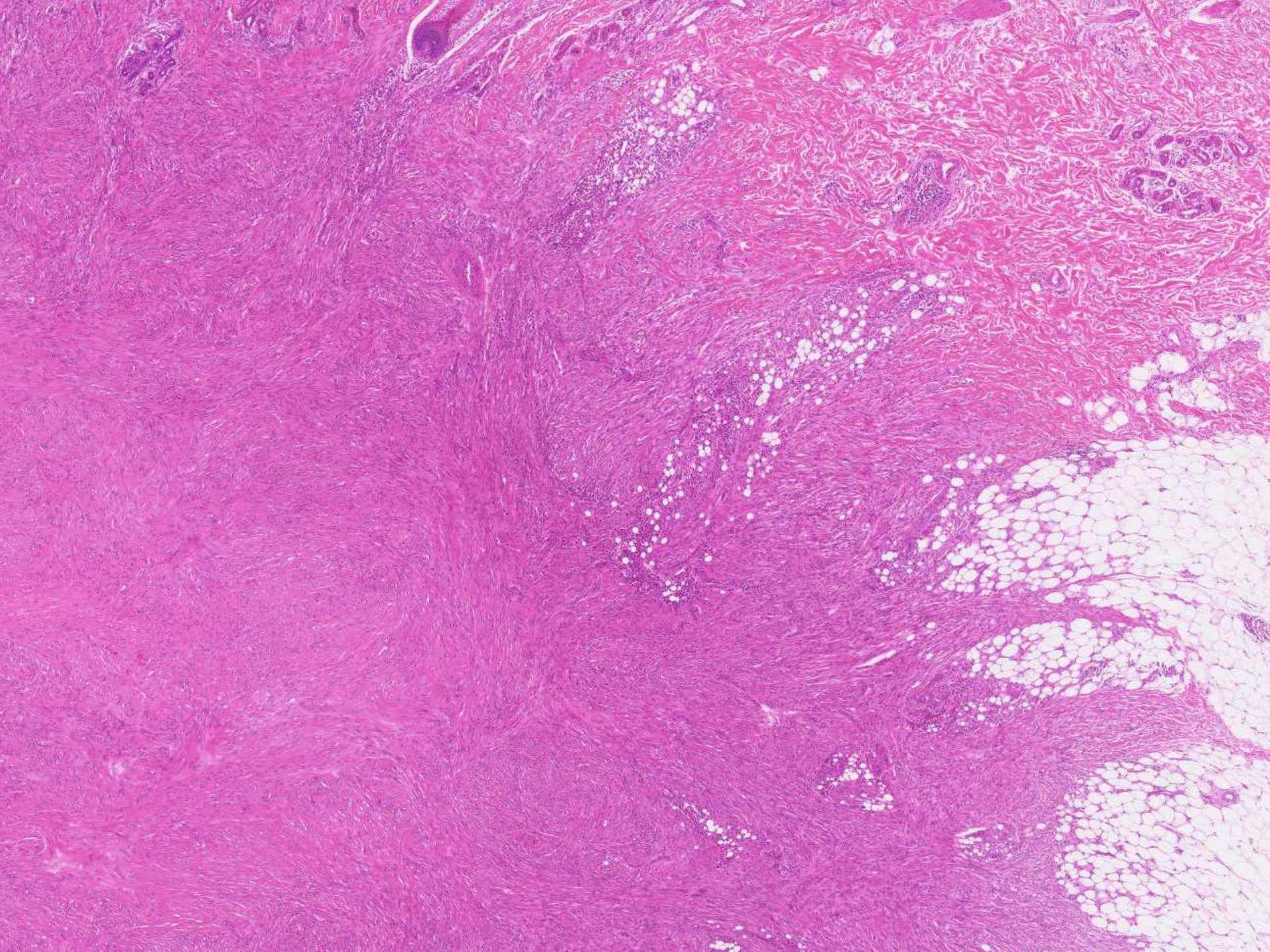
Ioannis Panagopoulos [a,b,*], Ludmila Gorunova [a,b], Bodil Bjerkehagen [c], Ingvild Lobmaier [c], Sverre Heim [a,b,d]

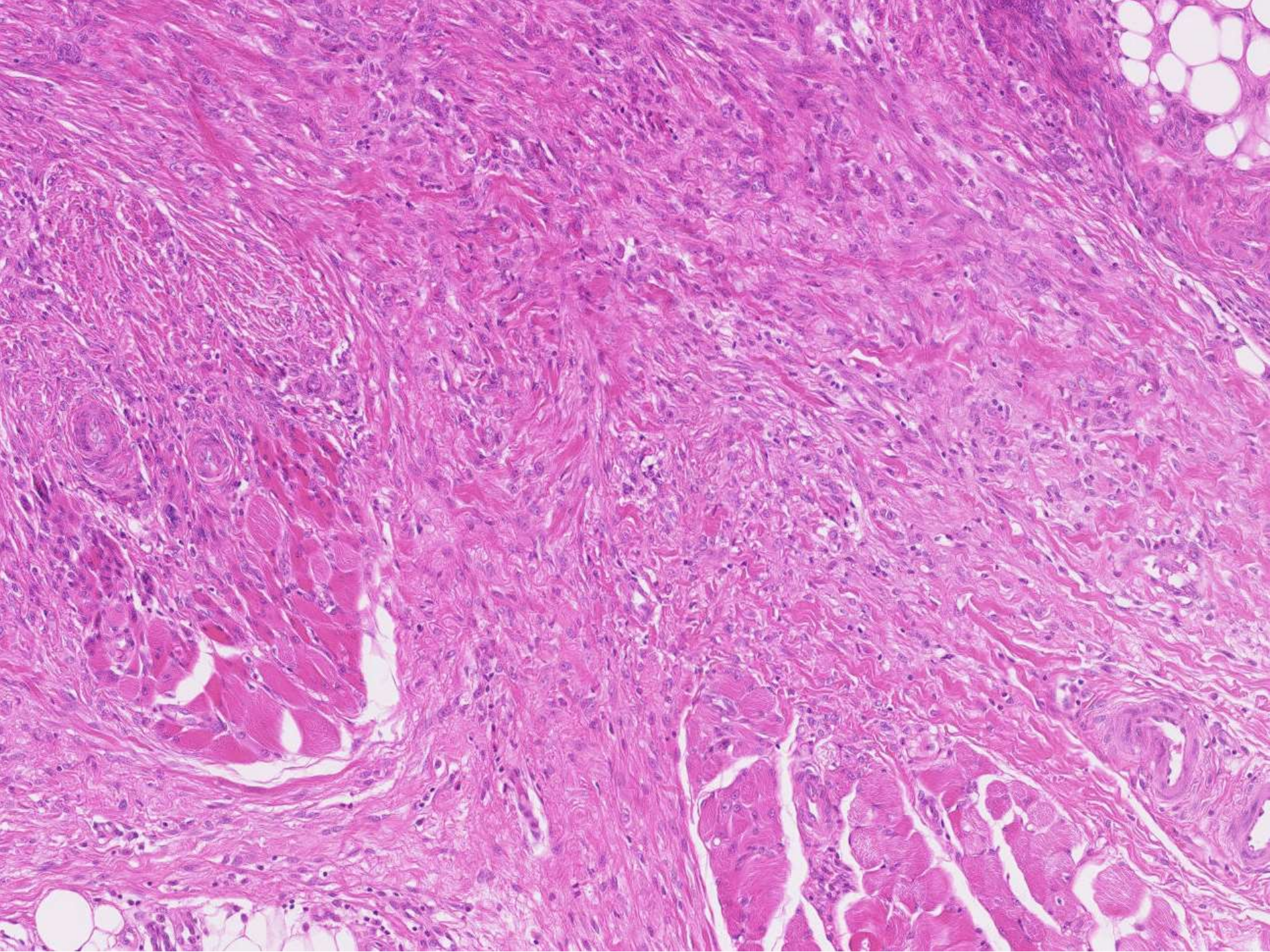
[a] *Section for Cancer Cytogenetics, Institute for Cancer Genetics and Informatics, The Norwegian Radium Hospital, Oslo University Hospital, Oslo, Norway;* [b] *Centre for Cancer Biomedicine, Faculty of Medicine, University of Oslo, Oslo, Norway;* [c] *Department of Pathology, The Norwegian Radium Hospital, Oslo University Hospital, Oslo, Norway;* [d] *Faculty of Medicine, University of Oslo, Oslo, Norway*

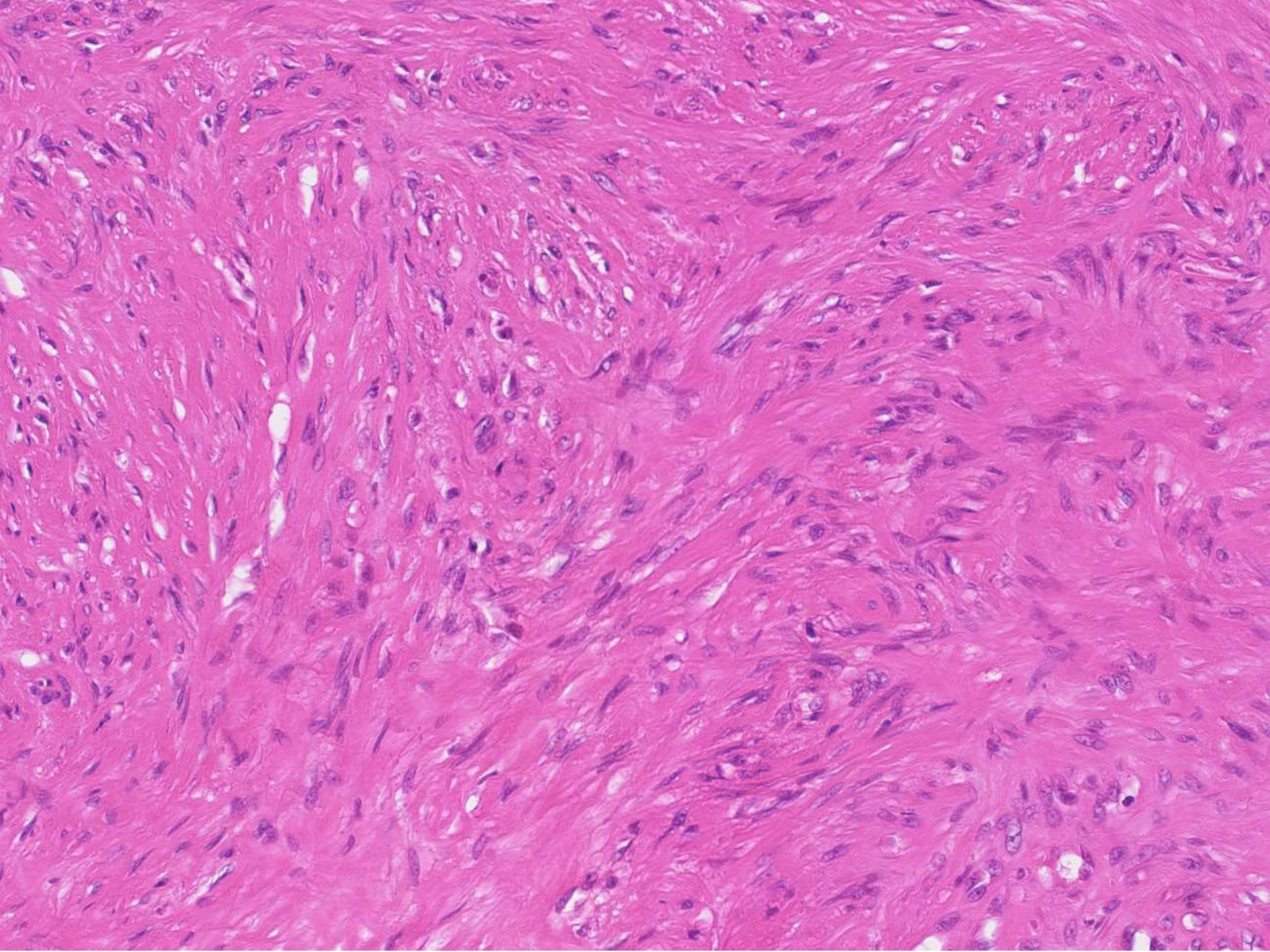


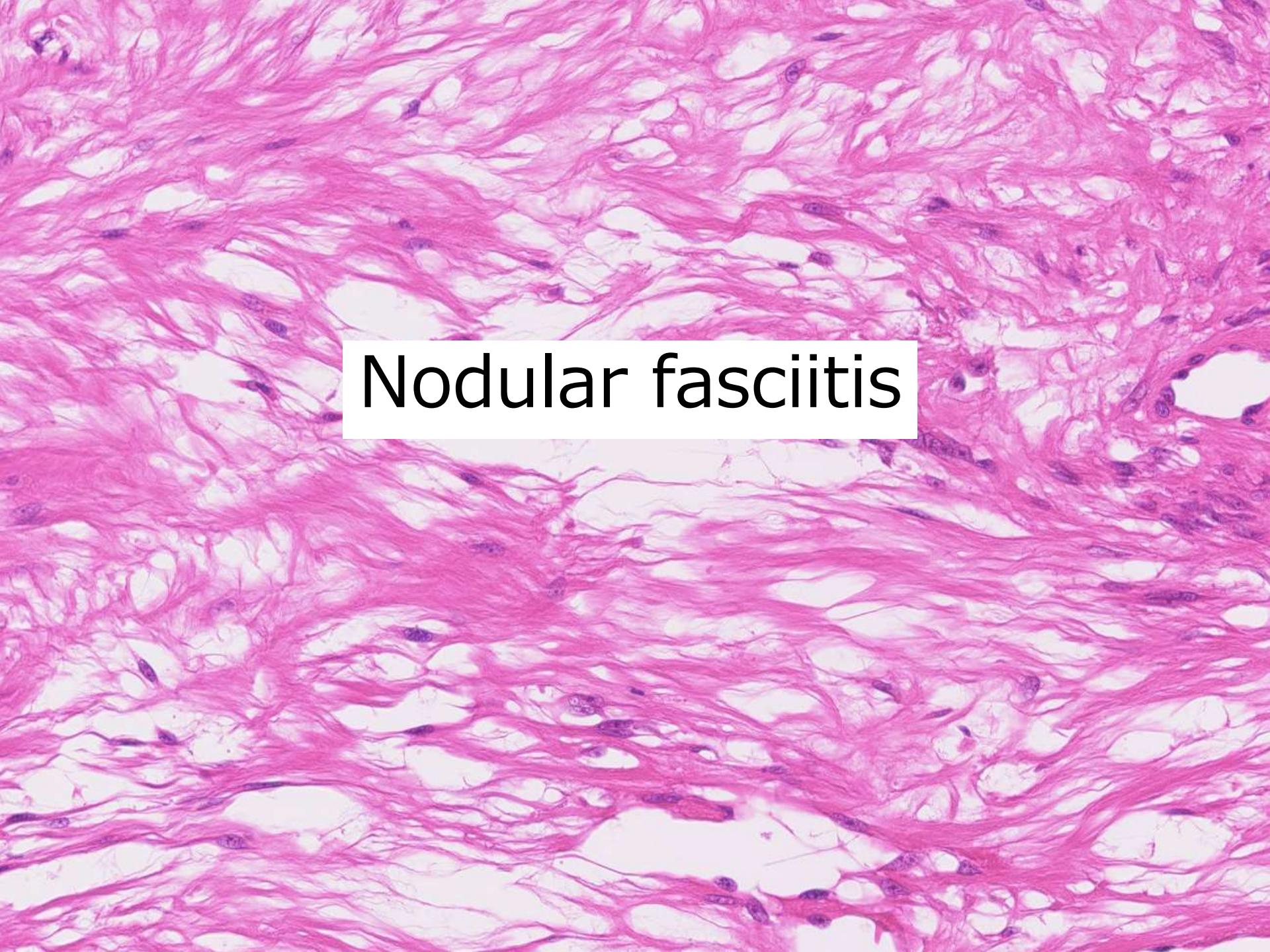
6M, face



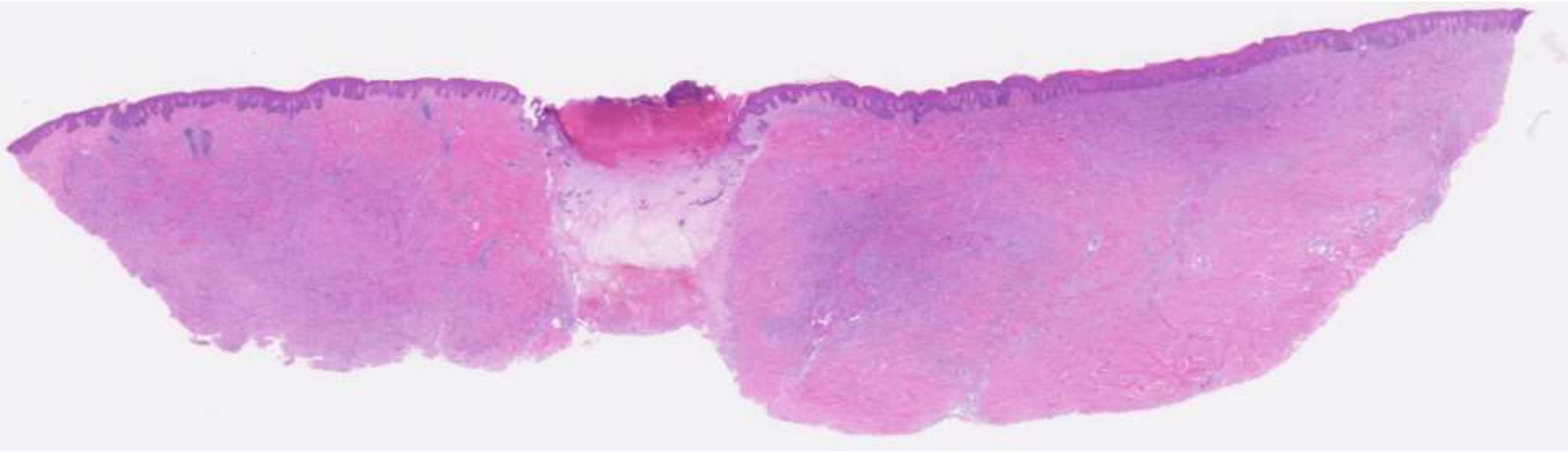


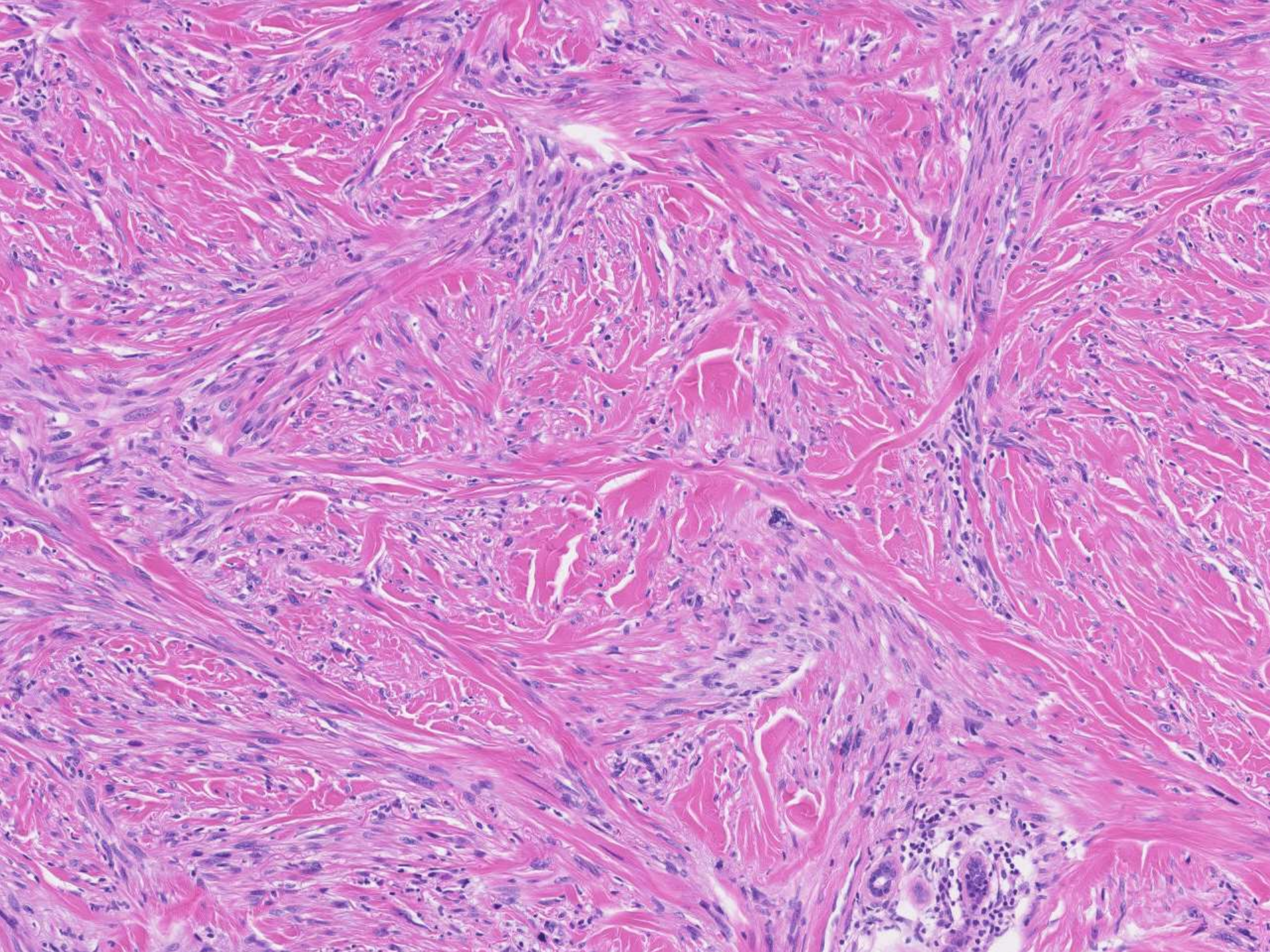


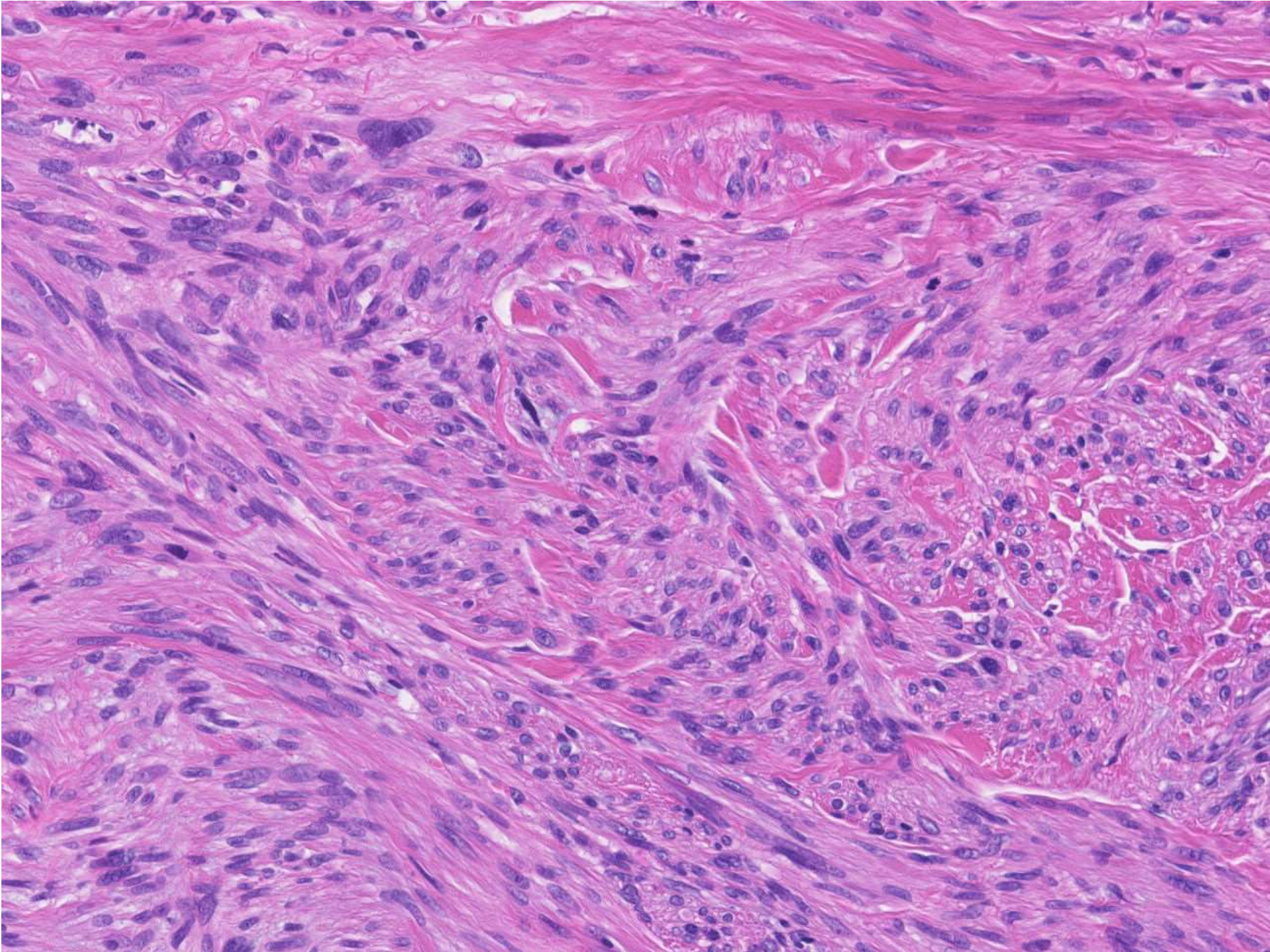


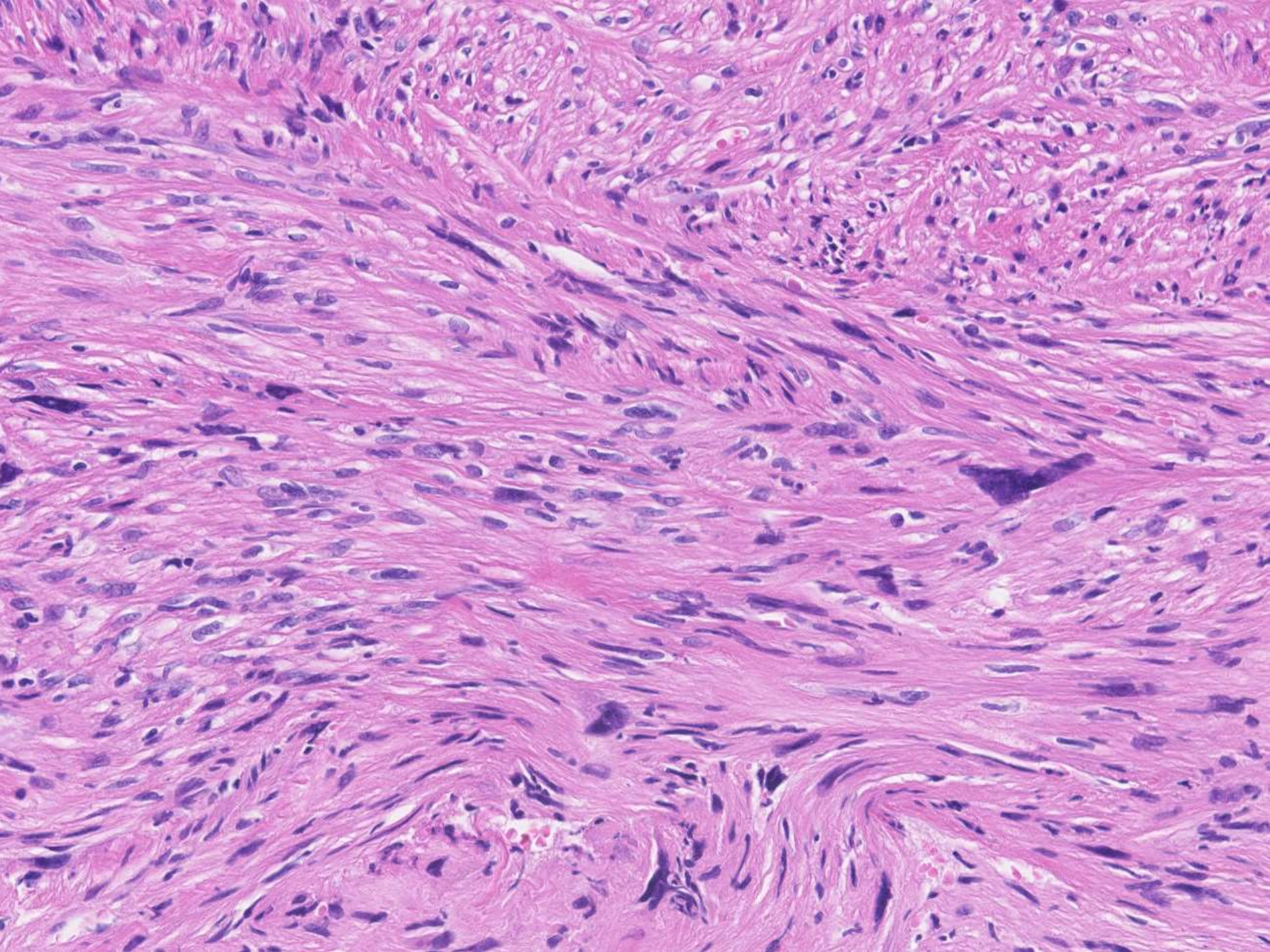

Nodular fasciitis

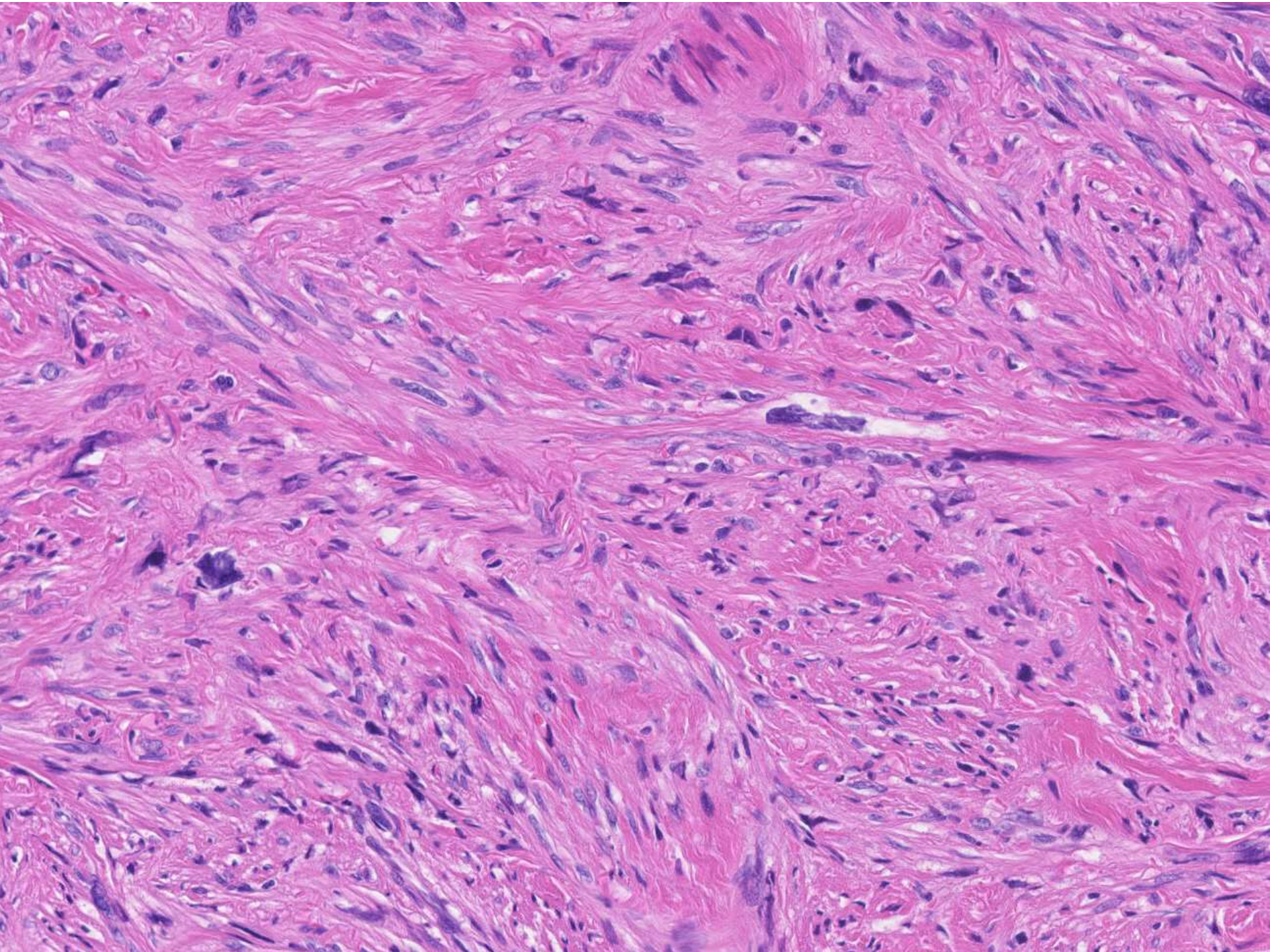
74M, back

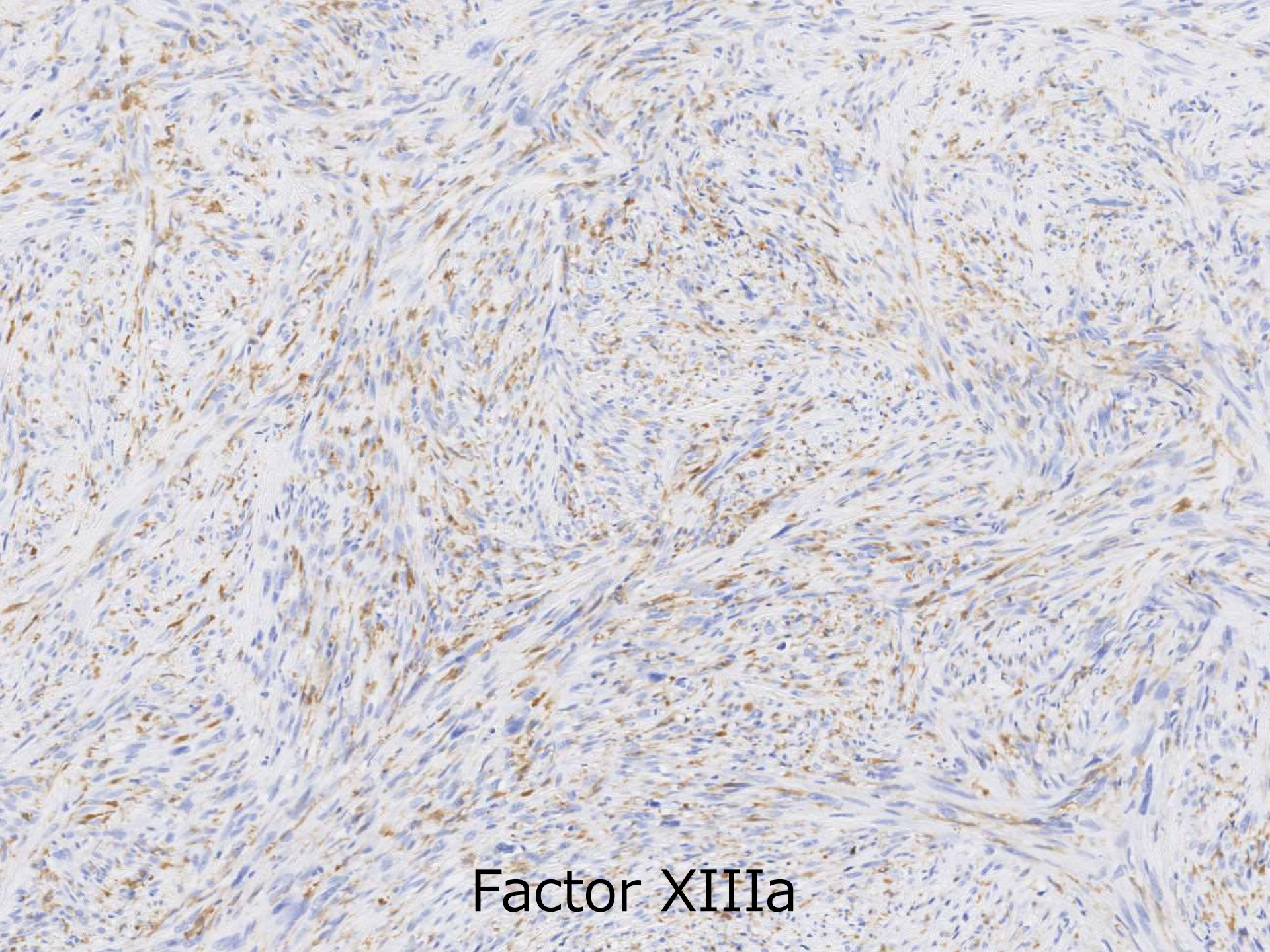


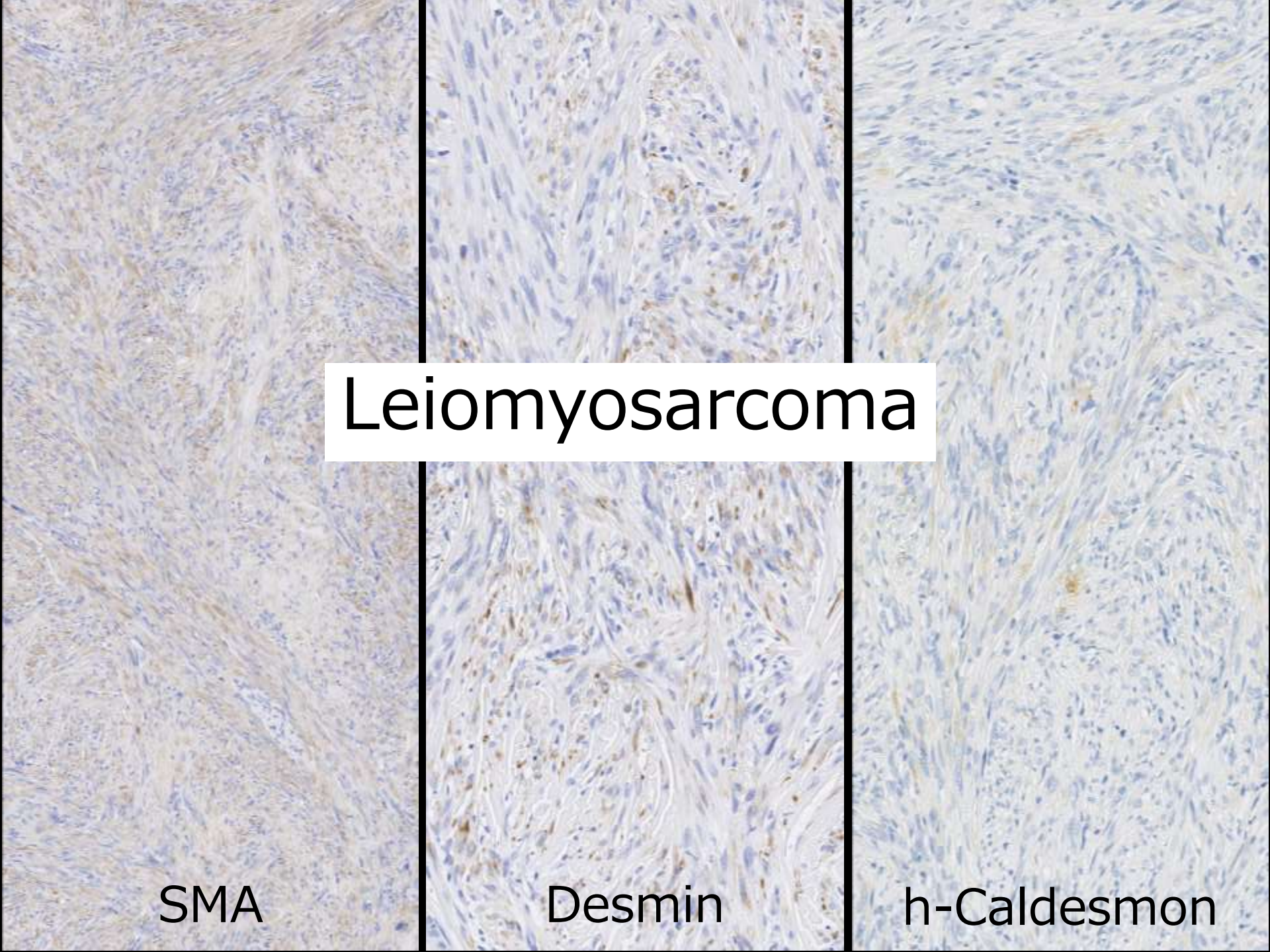




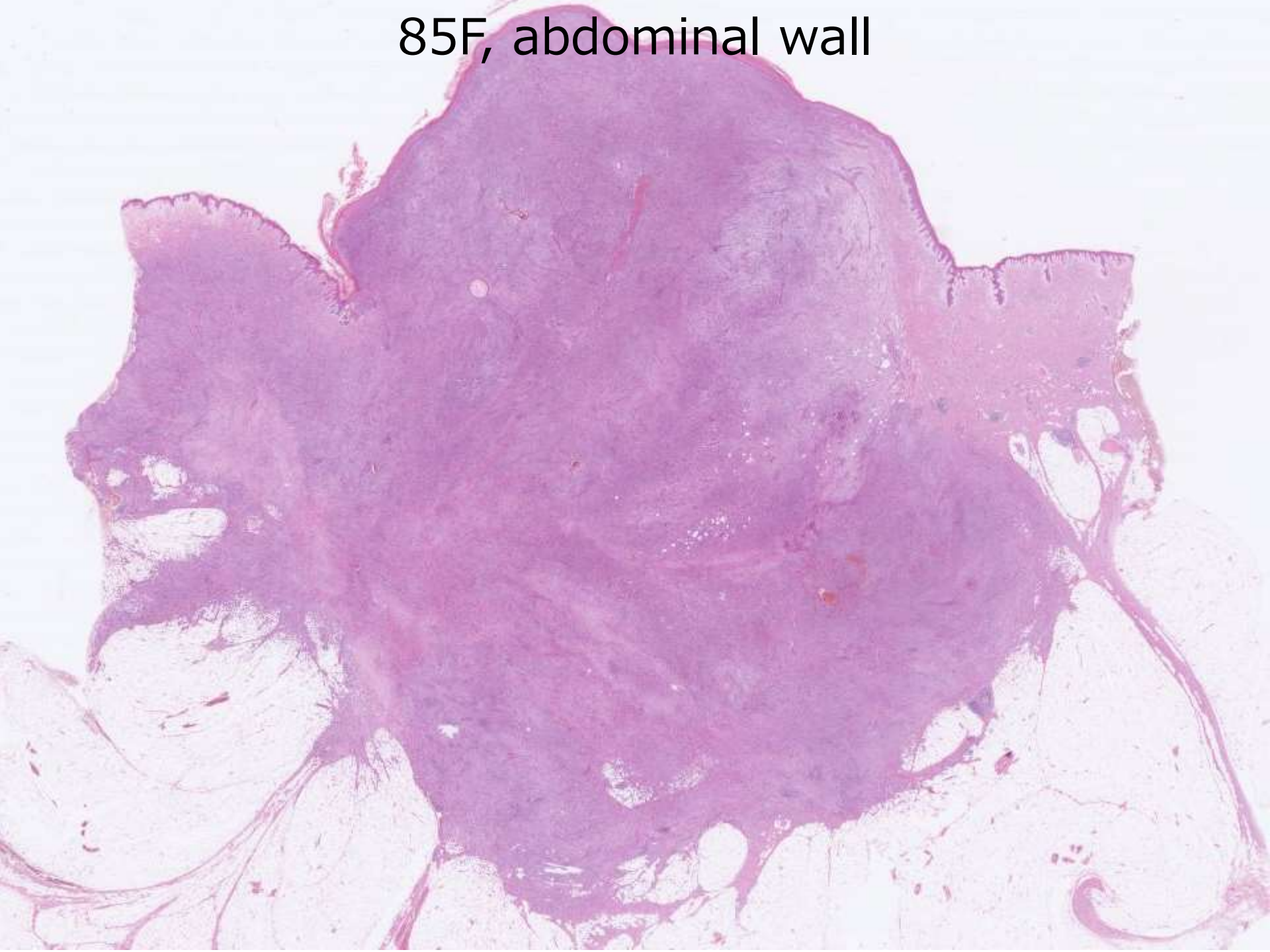


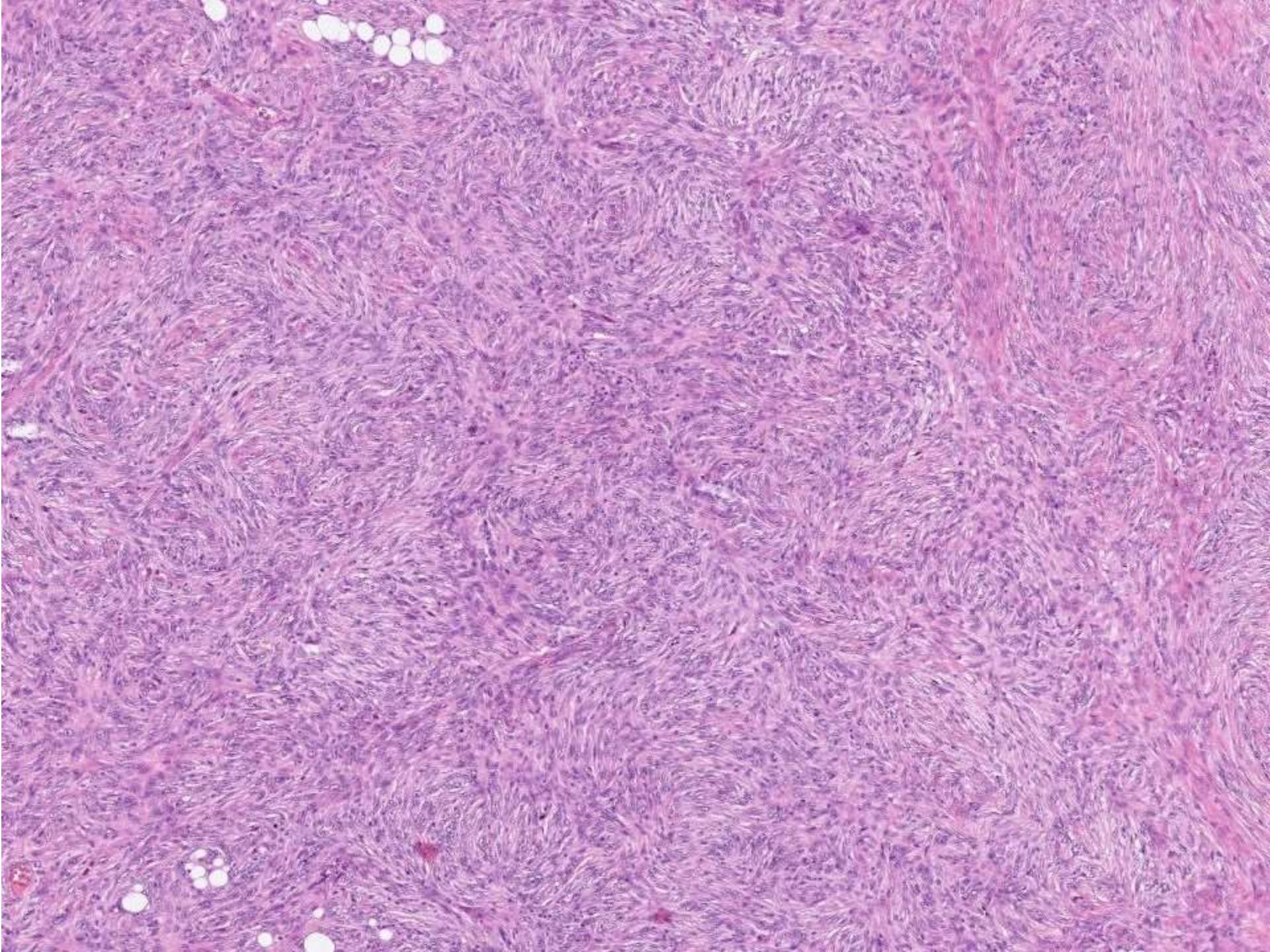


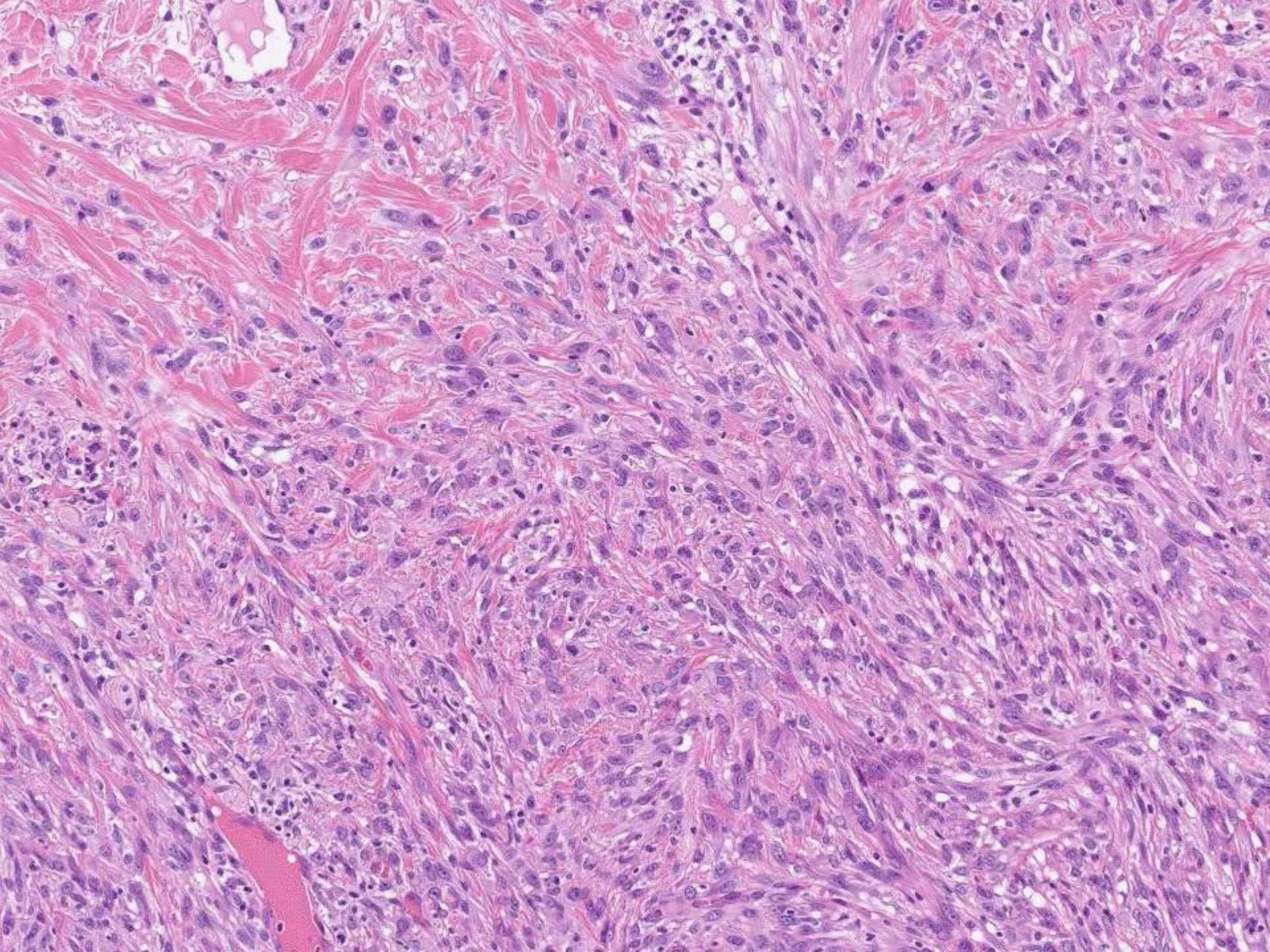

Factor XIIIa

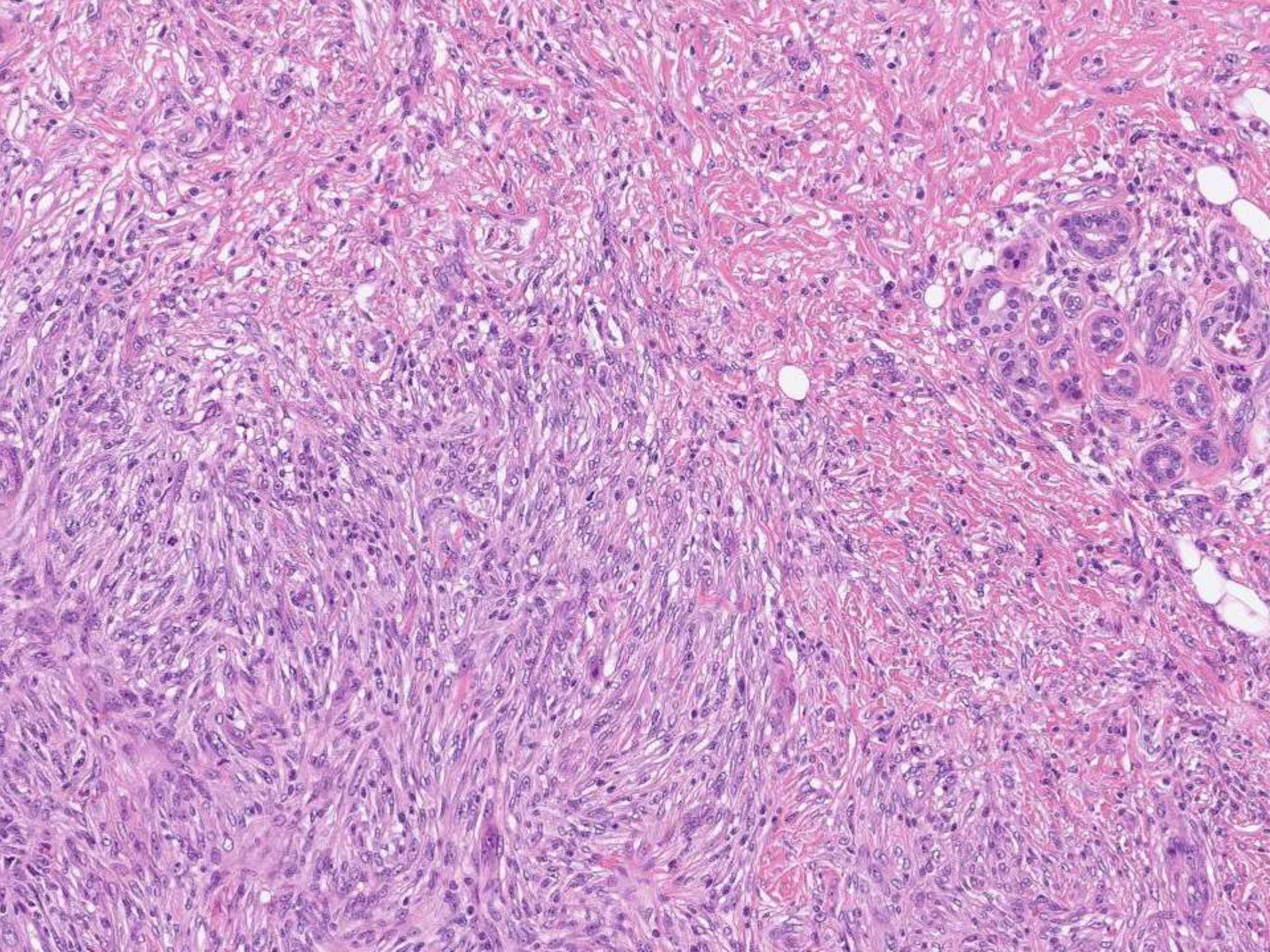

Leiomyosarcoma
SMA
Desmin
h-Caldesmon

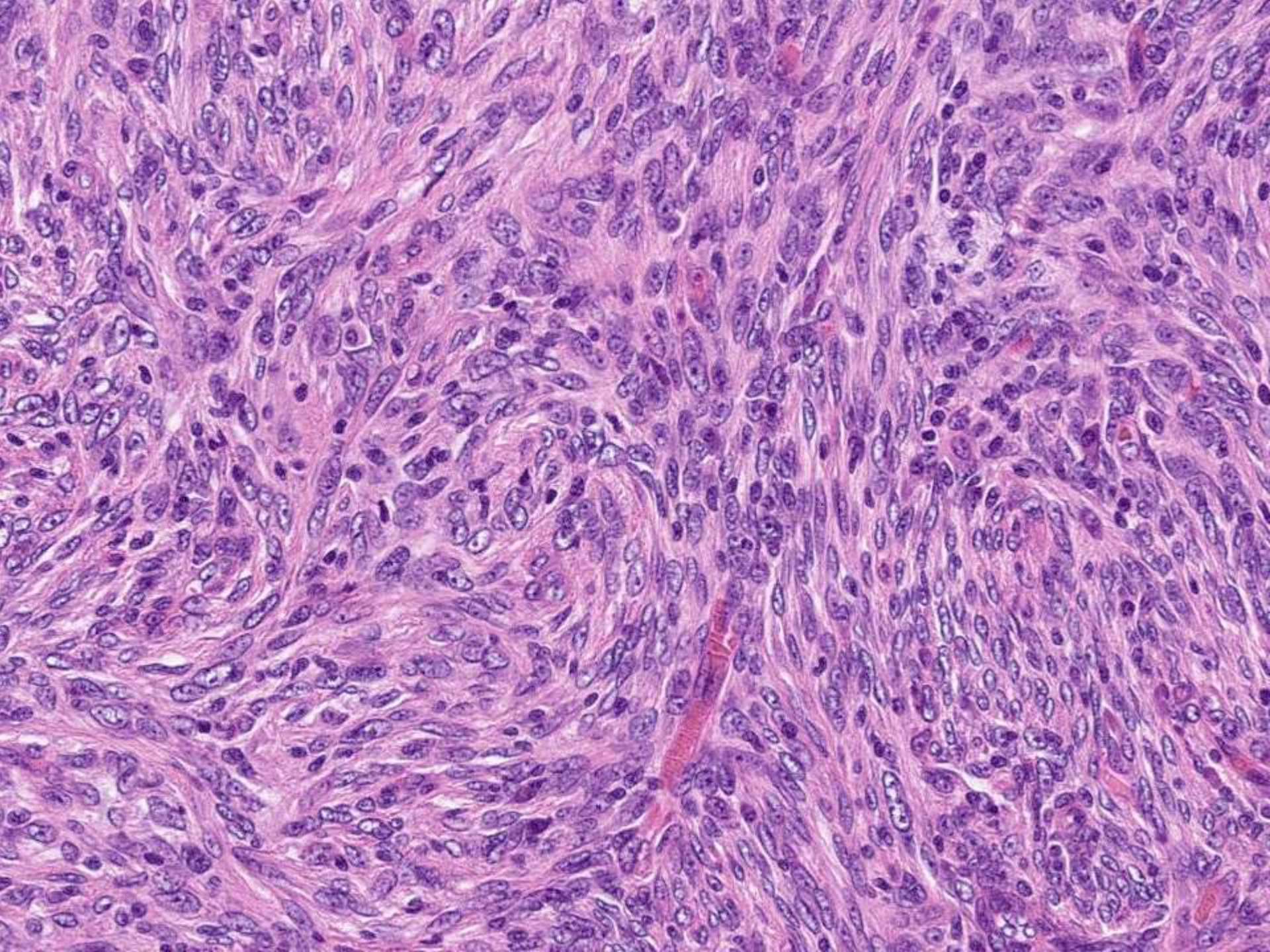
85F, abdominal wall

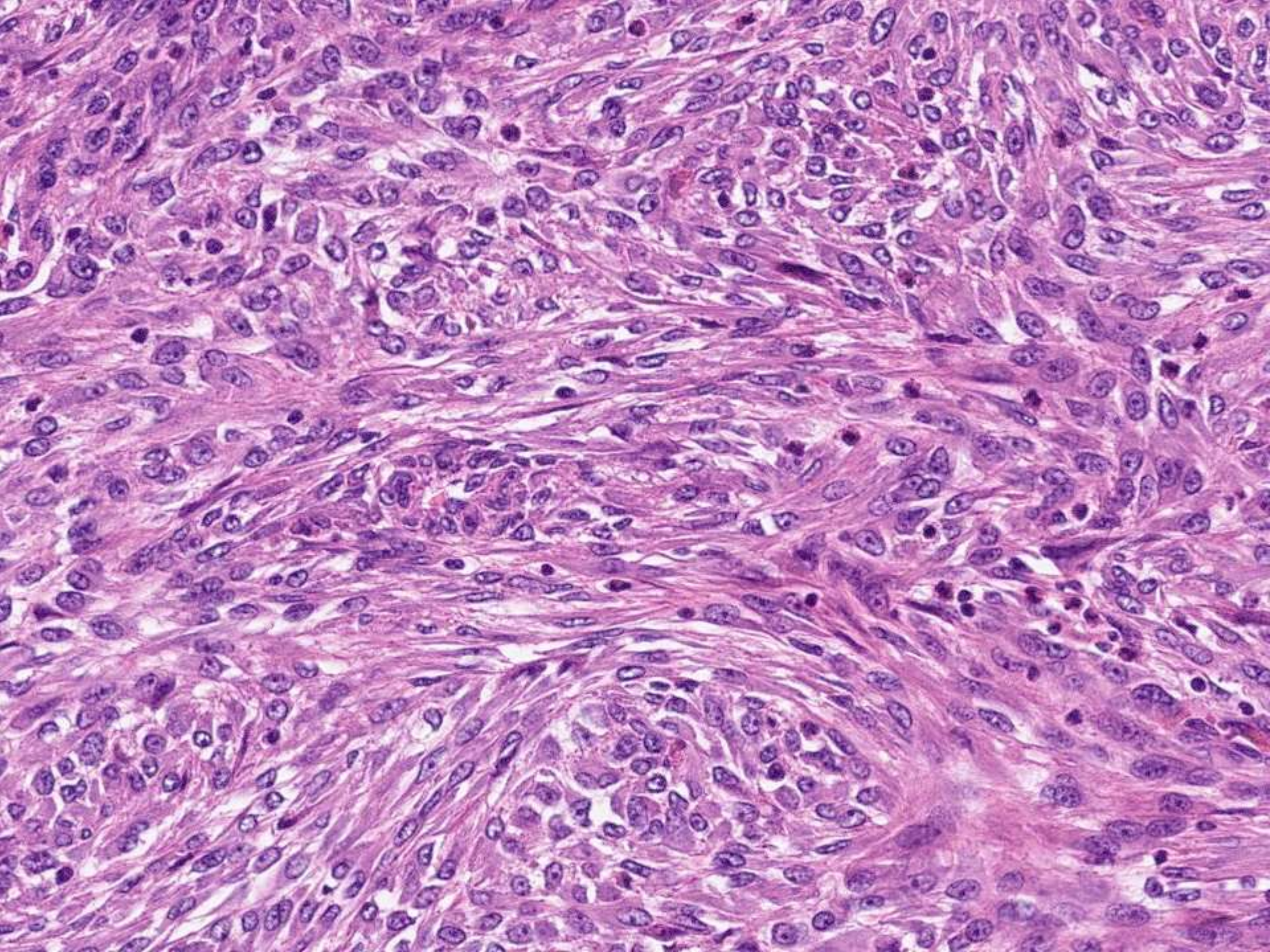


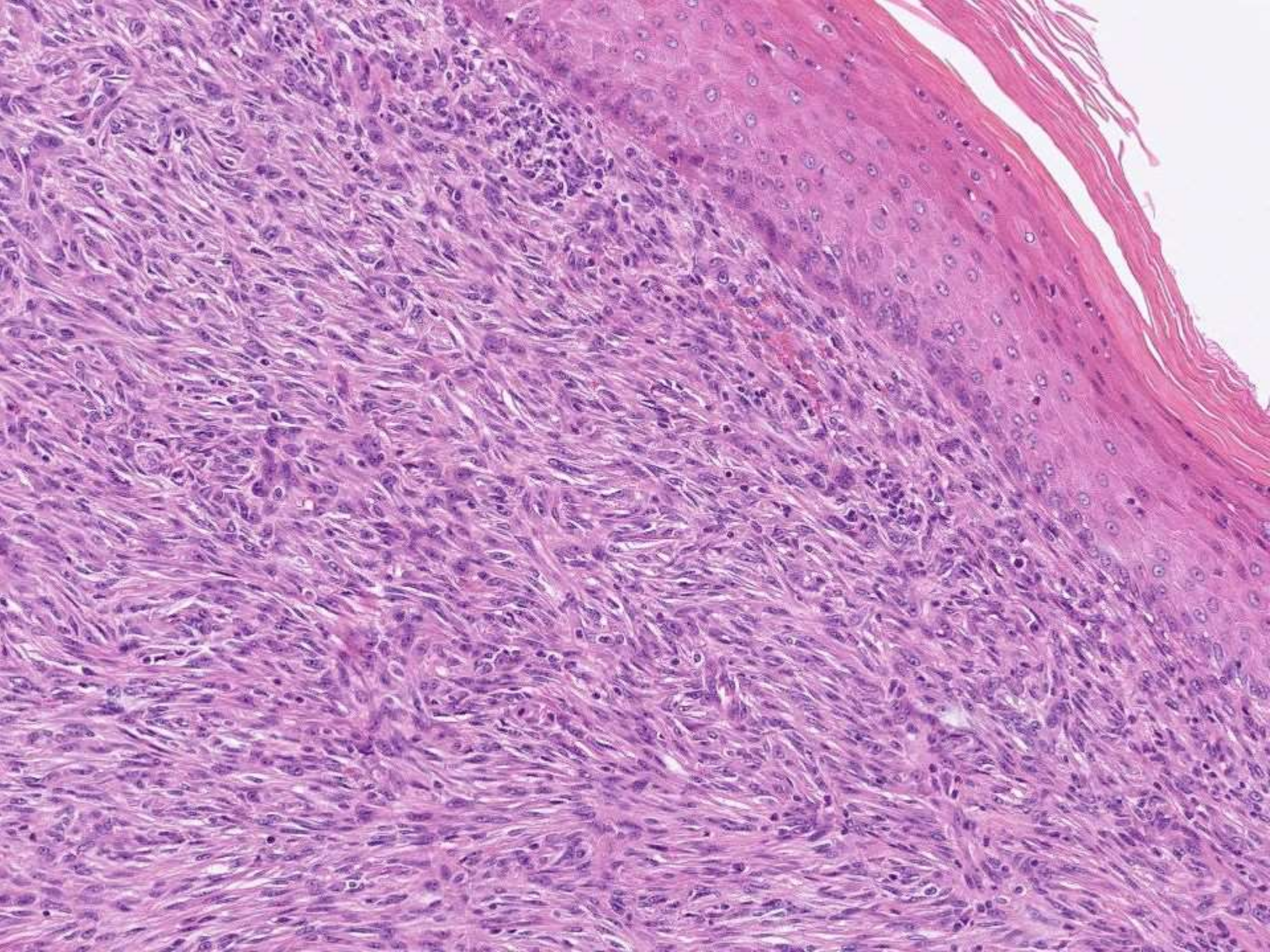


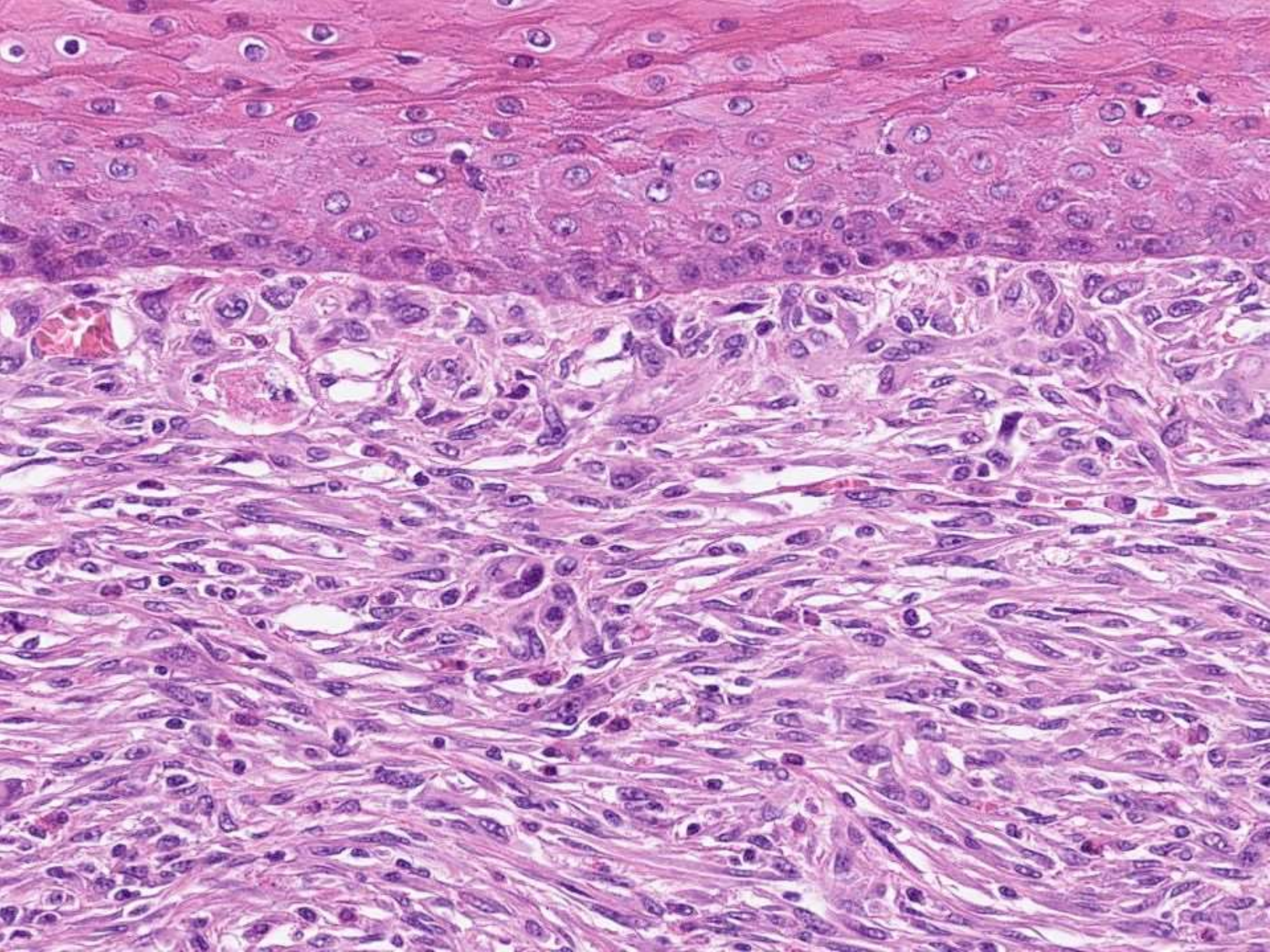


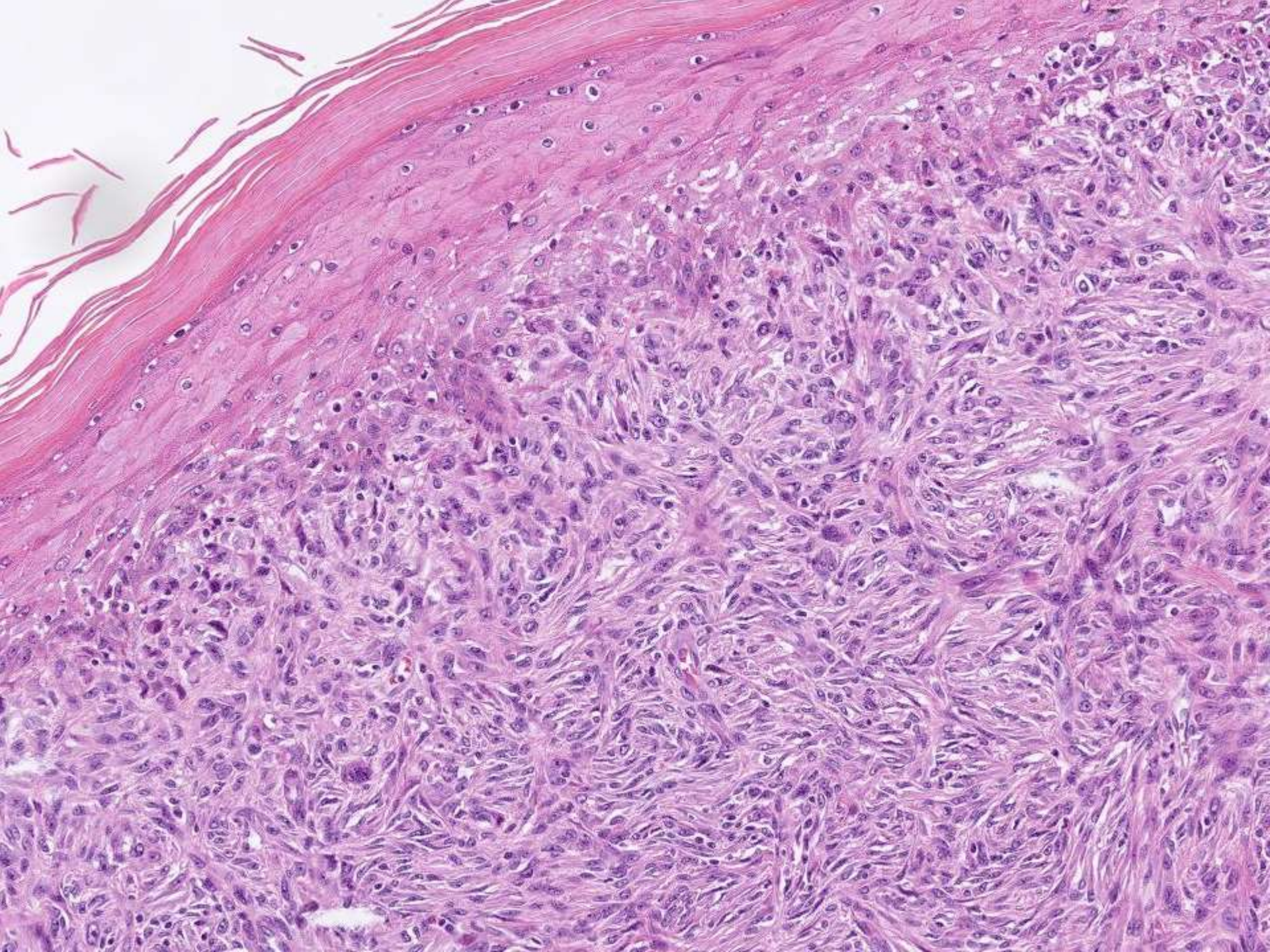


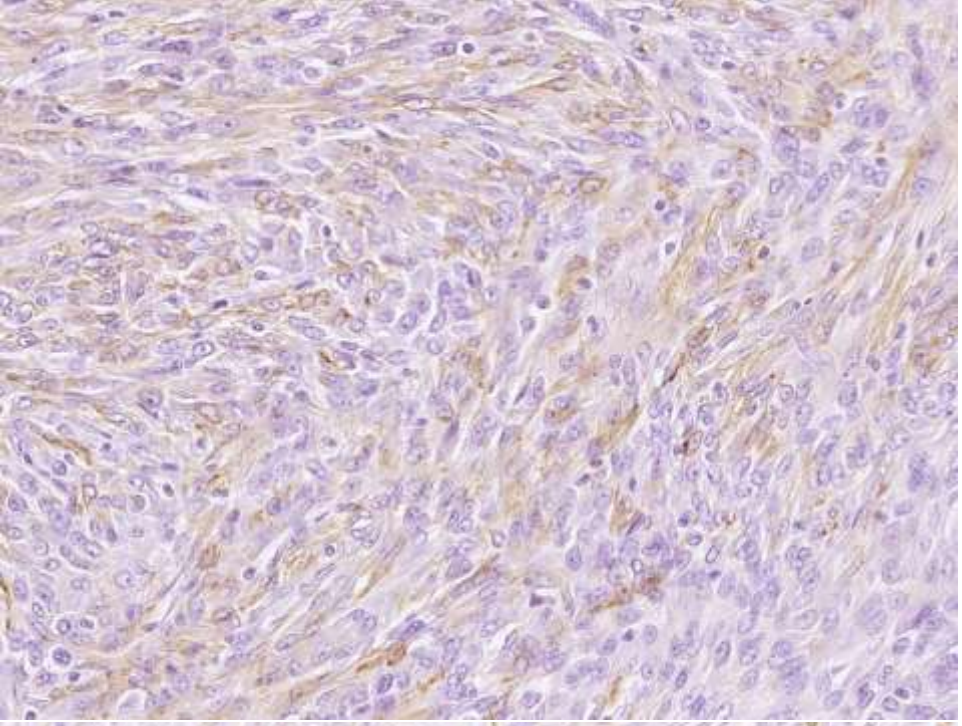


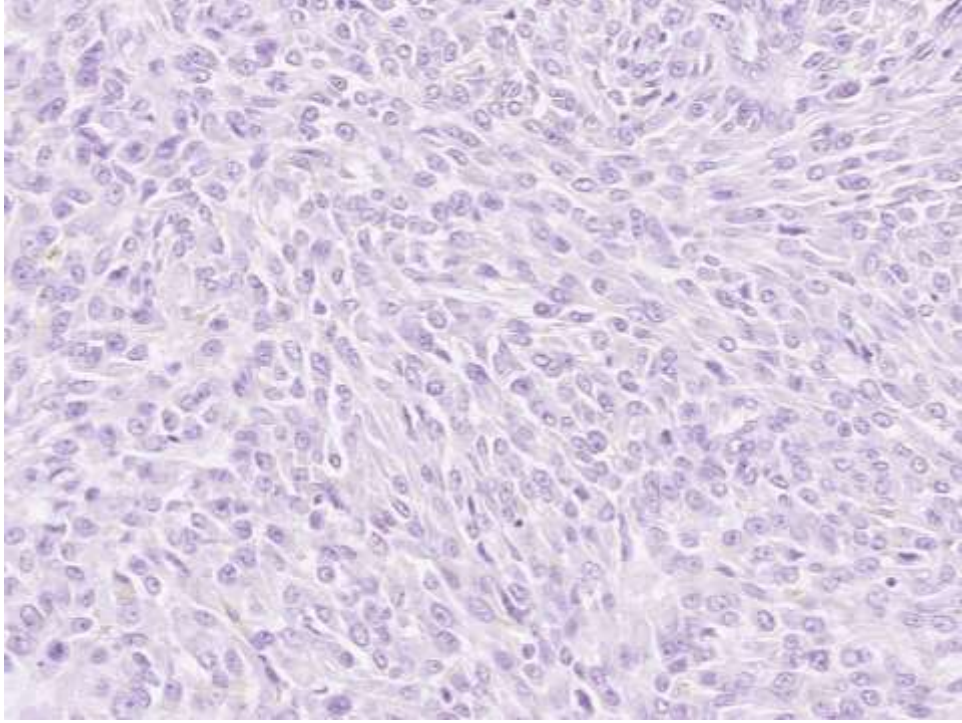


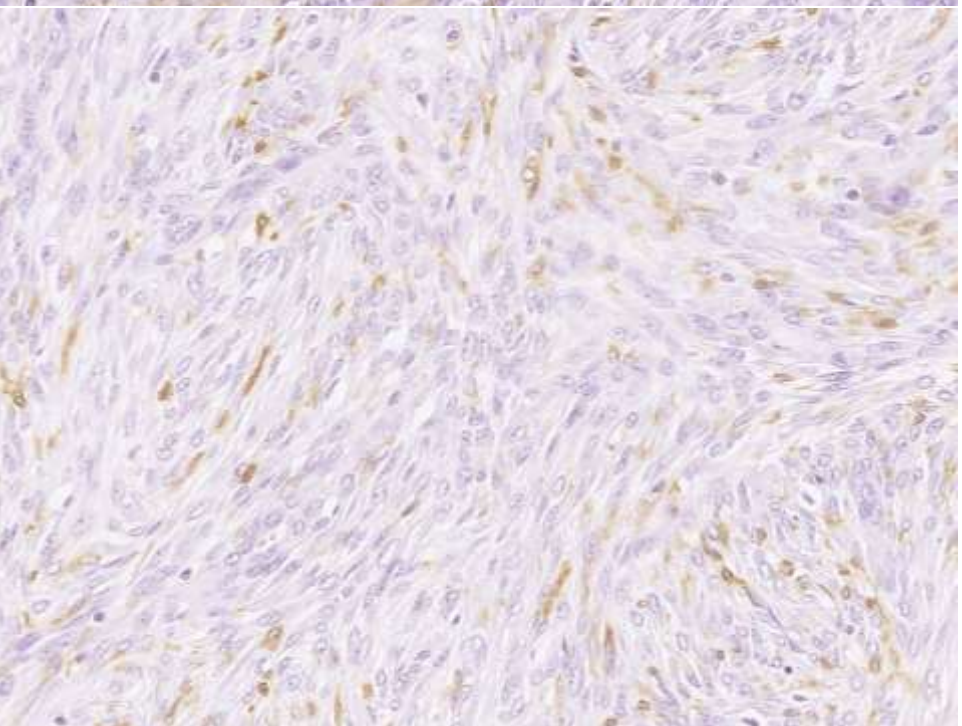






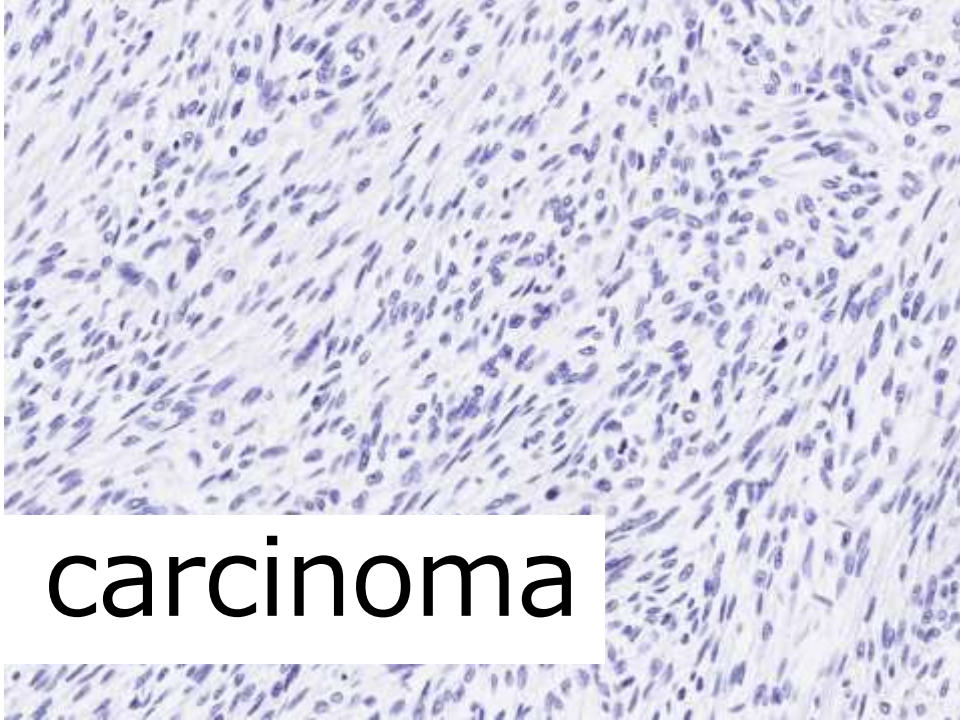


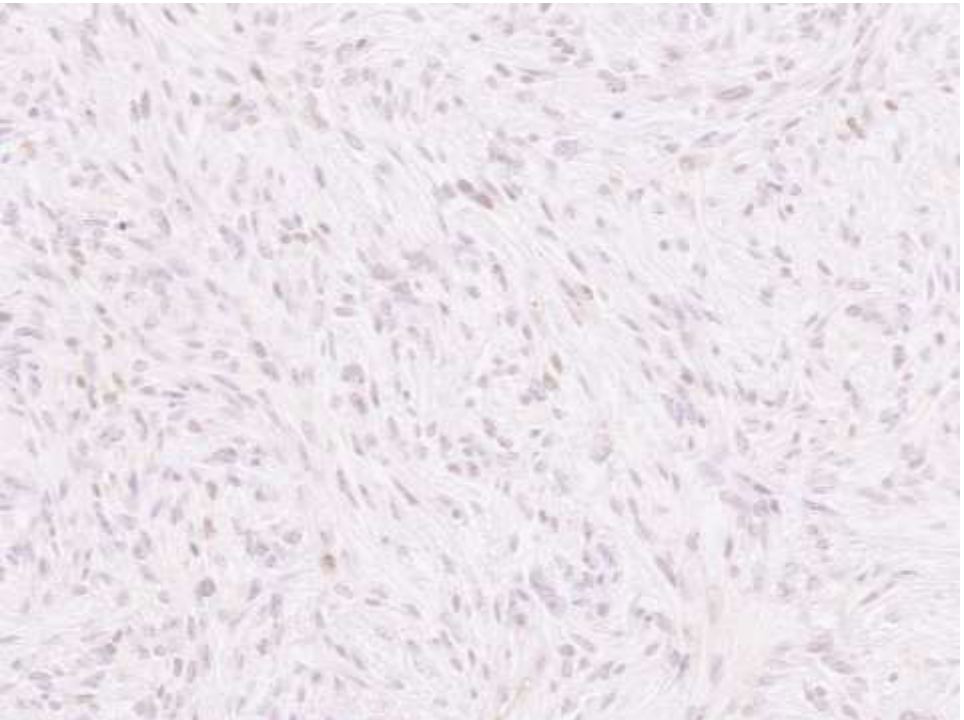




SMA	Desmin
Factor XIIIa	

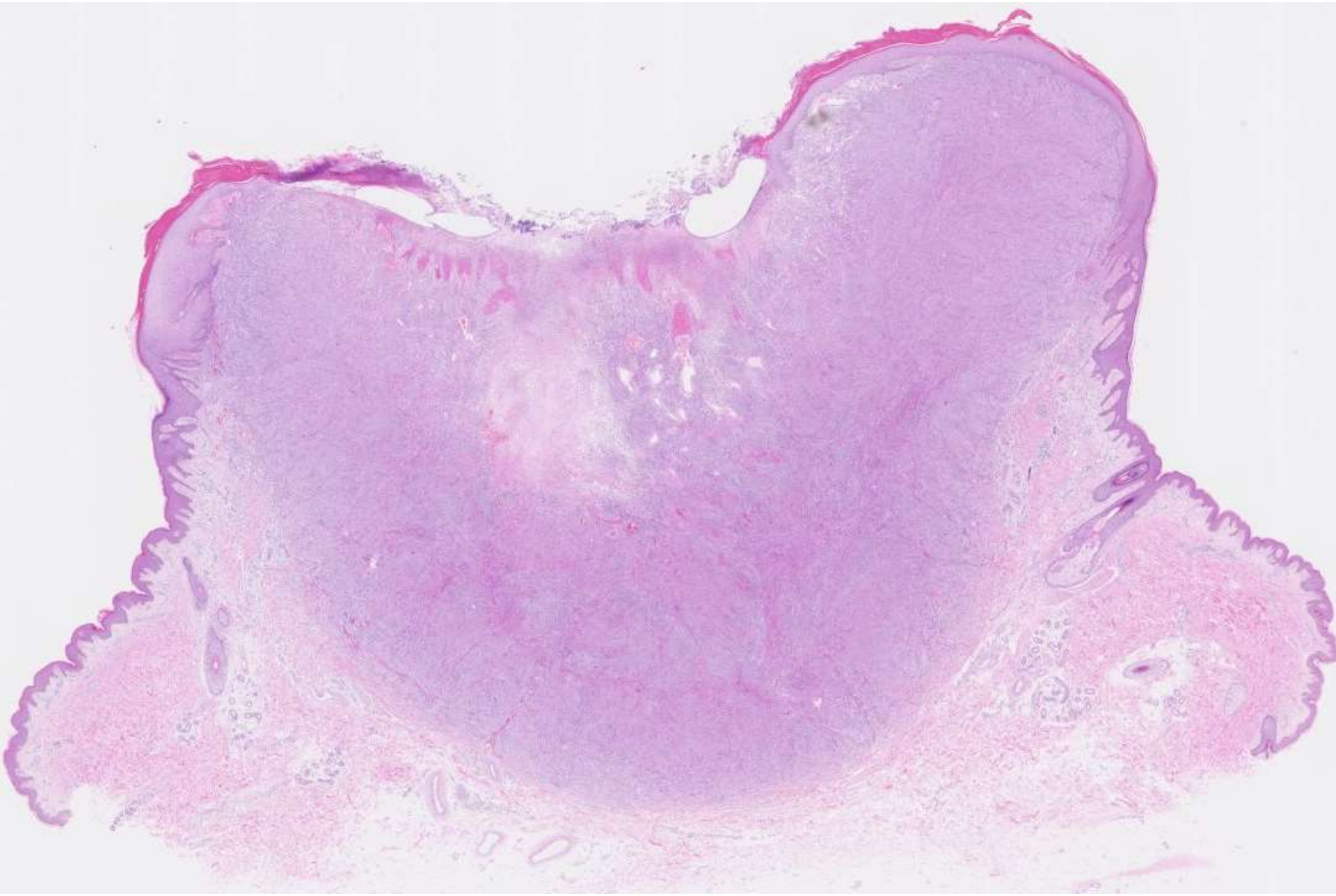
Spindle cell carcinoma





AE1/AE3	CAM5.2
p63	

27M, thigh



Tips 6

- Dermatofibromaはシルエットでほとんど診断可能(DFSPとの鑑別はあまり問題にならない)
- ただし、特異的な所見ではないため、非定型的な例では分化を確認し除外診断が必要
- 亜型を認識する

まとめ

- 紡錘形細胞腫瘍は恐れる必要はない
- 大まかな分化の方向を確認すれば、日常診療で遭遇する例のほとんどは形態診断が可能
- 免役染色の軽度の発現は重視しなくてよい場合もある
- 免役染色の発現をよく見て判断しなければならない場合もある
- 免役染色をやり過ぎない