第222回岡山外科病理研究会

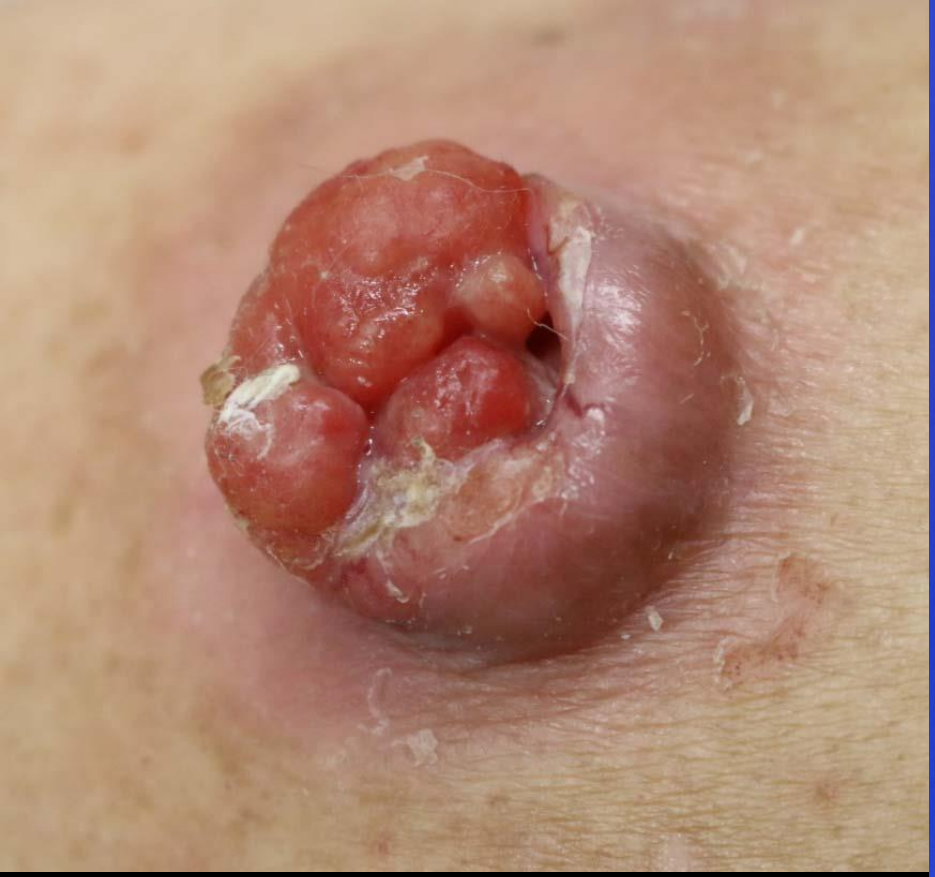
演題番号: 1424

症　　例: 80代後半、女性

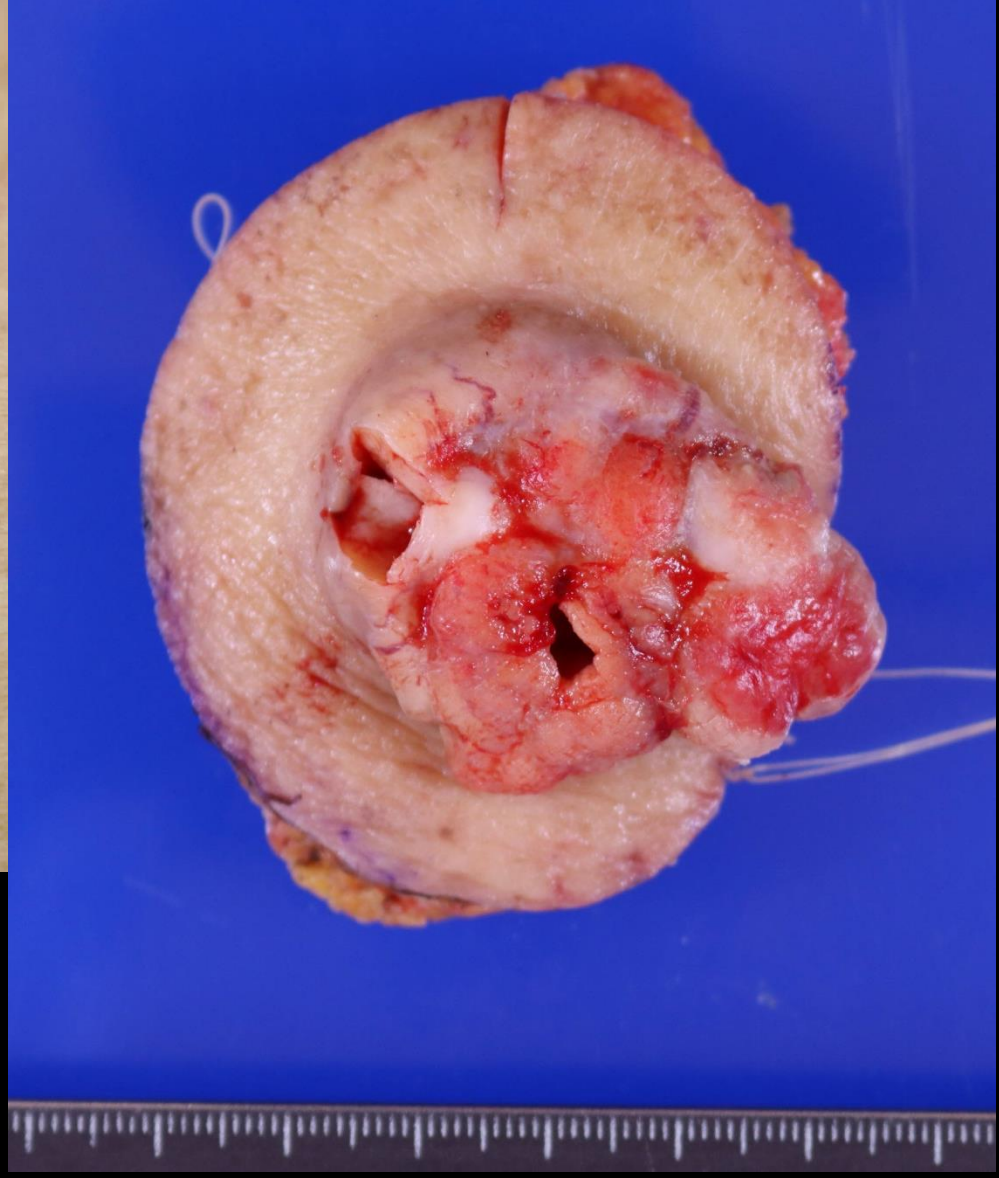
臨床診断: 右肩軟部腫瘍

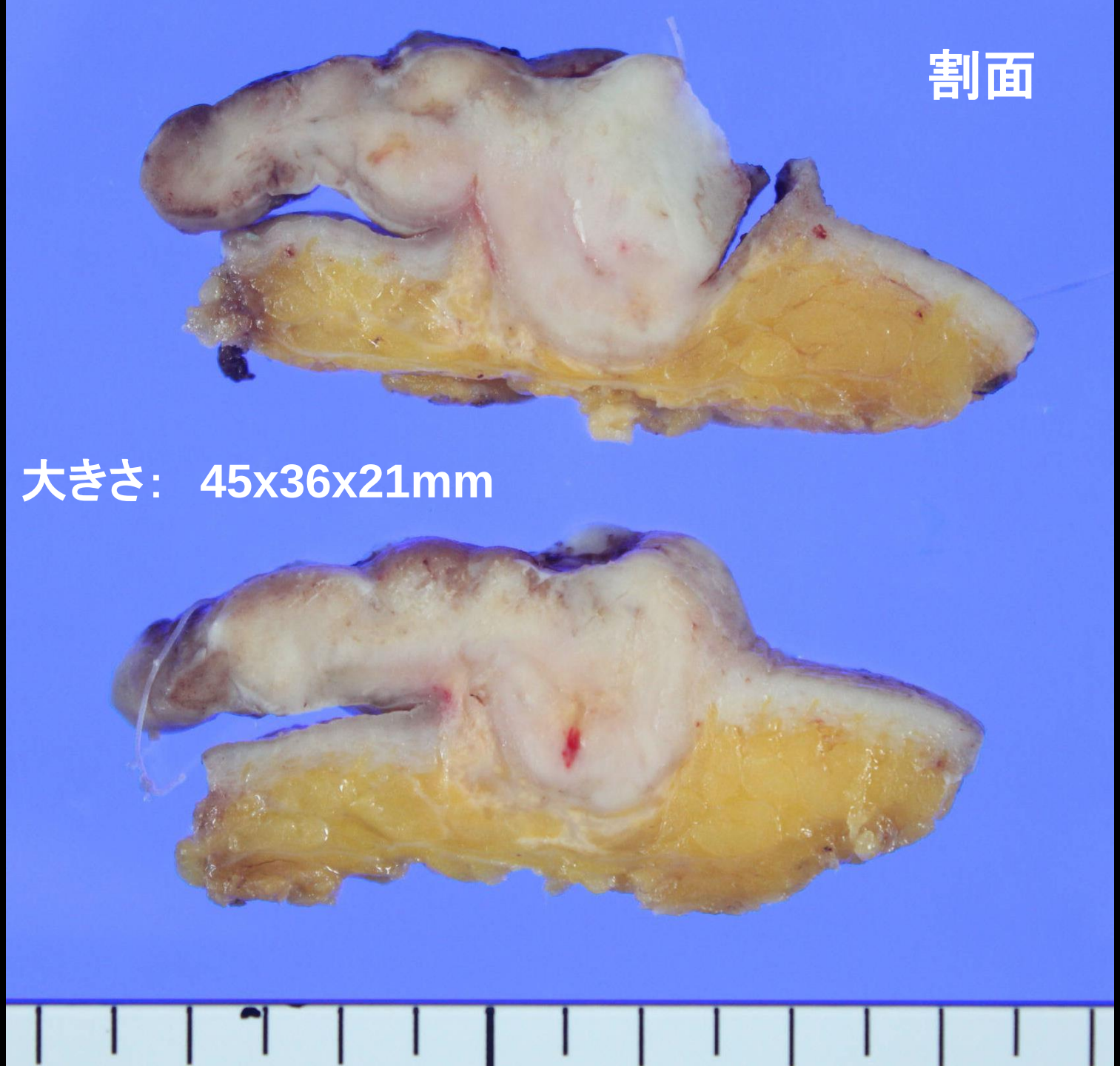
摘出部位: 右肩皮膚・軟部

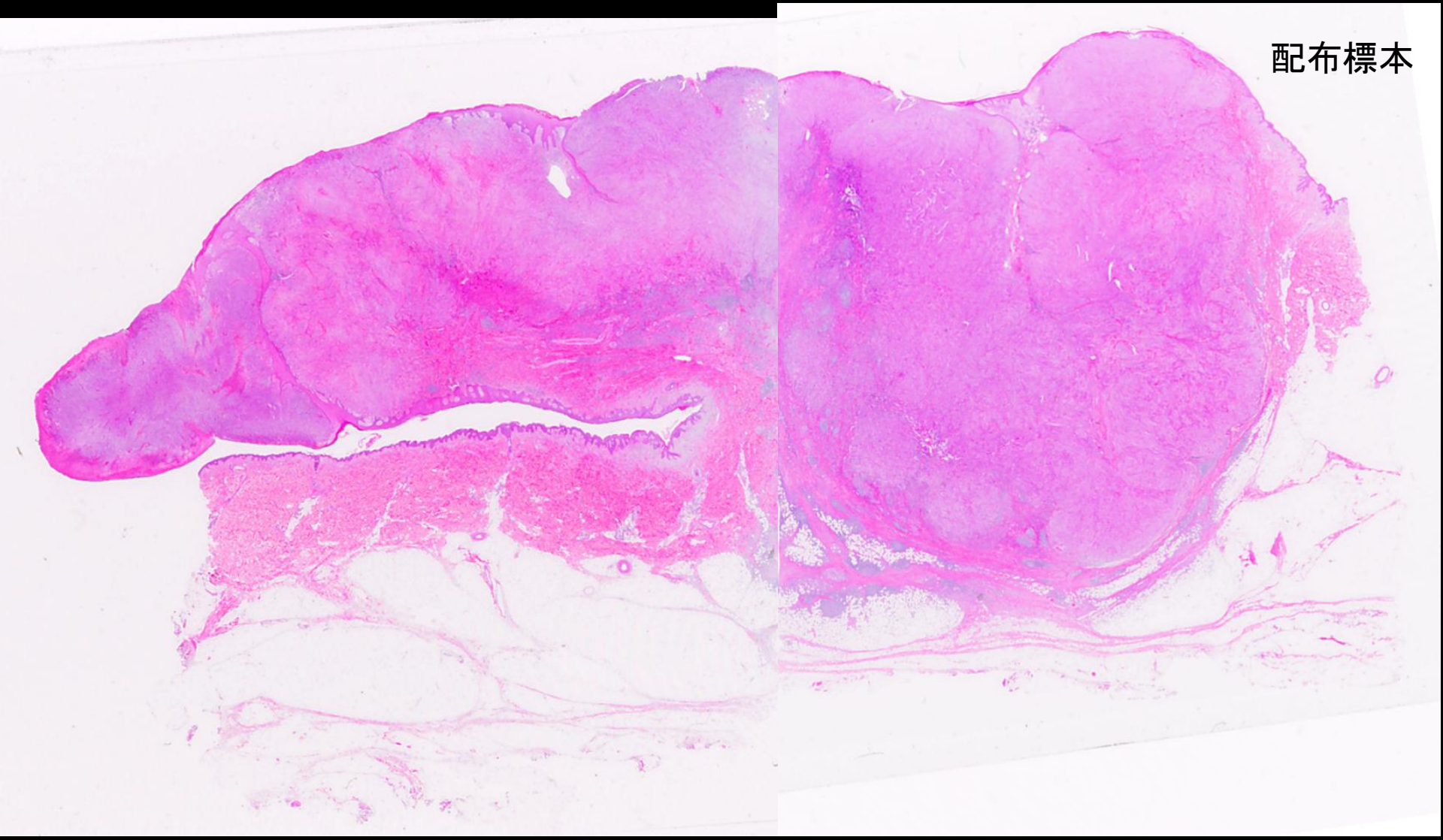
川崎医科大学総合医療センター
病理科　物部泰昌



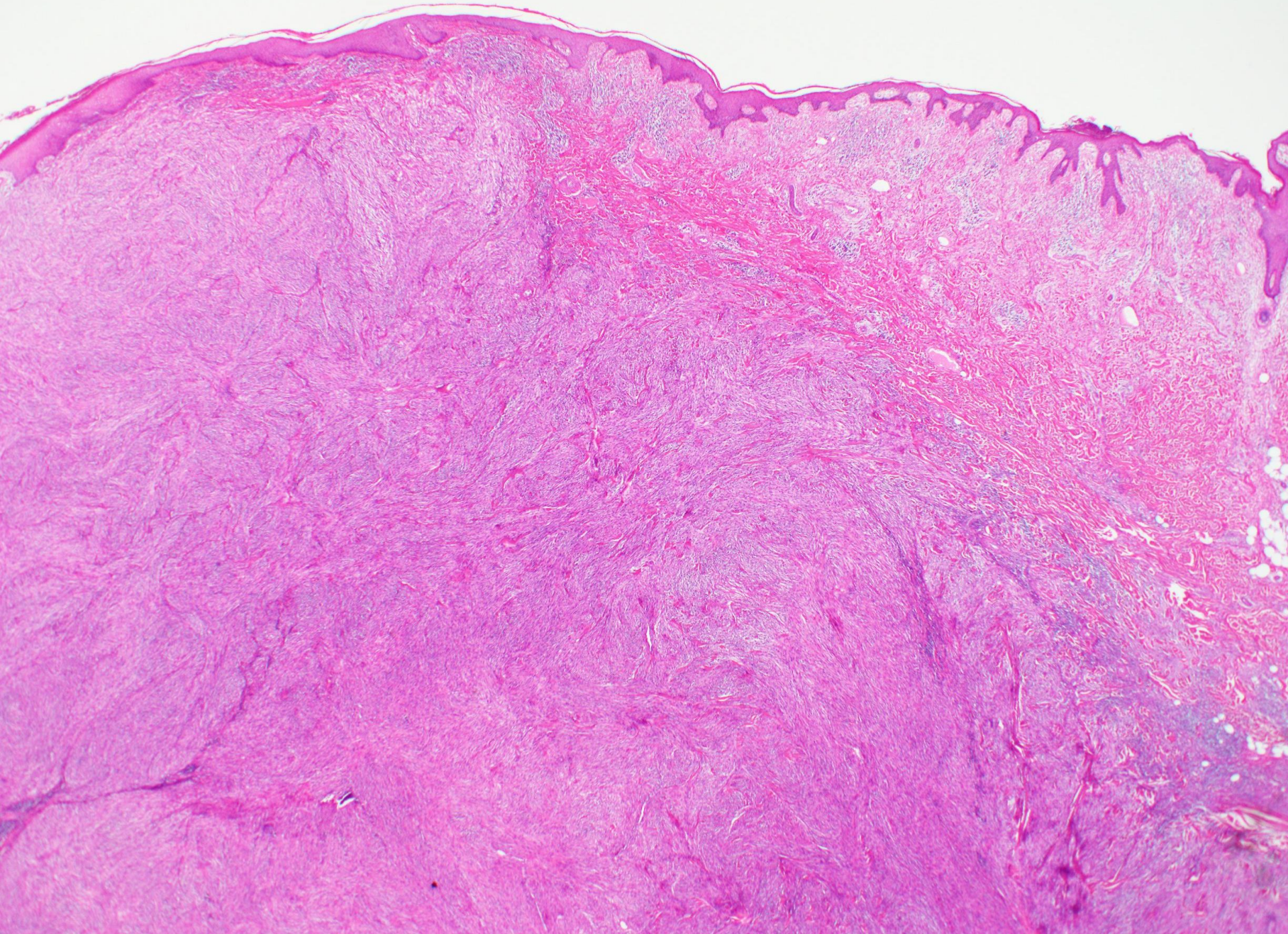
潰瘍を伴う隆起性腫瘍

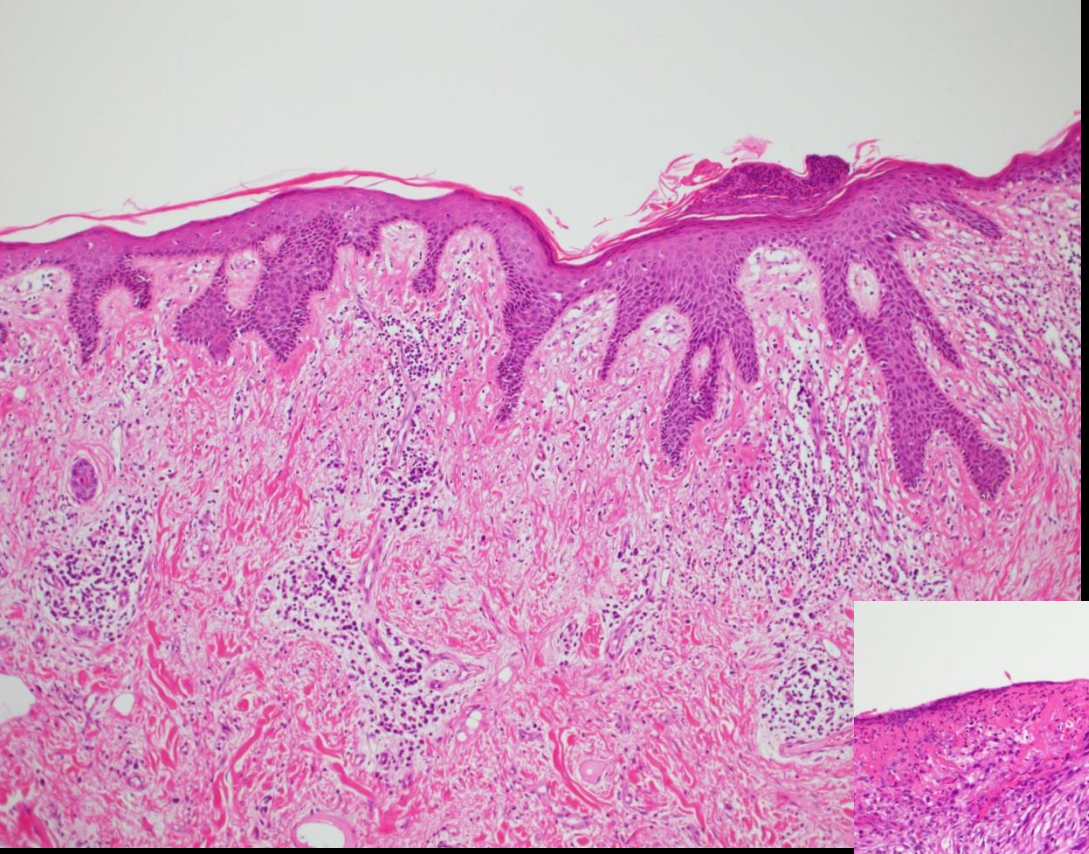



割面
大きさ: 45x36x21mm

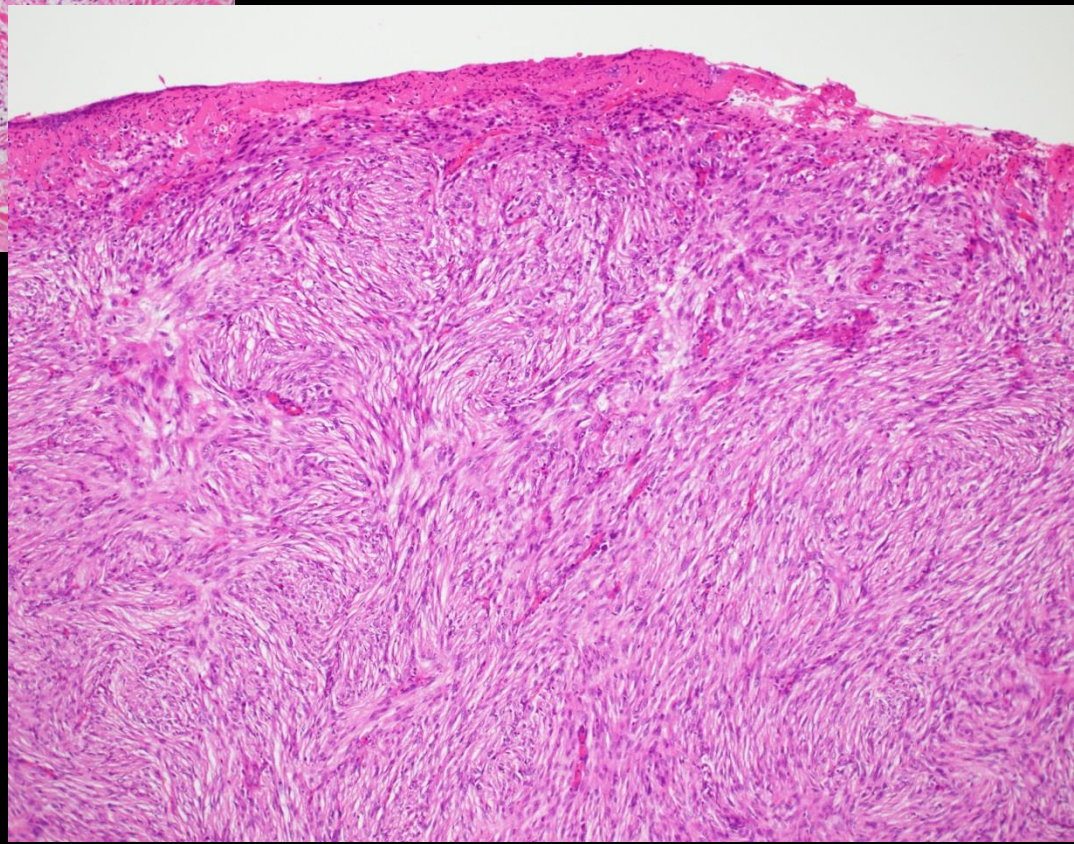


病変の主体は真皮

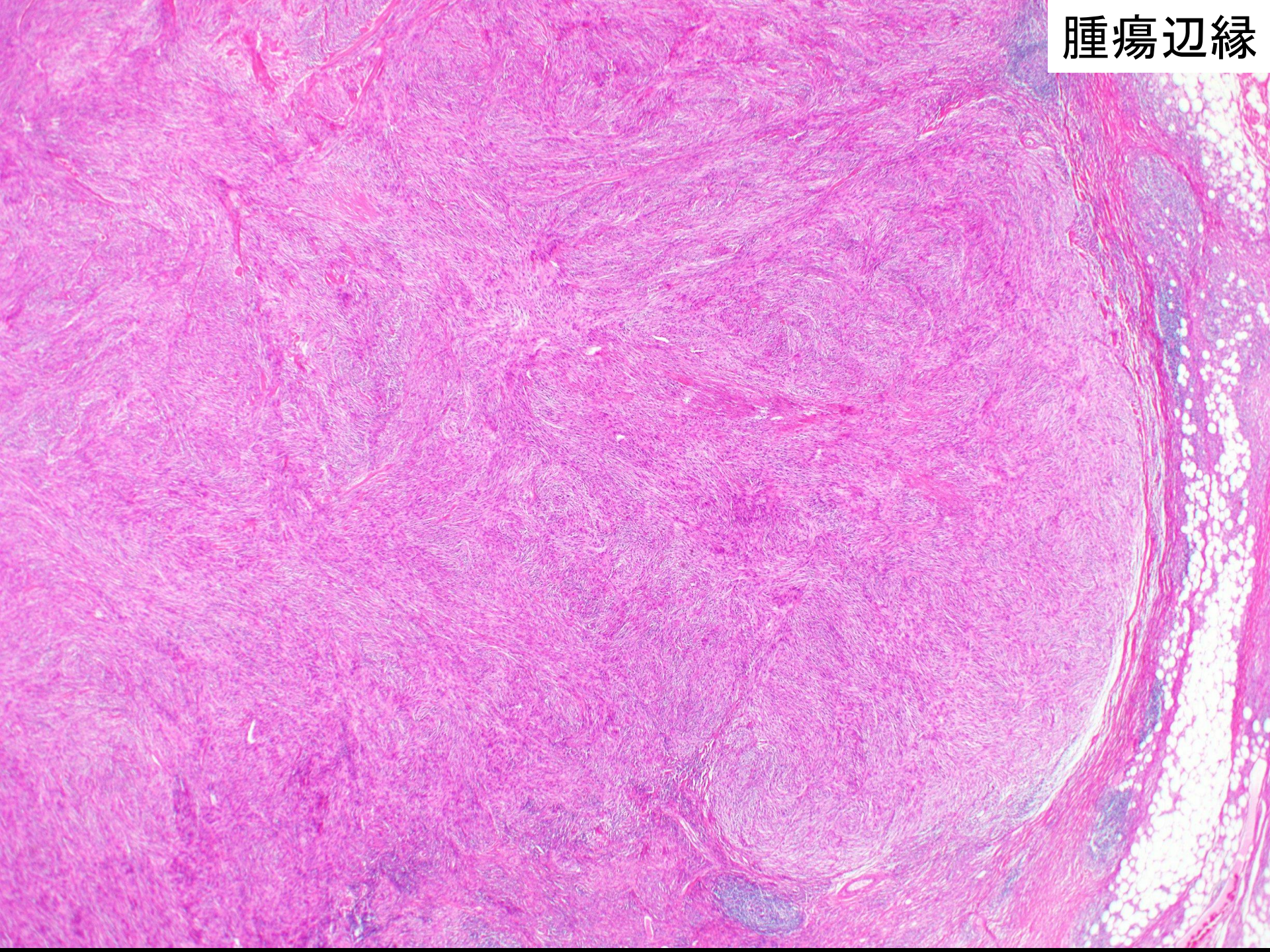


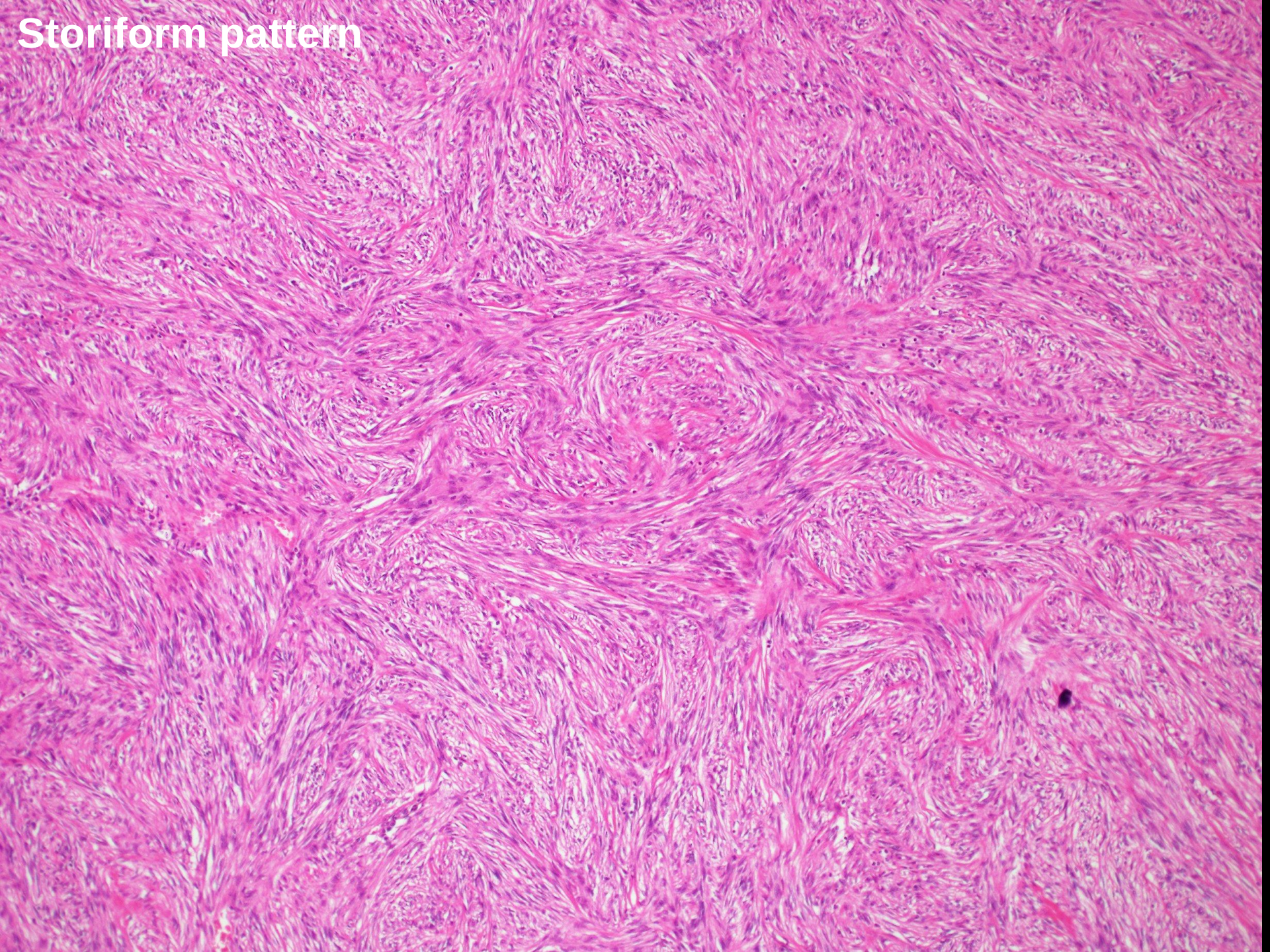


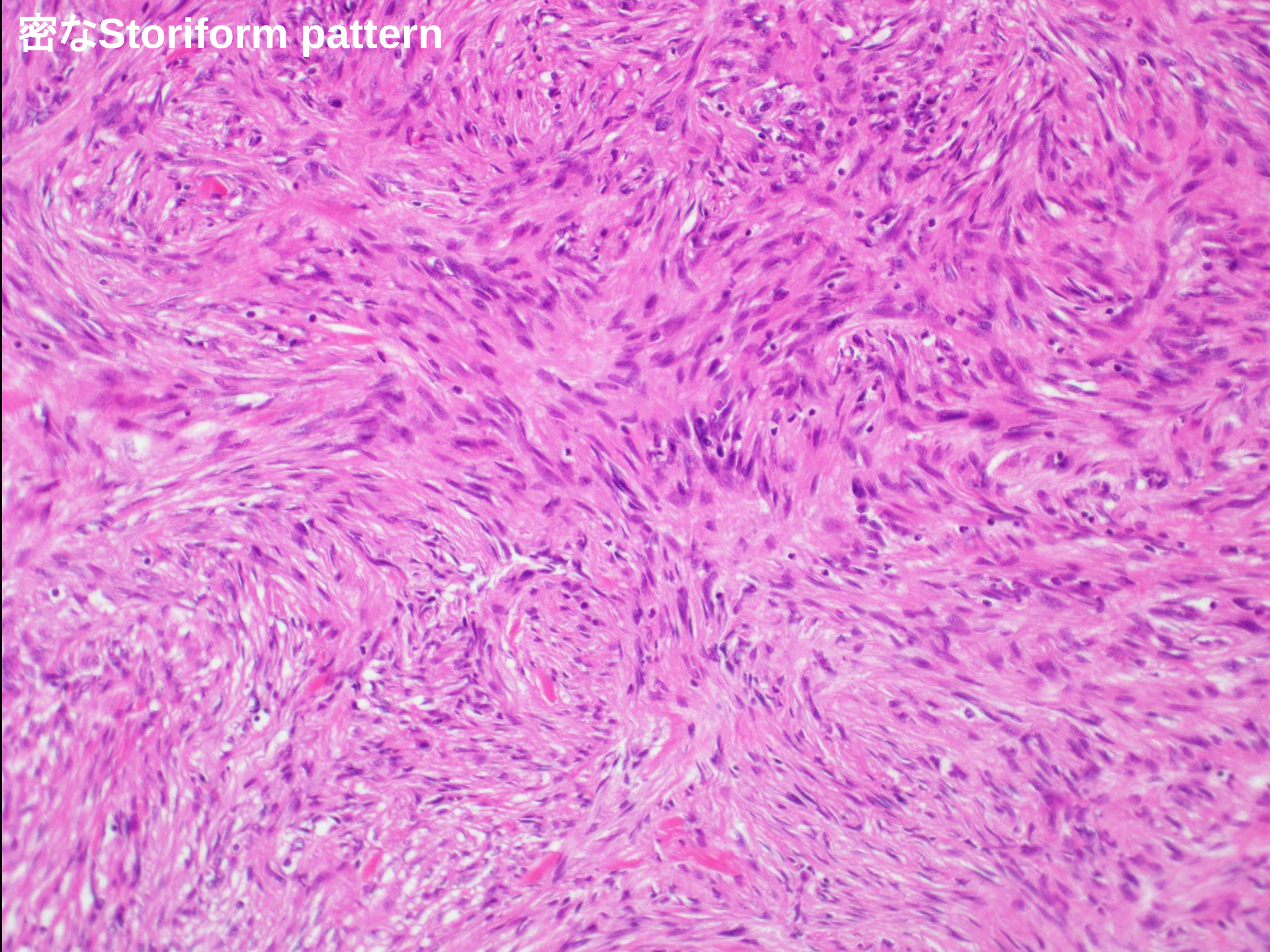
表面の表皮過形成変化
とメラニンの沈着



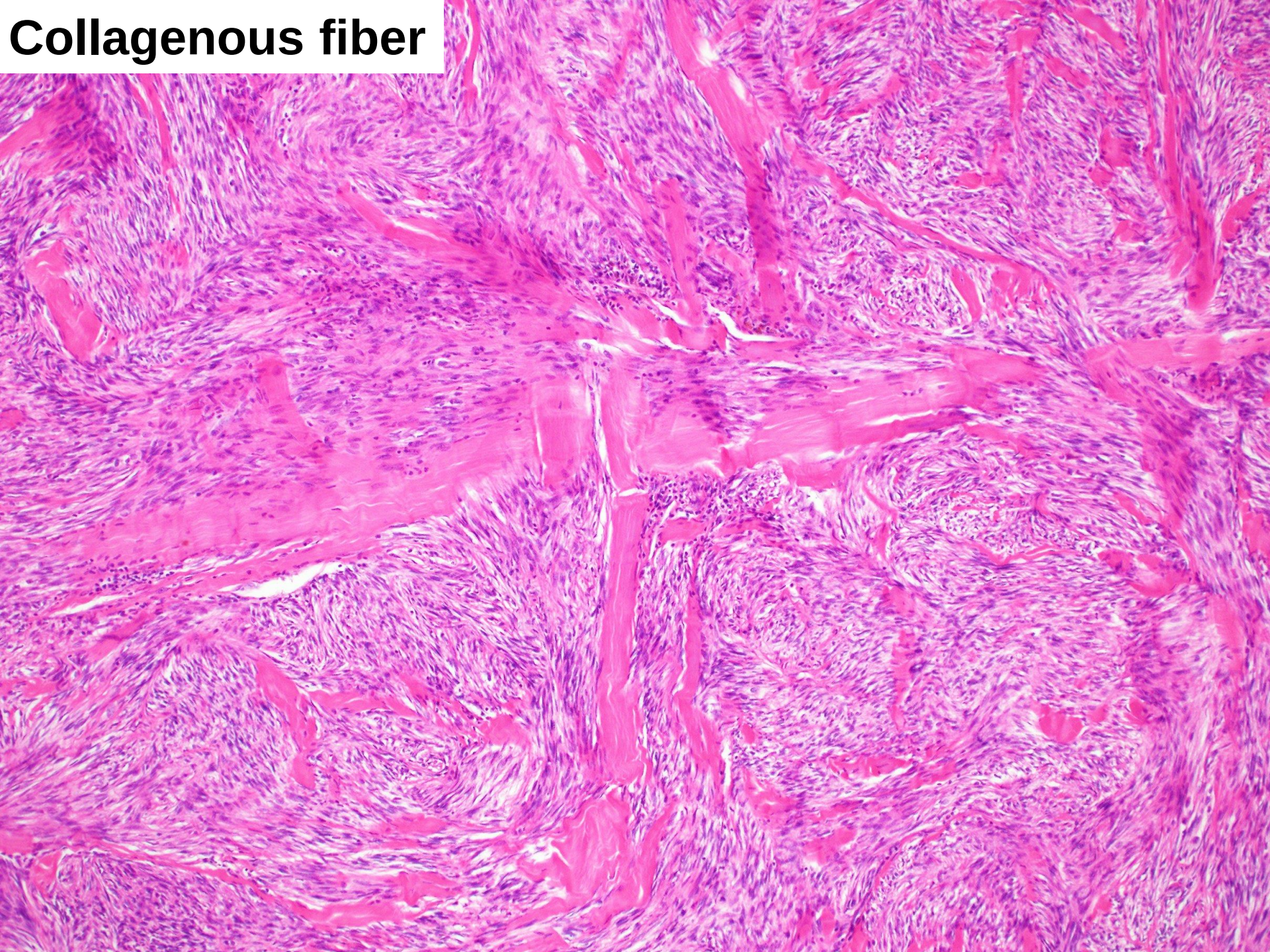
表皮の潰瘍

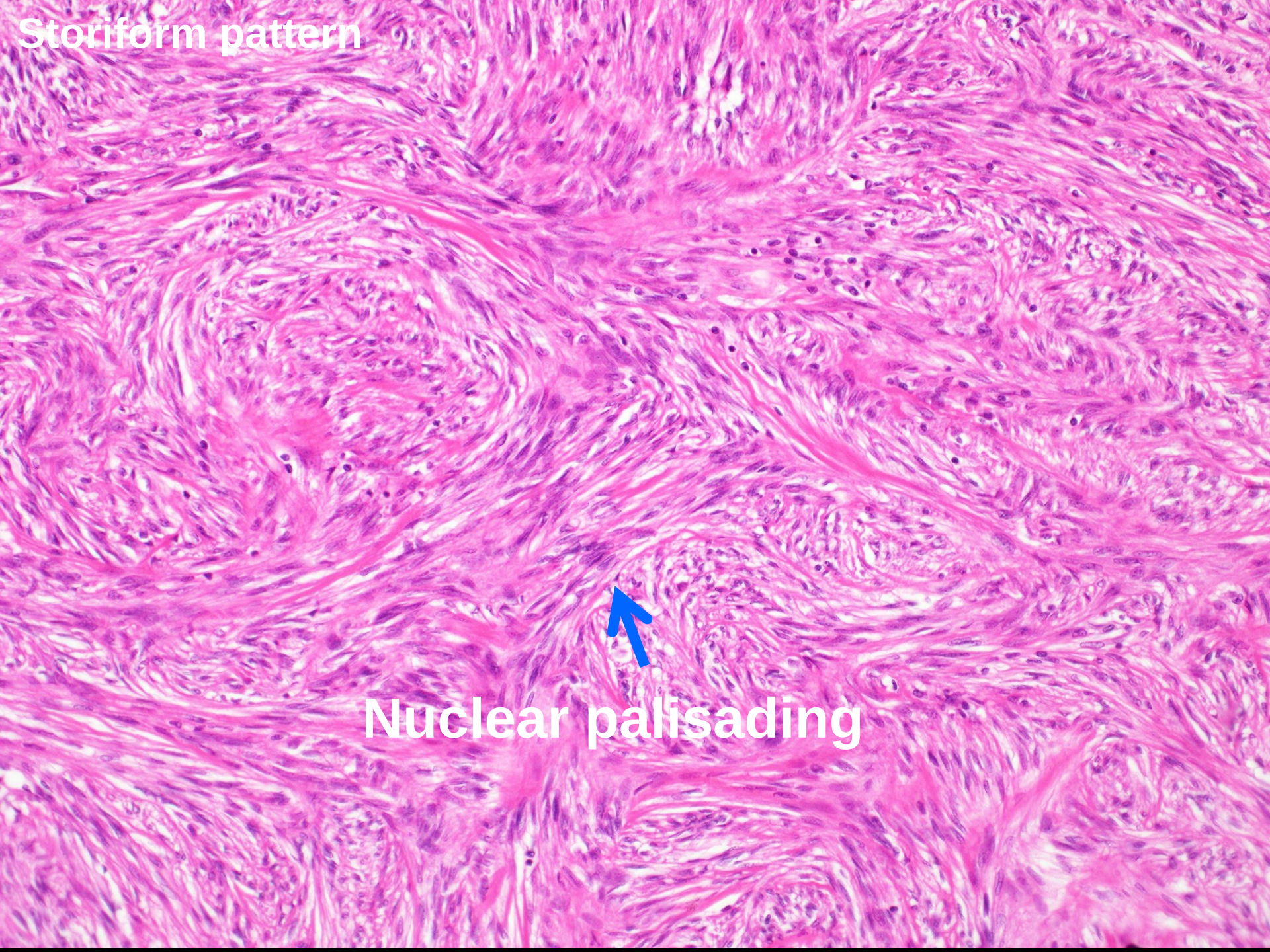

腫瘍辺縁

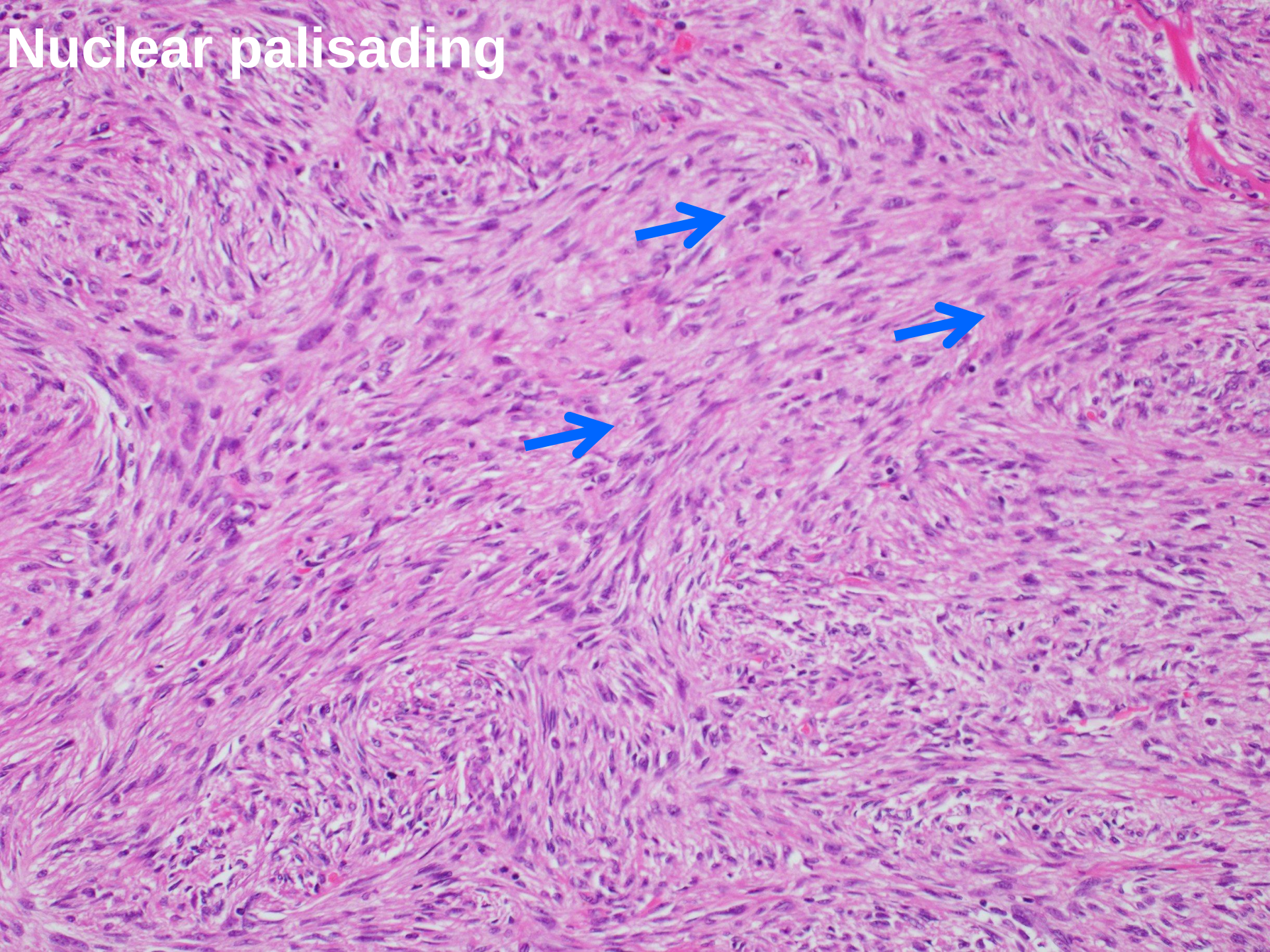

Storiform pattern


密なStoriform pattern

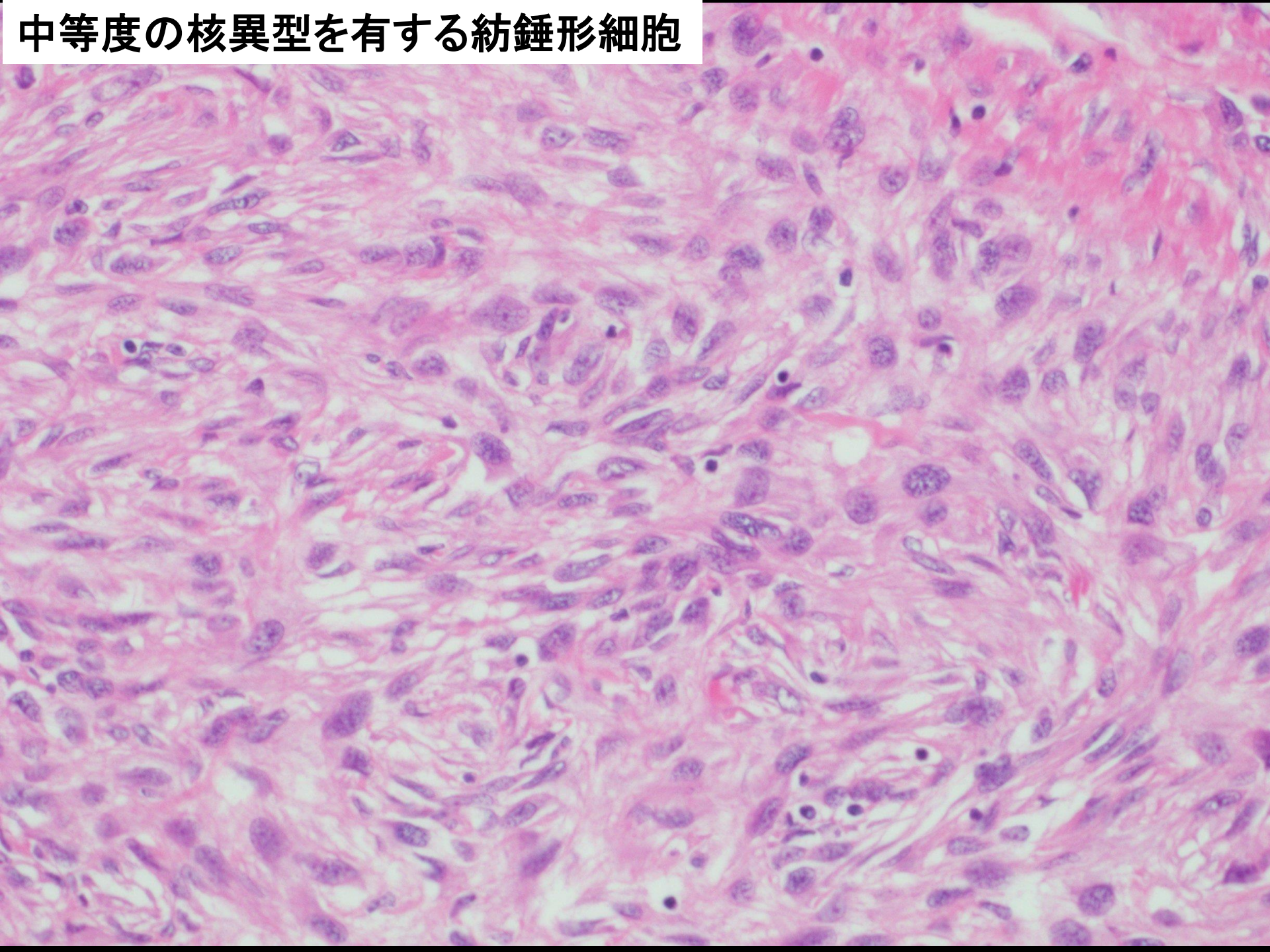
Collagenous fiber

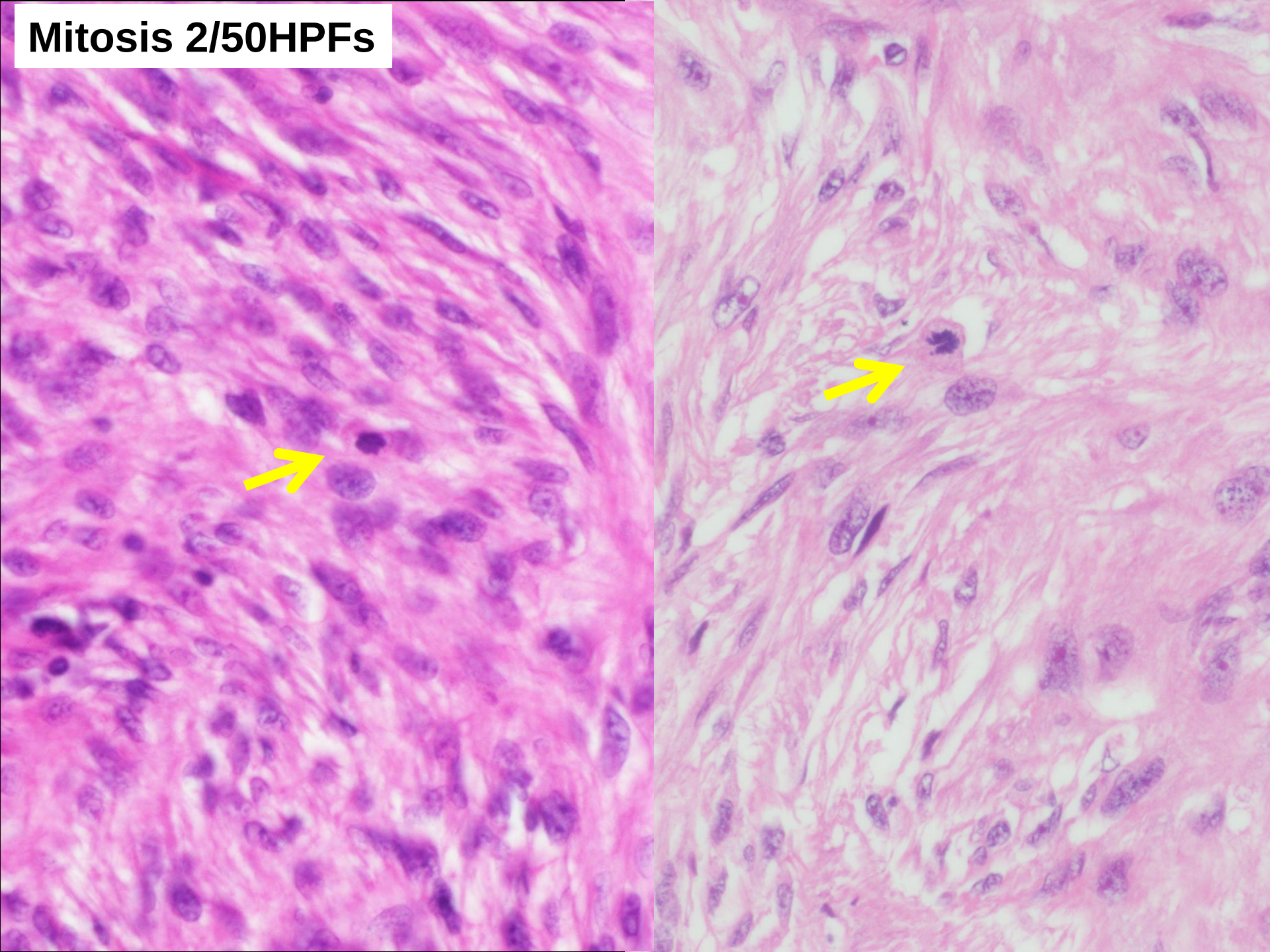


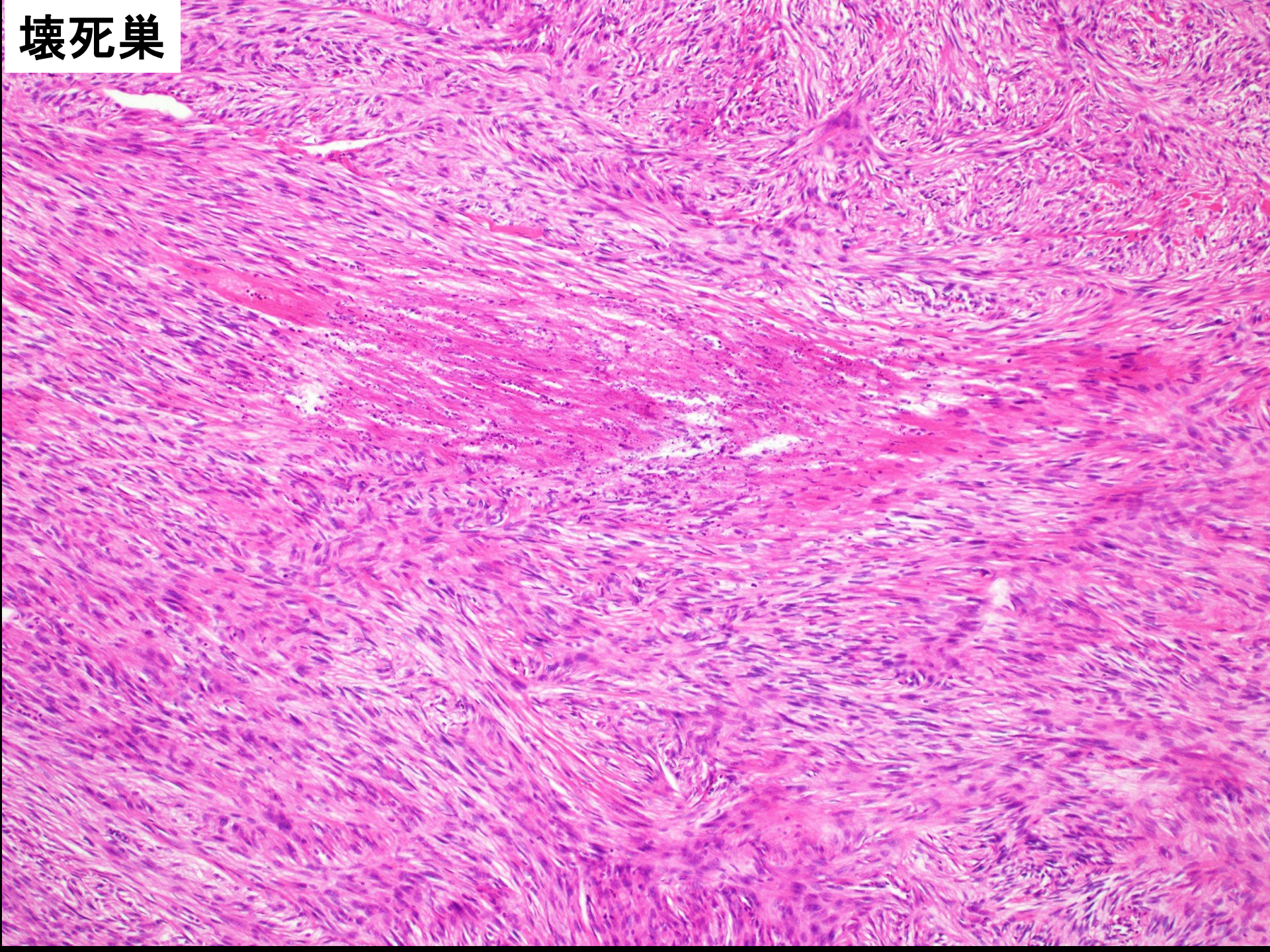

Storiform pattern
Nuclear palisading

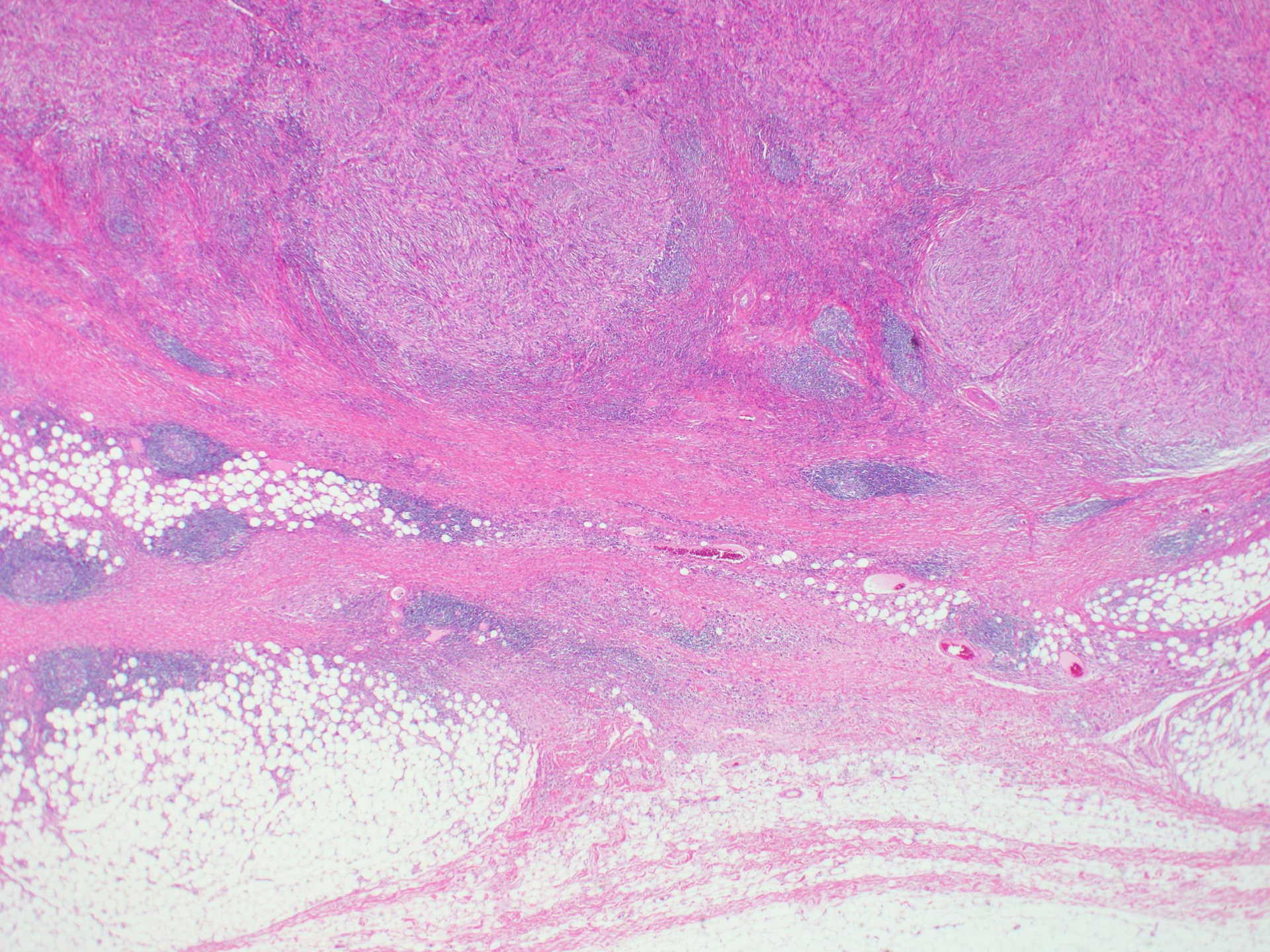

Nuclear palisading

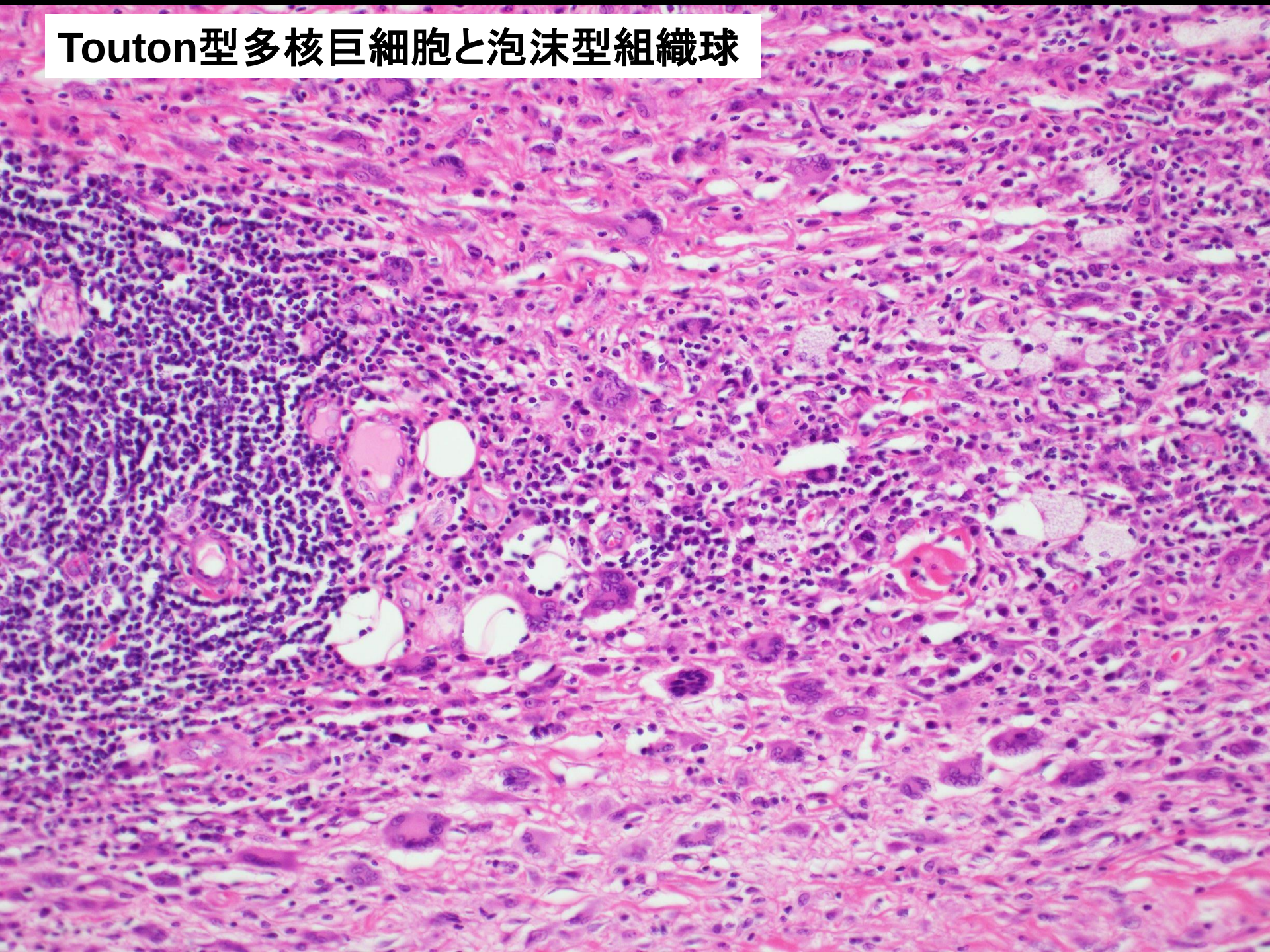
中等度の核異型を有する紡錘形細胞




Mitosis 2/50HPFs


壊死巣

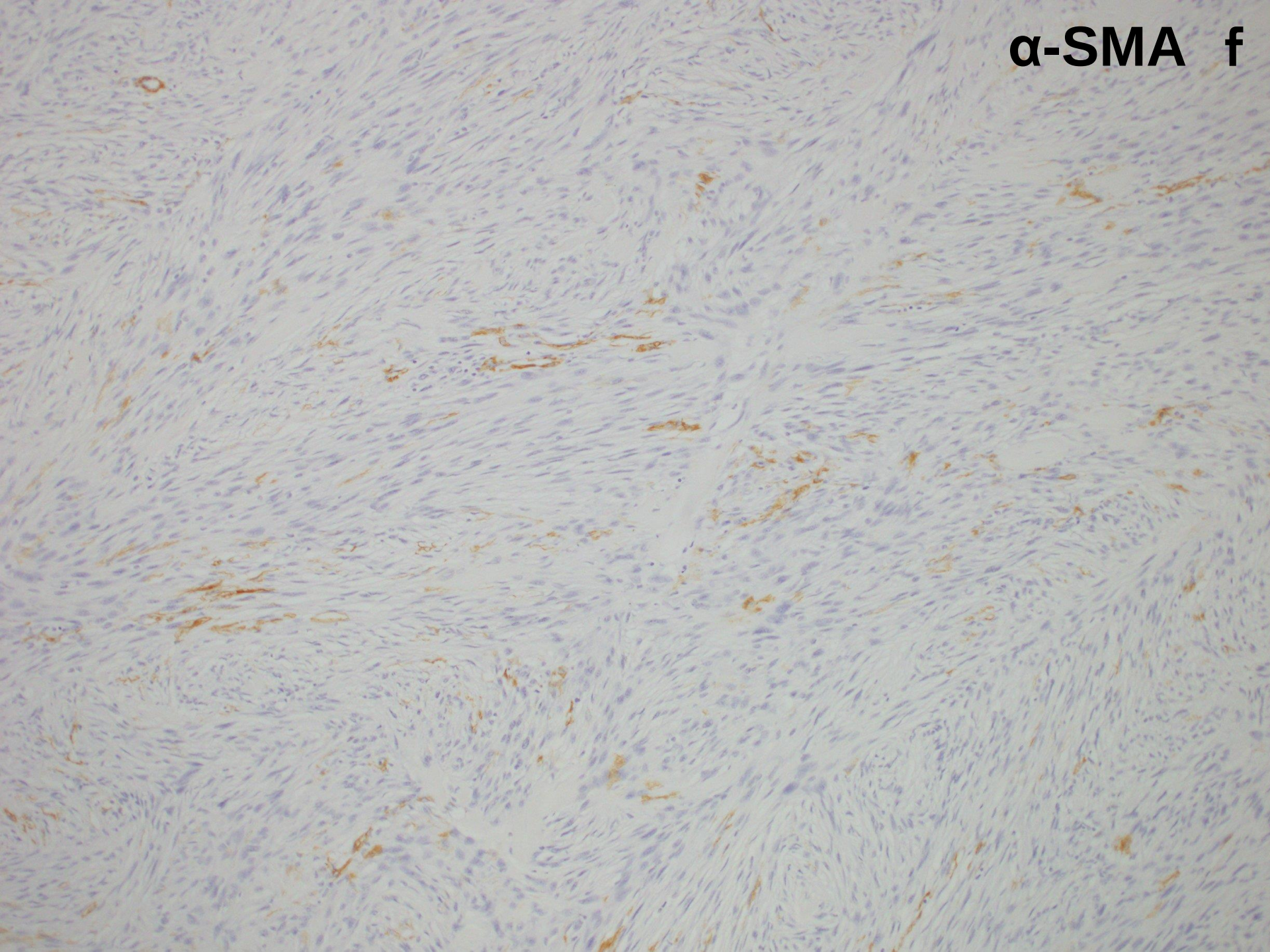



Touton型多核巨細胞と泡沫型組織球

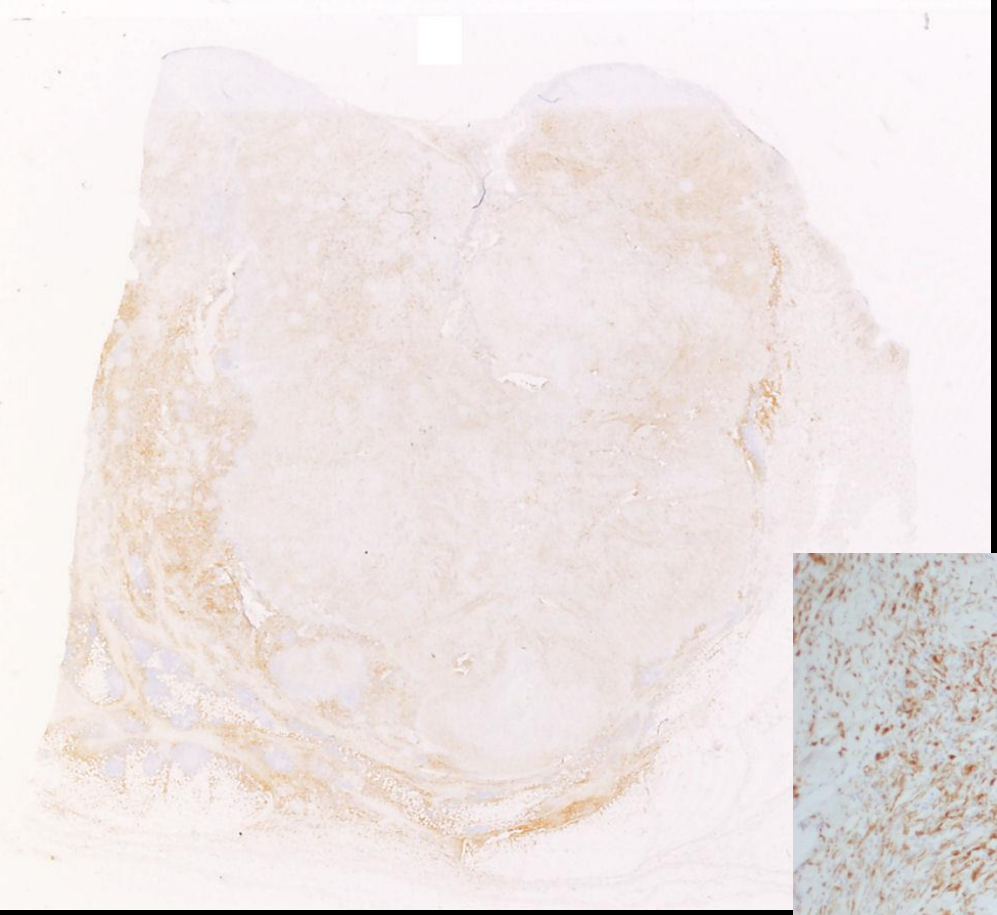
鑑別診断

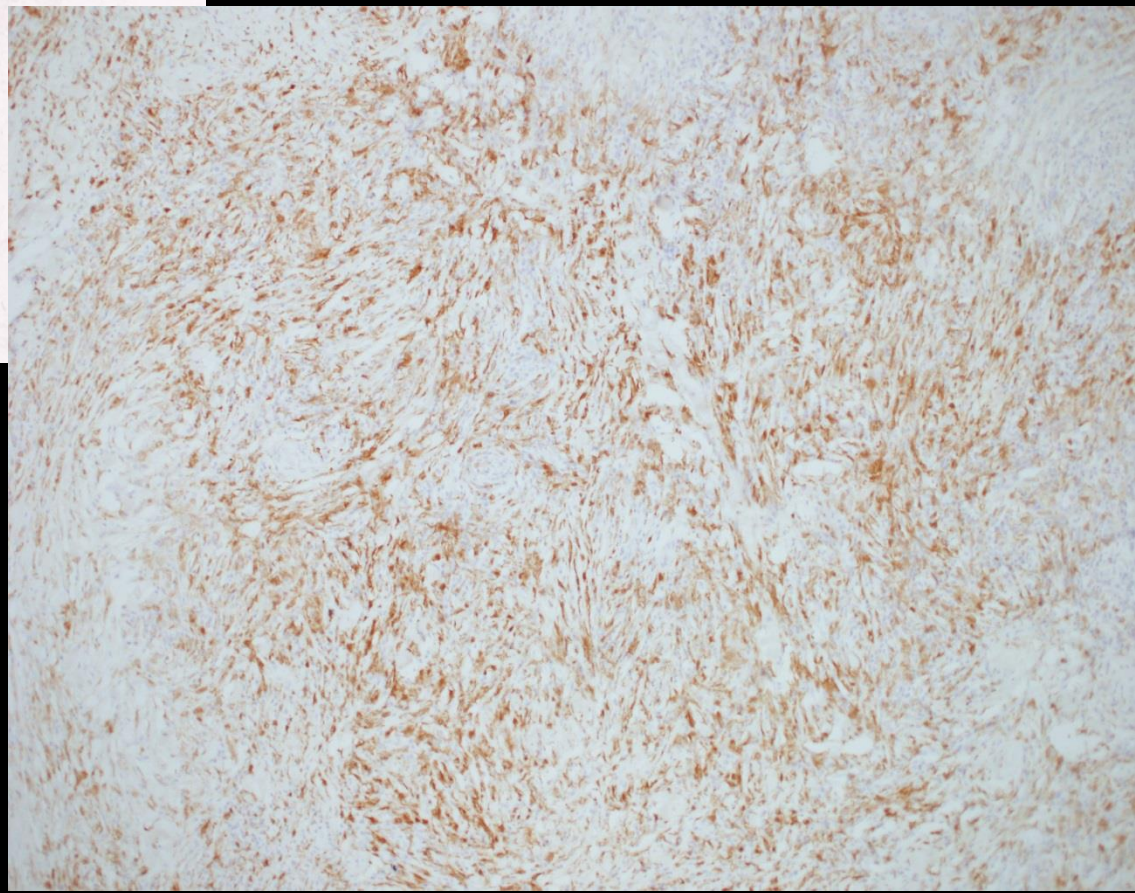
Cutaneous spindle cell tumors

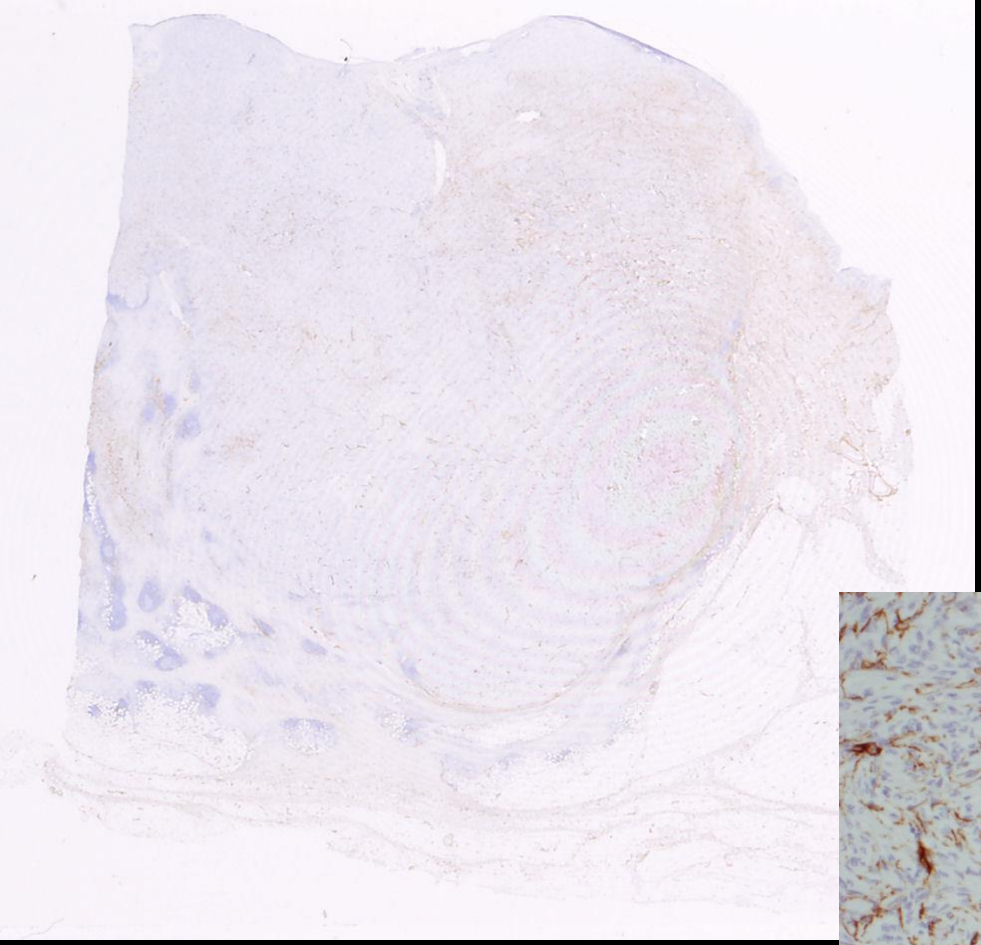
- Cellular fibrous histiocytoma (CFH)
- Dermatofibrosarcoma protuberans
- Atypical fibrous histiocytoma
- Atypical fibroxanthoma
- Inflammatory myofibroblastic tumor
- low grade MPNST
- Perineurioma
- Fibrosarcoma


α-SMA f

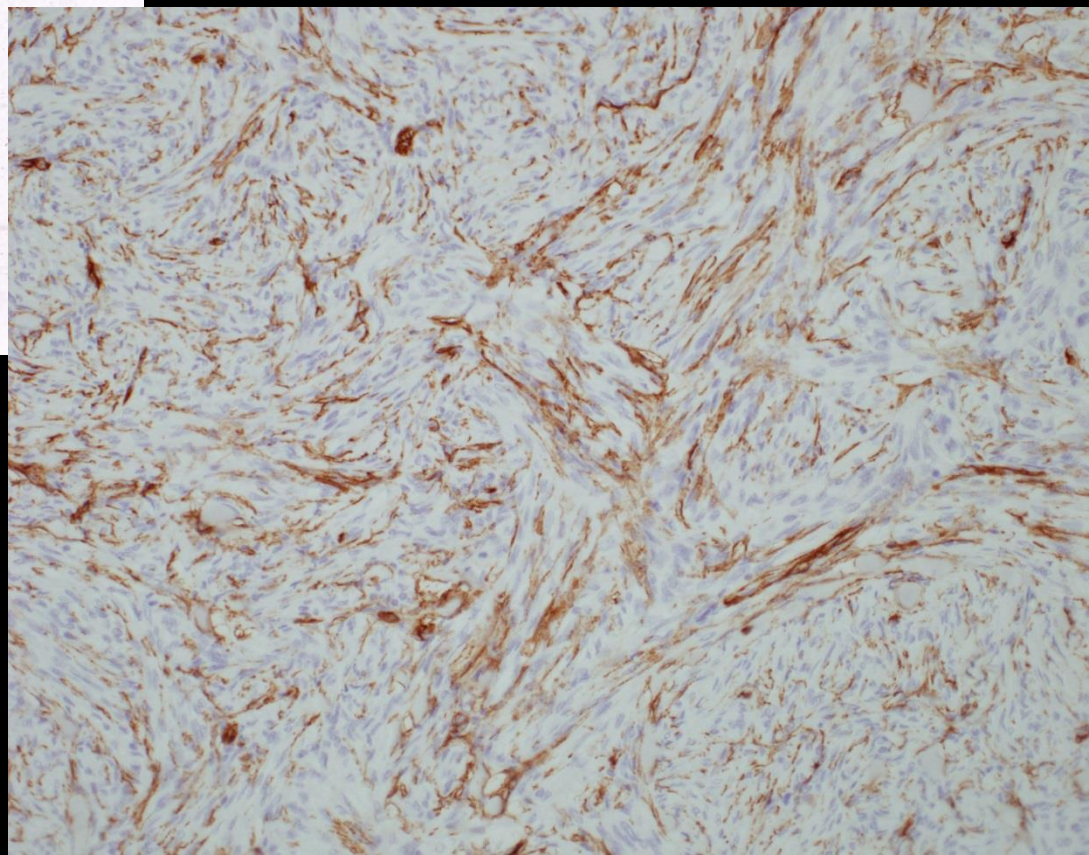
Factor XIIIa

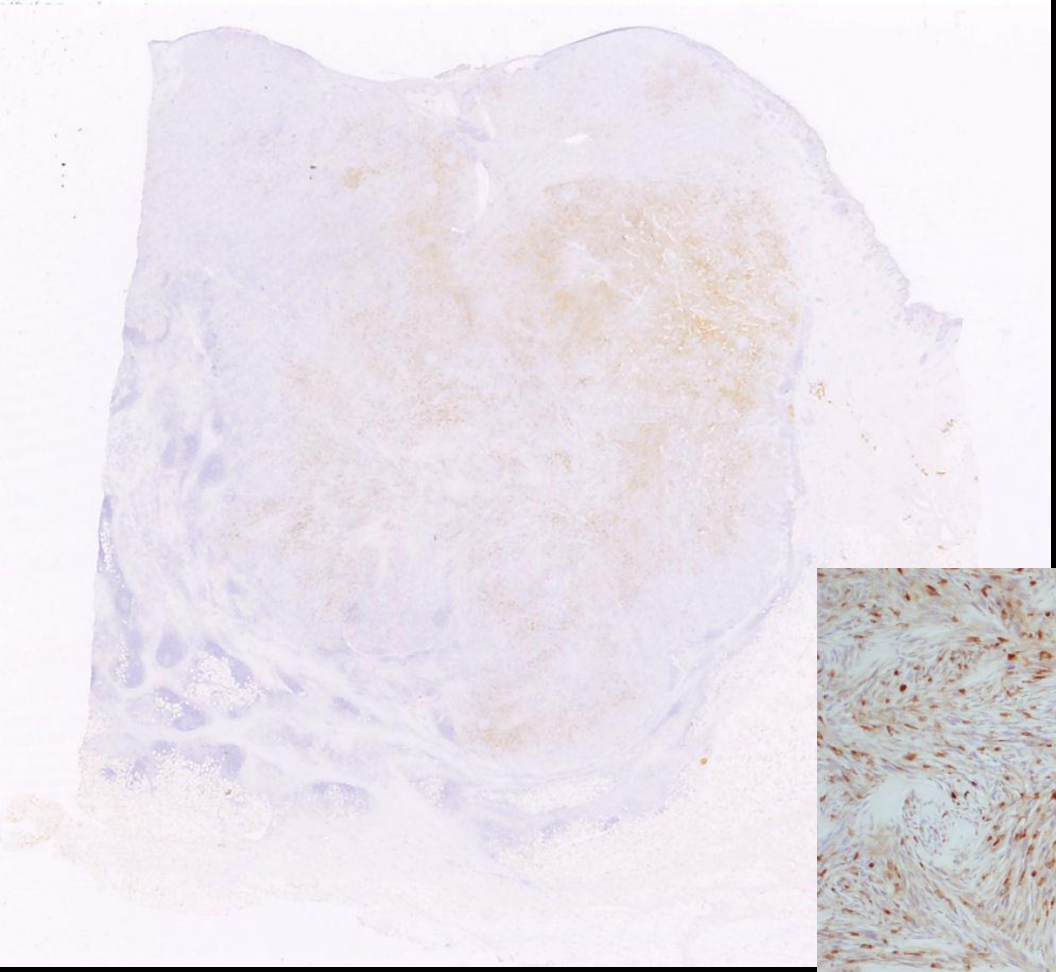




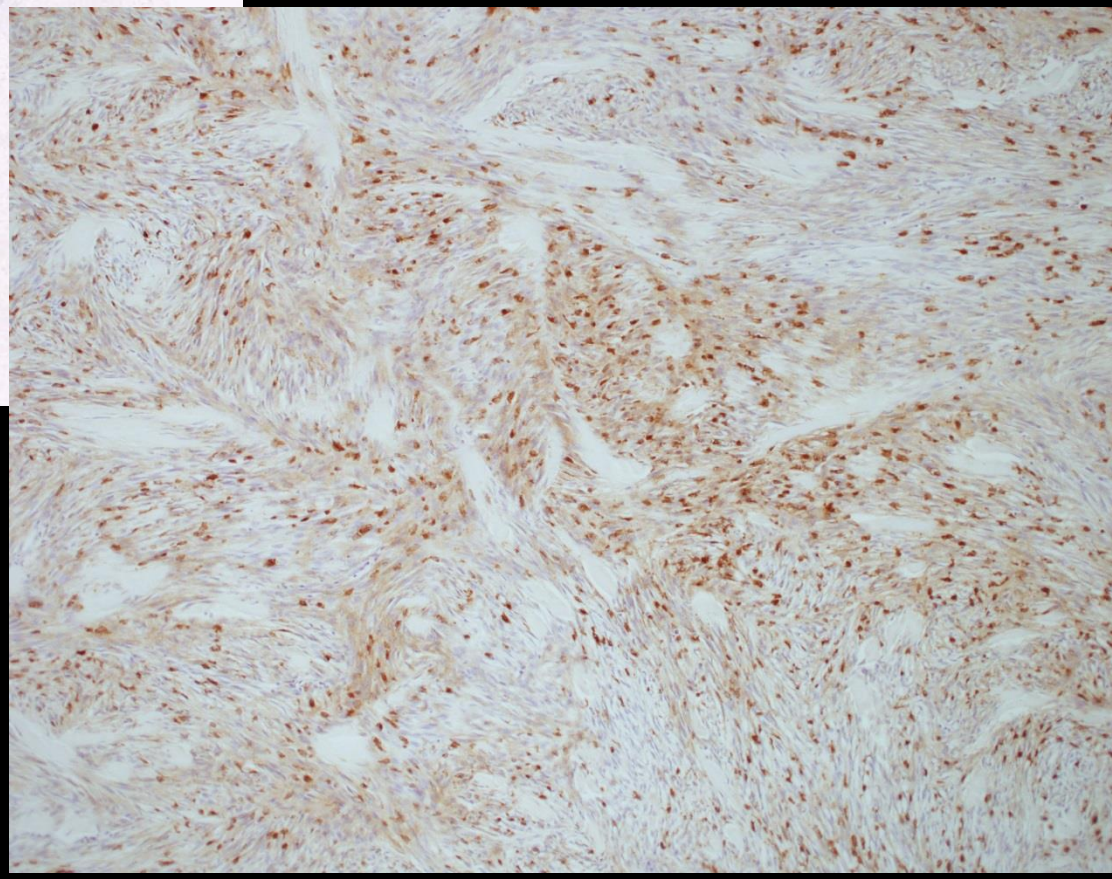


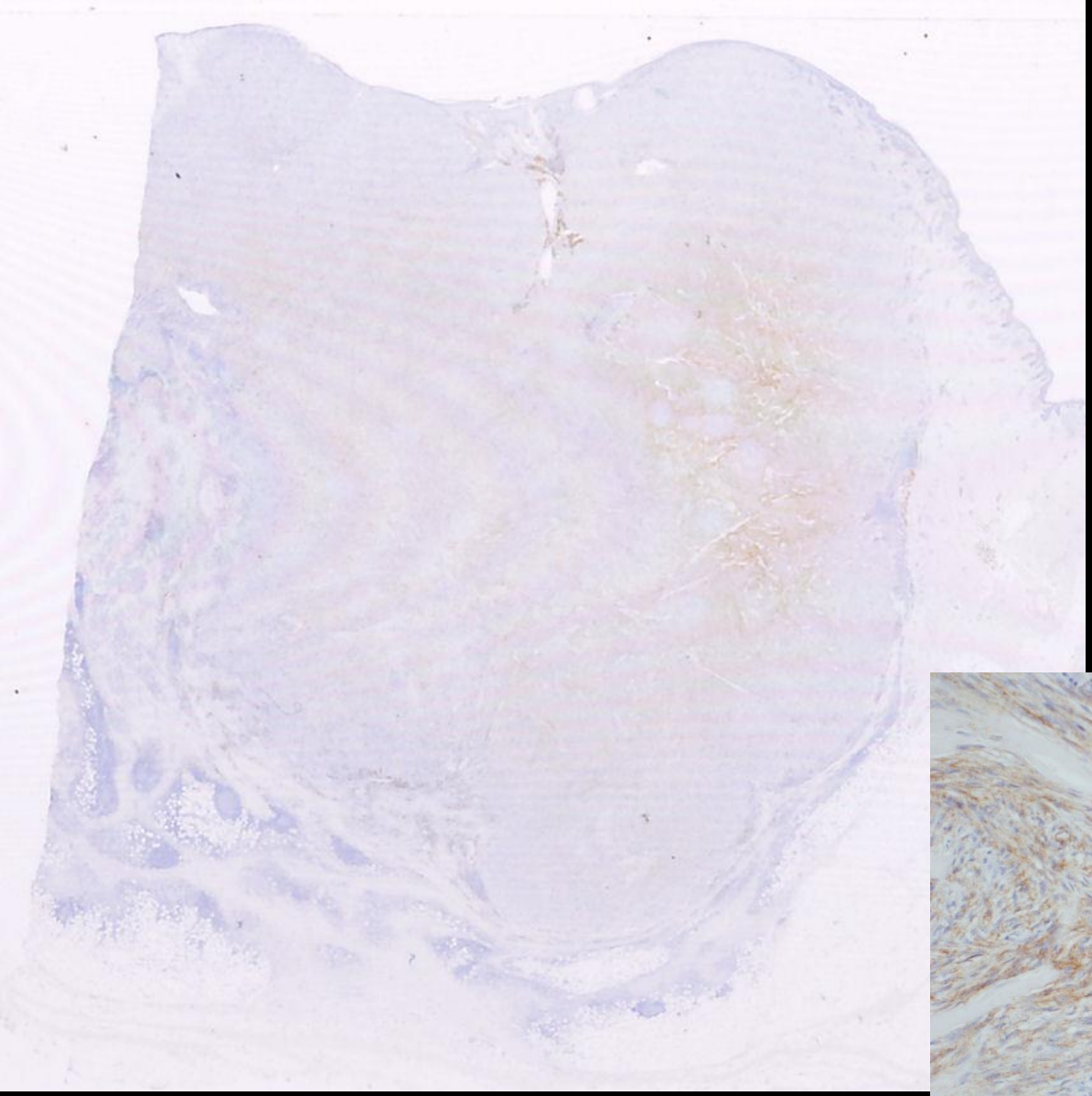
CD34 focal



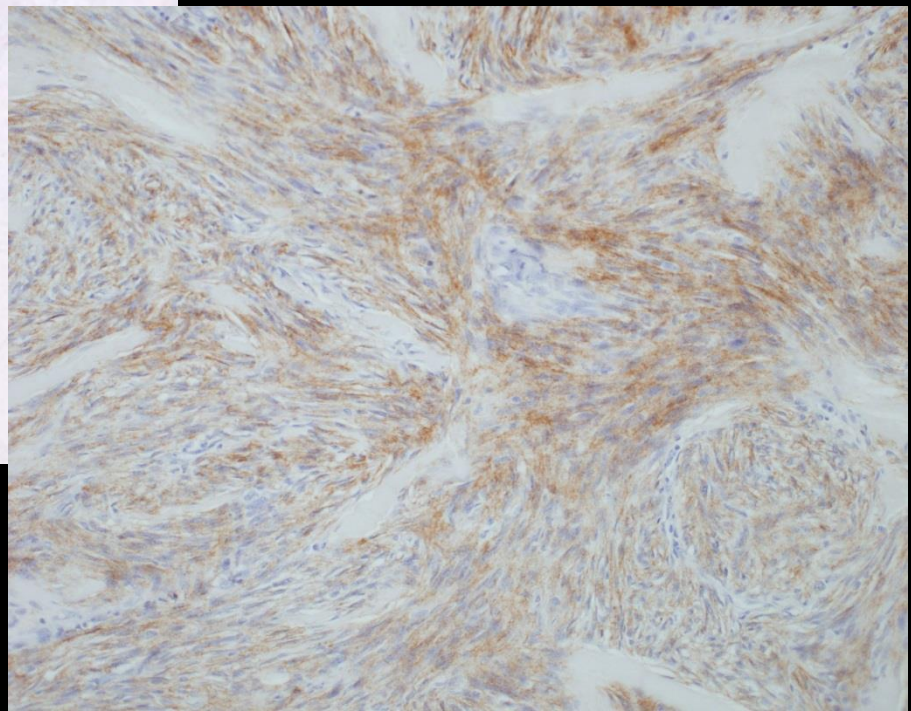


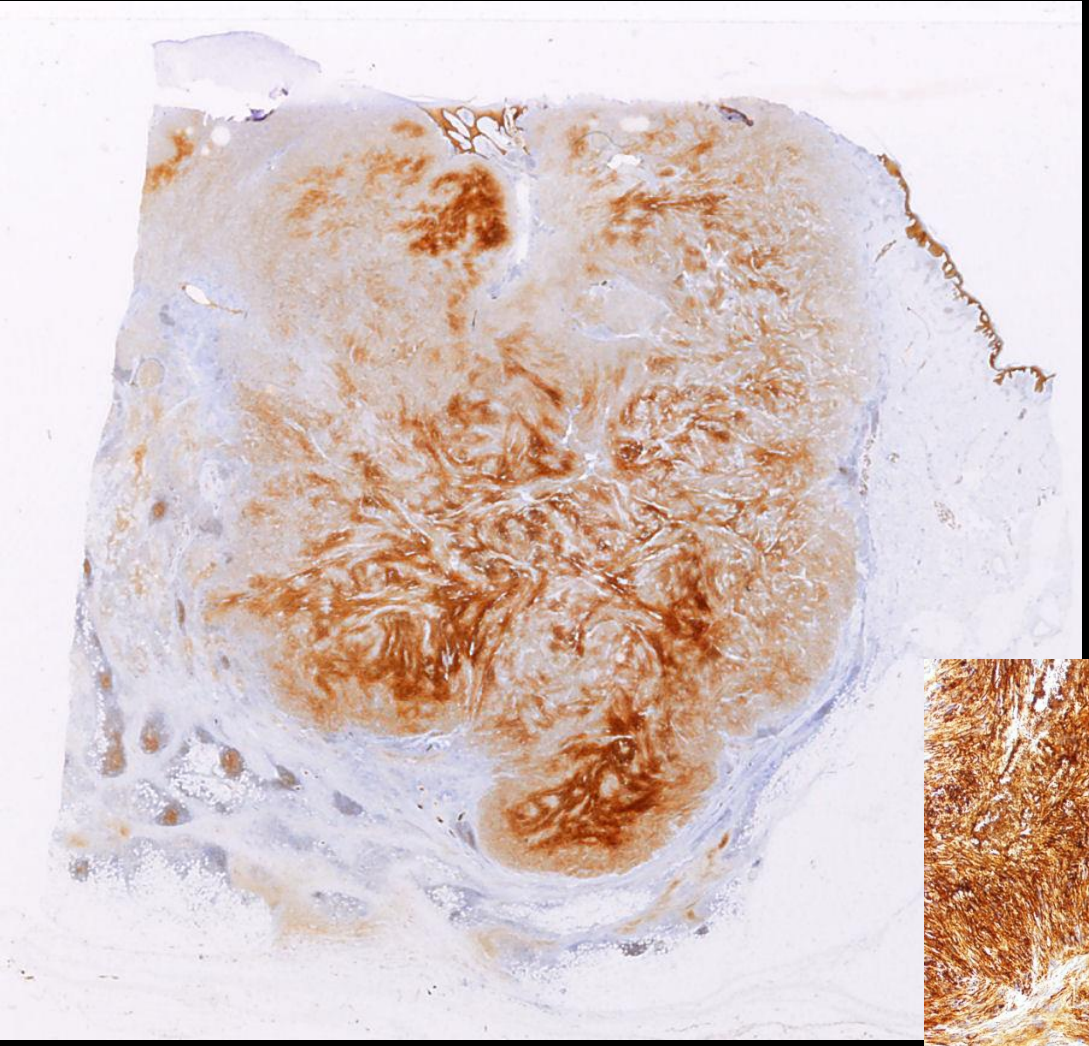
S100



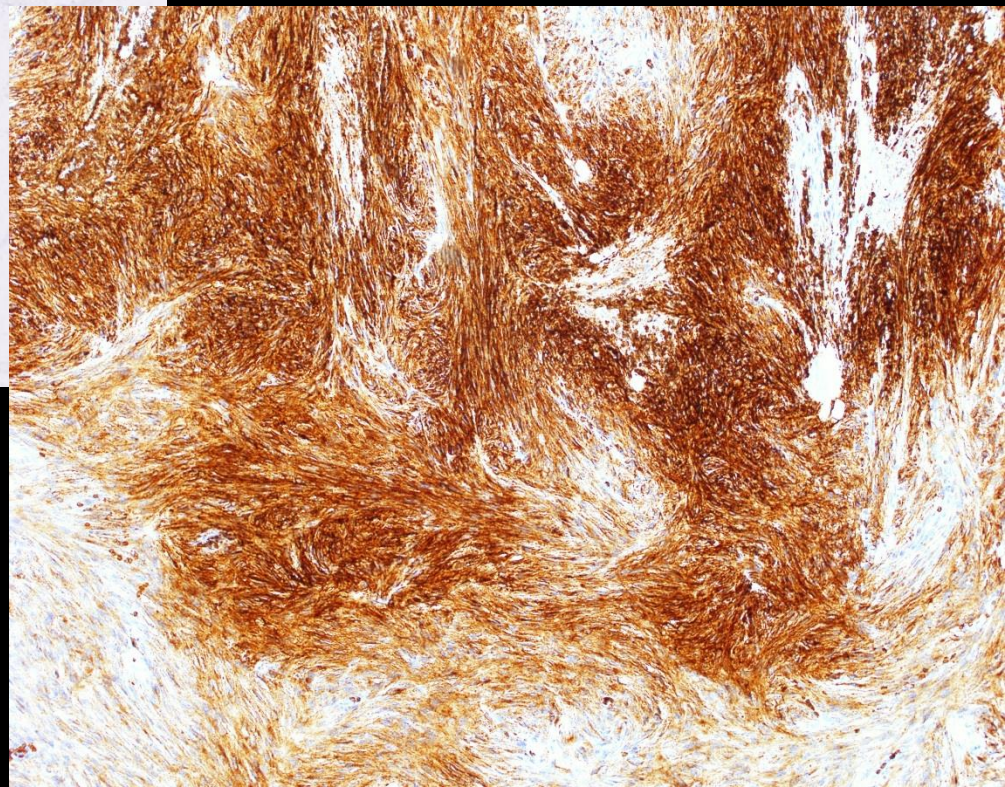


EMA





GLUT-1



D2-40, a novel immunohistochemical marker in differentiating dermatofibroma from dermatofibrosarcoma protuberans

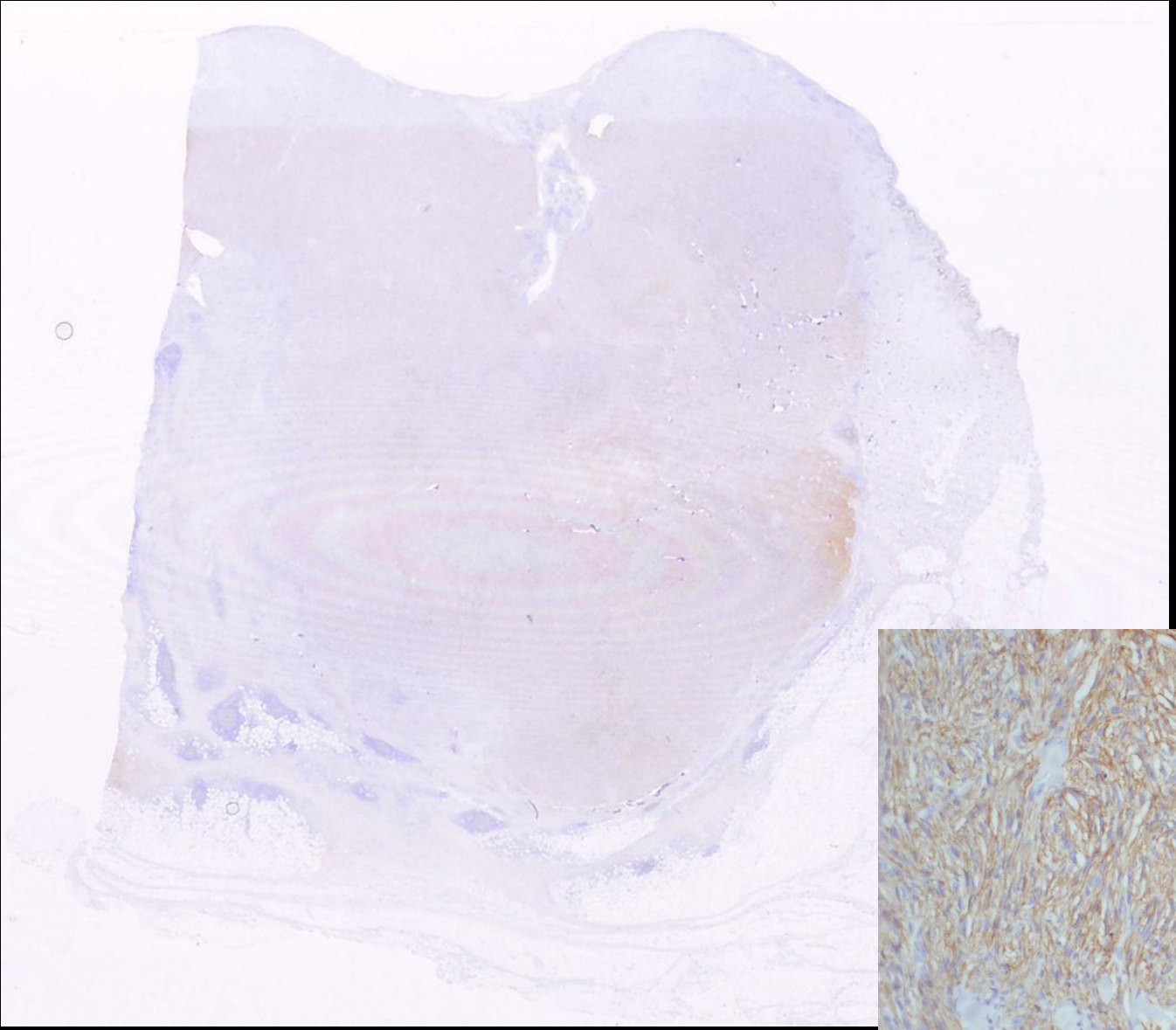
Bizhan Bandarchi[1], Linglei Ma[2,3], Celia Marginean[4], Sara Hafezi[1], Judit Zubovits[1] and Golnar Rasty[1]

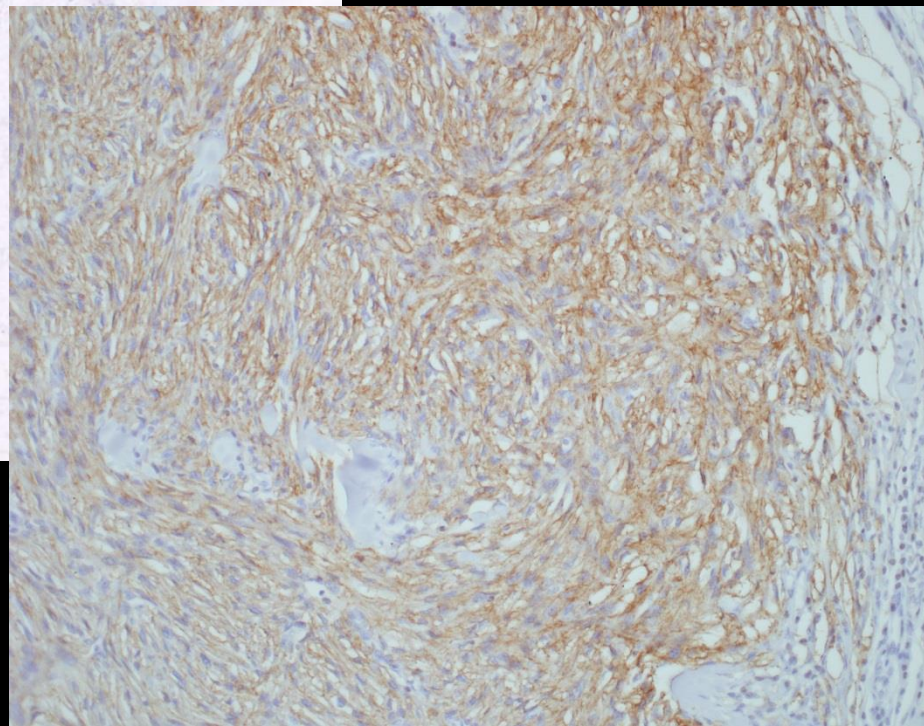
[1]*Department of Anatomic Pathology, Sunnybrook Health Sciences Center, University of Toronto, Toronto, ON, Canada;* [2]*Department of Pathology, University of Michigan Medical Center, Ann Arbor, MI, USA;* [3]*Department of Dermatology, University of Michigan Medical Center, Ann Arbor, MI, USA and* [4]*Department of Pathology, Ottawa Hospital, University of Ottawa, Ottawa, ON, Canada*

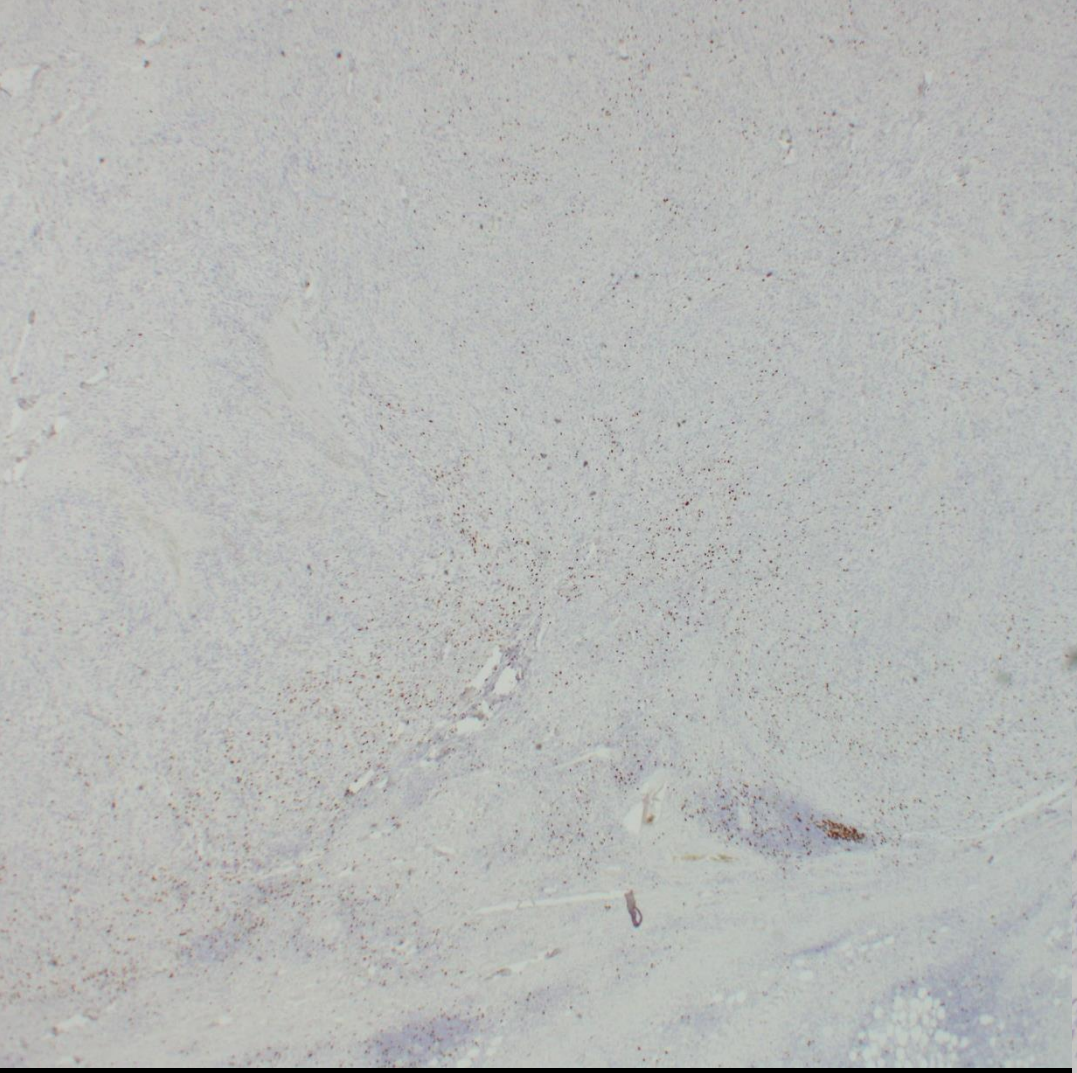
Modern Pathology(2010)23. 434-438

	症例数	CD34	Factor XIIIa	D2-40
Dermatofibroma (cellular variant)	56 (6)	1 (1)	56 (6)	56 (6)
DFSP	29	29	0	4 focal

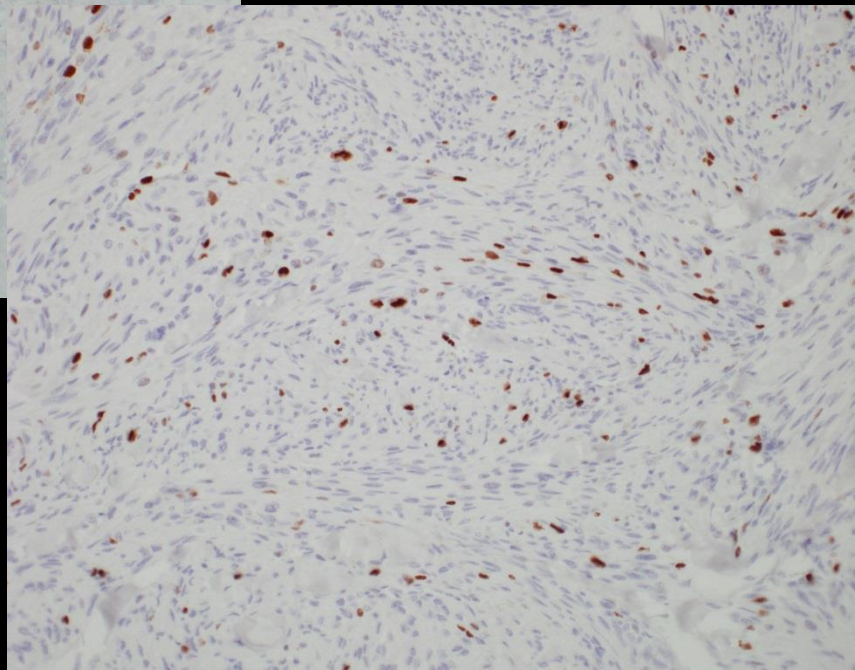
D2-40







MIB-1 index 12%



免疫染色結果

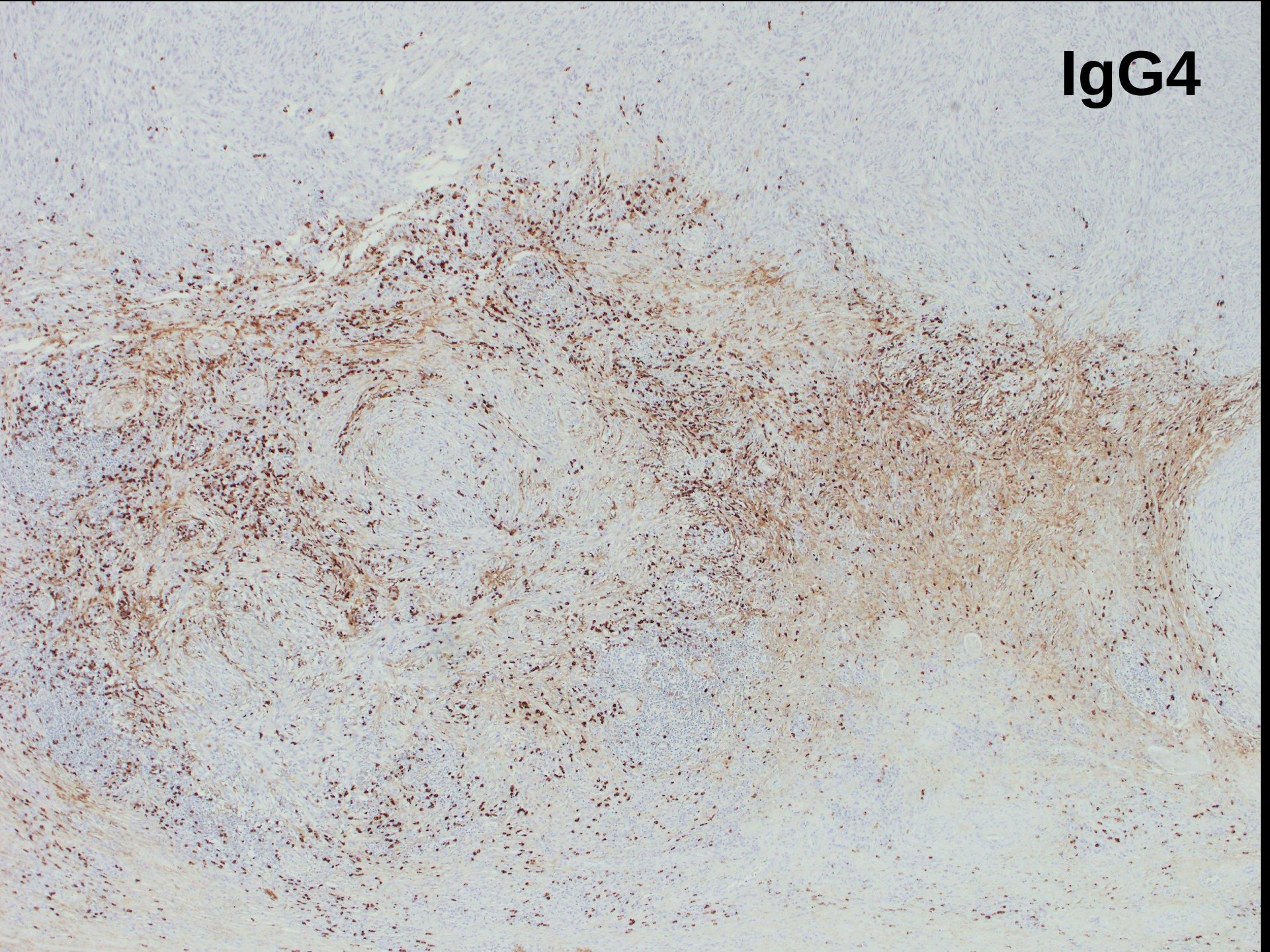
Positive

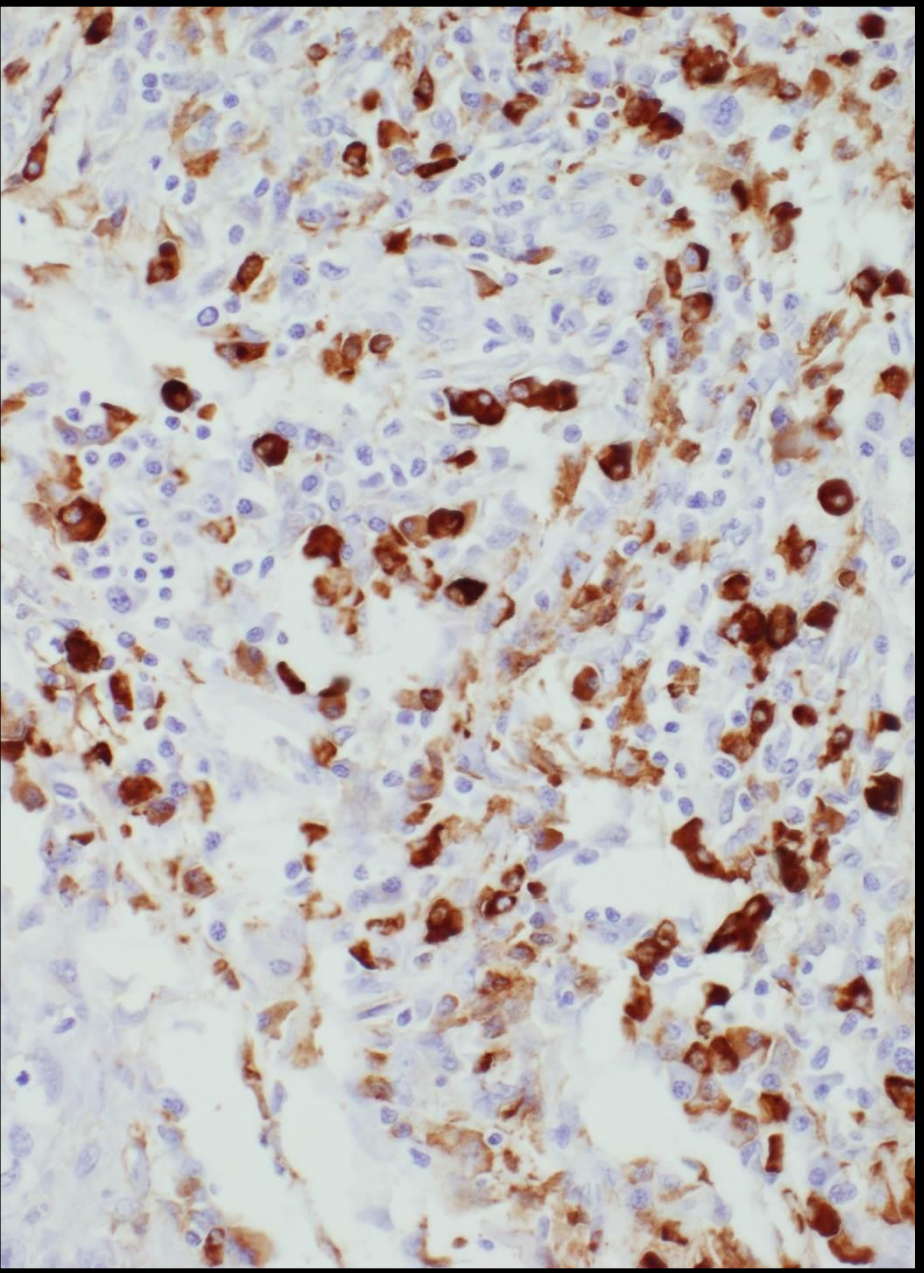
- Factor XIIIa ++
- S100 +
- Gult-1 ++
- D2-40 +
- CD34 + f
- α-SMA + f
- Bcl-2 + f
- EMA + f
- NSE + f
- MIC-2 + f
- MIB-1 index 12%

Negative

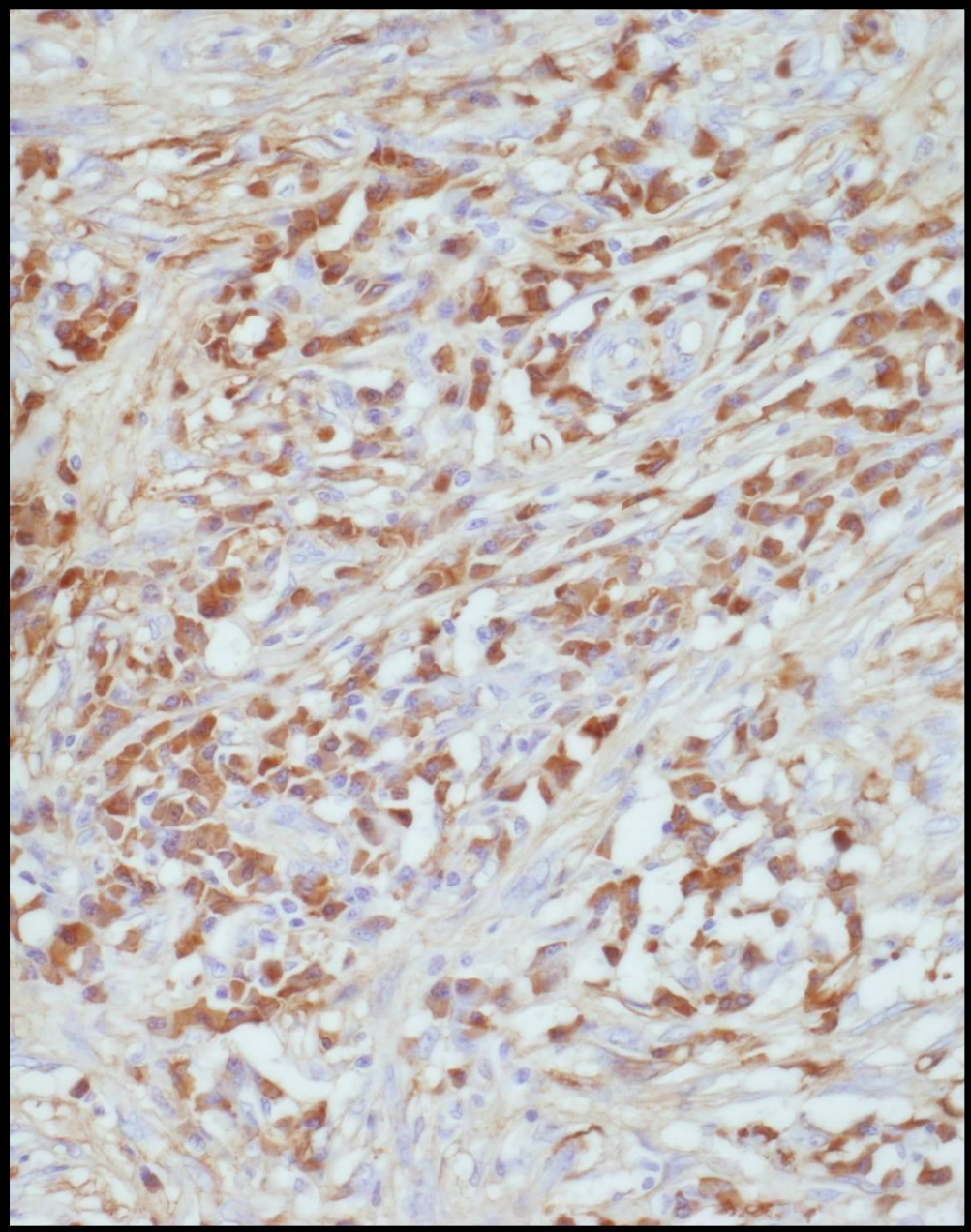
- Calponin
- Desmin
- NF

IgG4 : 109～132/HPF
IgG4/IgG : 37～41%


IgG4



IgG4



IgG

IgG4/IgG＝37-41%

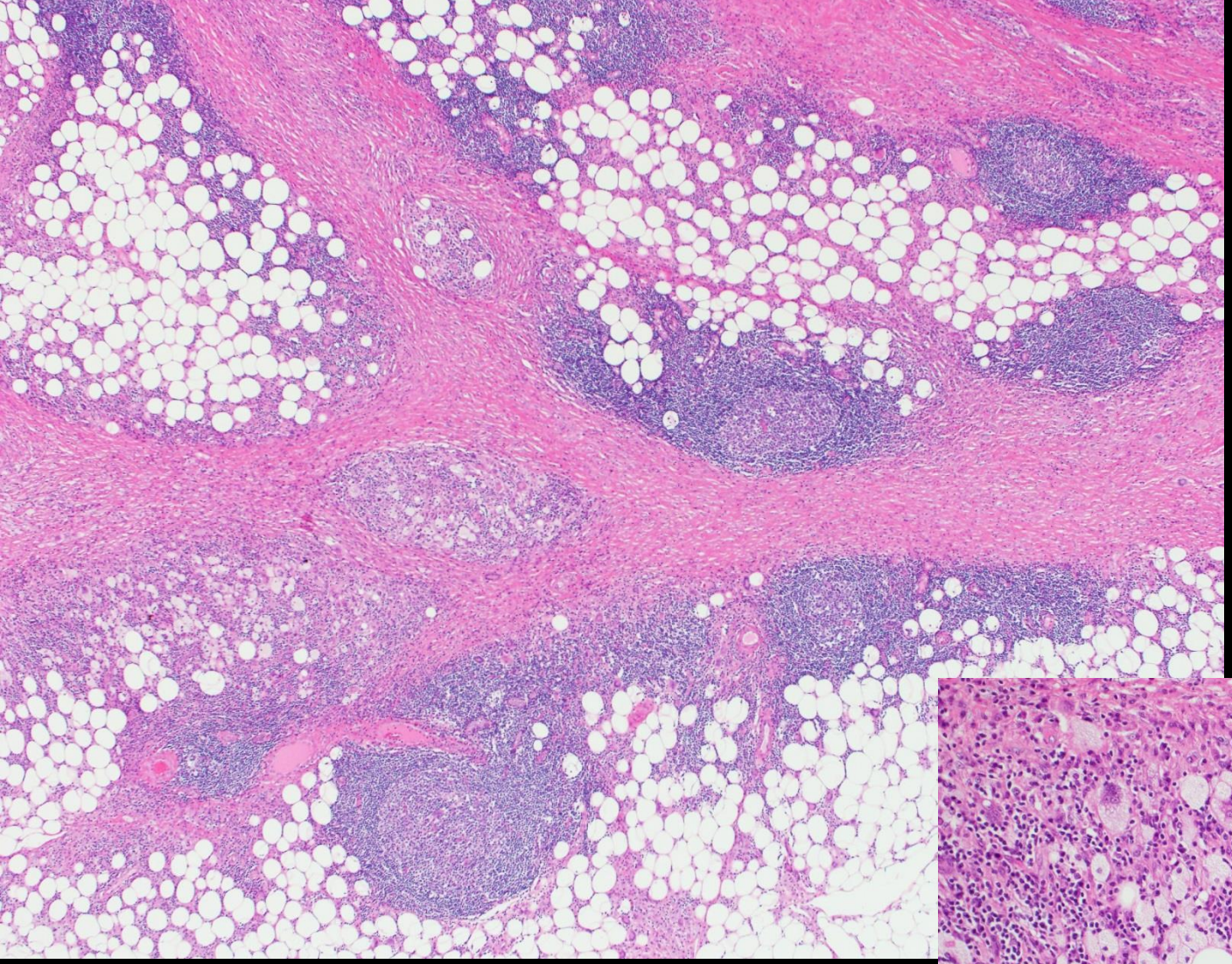
この腫瘍の特徴

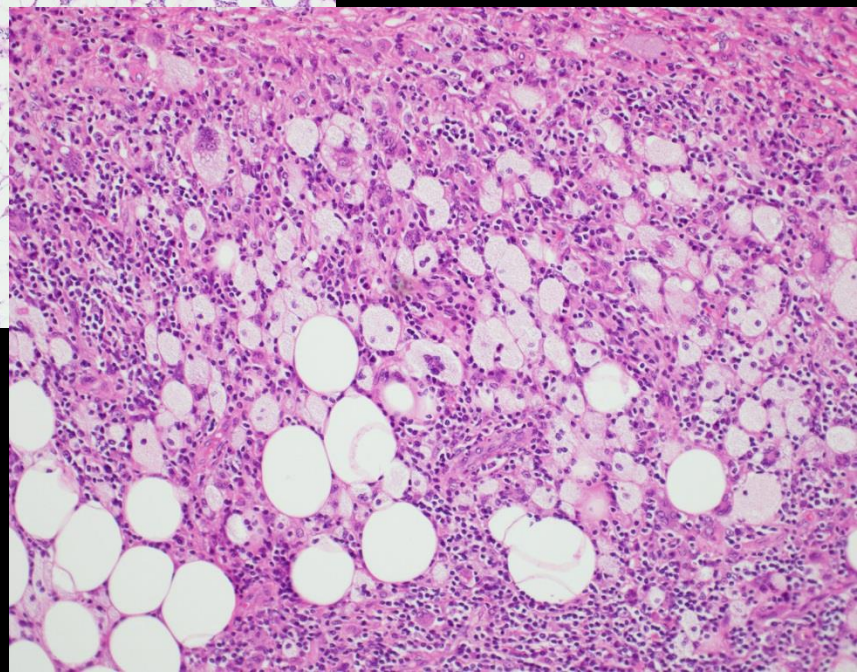
- ‘10年前’から存在する slow growing tumor
- 潰瘍を伴う隆起性腫瘤
- 表皮の過形成変化と色素沈着
- 比較的境界明瞭な白色調腫瘍
- 軽度~中等度異型を示す紡錘形細胞の増殖
- Storiform pattern, Fasicular patternが目立つ
- 核分裂像はまれ(2/50HPF)
- 辺縁に**Touton型多核巨細胞、泡沫型組織球**
- Factor XIIIa++, GULT-1++, D2-40+,　CD34 focal、
  S100 focal
- MIB-1 index　12%

Fibrous histiocytoma vs DFSP

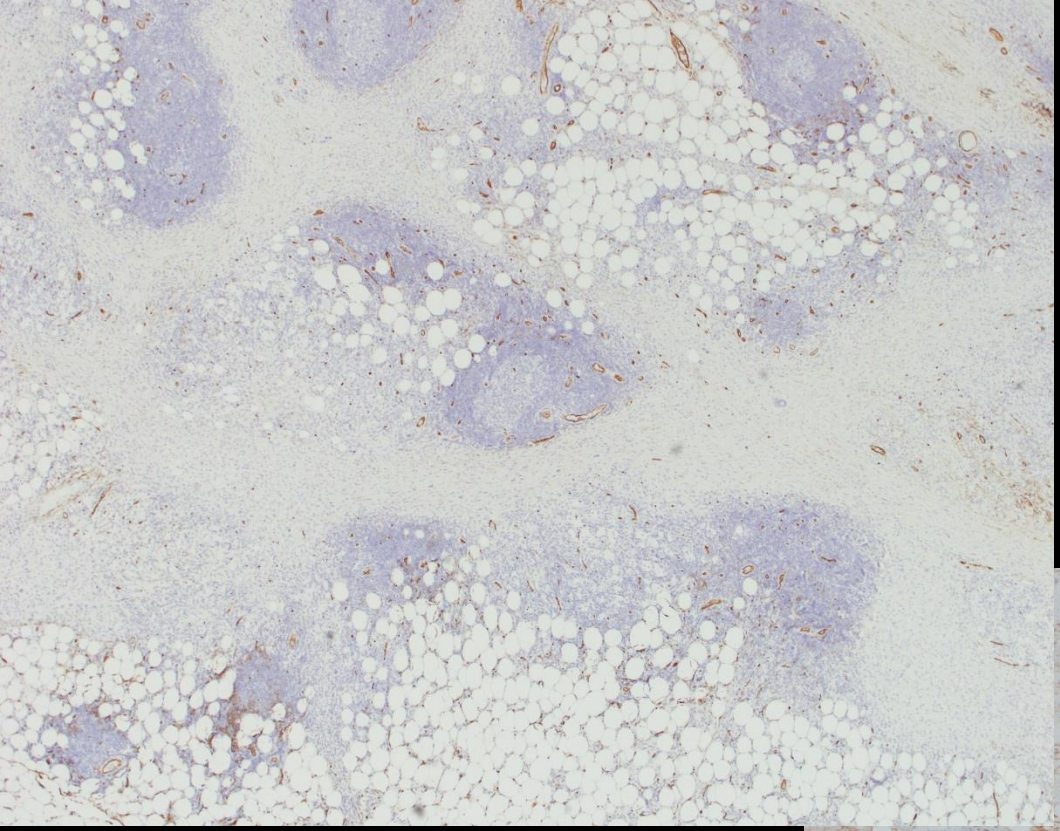
	皮膚線維腫	隆起性皮膚線維肉腫	異型線維黄色腫
性差	なし	男性優位	男性優位
好発年齢	特になし（30～40 歳に多い）	若年から中年	高齢
好発部位	四肢	体幹ないし四肢末端	頭頸部
病変の主座	真皮	真皮から皮下	真皮
構築	不規則な束状ないし花むしろ状	花むしろ状ないし車軸状	不規則な束状
対称性	対称	非対称	対称
境界	やや不明瞭	不明瞭	明瞭
核分裂	ほとんどみられない	みられる（多くの症例は＜5/10 HPF）	多い（異形核分裂あり）
Grenz zone	＋	±	－
被蓋表皮	肥厚，メラニン細胞の増生，毛包皮脂腺の増生	菲薄化ないし潰瘍化	菲薄化
再発	稀	しばしばみられる（術後 3 年以内）	時にみられる
転移	稀	稀	非常に稀
FXIIIa ＋	＋	－	一部＋
CD34	－	＋	－
p53	－	＋	±
αSMA ±	±	－	±
その他の免疫組織化学			CD10（＋），HMB-45（＋），vimentin（＋）

腫瘍病理鑑別アトラス　皮膚腫瘍Ⅰより





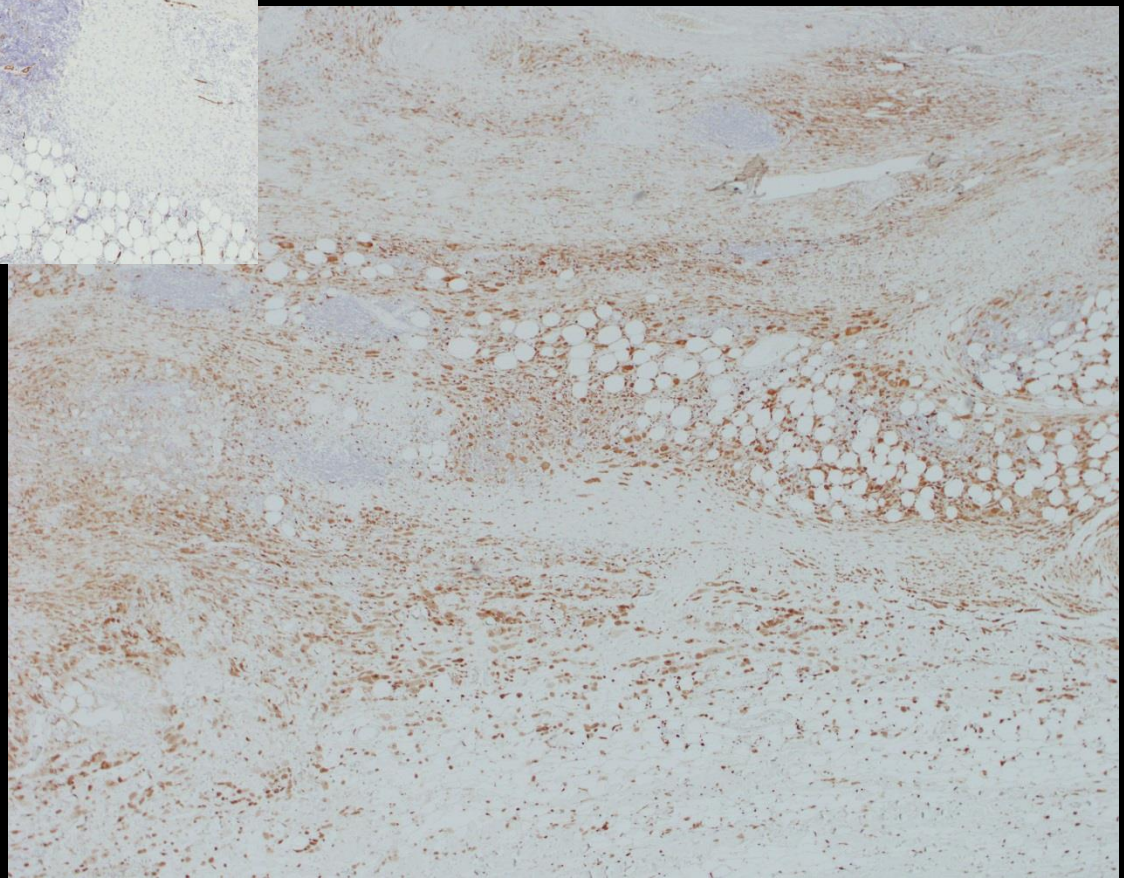
皮下脂肪組織の線維化巣



CD34

皮下脂肪組織の線維化巣

Factor XIIIa



鑑別診断

Cutaneous spindle cell tumors

- Cellular fibrous histiocytoma (CFH)
- Dermatofibrosarcoma protuberans
- Atypical fibrous histiocytoma
- Atypical fibroxanthoma
- Inflammatory myofibroblastic tumor
- low grade MPNST
- Perineurioma
- Fibrosarcoma

コンサルテーション結果

九州大学大学院　医学部研究科
形態機能病理　　小田　義直教授

Shoulder, lt. (resection): Cellular fibrous histiocytoma, suggestive

We reviewed this case and considered as above diagnosis.

The tumor located in the dermis and subcutis shows a cellular proliferation of oval to spindle-shaped cells having bland to hyperchromatic nuclei, arranged in fascicle pattern, accompanied by fibro-collagenous stroma. Touton-type multinucleated giant cells are also observed at the periphery. Mitotic figure is rarely seen.

Immunohistochemically, the tumor cells are positive for EMA,GLUT-1, claudin-1, alpha-SMA(focal), CD34(focal), and Factor XIIIa, but negative for CD68, CD163, AE1/AE3, CK903, p40, desmin, S-100 protein and collagen Type IV. H3K27me3 immunoexpression is preserved.

The feature is suggestive of cellular fibrous histiocytoma rather than perineurioma. There is no evidence of high-grade malignancy.

We would appreciate consulting for us.

コンサルテーション結果

九州大学大学院　医学部研究科
形態機能病理　　小田　義直教授

診断:　Cellular fibrous histiocytoma, suggestive

- 腫瘍は　皮膚・皮下組織に存在。
- 著明な fascicle pattern
- 核分裂像は希にみられる
- 辺縁でTouton type multinucleated giant cells
- Fibro-collagenous stroma
- 免疫染色

  陽性: EMA, GLUT-1, claudin-1, α-SMA, CD34f, Factor XIIIa

  陰性: CD68, CD163, S100, Desmin, AE1/AE3, CK903, p40, collagen typeⅣ, H3K27me3
- Perineurioma よりはCellular FHを疑う
- High grade malignancy ではない

病理診断

Left Shoulder, resection:

Cellular fibrous histiocytoma

Cellular fibrous histiocytoma

通常のdermatofibroma に比べて

- Increased cellularity and size
- Plump spindle cell のmonomorphic growth
- More fasicular pattern が目立つ
- 炎症細胞や多核巨細胞が少ない
- 皮下脂肪組織への進展が多い(30%)
- 核分裂像が多い(3/10HPFs)
- 中心部の壊死巣（10%）
- 局所再発が高い(25%)
- 稀に転移する

soft tissue tumorより