



Letný bioptický seminár SD IAP

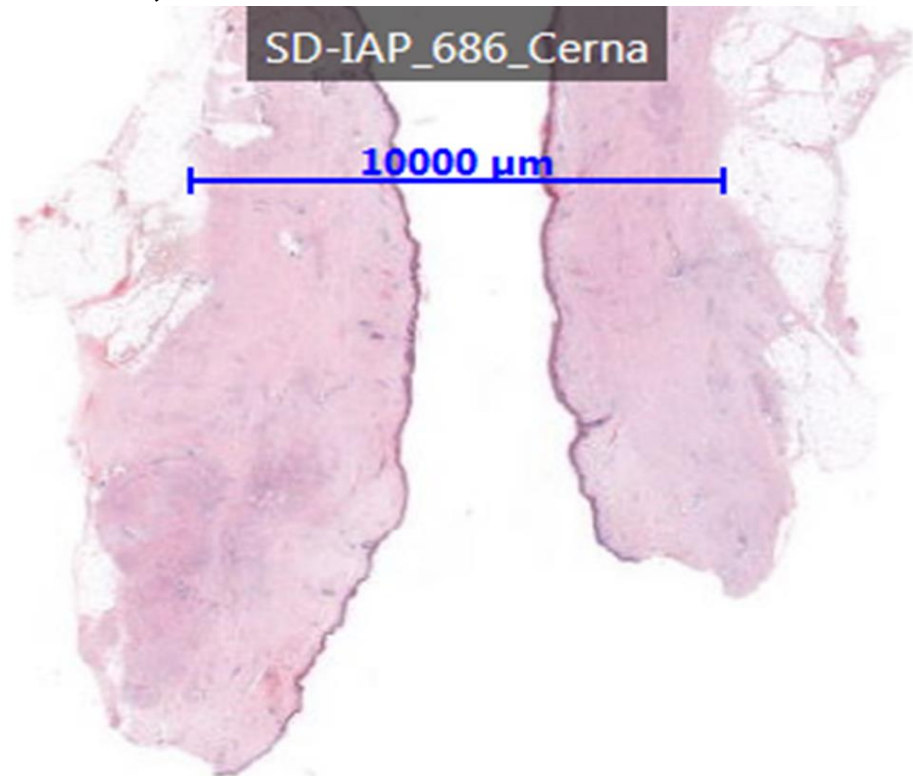
25.-26, mája 2018, Senec

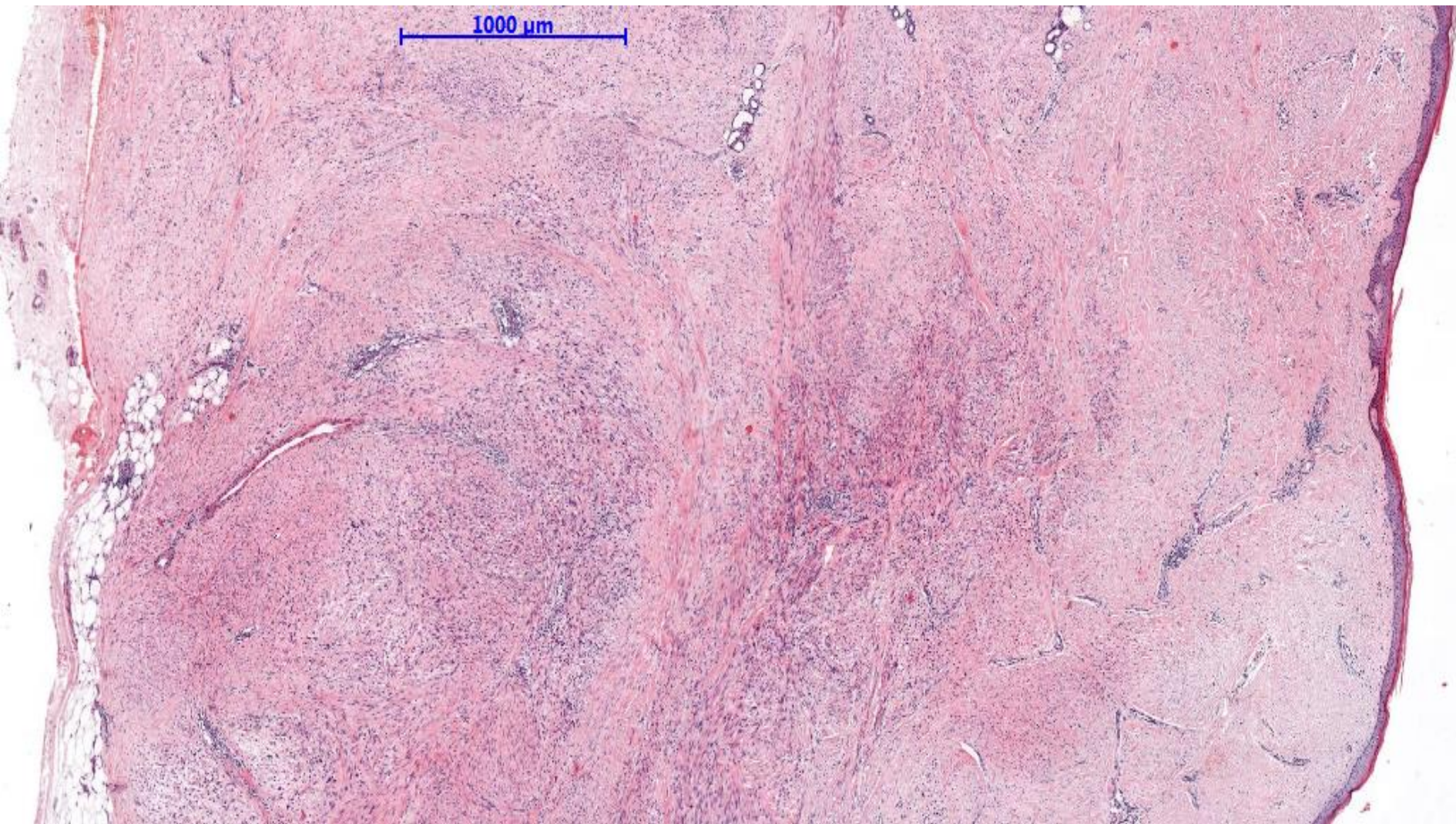
Prípad č. 686

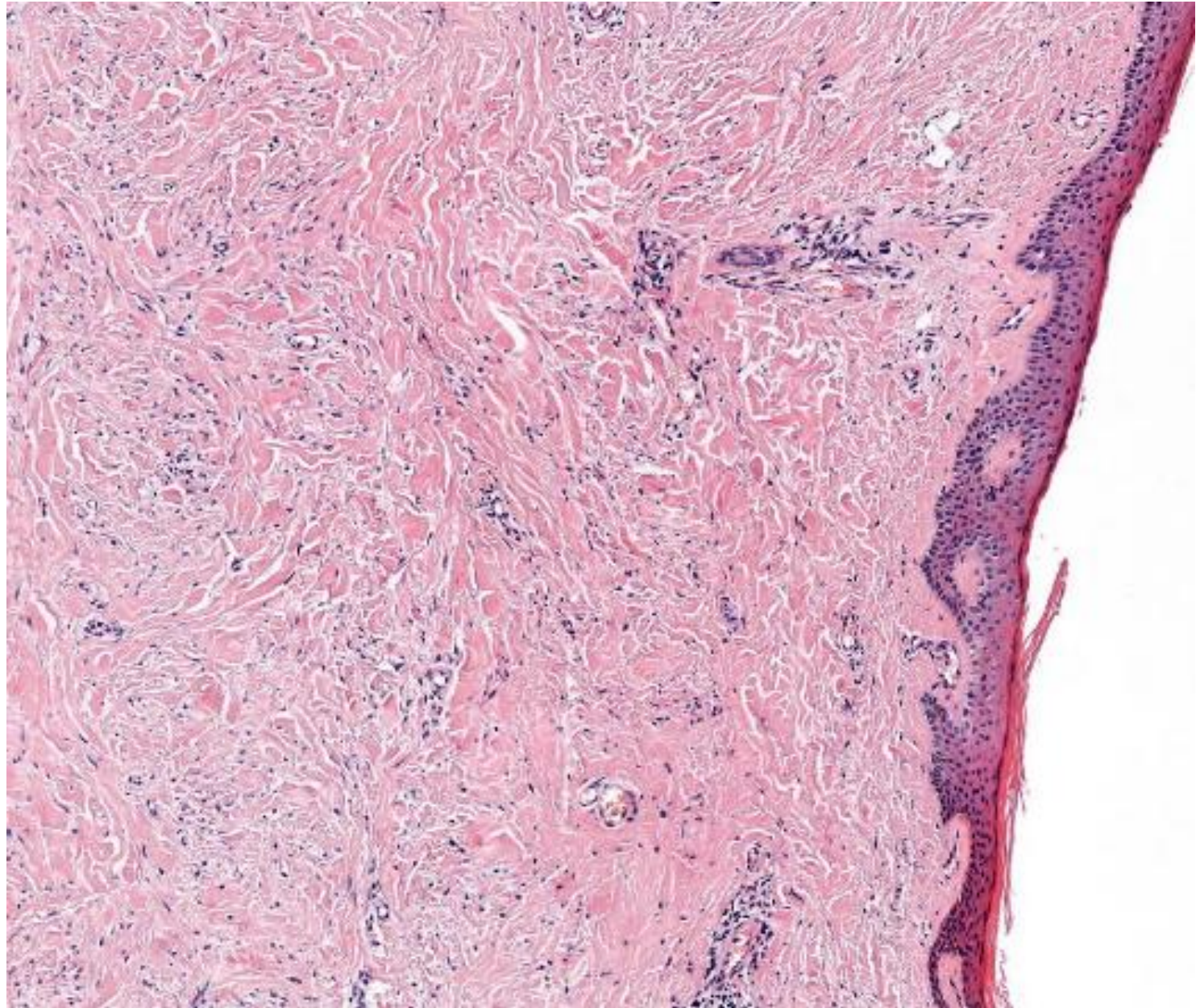
Andrea Černá

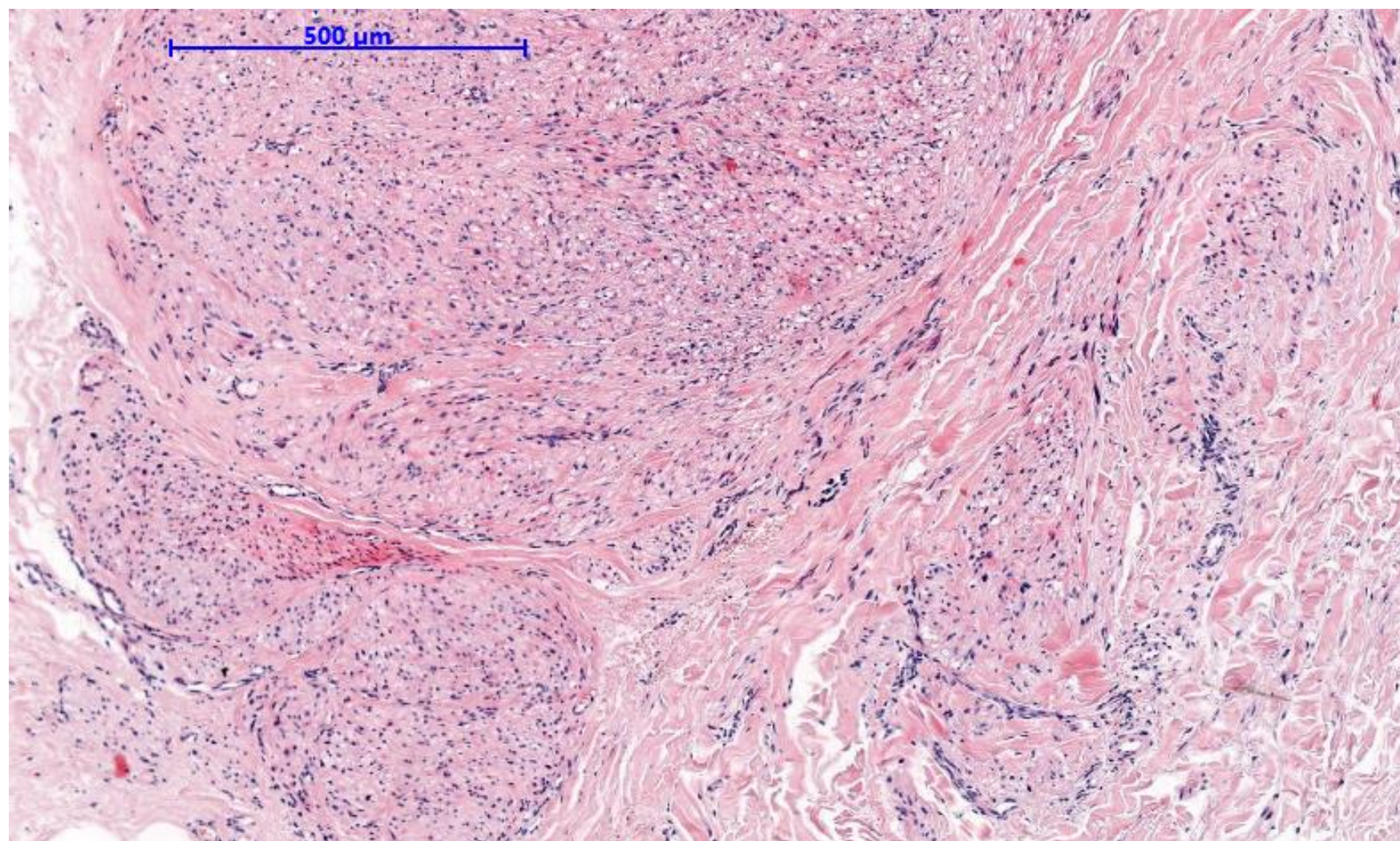
Klinické údaje

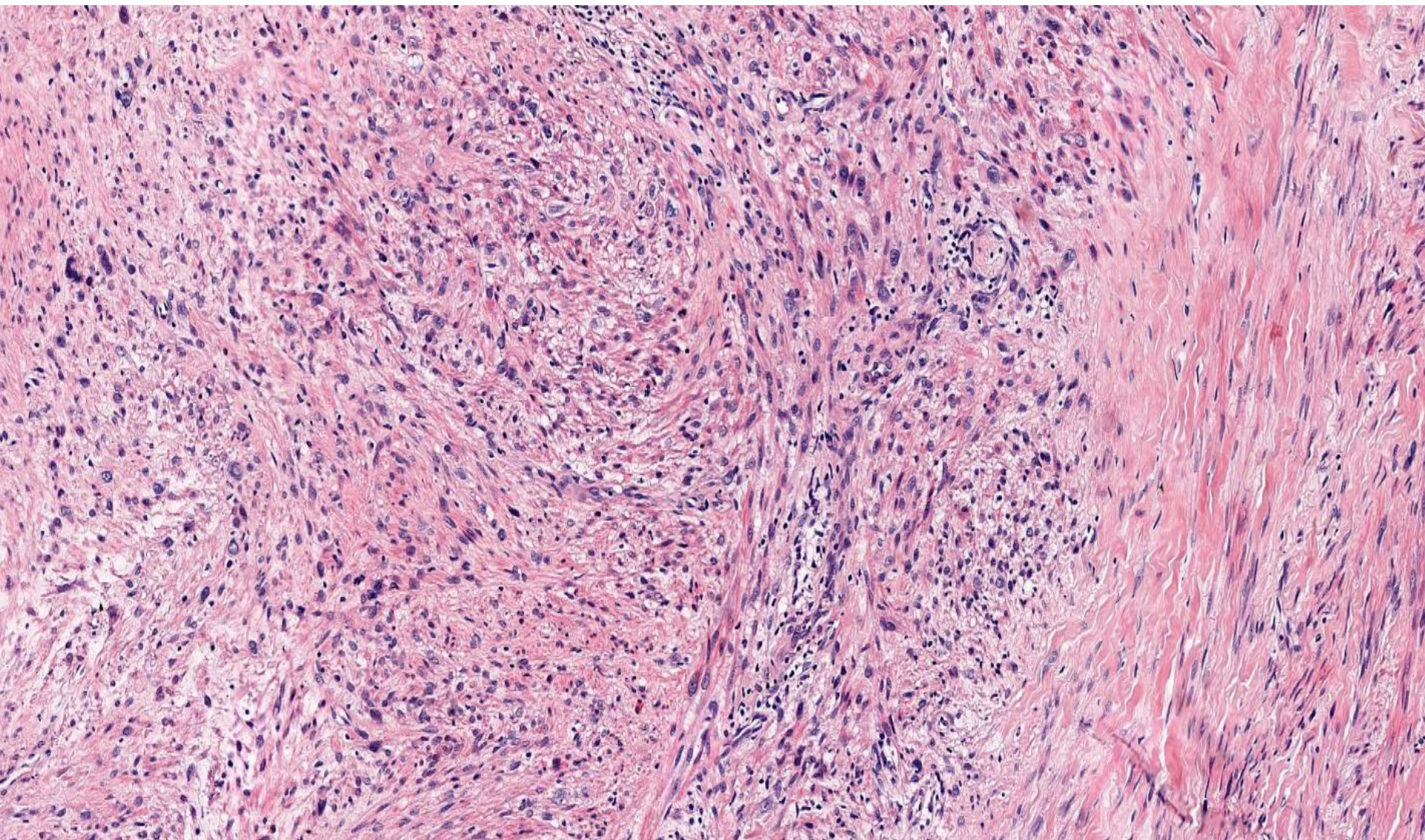
- 48 ročná žena
- bolestivá rezistencia pod pravým kolenom, posledný pol rok pozoruje zväčšovanie a bolestivosť pri pohybe
- excízia kože a podkožia 18 x 10 mm,
- kl.dg.: TU cutis

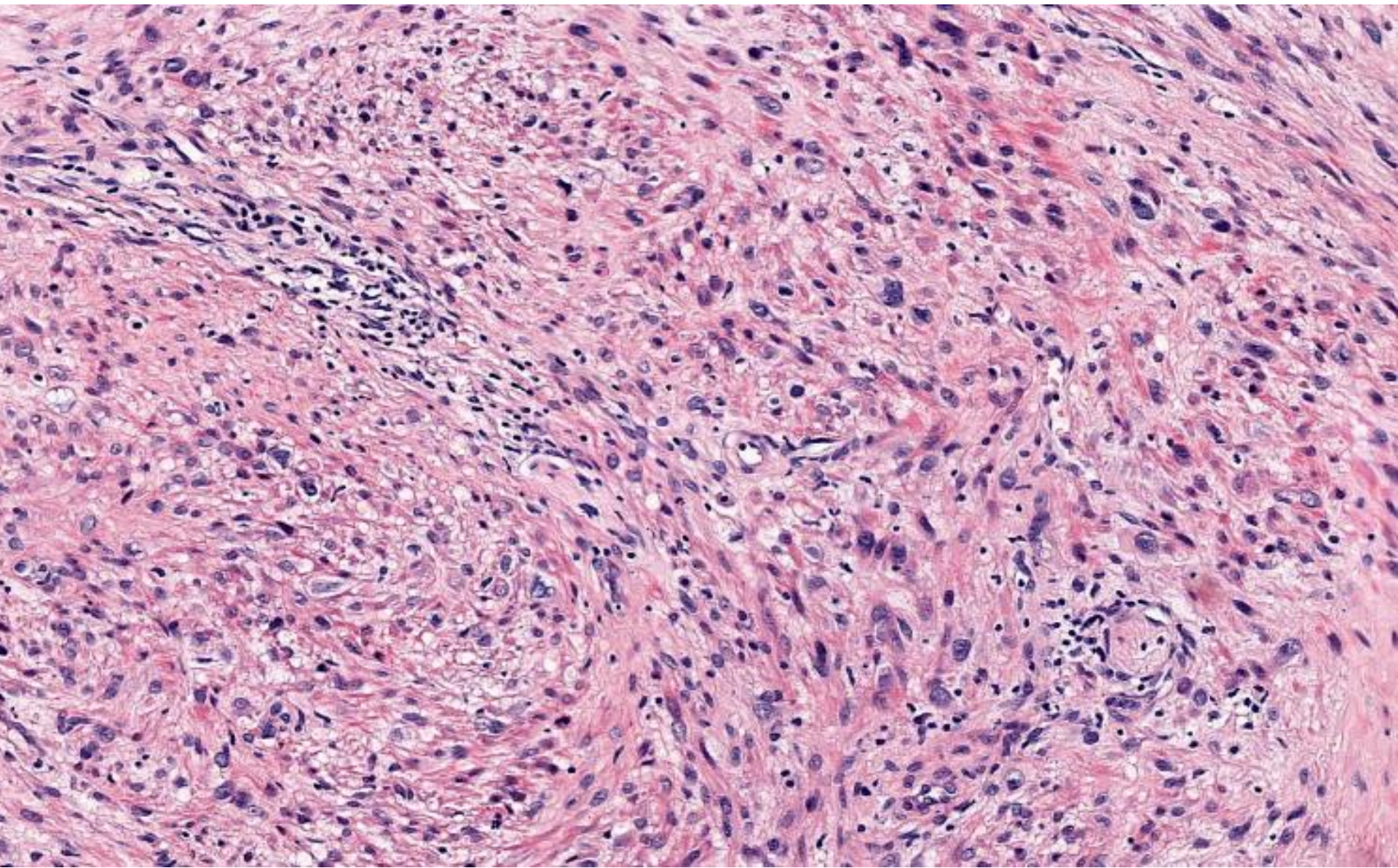


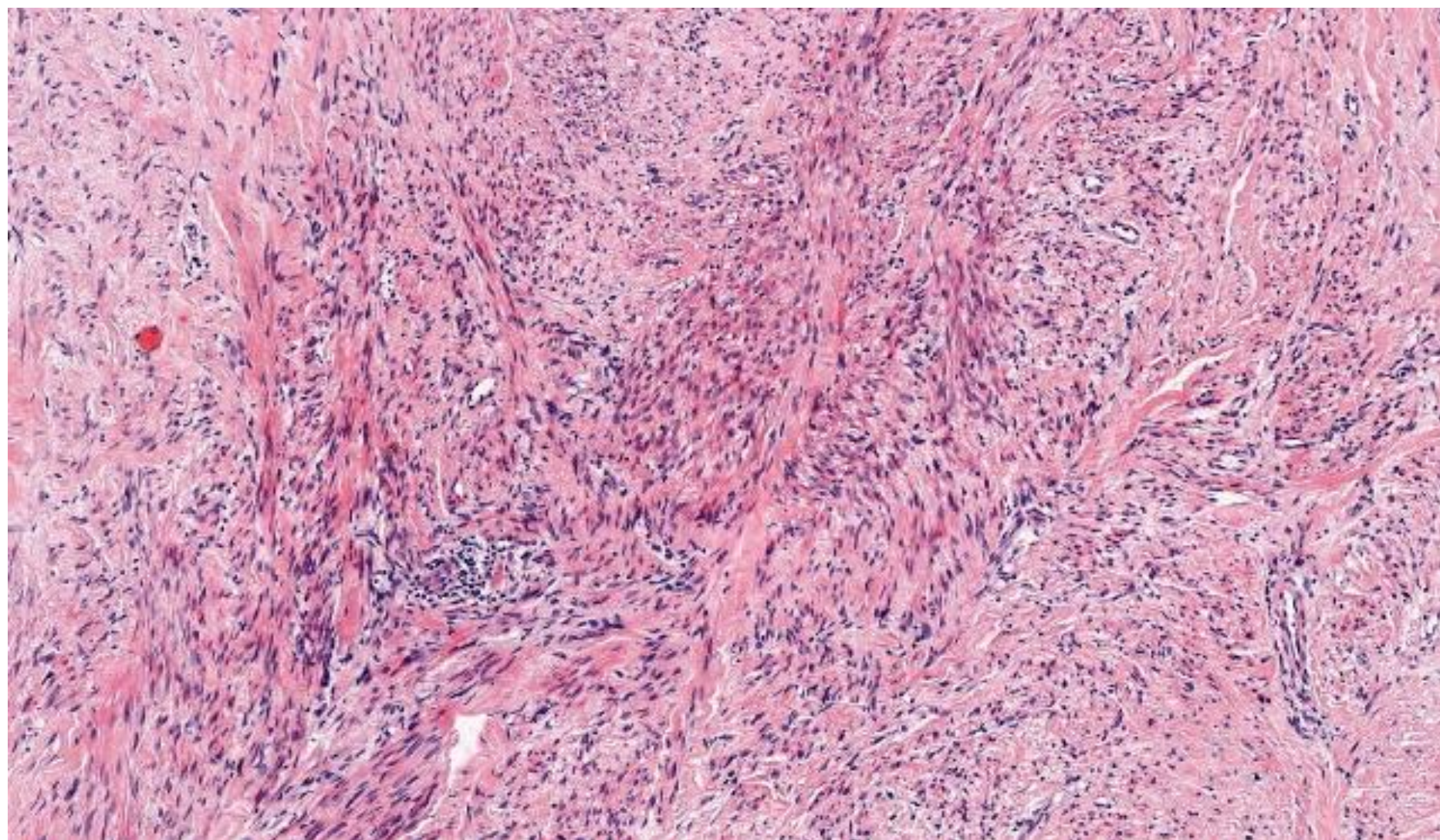


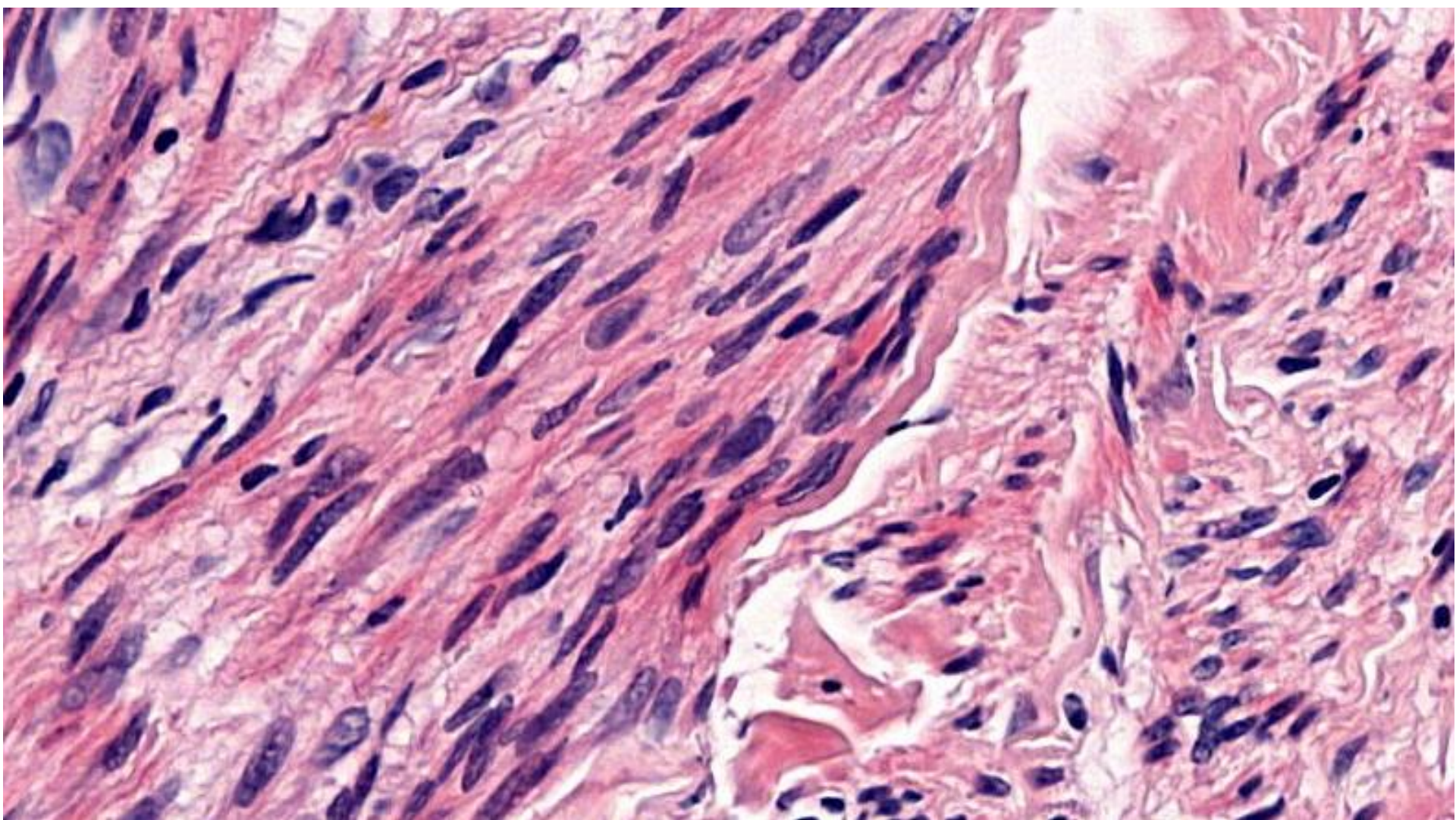


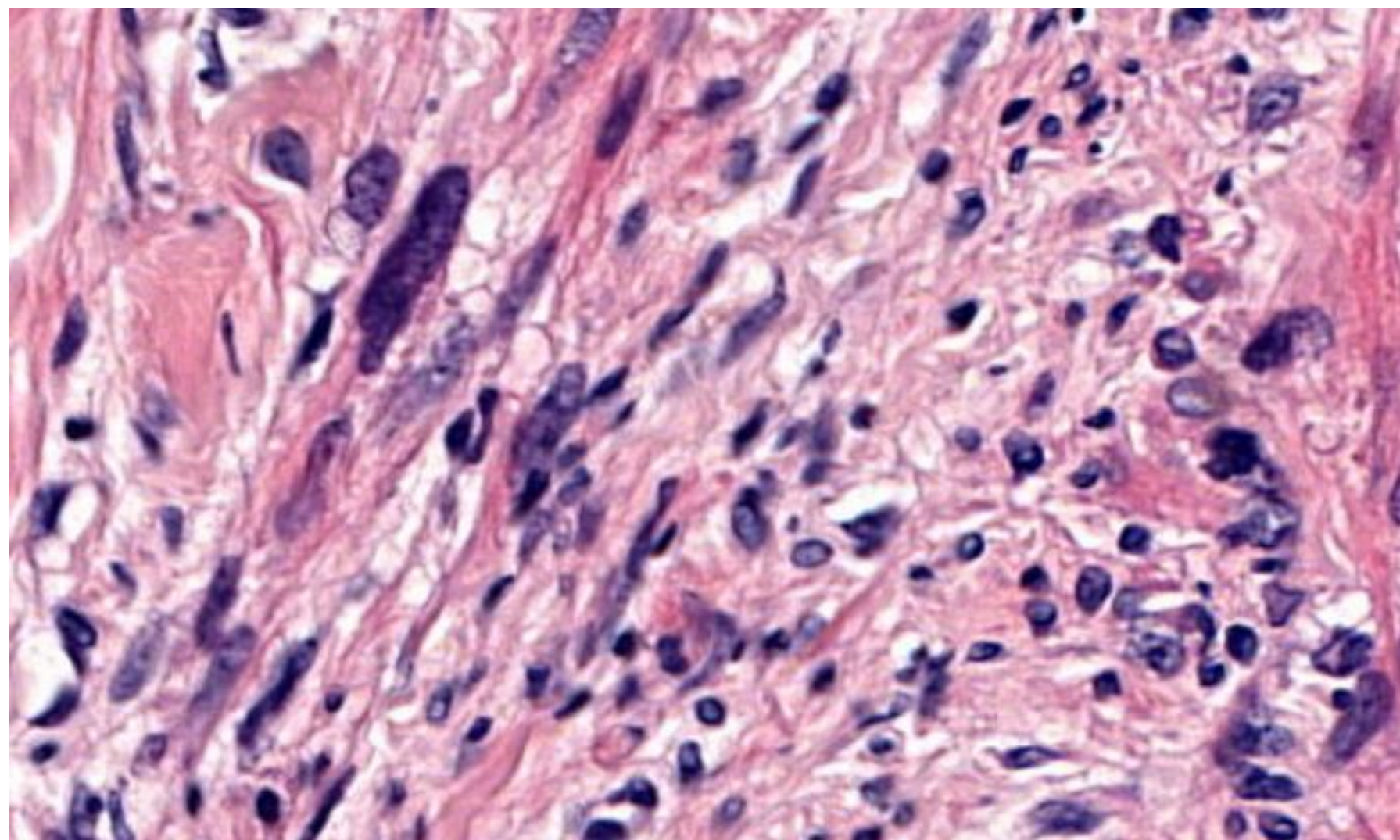


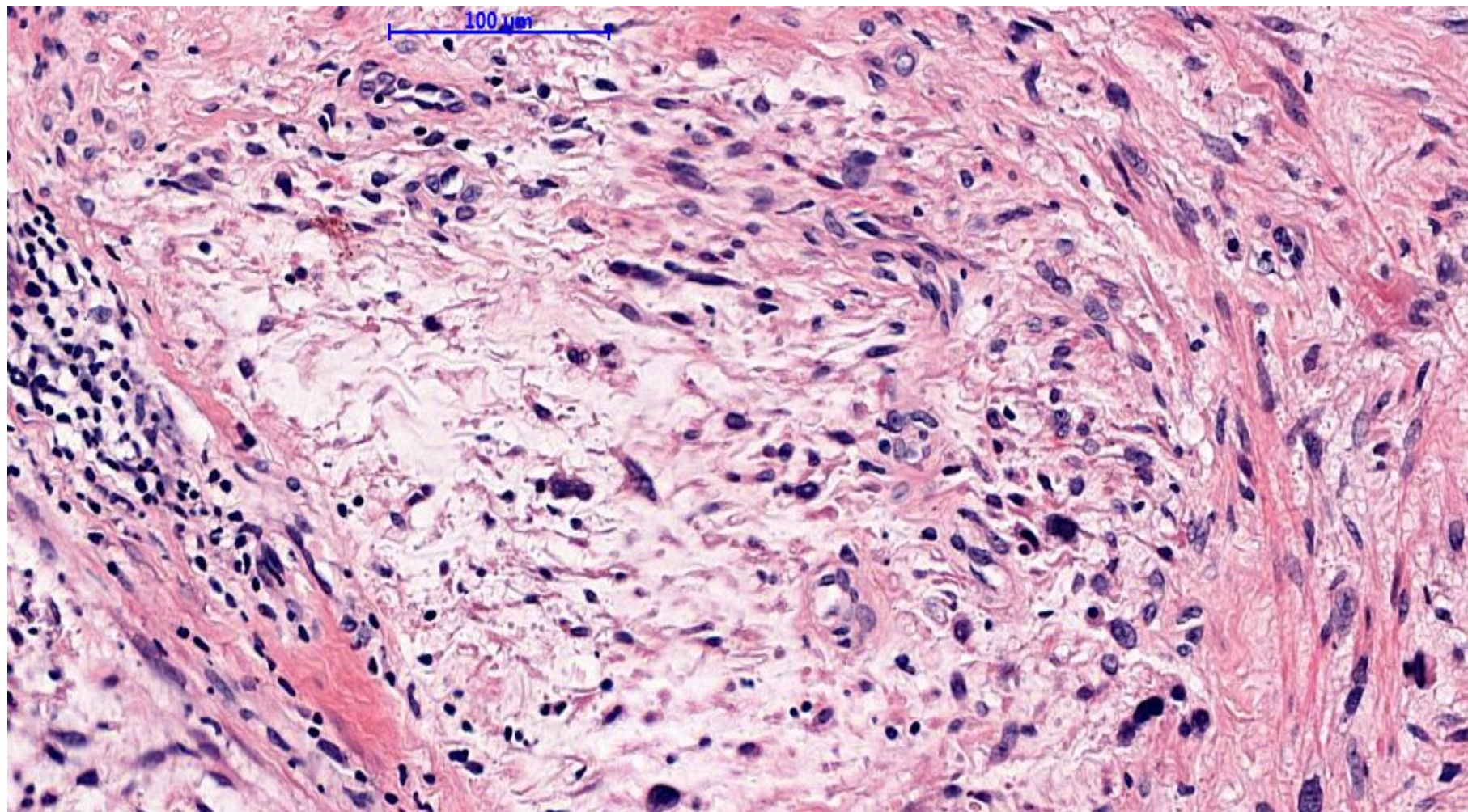


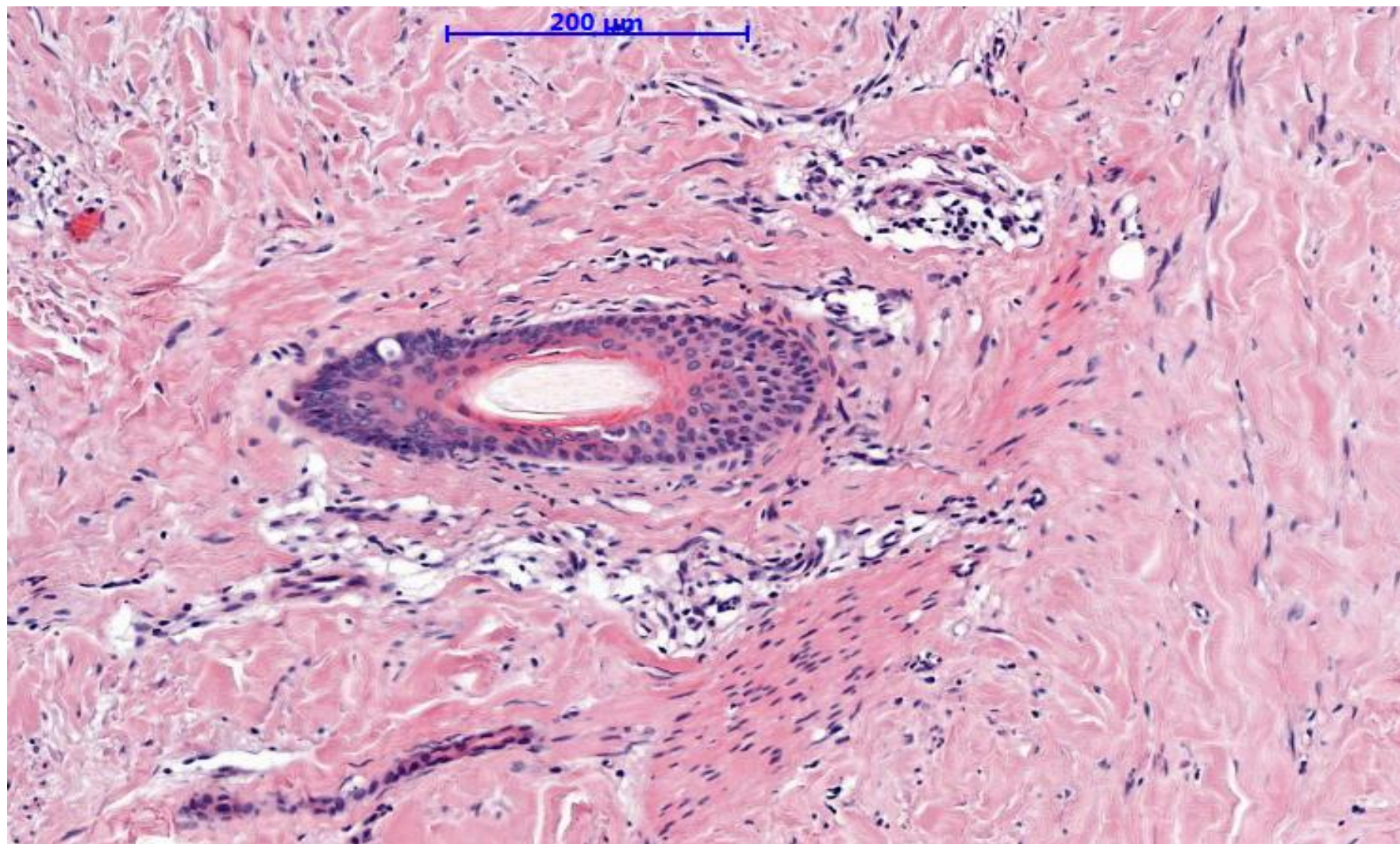


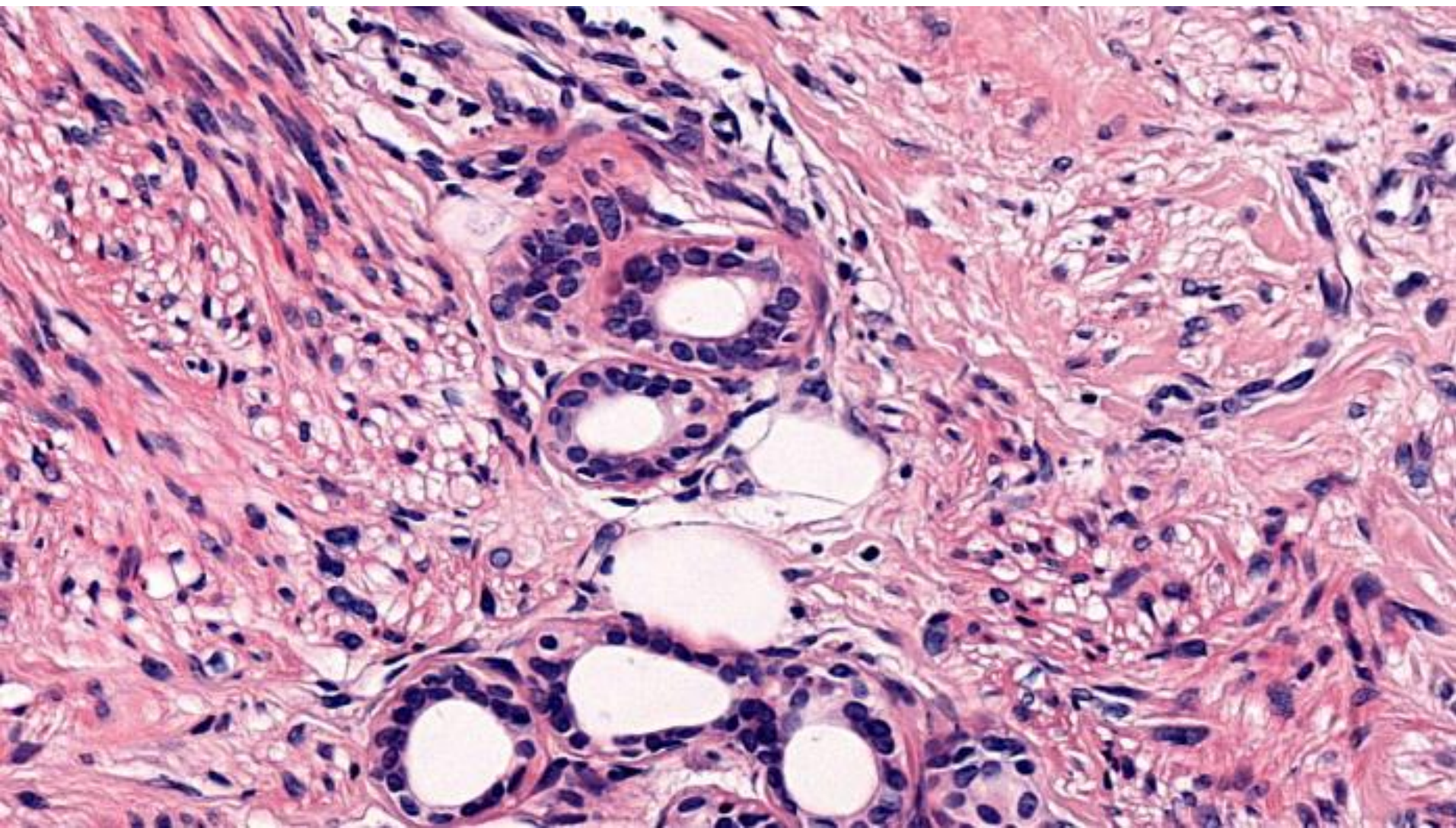


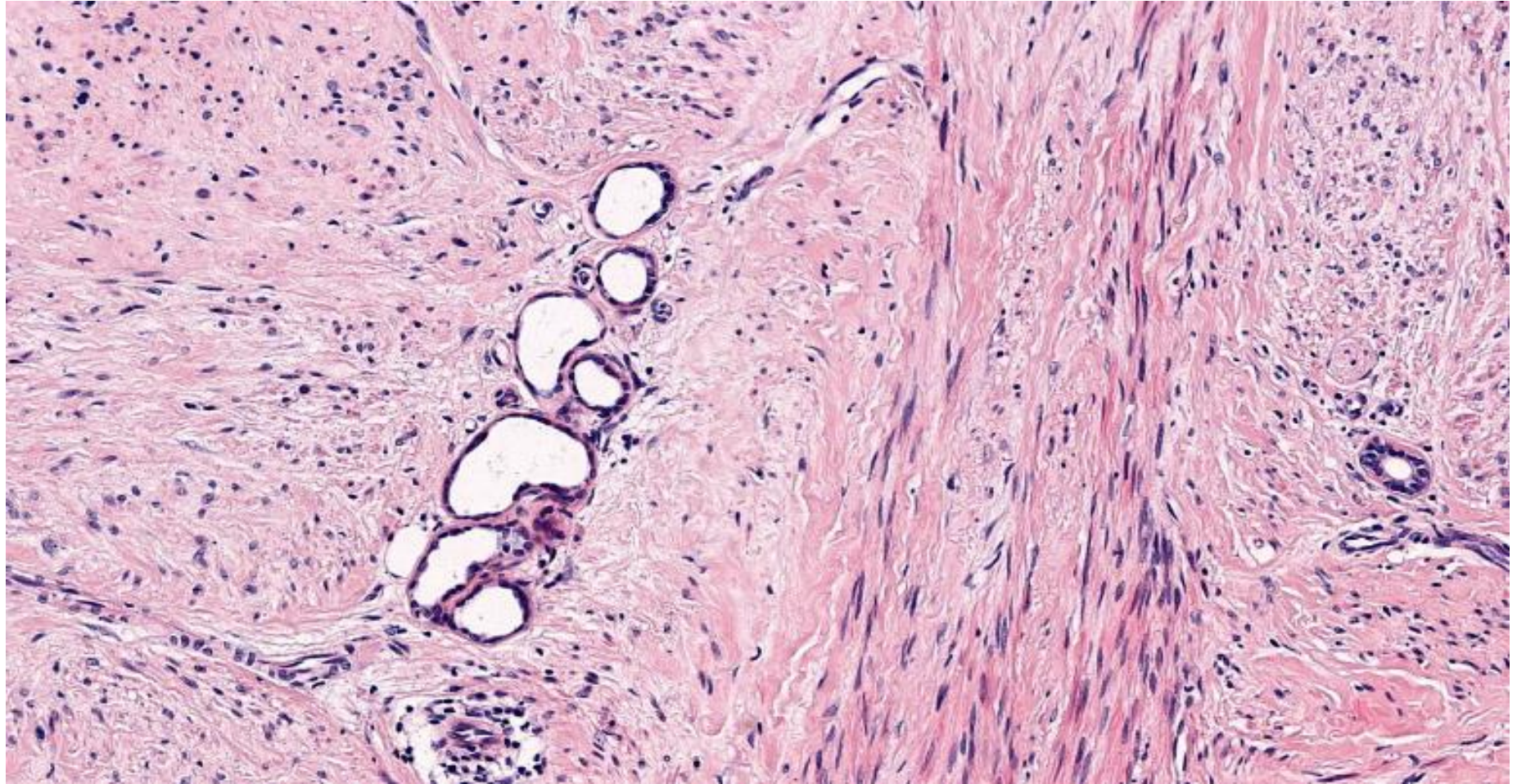


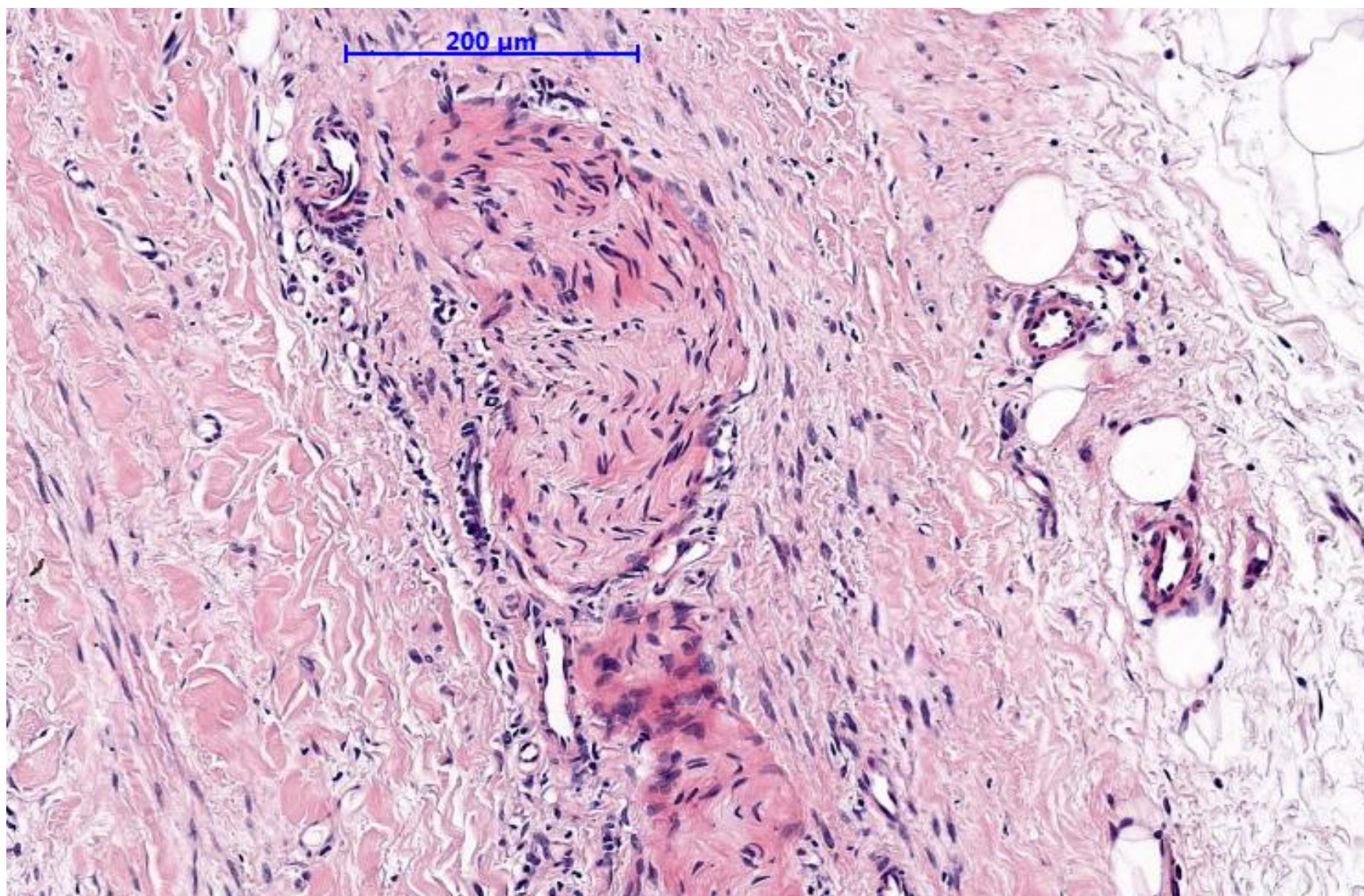


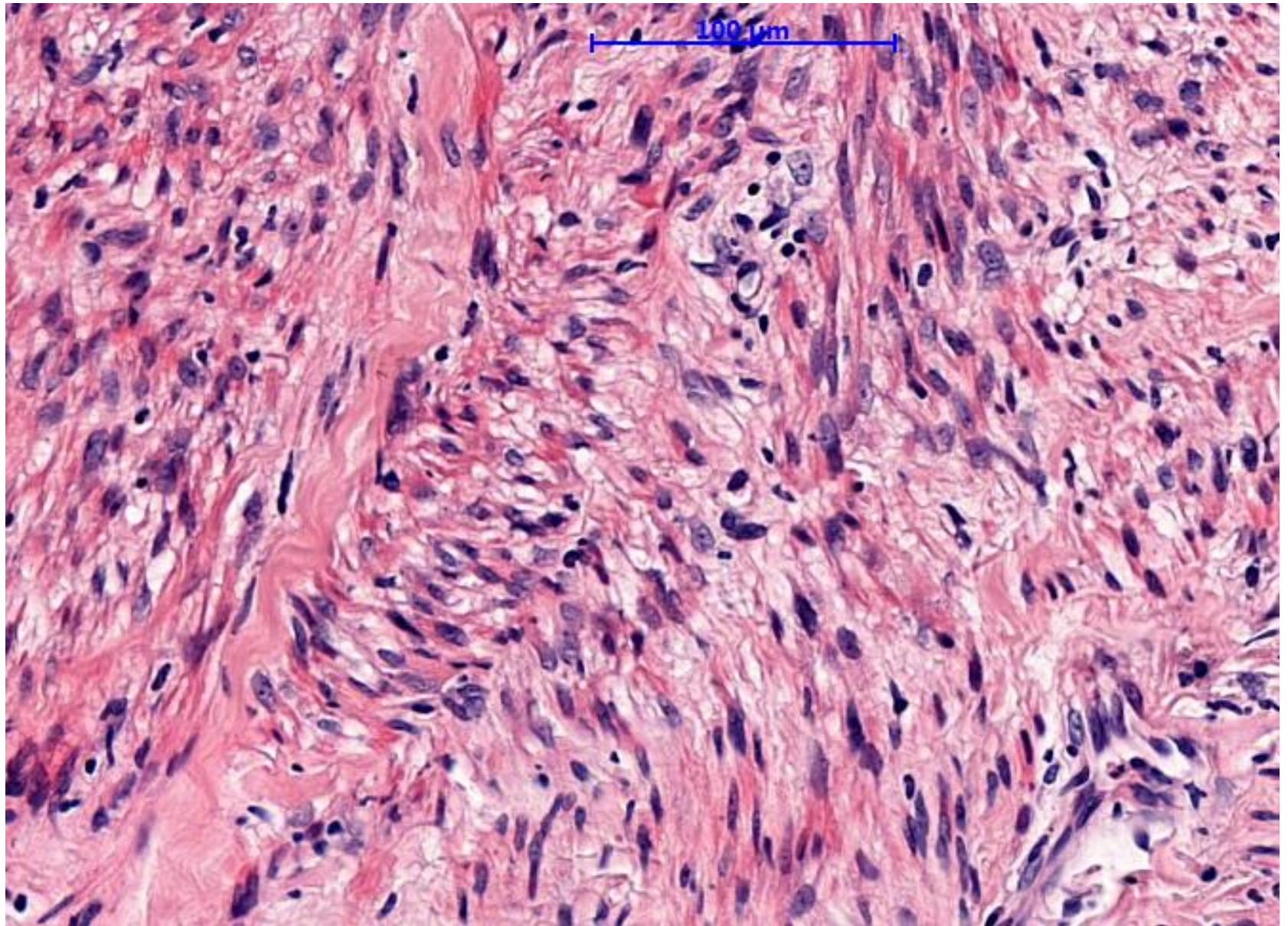


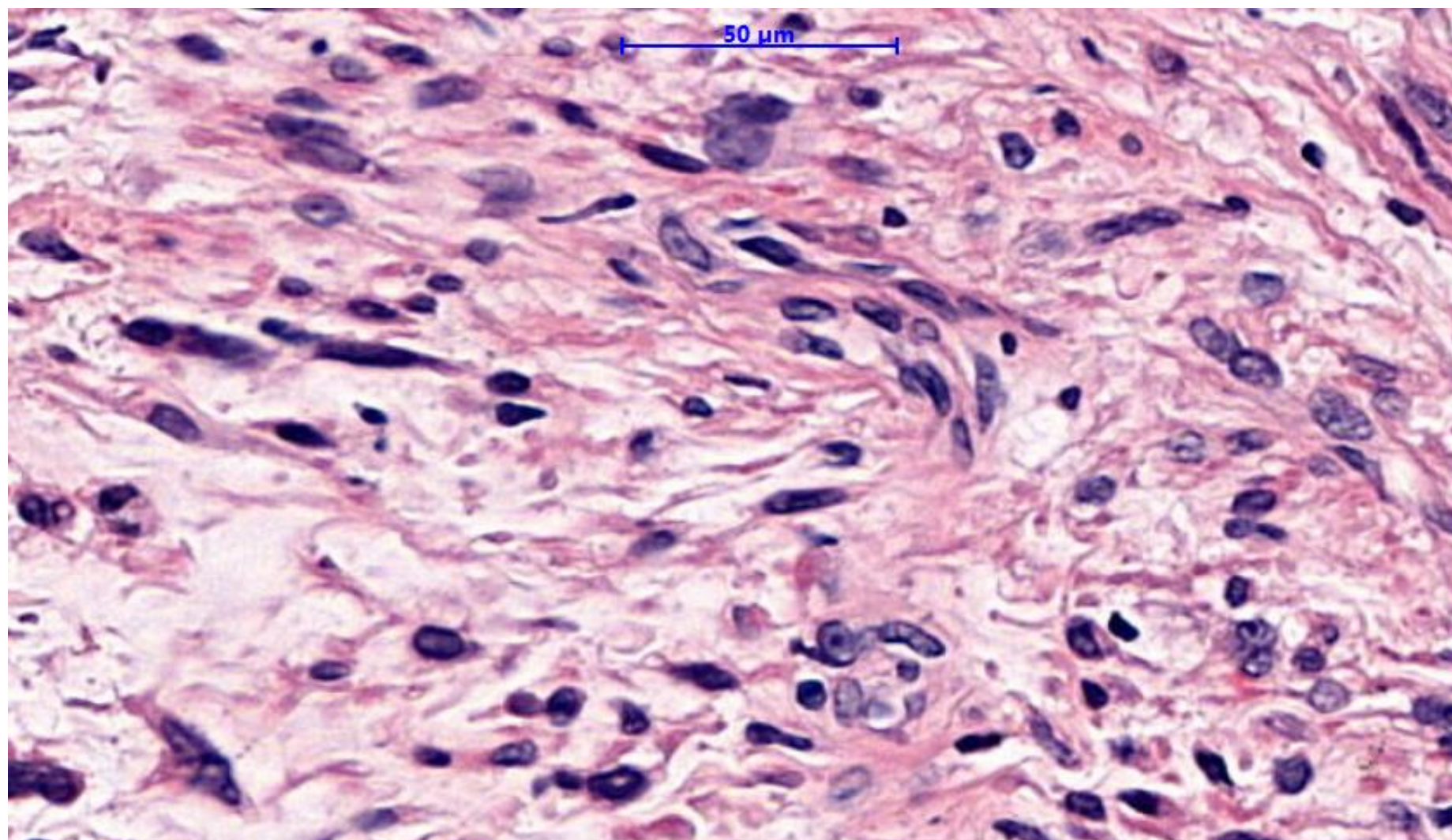


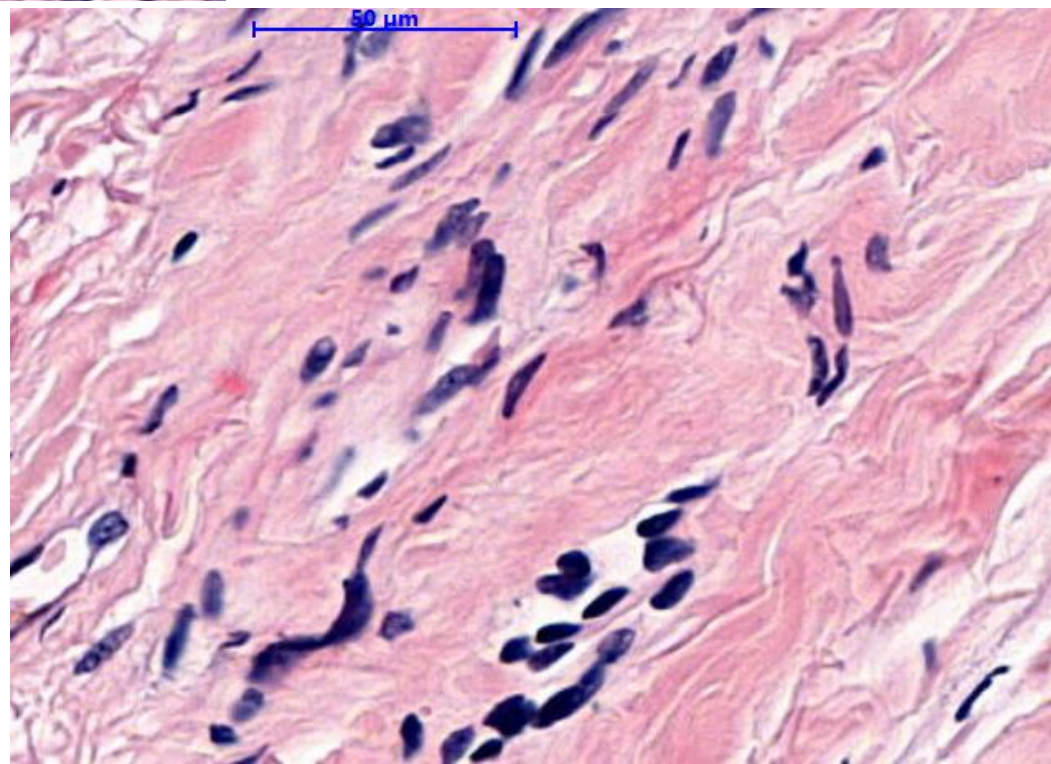
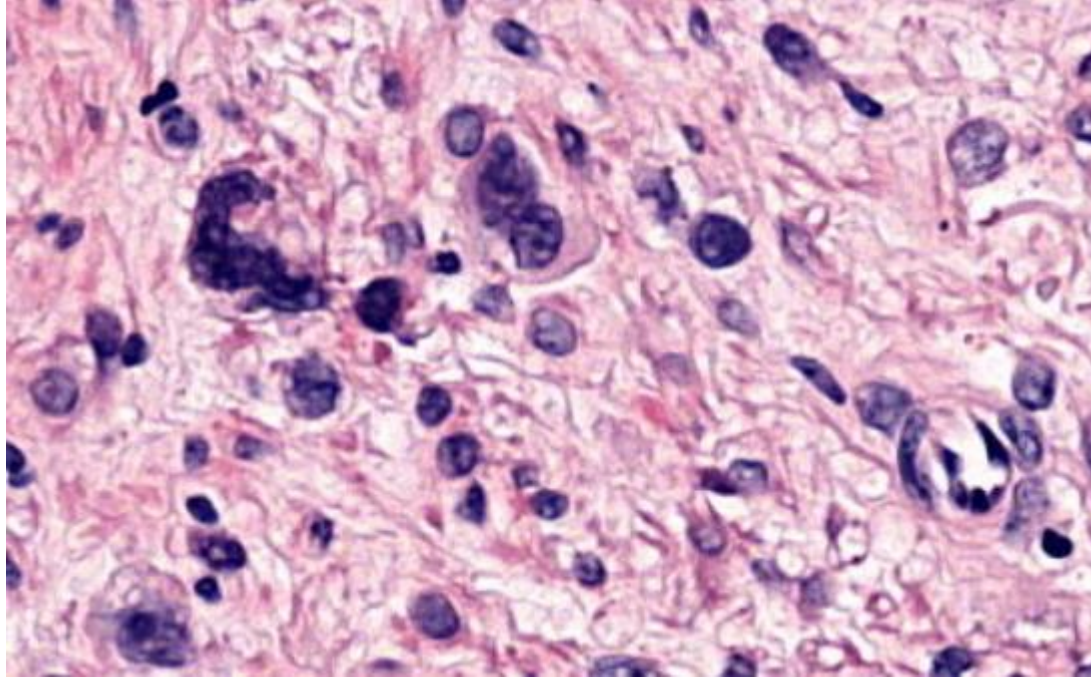


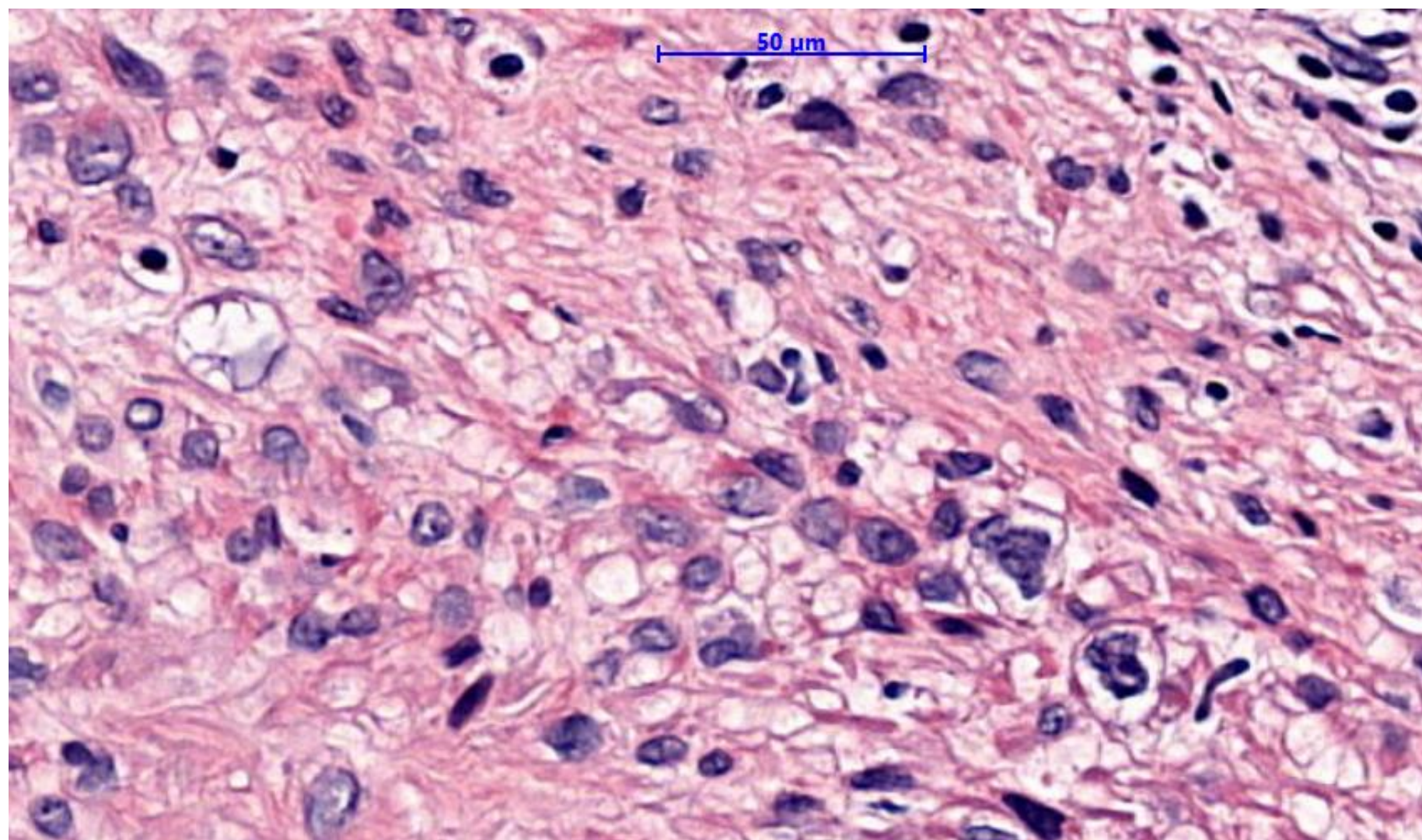


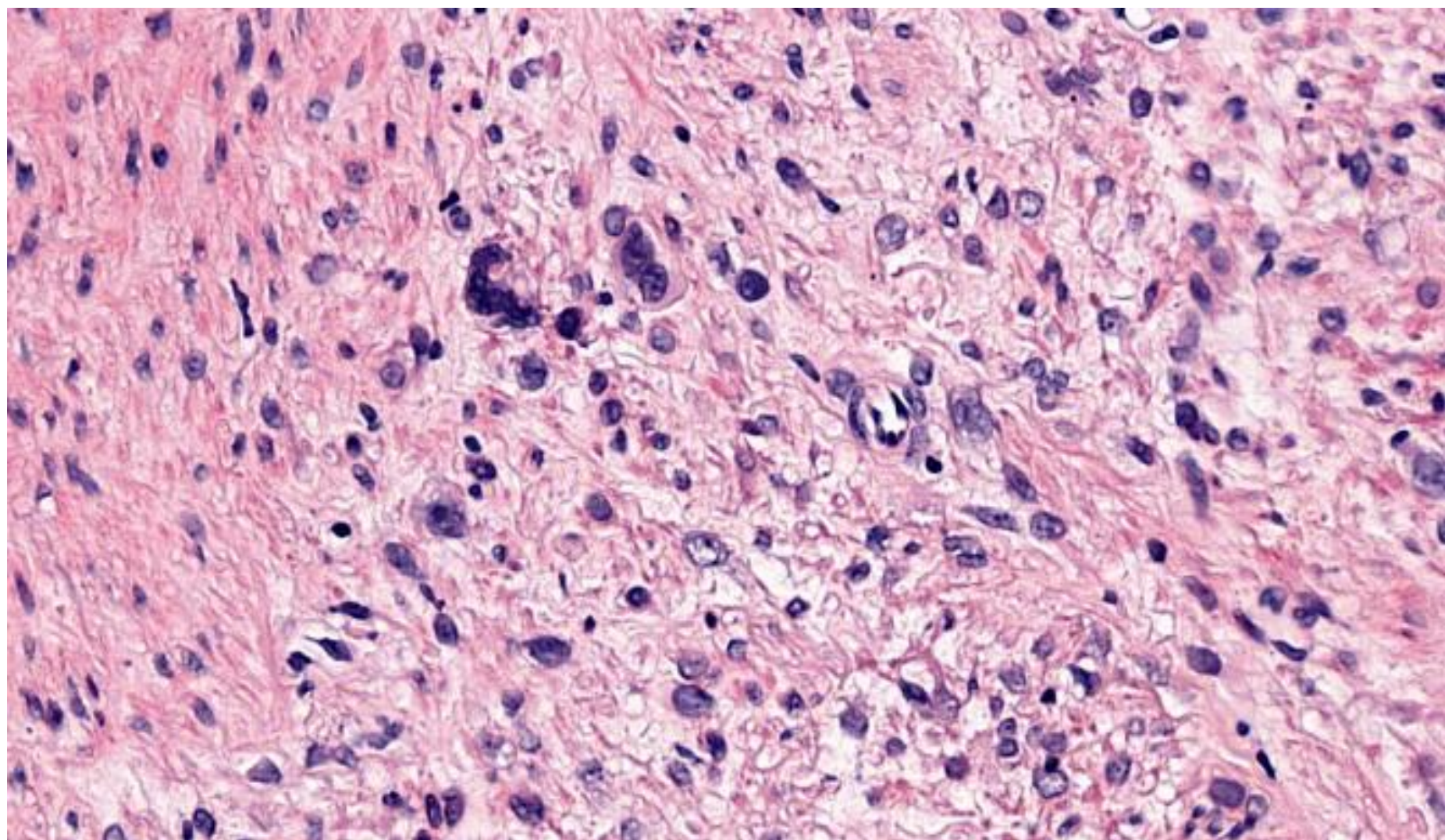














IHCH

Pozitívne: desmin,
SMA
h-caldesmon
HHF-35
p53 (100% jadier)
ki67 do 30%

Negatívne: S100
CD68
beta-cathenin
SOX-10
cdk-4
EMA
CD 34
CKAE1/3

Naša diagnóza

**Atypická intradermálna hladkosvalová neoplázia
(kožný leiomyosarkóm)**



Atypická intradermálna hladkosvalová neoplázia

- zriedkavá lézia, predtým nazývaná ako kožný leiomyosarkóm
- vychádza z dermy, až sekundárna invázia do podkožia
- pôvod musculus arrector pilii (v skrúte tunica dartos) , stena cievy
- výskyt muži: ženy 4,3:1,
- priemerný vek 56 rokov
- dolné končatiny, trup, menej často hlava a krk
- zmeny povrchu kože – hyperplázia epidermy, ulcerácia, strata adnex
- častá bolestivosť postihnutej oblasti
- low risk agresívneho správania
- veľkosť od 0,3-5 cm
- nemetastazujú, pokiaľ sú obmedzené na dermu
- excelentná prognóza
- kompletne chirurgické odstránenie
- bez regresívnych a degeneratívnych zmien
- veľmi zriedka hemorágie, nekrózy alebo myxoidné zmeny
- -80% tumorov 2 a viac mitóz /10 HPF

Am J Surg Pathol. 2011 Apr;35(4):599-607. doi: 10.1097/PAS.0b013e31820e6093.

Atypical intradermal smooth muscle neoplasms: clinicopathologic analysis of 84 cases and a reappraisal of cutaneous "leiomyosarcoma".

Kraft S¹, Fletcher CD.

 **Author information**

Ann Diagn Pathol. 2016 Oct;24:25-9. doi: 10.1016/j.anndiagpath.2016.05.008. Epub 2016 Aug 16.

Immunoexpression of p53 in cutaneous and subcutaneous leiomyosarcomas.

Fernandez-Flores A¹, Monteaquedo C².

 **Author information**

Actas Dermosifiliogr. 2018 Mar;109(2):140-147. doi: 10.1016/j.ad.2017.08.005. Epub 2017 Nov 1.

Clinical and Histopathologic Findings of Cutaneous Leiomyosarcoma: Correlation with Prognosis in 12 Patients.

[Article in English, Spanish]

Rodríguez-Lomba E¹, Molina-López I², Parra-Blanco V³, Suárez-Fernández R², Pulido-Pérez A².

 **Author information**

Diferenciálna diagnostika: „spindle cell“ proliferácie dermy“

- leiomyóm
- celulárny schwanóm
- dezmoplastický malígny melanóm
- spindle cell forma skvamocelulárneho karcinómu
- dermatofibrosarcoma protuberans
- spindle cell atypický fibroxantóm
- nediferencovaný sarkóm (MFH)
- MPNST

Common Spindle Cell Malignant Neoplasms of the Skin: Differential Diagnosis and Review of the Literature

S Repertinger, B Teruya, D Sarma

Citation

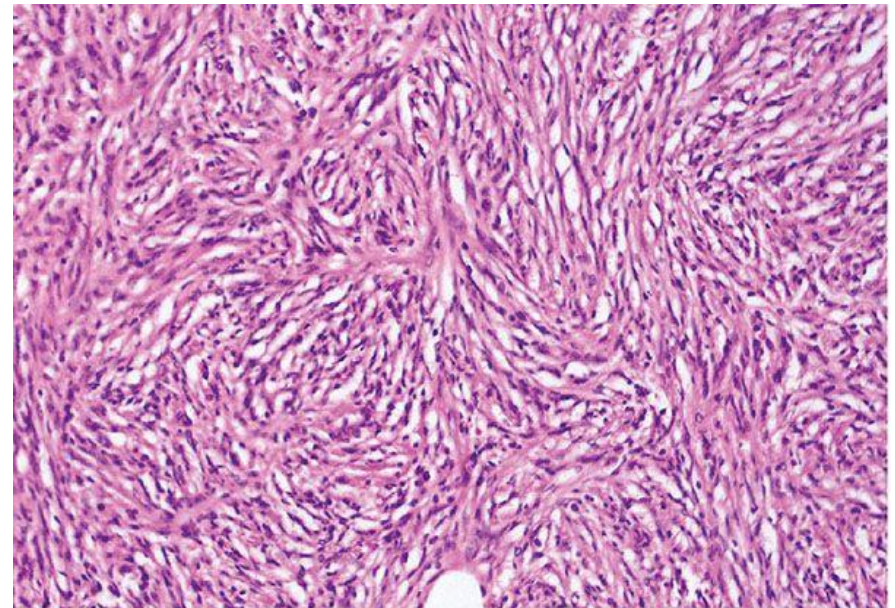
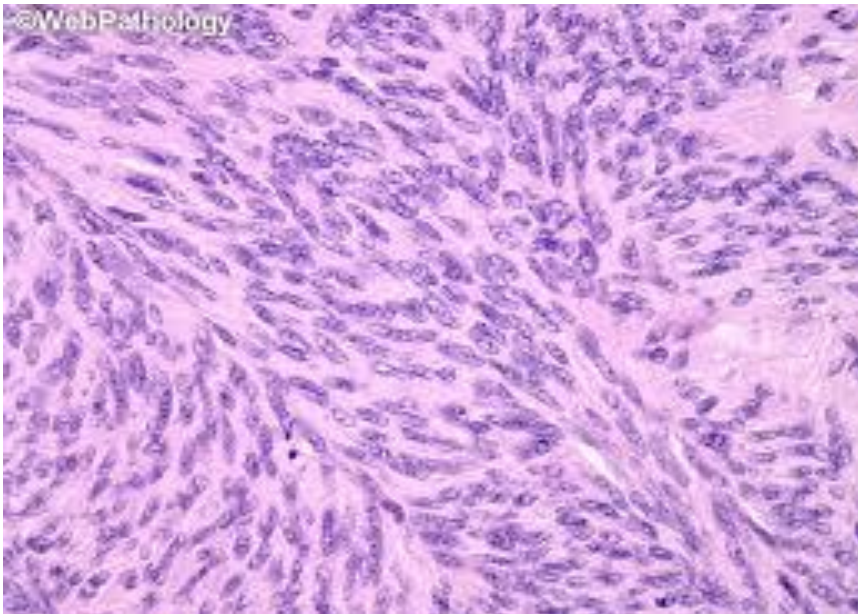
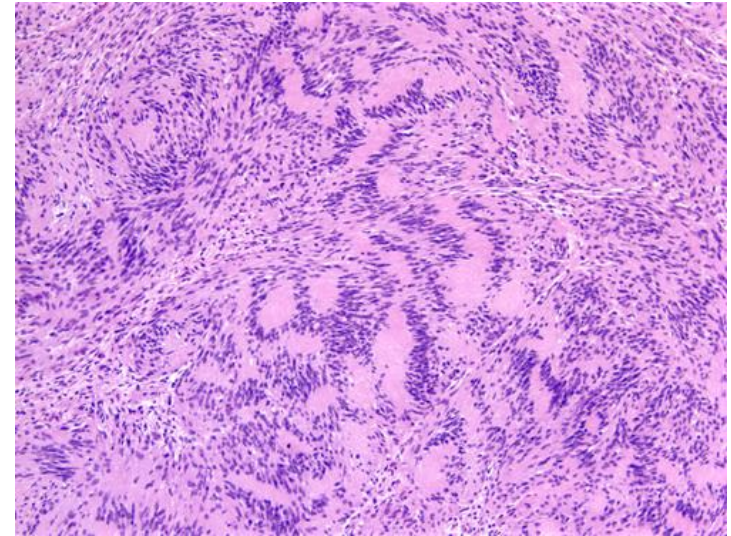
S Repertinger, B Teruya, D Sarma. *Common Spindle Cell Malignant Neoplasms of the Skin: Differential Diagnosis and Review of the Literature*. The Internet Journal of Dermatology. 2008 Volume 7 Number 2.

Abstract

Malignant and borderline-malignant neoplasms of the dermis can pose diagnostic challenges. Because these tumors can share similar clinical and histologic features, including a predominantly spindle cell morphology, the pathologist must be familiar with these entities in order to facilitate accurate diagnosis, as treatment for these tumors may be different. We review several of these lesions with respect to clinical and histologic features: desmoplastic melanoma, spindle cell carcinoma, spindle cell atypical fibroxanthoma, dermatofibrosarcoma protuberans, and cutaneous leiomyosarcoma.

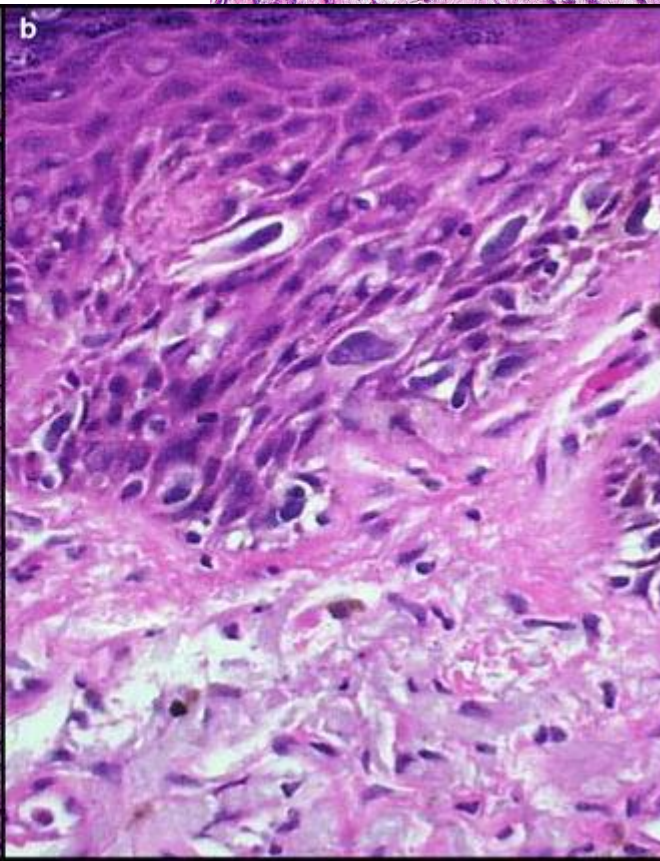
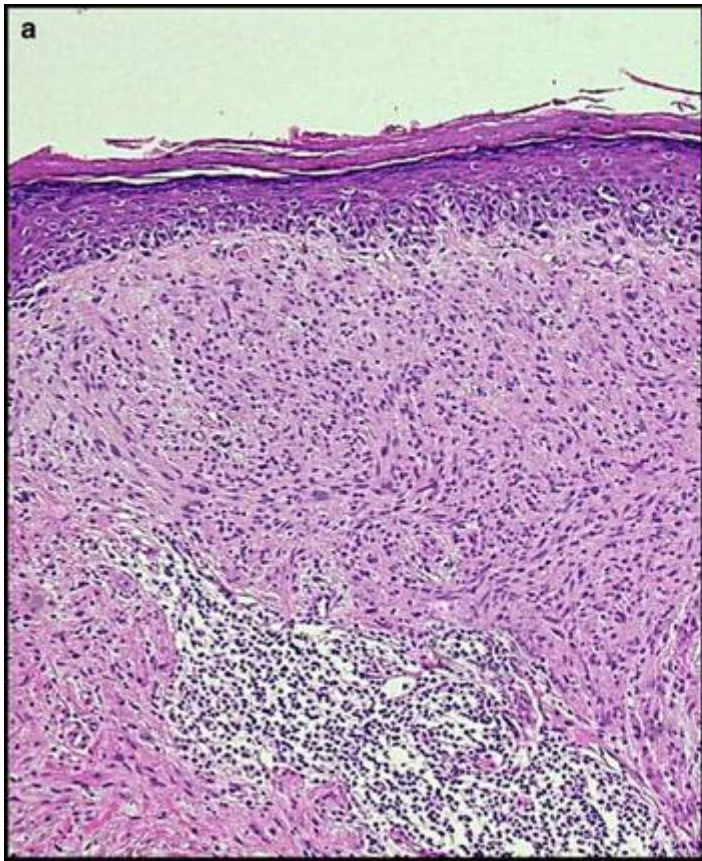
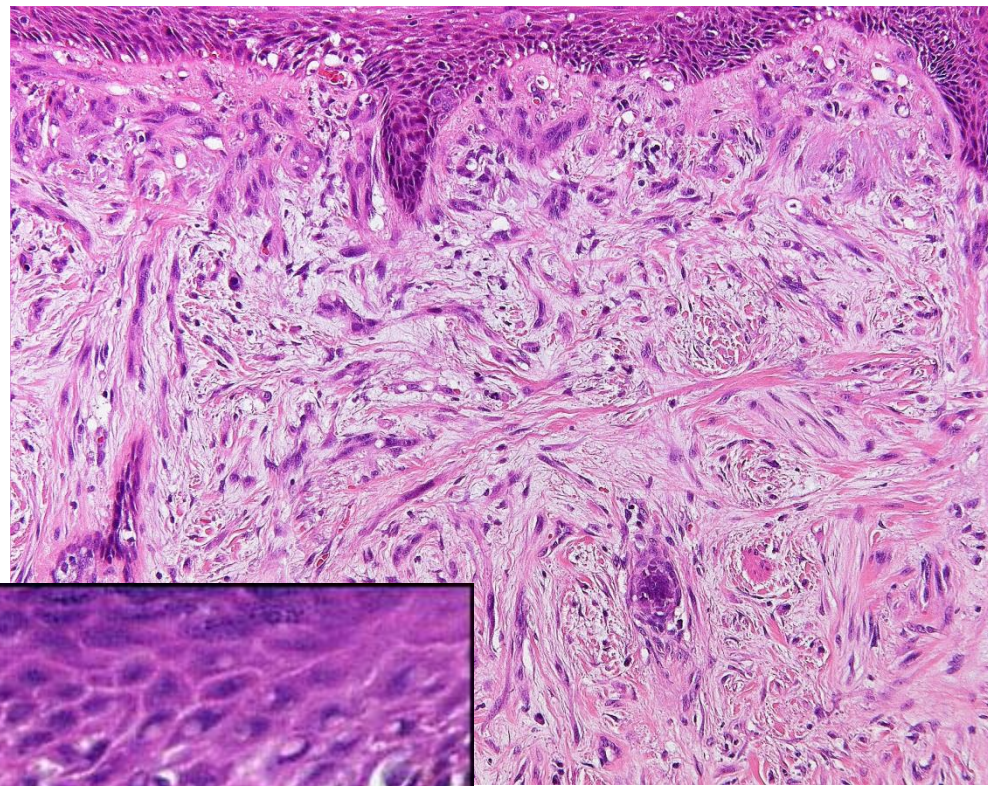
Celulárny schwanóm

- enkapsulovaný
- vysoko celulárny
- fascikulárna proliferácia je dobre vyznačená
- na periférii lymfoidné agregáty
- intratumorálne penovité makrofágy
- S100+, desmin -, actin -



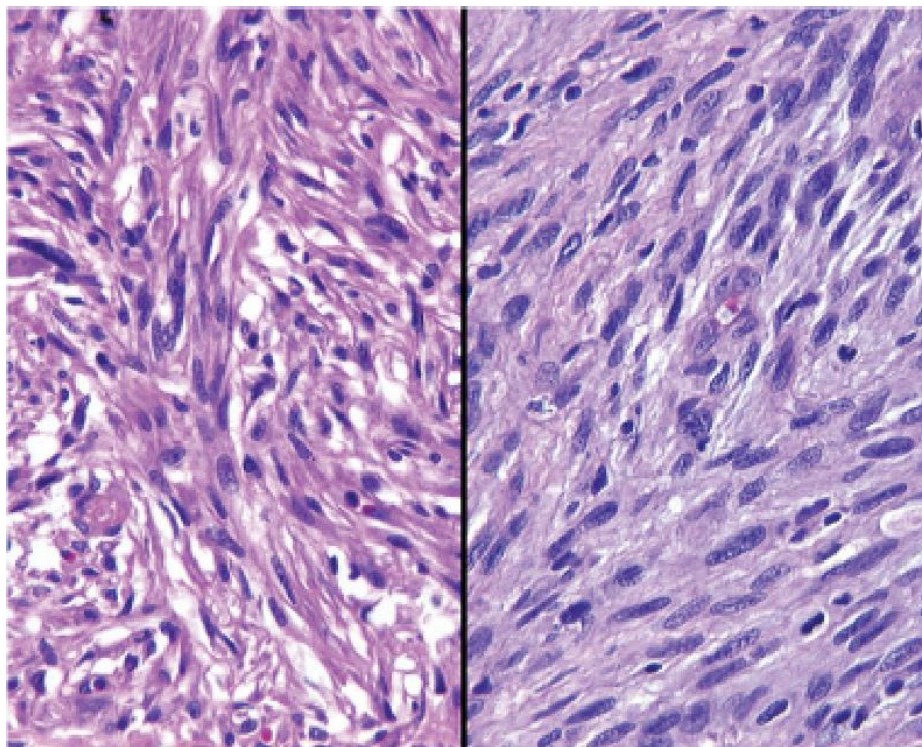
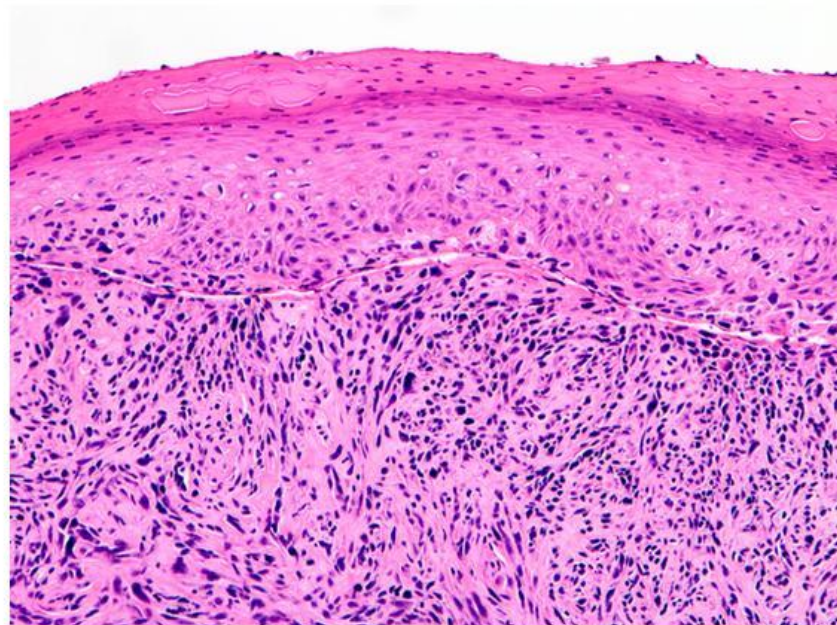
Dezmoplastický malígny melanóm

- S100 +, HMB45 -, Melan A –
- bez pigmentu
- slnkom exponovaná koža
- -neurotropizmus
- stromálna fibróza
- fokálne SMA + !!!

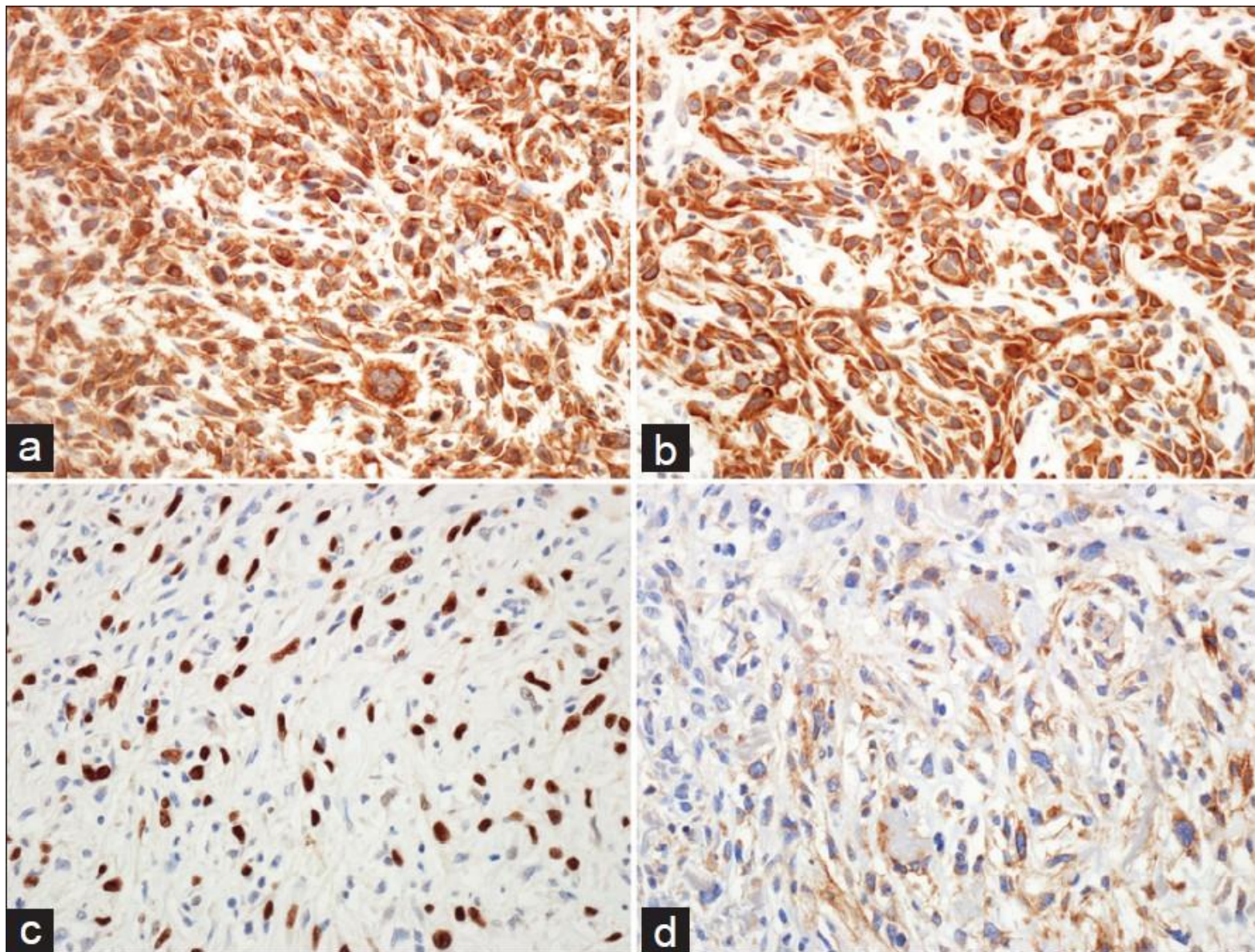


„Spindle cell“ skvamózný karcinóm kože

- slnkom exponovaná koža
- častá ulcerácia a krusta na povrchu
- po rádioterapii alebo popáleninách
- P63+, CK5/6+
- možná vimentín +, SMA +!!!



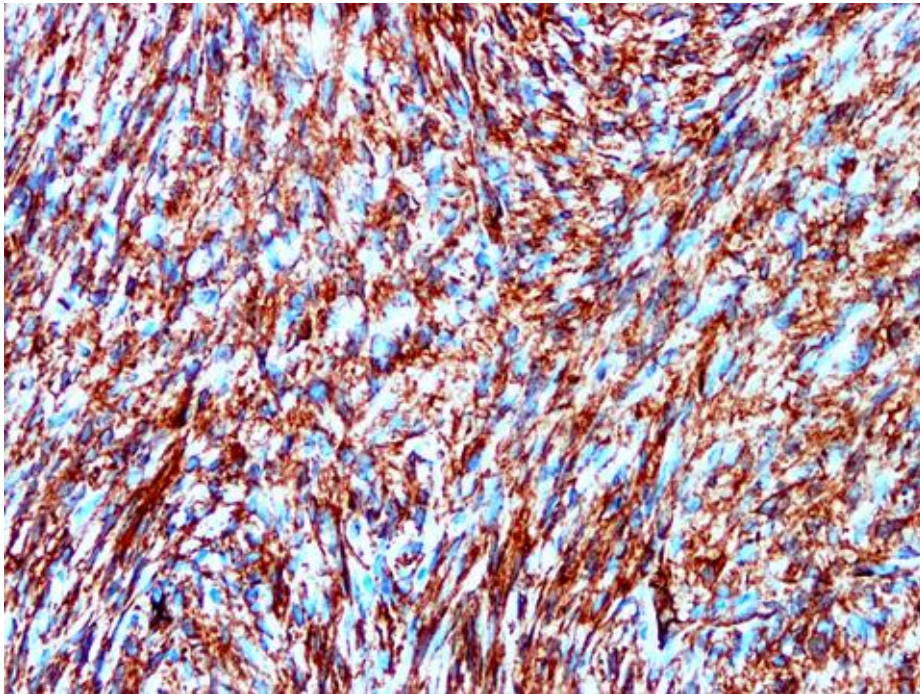
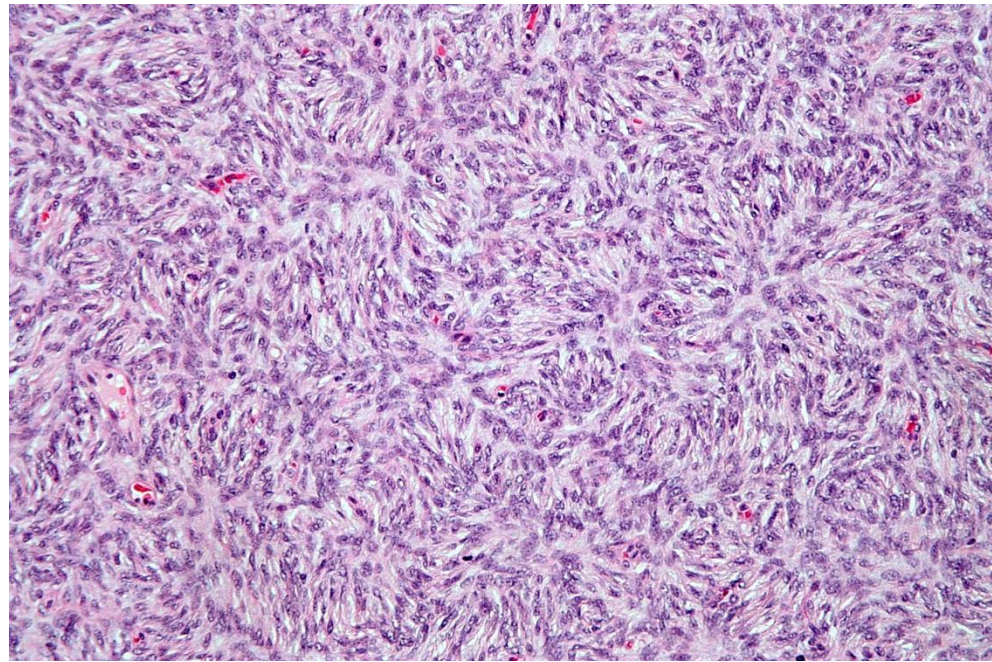
Primárny kožný sarkomatoidný skvamocelulárny karcinóm



a.vimentín b.CK5/6 c. p63 d. EMA

Dermatofibrosarcoma protuberans

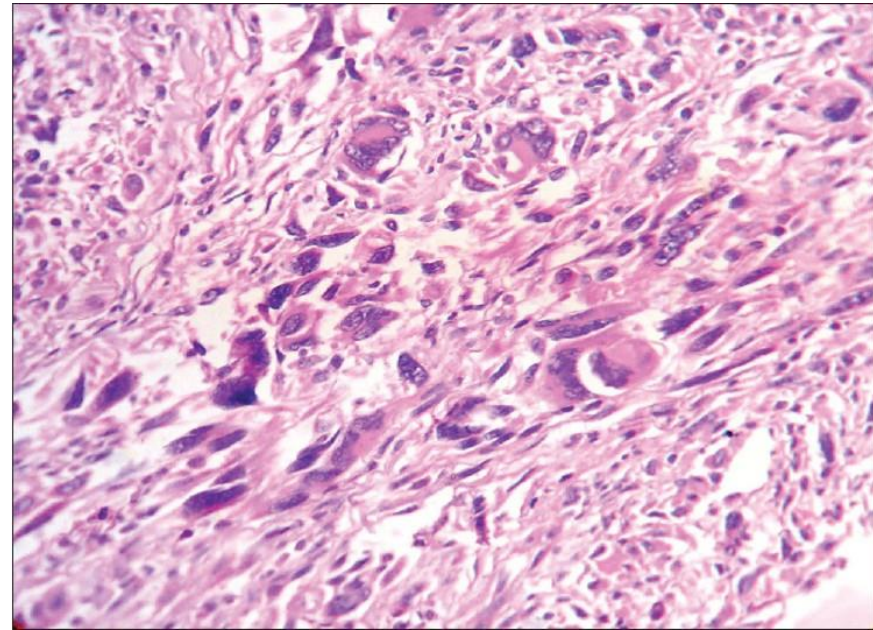
- mladší vek (aj deti)
- trup a horné končatiny
- storiformné usporiadanie
- CD34 + (vysoko senzitívny marker)
- v derme rast pozdĺž sept a medzi lipocyty „honeycomb“ vzhľad
infiltrácie



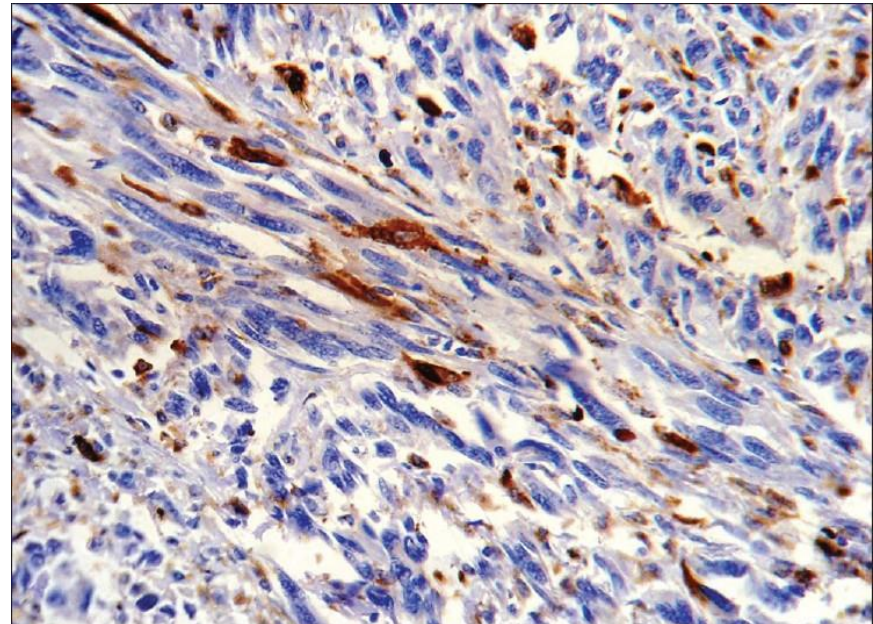
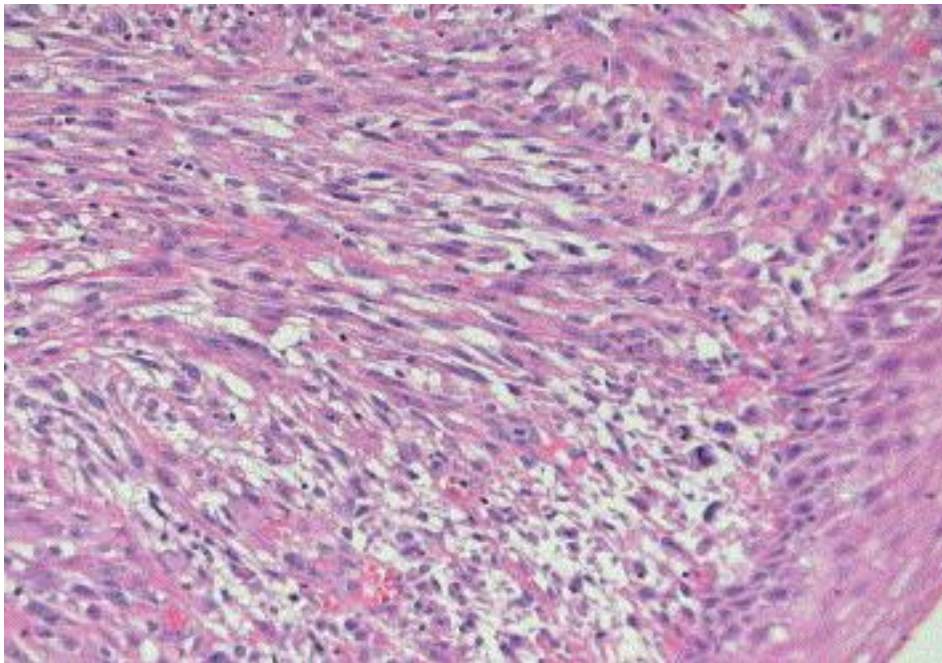
CD34 pozitivita

Atypický fibroxantóm, spindle variant

- hlava a krk
- koža poškodená slnkom
- rýchly rast
- lokálna invázia
- ulcerácia častá
- hemorágie časté (siderofágy)
- CD68 +, Cd117+, SMA-, desmin-, S100-



CD68 +



Ďakujem za pozornosť

