

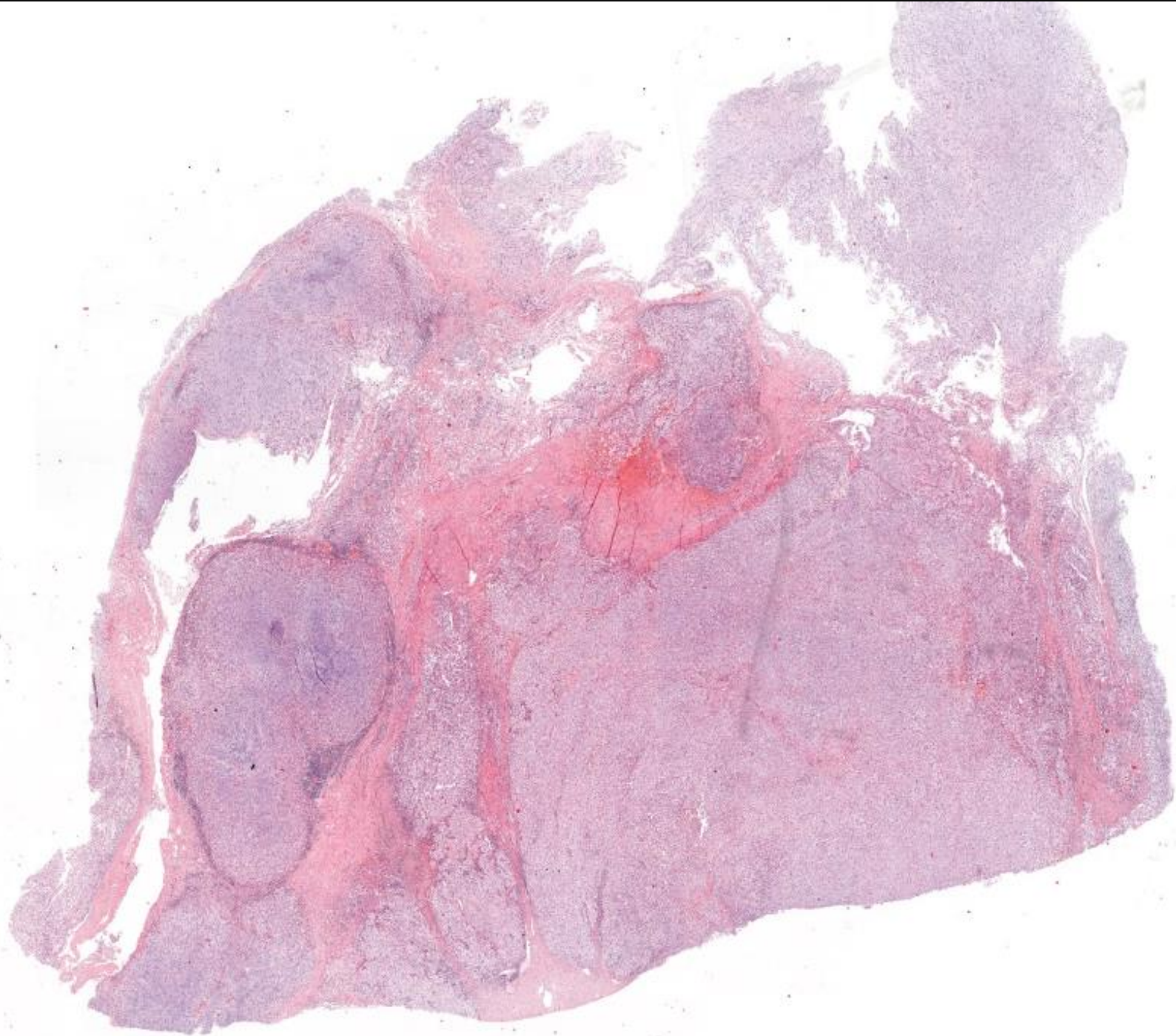
Prípád SD-IAP č. 668

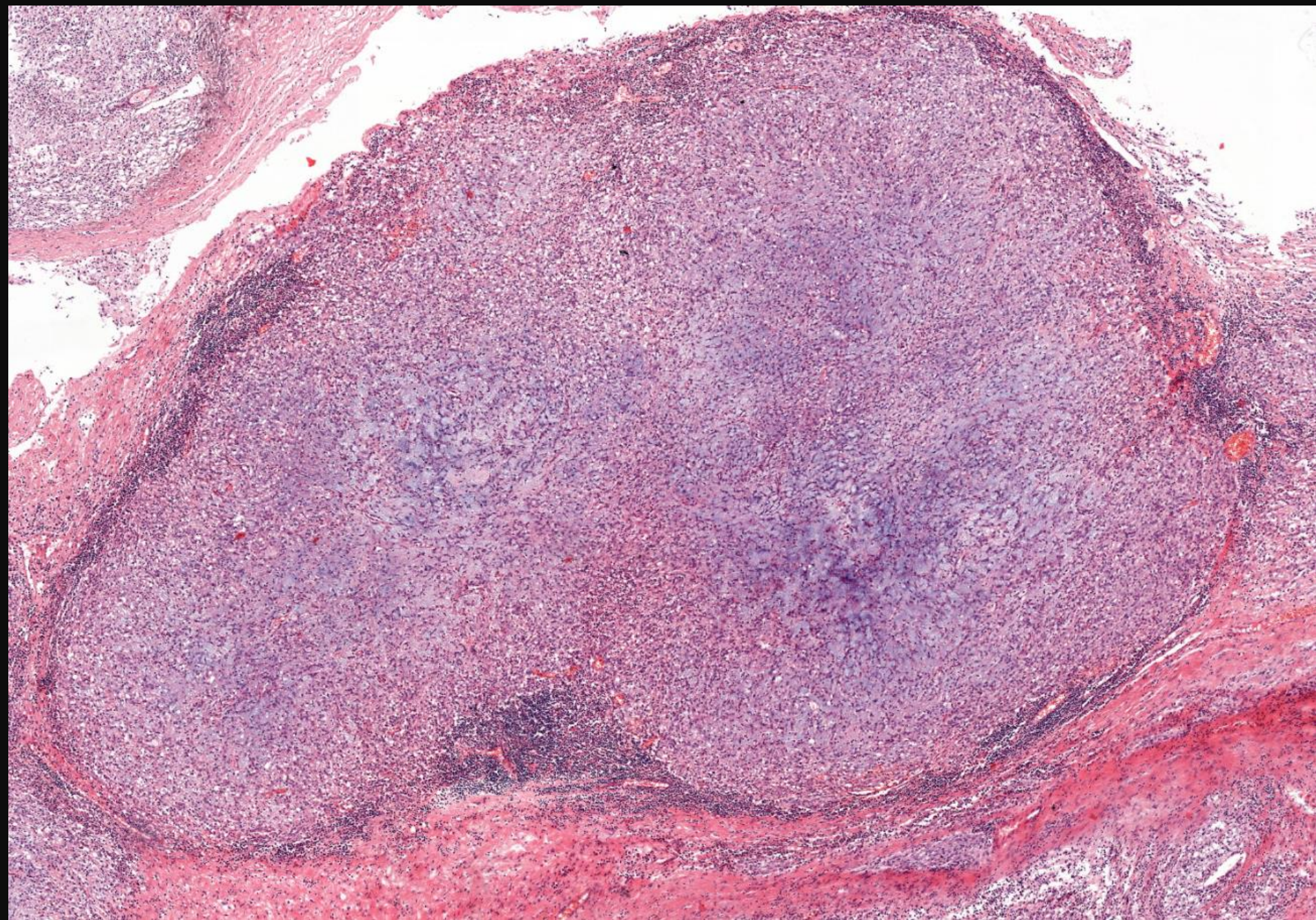
Boris Rychlý
Cytopathos s.r.o. Bratislava

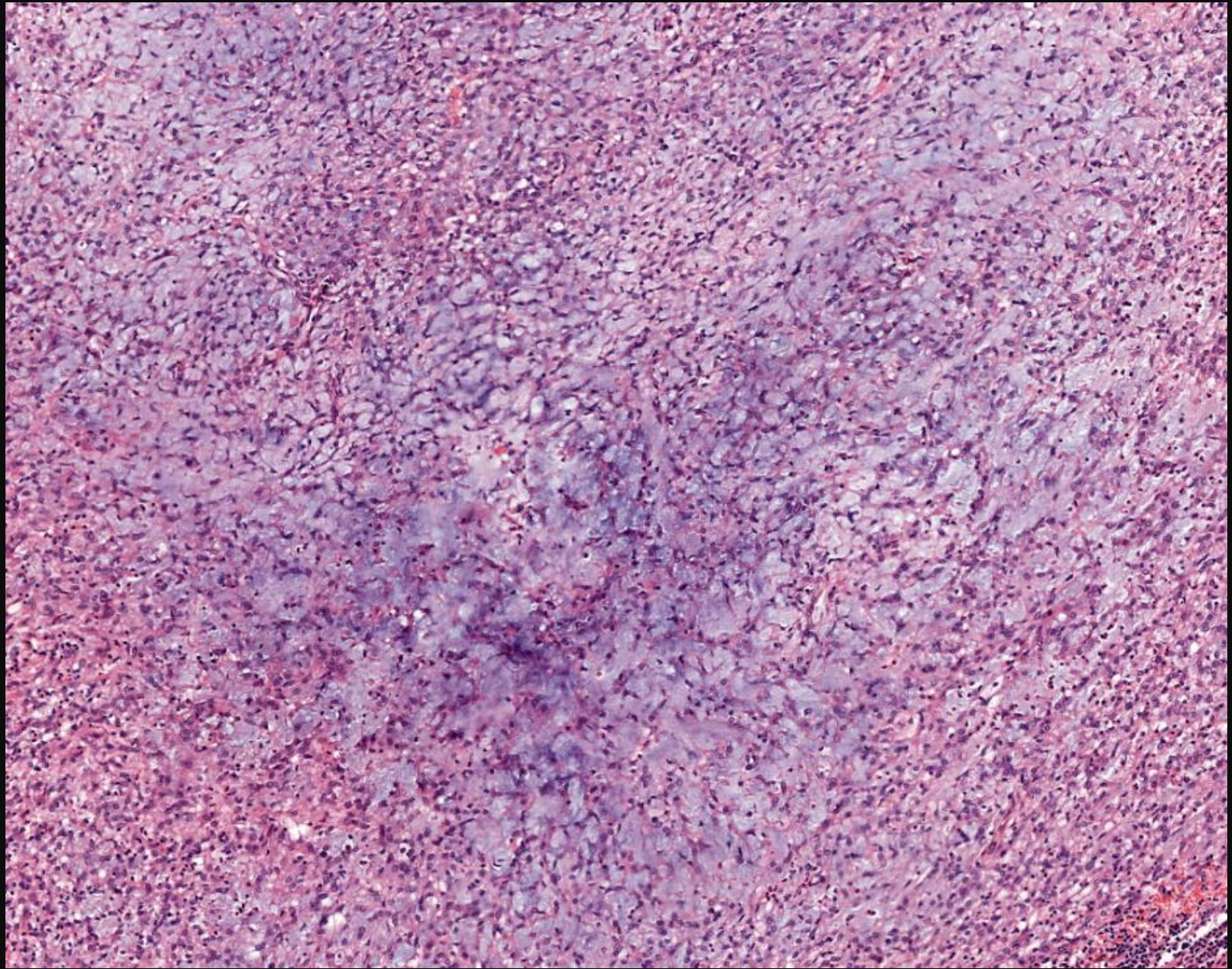
Adrian Kališ
ÚVN, Ružomberok

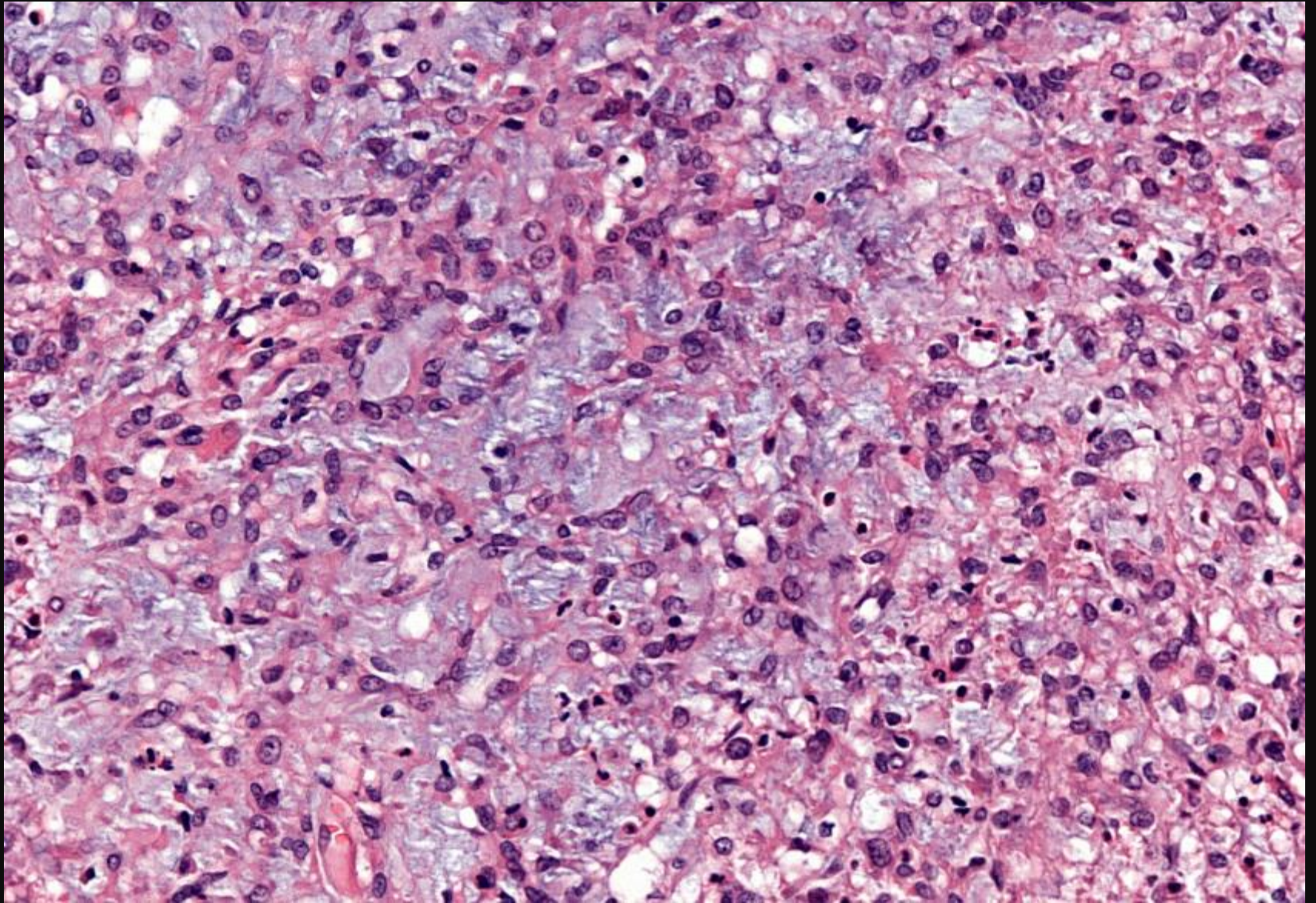
Klinické údaje

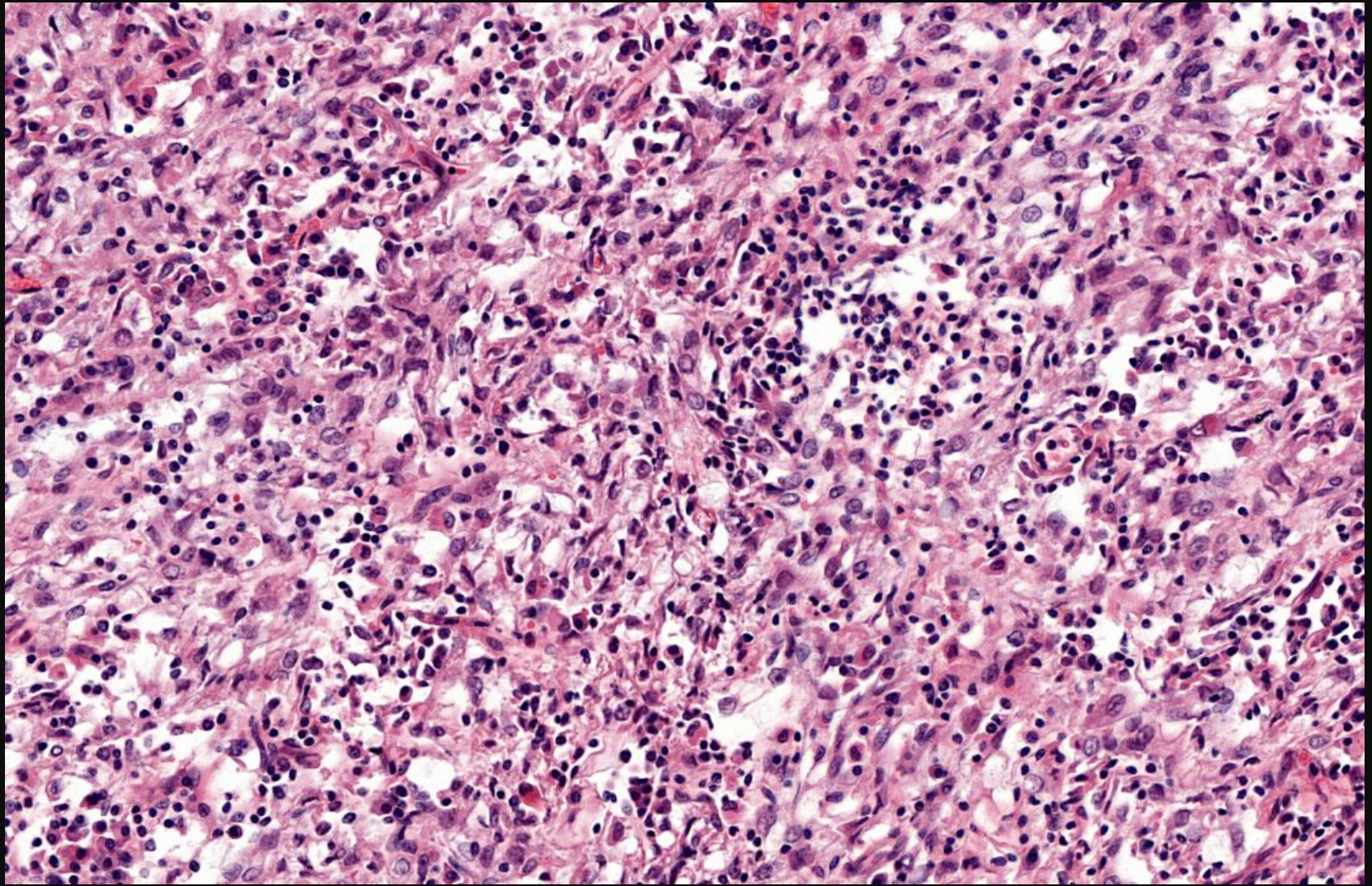
- 21-ročný muž, odobratá časť intra- a paraspinálneho tumoru v úrovni L5 - v. s. Schwannóm
- RTG popis - deštruktívny proces stavca

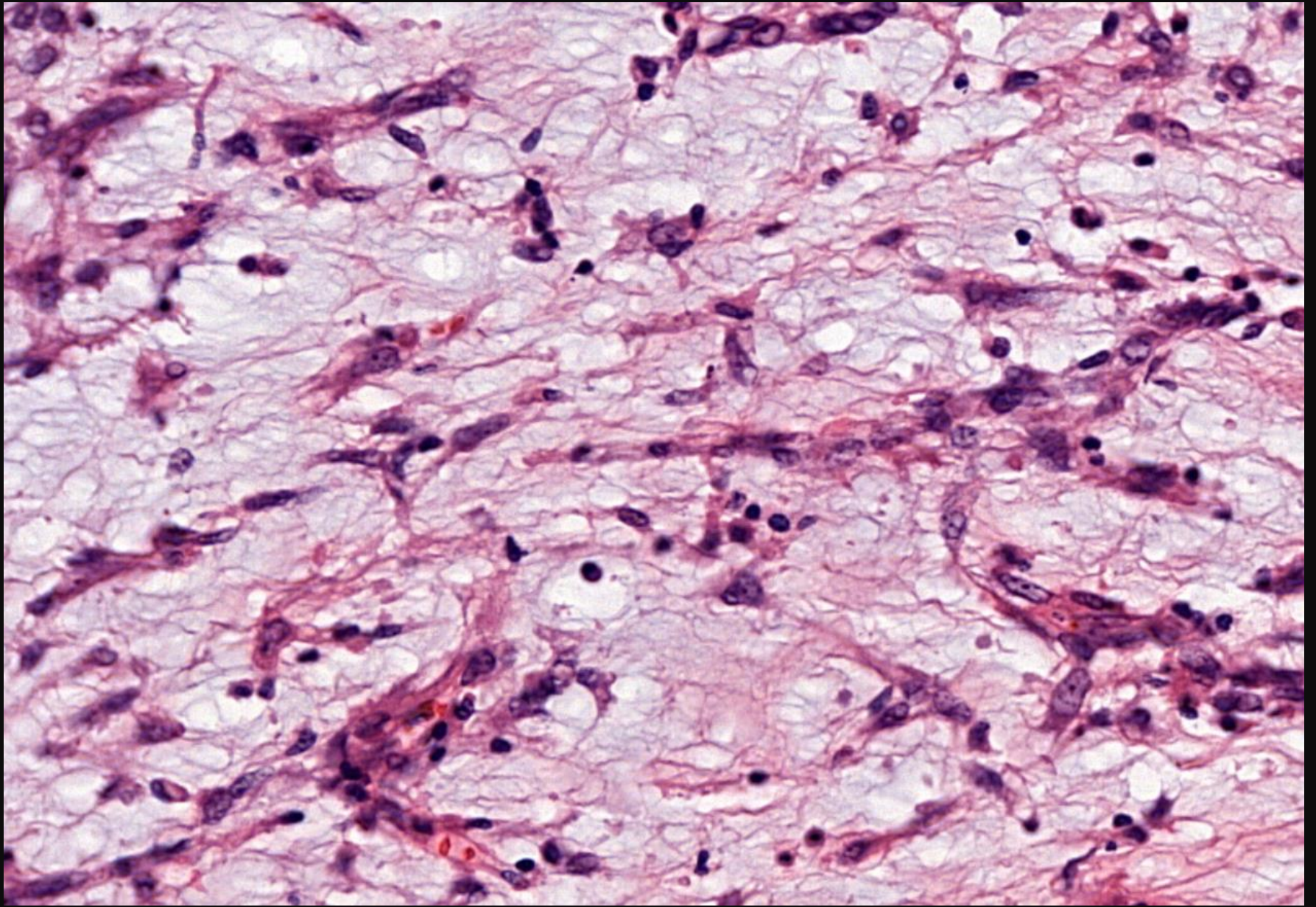


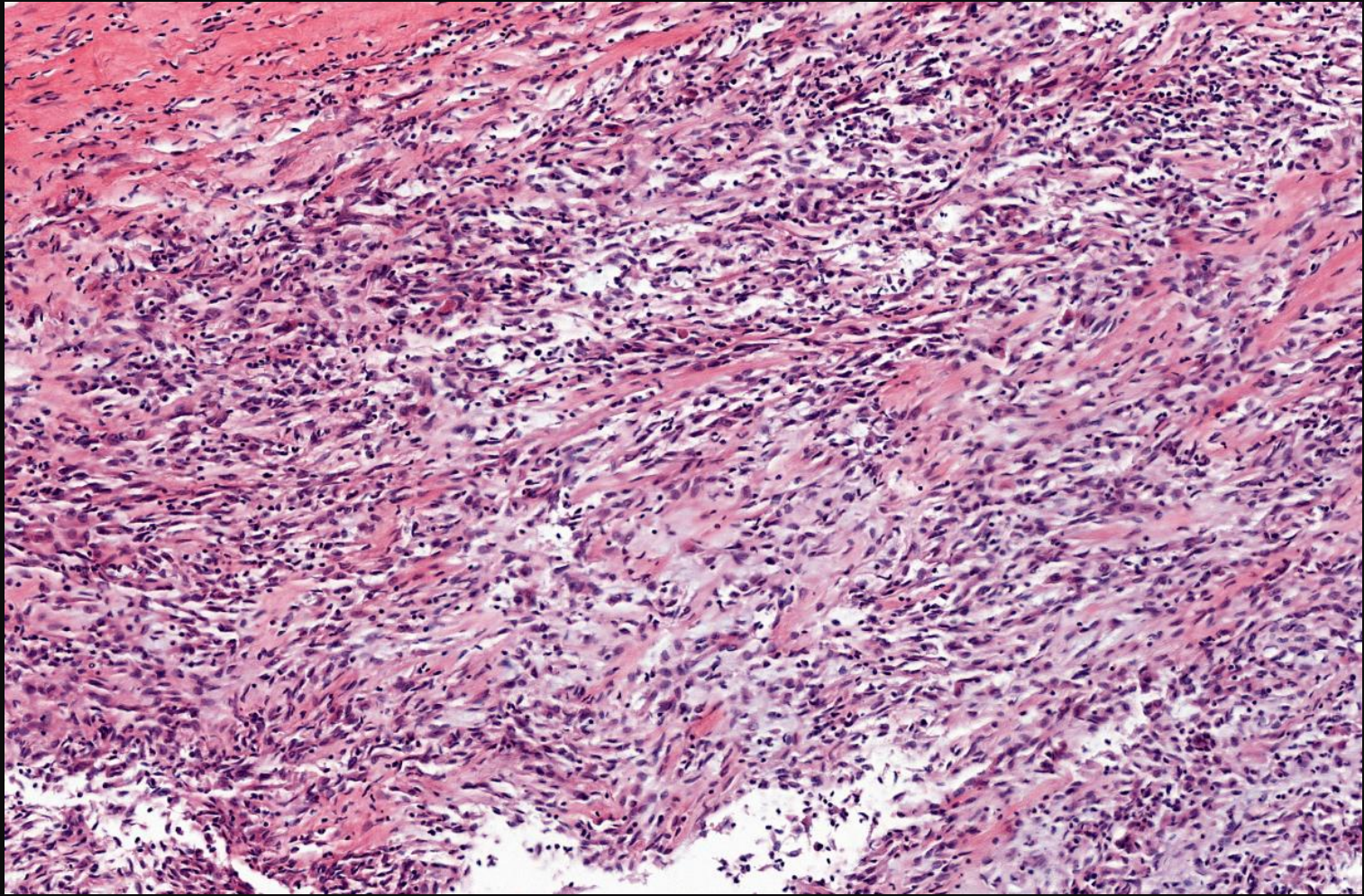


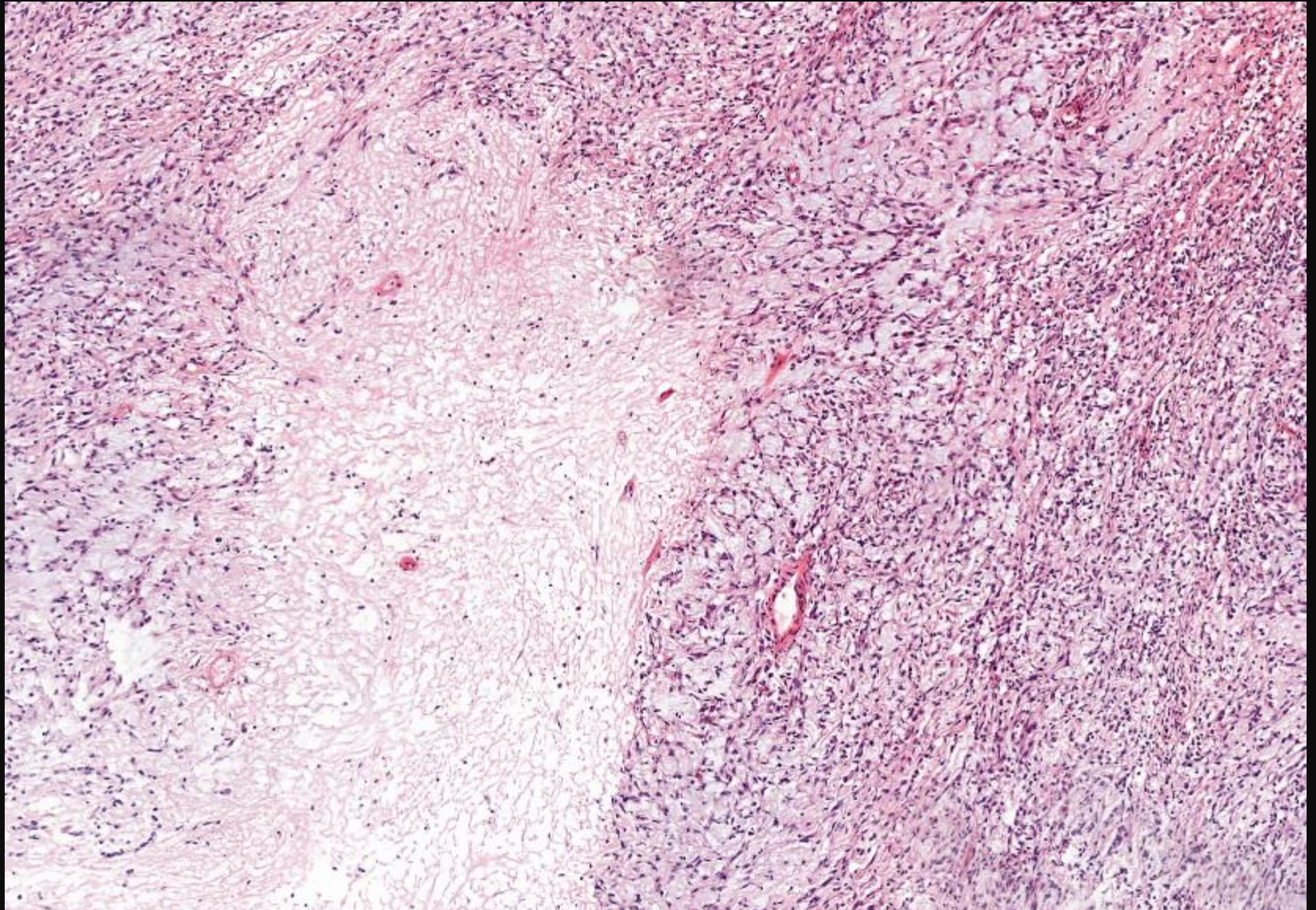


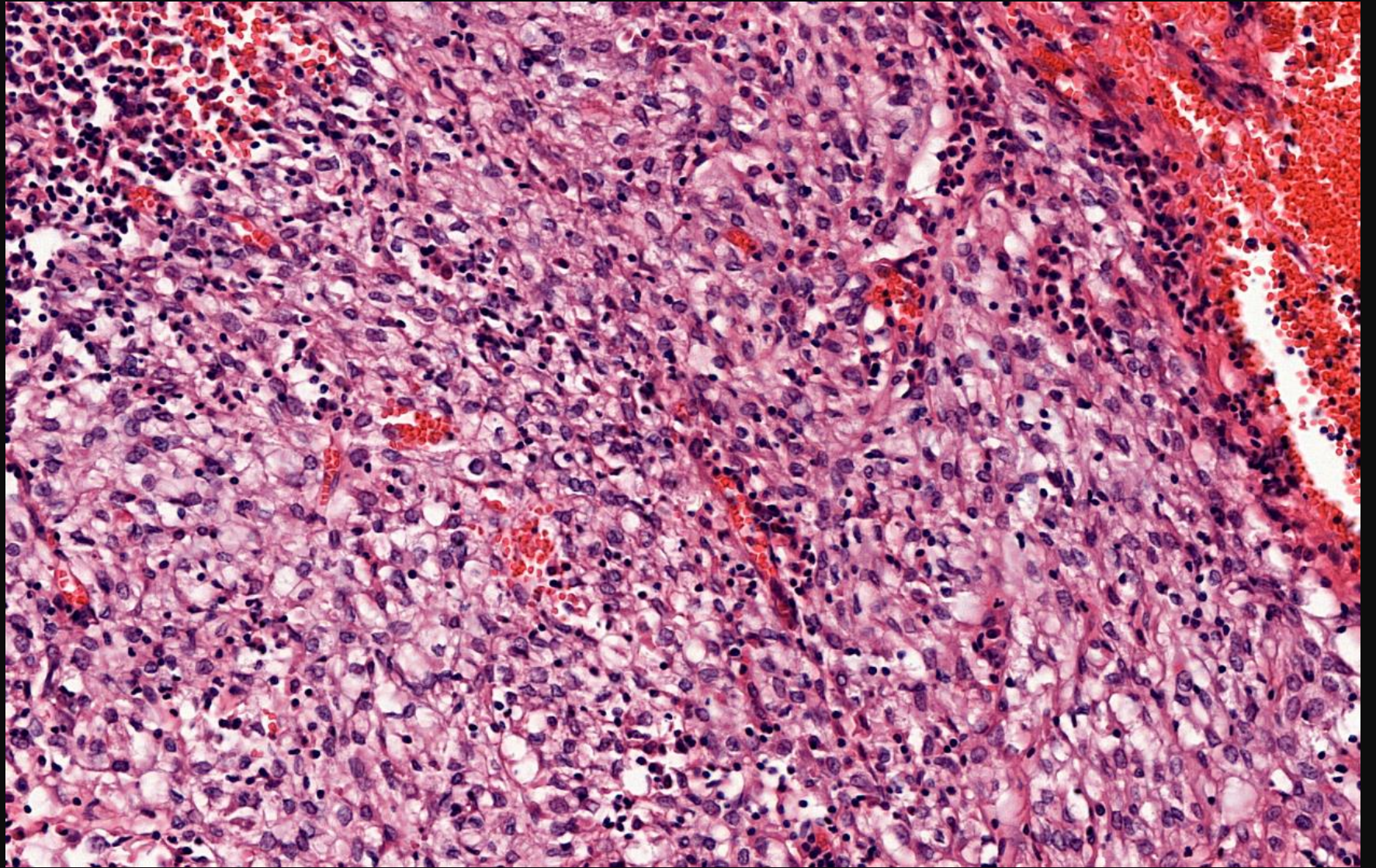


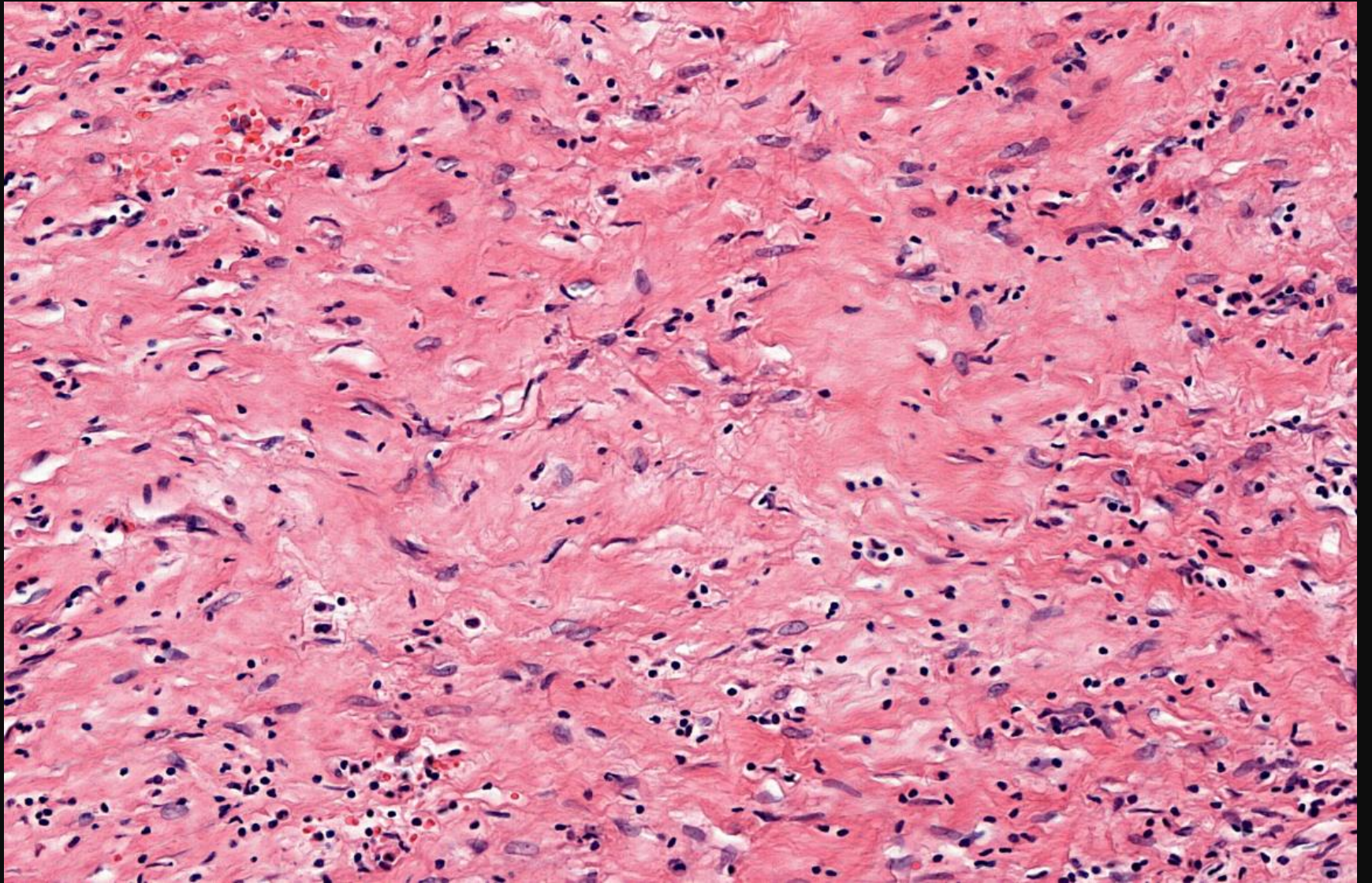


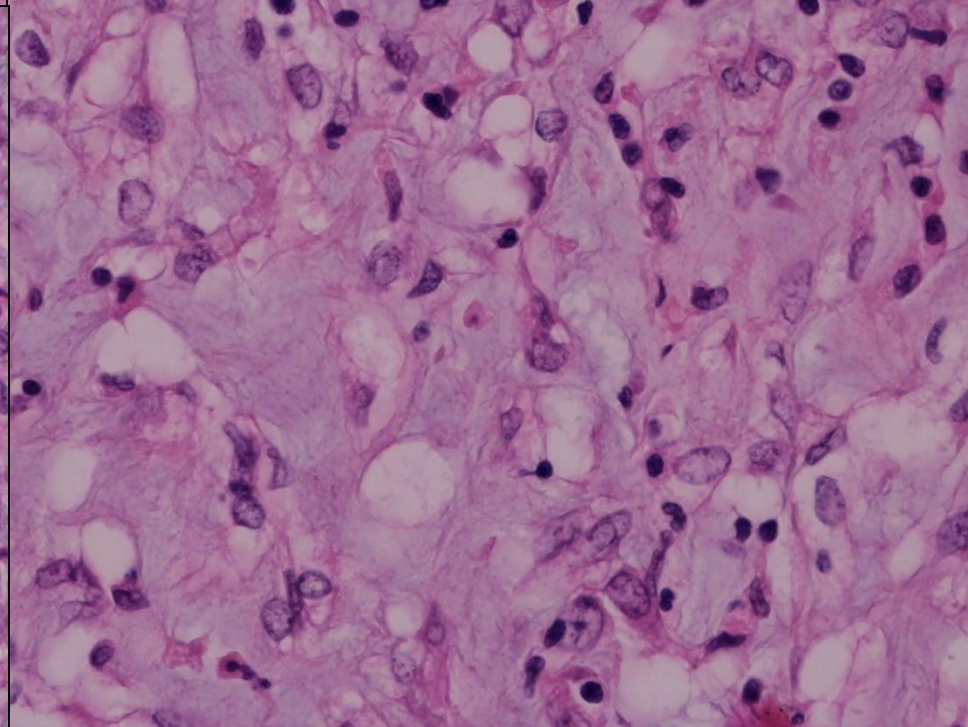
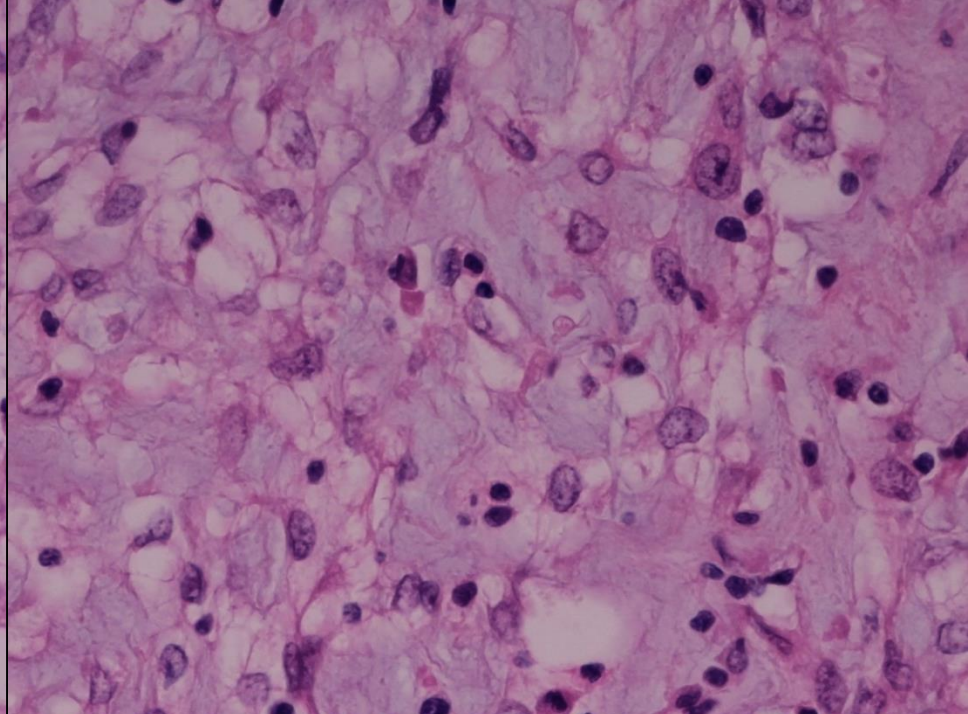
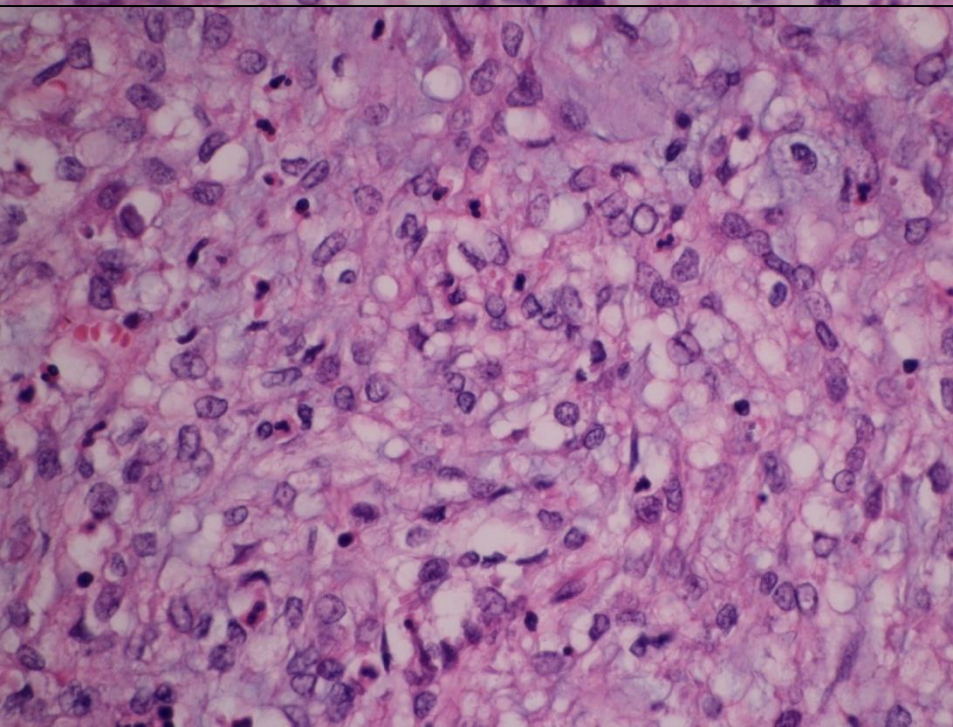
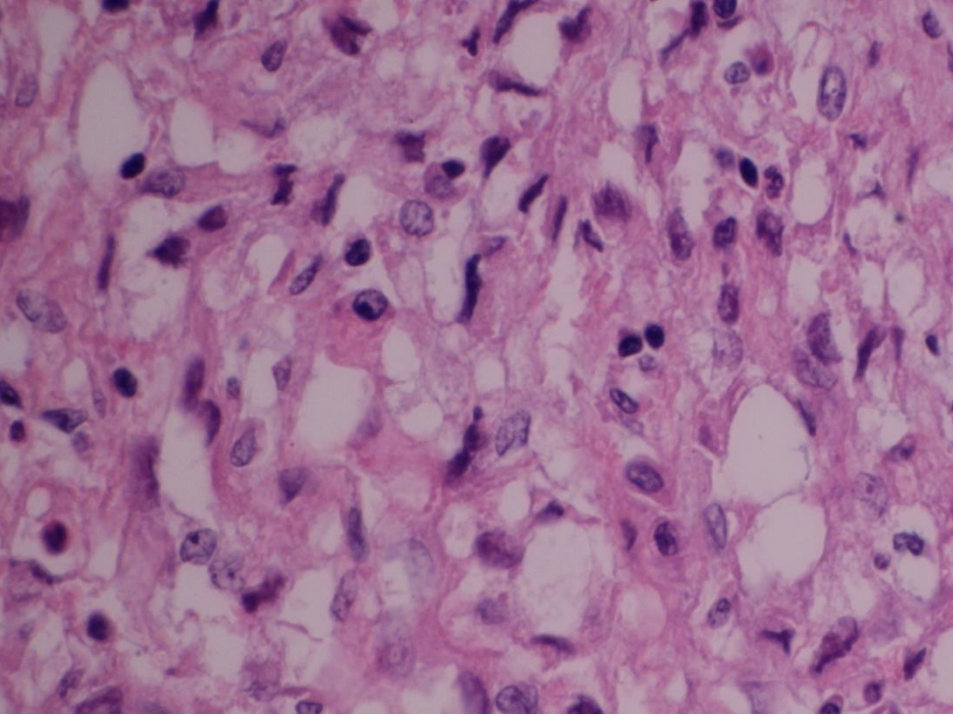


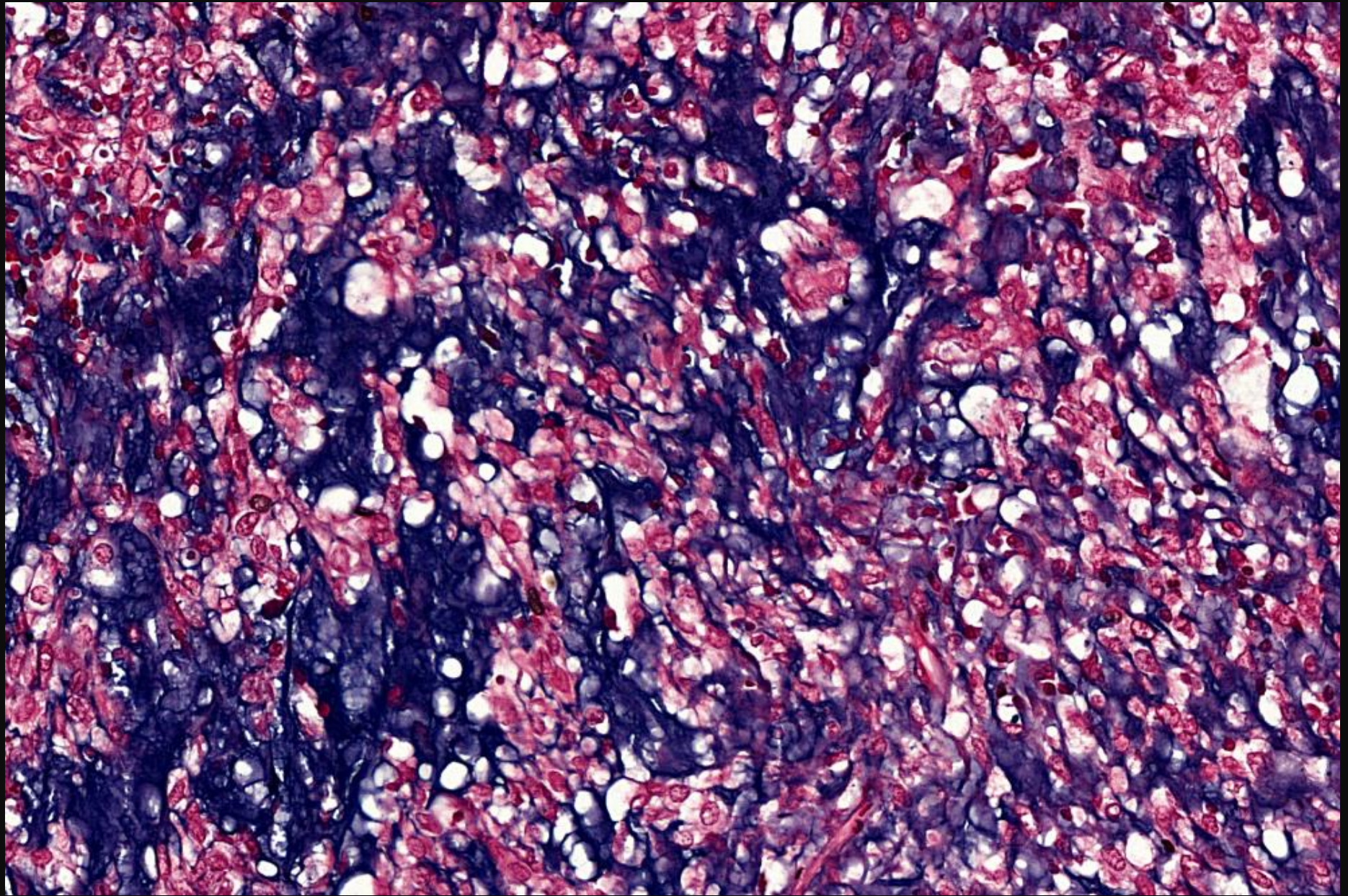










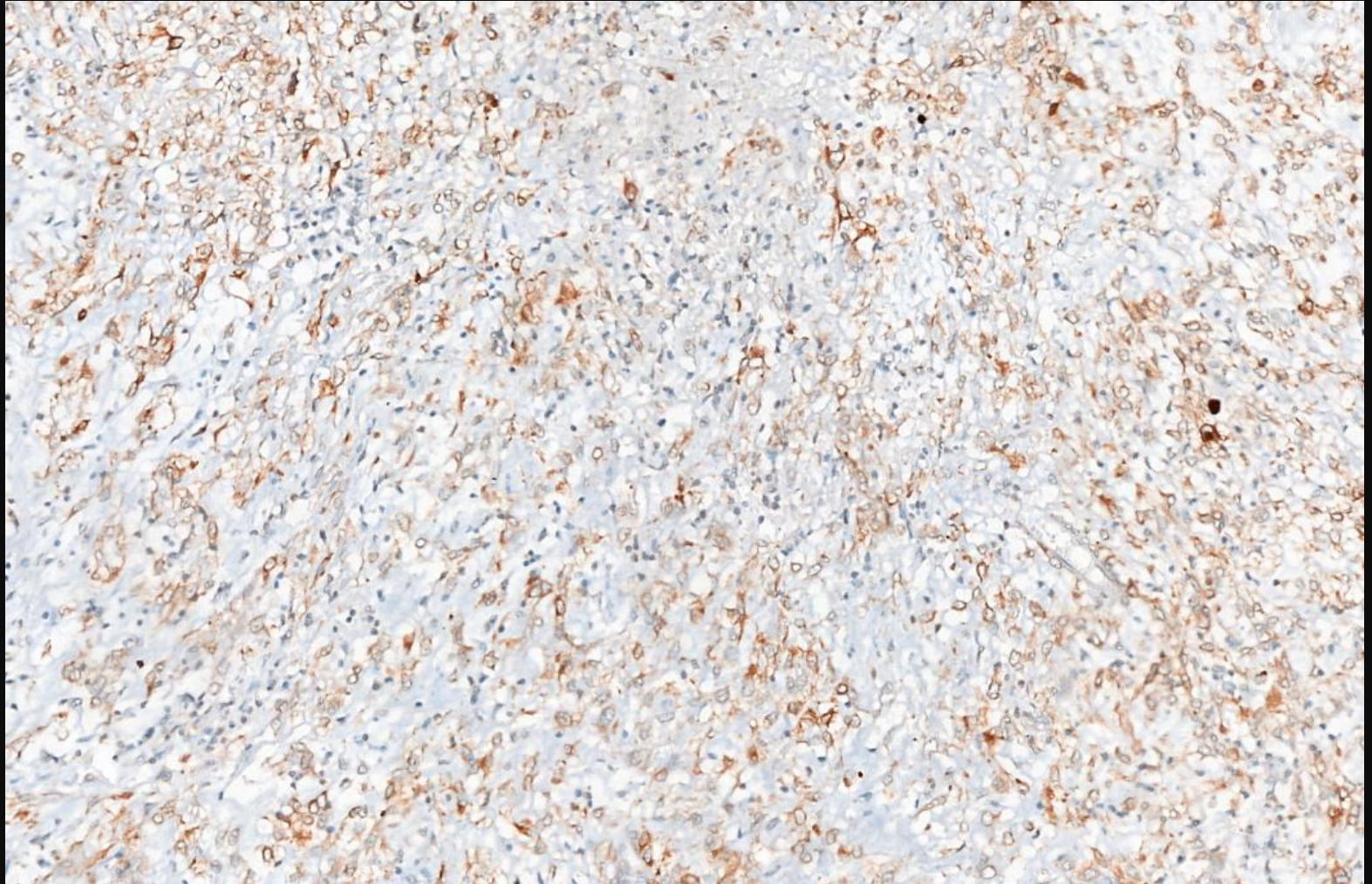


???

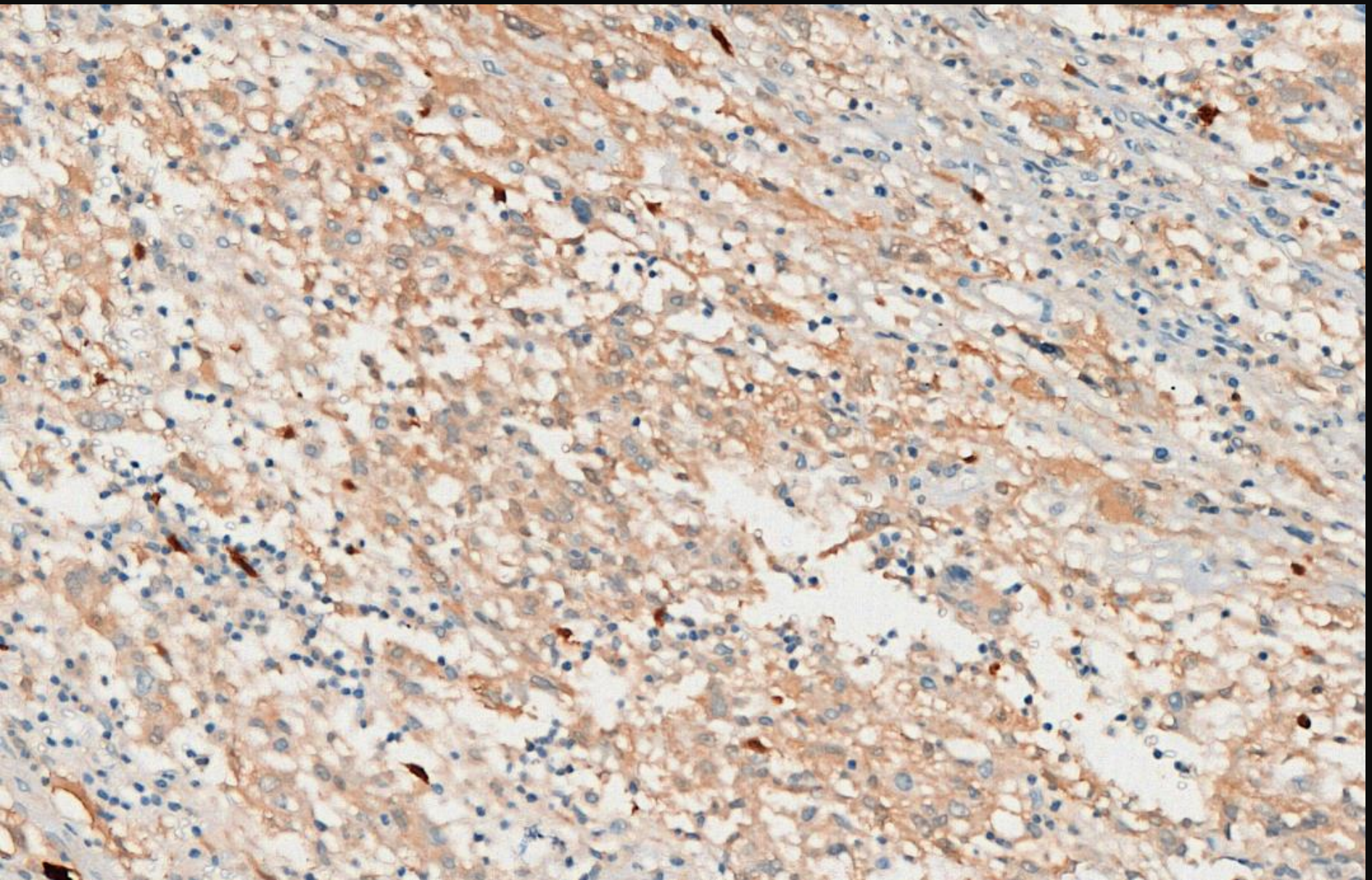
EMA



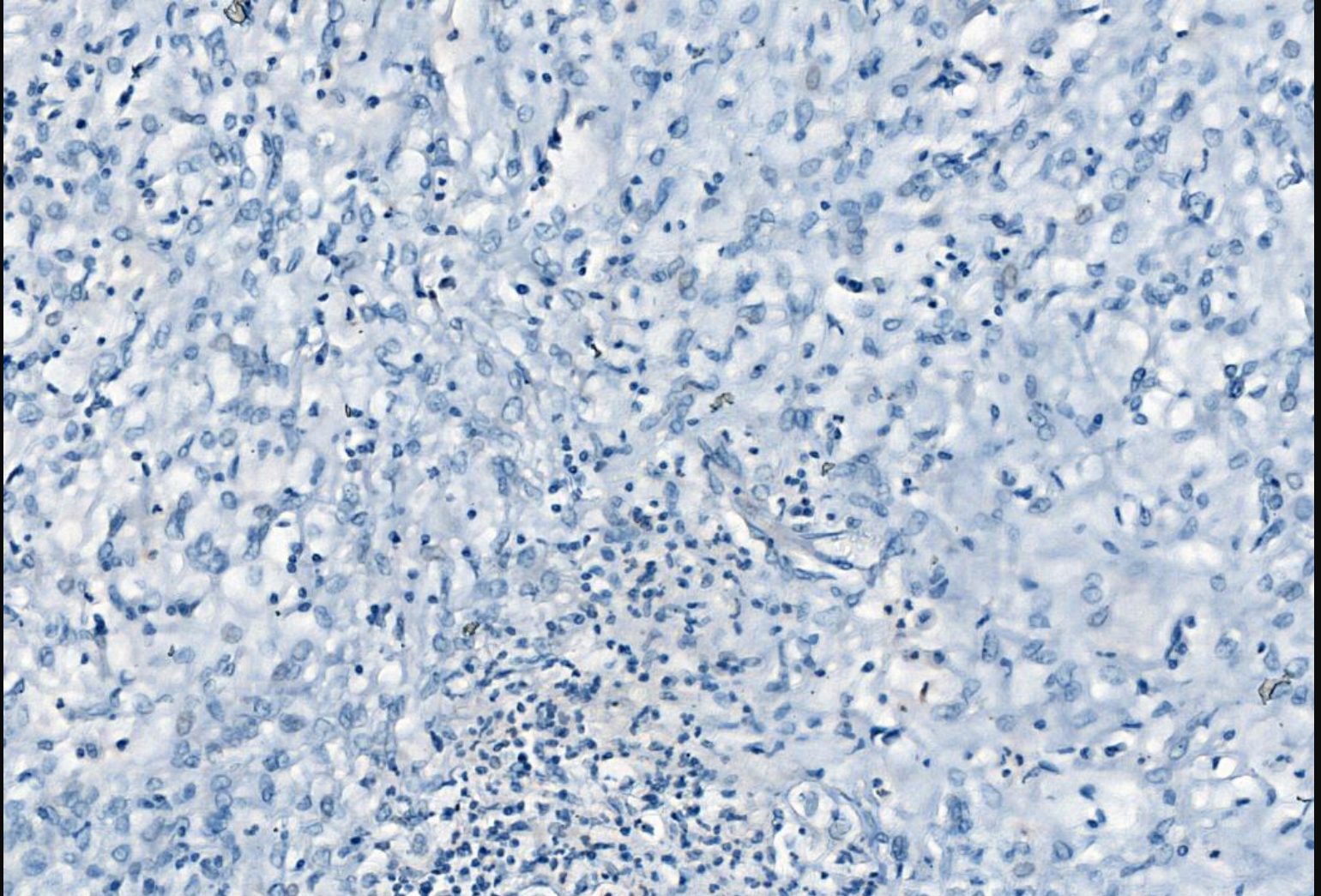
AE1/3



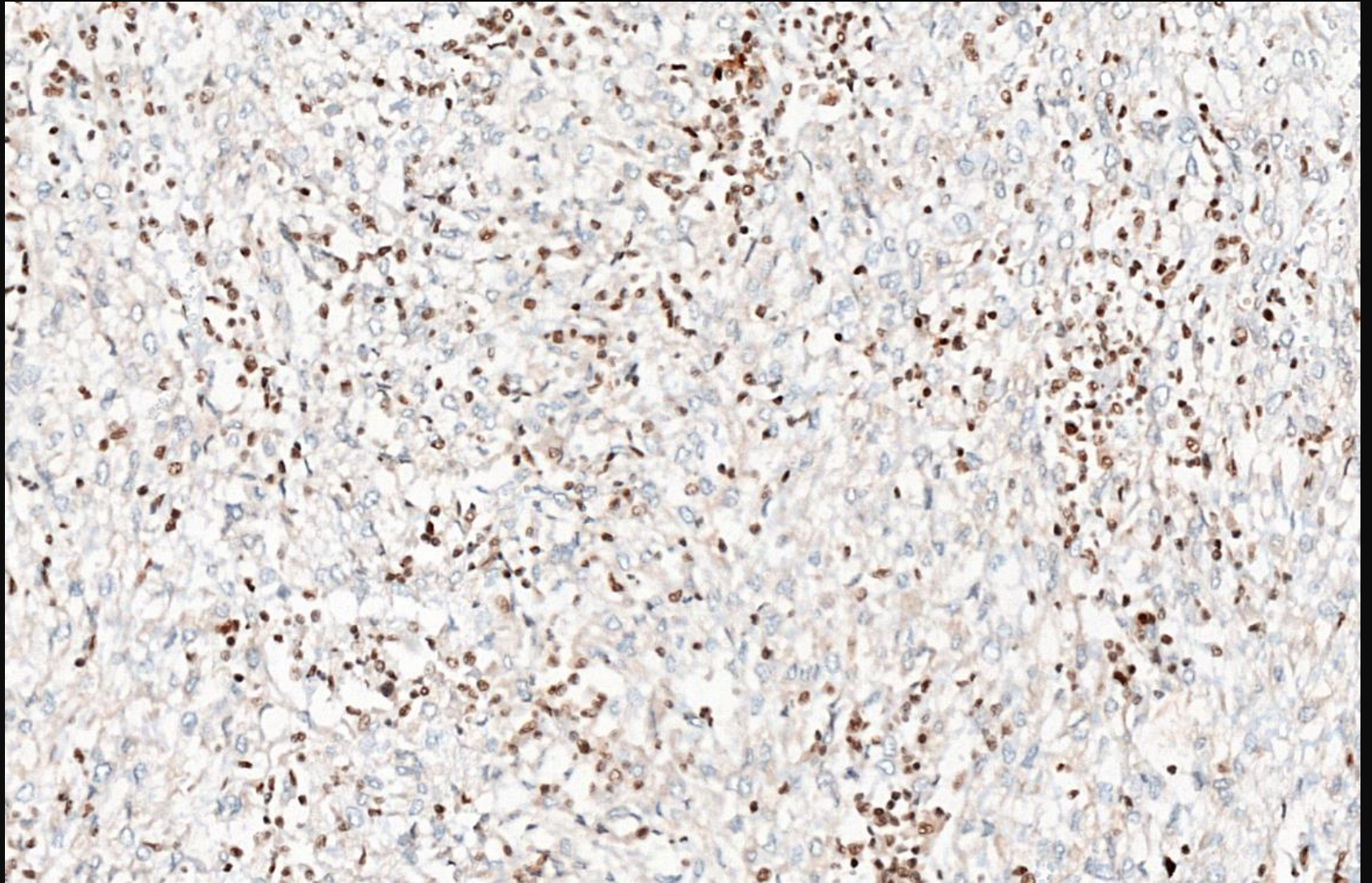
S100



GFAP

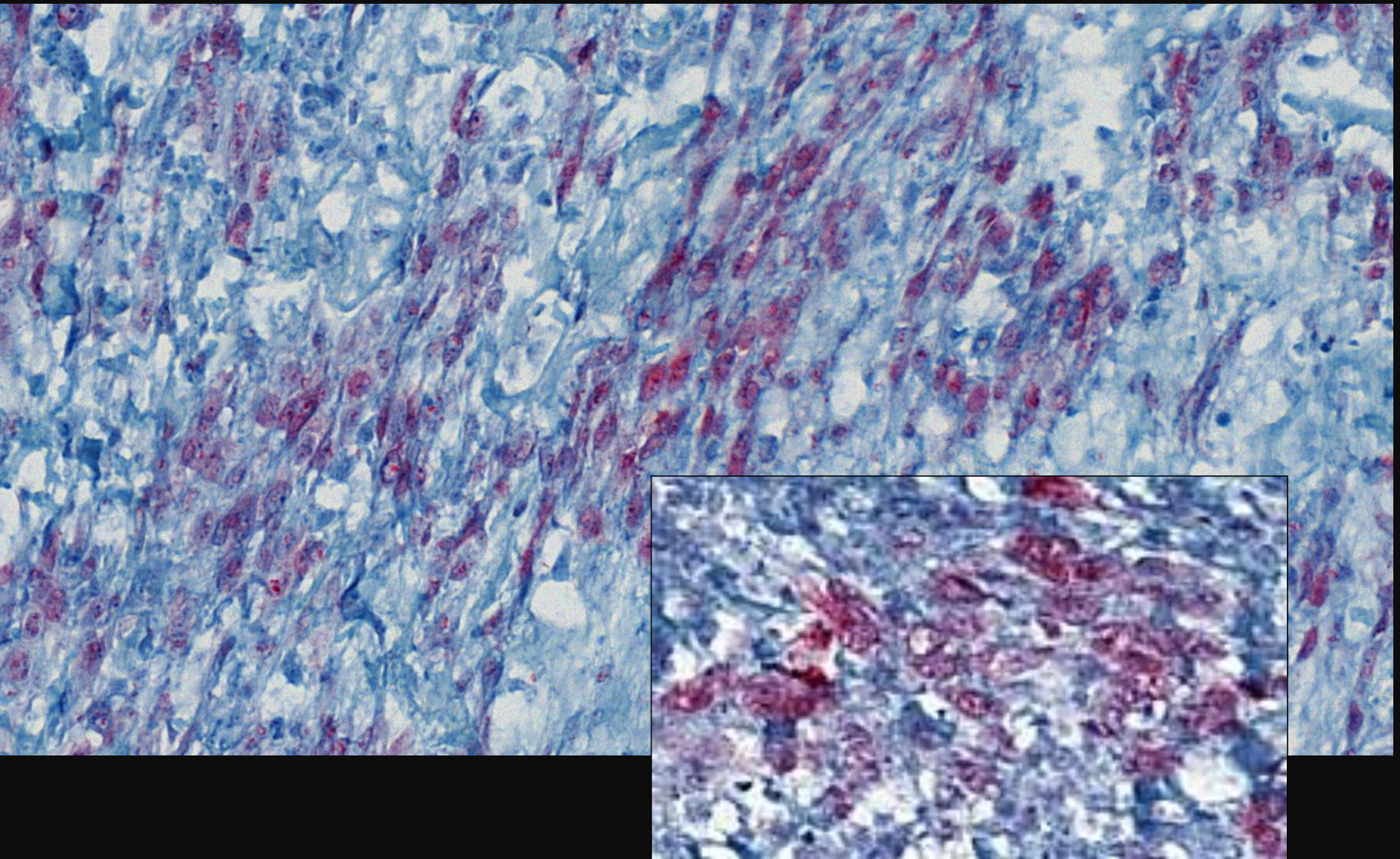


INI1



???

Brachyury

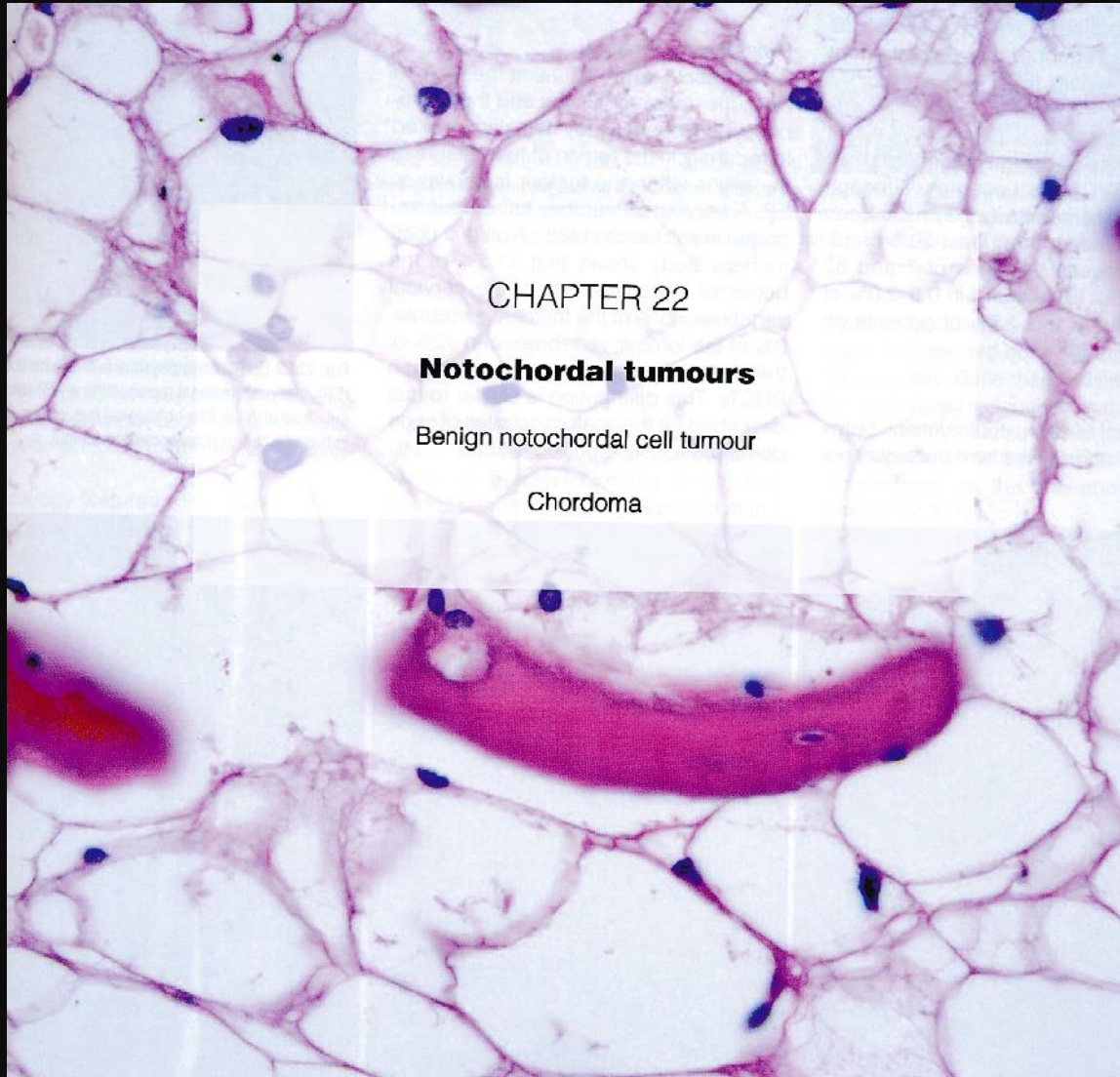


Naša diagnóza

Chordóm

(nízko diferencovaný)

WHO 2013



CHAPTER 22

Notochordal tumours

Benign notochordal cell tumour

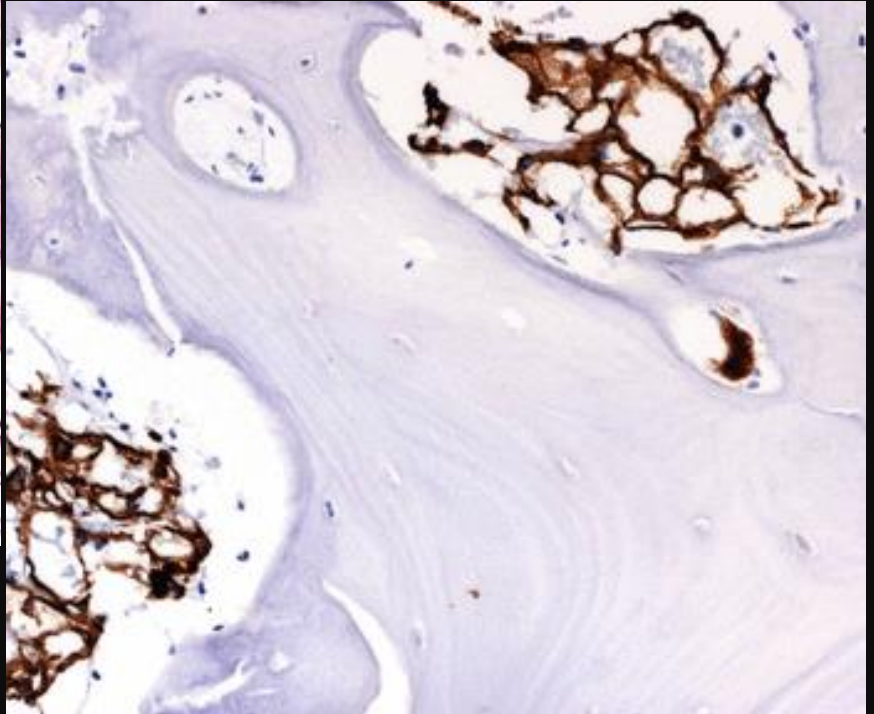
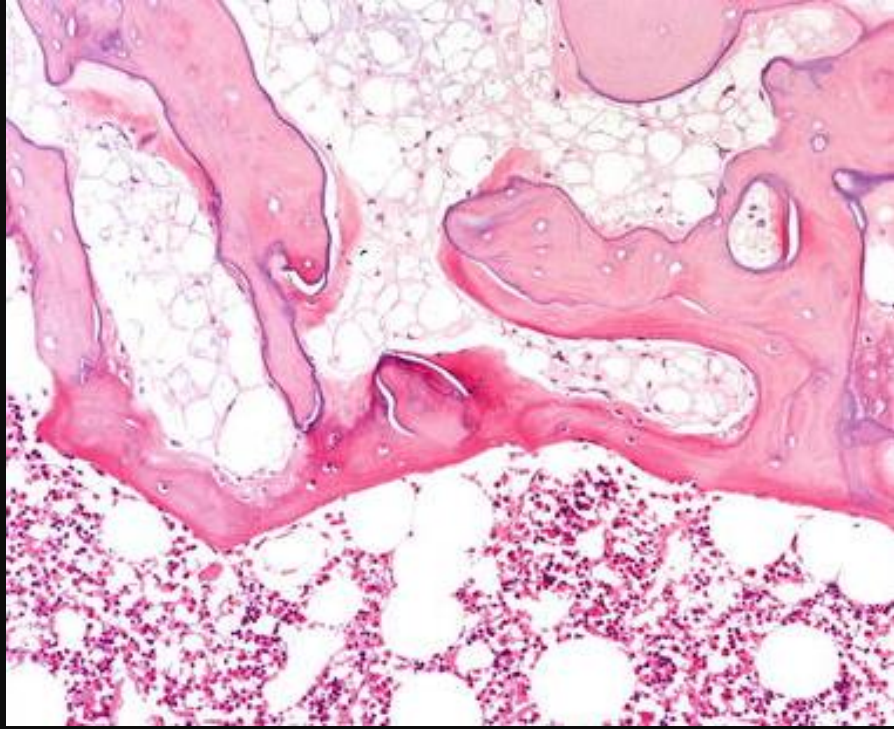
Chordoma

Notochorda – notochordálne TU

- notochorda – nucleus pulposus intravert. diskov
- intraoseál. notochordálne zvyšky
(náhodný nález, menej ako 10mm)
- ecchordosis physaliphora (zvyšky v clive)
- benígny notochordálny TU
- atypický notochordálny TU (incipientný chordóm)
- konvenčný chordóm
- nízko diferencovaný chordóm (atypický chordóm)
- dediferencovaný chordóm

Benígny notochordal cell tumor

- 2001(Mirra and Brien) 2 prípady giant notochordal hamartoma
- giant notochord rest, benign notochordal cell lesion, notochord. hamartóm, benign notochordal cell tumor (WHO)
- od konvenčného – histologicky a rádiologicky – náhodne na MR z iných príčin (na RTG väčšinou 0)



Benígny notochordal cell tumor

- oproti chordómu bez lobulácie, „kordingu“, myxoidného pozadia, fibróznych sept, mitóz, nekróz, nukleárnych atypií
- 7% CH má aj BNCT zložku
- BNCT – 20% pri autopsii
- CH – 8,4 / 10 000 000 (USA)

Atypical Notochordal Cell Tumors

A Series of Notochordal-derived Tumors That Defy Current Classification Schemes

Jodi M. Carter, MD, PhD, Doris E. Wenger, MD,† Peter S. Rose, MD,‡
and Carrie Y. Inwards, MD**

(Am J Surg Pathol 2017;41:39–48)

- histologicky benígny (prípadne fokálne myxoidná zmena) ale rádiologicky minimálna kortikálna permeácia / soft tissue extenzia

Modern Pathology (2005) 18, 1005–1010
© 2005 USCAP, Inc. All rights reserved 0893-3952/05 \$30.00
www.modernpathology.org



Incipient chordoma: a report of two cases of early-stage chordoma arising from benign notochordal cell tumors

Takehiko Yamaguchi¹, Hiroaki Watanabe-Ishiiwa², Seiichiro Suzuki³, Yoshihiko Igarashi⁴
and Yoshihiko Ueda¹

Konvenčný chordóm

- chordóm – malígný, osteolytický, deštruktívny, cez kortikalis s extenziou do mäkkých tkanív
- lobulárny, myxoidná matrix, pruhy buniek
- nukleárne atypie
- 50% sakrálnych, 30% lebka, 20% mobilná chrbtica

Nízko diferencovaný chordóm

- solídny rast, zvýšená celularita, mitózy, atypie, nekrózy
- delécia zasahujúca SMARCB1
- strata INI1 expresie
- biologicky odlišný od konvenčného chordómu

Acta Neuropathol (2010) 120:745–753
DOI 10.1007/s00401-010-0767-x


ORIGINAL PAPER

Loss of SMARCB1/INI1 expression in poorly differentiated chordomas


Bret C. Mobley · Jesse K. McKenney · Charles D. Bangs · Katherine Callahan ·
Kristen W. Yeom · Reinhard Schneppenheim · Melanie G. Hayden · Athena M. Cherry ·
Murat Gokden · Michael S. B. Edwards · Paul G. Fisher · Hannes Vogel


107TH ANNUAL MEETING
GEARED TO LEARN




 **Bone and Soft Tissue Pathology**

Clinicopathologic Characteristics of Poorly Differentiated Chordoma

 Tue, March 20

 8:00 AM - 8:15 AM

 VCC West 205 - 206

 Platform Presentation

Part of:

Bone & Soft Tissue - Tuesday AM Platform

8:00 AM - 12:00 PM

Authors



Angela Shih, MD

Dediferencovaný chordóm

- konvenčný chordóm +
nediferencovaný sarkóm
(zachováva brachyury)

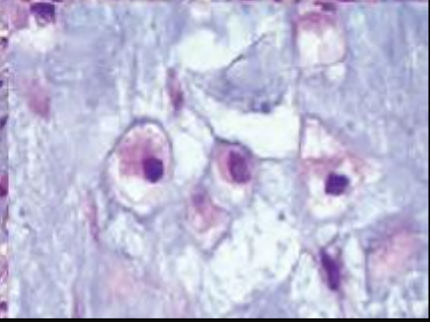
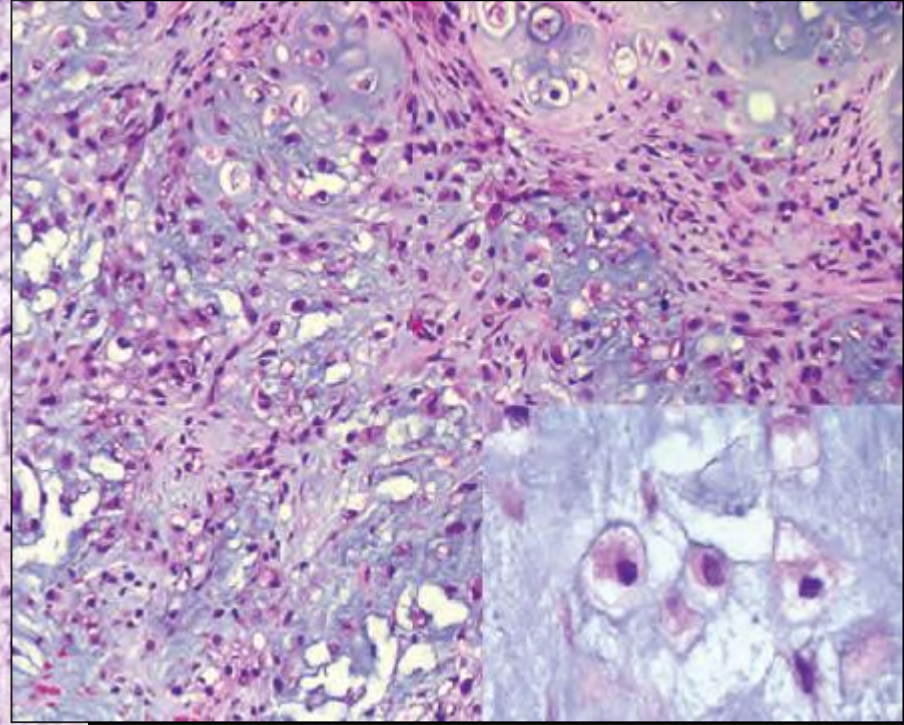
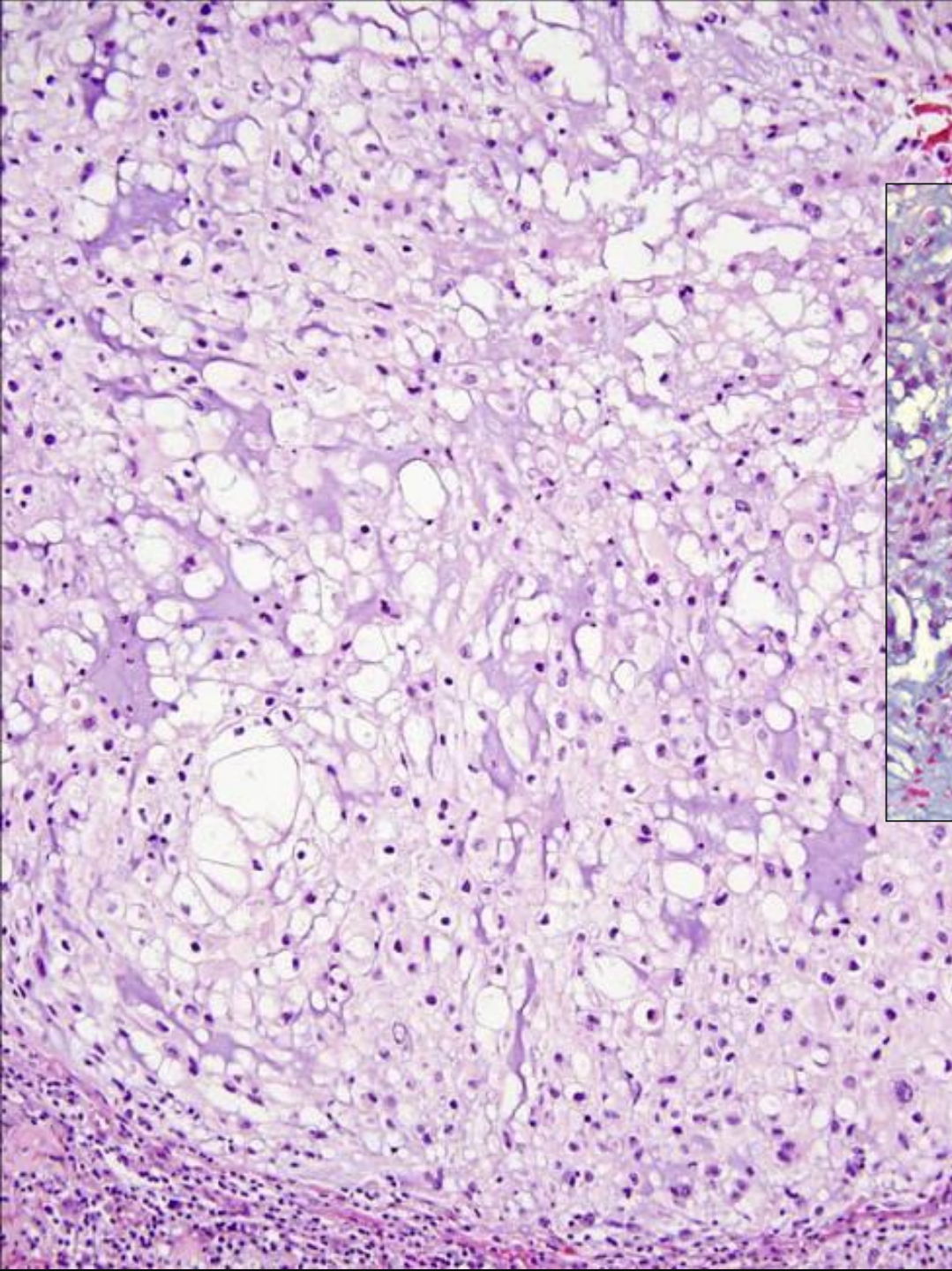
Existuje soft tissue chordóm? (alebo kostný extraaxiálny)

- Dabská – parachodróóm
(Jozef Laskowski chordoma periphericum)
- soft tissue myoepitelióm
- SF chordóm (s brachyury)
– extrémne zriedkavý
(vylúčiť MTS)
- v literatúre len 2 soft tissue
+ 9 v extraaxiálnom skelete



Brachyury

- transkripčný faktor kľúčový pre vývoj notochordy
- senzitivita 89,7 – 100% (vrátane dediferencovaných a metastatických)
- špecificita prakticky 100%
- z iných nádorov len chytoplazmatická pozitivita stromálnych buniek hemangioblastómu
- spermatogónia a niektoré testikulárne GCT



Dif. dg.

- myxoidný chondrosarkóm,
chondroblastický osteosarkóm,
mucinózny adenokarcinóm, chordoidný
meningióm, chordoidný glióm, ...
- **extraskeletálny myxoidný
chondrosarkóm, soft tissue
myoepitelióm**

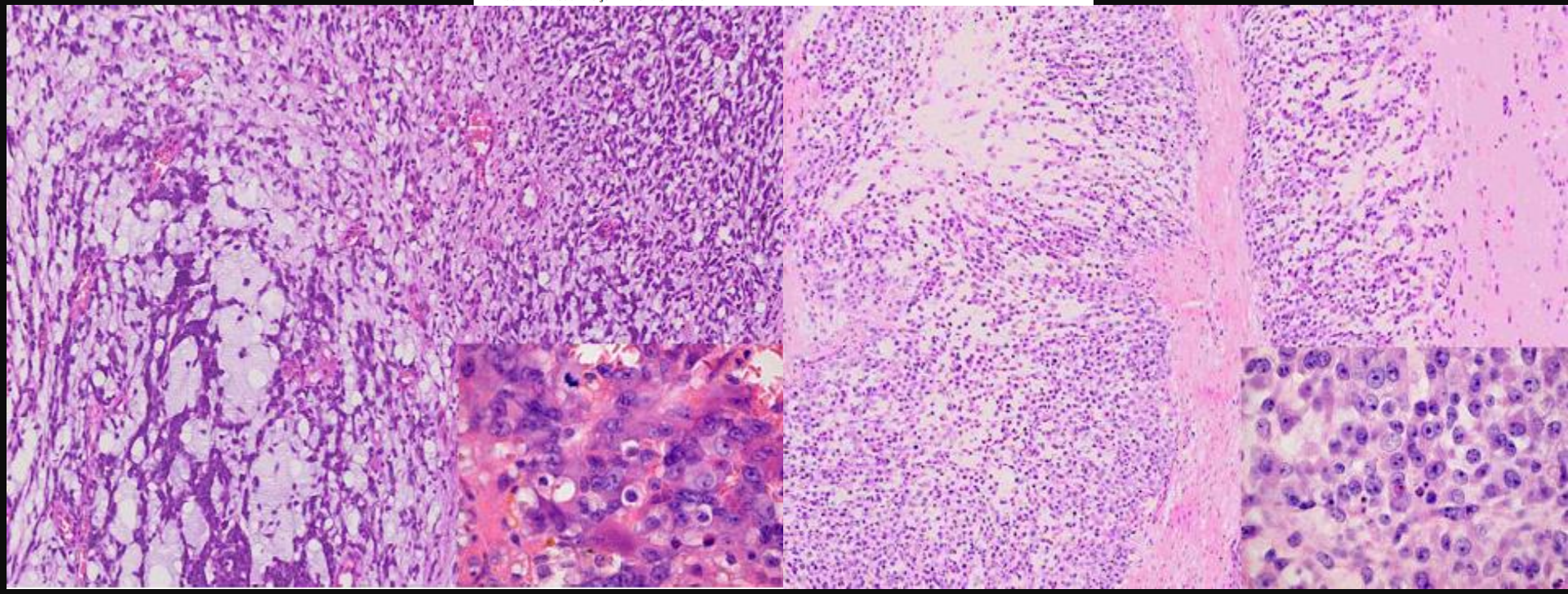
Extraskelletálny myxoidný chondrosarkóm

Virchows Arch (2012) 460:621–628
DOI 10.1007/s00428-012-1240-0

ORIGINAL ARTICLE

NR4A3 rearrangement reliably distinguishes between the clinicopathologically overlapping entities myoepithelial carcinoma of soft tissue and cellular extraskelletálny myxoid chondrosarcoma

Uta Flucke · Bastiaan B. J. Tops · Marian A. J. Verdijk ·
Patricia J. H. van Cleef · Peter H. van Zwam ·
Pieter J. Slootweg · Judith V. M. G. Bovée ·
Robert G. Riedl · David H. Creyten ·
Albert J. H. Suurmeijer · Thomas Mentzel



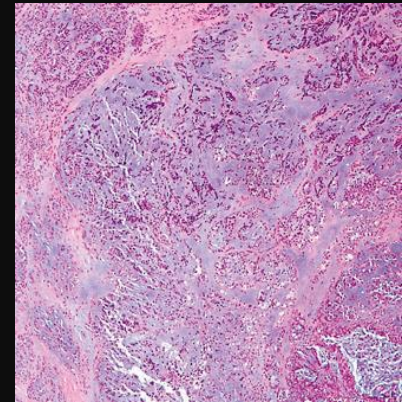
Extraskeletálny myxoidný chondrosarkóm

- lokalita, hlboké mäkké tkanivá DK
- skeletálny extraskeletálny myxoidný chondrosarkóm? 😊
- S100 (50%)
- neuroendokrinné markery +
- epitelové markery (zriedkavo, slabo, fokálne)
- fúzia NR4A3-EWSR1 alebo NR4A3-TAF15
- brachyury

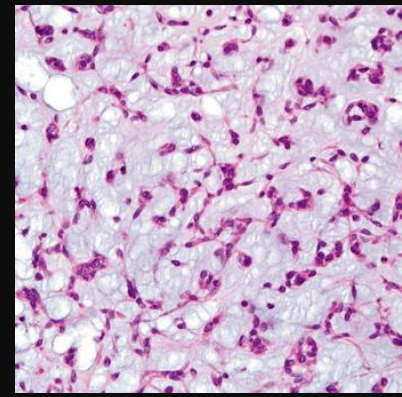
Soft tissue myoepitelióm

- lokalita
- epitelové markery + S100 + ďalšie myoepitelové markery
- niektoré strata INI1
- prestavba EWSR1 v necelých 50%

myoepitelióm s vakuolizovanými
bb = parachordóm



Lindberg 2016



Clival chordoma associated with pathological laughter

Case report

**SHIGEAKI MATSUOKA, M.D., AKIRA YOKOTA, M.D., HIDEOKI YASUKOUCHI, M.D.,
ATSUKUNI HARADA, M.D., CHITOSHI KADOYA, M.D., SHIN-ICHI WADA, M.D.,
TADAHIRO ISHIKAWA, M.D., AND SHINYA OKUDA, M.D.**

*Departments of Neurosurgery, University of Occupational and Environmental Health School of
Medicine, and Kitakyushu Chuo-Hospital, Kitakyushu, Japan*













d'akujem za pozornost'