



Çocukluk çağı yuvarlak hücreli  
tümörleri

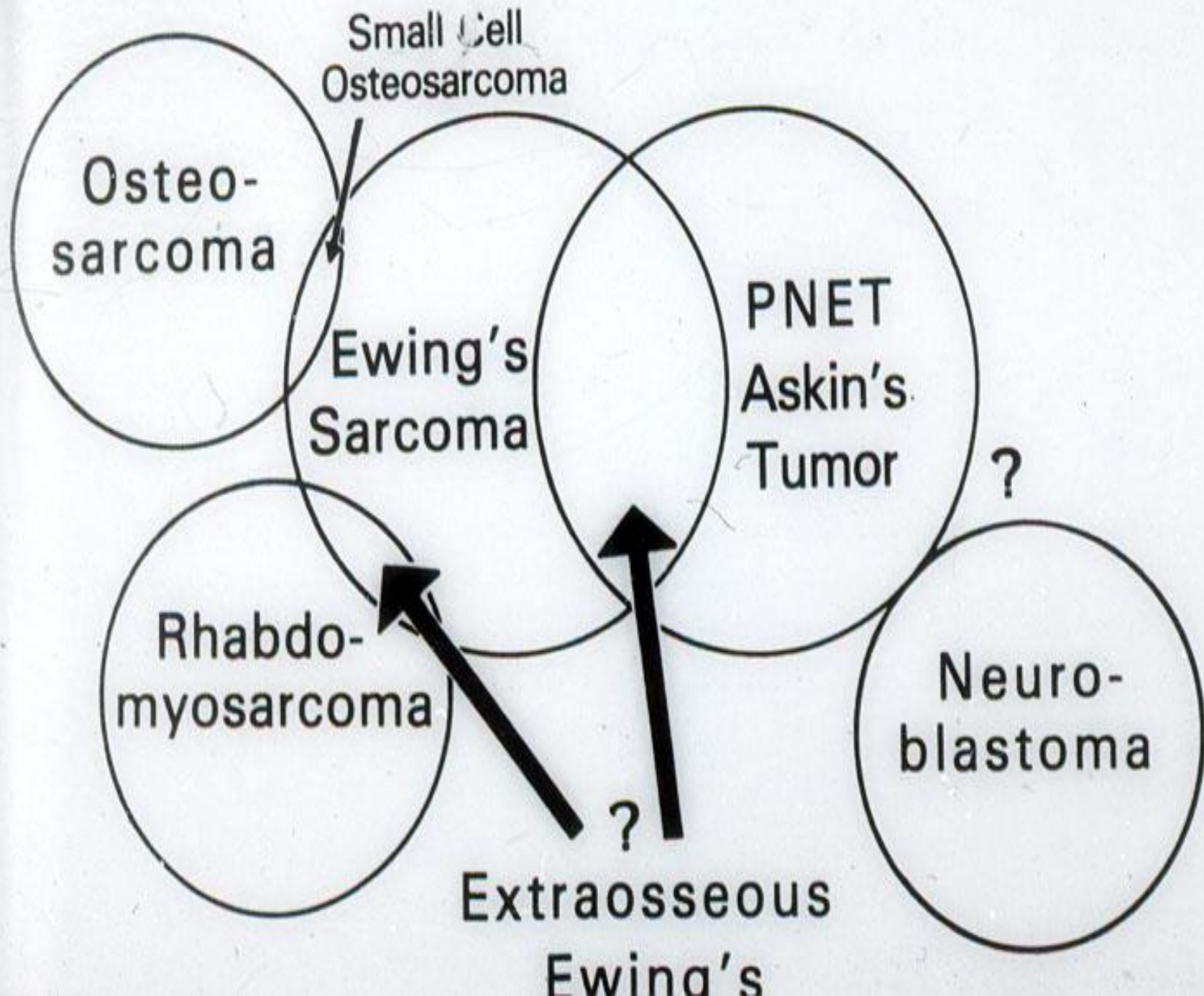
Slayt semineri

Prof. Dr. Sergülen Dervişoğlu

21. Ulusal Patoloji Kongresi

16-20 Kasım 2011, İzmir





## OLGU

- 15 yaşında erkek çocuk
- Haziran 2002 de
- Sızlama tarzı yan/sırt ağrısı, halsizlik
- 15 gün sonra skrotal ödem, sol bacakta yaygın şişlik
- US bilateral üriner nefroz
- Üre, kreatinin yüksekliği
- Rektal tuşede kitle



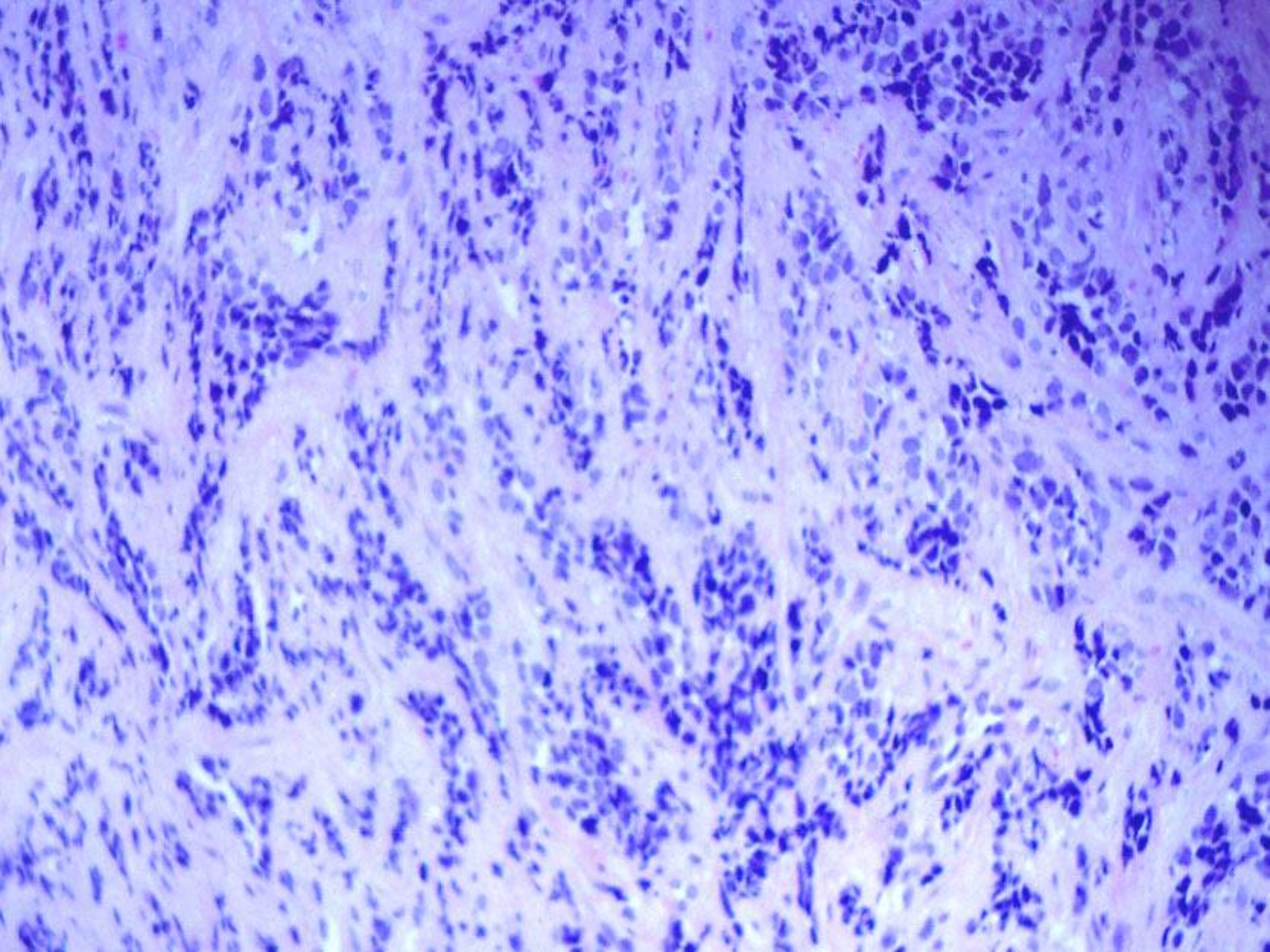
# MUAYENE BULGULARI

- Sol bacak şiş, ödemli
- Batında yaygın asit
- Sol inguinal bölgede 3X1 cm sert LAP?
- Splenomegali
- Rektal tuşede 10x10 cm kitle
- Bilateral testiküler en büyüğü 1 cm çapta kitleler
- Prostat, rektum tutulumu



- Multipl pelvik/abdominal/paraaortik LAP
- Her iki böbrekte parankim tutulumu, hidronefroz
- Mesane duvarında kalınlaşma --tutulum
- Akut böbrek yetmezliği--hemodializ
- Kemik iliği tutulumu %47 patolojik hücre
- ( kitlesel indiferansiye hücreler )
- **Lenfoma** ön tanısı ile US de saptanan heterojen kitleden prostatik biopsi



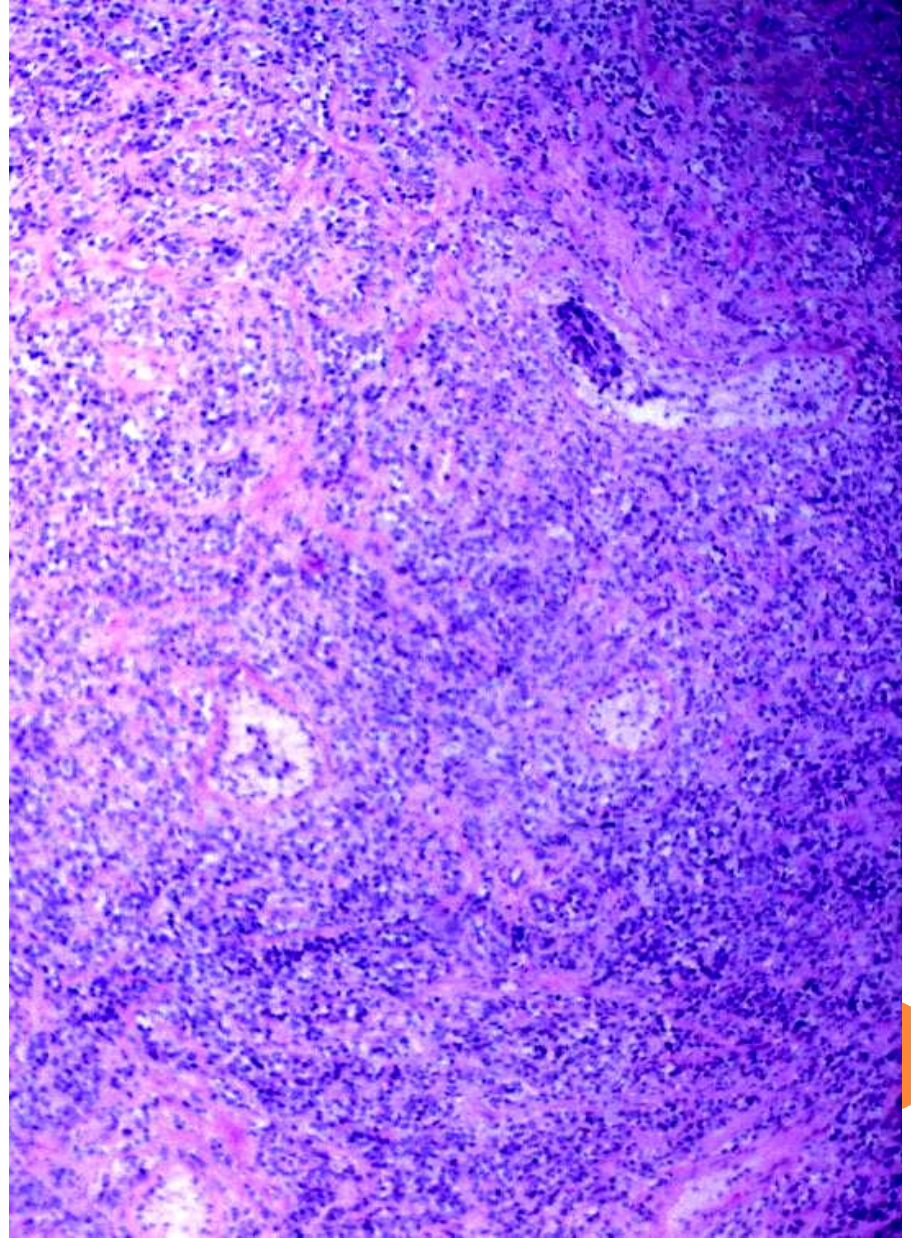


- Biopsi tanısı---Az diferansiye malign tümör---Ca?
- Revizyon---İndiferansiye malign tümör
- Sinaptofizin+/-, Desmin -, ---PNET?
- Klinik ön tanı lenfoma/Lösemik tutulum??
- Anemi, trombositopeni
- Granülositik sarkom??

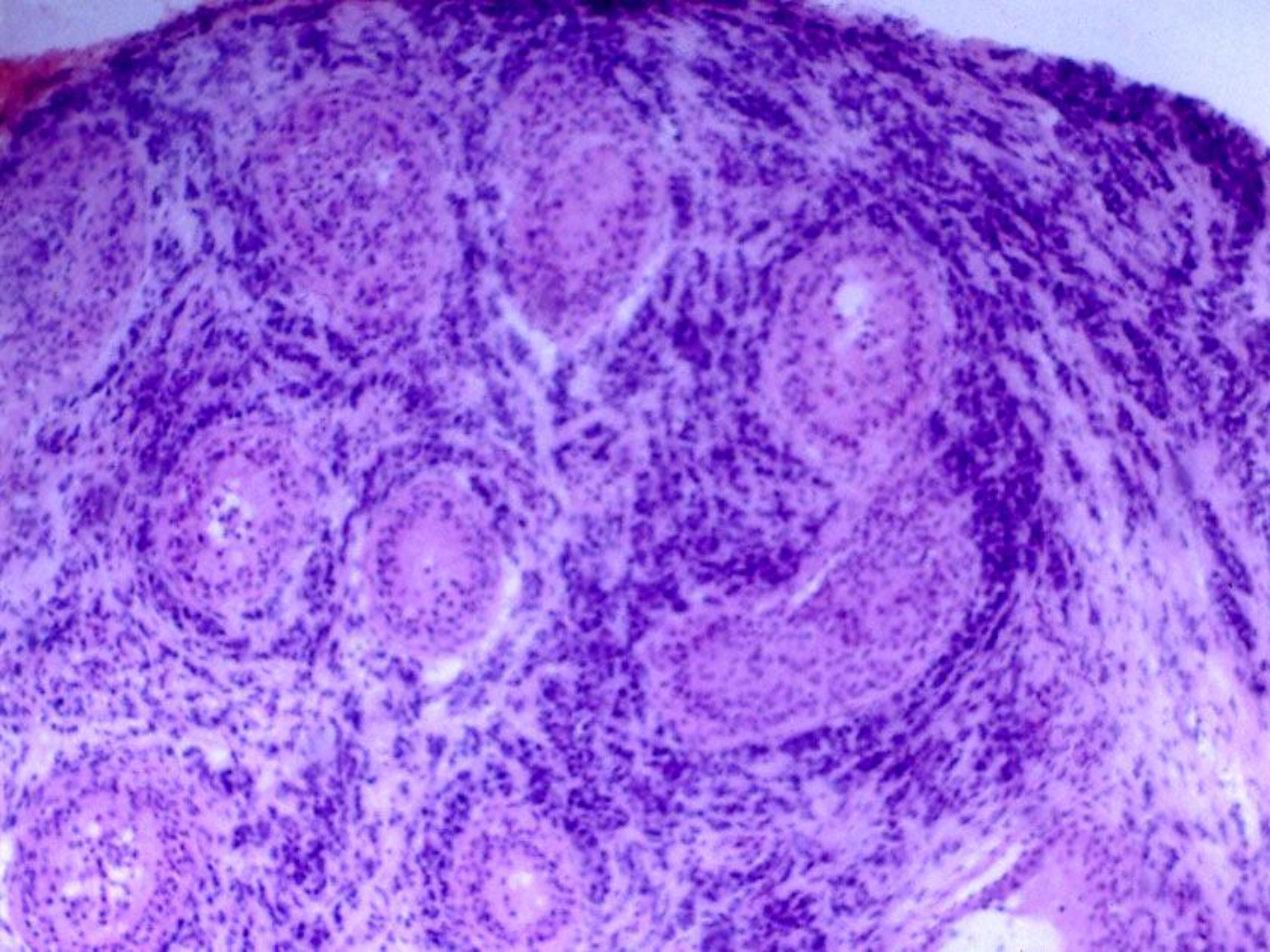


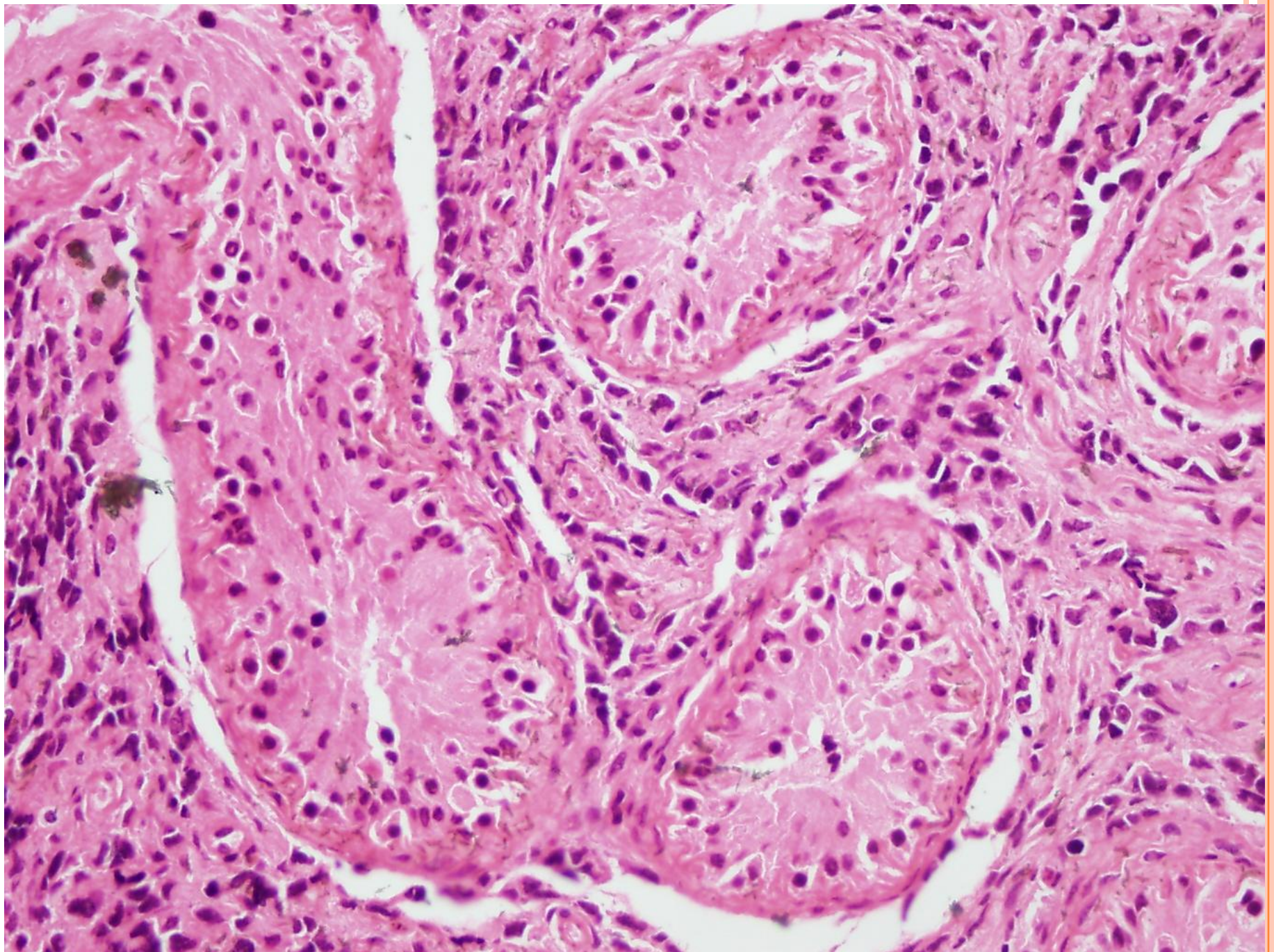
# Bx REVİZYONU VE BİLATERAL ORŞİEKTOMİ

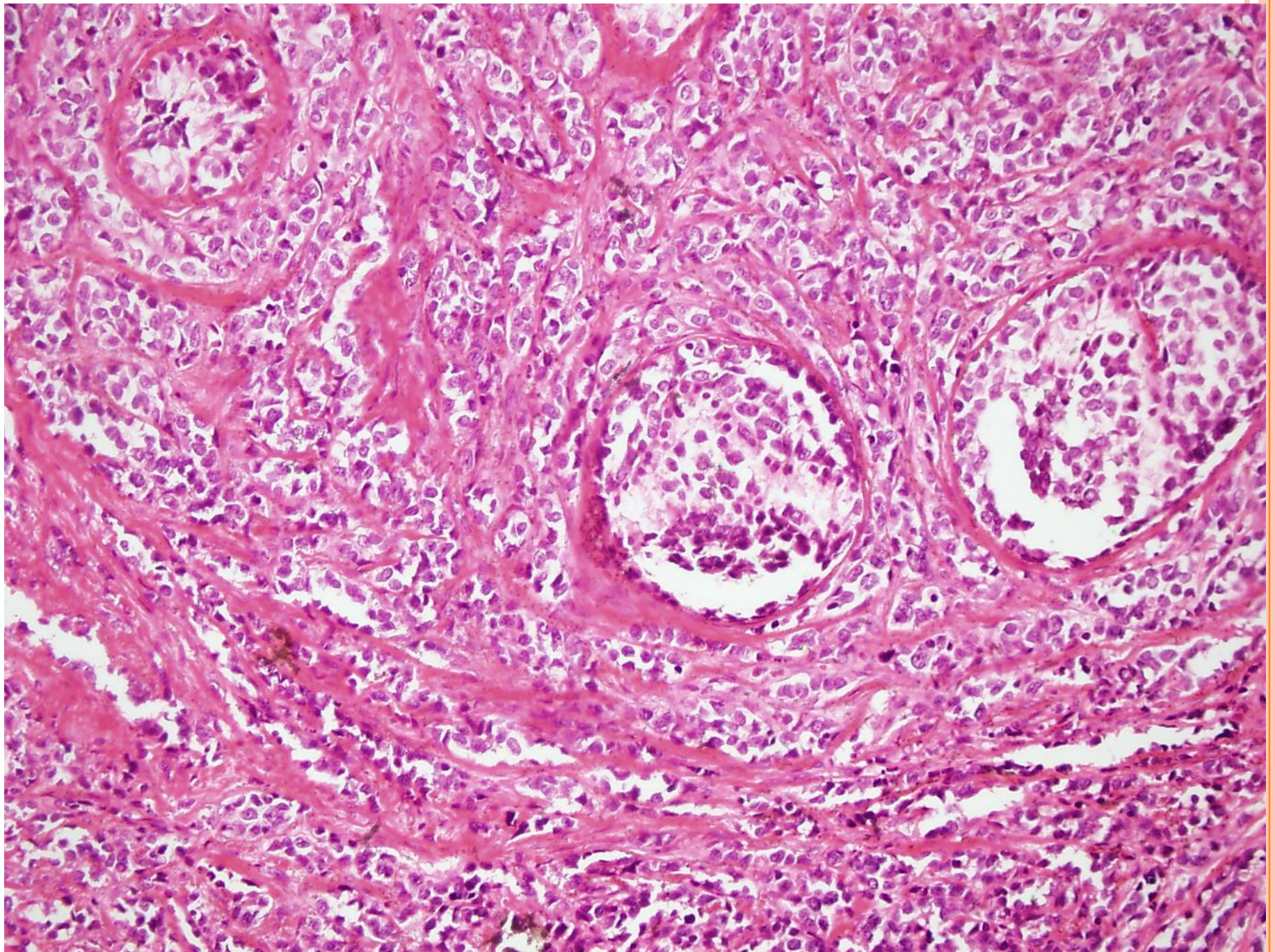
- İndiferansiye malign tümör
- Fokal alveoler dizilim
- İntertübüler alanda infiltratif yayılım
- RMS? PNET?  
Lenfoma?  
Granülositik sarkom?

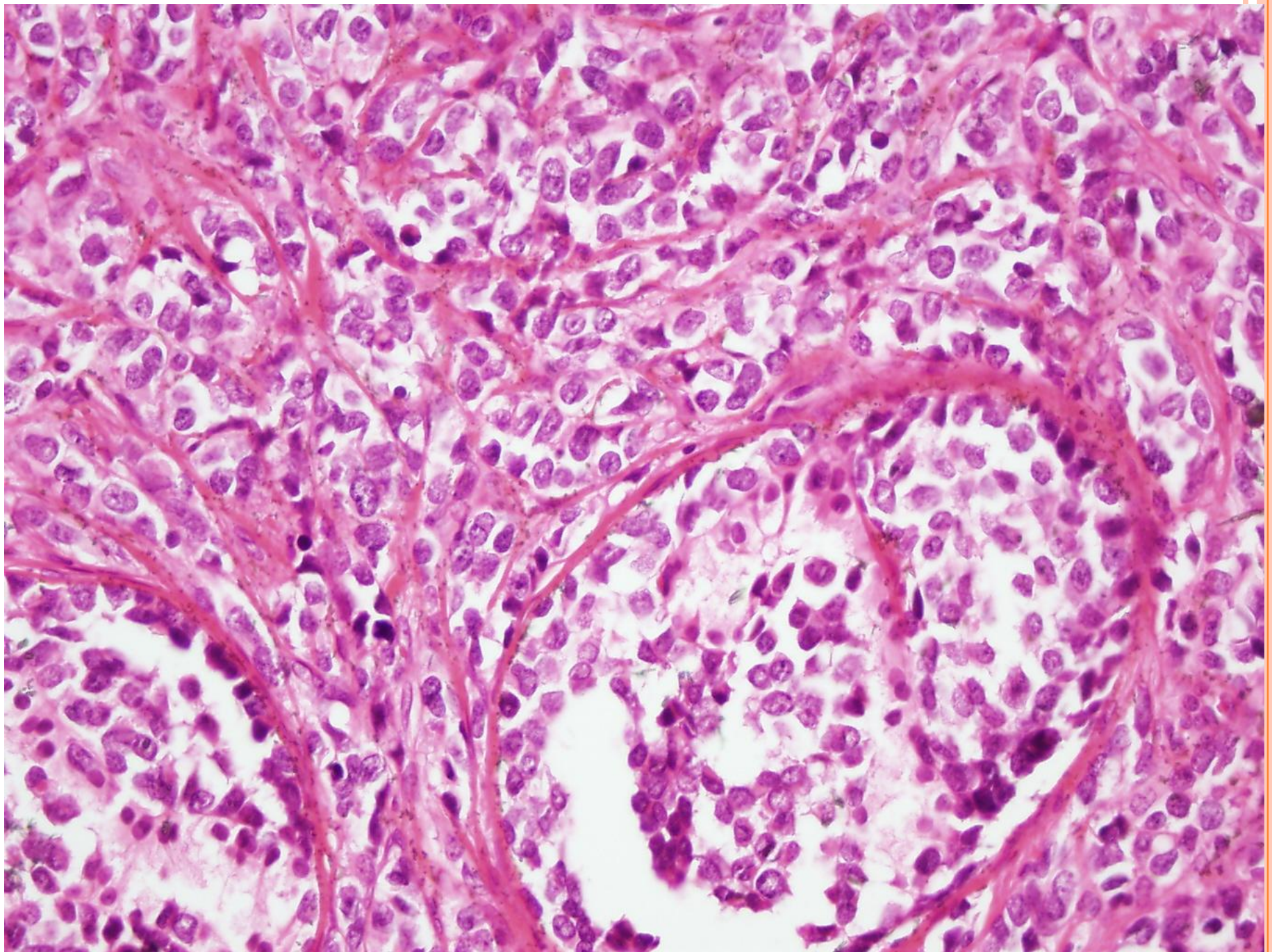


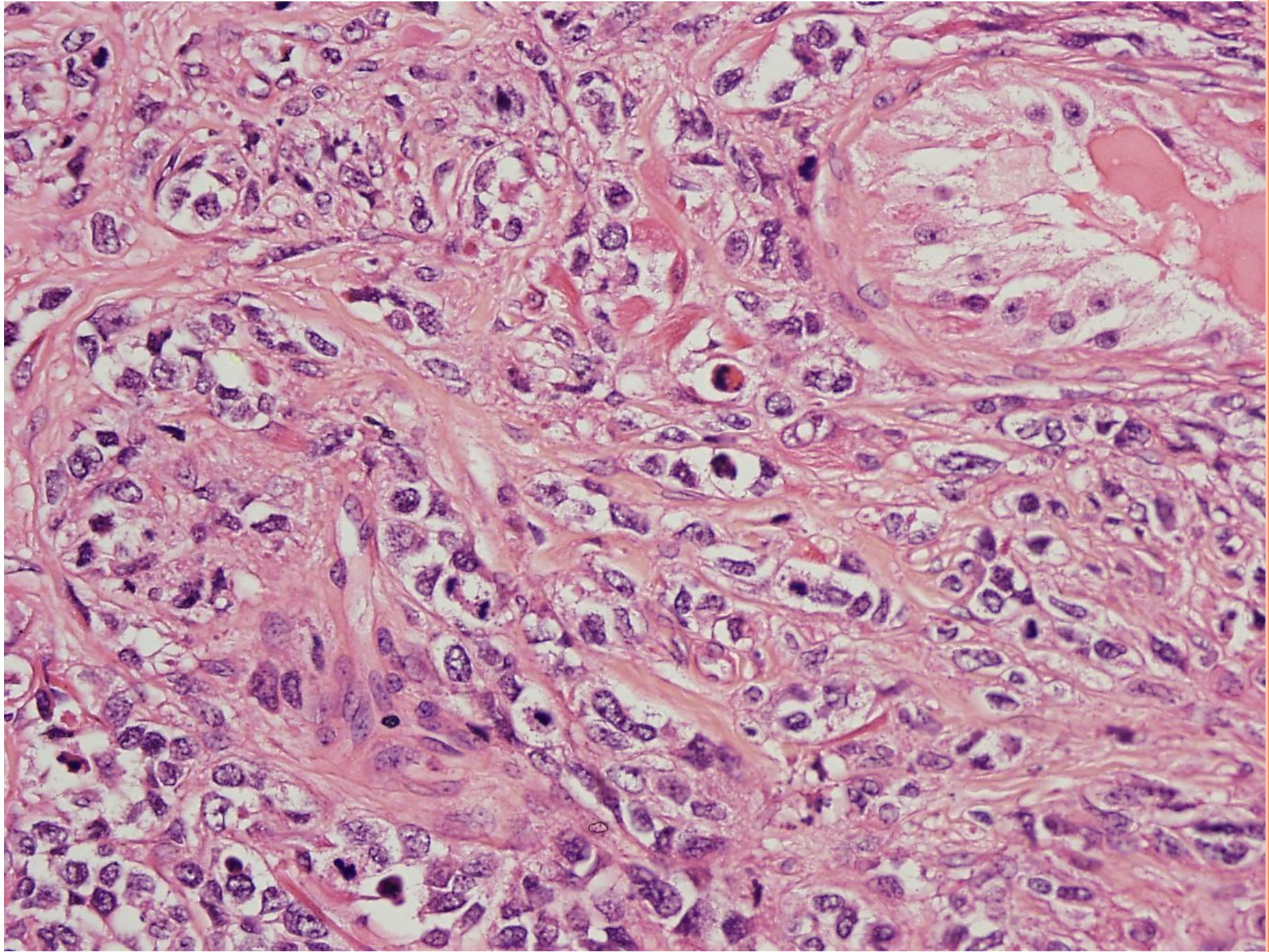


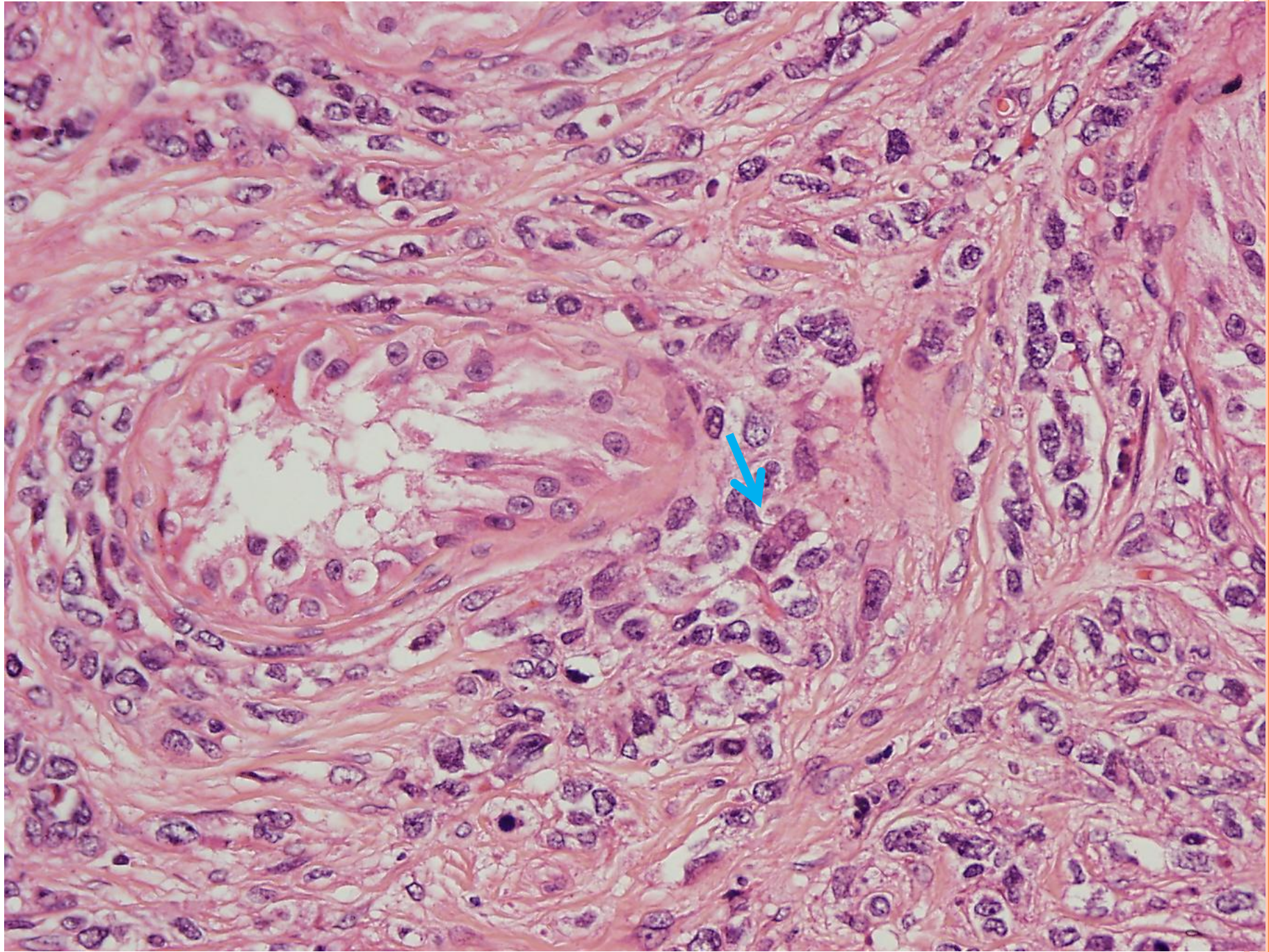










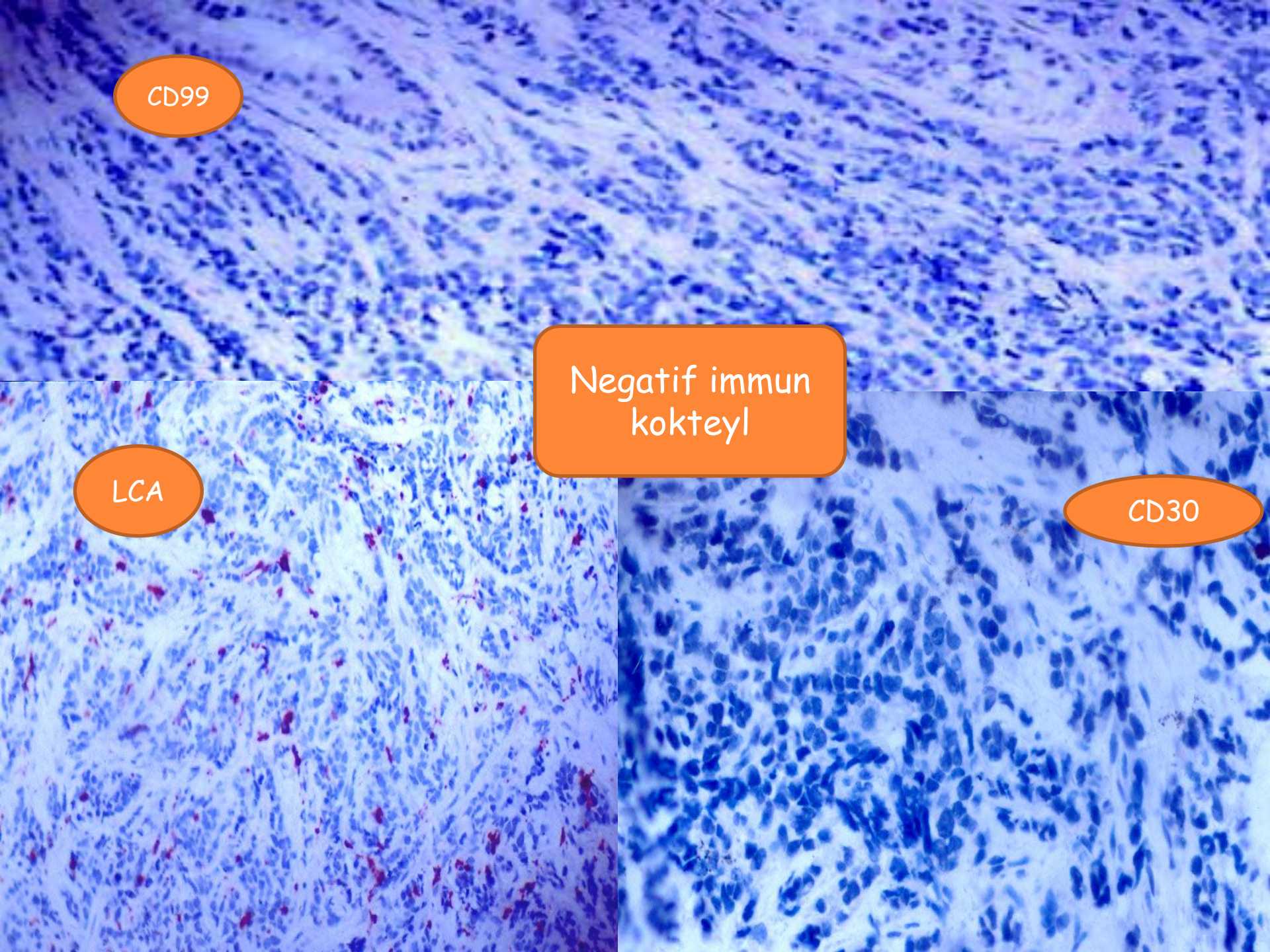


CD99

Negatif immun  
kokteyl

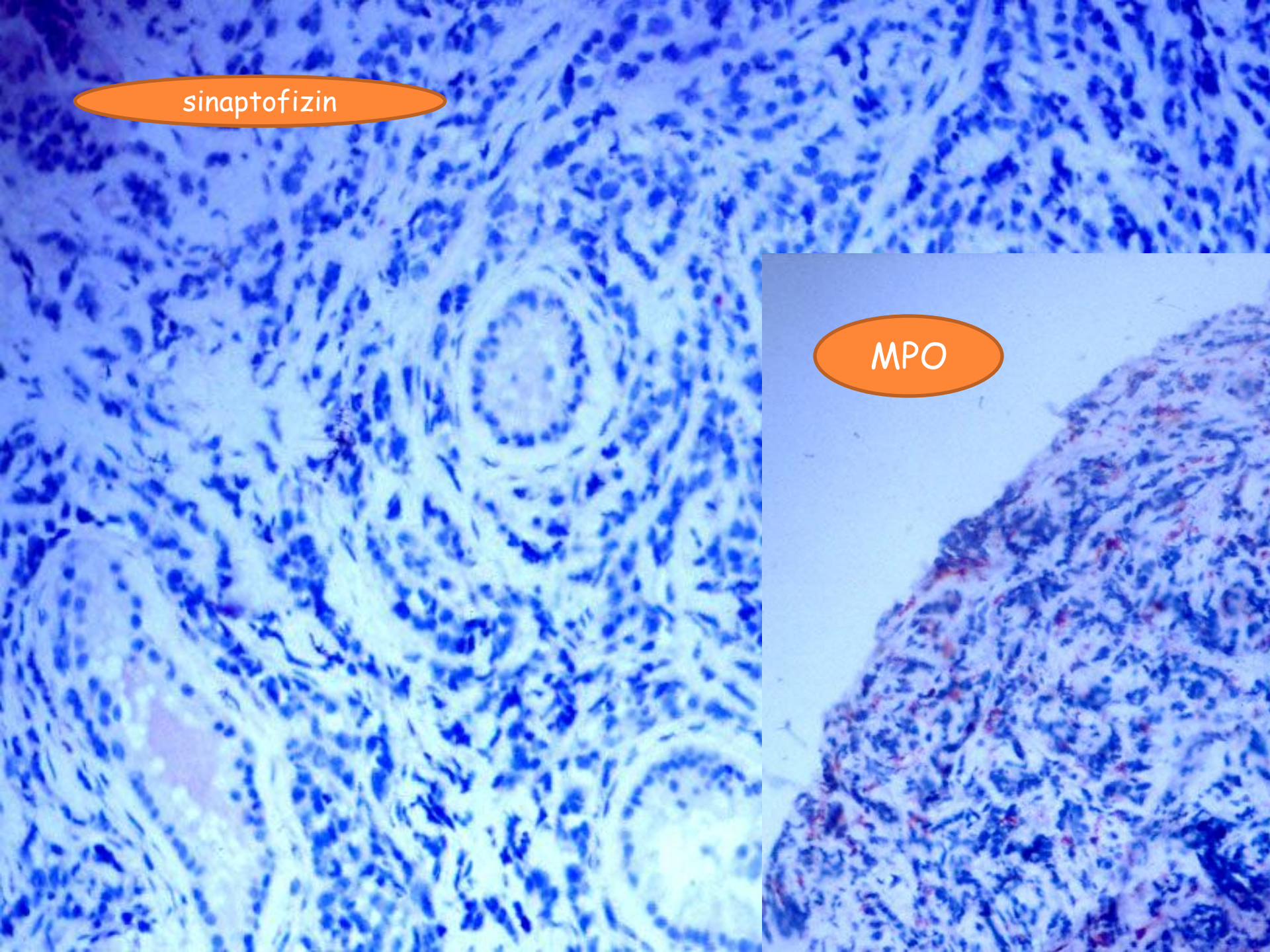
LCA

CD30



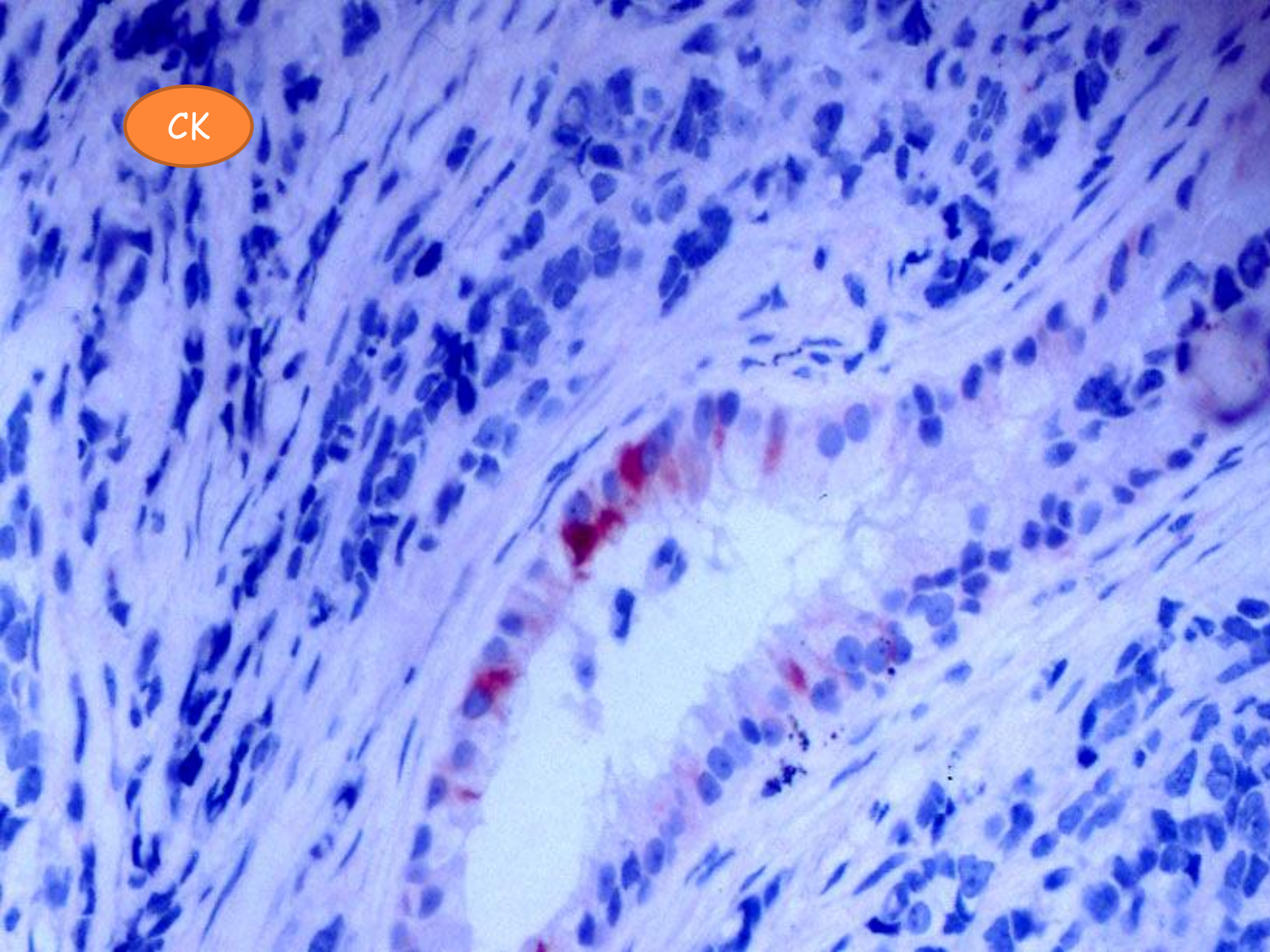
sinaptofizin

MPO





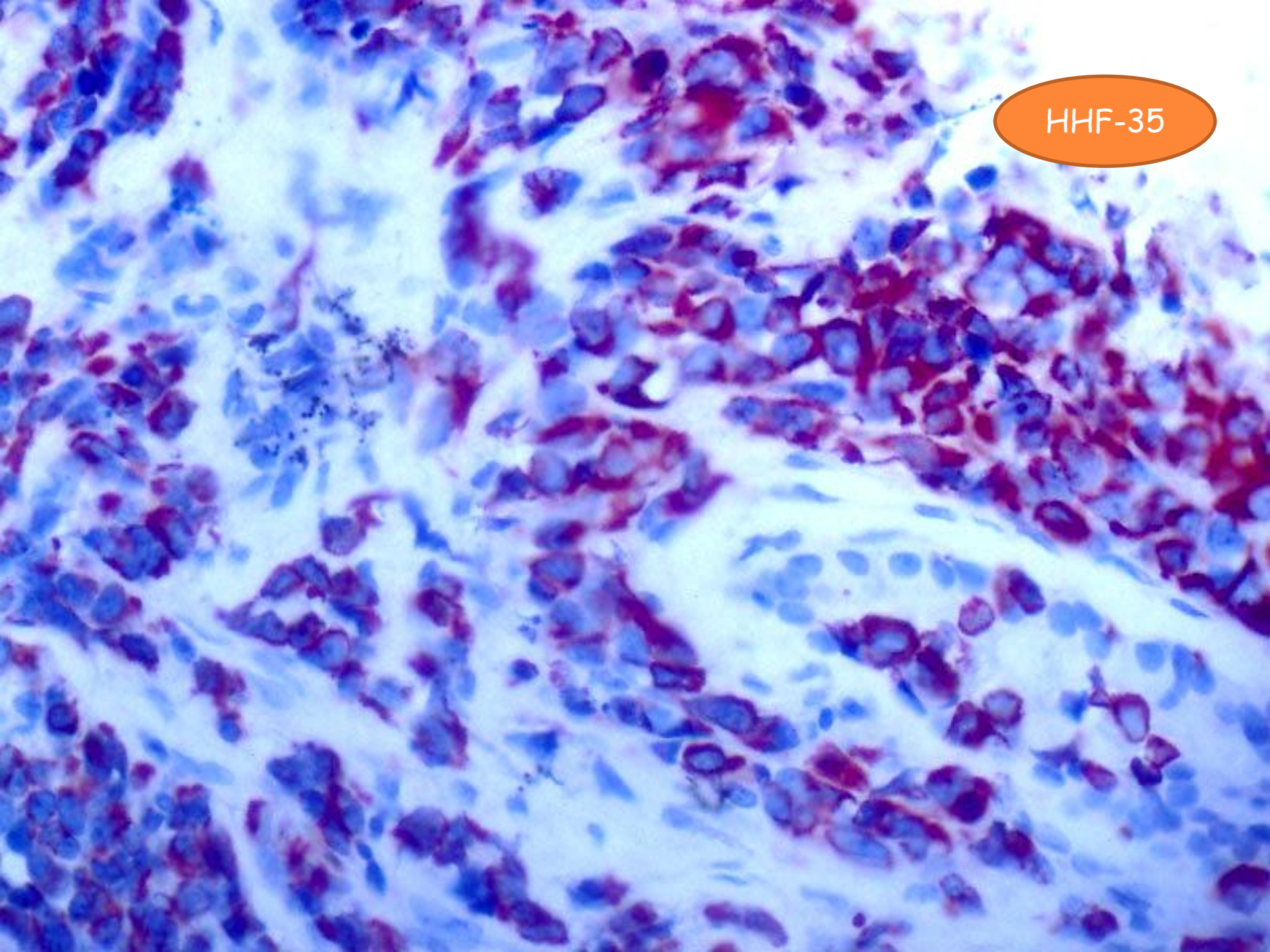
CK

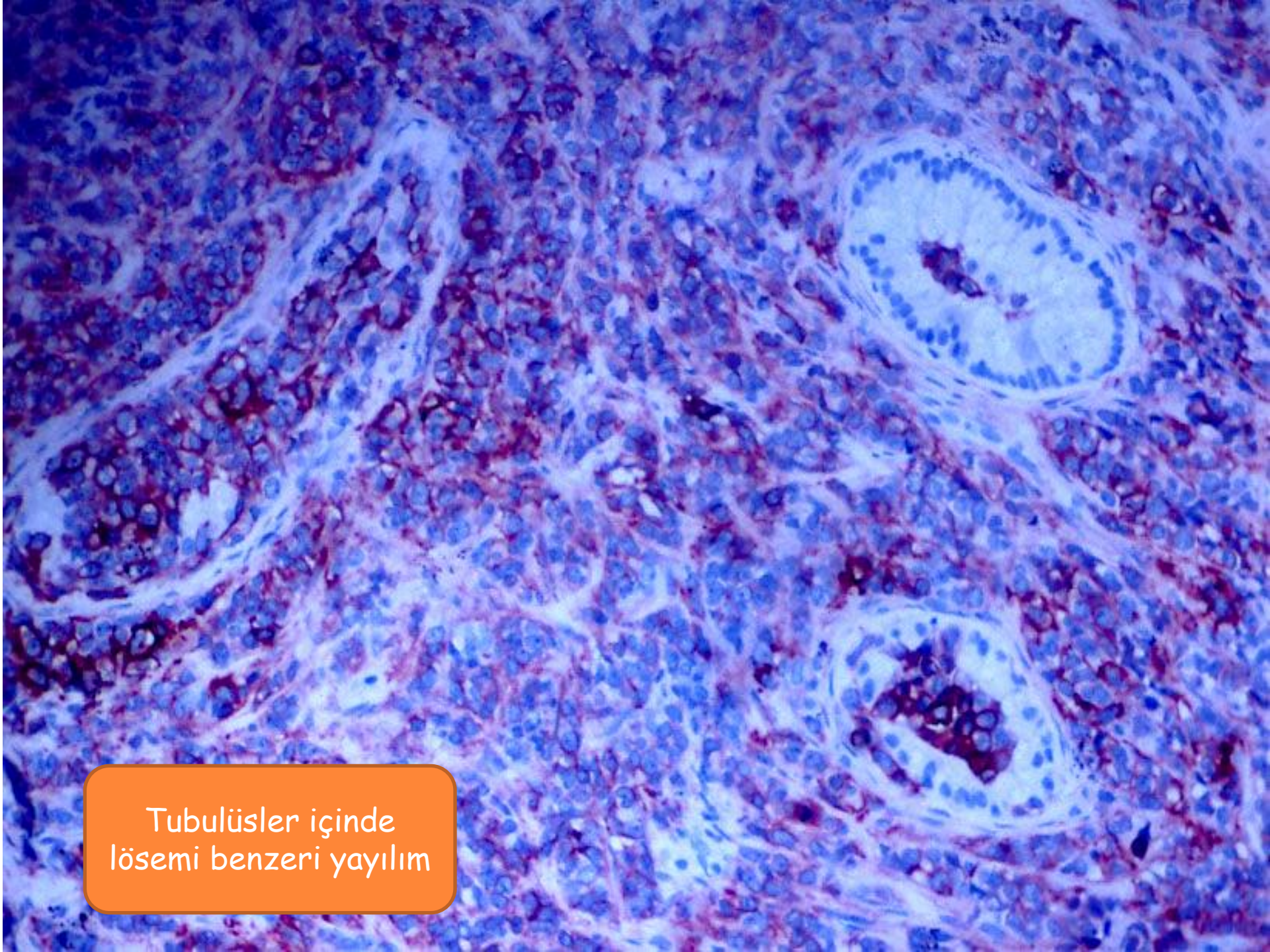


A histological slide showing muscle tissue stained for Desmin. The tissue is stained with hematoxylin and eosin (H&E), showing a dense network of muscle fibers. The nuclei are stained blue, and the cytoplasm and extracellular matrix are stained pink. The overall appearance is that of a highly cellular, fibrous tissue.

Desmin

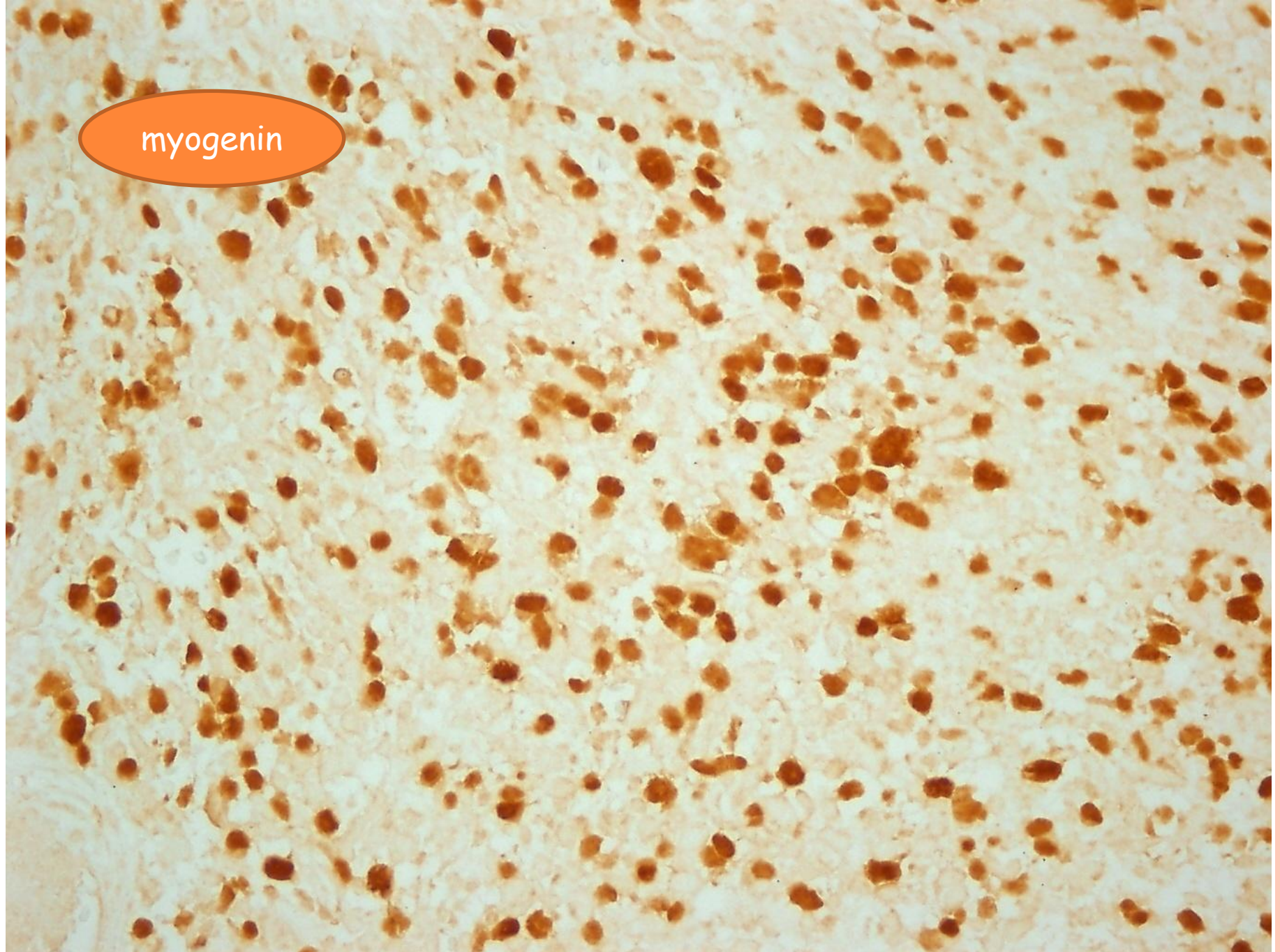
HHF-35

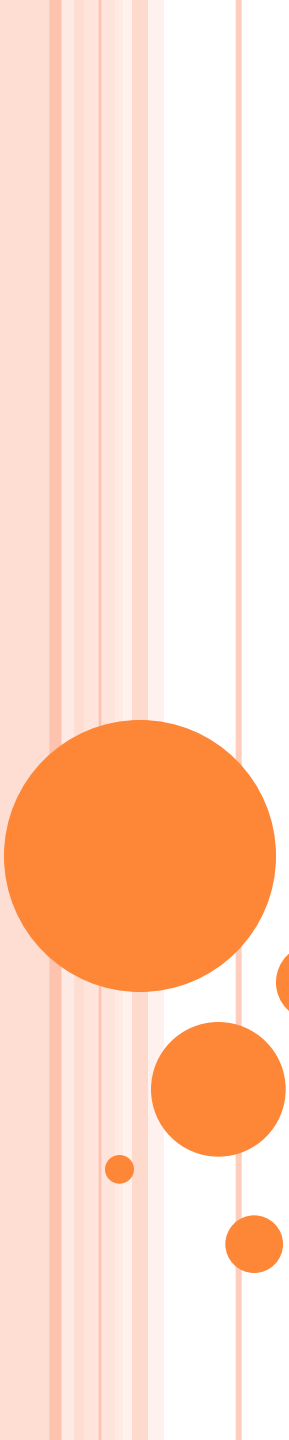




Tubulüsler içinde  
lösemi benzeri yayılım

myogenin

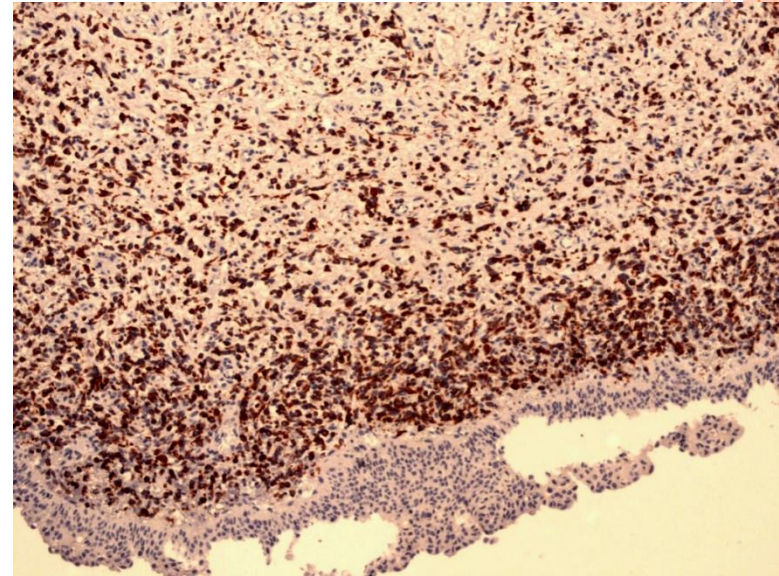




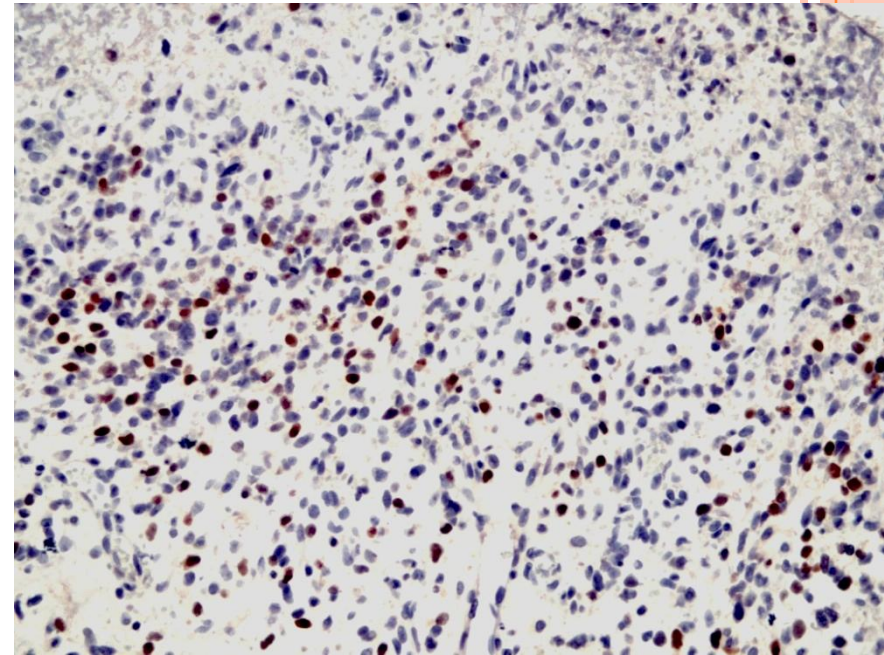
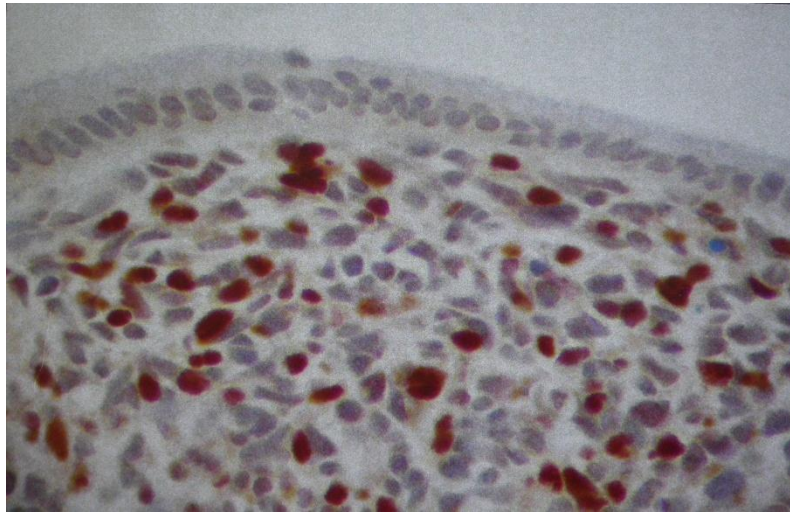
LÖSEMİ BENZERİ YAYILIM  
GÖSTEREN EMBRİYONEL  
RABDOMYOSARKOM

# RABDOMYOSARKOM TANISINDA İMMUNHİSTOKİMYA

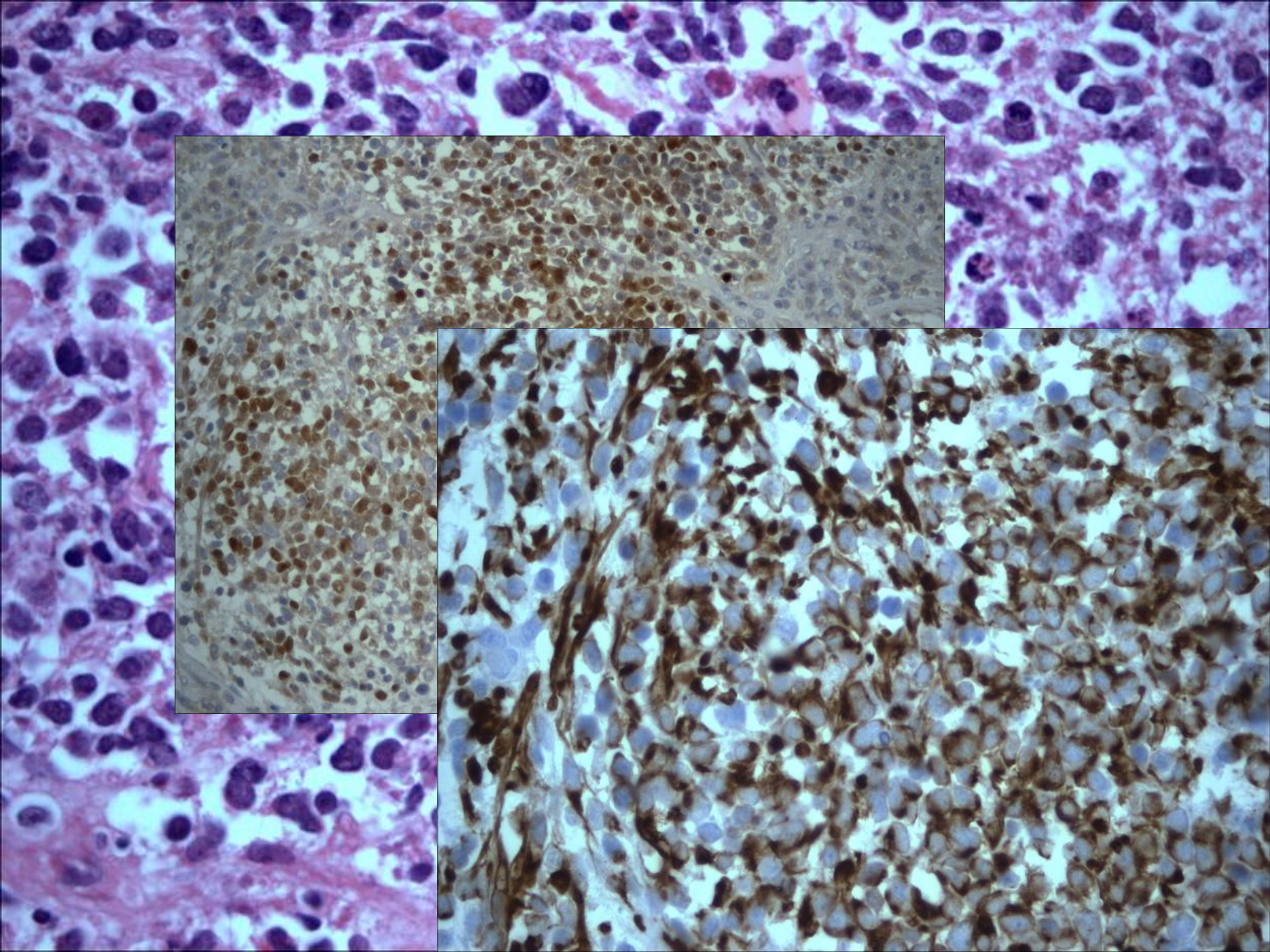
- Desmin
- HHF-35, kasa özgü aktin
- Düz kas, kalp kası, myofibroblast,
- Perisit, myoepitel, perisit
- Myozin
- Myoglobin
- Kreatin kinaz
- İskelet kası farklılaşması



- Myogenin
- Myo-D1
- Nükleer boyanma
- RMS için yüksek sensitivite ve spesivite

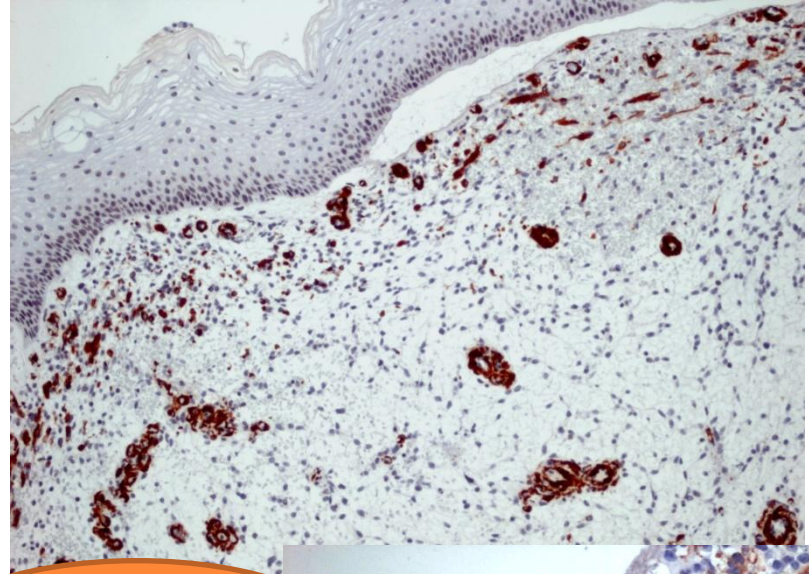




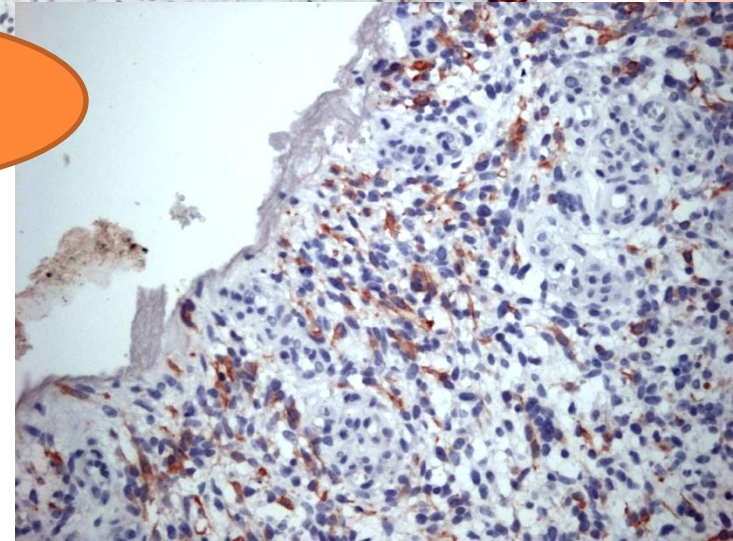


# RABDOMYOSARKOMDA ABERAN POZITIVİTE

- Sitokeratin
- S-100
- Nörofilaman
- CD 20
- SMA ( düz kas aktin )
- İmmunglobülinler
- NSE



SMA  
Sarkomerik  
aktin



# İMMUNHİSTOKİMYA YORUMUNDA TUZAKLAR

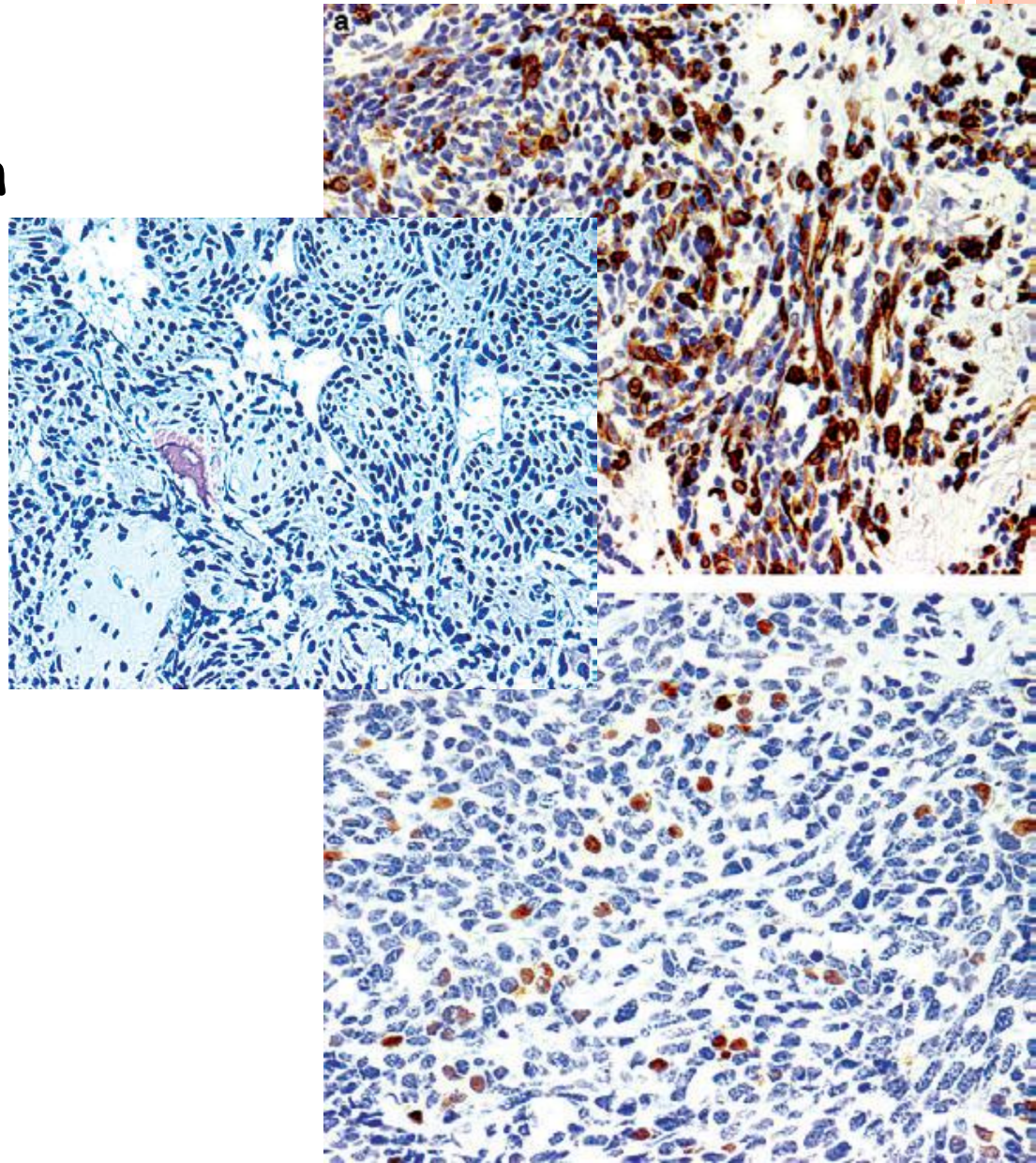
- Arada kalmış **atrofik tümör dışı iskelet kasında** yalancı pozitiflik
- Desmin, HHF35 + stromal **reaktif myofibroblastlar**/myofibroblastik tümör
- Rabdomyoplastik **heterolog** eleman
- **İndiferansiye tümör**/rabdomyoplastik diferansiasyon azlığı
- **İskelet kasını invaze etmiş nonmüsküler** tümör hücrelerinde difuzyonla tutulum



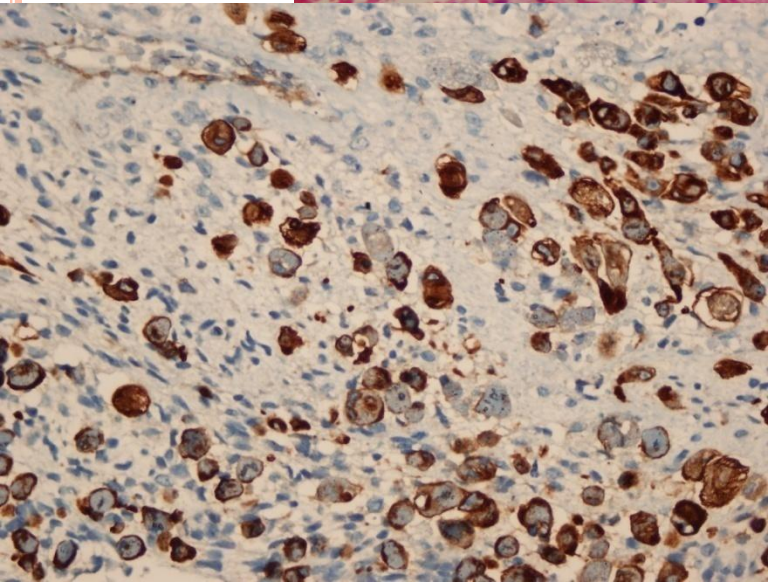
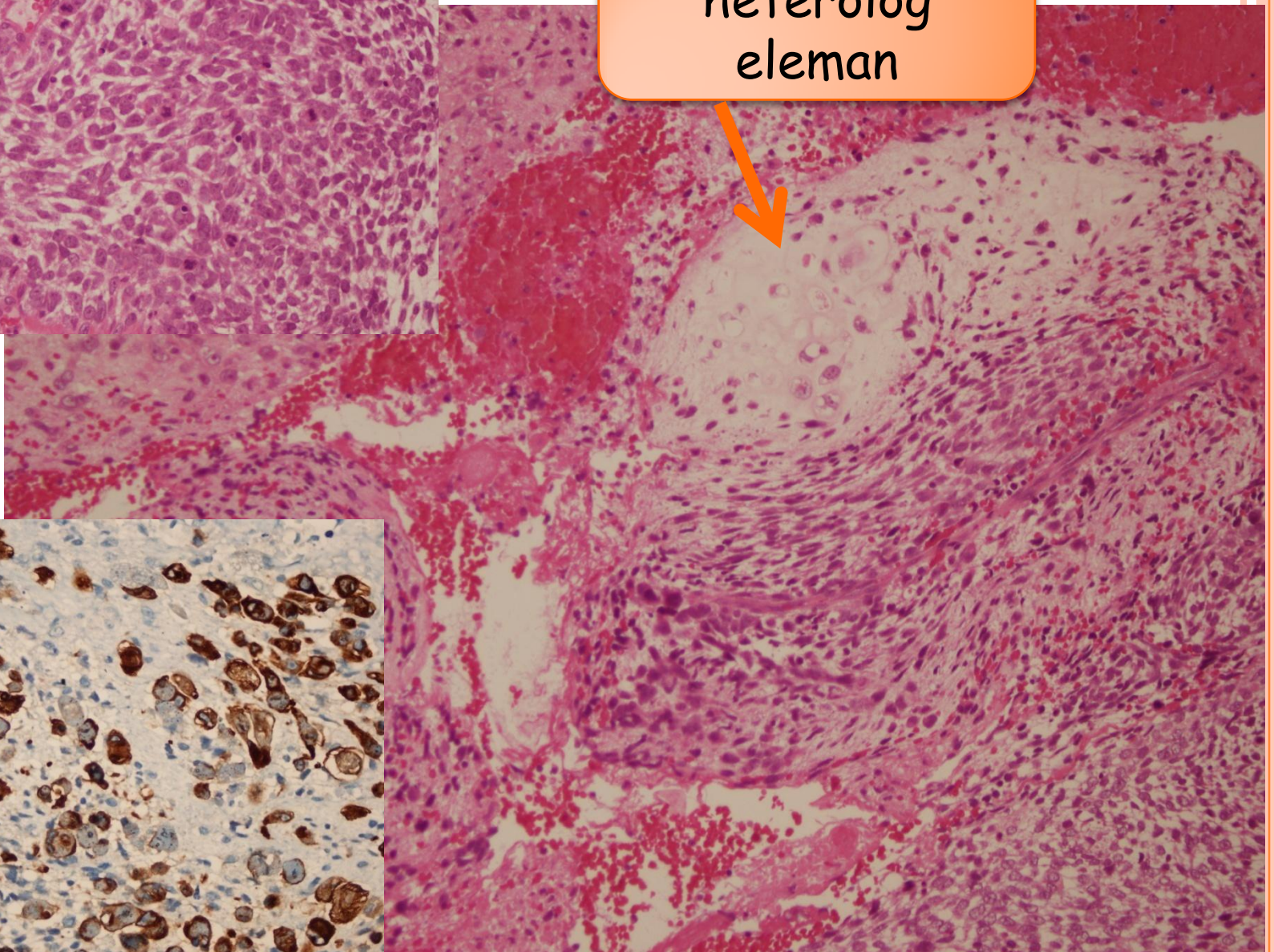
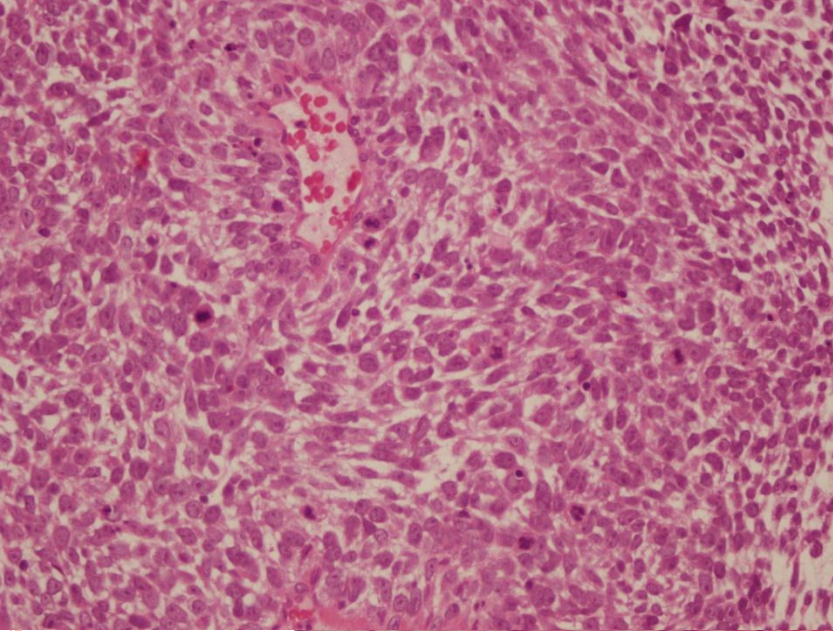
- Desmin and myogenin reactivity in mesenchymal chondrosarcoma: a potential diagnostic pitfall.

Gengler C, Letovanec I, Taminelli L, Egger JF, Guillou L.

Histopathology. 2006  
Jan;48(2):201-3.



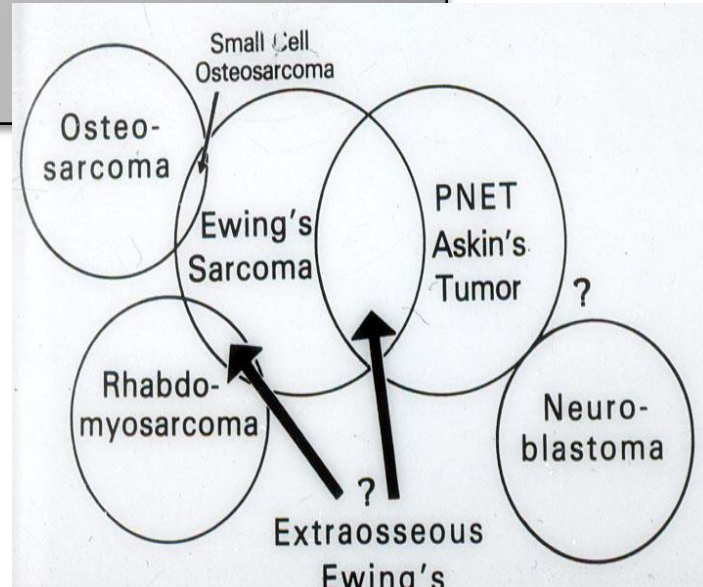
RMS de heterolog eleman



Hoang et al. 21 mezenkimal KS  
8 olguda fokal/yaygın desmin pozitivitesi  
2 olguda fokal myoD1 pozitivitesi

Mezenkimal kondrosarkomda  
bulunan stem hücrelerde  
iskelet kası yönüne farklılaşma

Polifenotipik SBRCTm  
'heterolog'  
Çizgili kas ve epitelial farklılaşma  
Ewing's sarkom / PNET  
pleuropulmoner blastom



Lösemi benzeri RMS çok nadir  
Solid tm olmaksızın kemik  
iliğinde %95 blast-like atipik  
hücreler  
Myeloid/lenfoid seri yüzey  
antijenleri ile negatif  
Hücrelerde aktin, desmin ve  
myoglobin pozitifliği

Kemik iliği örneklerinde iHK önemli  
Özellikle flow sitometri sıradışı ise

Pediatr Blood Cancer. 2009 Feb;52(2):286-7.

**Rhabdomyosarcoma masquerading as acute leukemia.**

Shinkoda Y, Nagatoshi Y, Fukano R, Nishiyama K, Okamura J.



Mod Pathol. 1988 Jan;1(1):23-8.

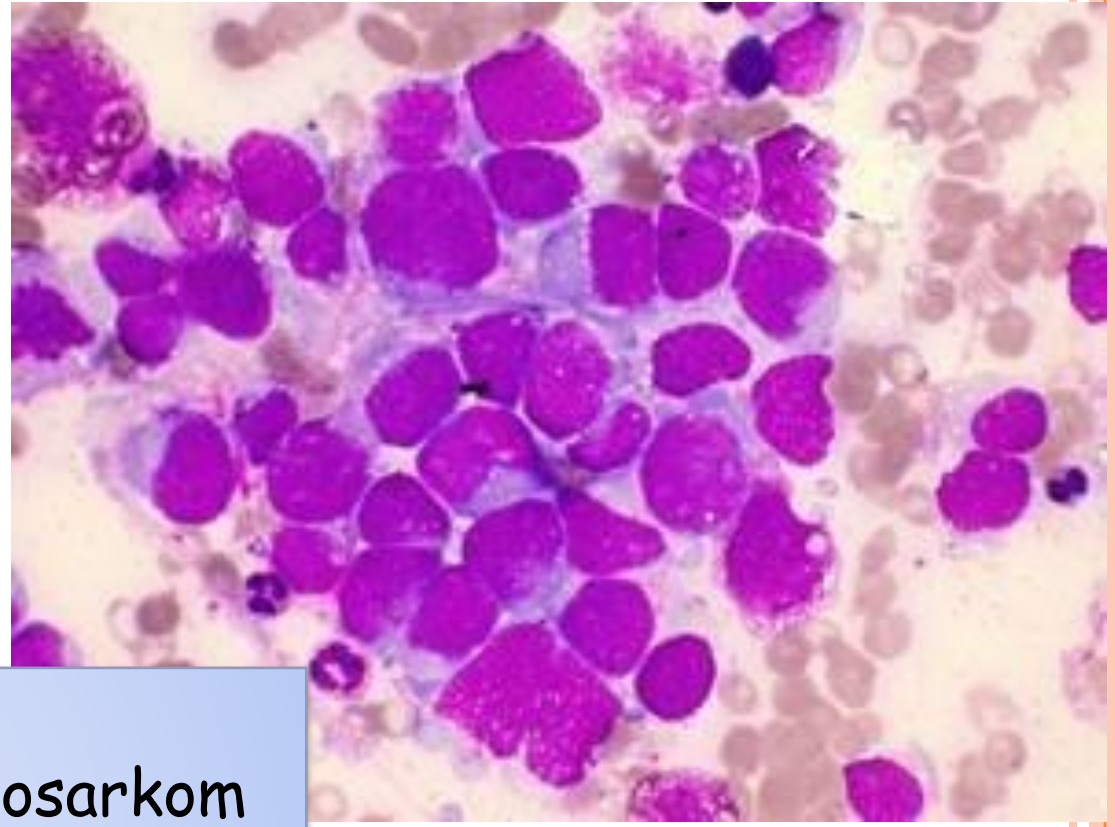
Primitive rhabdomyosarcoma presenting with diffuse bone marrow involvement: an immunohistochemical and ultrastructural study.

Cho KR, Olson JL, Epstein JI.

Yaygın kemik iliđi tutulumu  
Belirgin yumuřak doku kitlesi  
yok  
Akut lösemi ön tanısı  
İHK ve EM ile RMS







12 yaş erkek çocuk  
Kİ alveolar rhabdomyosarkom  
sitogenetik t(2;13)(q25;q14)  
Primer tümör olmaksızın

Am J Hematol. 2001 Sep;68(1):51-7.

**Hematologic masquerade of  
rhabdomyosarcoma.**

Sandberg AA, Stone JF, Czarnecki L, Cohen  
JD.



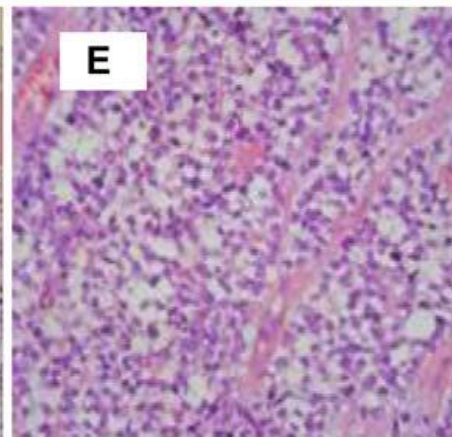
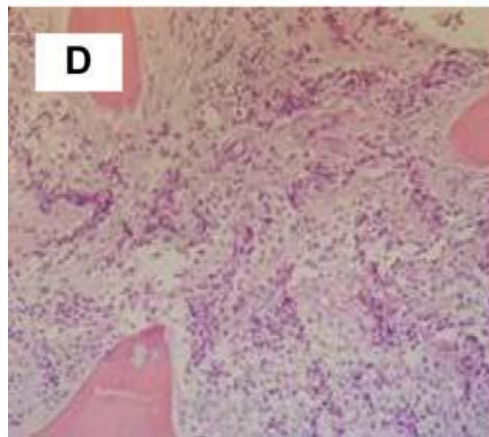
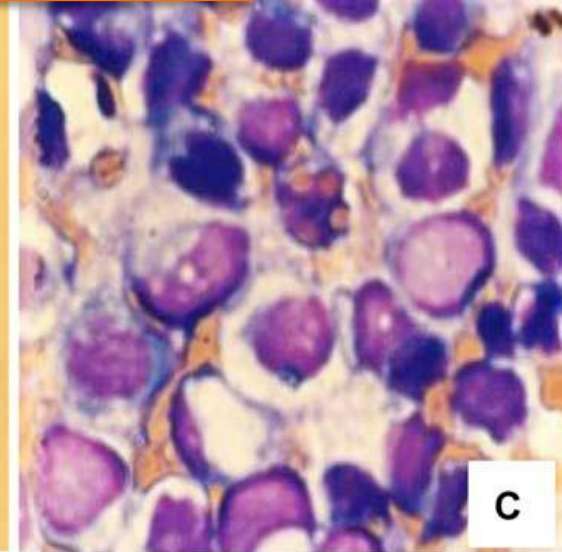
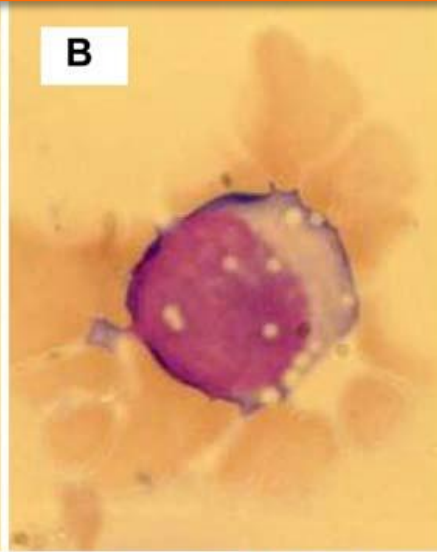
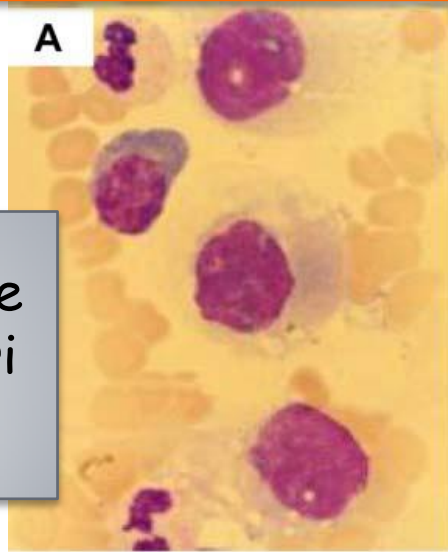
Ann Hematol (2004) 83: 729-730

## LETTER TO THE EDITOR

Rıdvan Ali . Fahir Özkalemkaş . Ülkü Ozan .  
Tülay Özçelik . Vildan Özkocaman . Gülaydan Filiz .  
Osman Manavoğlu . Ahmet Tunalı

# Rhabdomyosarcoma of the perianal region presenting as acute leukemia

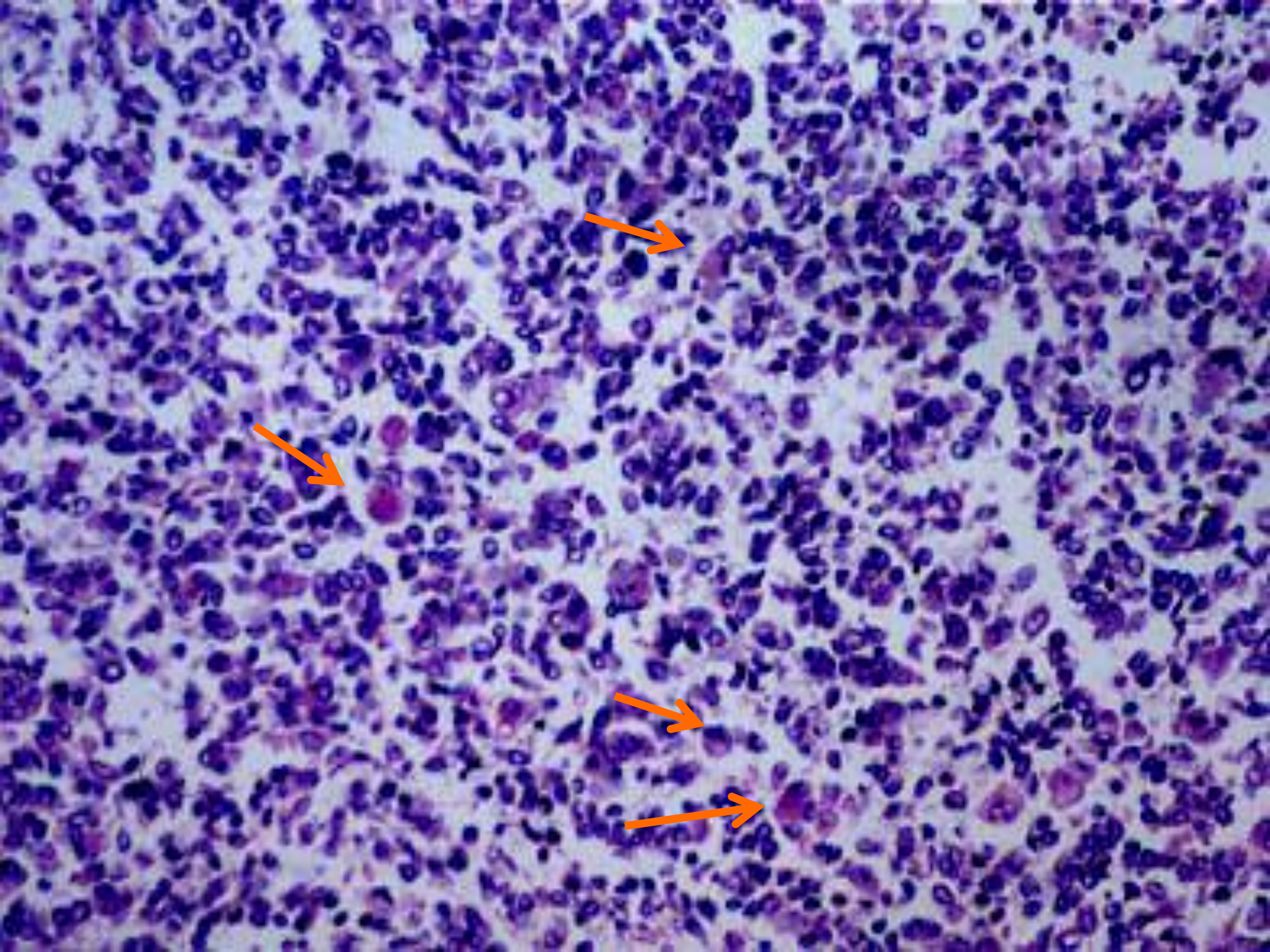
Kemik iliğinde  
blast benzeri  
hücreler

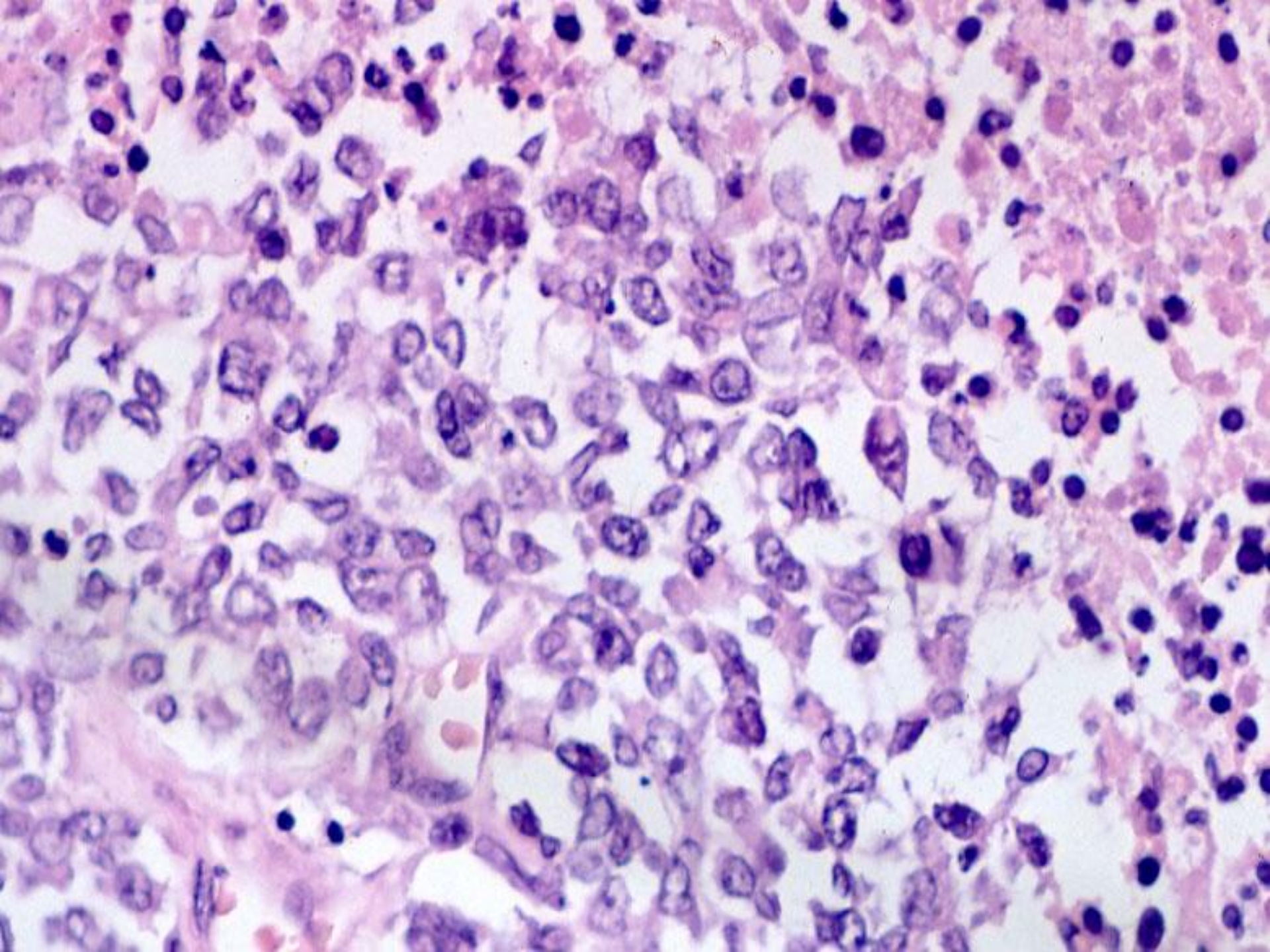


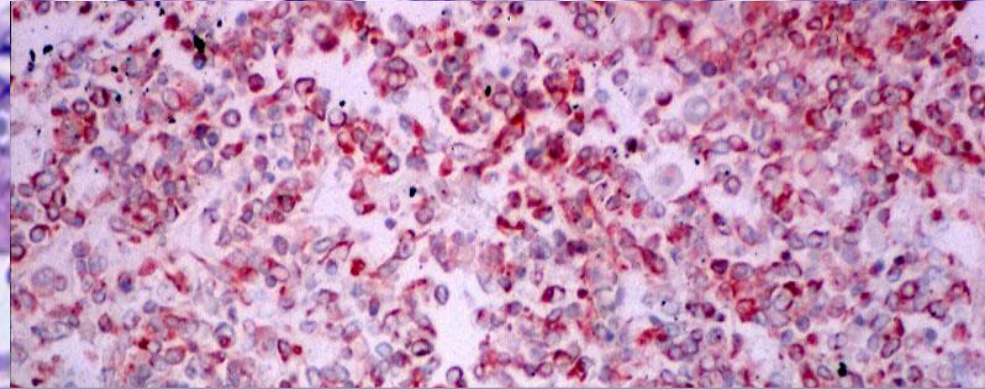
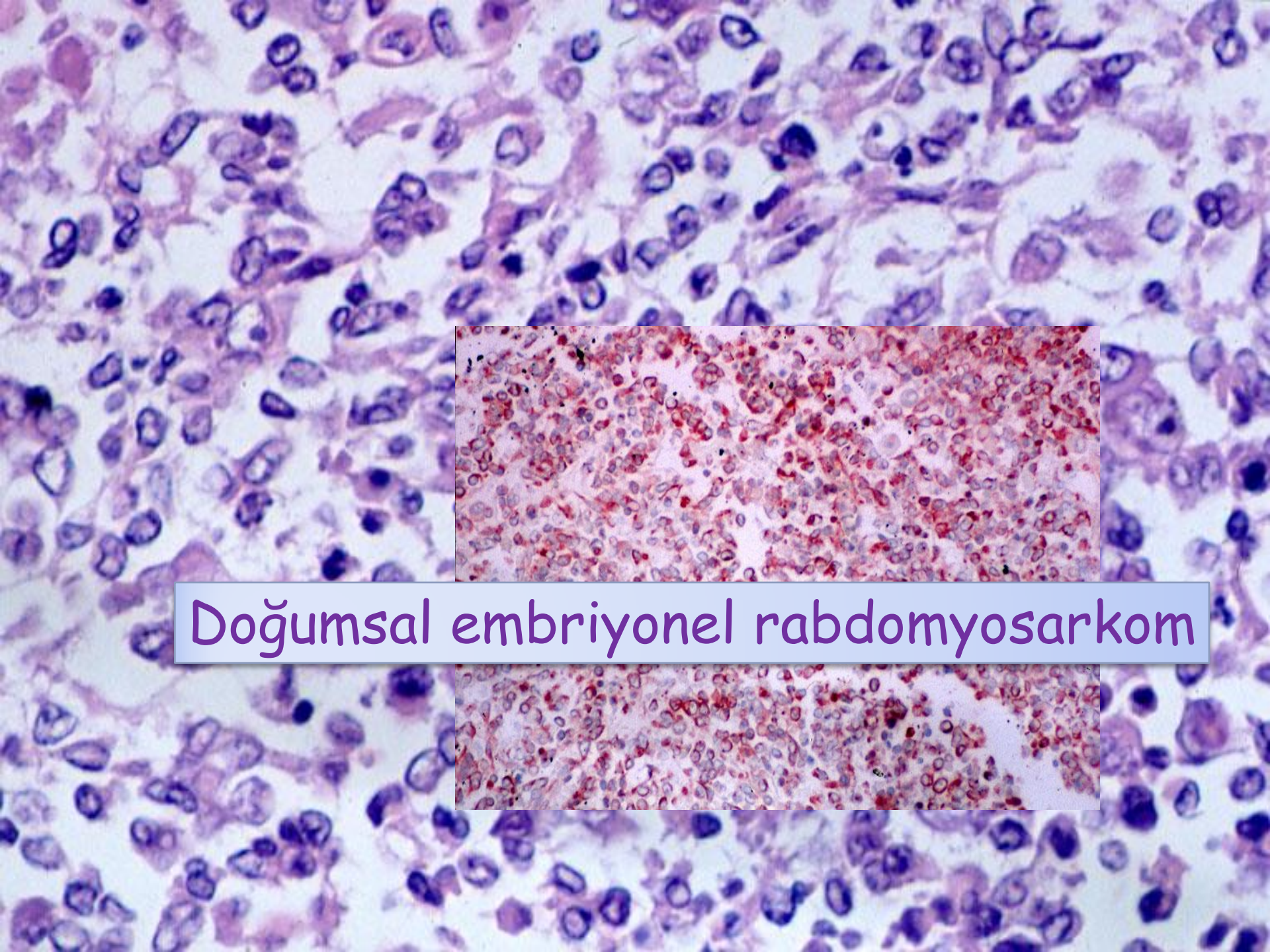




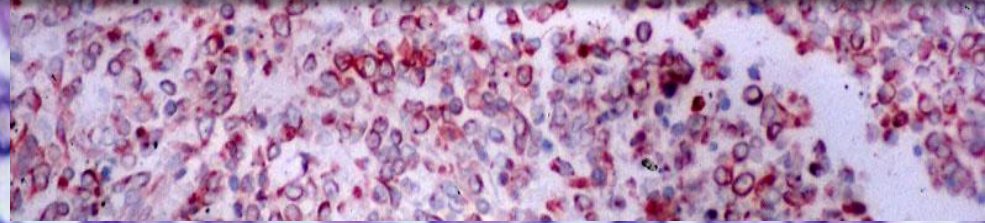




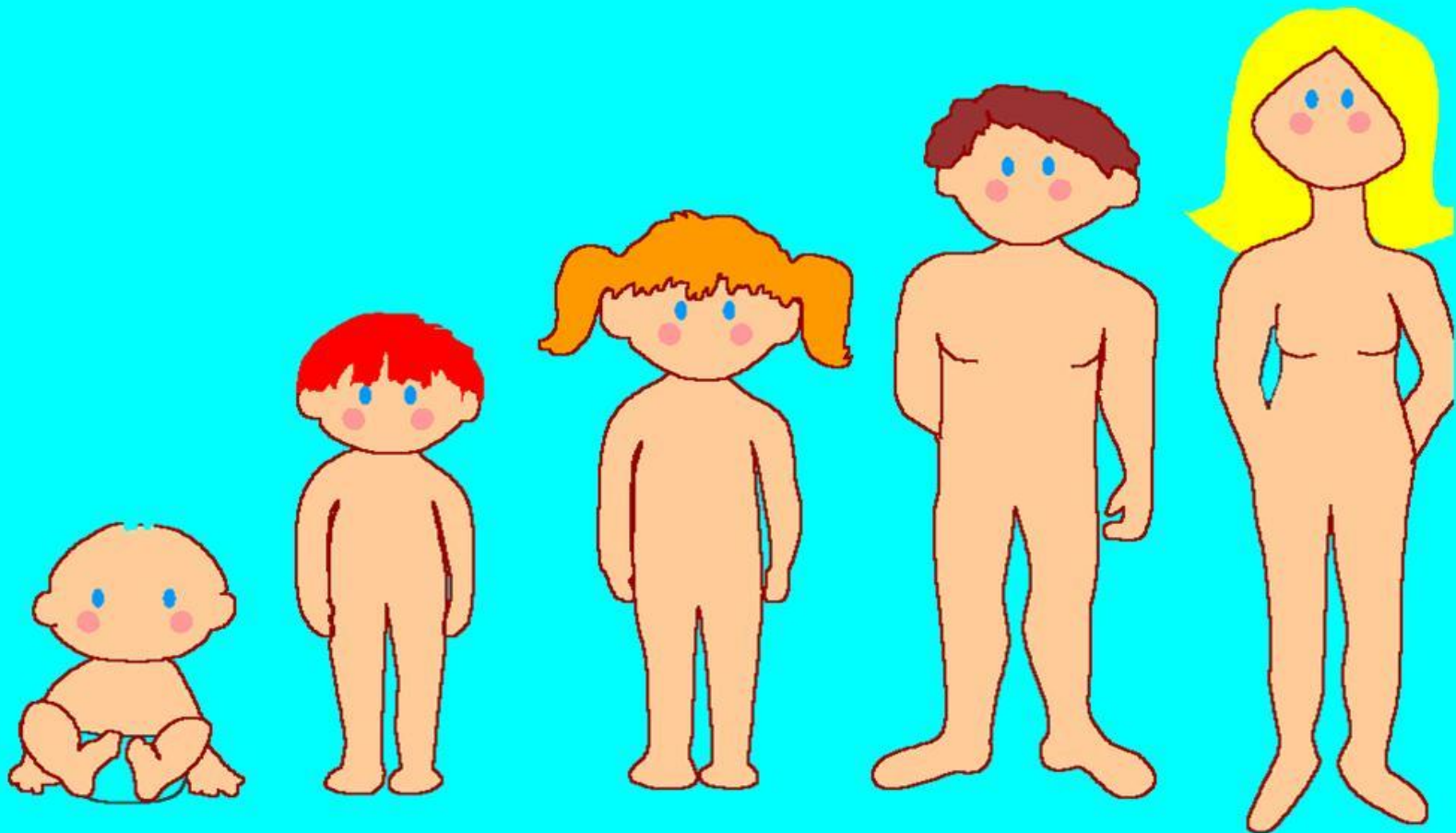




Doğumsal embriyonel rabdomyosarkom







<1  
4%

1-4  
34%

5-9  
25%

10-14  
22%

15-21  
15%

Age (in years)



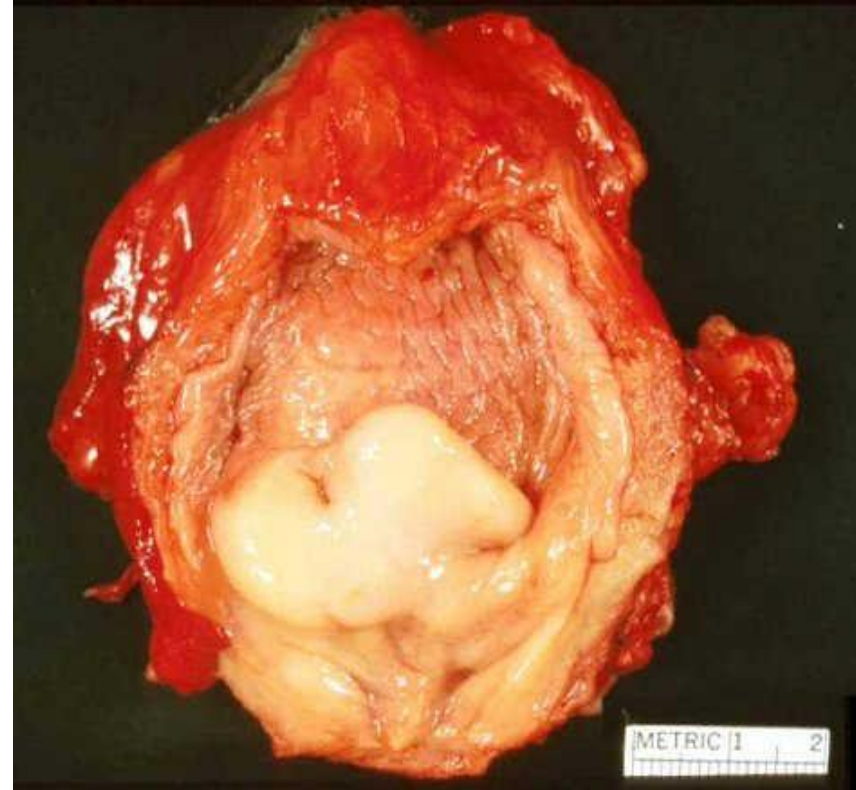
# İNFANTİL DÖNEMDE RMS

- Embriyonel ve botrioid tip sık
- İndiferansiye sarkom sık
- Ayırıcı tanıda nöroblastom, lenfoma, Ewing/PNET ailesi tümör
- Myojen farklılaşma yok---primitif yumuşak doku sarkomu, ES benzeri yumuşak doku sarkomu



# RMS İNTERNASYONAL SINIFLAMA

- En iyi prognoz
- Botrioid RMS
- İğsi hücreli RMS
- Orta prognoz
- Embriyonel RMS
- Kötü prognoz
- Alveoler RMS
- İndifferansiye sarkom
- Gidişi ?—RMS rabdoid özellikli



- Subtype and prognostic classification of rhabdomyosarcoma by immunohistochemistry.

Wachtel M, Runge T, Leuschner I, Stegmaier S, Koscielniak E, Treuner J, Odermatt B, Behnke S, Niggli FK, Schafer BW.

J Clin Oncol. 2006 Feb 10;24(5):816-22. Epub 2006 Jan 3

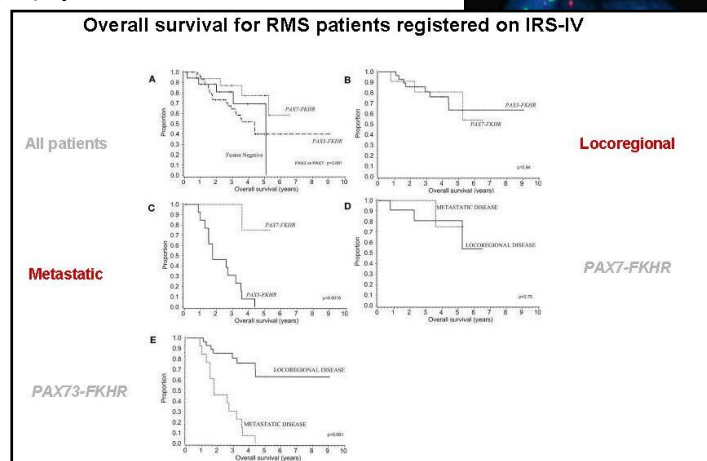
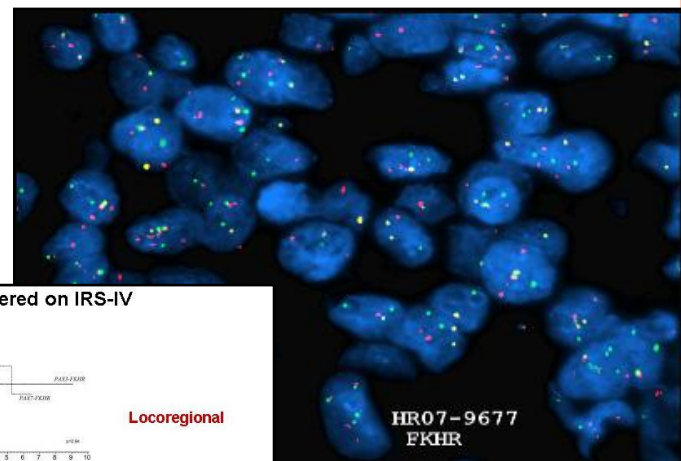
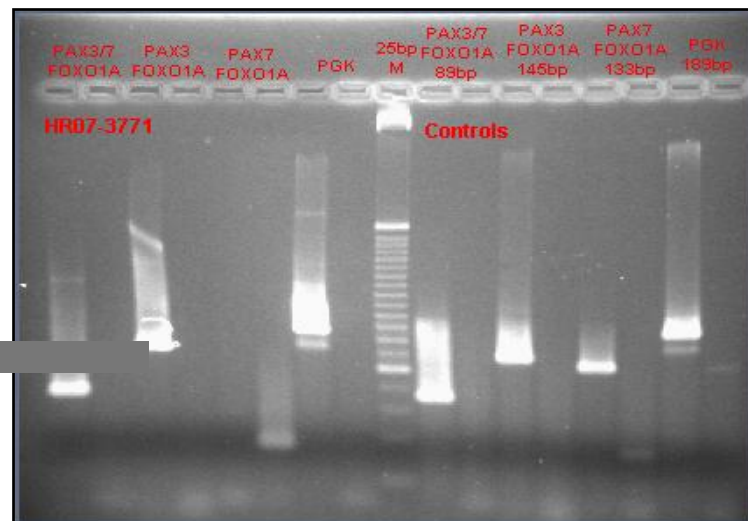


- Embriyonel ve alveoler alt tipler
- Alveoler spesifik translokasyon
- İmmunohistokimyasal alt tipleme
- Tissue array
- AP2beta ve P-Cadherin AlvRMS
- EGFR ve fibrillin-2 EmbRMS
- Yanlış alt tiplerede yardımcı
- Sonuçlar iyi klinik gidiş ile ilişkili



# ALVEOLER RMS

- t (2;13)(q35;q14) (*PAX3-FKHR*): 75%
- t (1;13) (p36;q14)(*PAX7-FKHR*): 10%/metastatik formda daha iyi prognoz
- <15% fusion-negatif
- ARMS ilkel form ERMS den ayırmda kullanılabilir



Sorensen, P. H.B. et al. J Clin Oncol; 20:2672-2679 2002

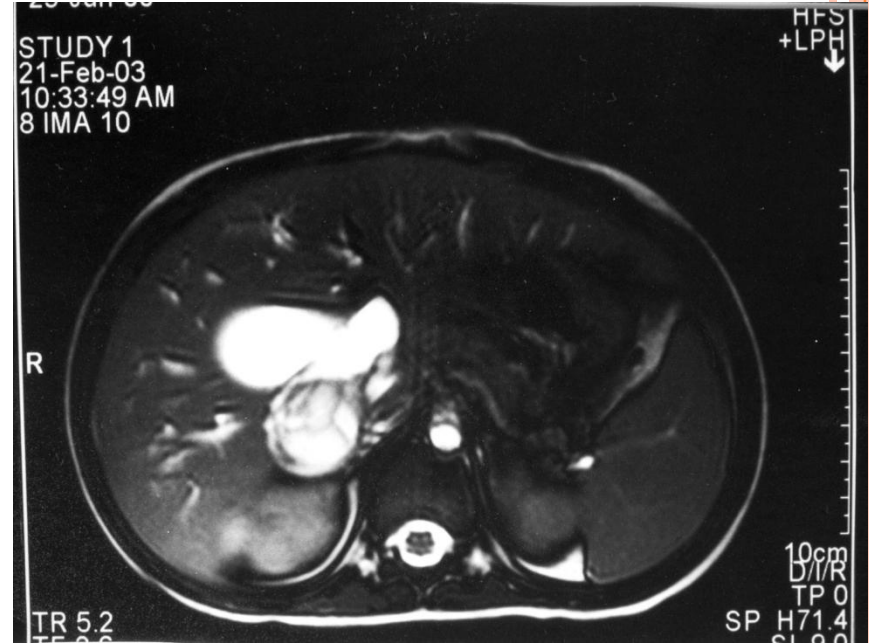
## CERRAHPAŞA 2000-2005 RMS

- 36 olgu----tüm maligniteler içinde sarkomlar %11, bunun %1,8'i RMS
- Cins ayrımı yok, baş/boyun yerleşimi sık
- %57----- 10 yaş altı
- Emb RMS en sık ve en iyi gidişli
- Emb RMS medyan sağkalım 41 ay alvRMS 28 ay (p<0,002)
- %31 nüks ve metastaz

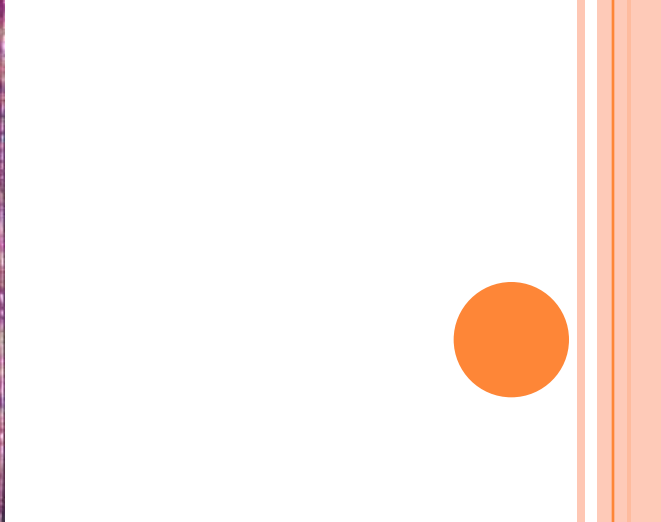
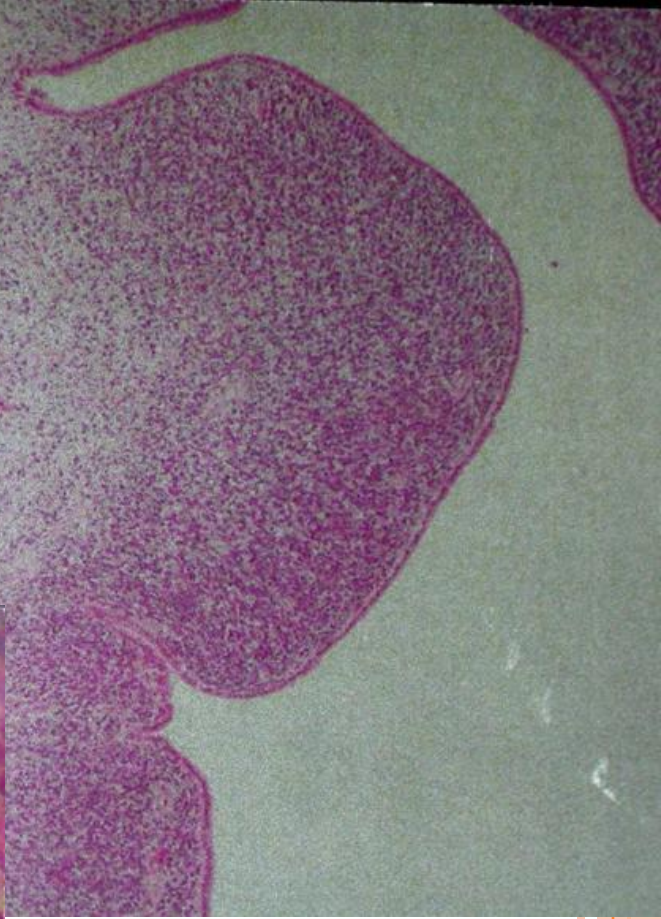
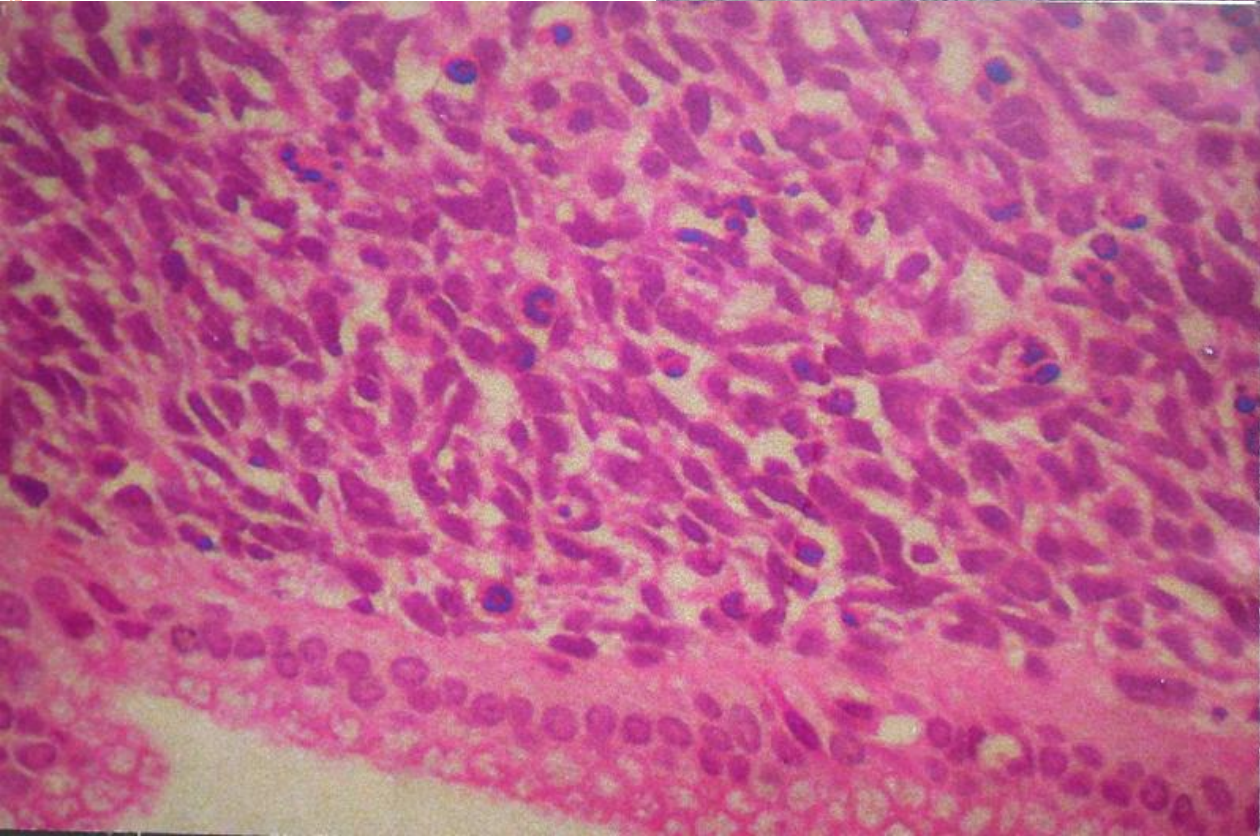
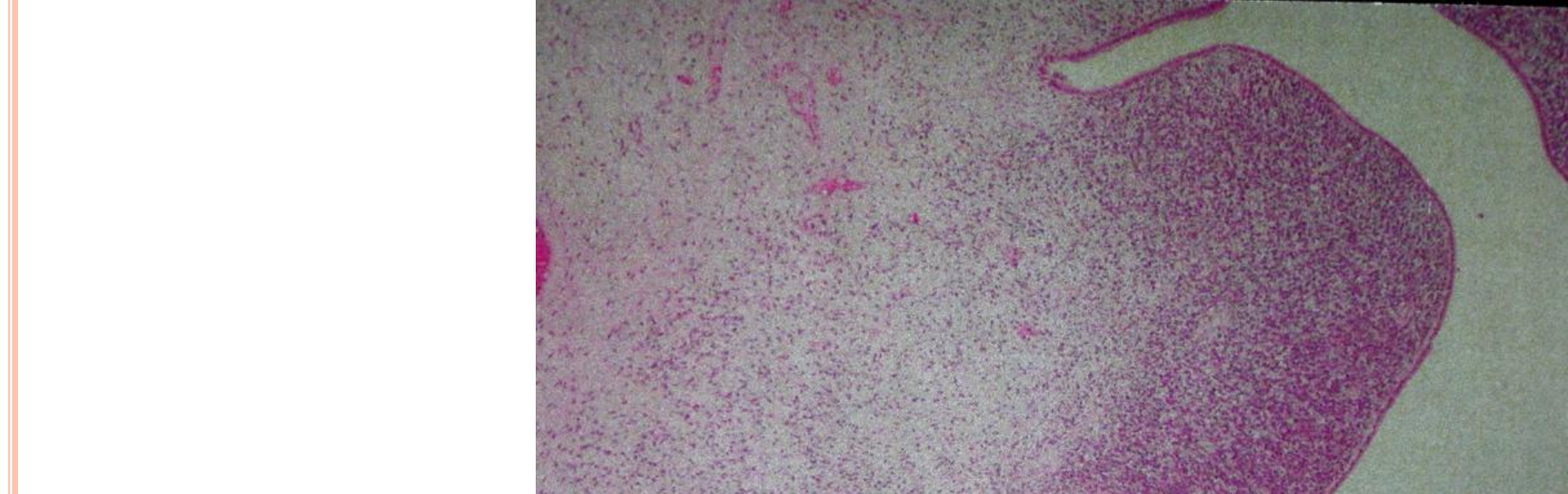


# RMS YERLEŐİM

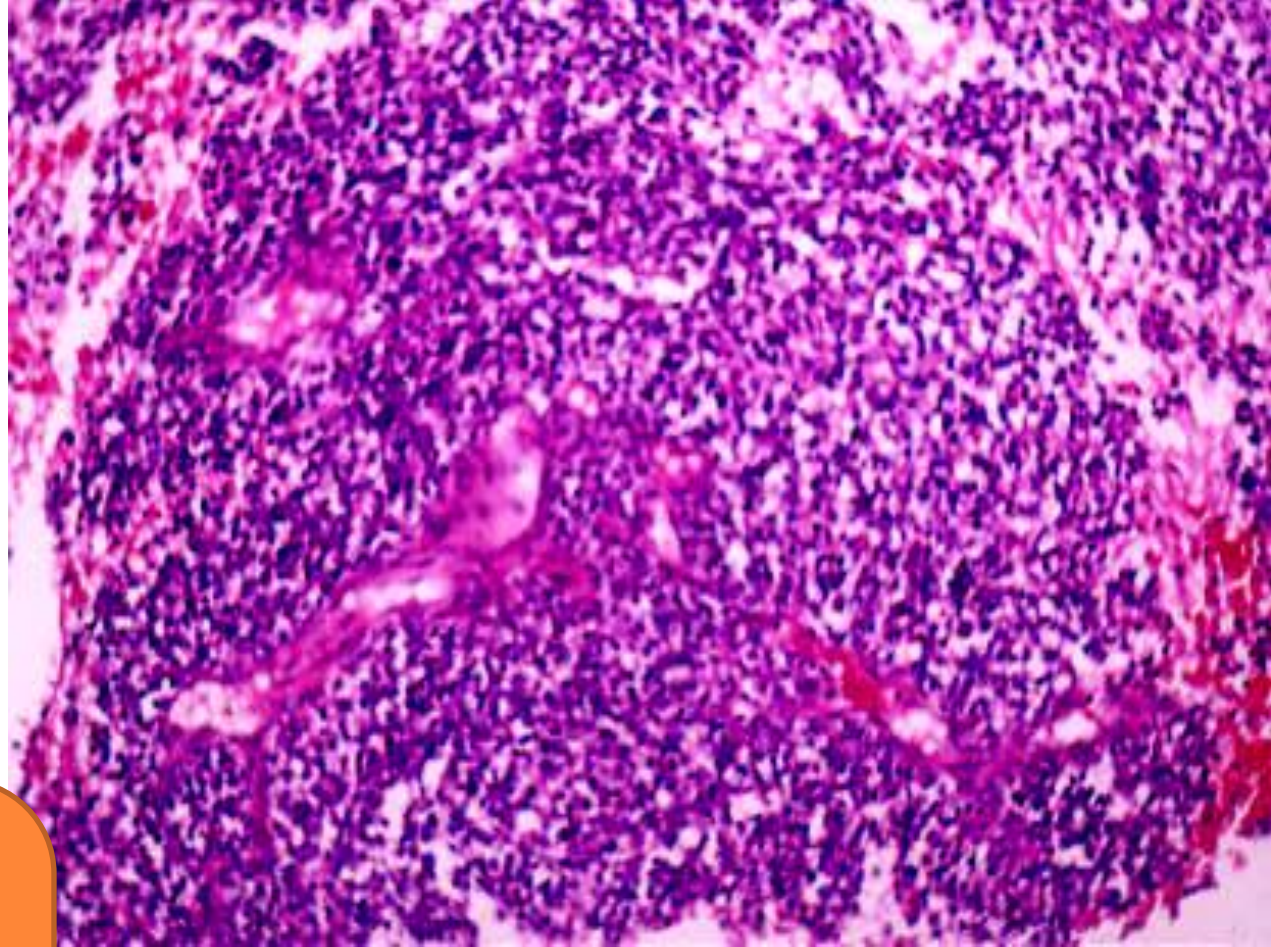
- BaŐ/boyun ( orbita, nazal kavite, oral, sinüsler, kulak, mastoid )
- Gövde ( paratestiküler, retroperiton, pelvis, göğüs-karın duvarı, mesane, prostat, vajen, vulva)
- Ekstremiteler





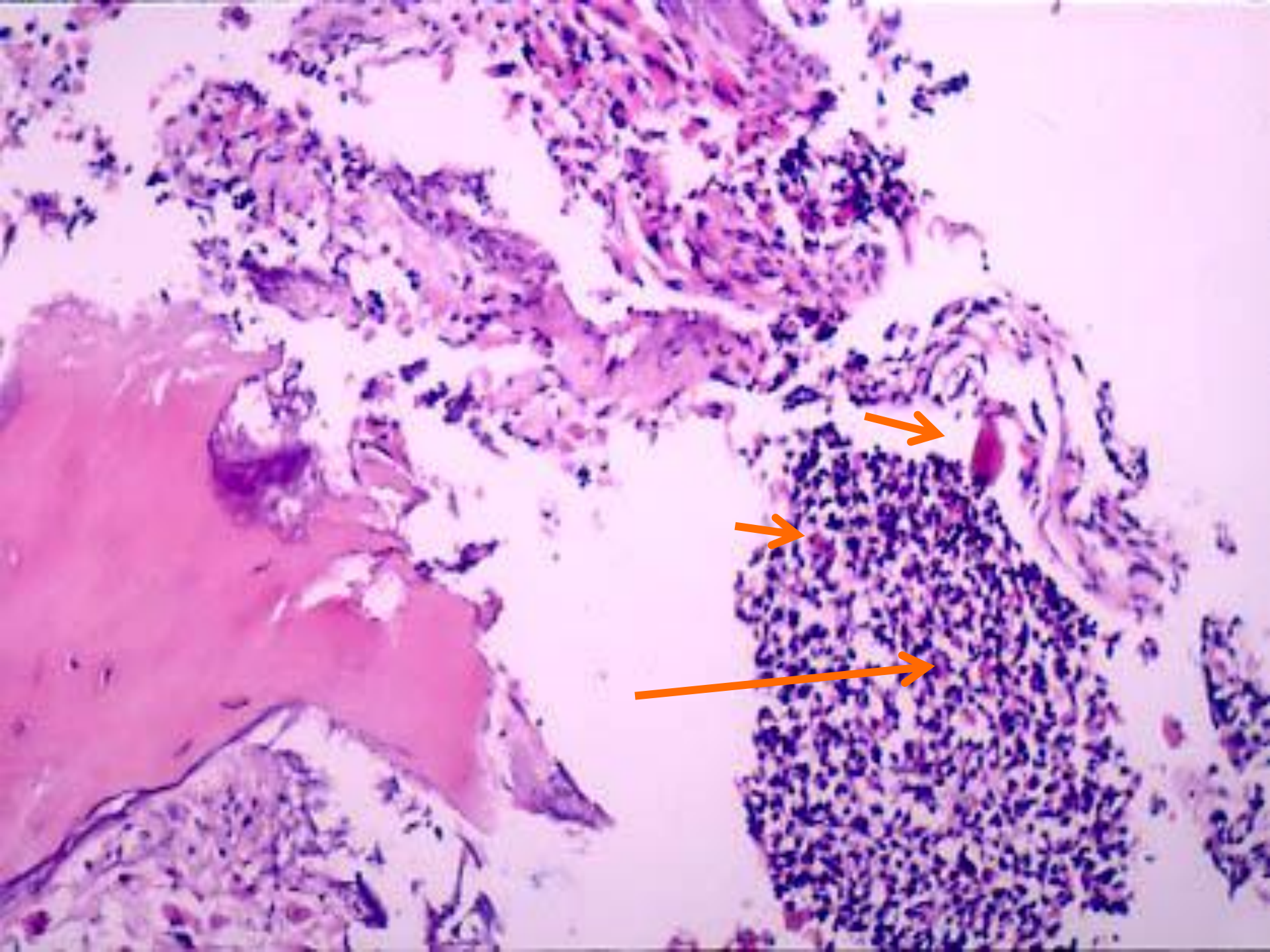


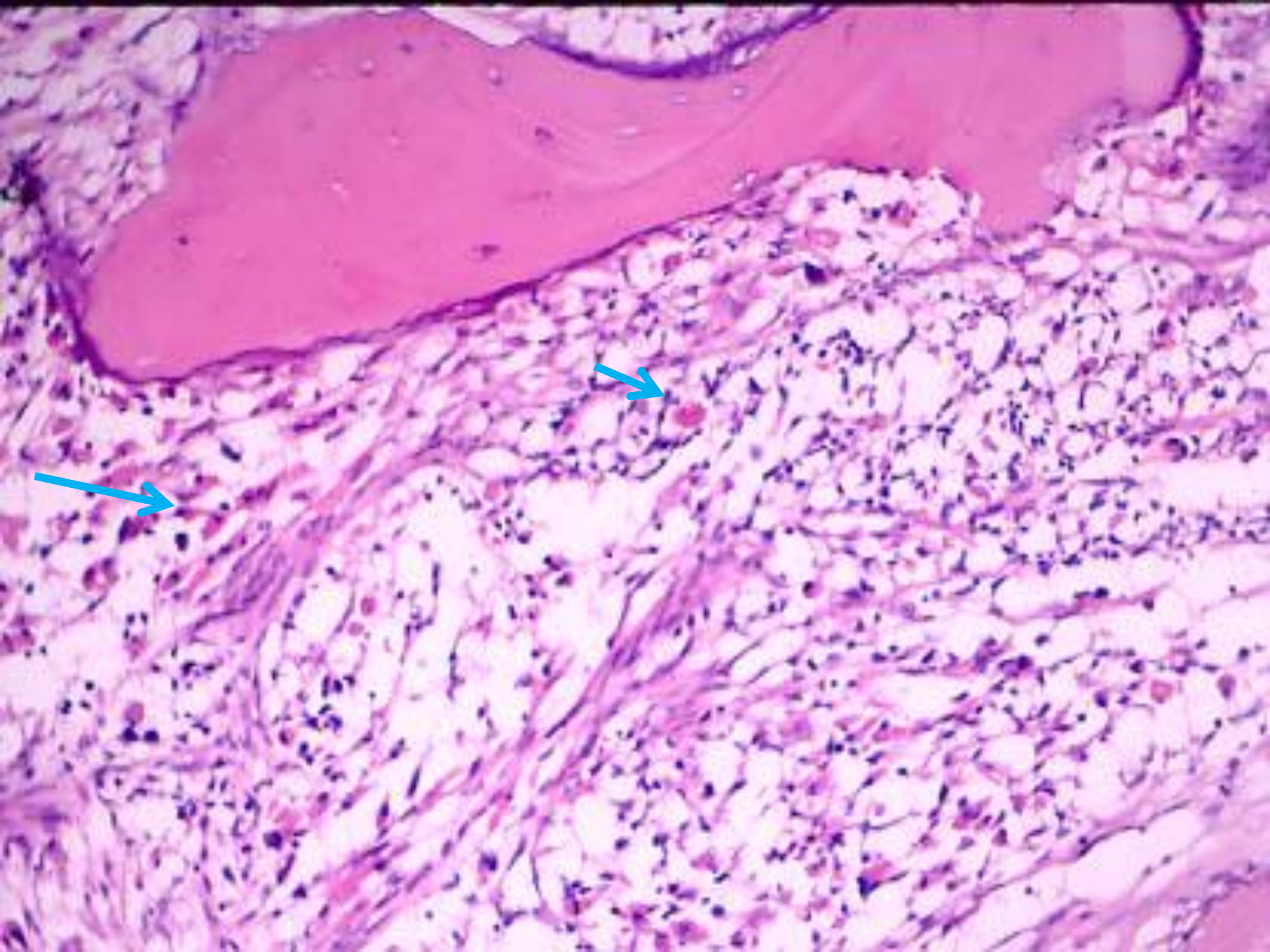




Küçük hücreli  
osteosarkom?  
Ewing/PNET?



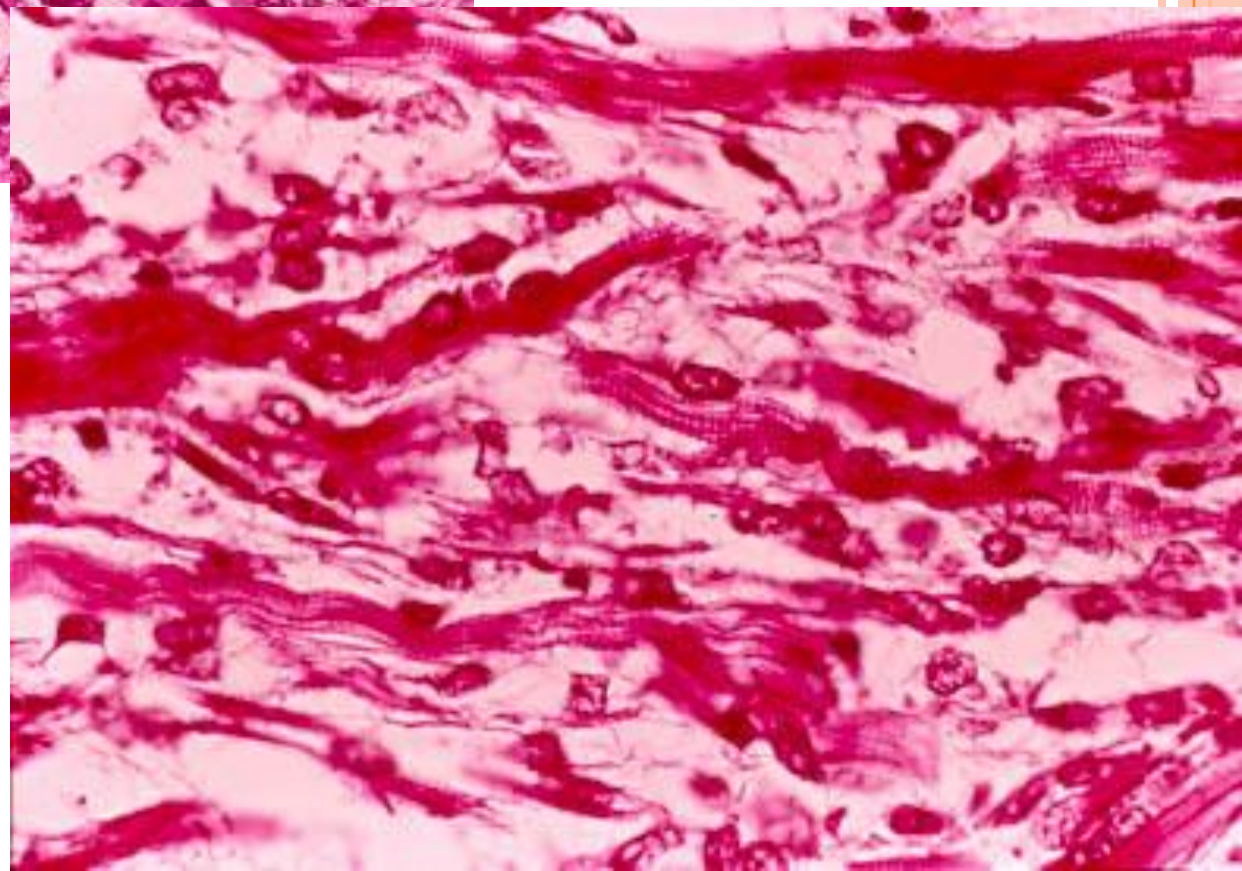
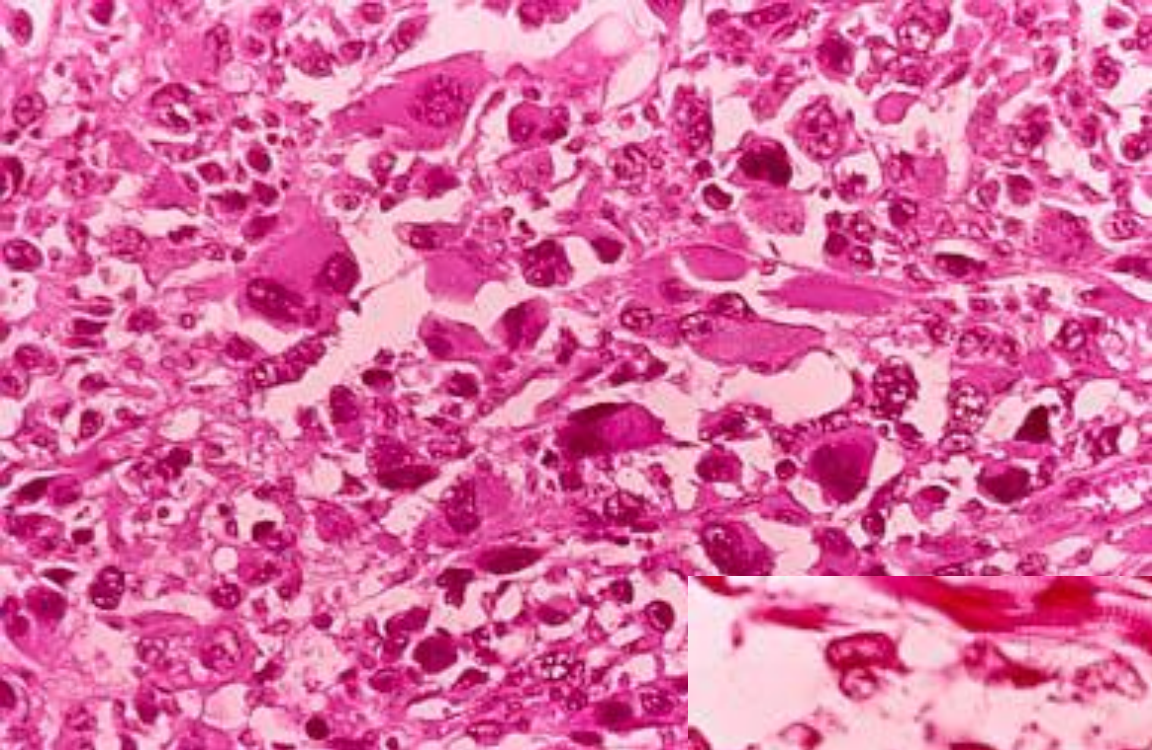




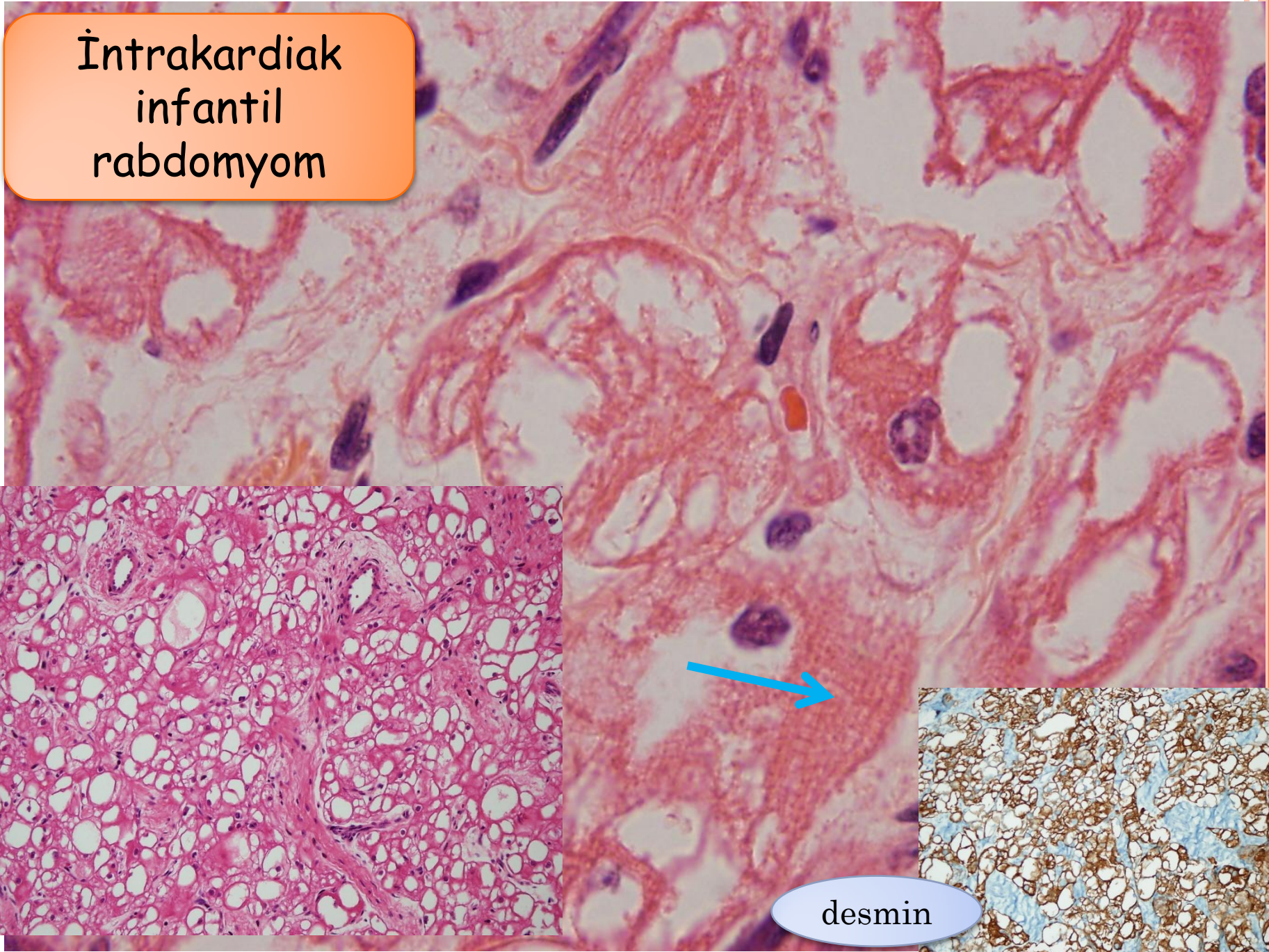
# RMS TANISINDA ANA PROBLEMLER

- İndiferansiye tipin diğer küçük hücreli tümörlerden ayrımı
- İğsi hücreli tipin FS, LMS, MFH ve diğer sarkomlardan ayrımı
- Küçük biopsi, iğne biopsi ve frozen materyellerinde tanı kriterlerinin minimal oluşu, örtüşebilen paternler, rabdoid hücreler
- Olağandışı yerleşimlerde tanı güçlüğü



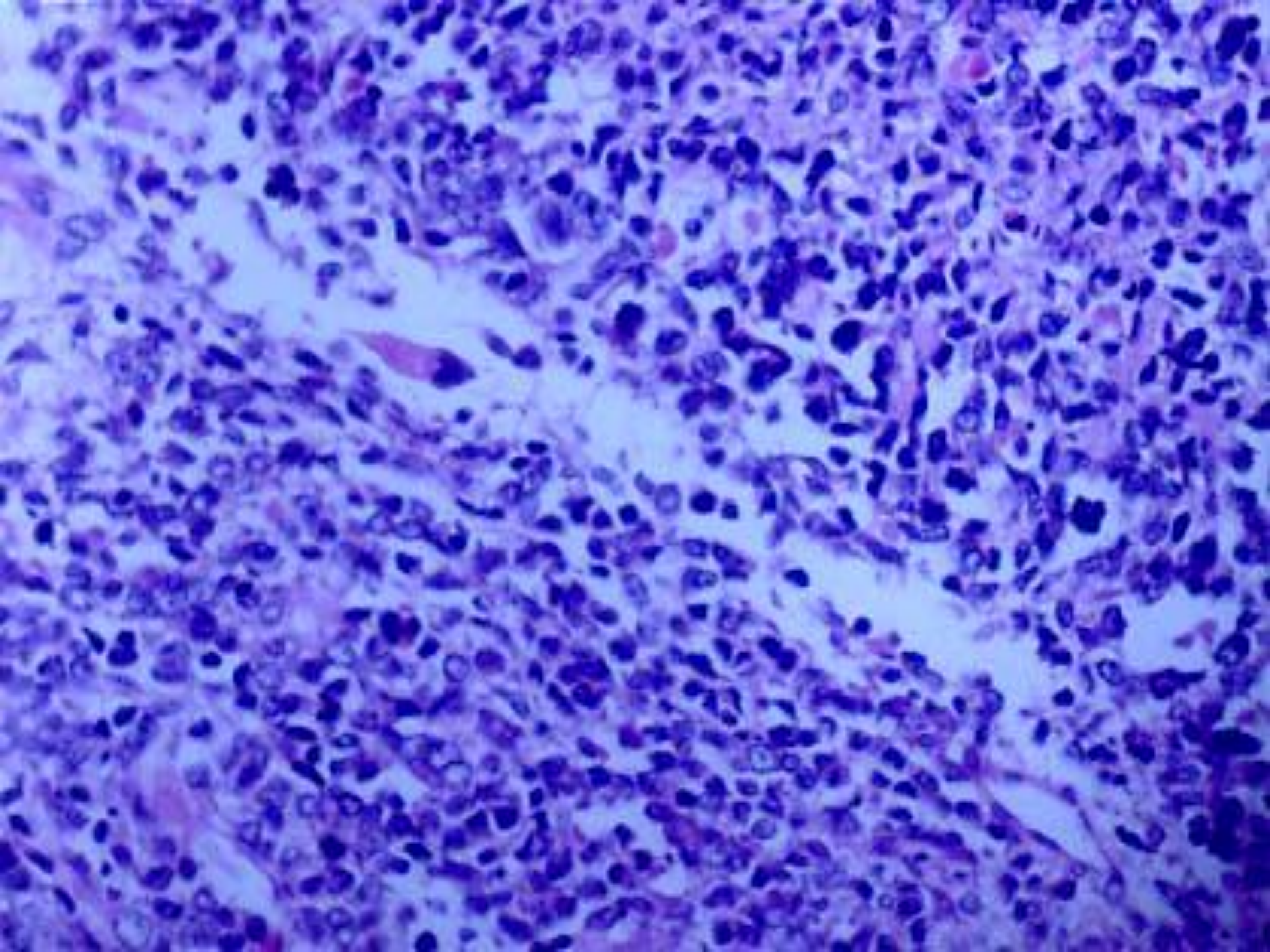


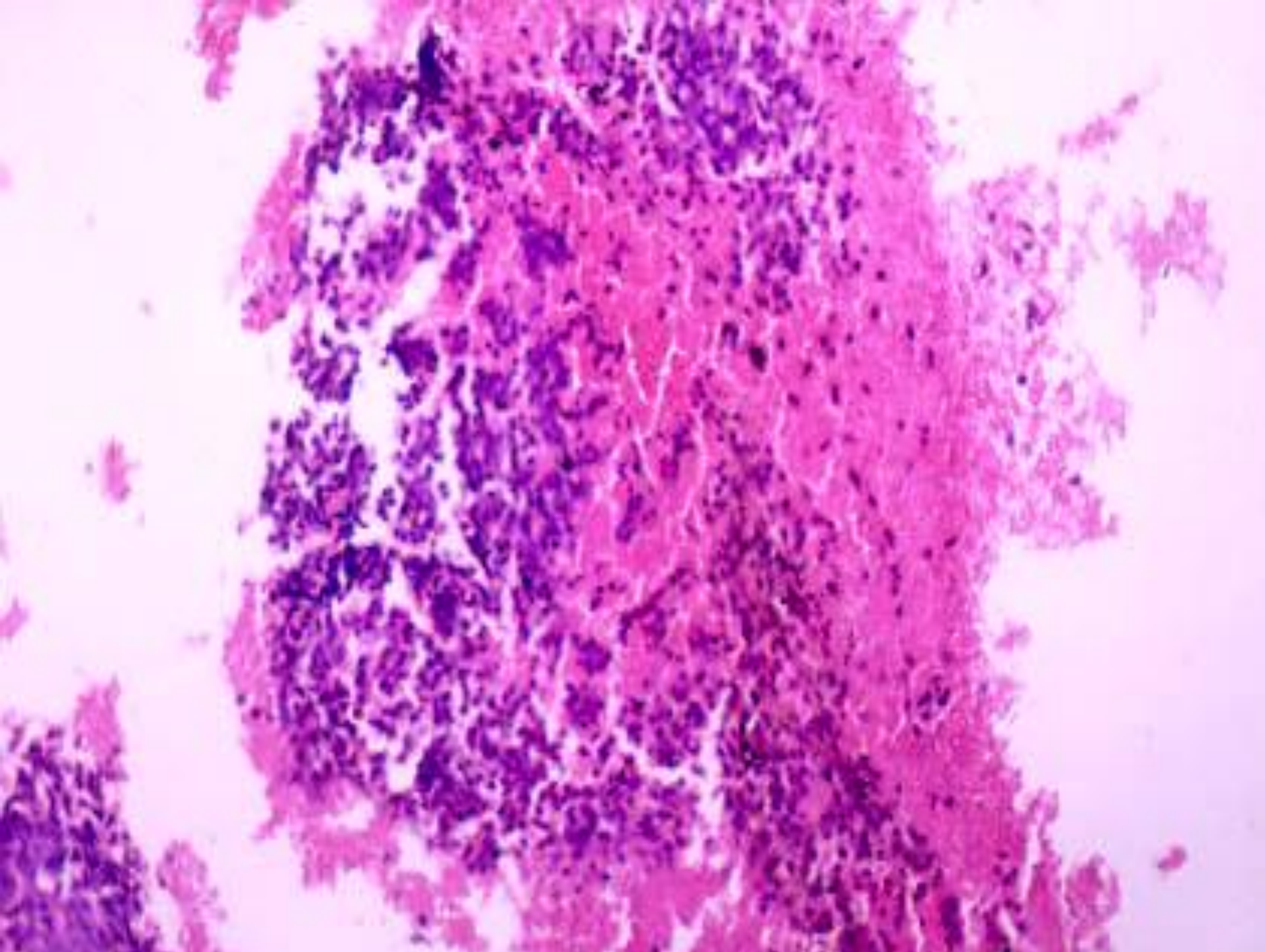
Intrakardiak  
infantil  
rabdomyom

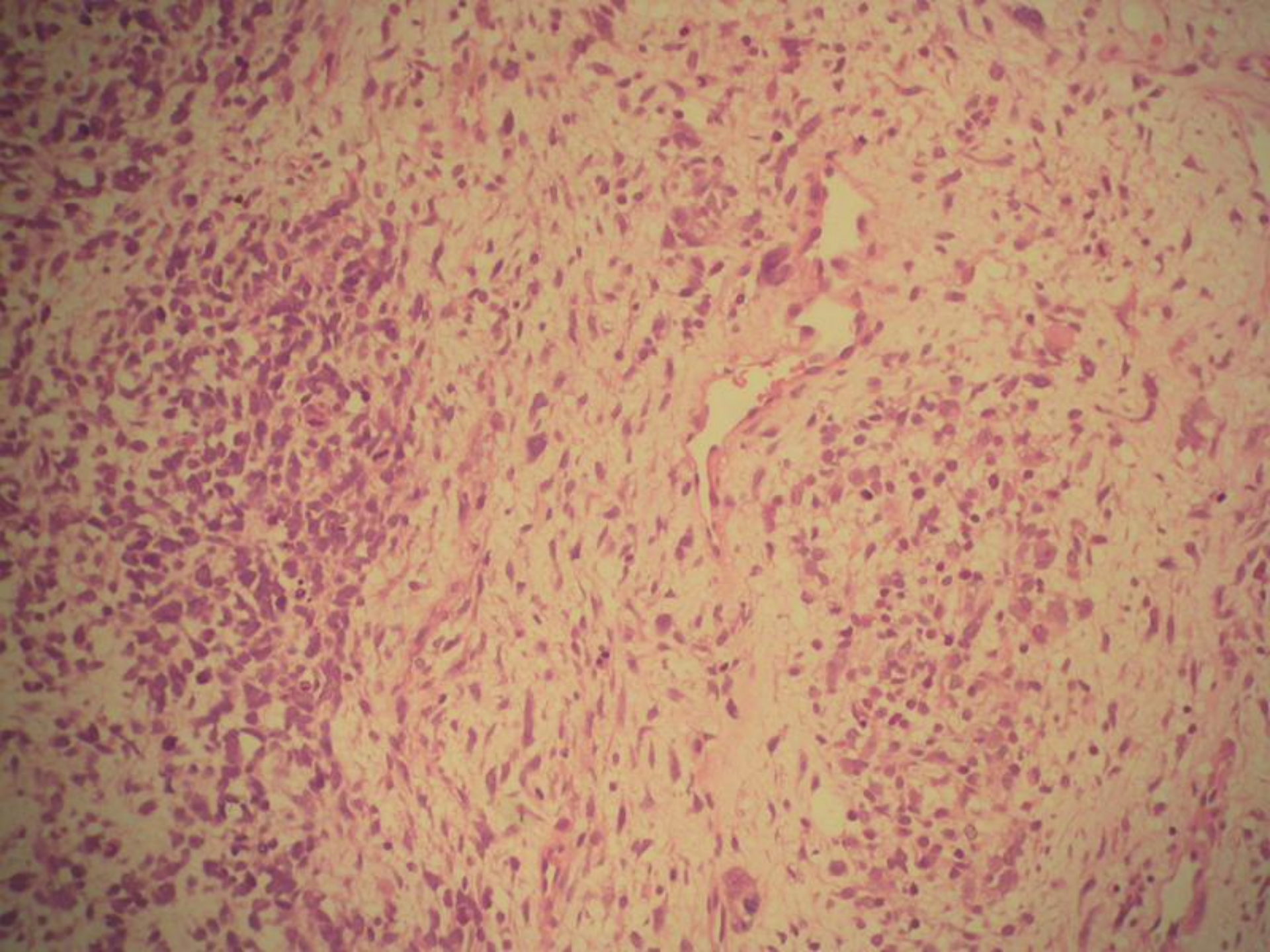


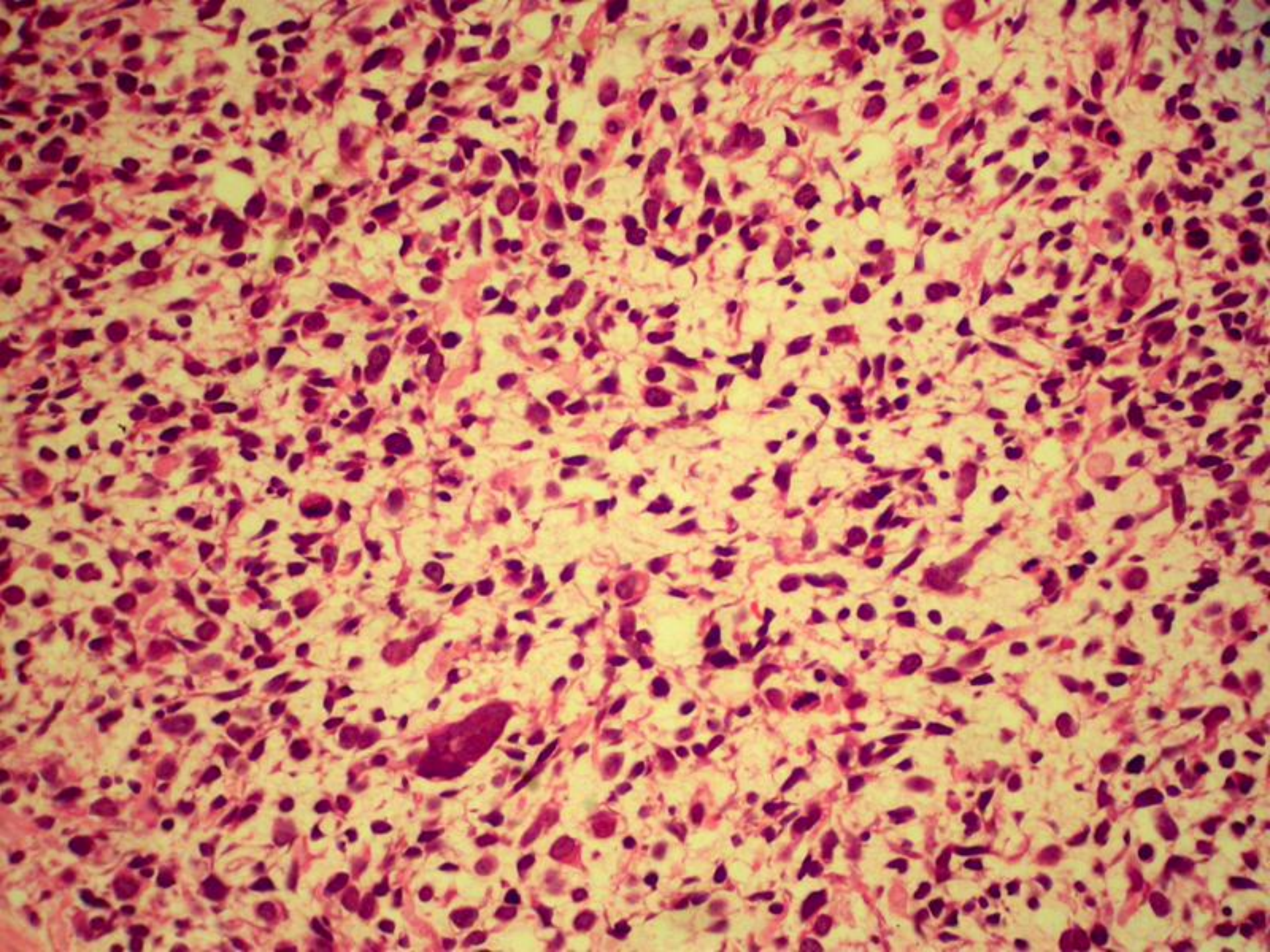
desmin

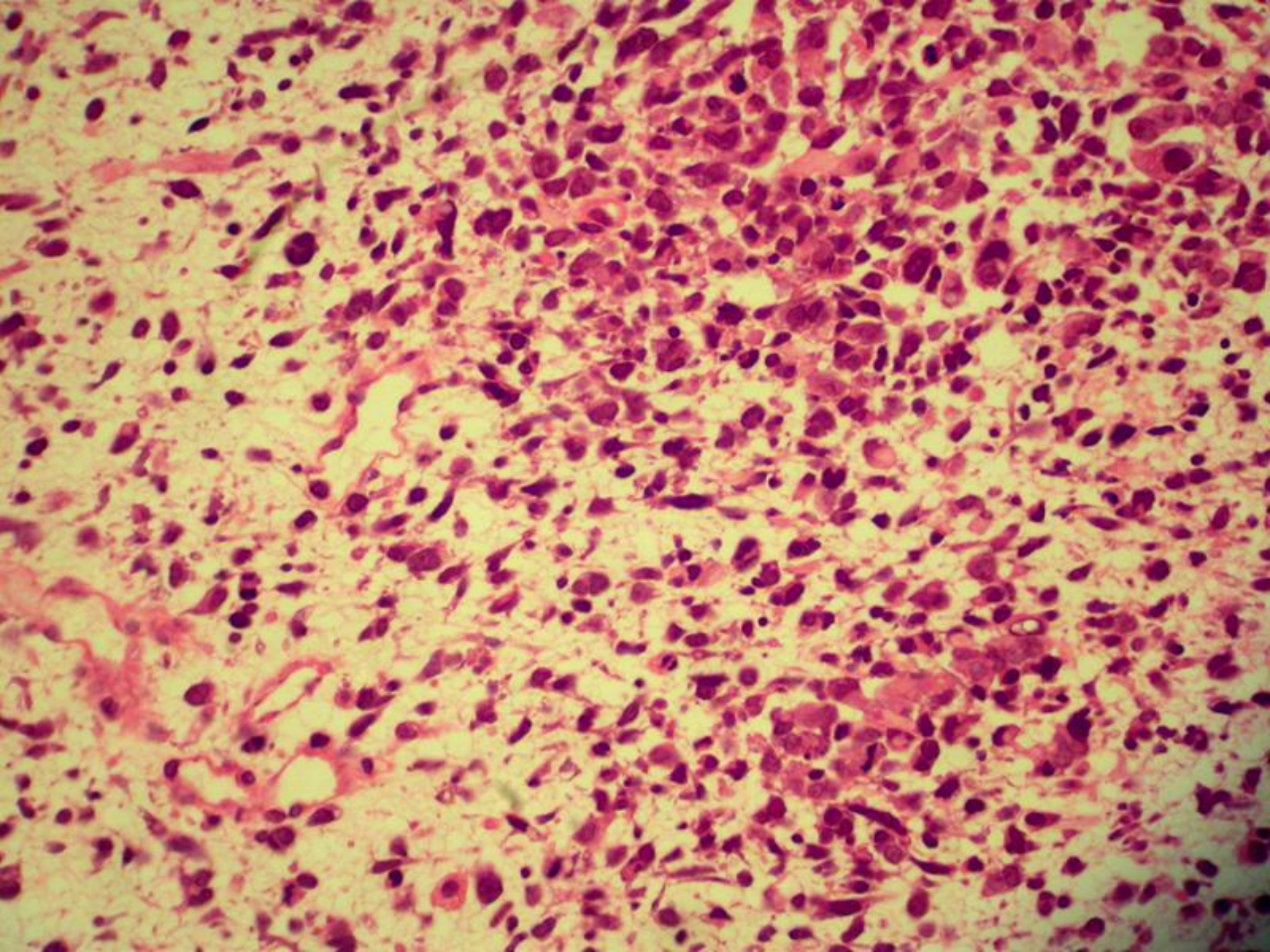


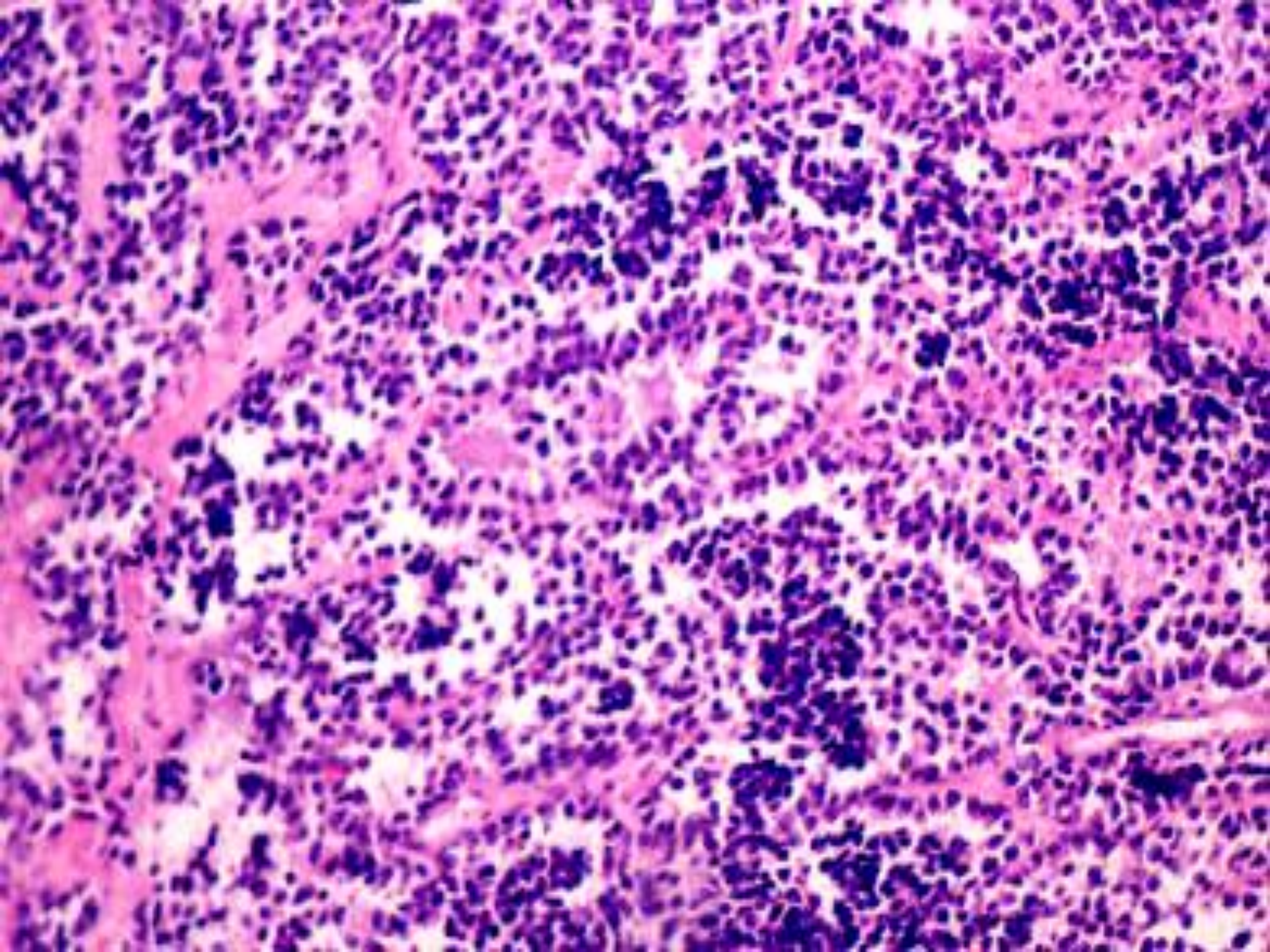










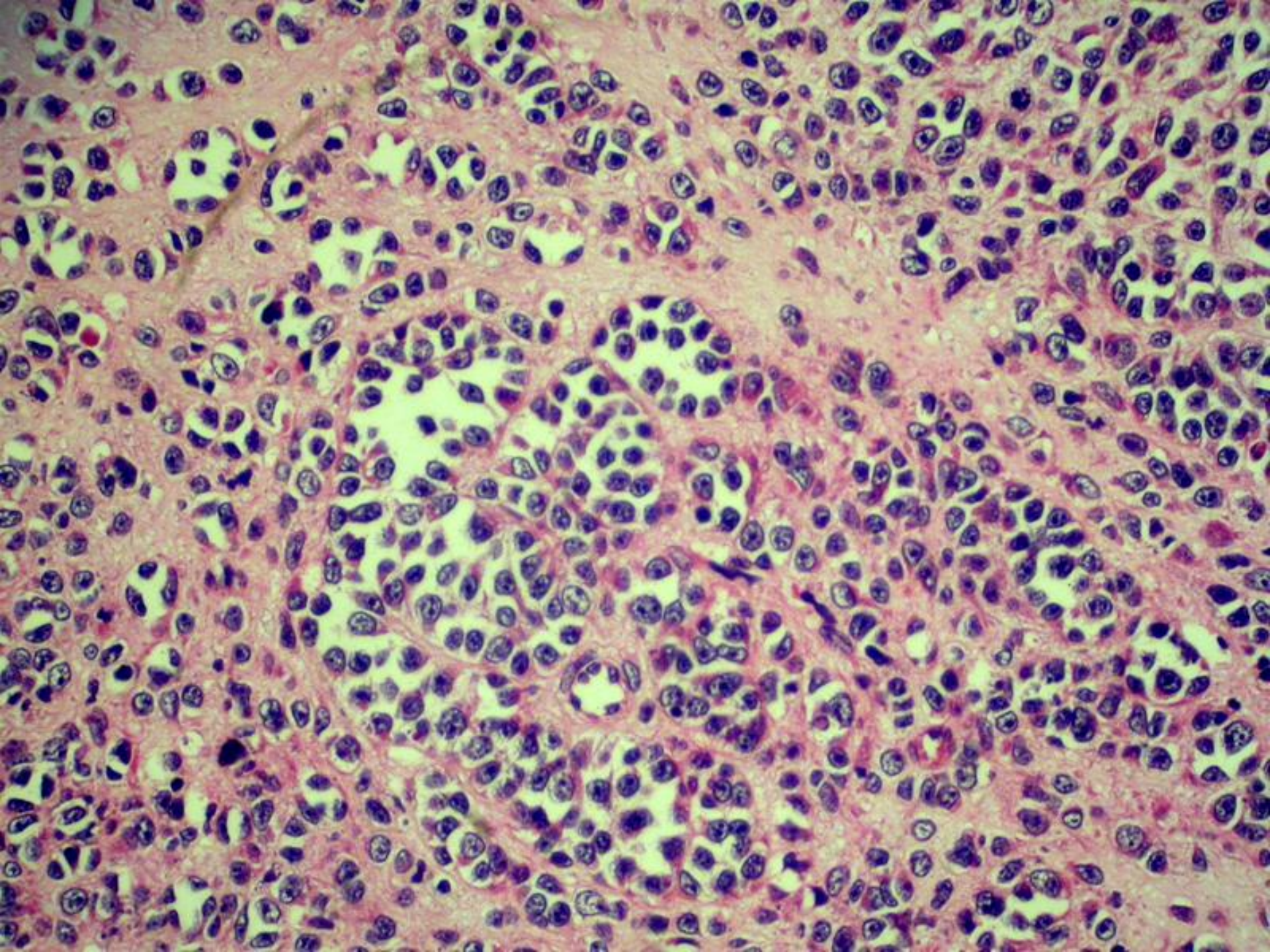


- Sclerosing rhabdomyosarcoma in adults: report of four cases of a hyalinizing, matrix-rich variant of rhabdomyosarcoma that may be confused with osteosarcoma, chondrosarcoma, or angiosarcoma.

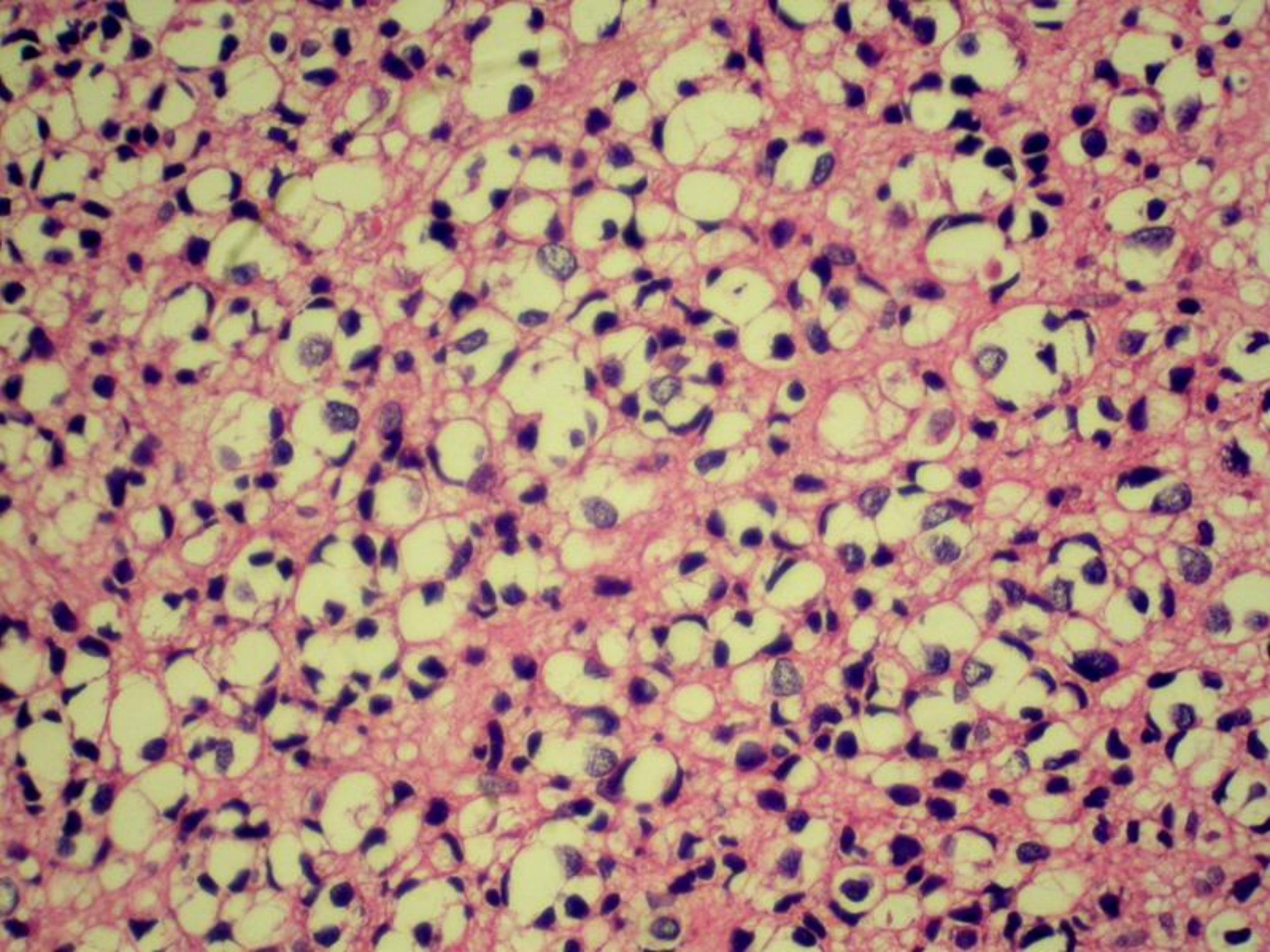
Folpe AL, McKenney JK, Bridge JA, Weiss SW.

Am J Surg Pathol. 2002 Sep;26(9):1175-83.

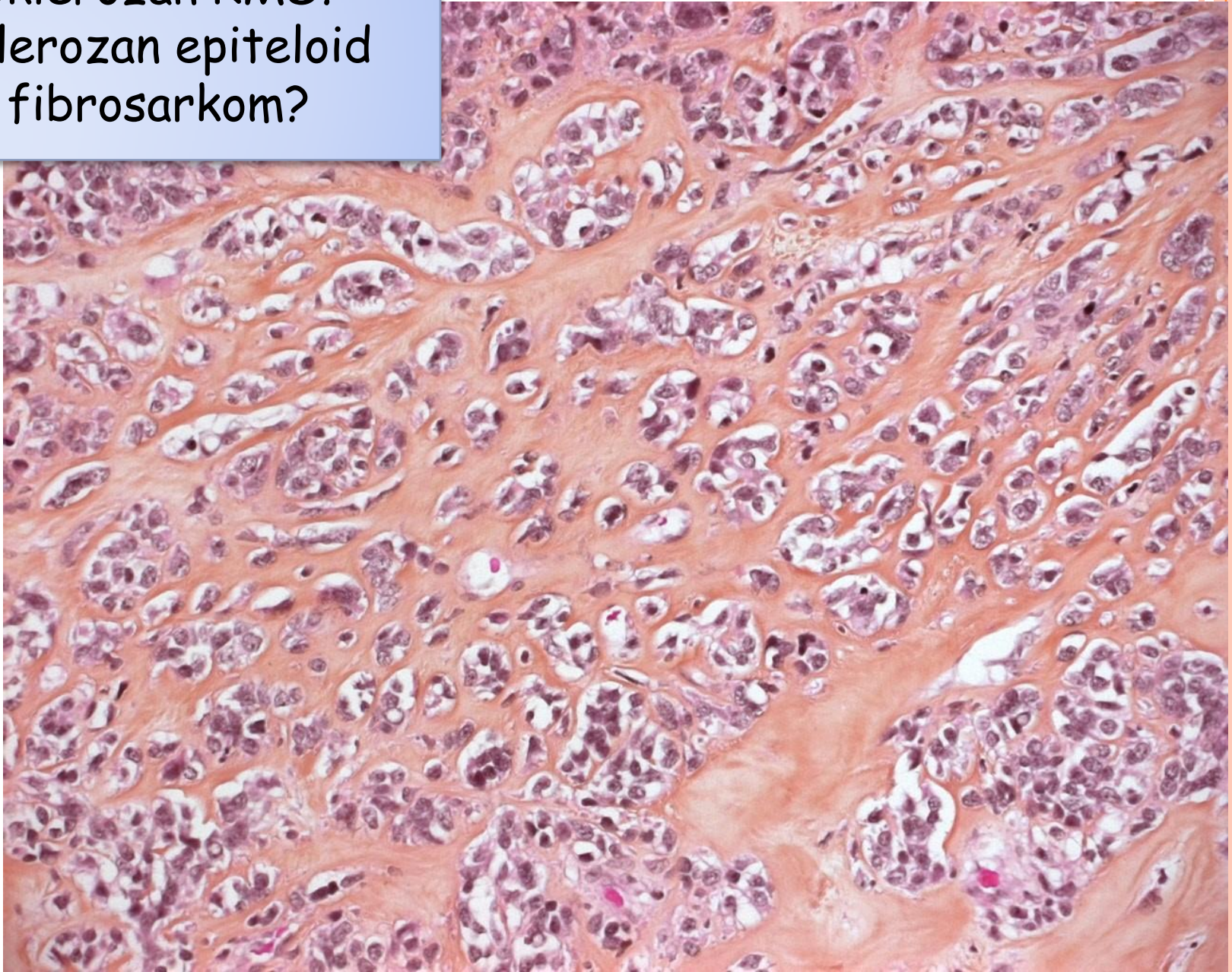


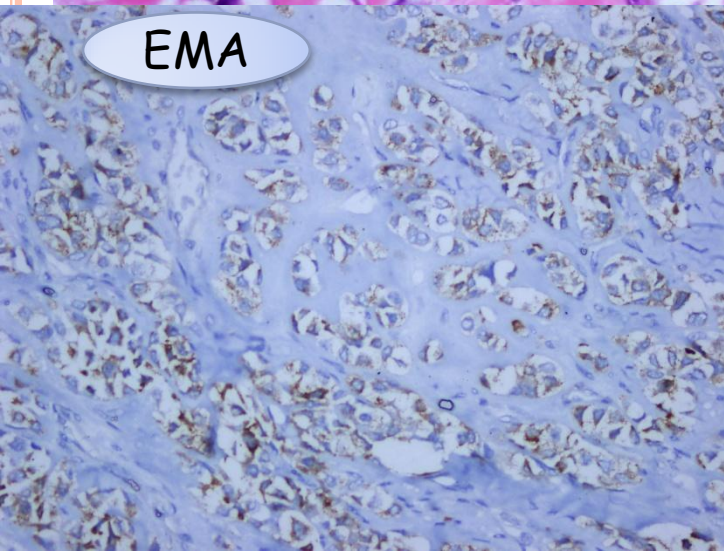
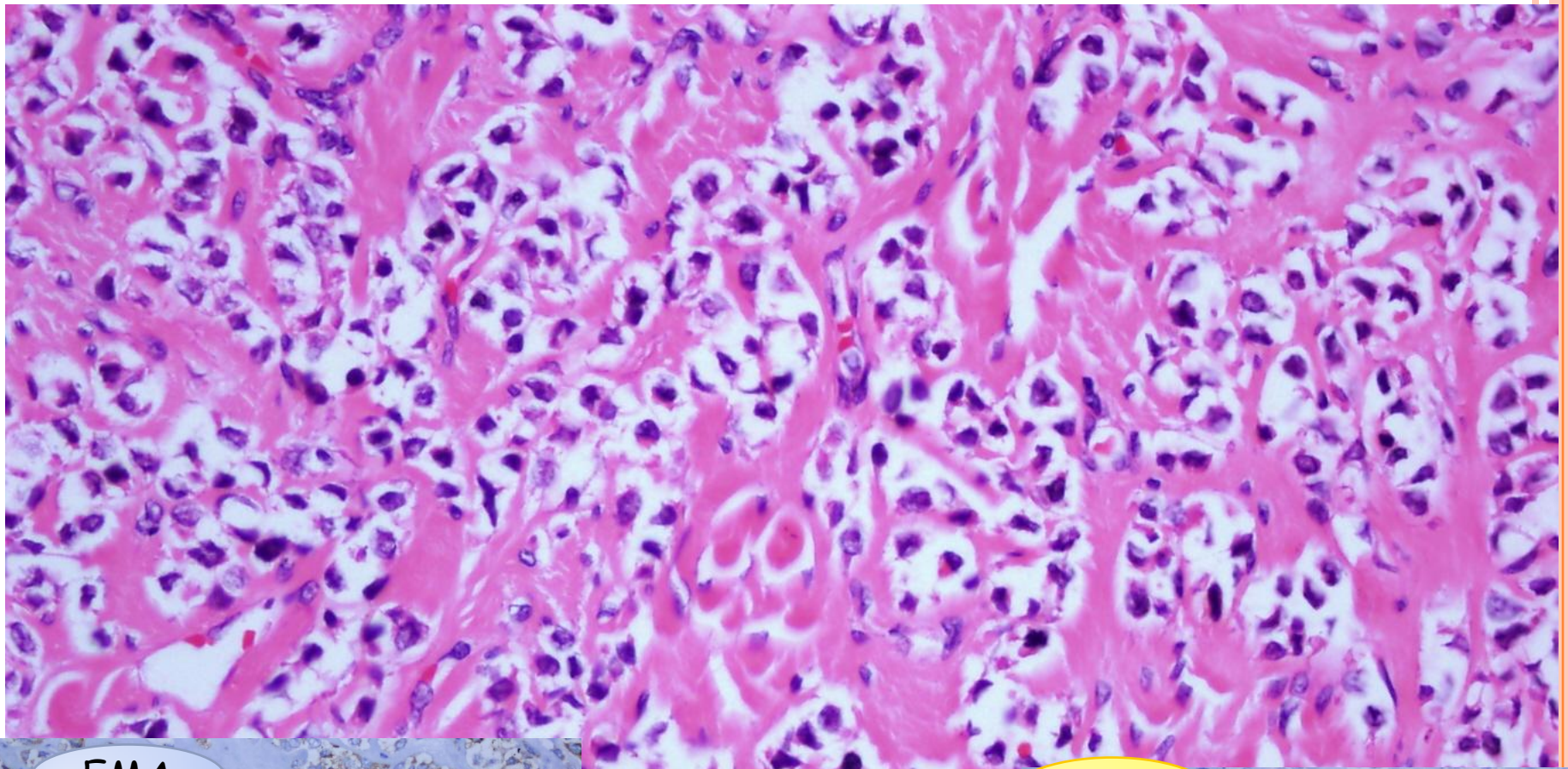




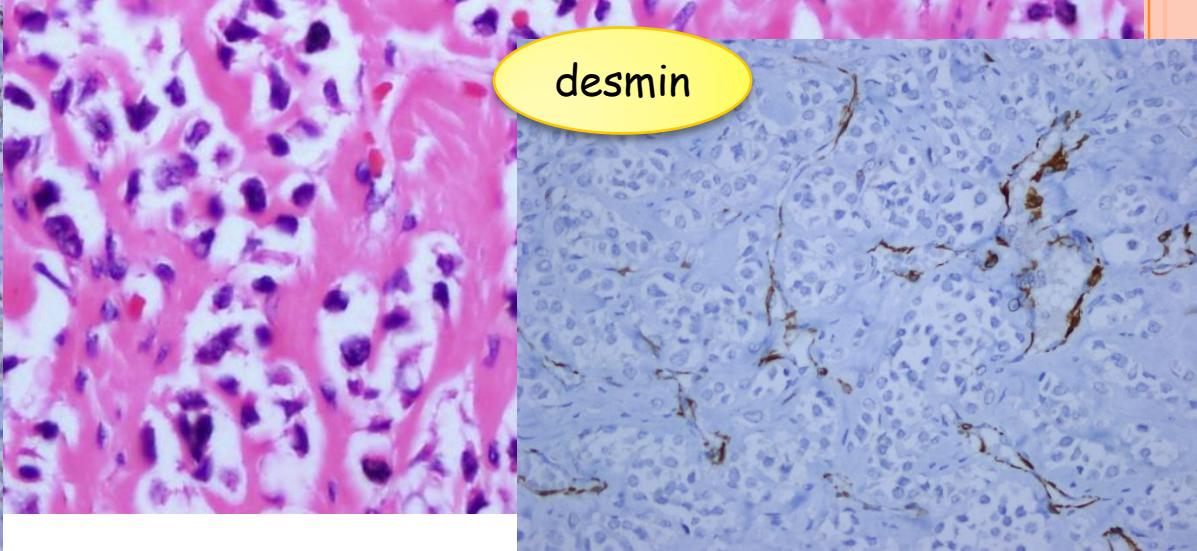


Sklerozan RMS?  
sklerozan epiteloid  
fibrosarkom?



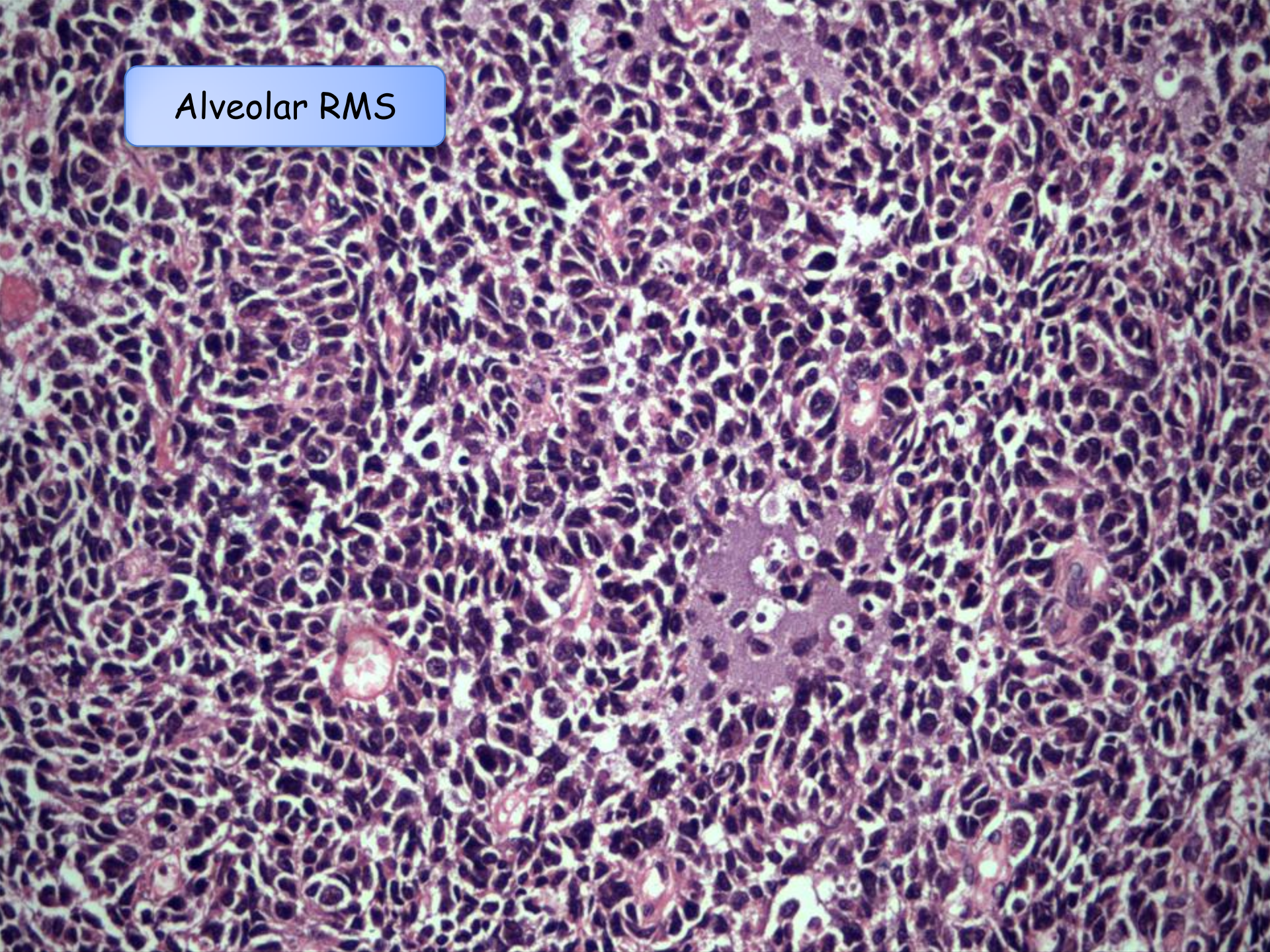


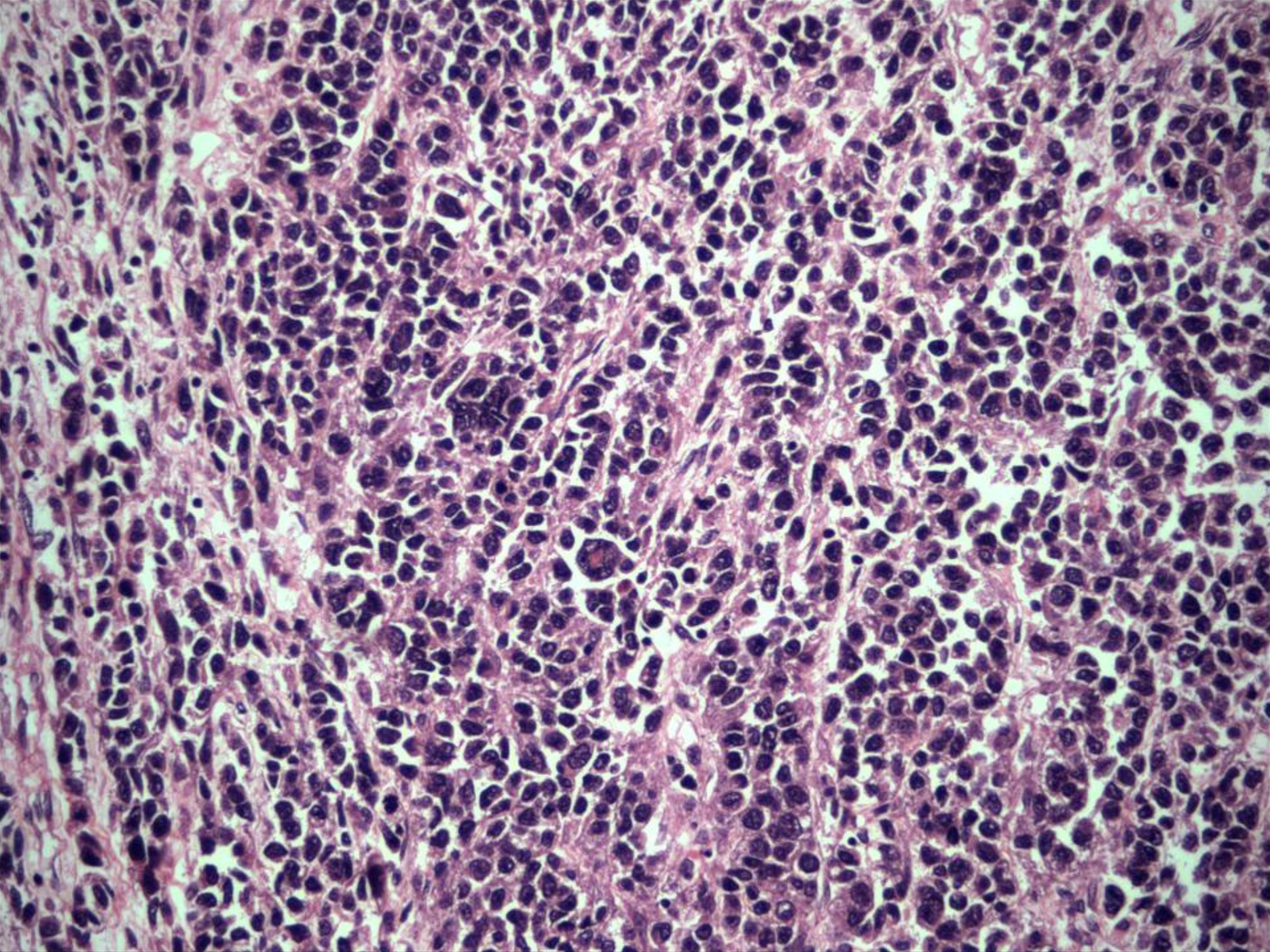
EMA

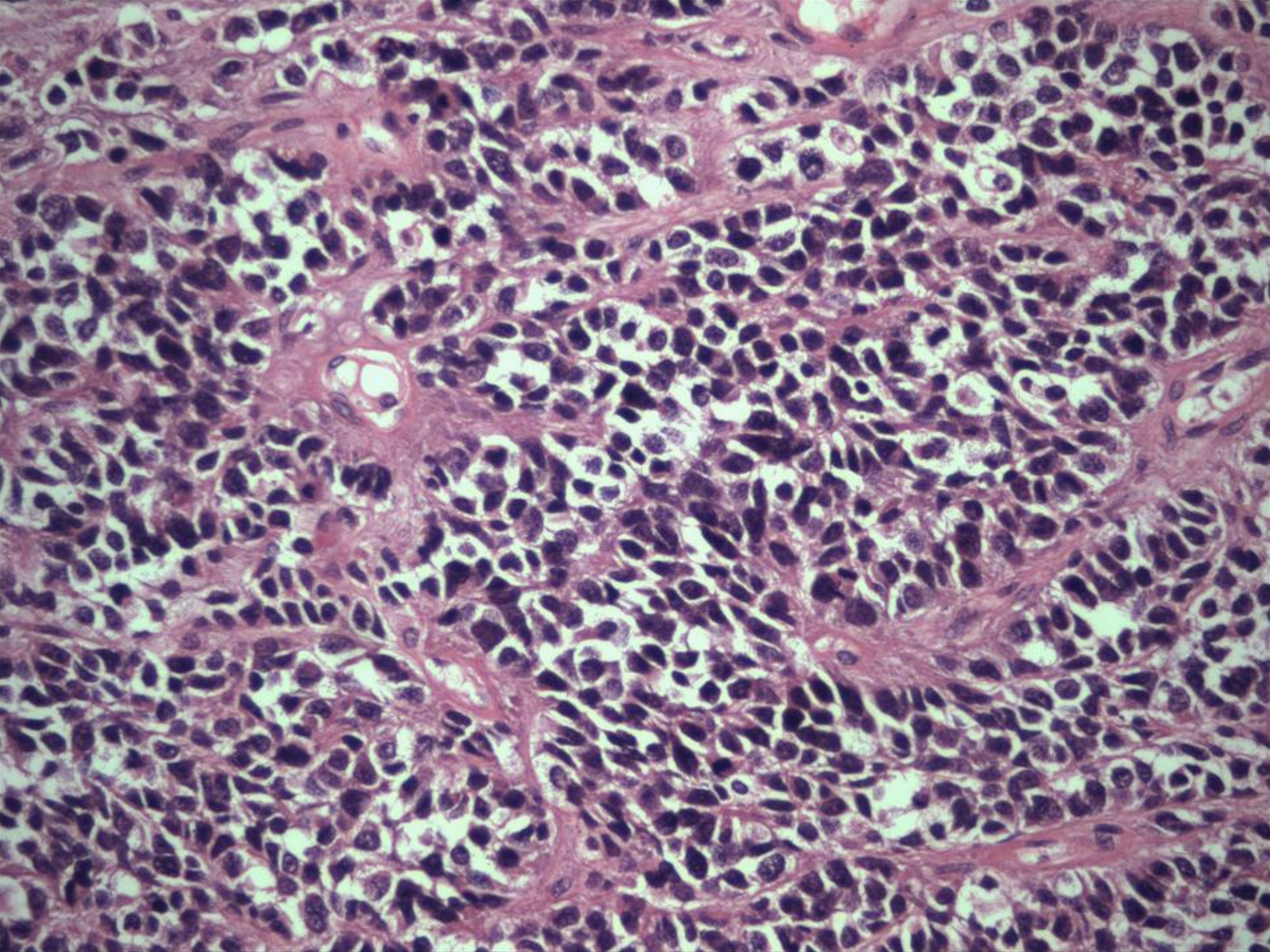


desmin

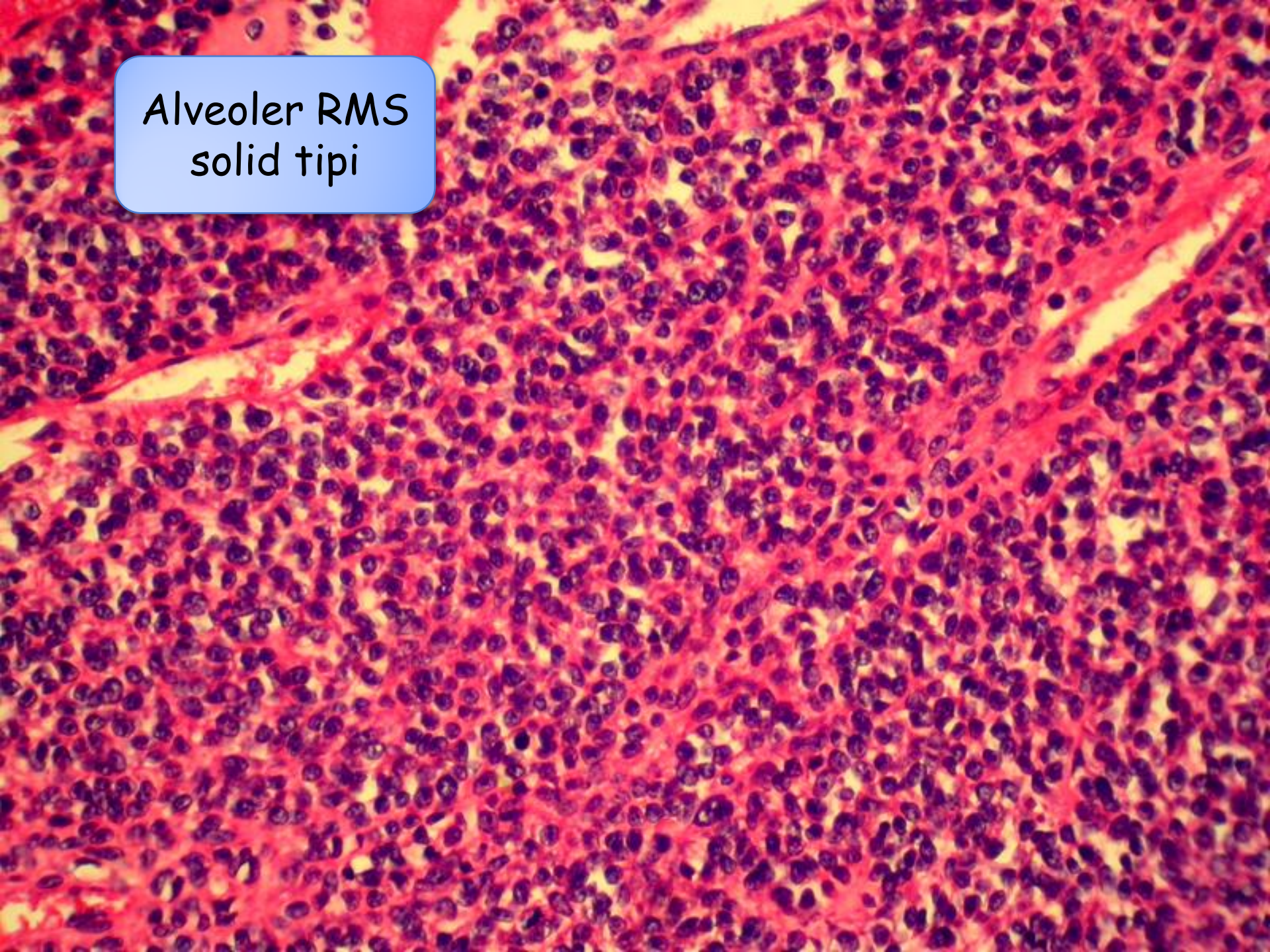
Alveolar RMS







Alveoler RMS  
solid tipi





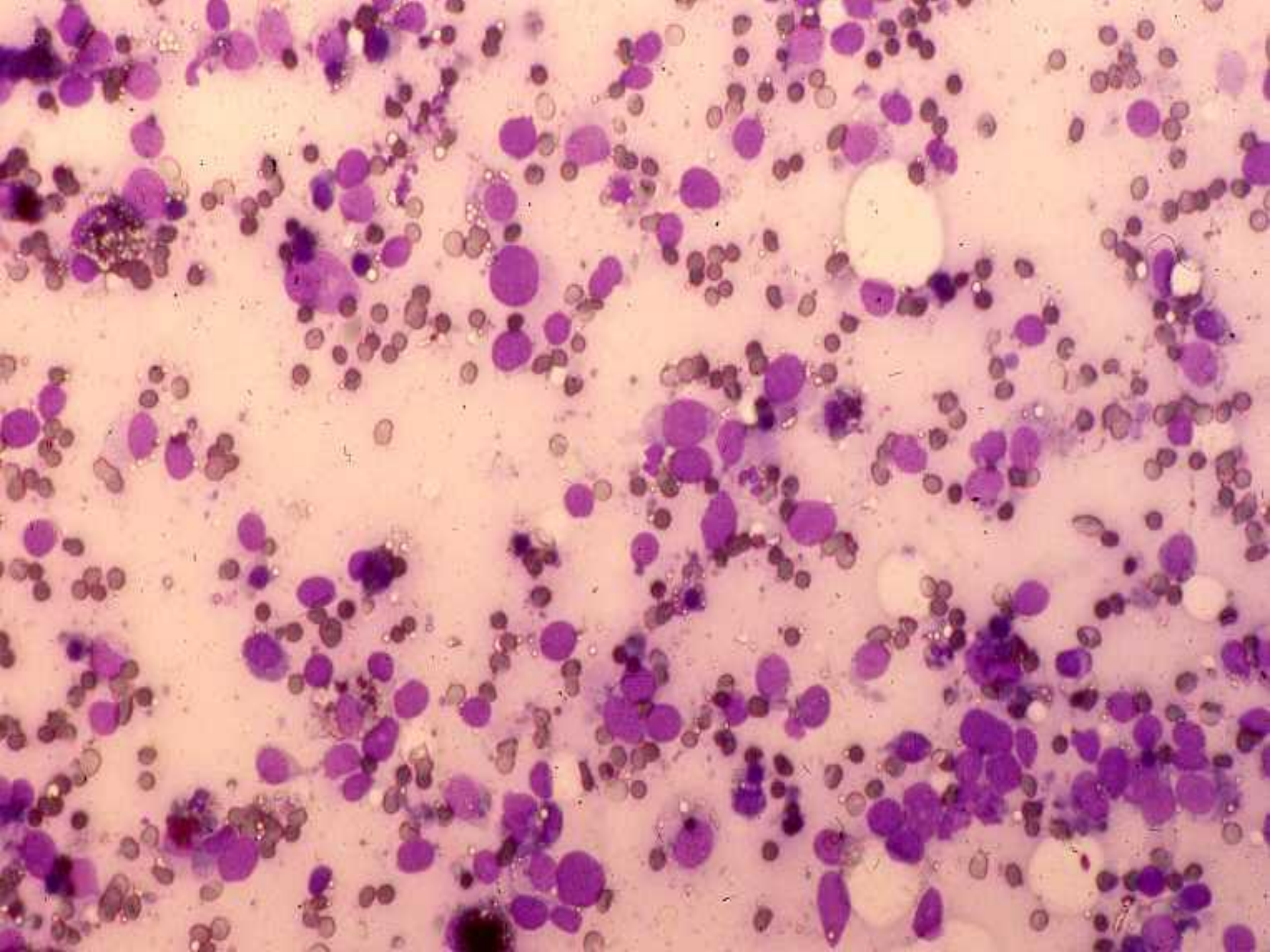
Malign rhabdoid tümör

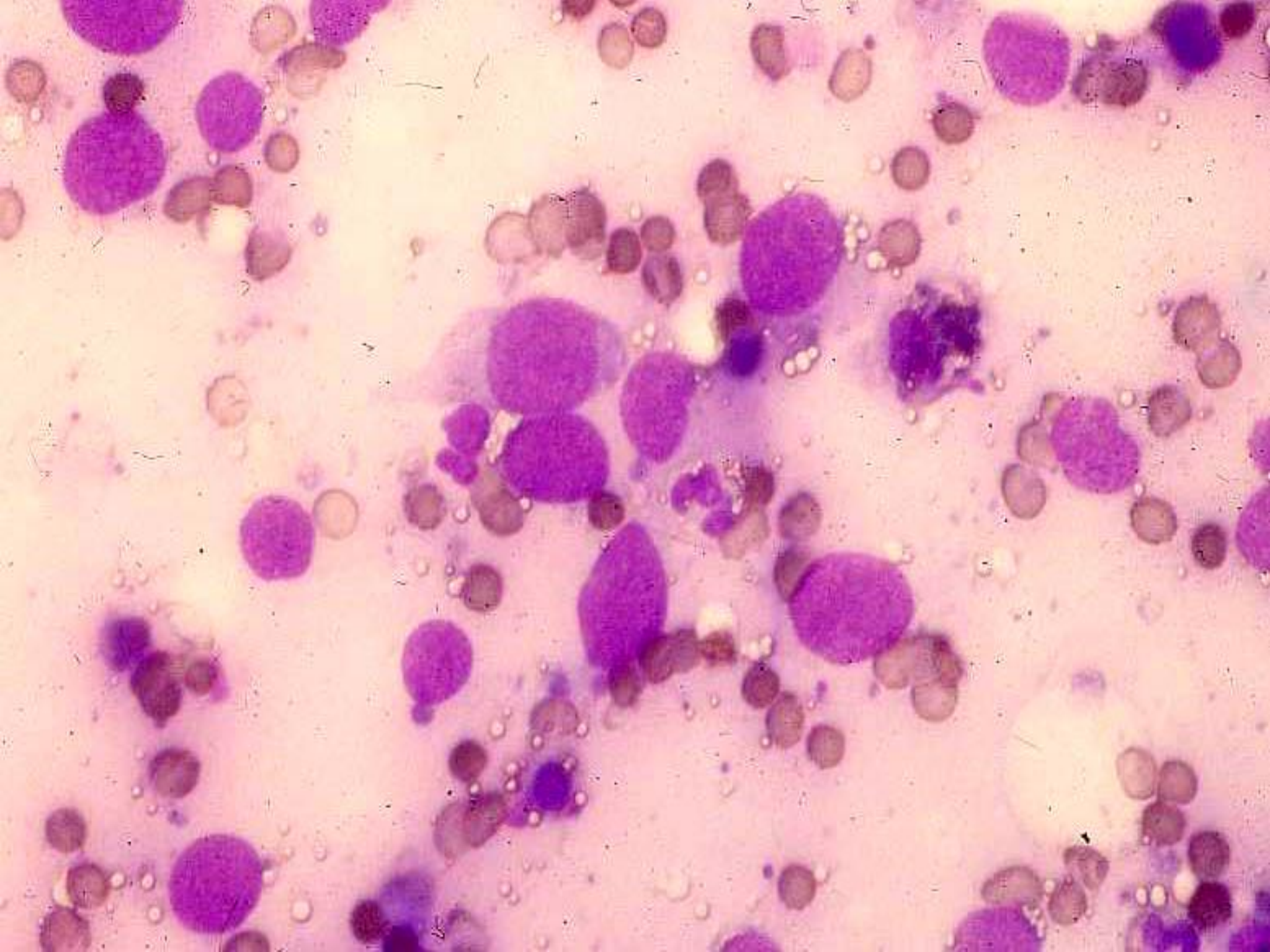
This image shows a microscopic view of a malignant rhabdoid tumor. The background is a dense field of rhabdoid cells, which are characterized by their large, eccentric nuclei and prominent, eosinophilic nucleoli. The cells are arranged in a disorganized pattern, typical of a malignant neoplasm. A central inset provides a higher magnification of the tumor cells, highlighting their characteristic morphology and the presence of numerous mitotic figures, indicating high cellular turnover and malignancy.

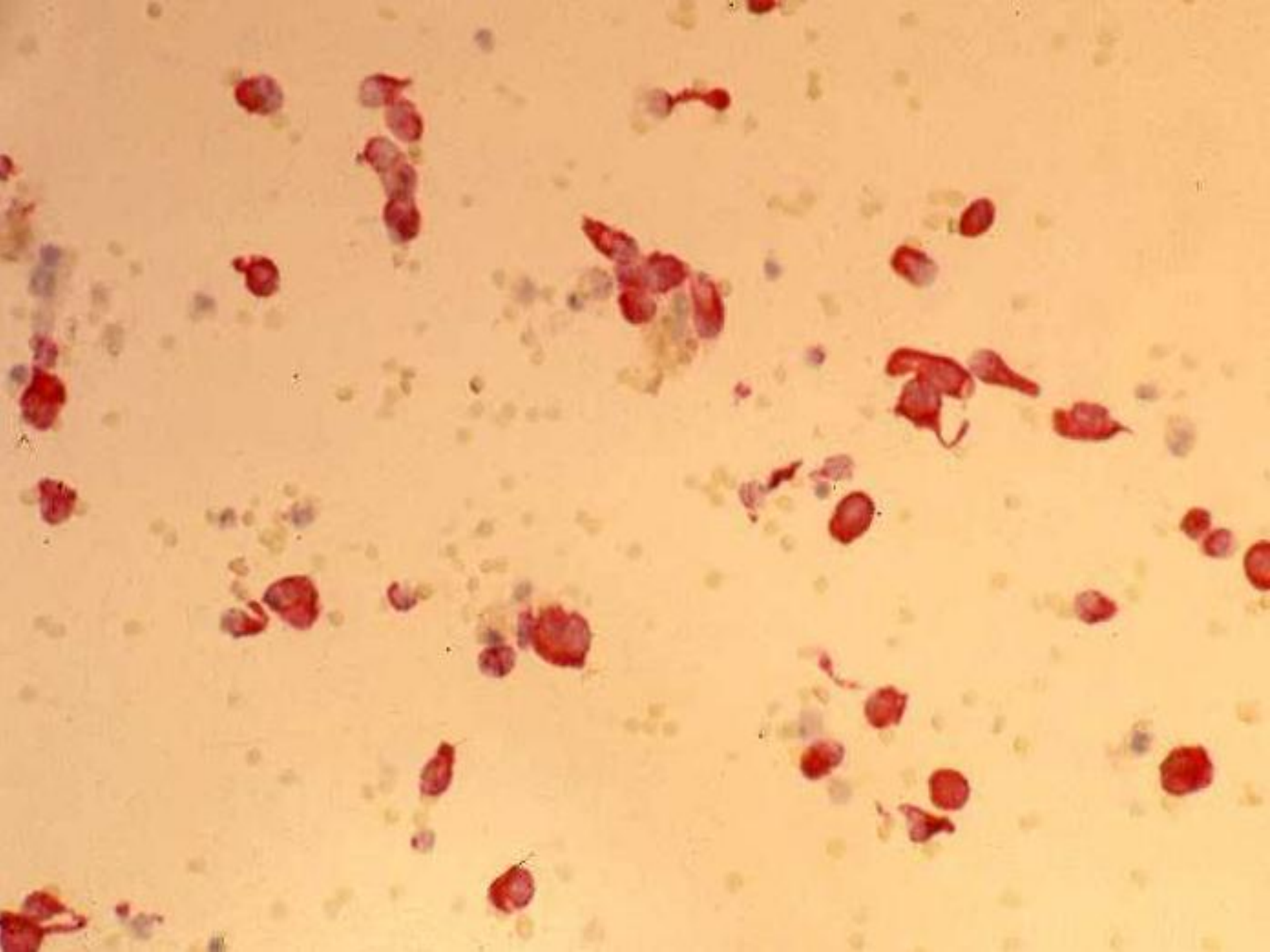


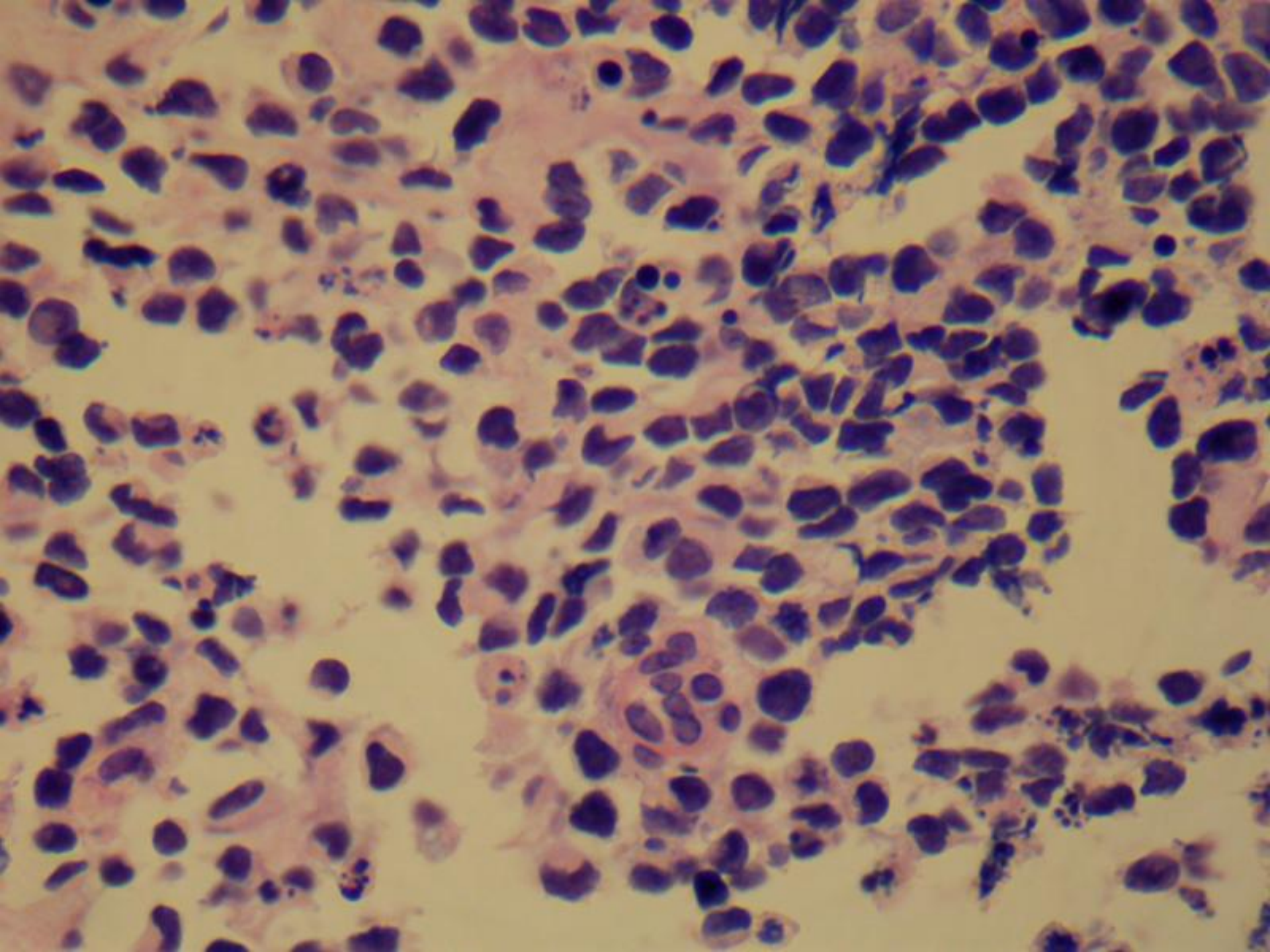
- 16, erkek
- 1 sene önce dudakta alveoler RMS öyküsü
- Mediasten üst bölümde 7cm çaplı konglomere kitle
- Klinik ön tanı: Lenfoma? RMS?

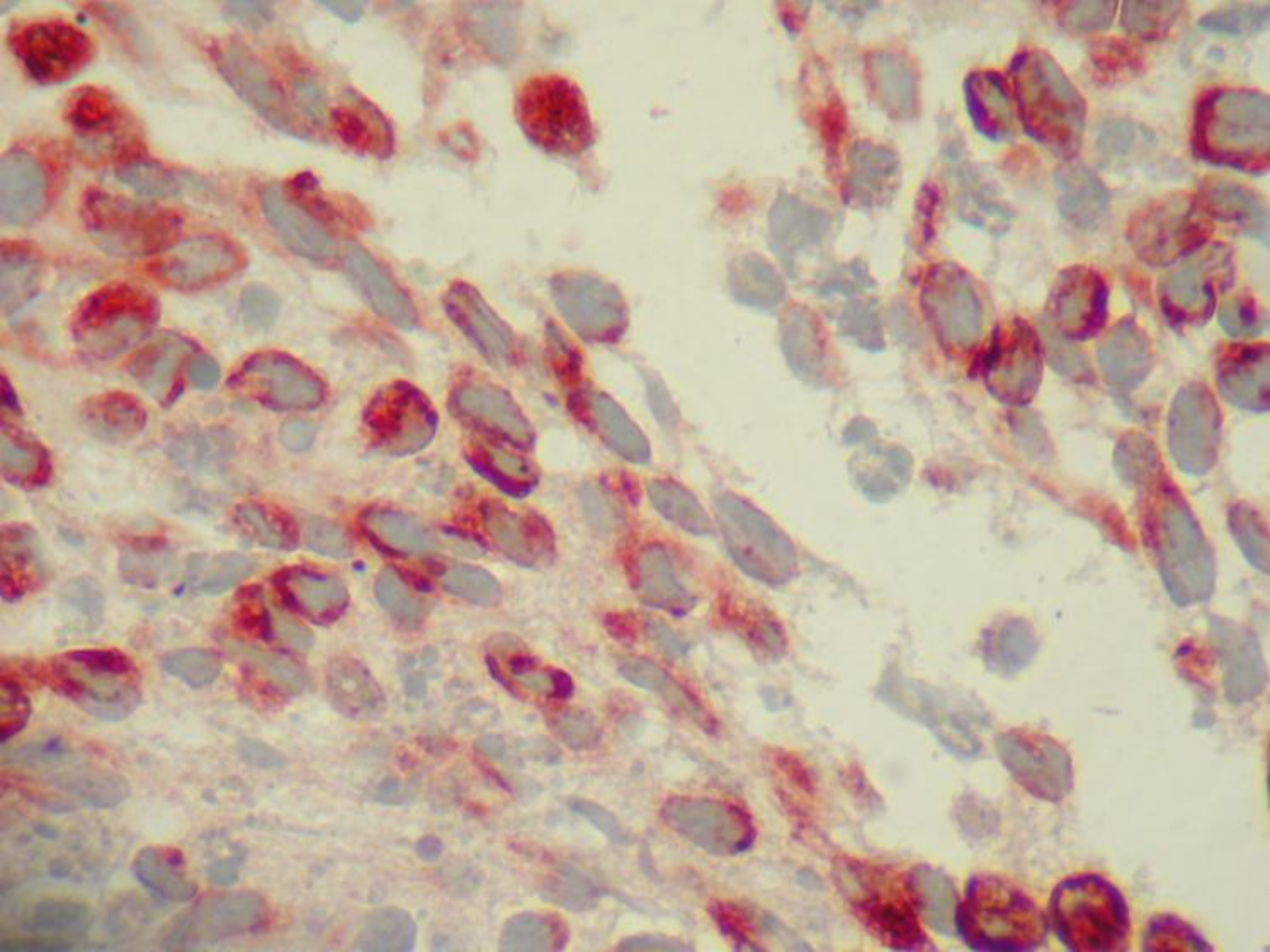




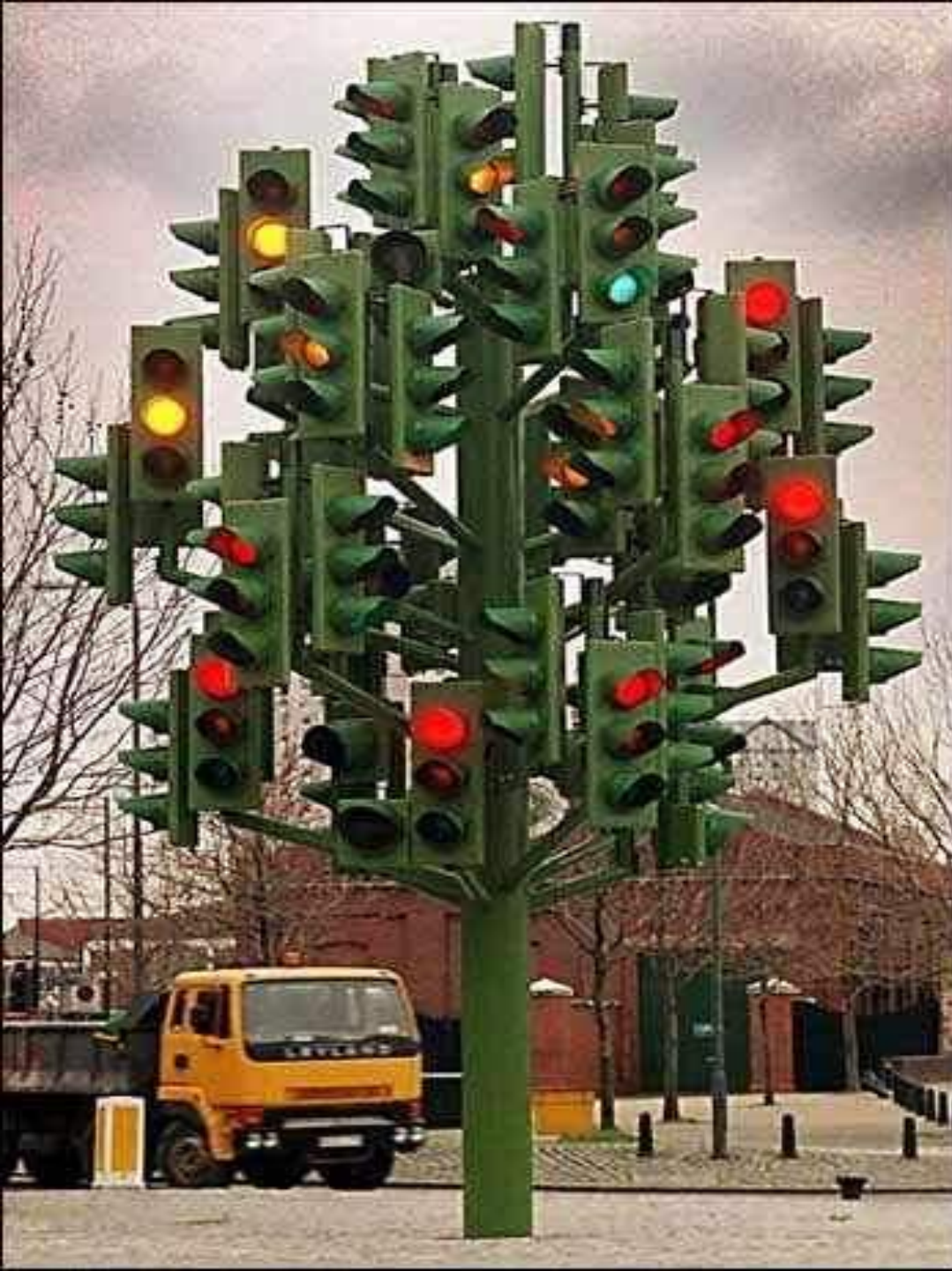










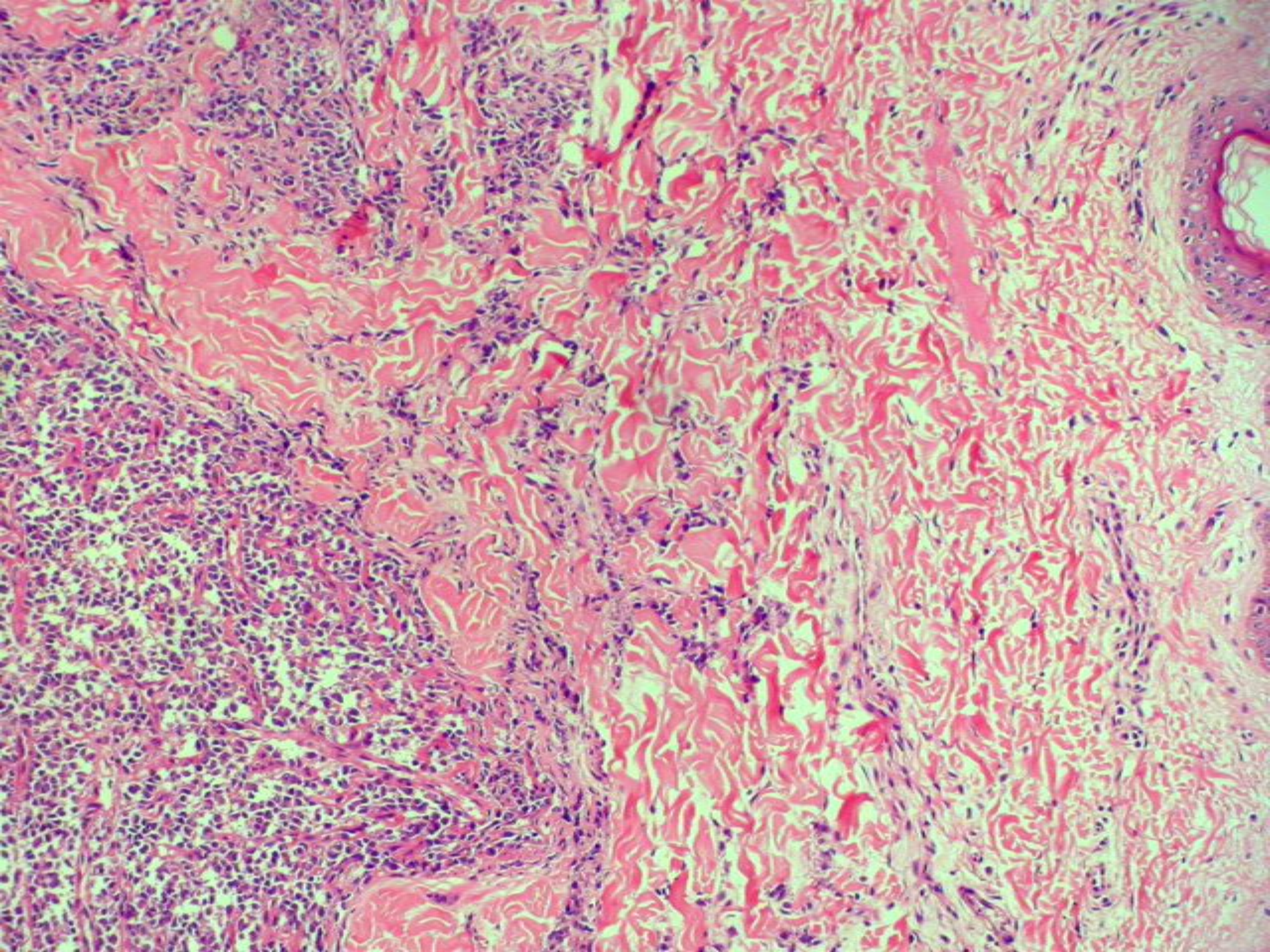


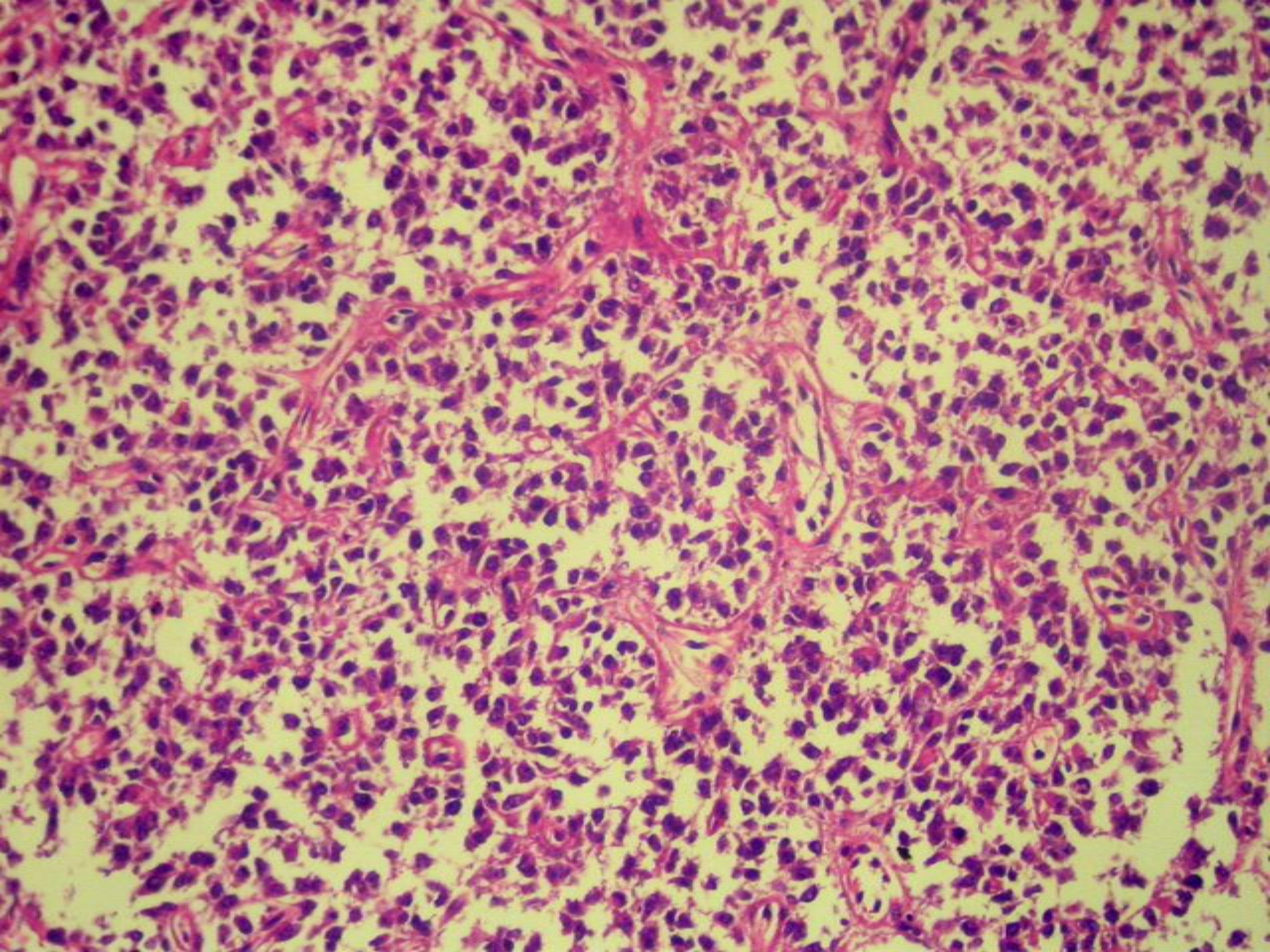


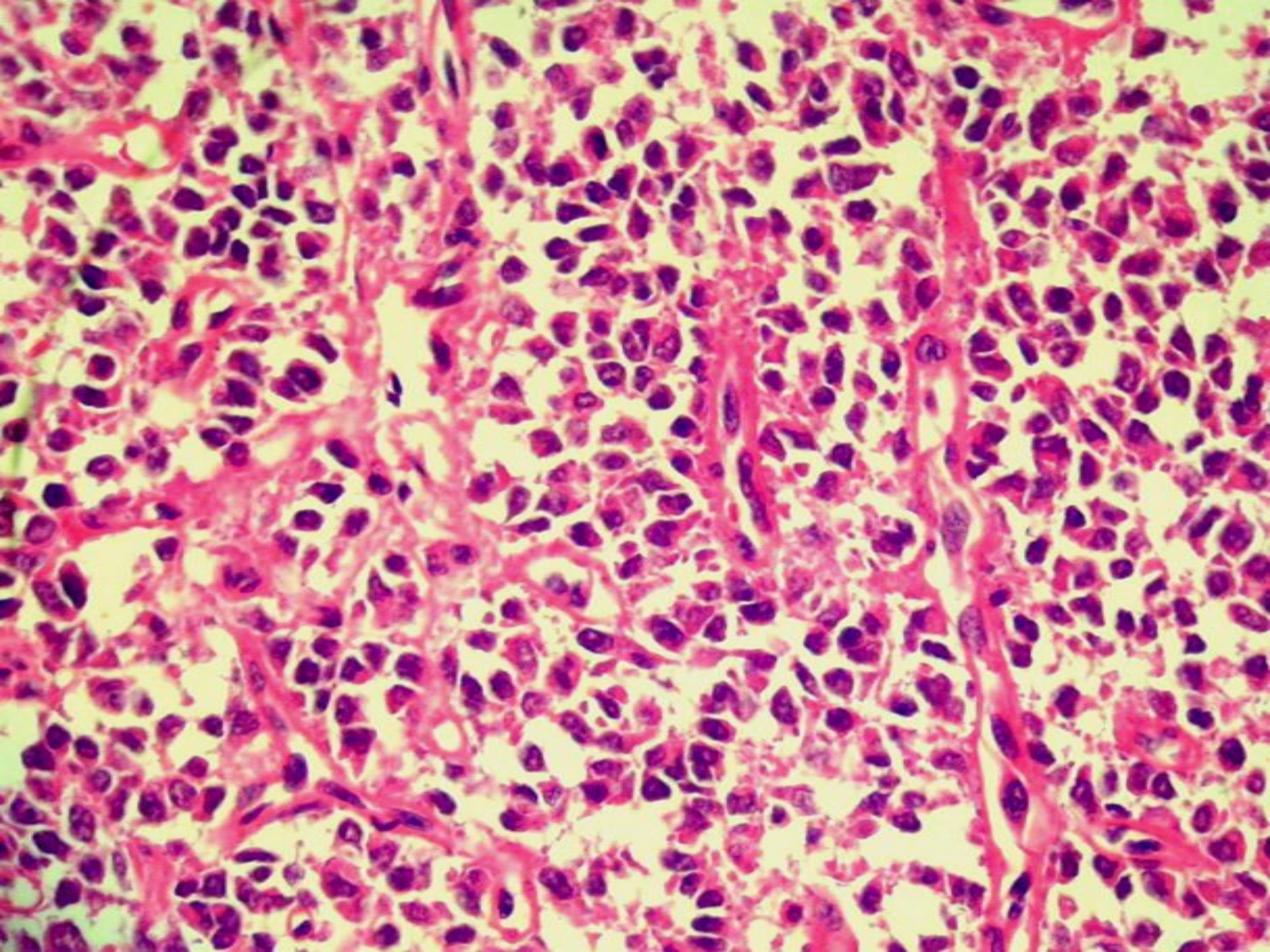
## PROBLEM OLGU

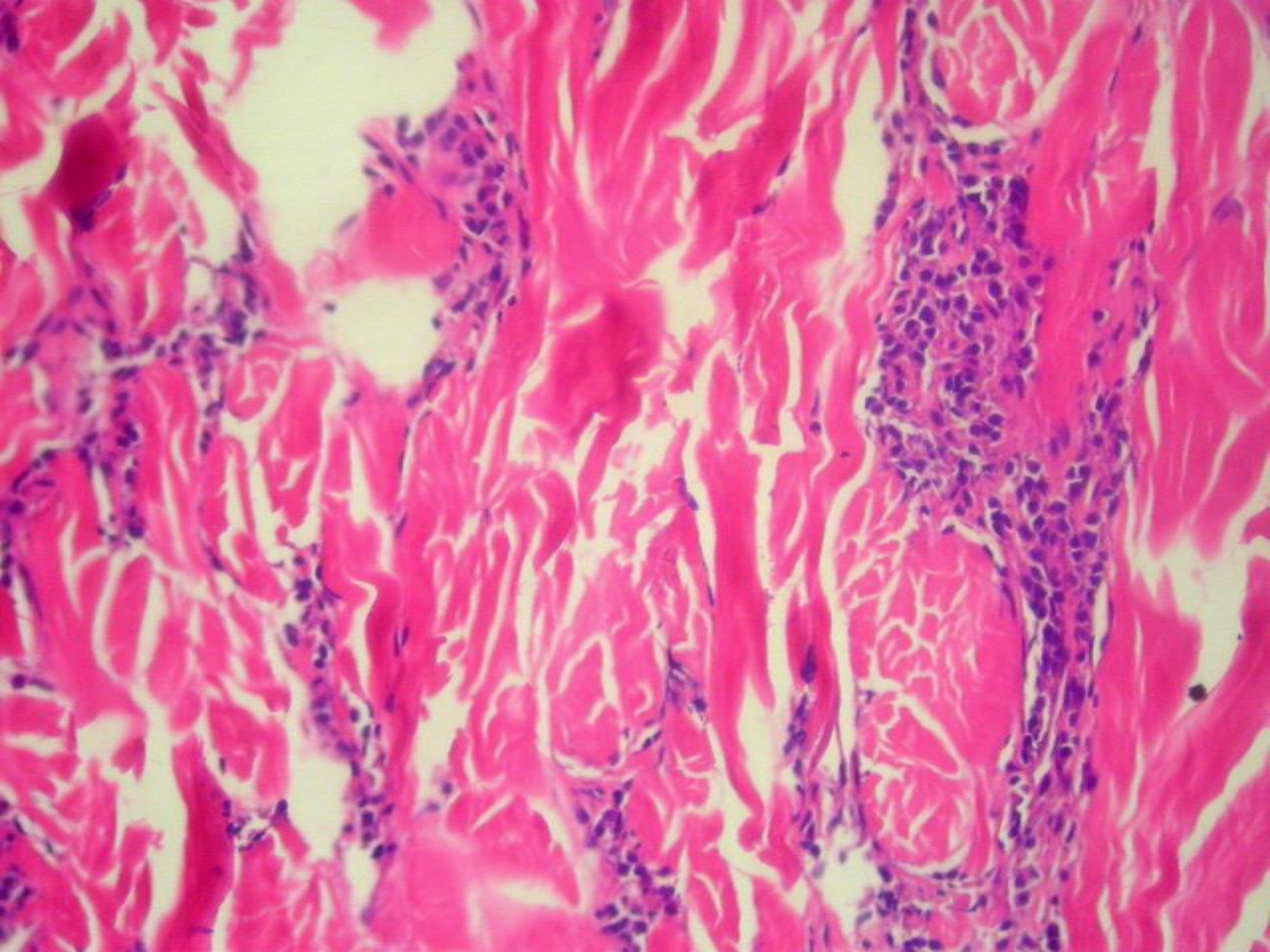
- 22 yaş bayan hasta
- Sağ boyunda yumuşak doku kitlesi
- Yüzeyel yerleşim
- Son zamanda büyüme hızında artış
- Eksizyonel biopsi

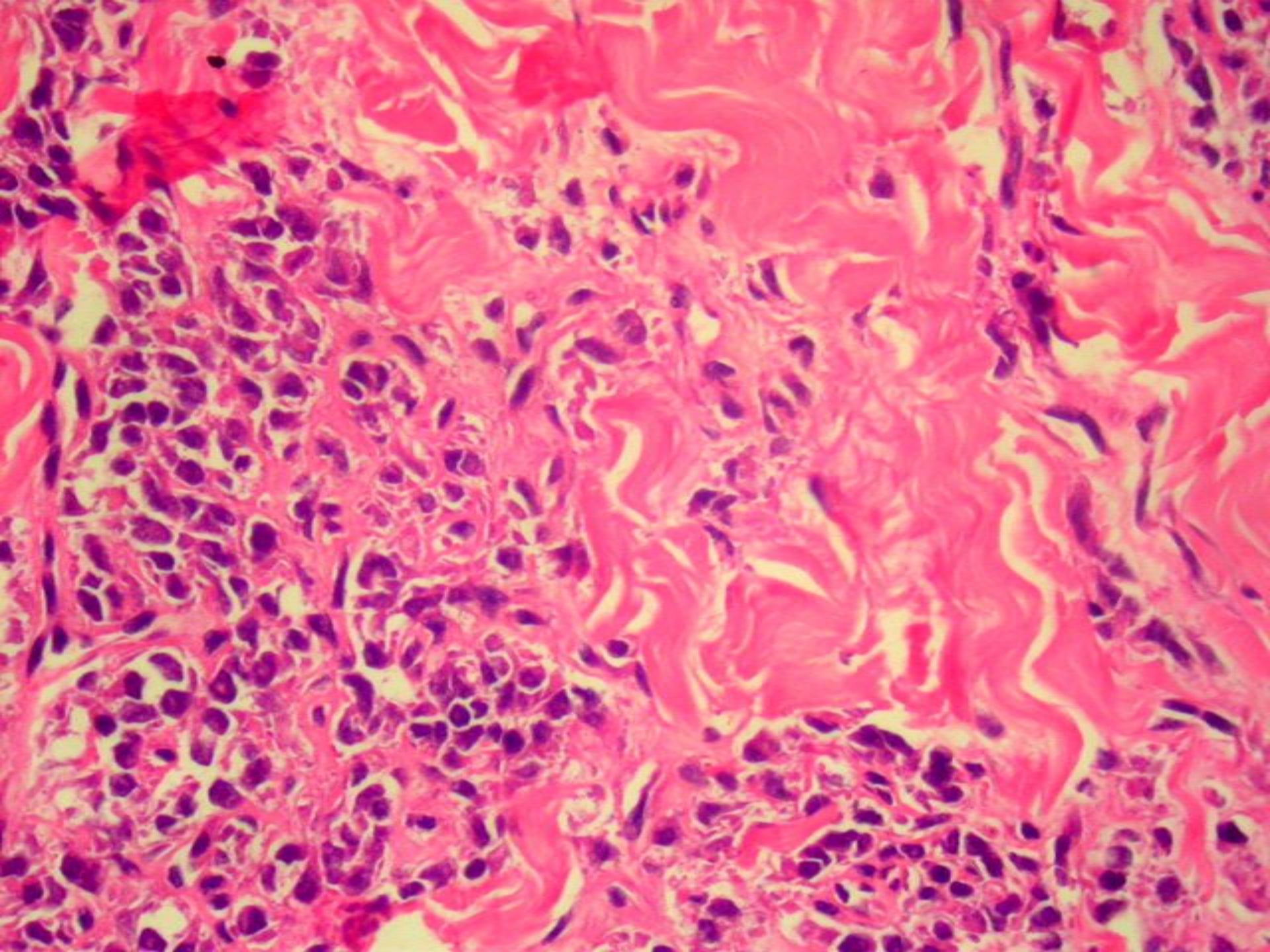




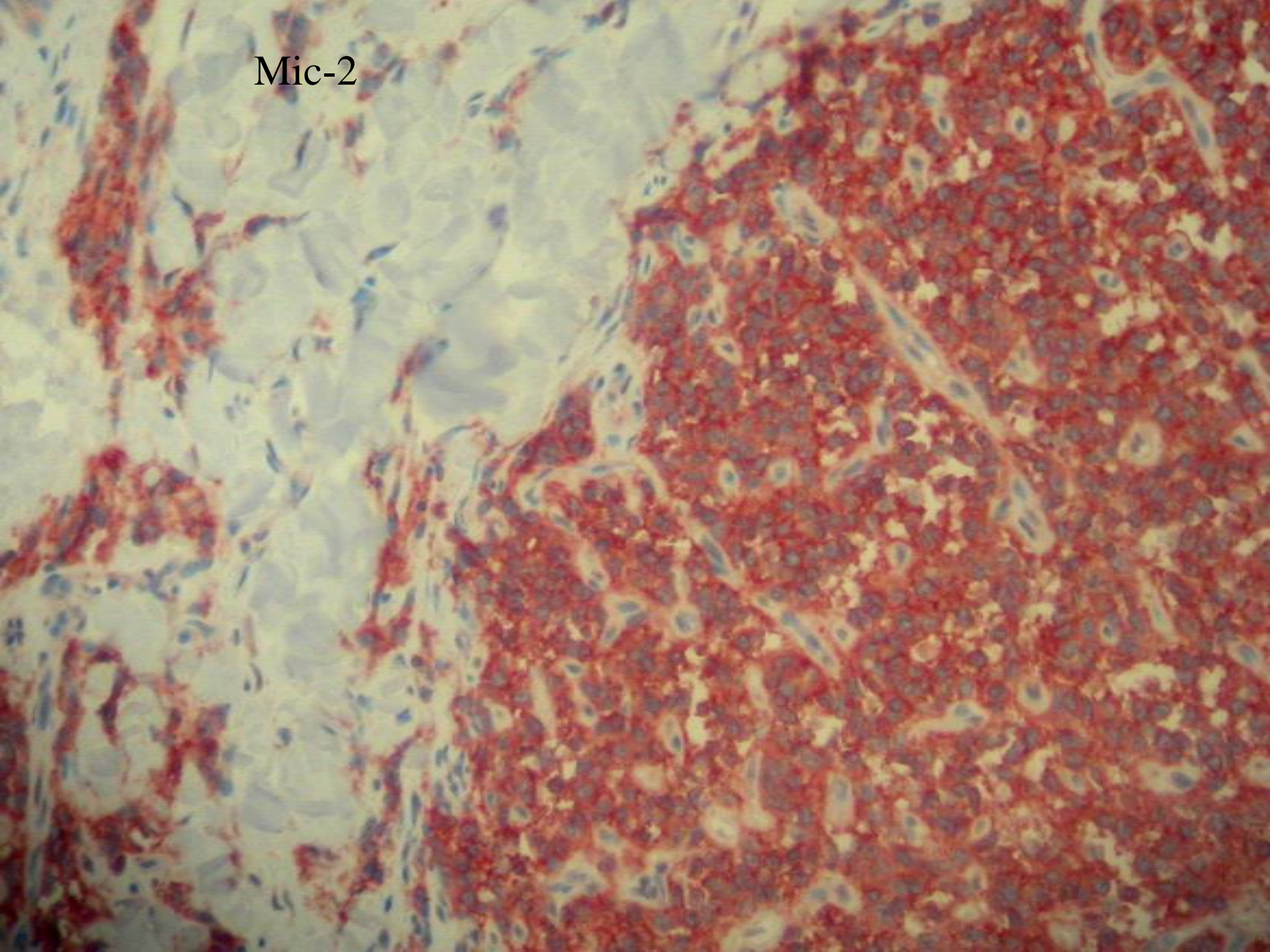


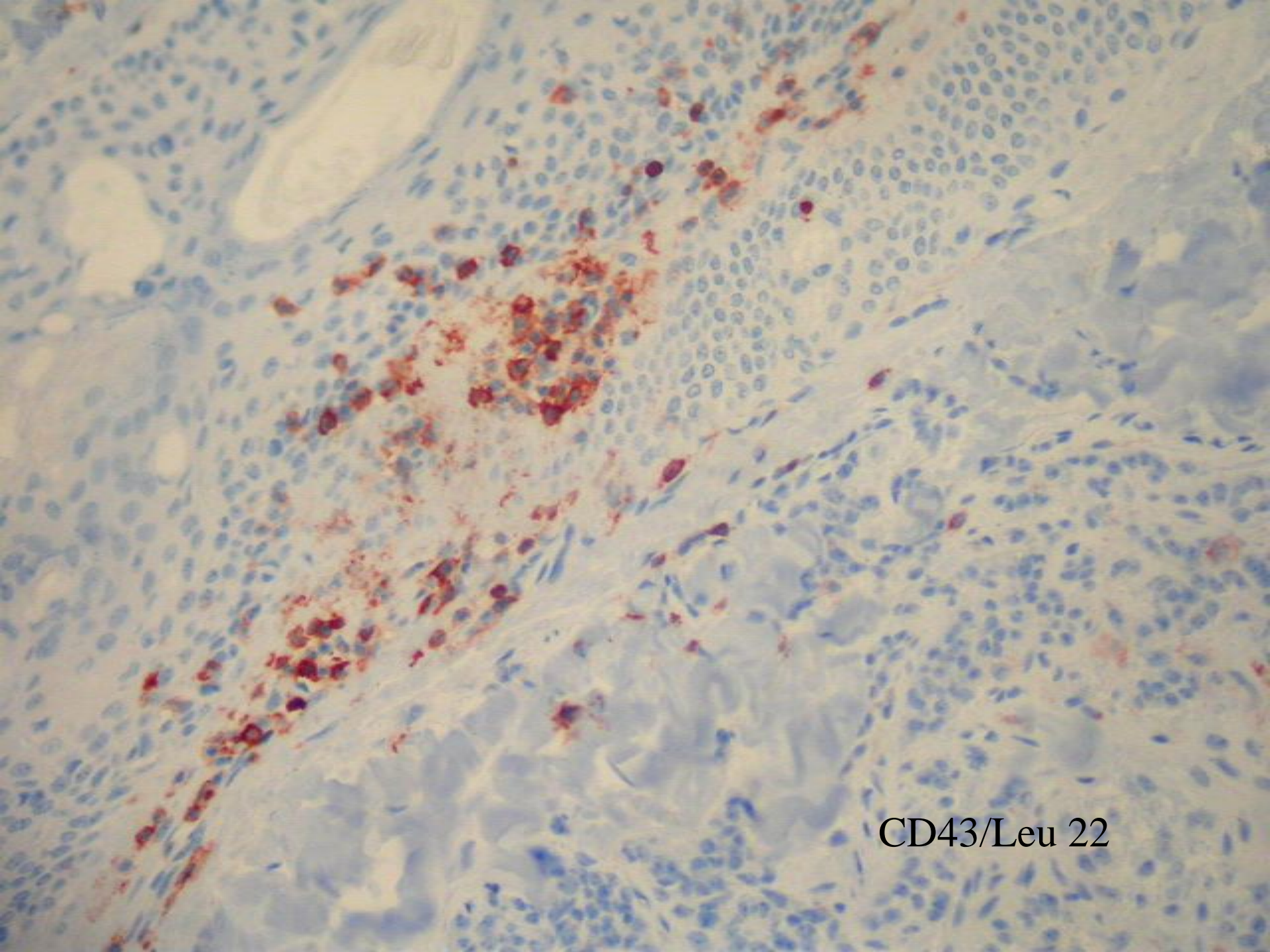






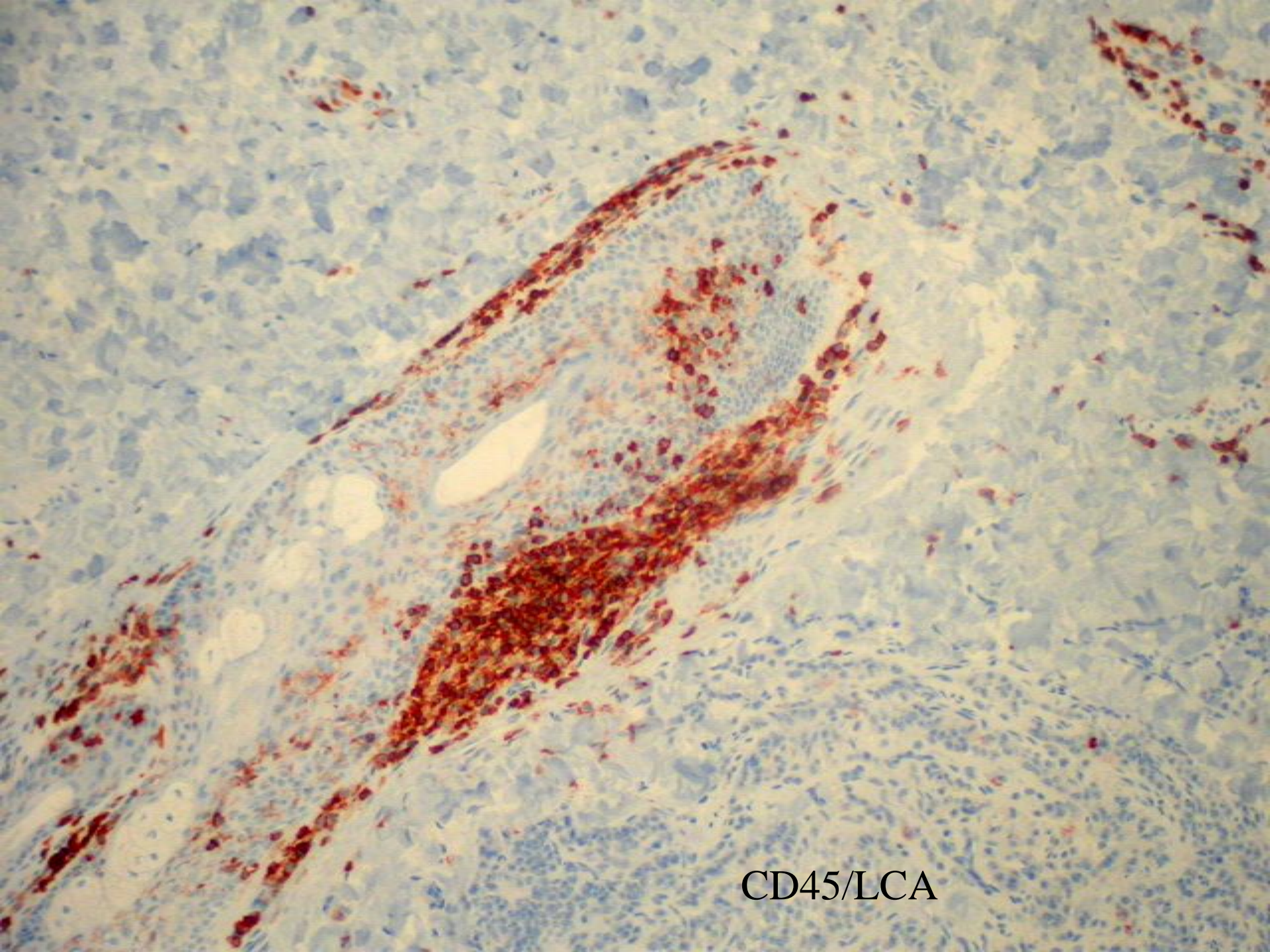
Mic-2





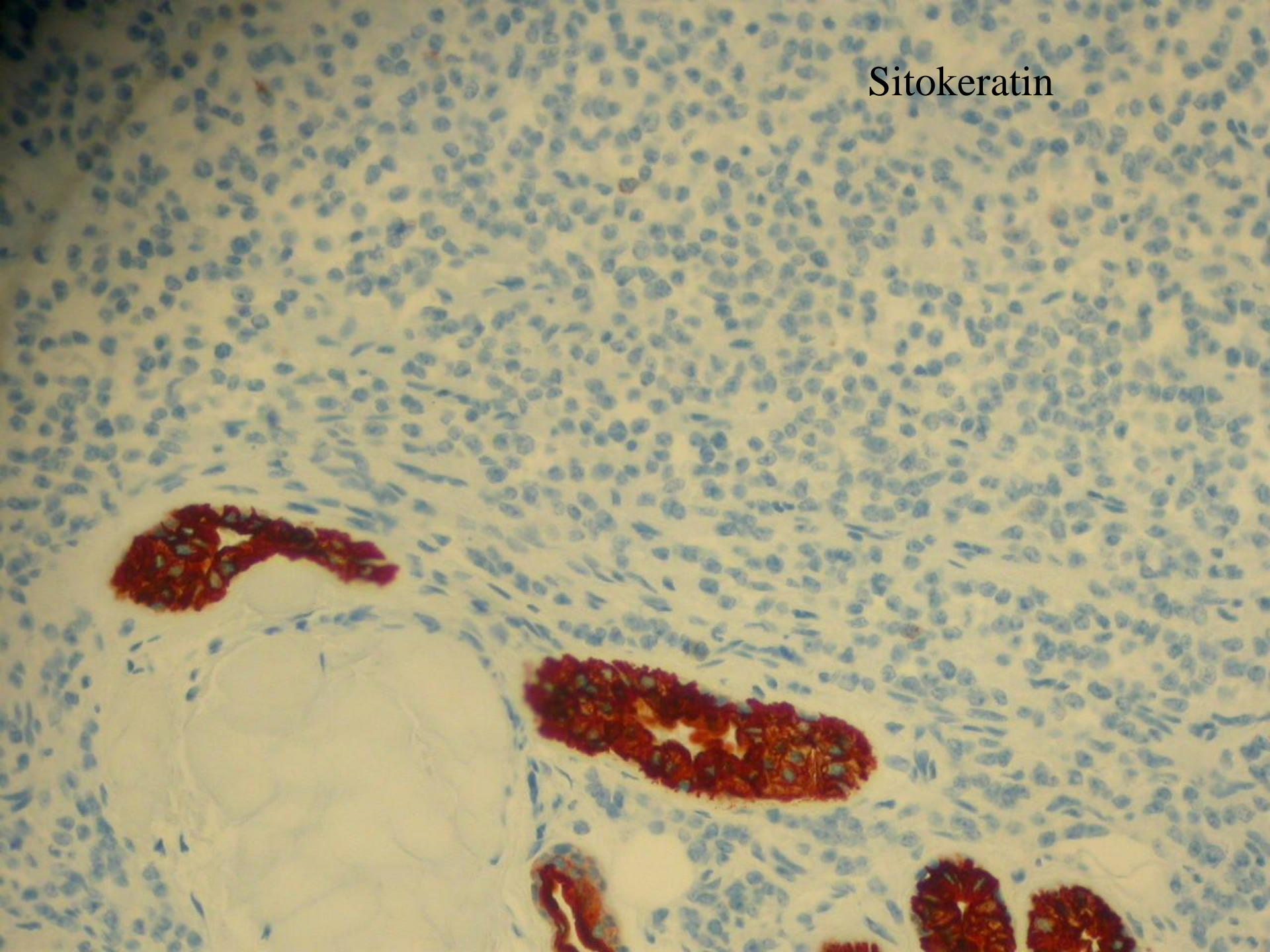
CD43/Leu 22

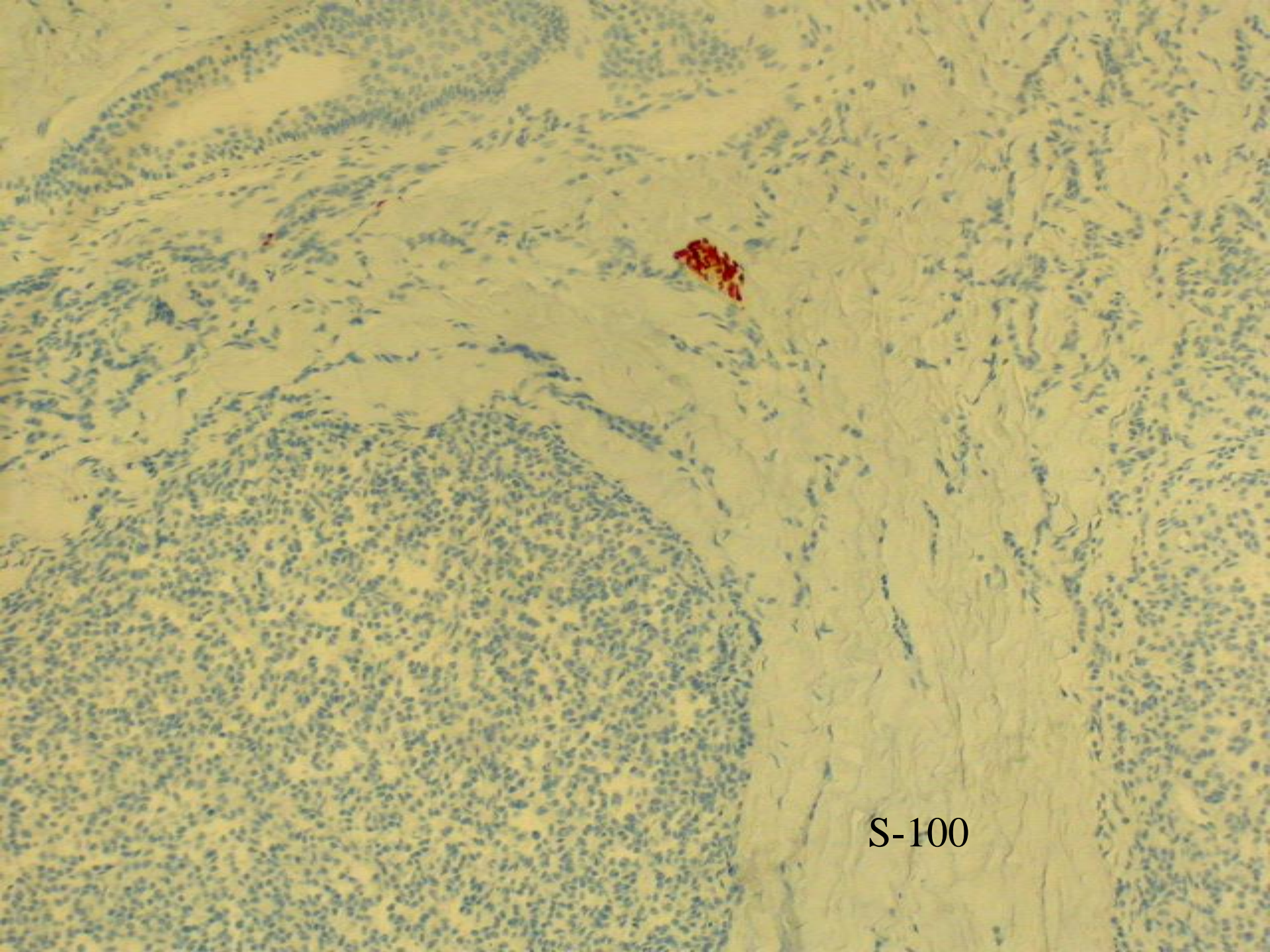




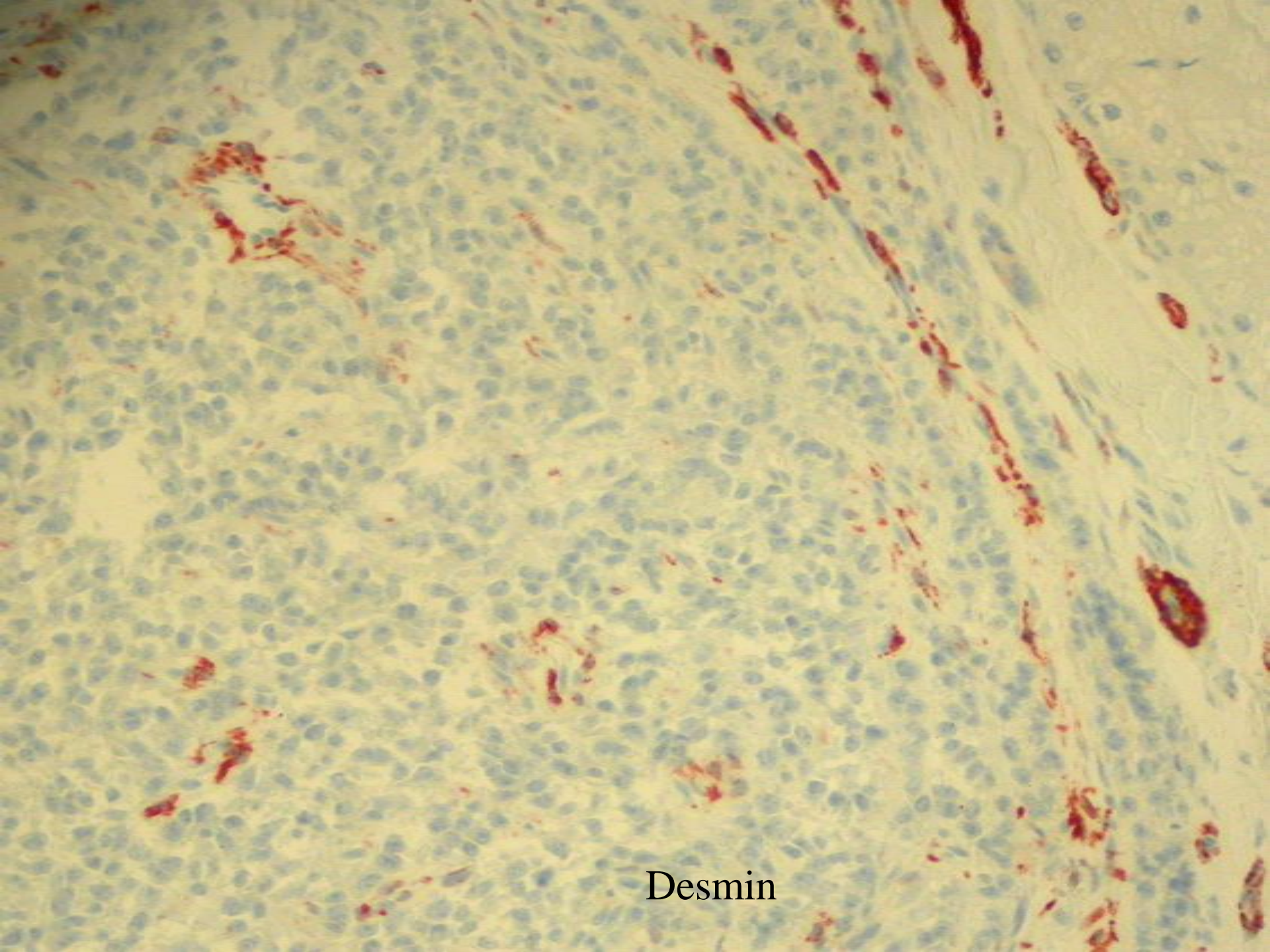
CD45/LCA

Sitokeratin

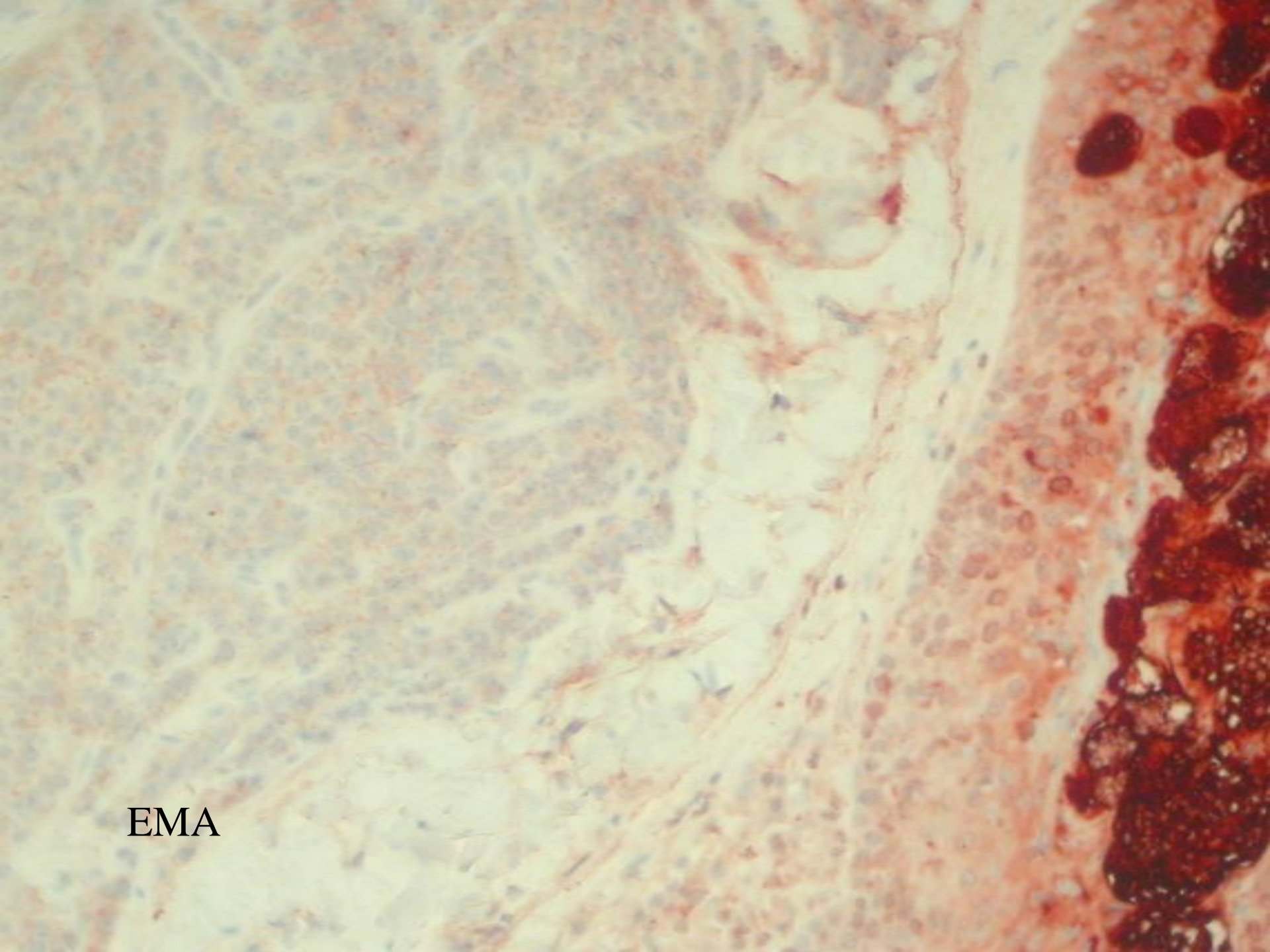




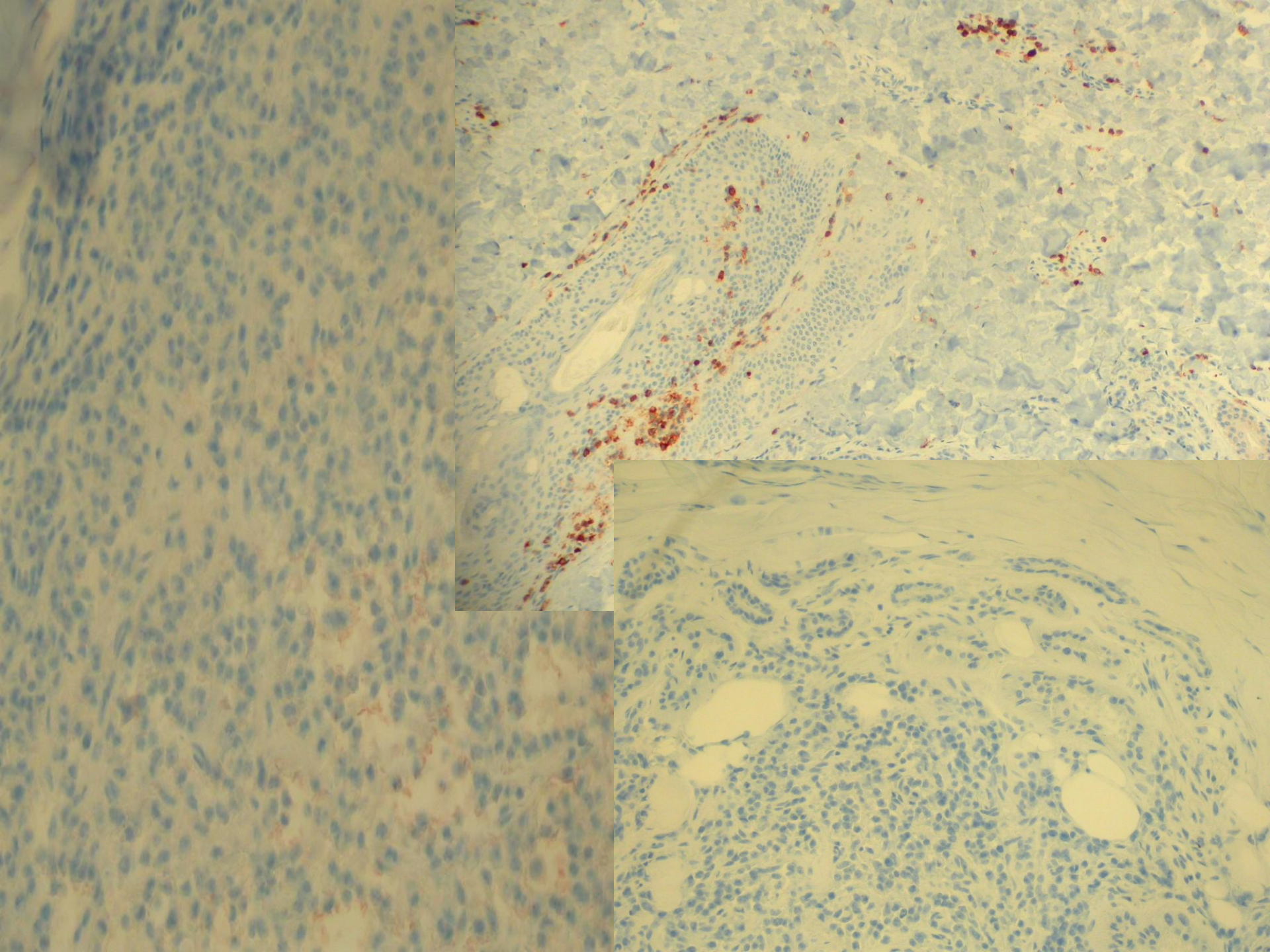
S-100



Desmin



EMA





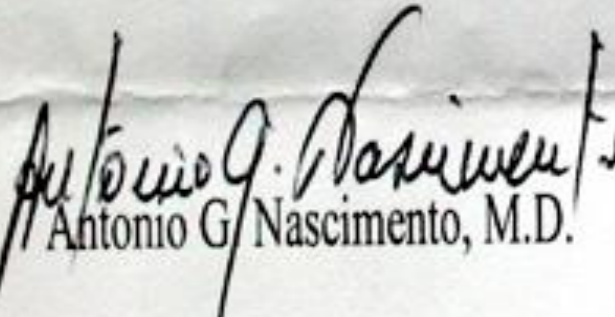
Dear Dr. Dervisoglu:

Thank you very much for sharing with us the materials representing tissue from the neck of this 22-year-old patient, Deniz Oral.

Our pathology report reads as follows: "Skin, right neck, biopsy (2324/03): Undifferentiated small cell neoplasm, superficially located. Immunostain for MIC-2 is positive, but all other markers are negative. Molecular genetics studies for Ewing's sarcoma/PNET and alveolar rhabdomyosarcoma are negative."

Again, thank you very much for sharing this interesting diagnostic problem with me. If I can be of any further assistance in the care of this patient, please do not hesitate to contact me. Your materials are enclosed.

Sincerely yours,

  
Antonio G. Nascimento, M.D.

MD Anderson  
Her şey negatif ama  
alvRMS kabul edin





Tuzaklara  
düşmeden  
değerlendirme