

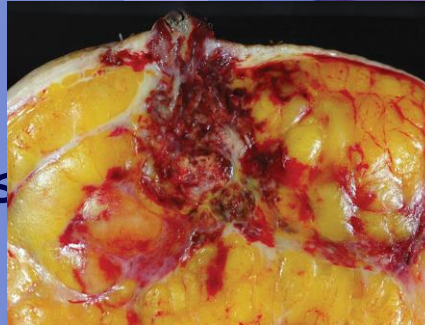
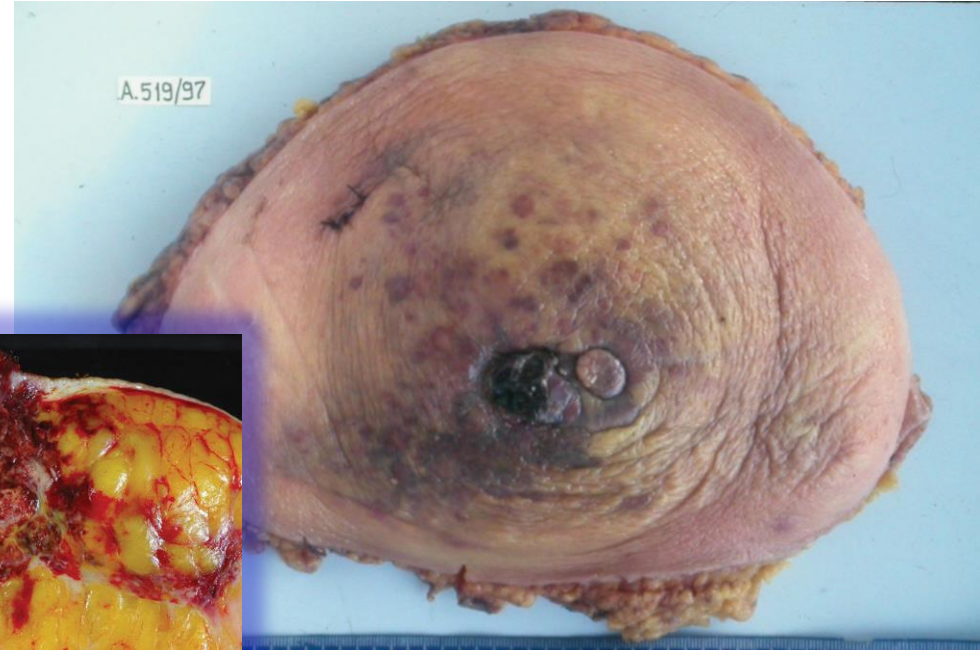


Epiteloid sarkom morfolojik spektrum ve ayırıcı tanısı

Prof Dr Sergülen Dervişođlu
İ.Ü. Cerrahpaşa Tıp Fakóltesi
Patoloji Anabilim Dalı

Yüzeyel yerleşimli sarkomlar

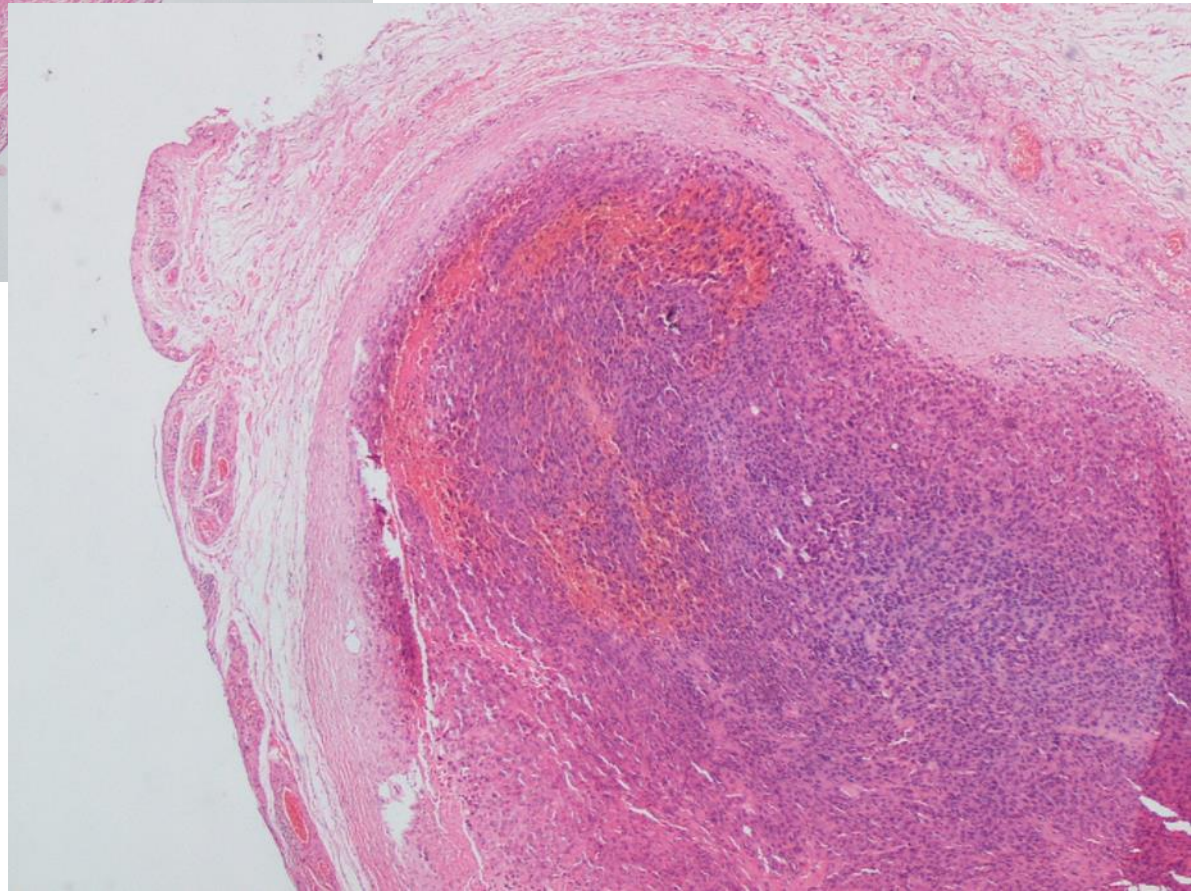
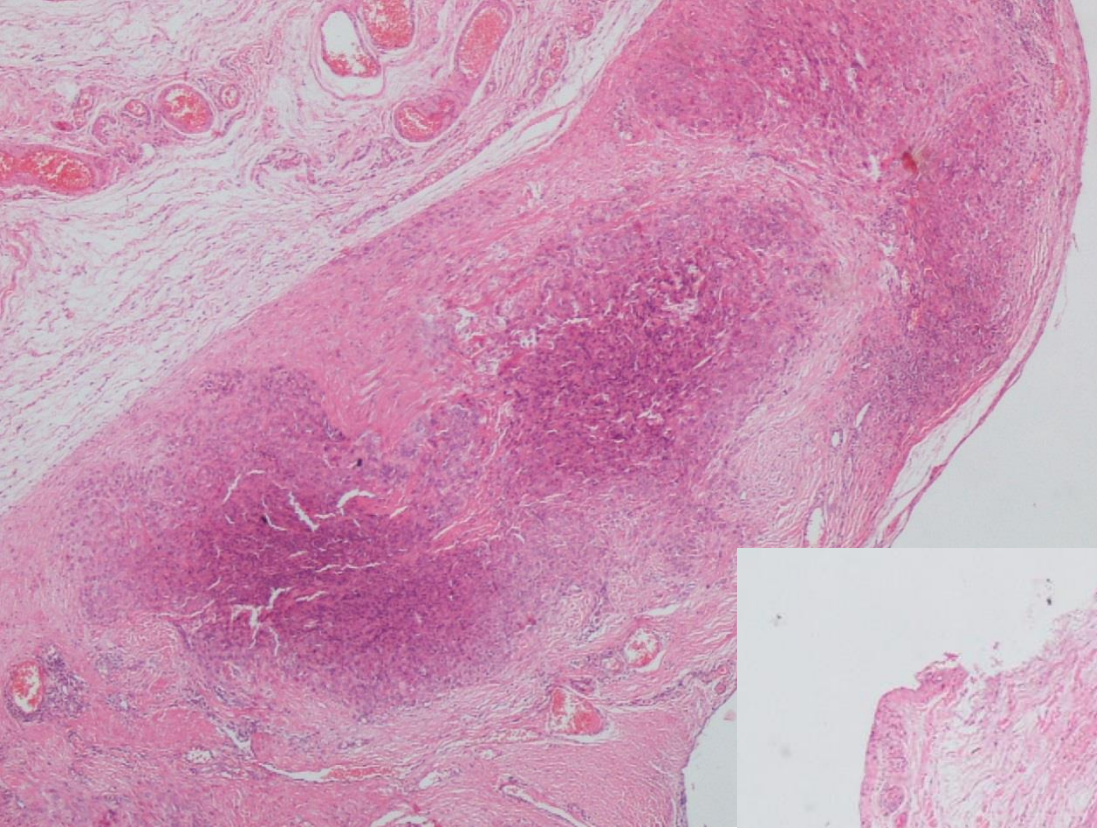
- Dermatofibrosarkoma protuberans
- Epiteloid sarkom
- Angiomatoid fibröz histiyositom
- Pleksiform fibrohistiyositom
- Miksofibrosarkom /miksoid MFH
- Anjiosarkom
- Kaposi sarkomu
- Atipik fibroksantom
- Kutanöz epiteloid rabdomyosarkom

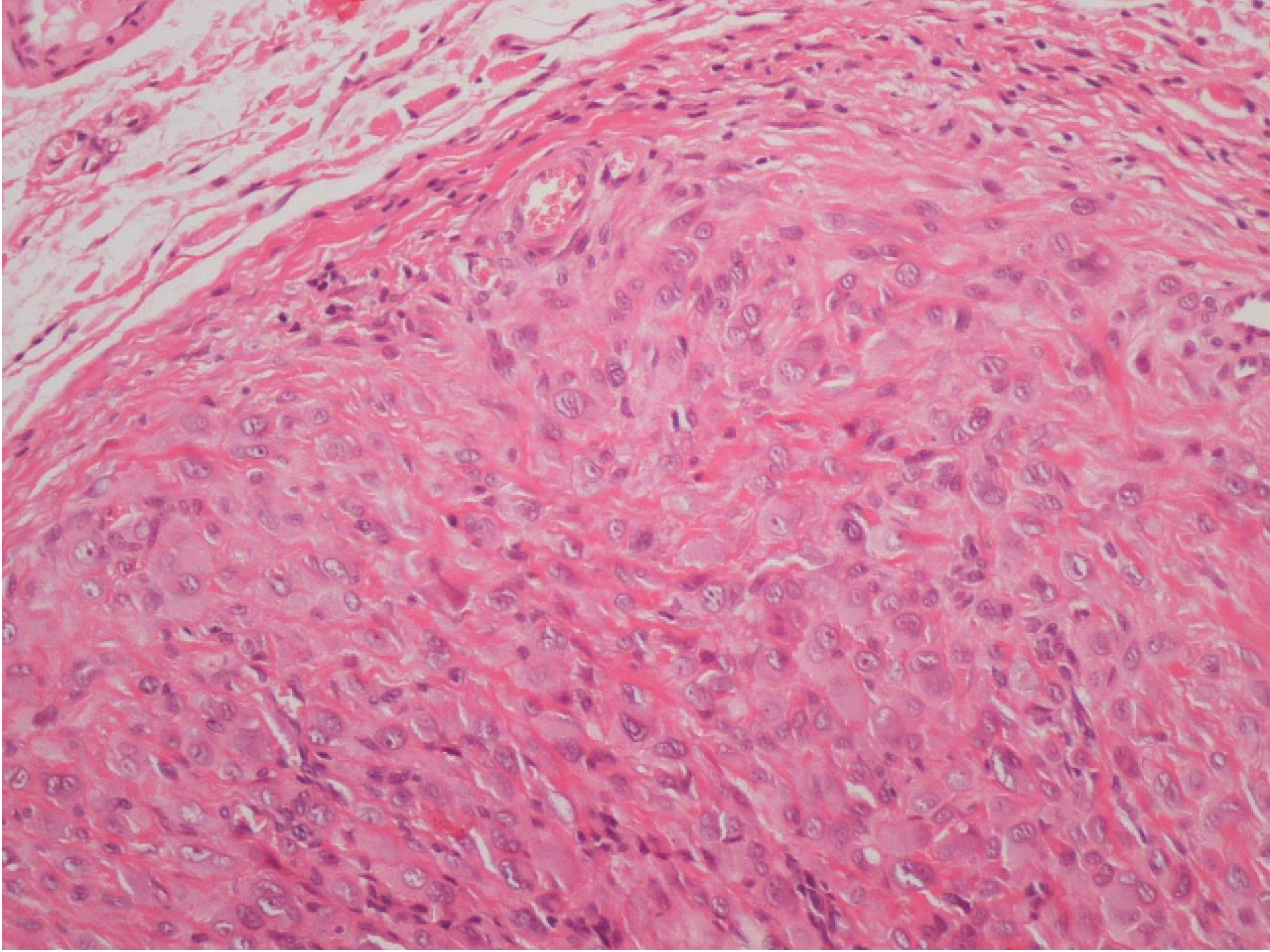


- Genç erişkin hastada, 1 senedir elde şişlik
- Ne olabilir?
- Tendon tümörü, **epiteloid sarkom**, sinoviyal sarkom, rabdomyosarkom, anjiyosarkom..vs

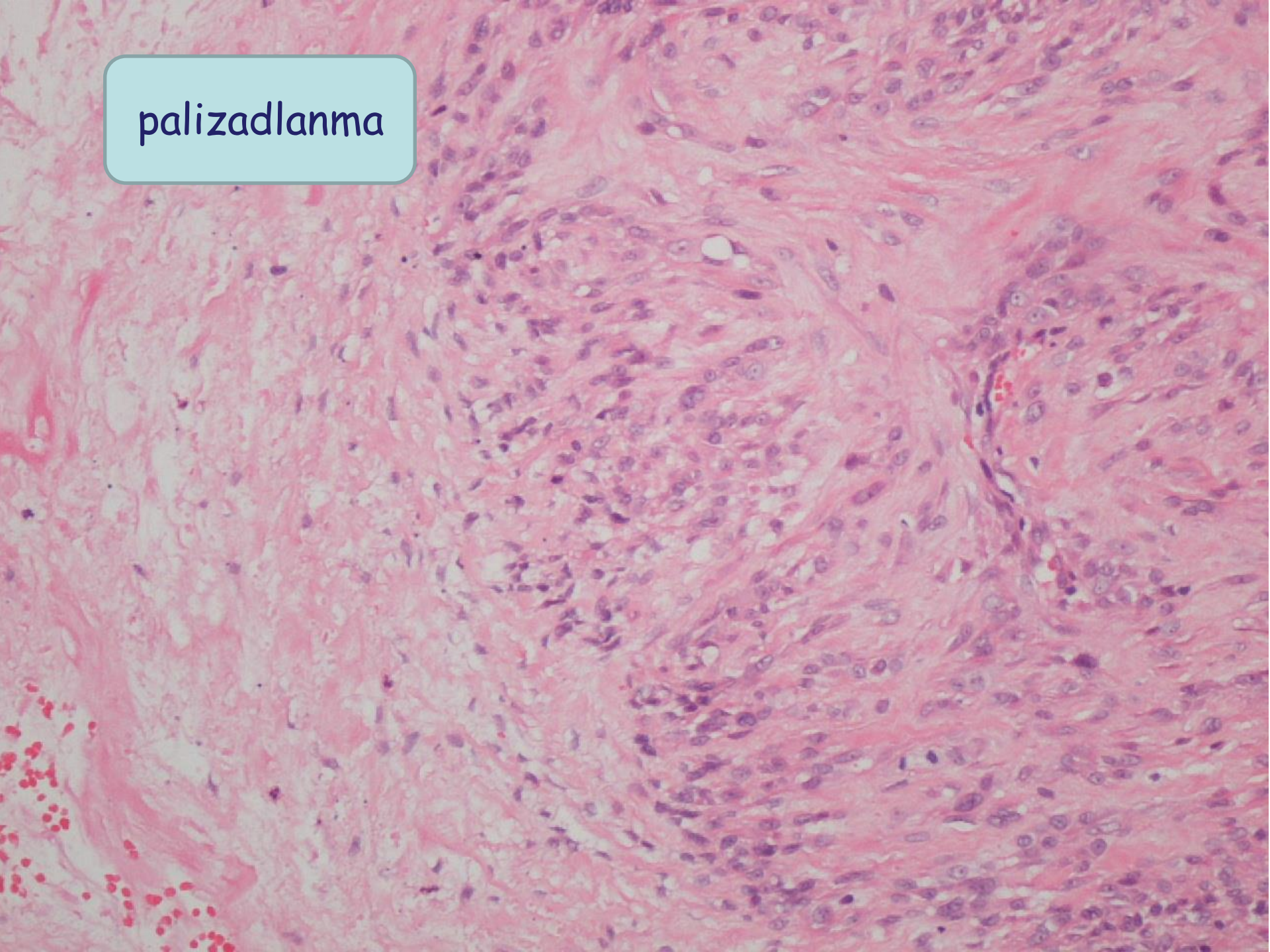


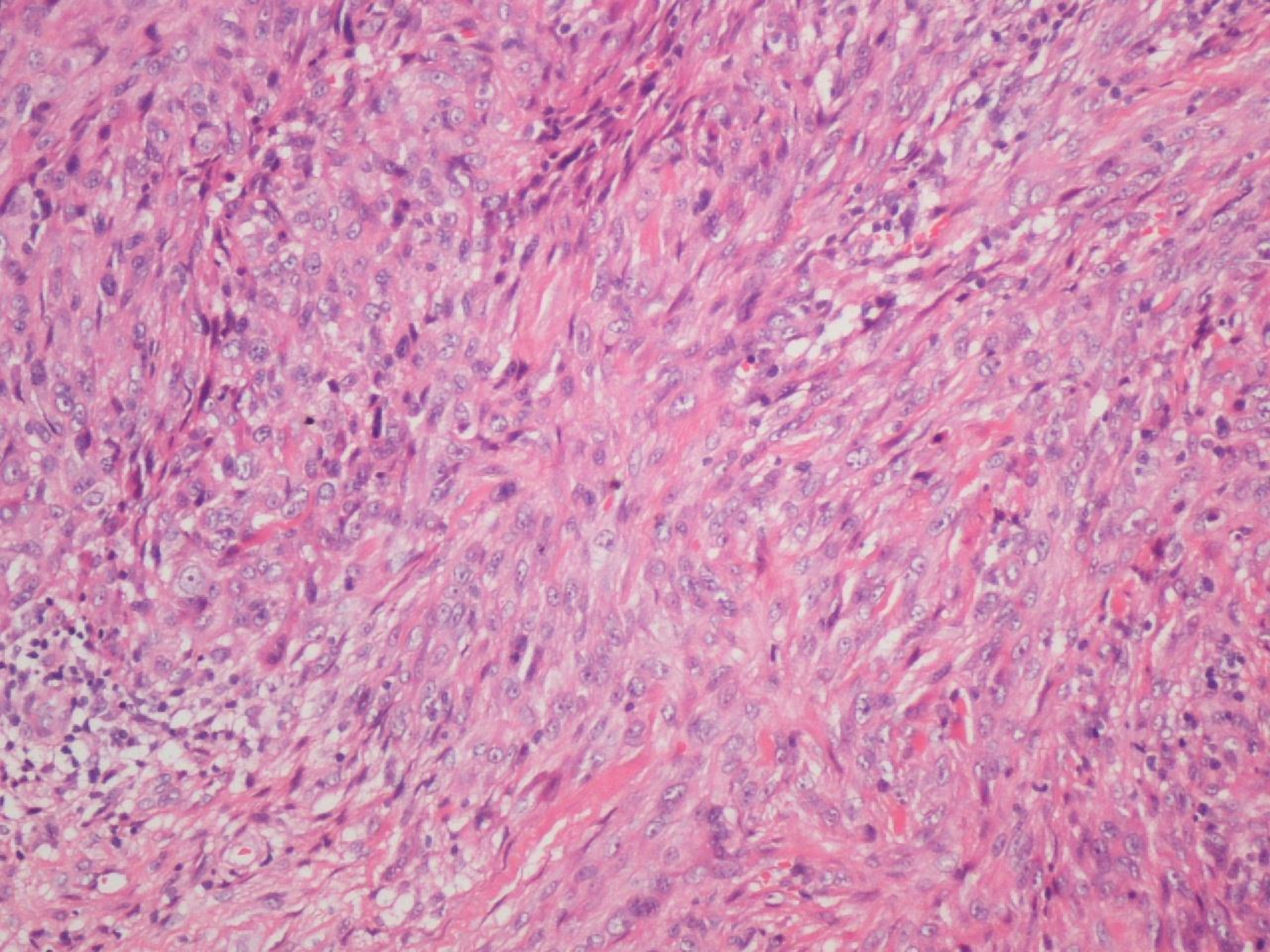
Prof Dr
Bilge Bilgiç
arşivi

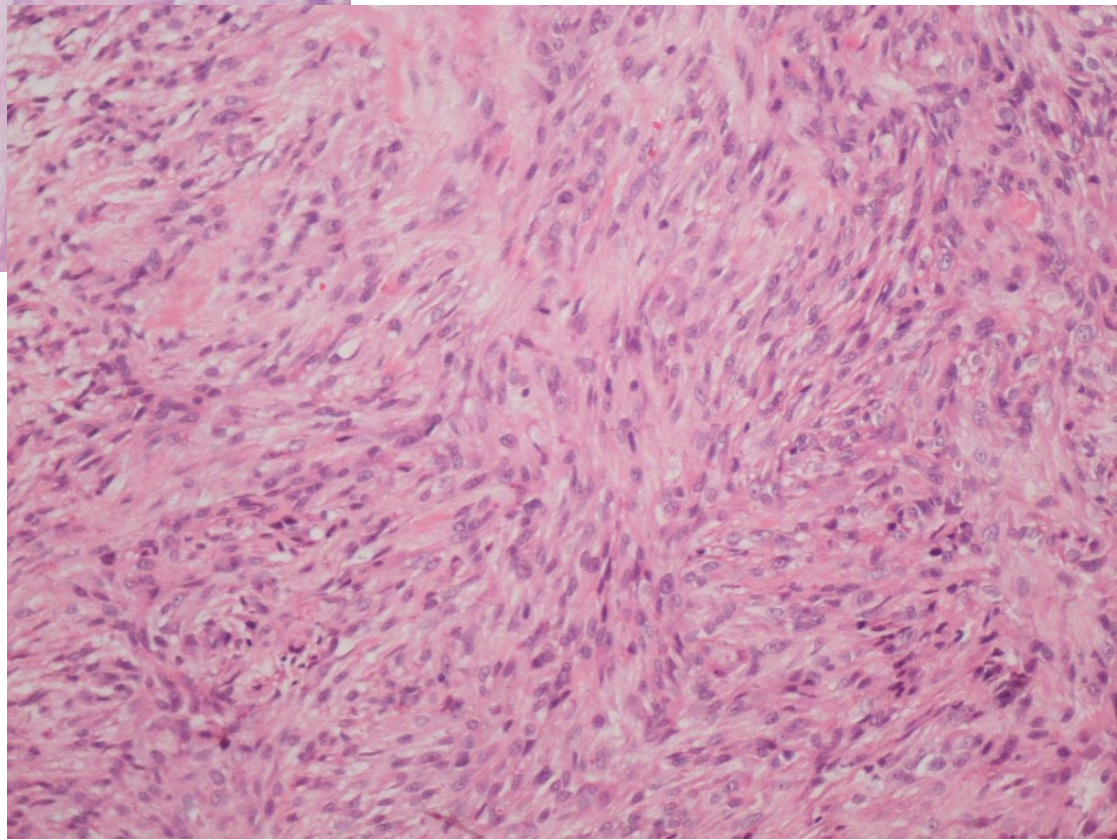
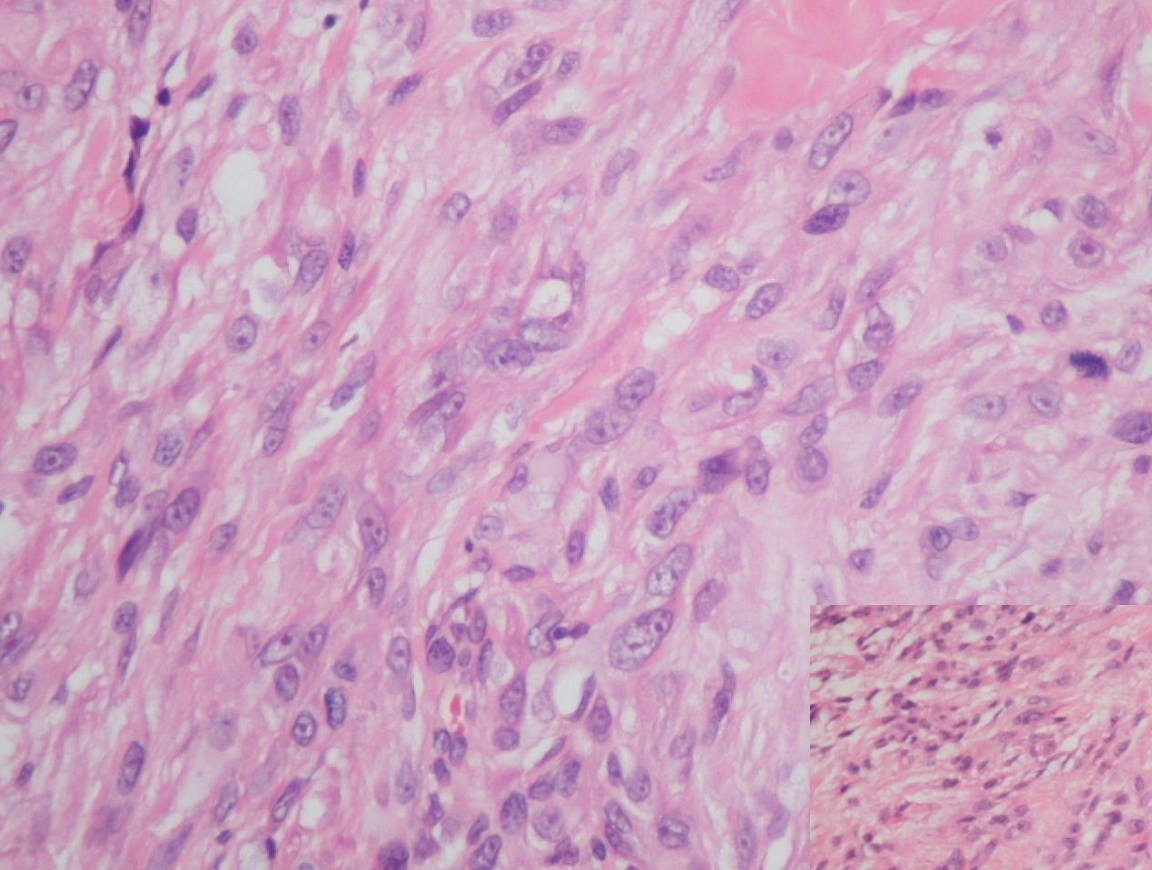


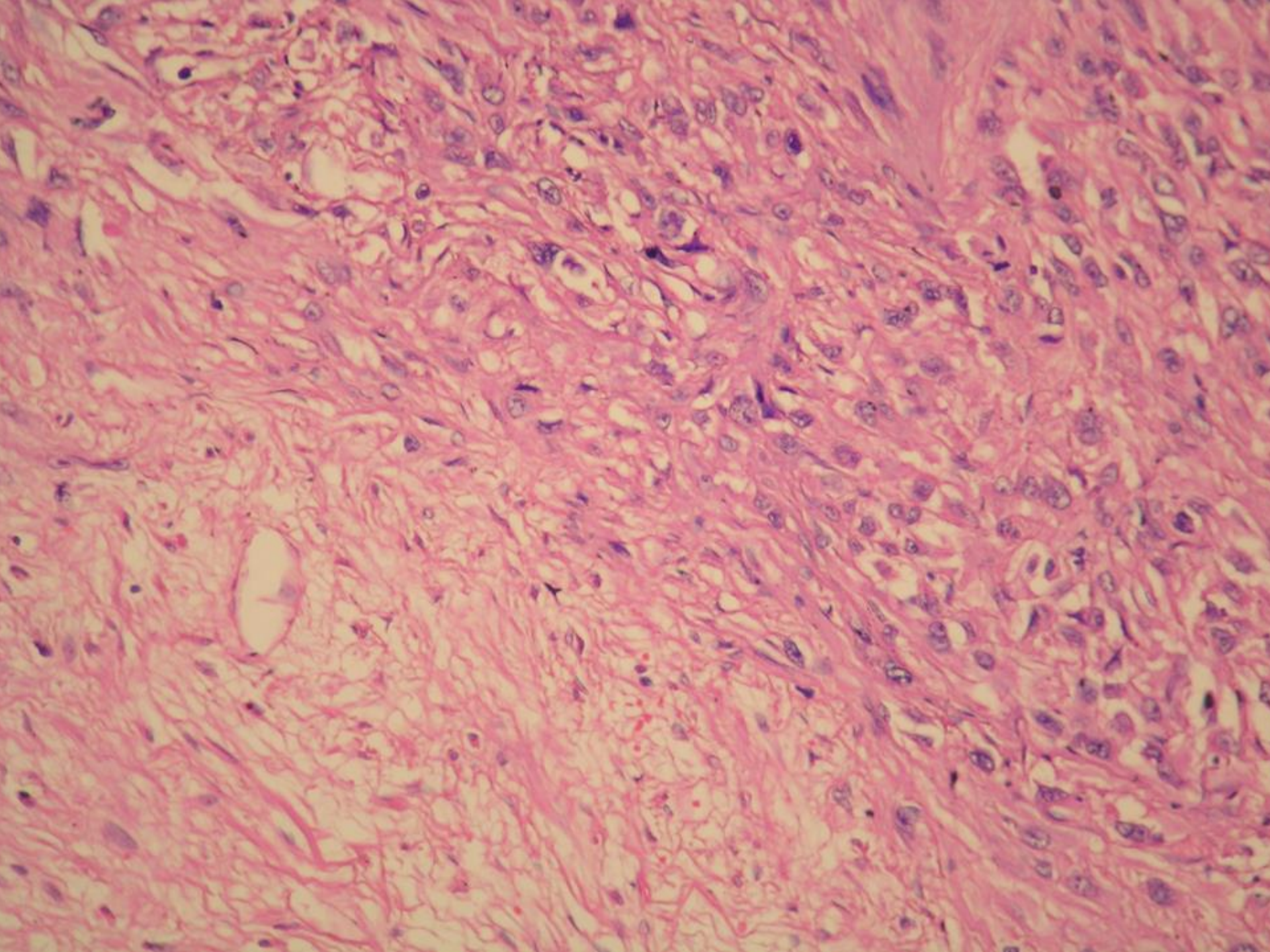


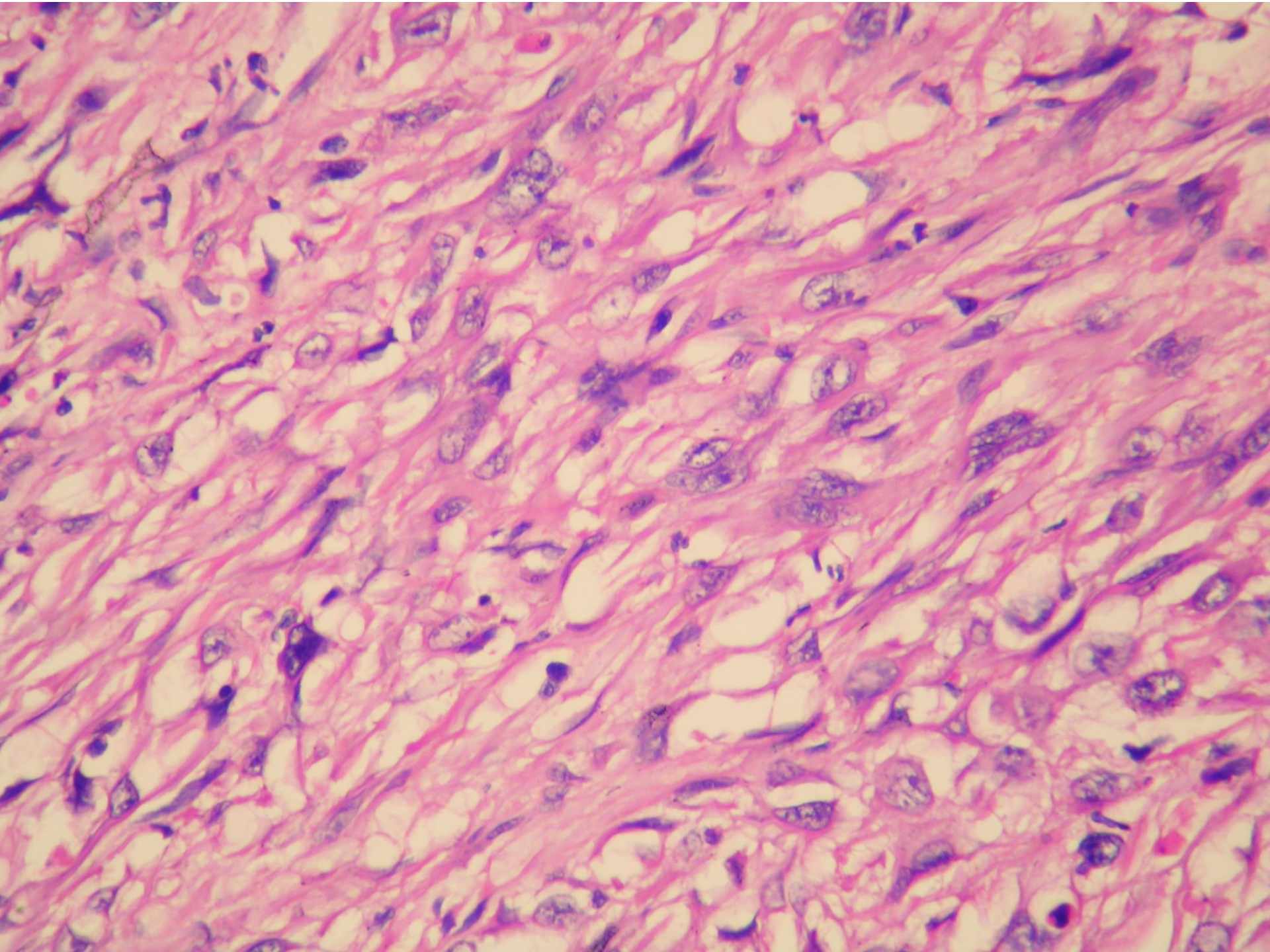
palizadlanma

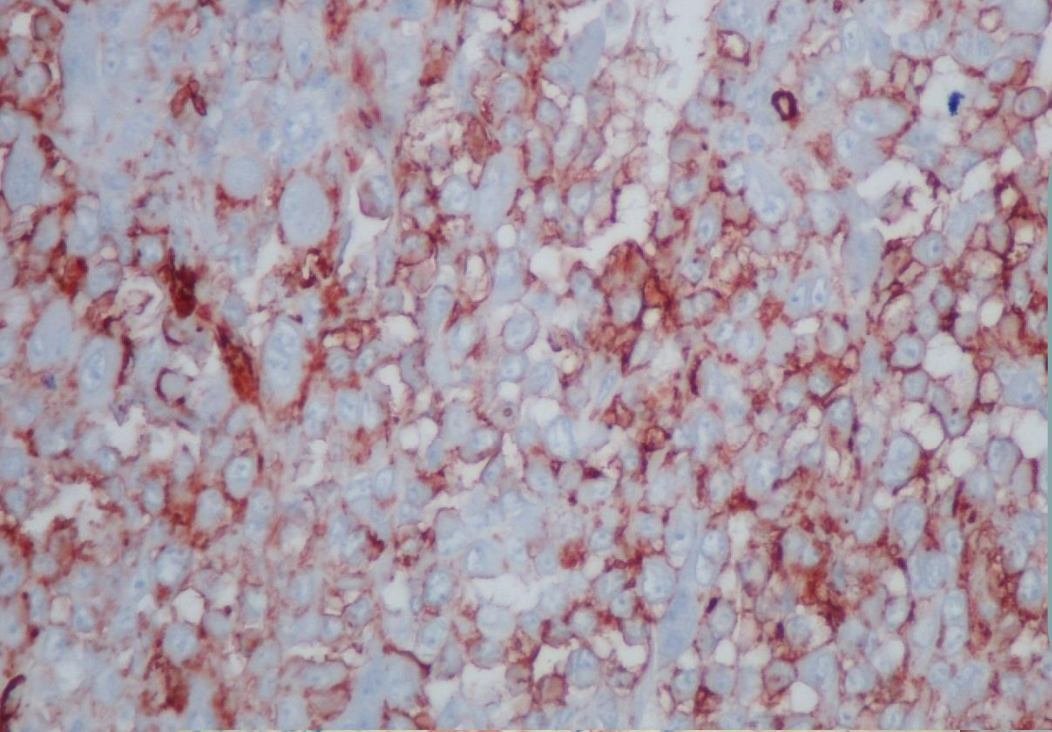




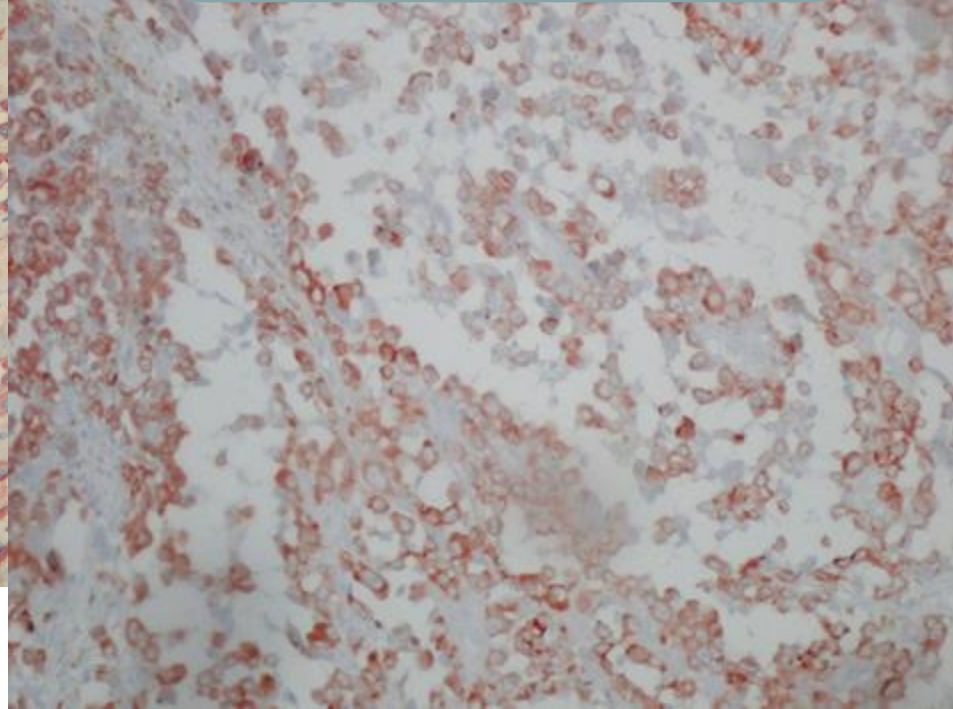
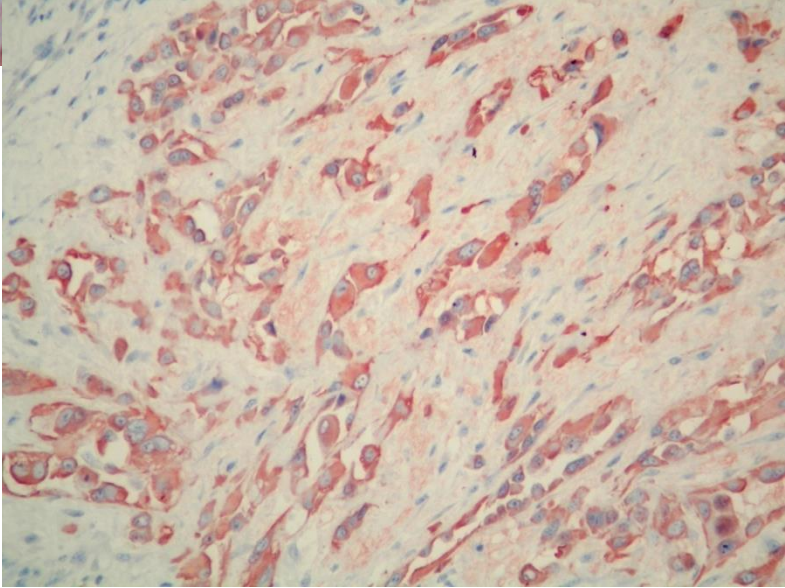








İHK: PSK, EMA ve vimentin (+)
CD34 %50(+)
SMA üçte bir olguda (+)
INI-1 (-)

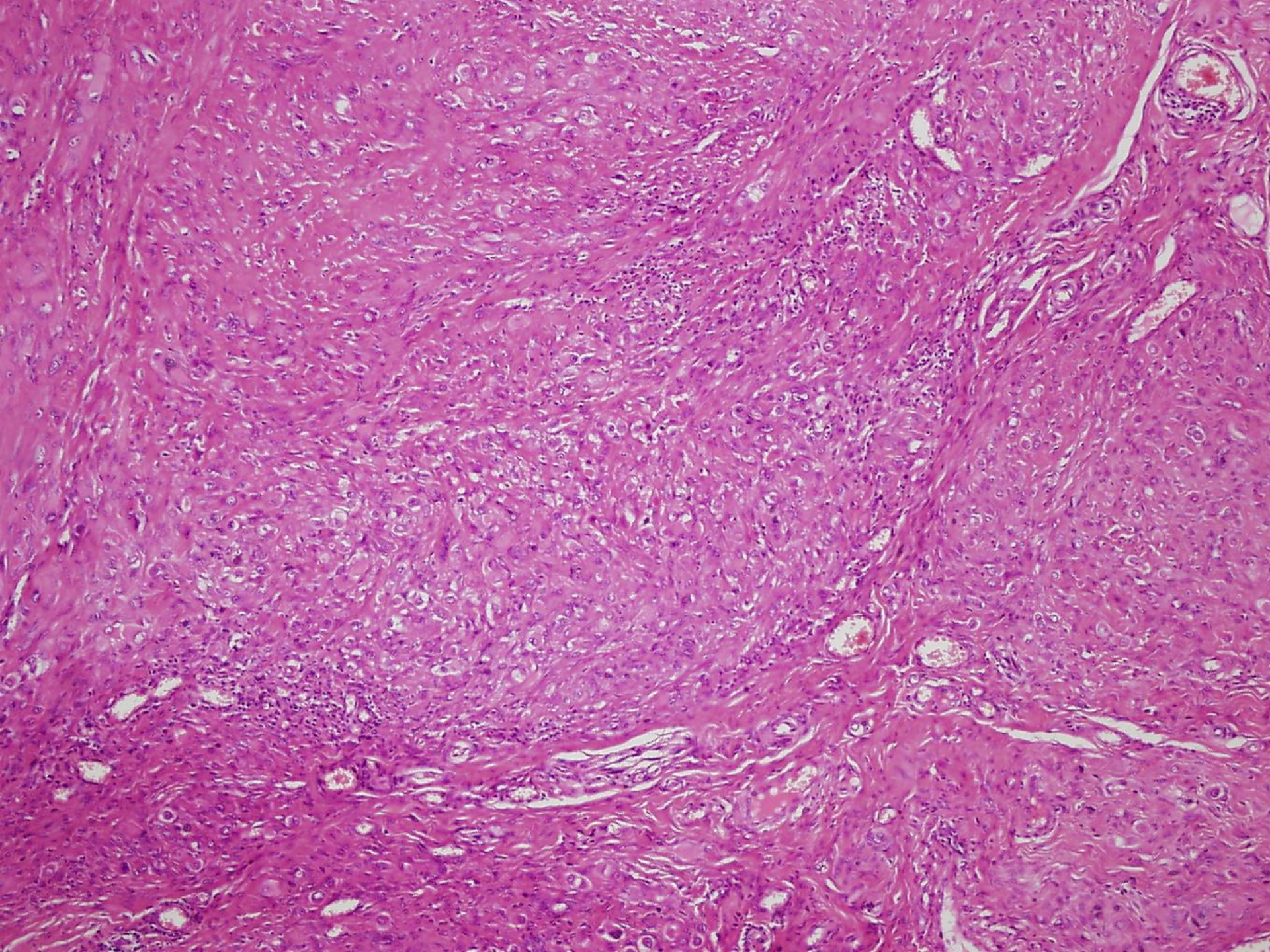


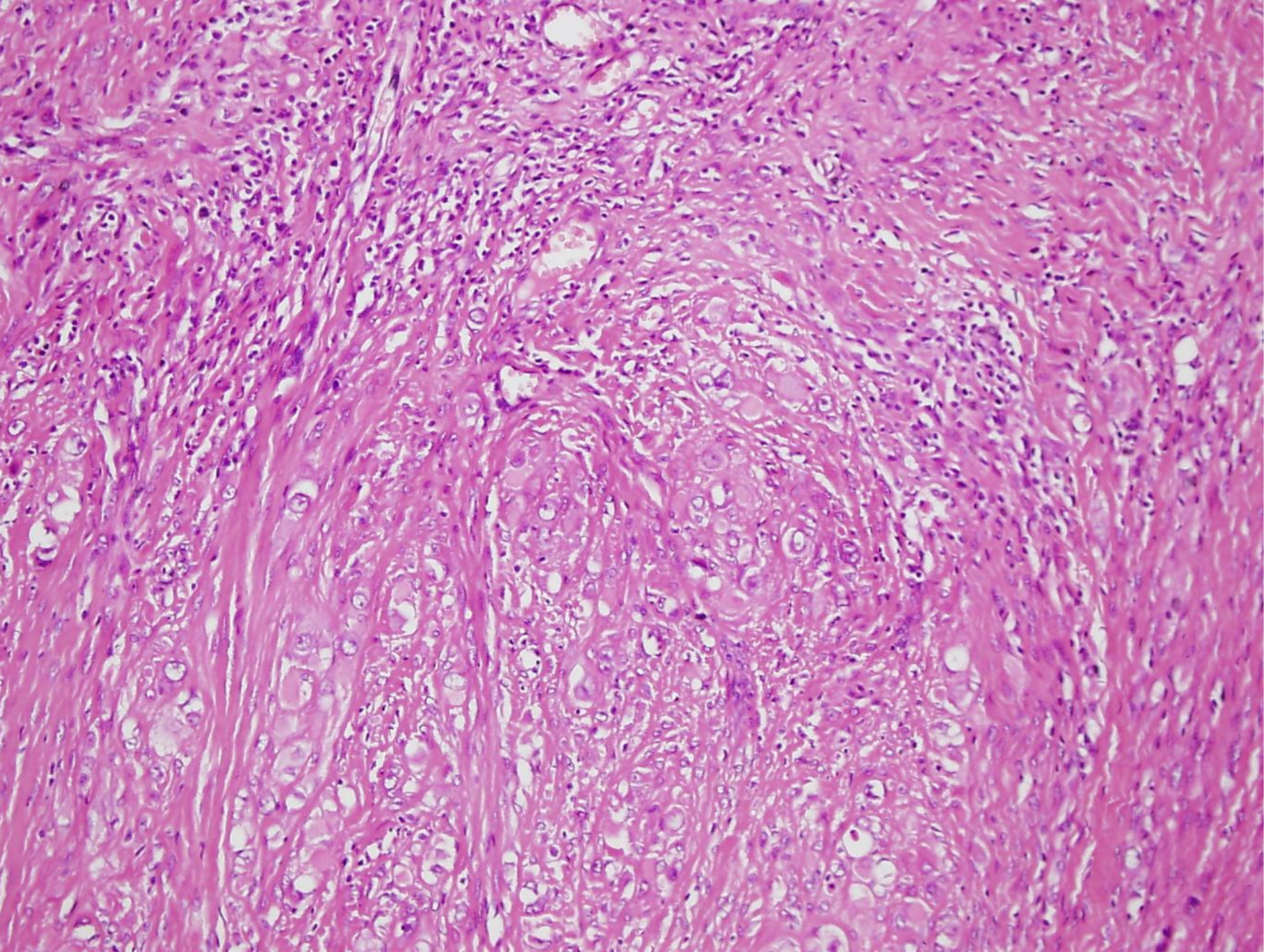
ZGÇ
23 K

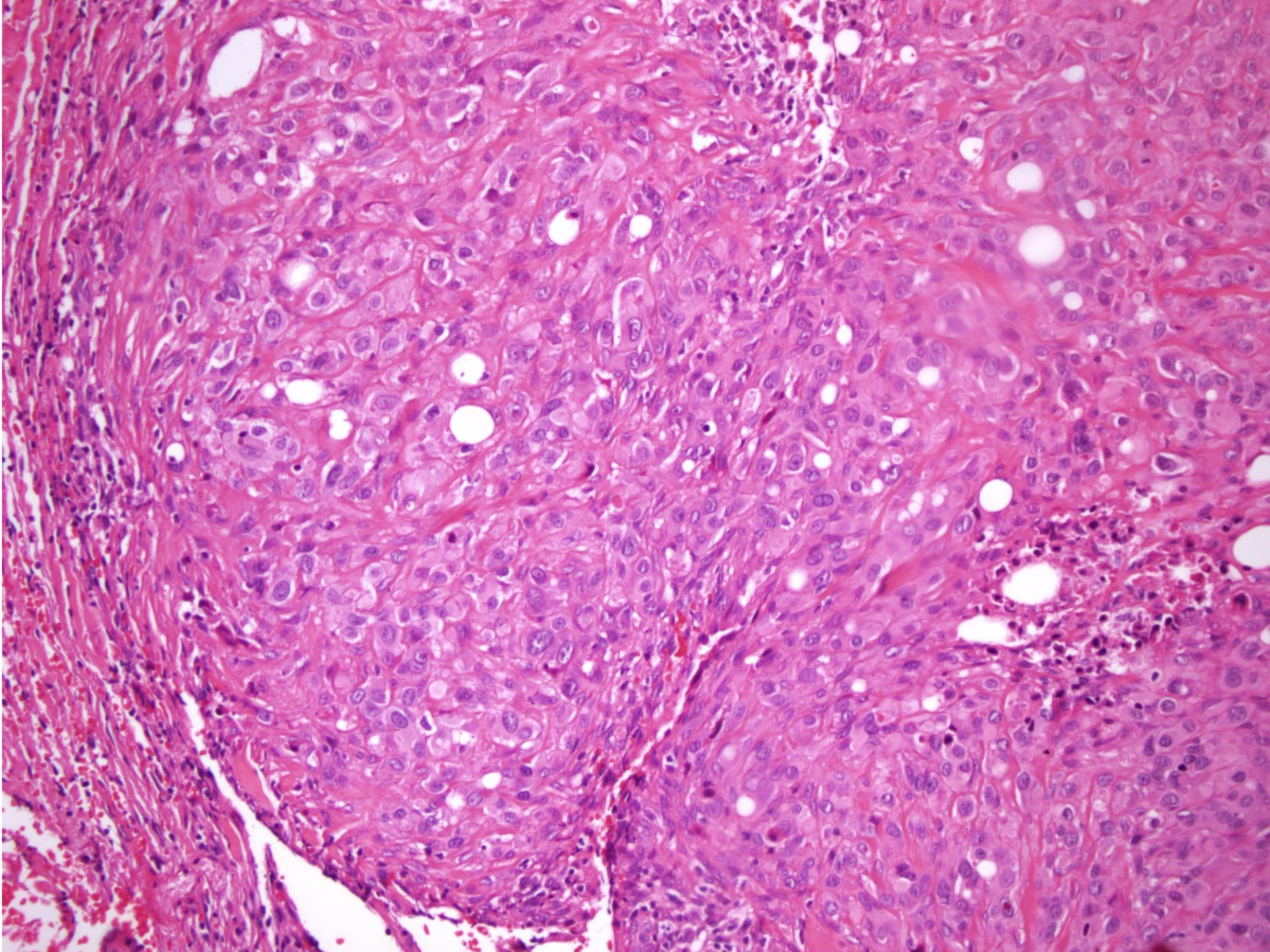


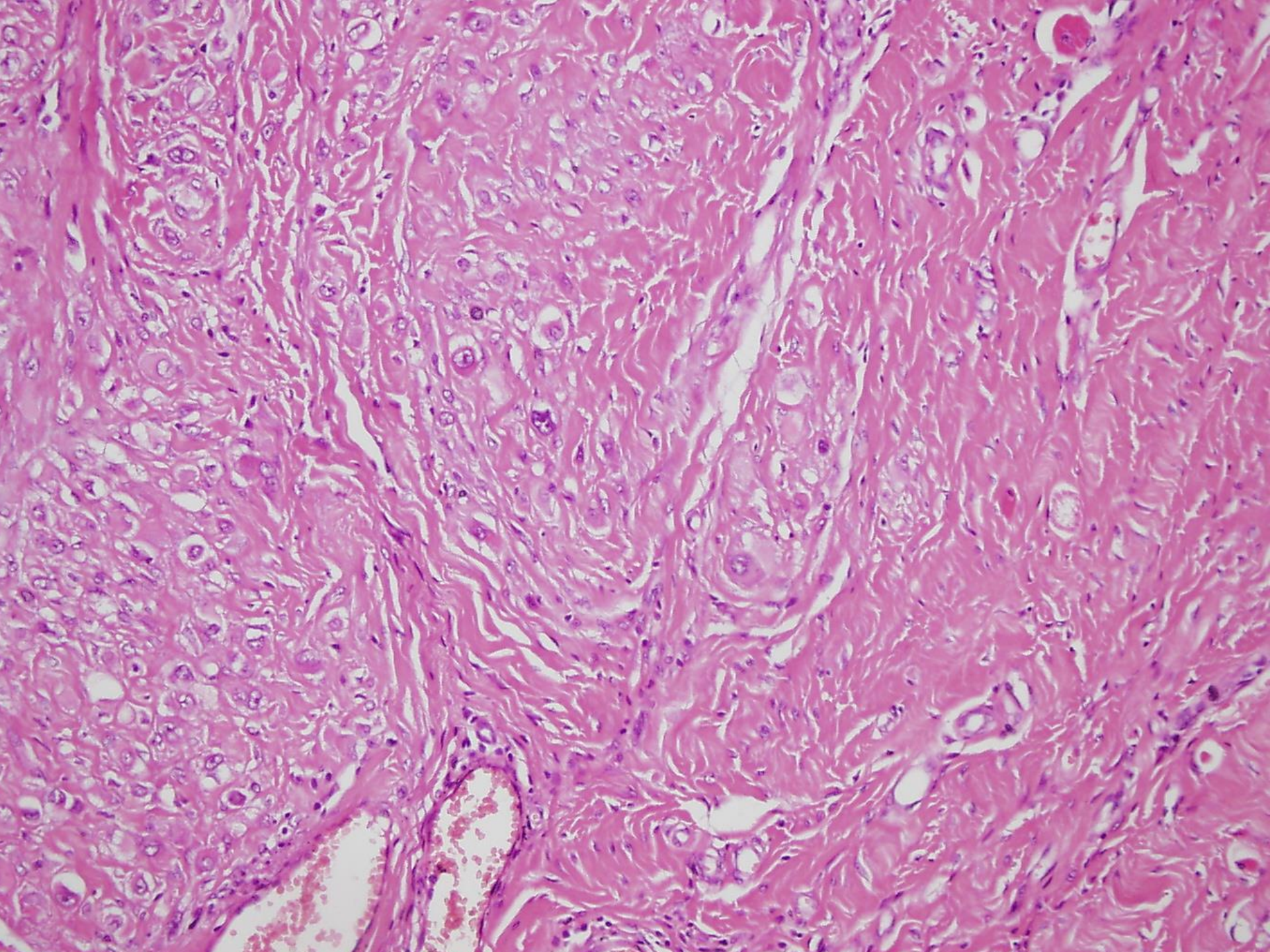
Epiteloid
sarkom

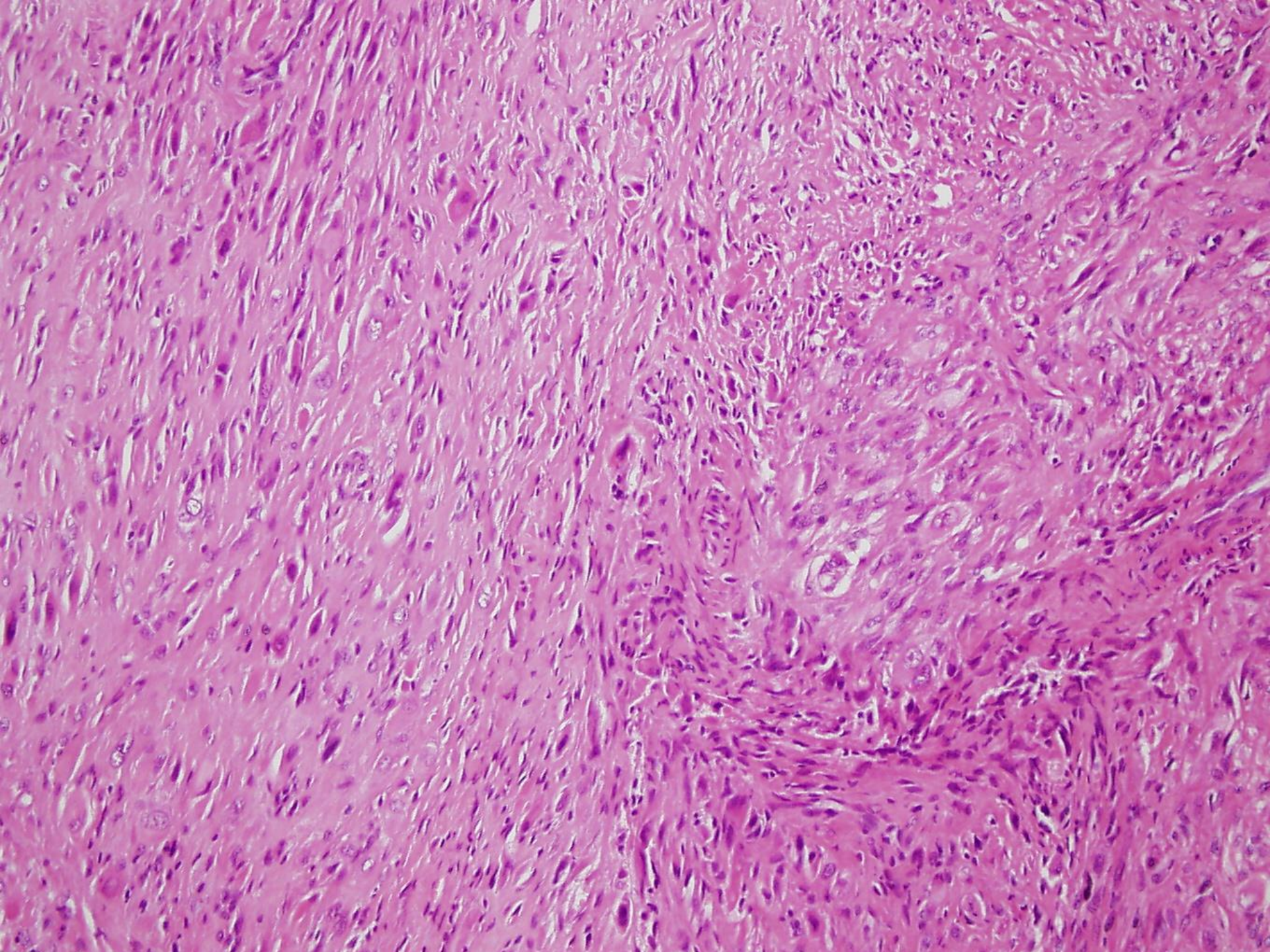


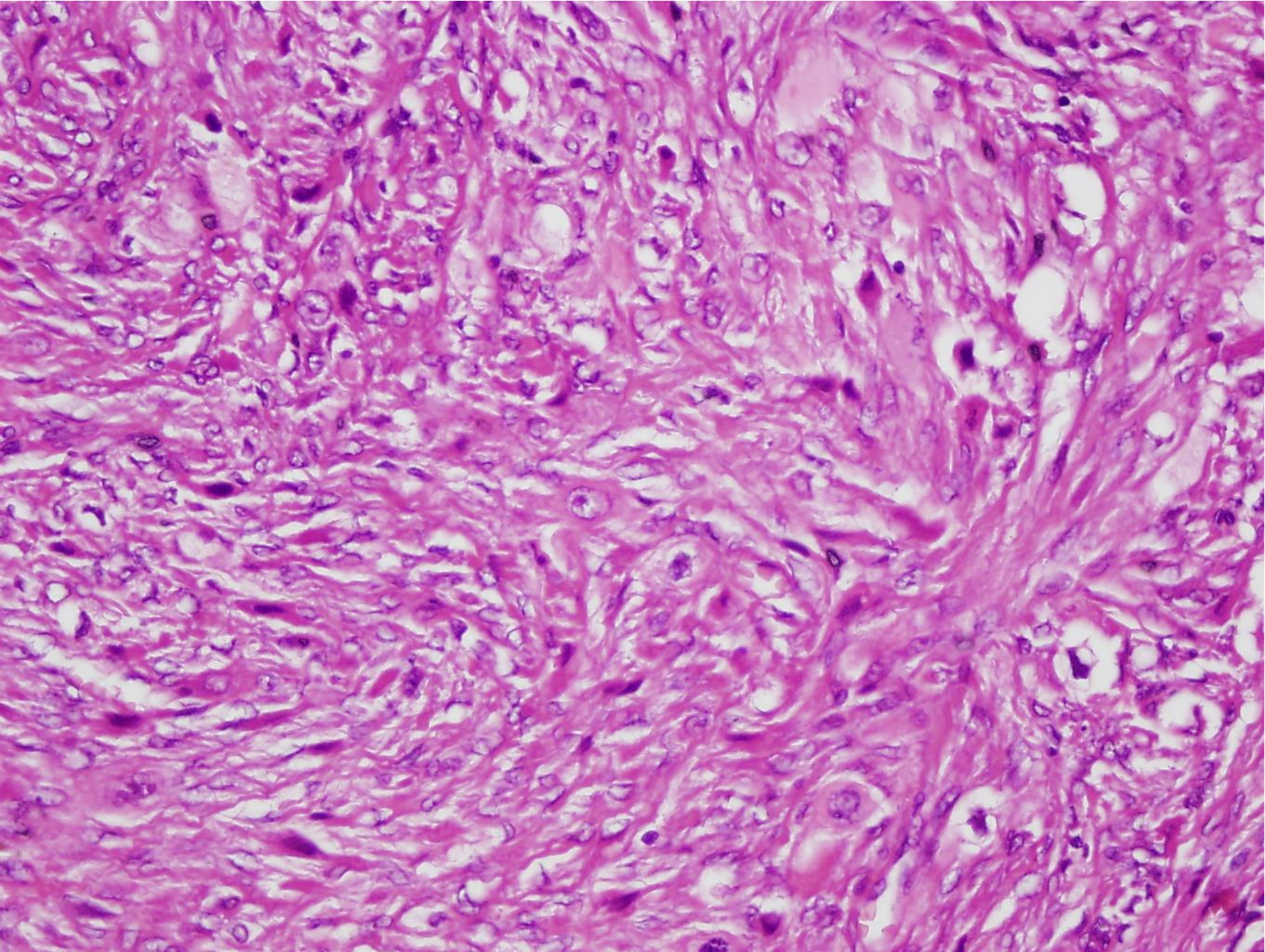


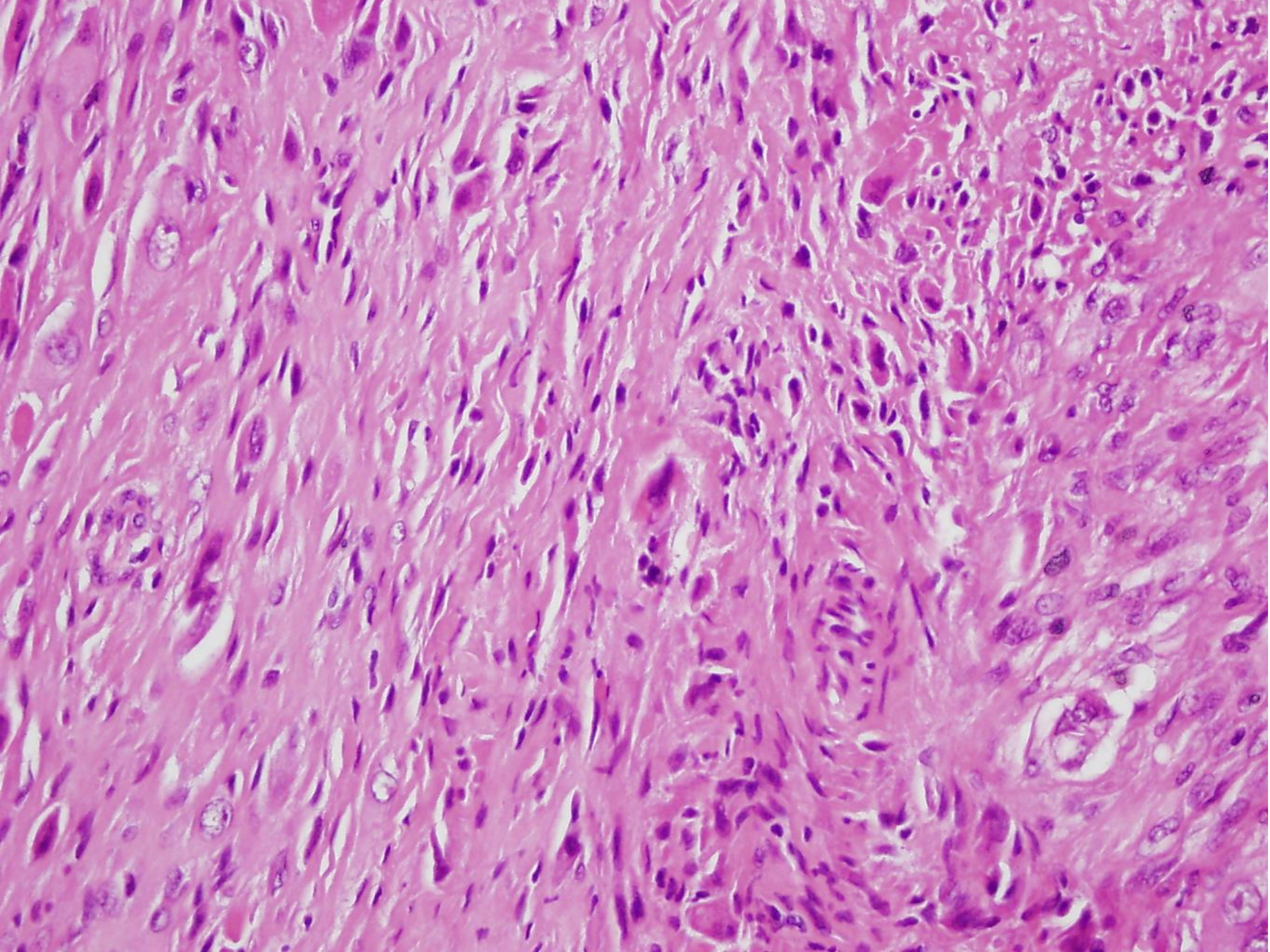




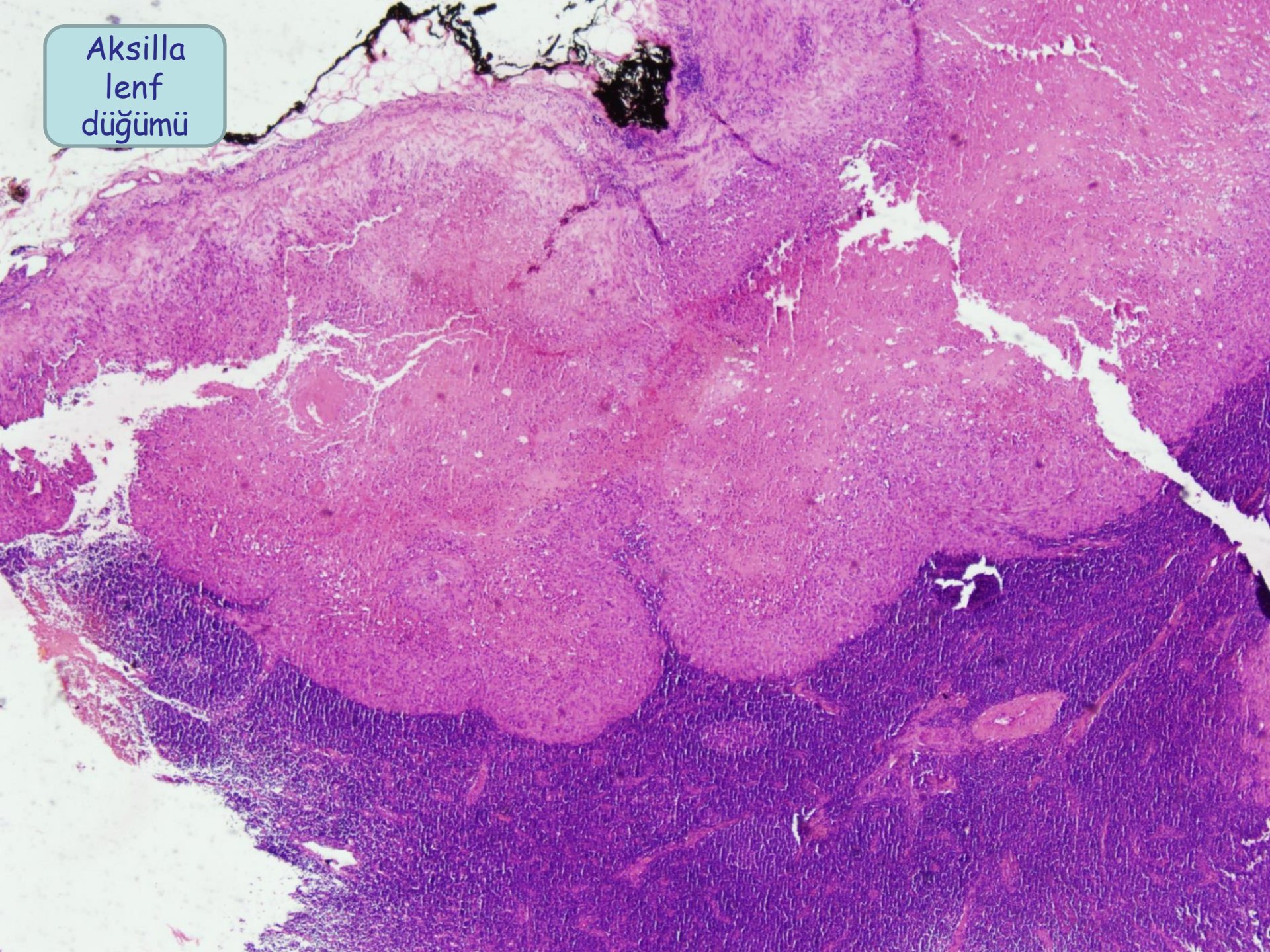


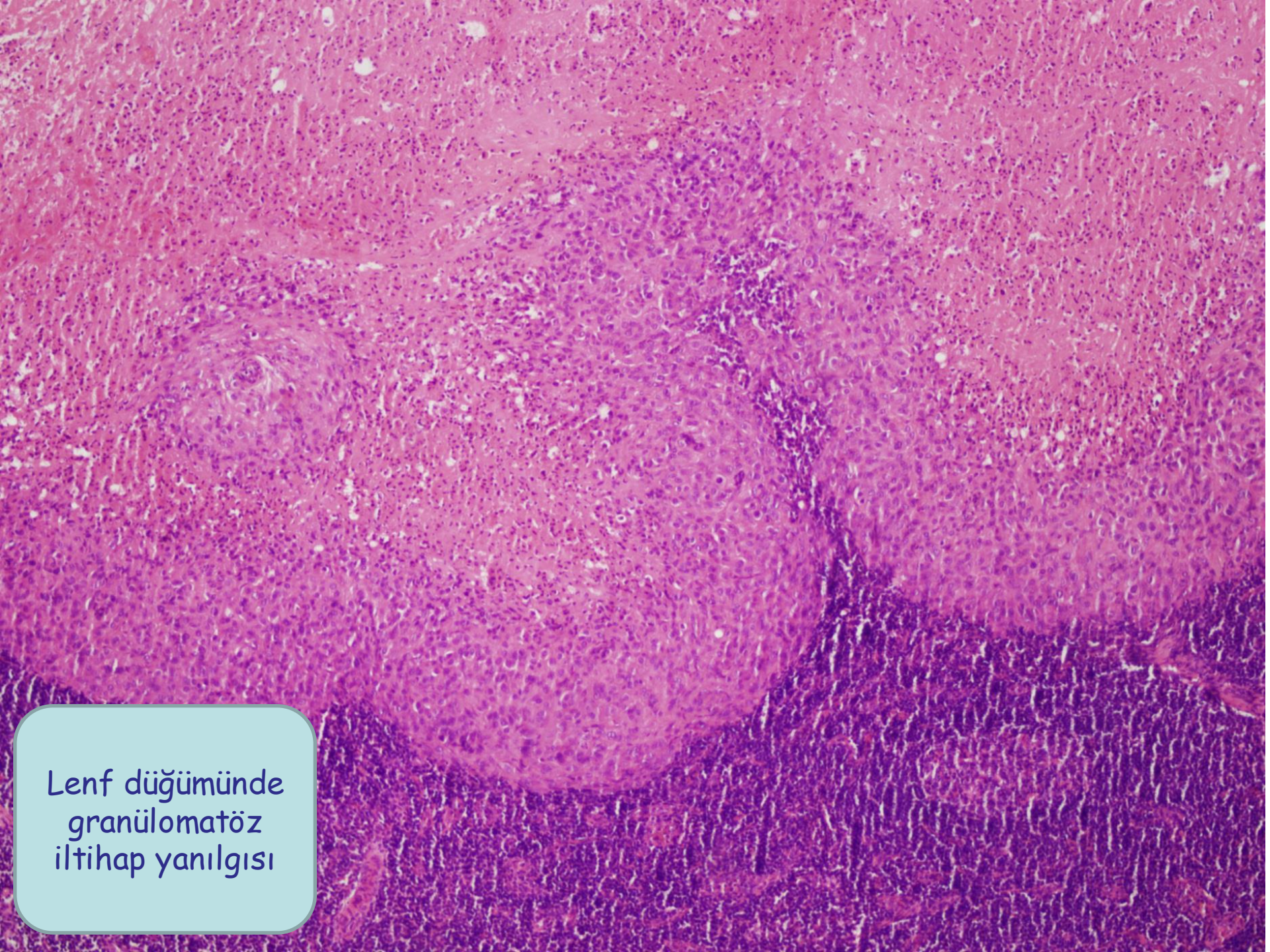






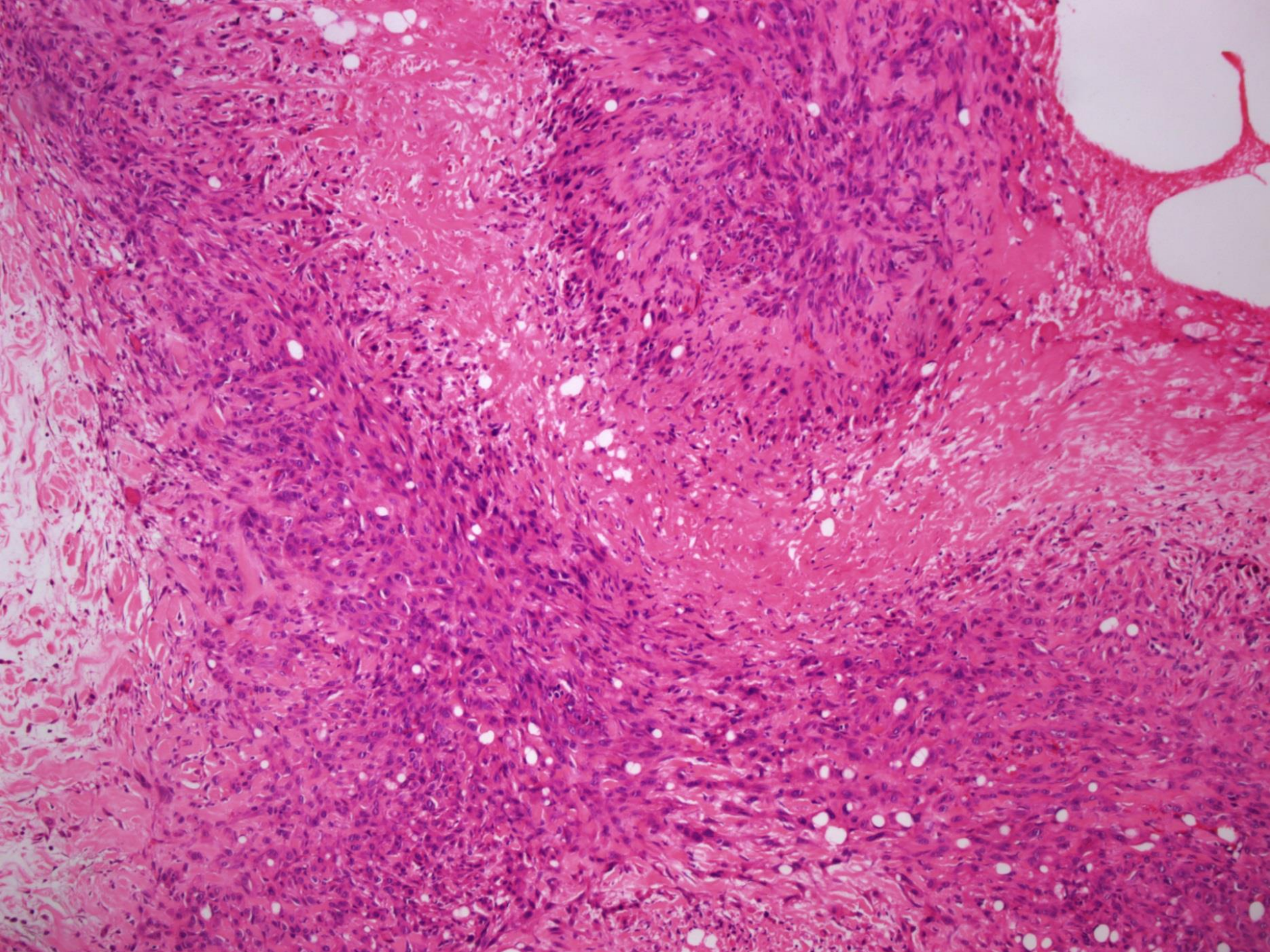
Aksilla
lenf
düğümü

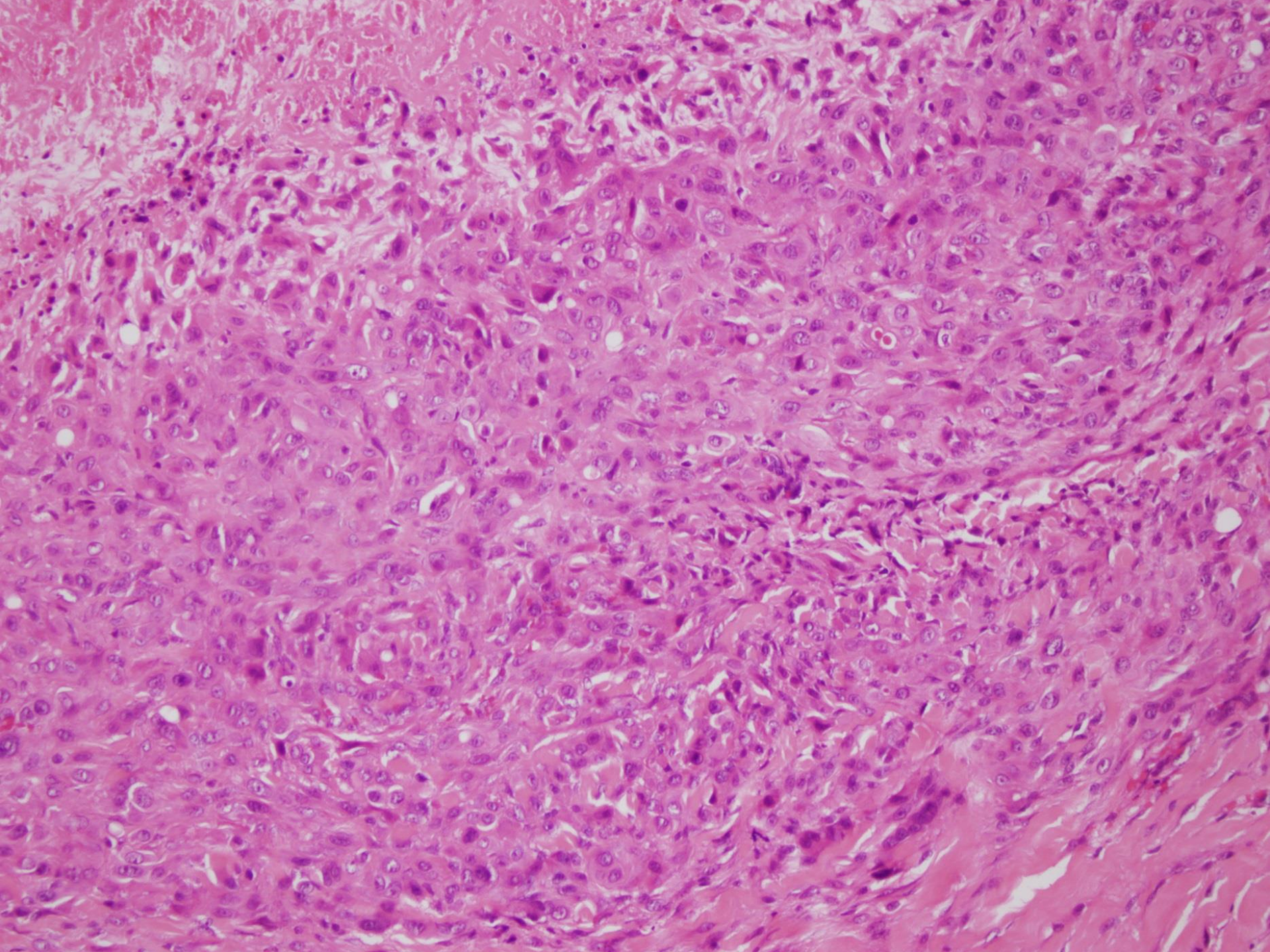


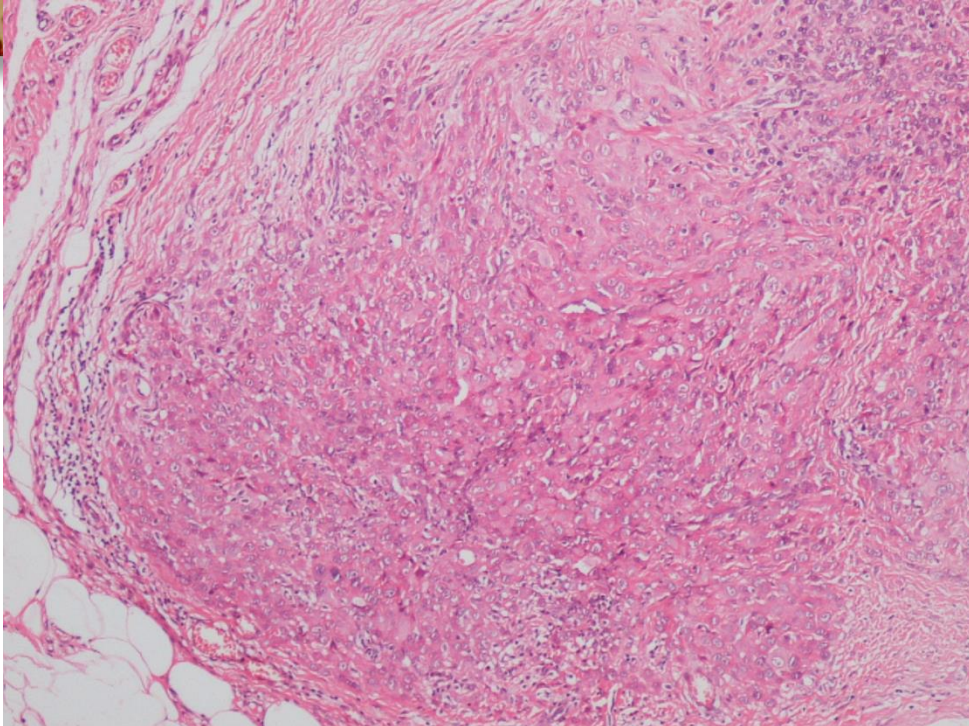
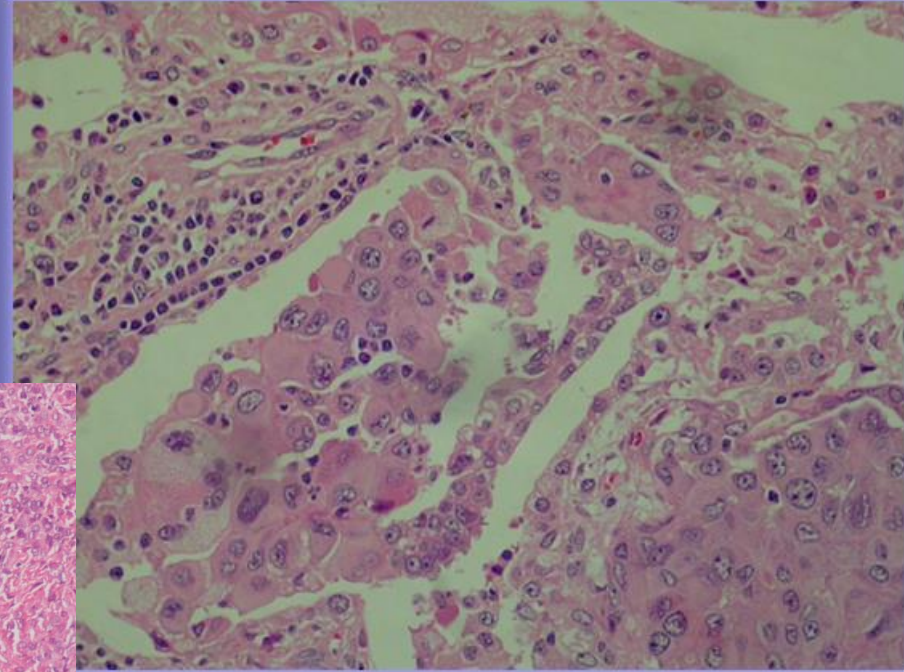
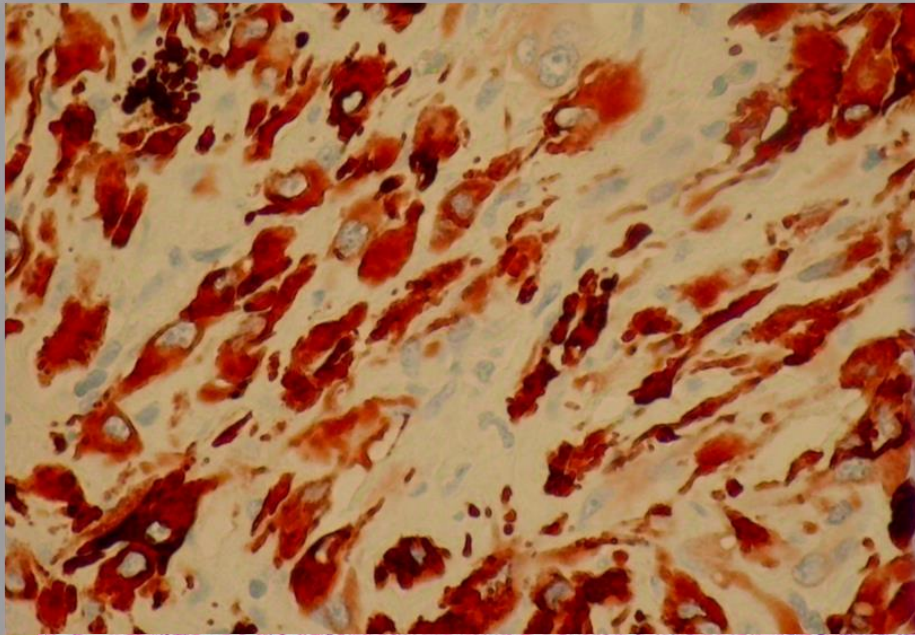


Lenf düğümünde
granülomatöz
iltihap yanılığı

This histological image shows a lymph node with granulomatous inflammation. The central feature is a large, well-defined granuloma, which is a collection of immune cells, including macrophages, T lymphocytes, and multinucleated giant cells, arranged in a circular pattern. The surrounding lymphoid tissue shows a dense population of lymphocytes. The overall appearance is characteristic of a chronic inflammatory response, such as that seen in tuberculosis or other granulomatous diseases.



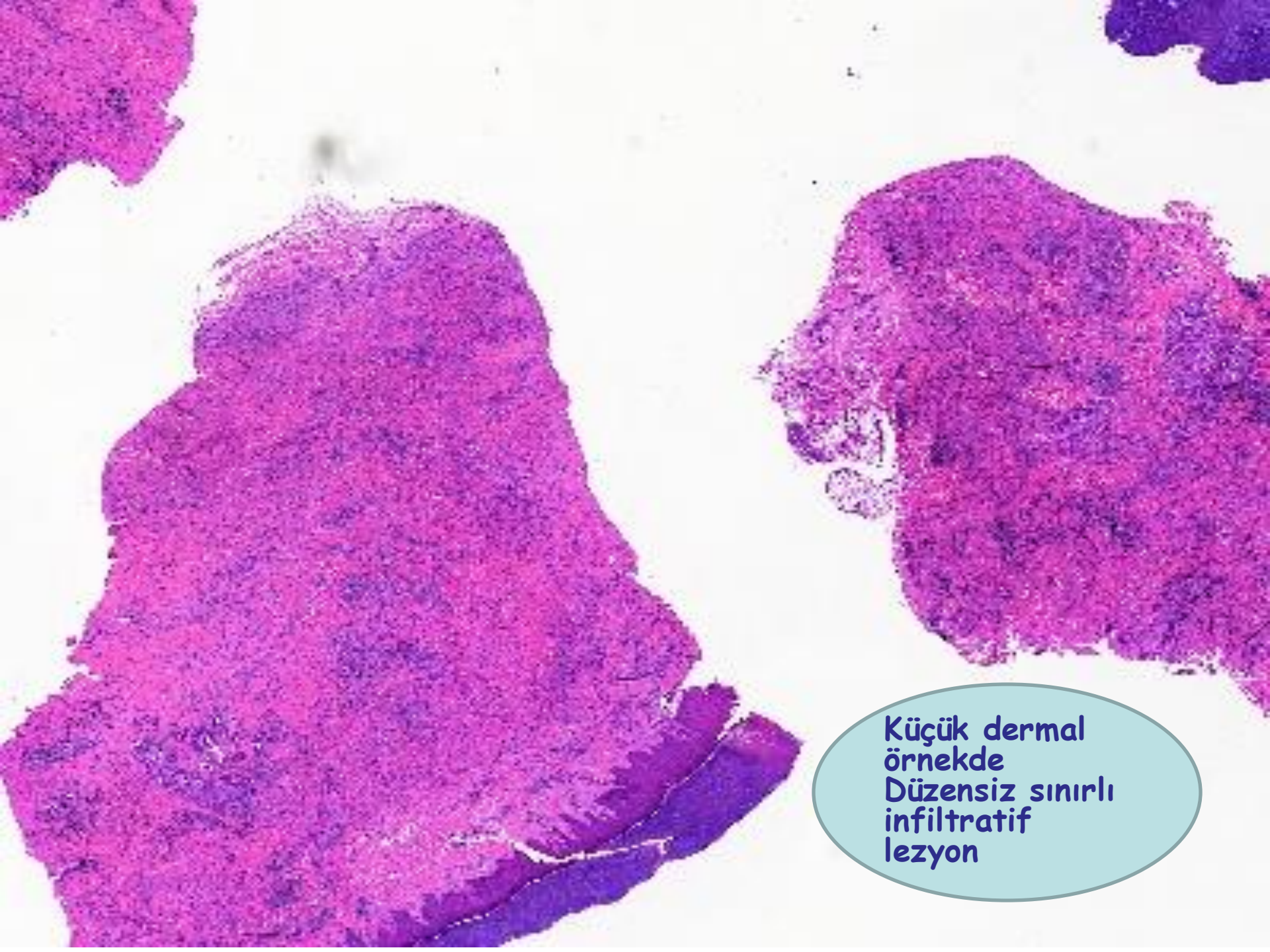




22 yaşında erkeek
Sağ elde endüre
lezyon
Tanı sonrası
eksizyon

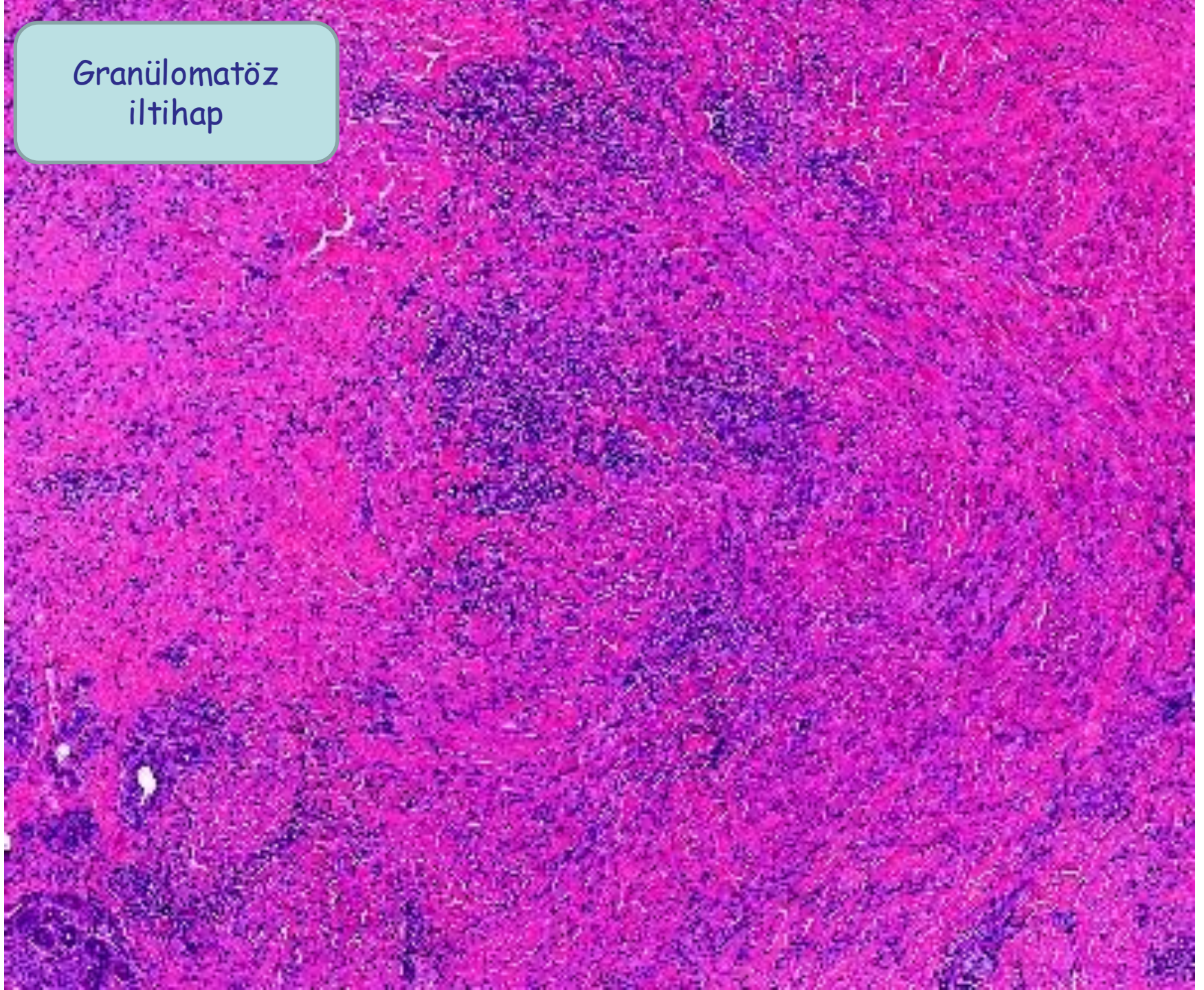


Dr. Mentzel Thomas,
Friedrichshafen,
Germany

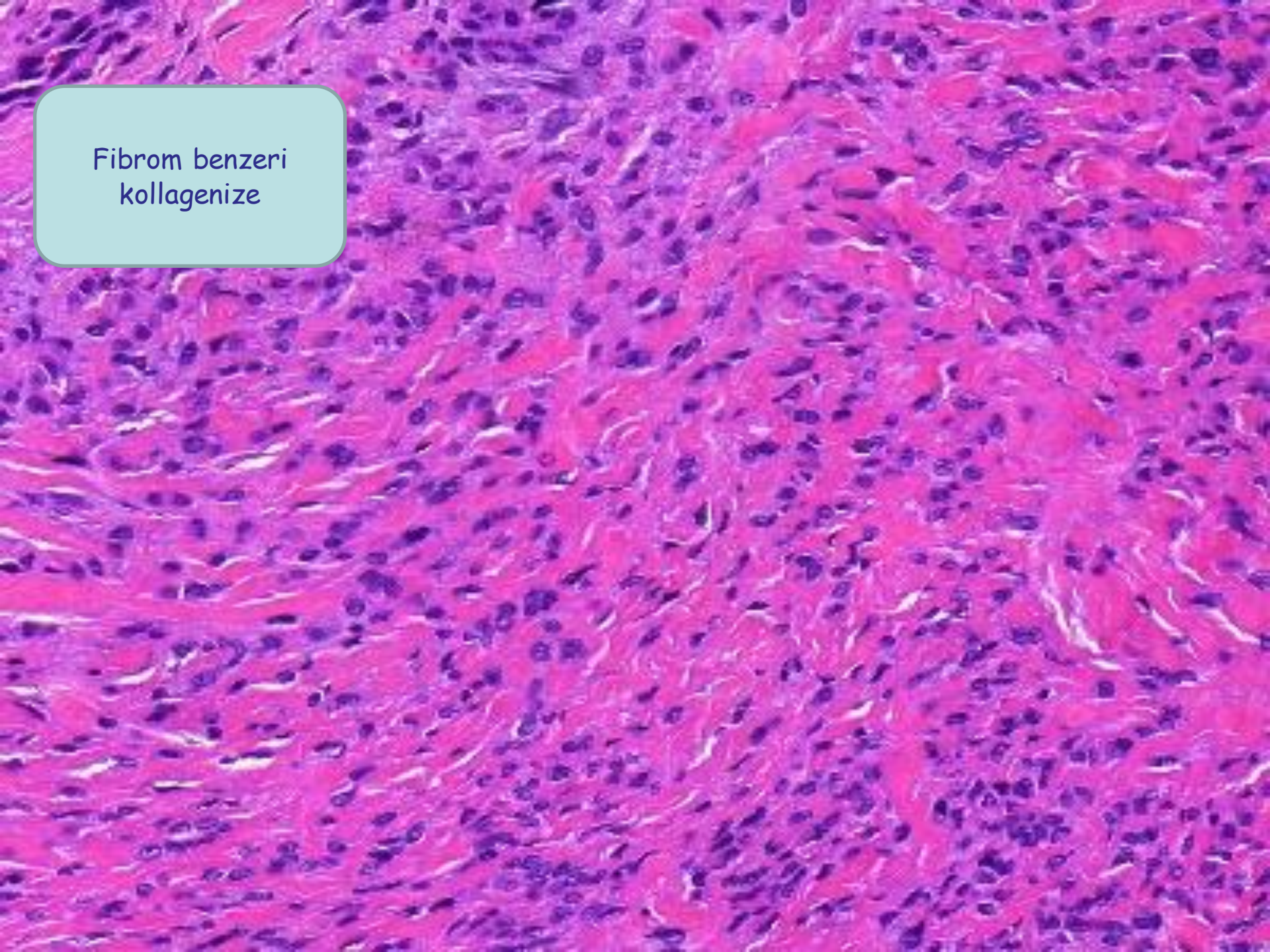


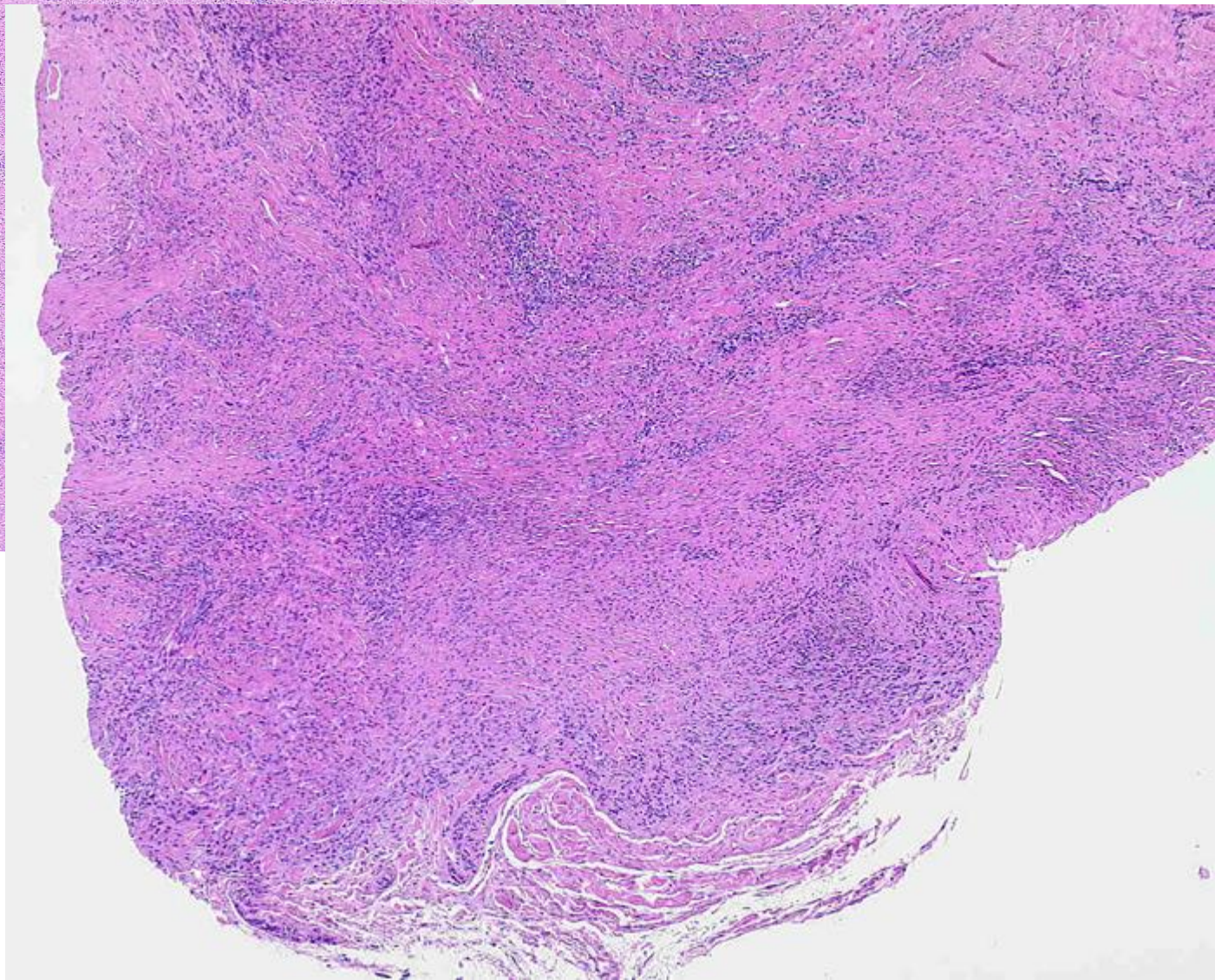
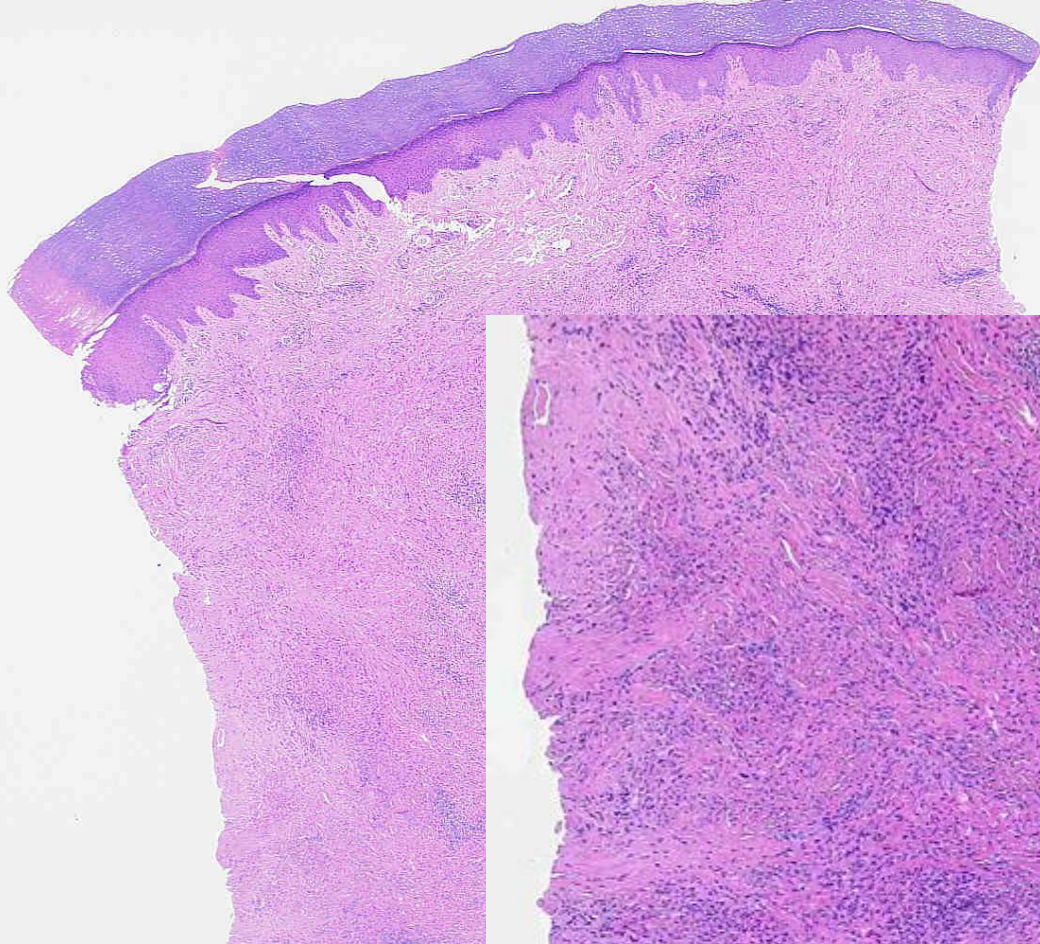
Küçük dermal
örnekde
Düzensiz sınırlı
infiltratif
lezyon

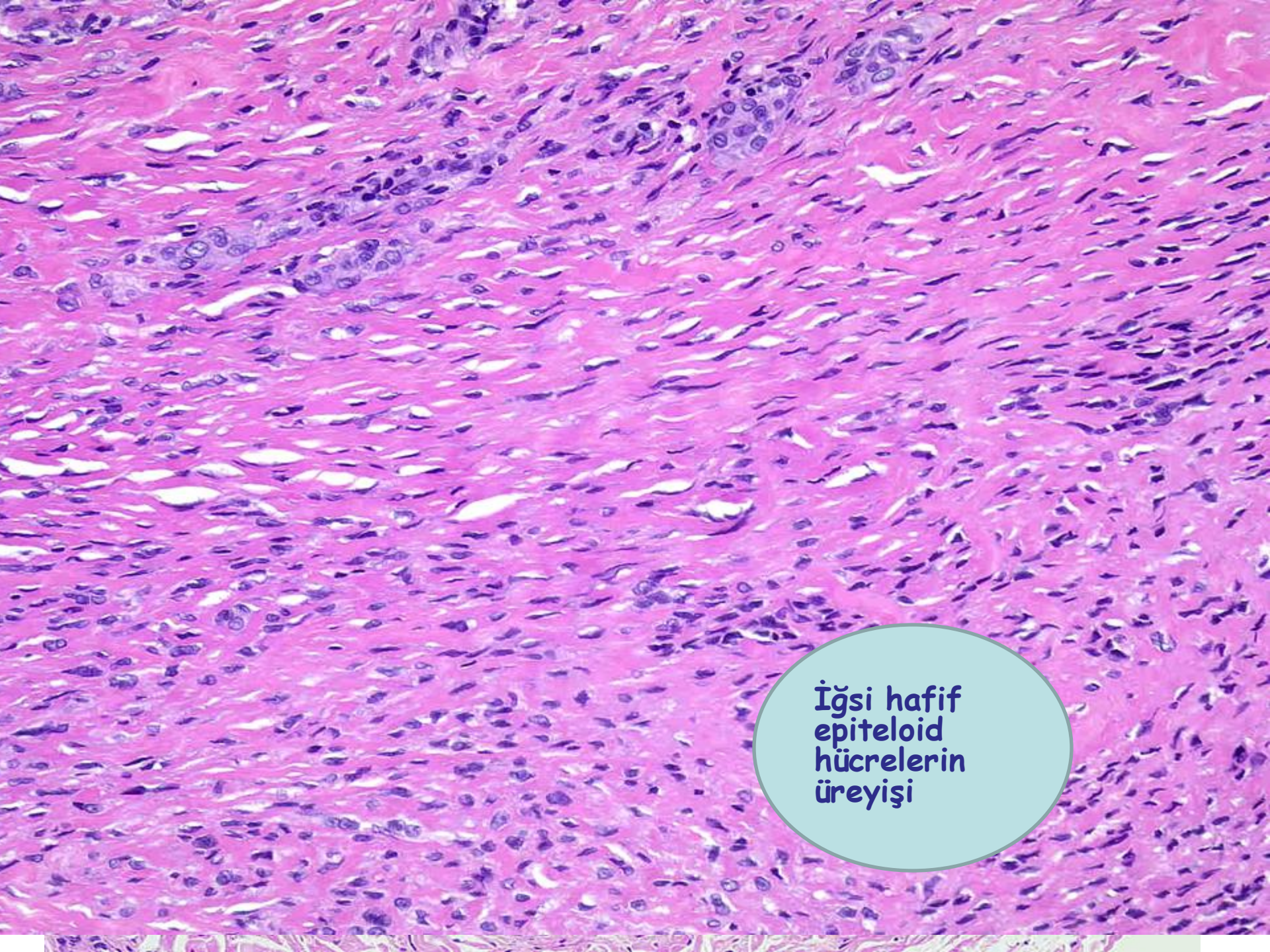
Granülomatöz
iltihap



Fibrom benzeri
kollagenize



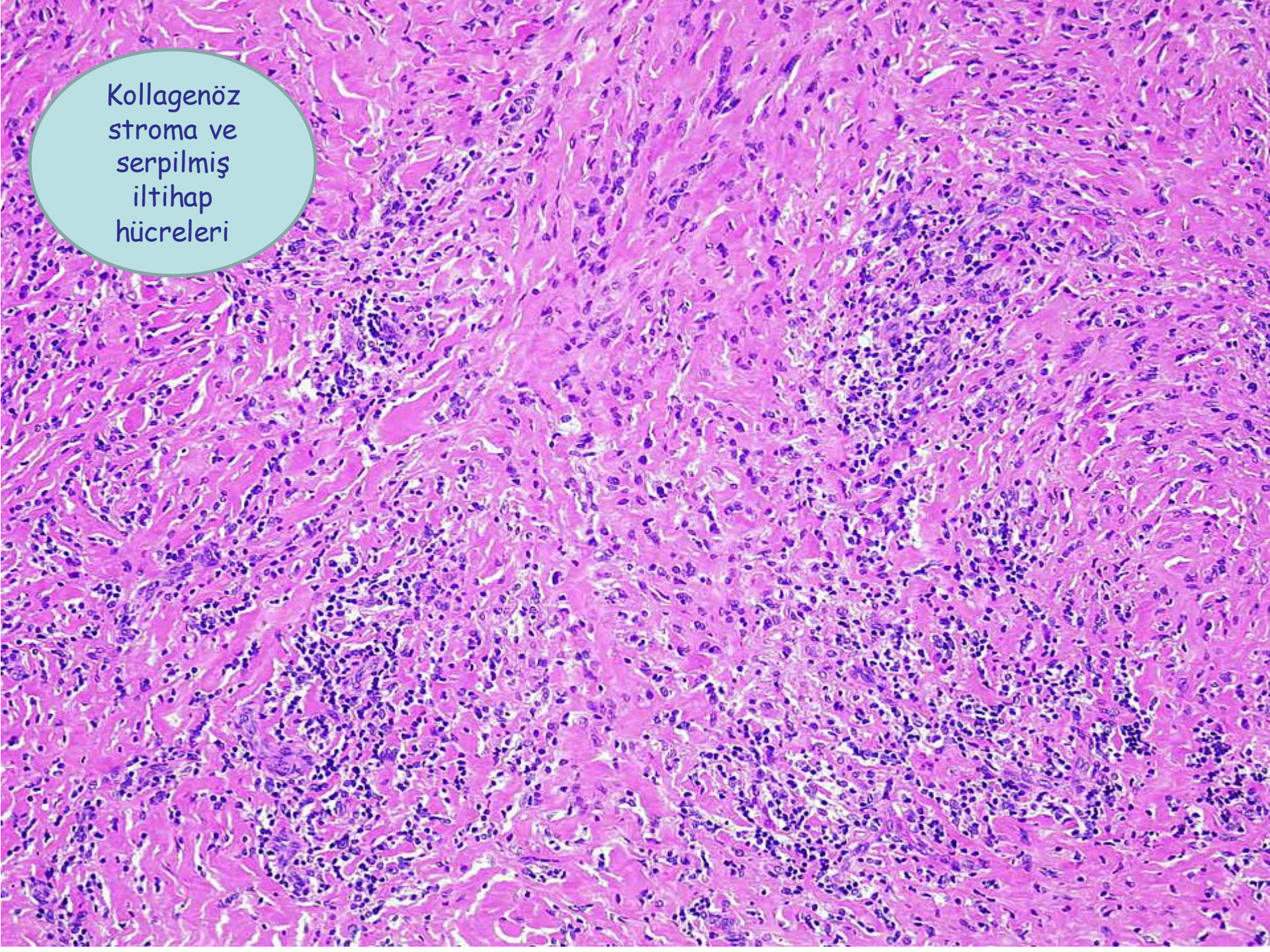


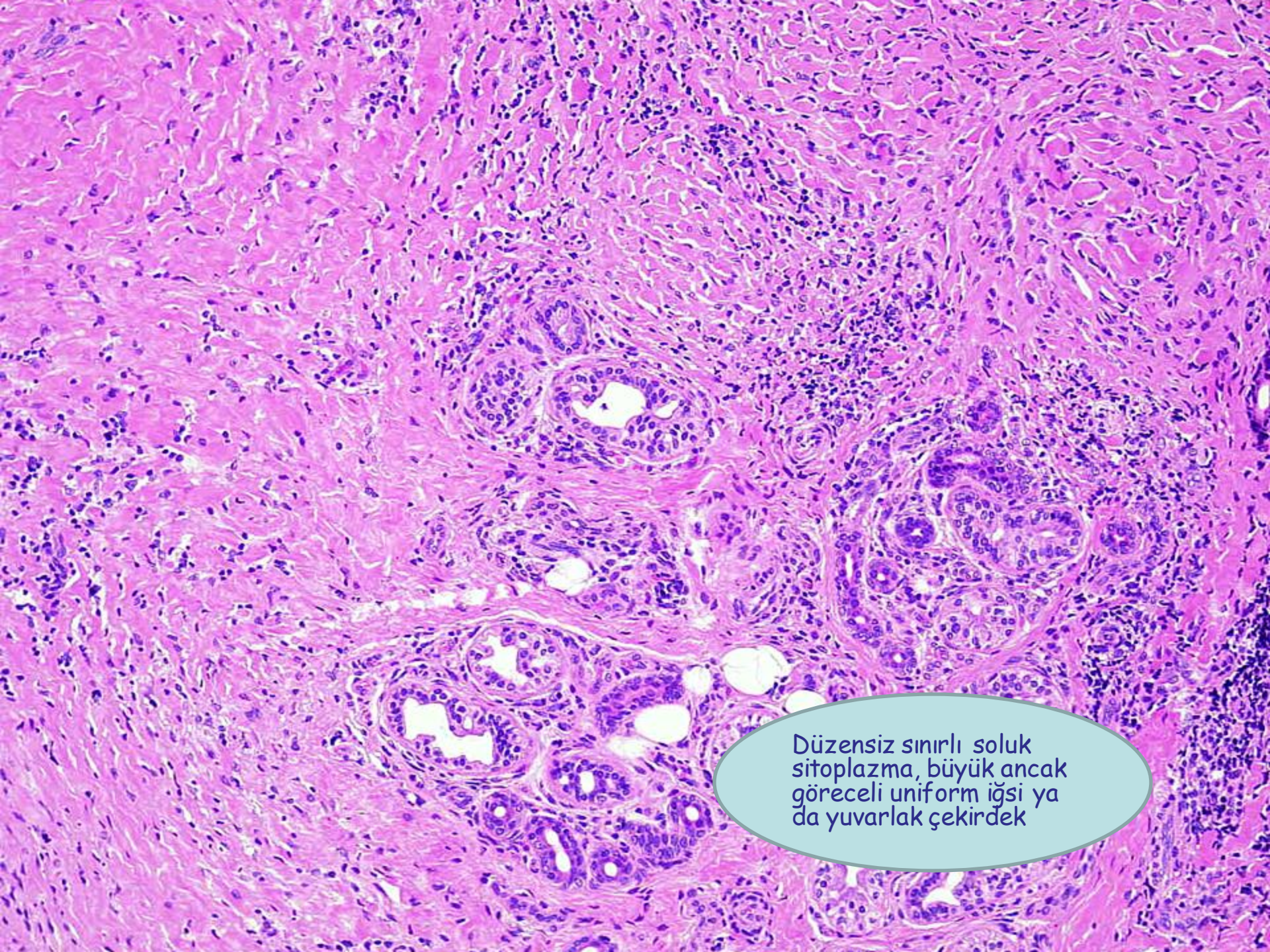


The image displays a histological section of the endometrium, stained with hematoxylin and eosin (H&E). The tissue is characterized by a dense population of cells with prominent, darkly stained nuclei and a pinkish cytoplasm and extracellular matrix. The overall appearance is that of a highly cellular, proliferative state. The cells are arranged in a somewhat disorganized pattern, with many nuclei appearing to be in various stages of the cell cycle. The background is a rich pink color, indicating the presence of eosinophilic cytoplasm and extracellular matrix components.

**İçsi hafif
epiteloid
hücrelerin
üreyişi**

Kollagenöz
stroma ve
serpilmiş
iltihap
hücreleri

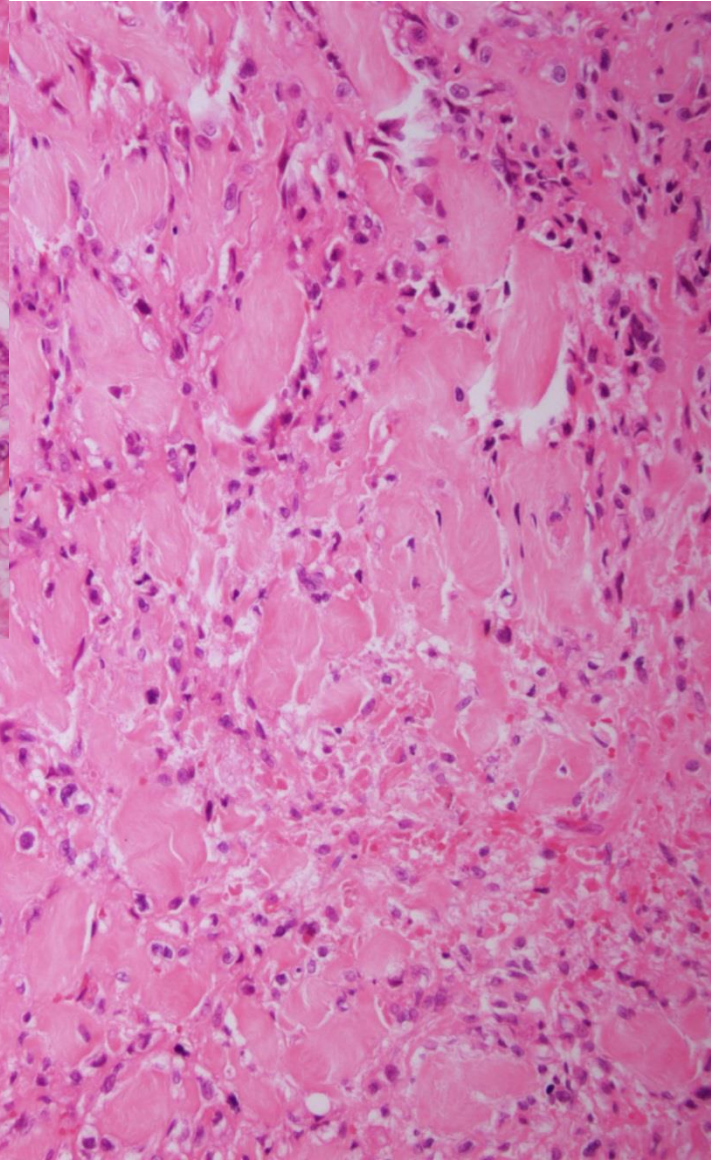
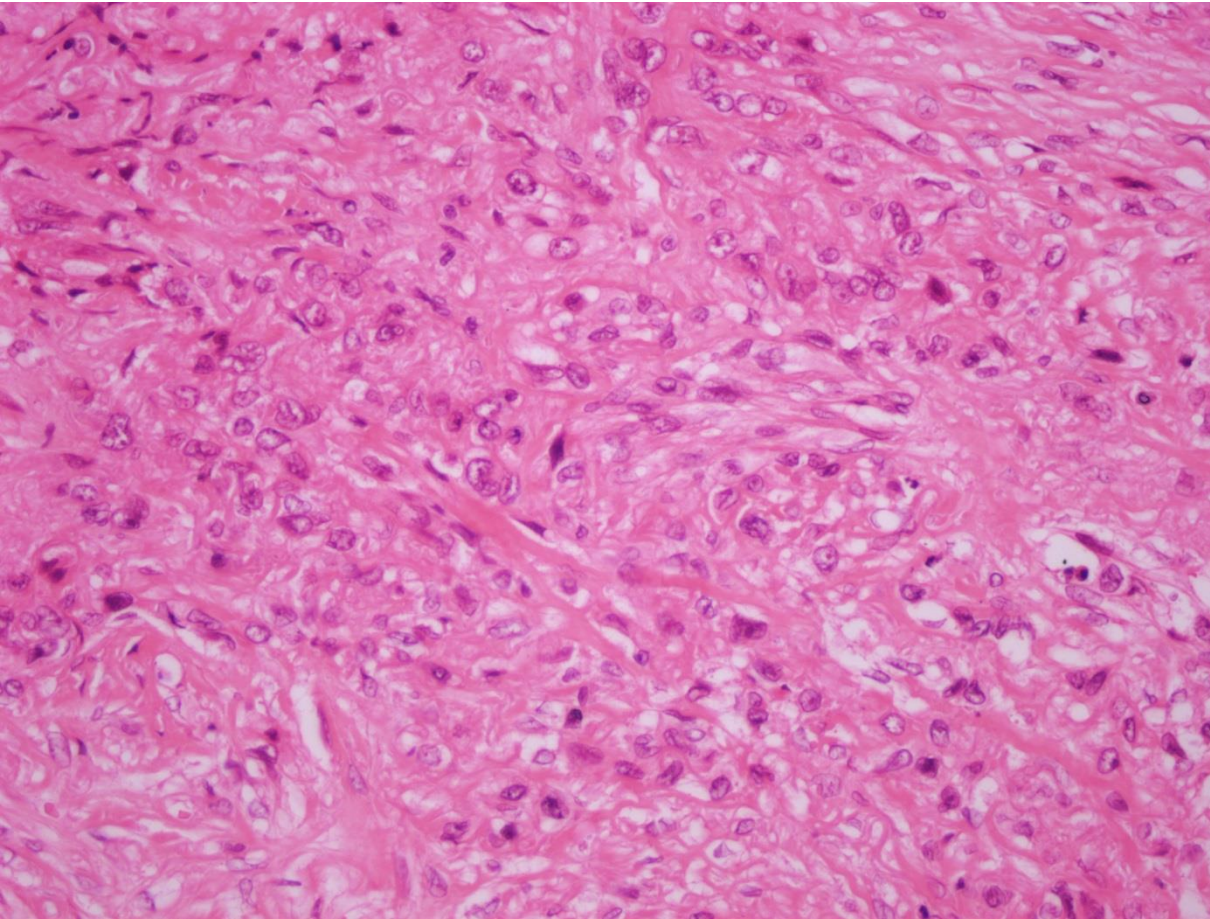




Düzensiz sınırlı soluk sitoplazma, büyük ancak göreceli uniform içi ya da da yuvarlak çekirdek

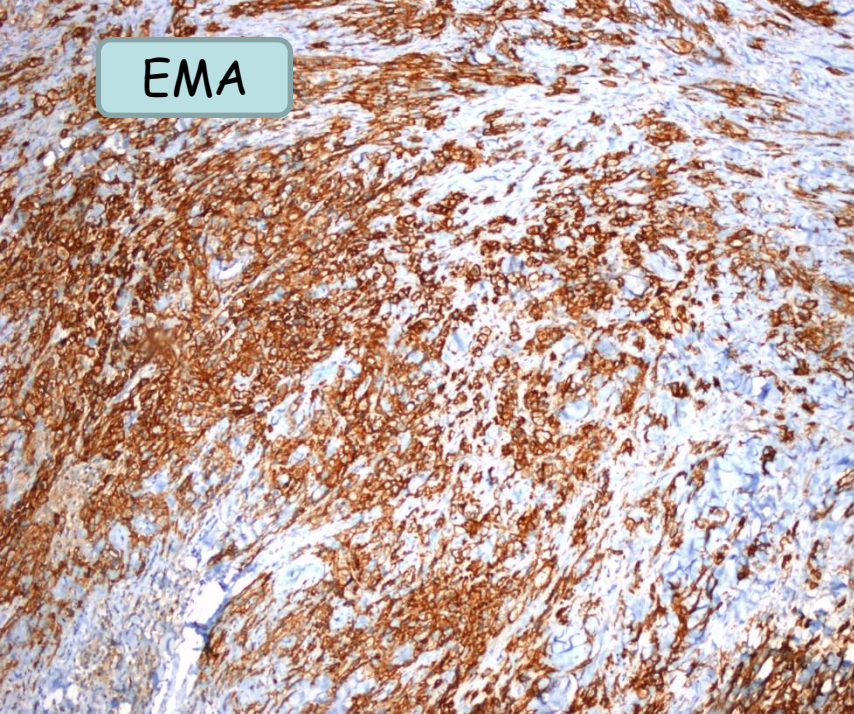
- Proliferatif aktivite çarpıcı değil
- Tümör nekrozu yok
- İmmunprofil
- Vimentin,
- Pansitokeratin
- EMA
- Fokal S-100 pozitifliği
- INI1 dışavurumu kaybı
- CD34 ve myojenik belirleyiciler negatif

Epiteloid sarkom
fibrom benzeri tip

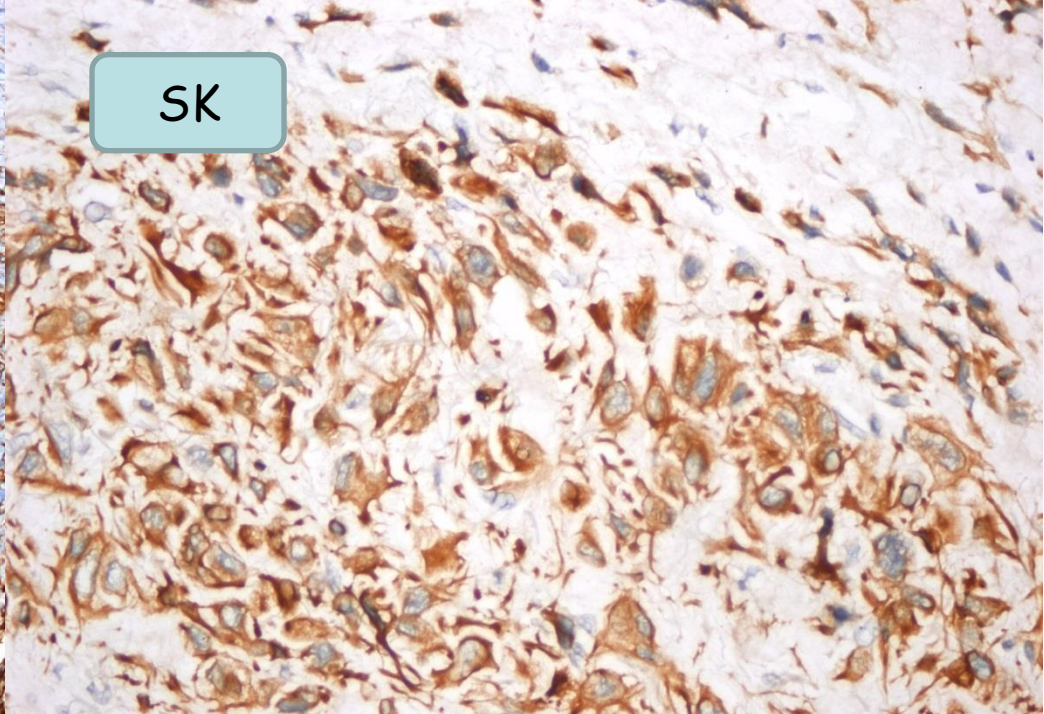


75K
Kulak tragus

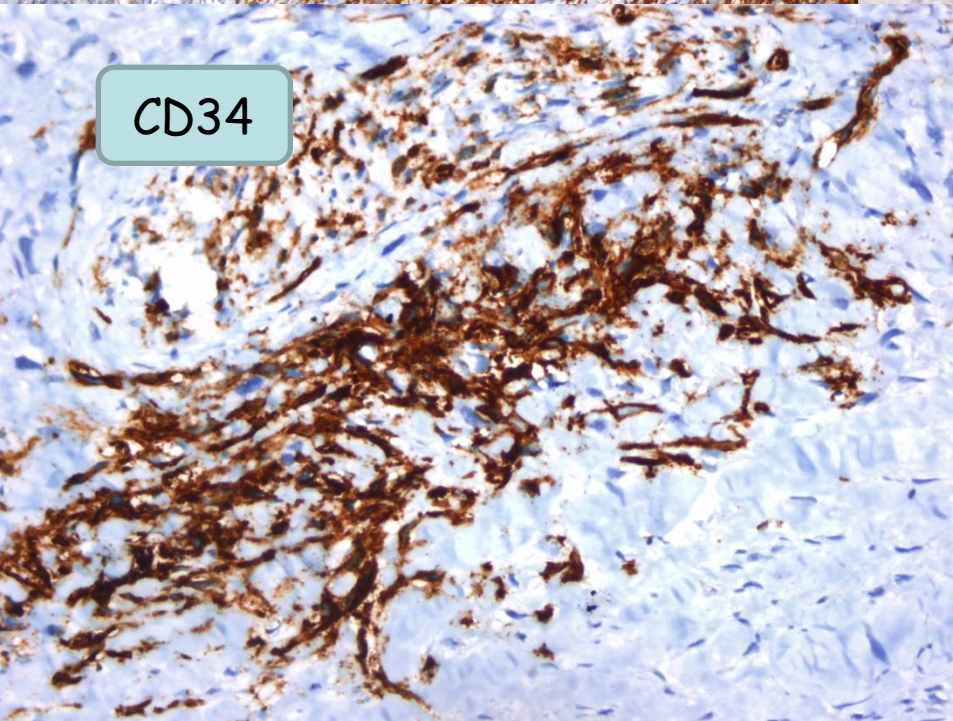
EMA



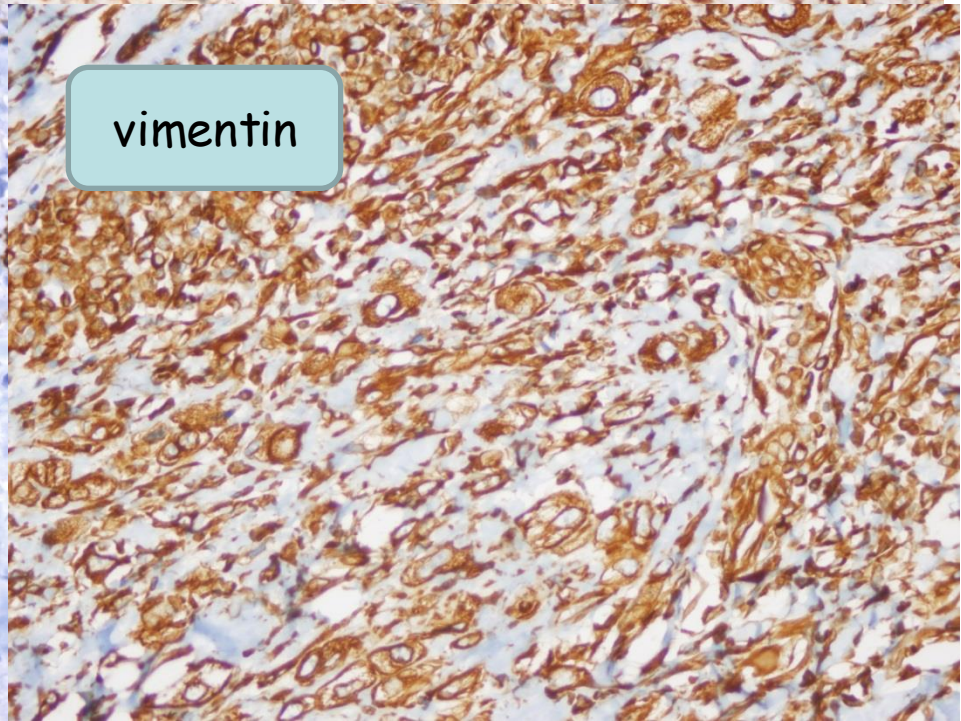
SK



CD34

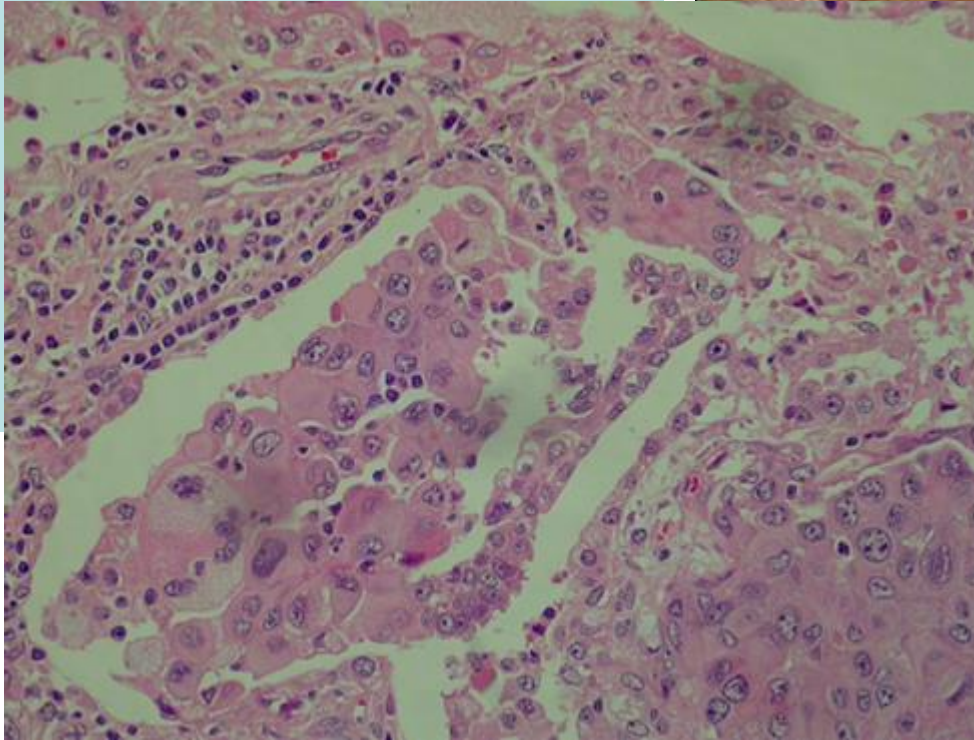


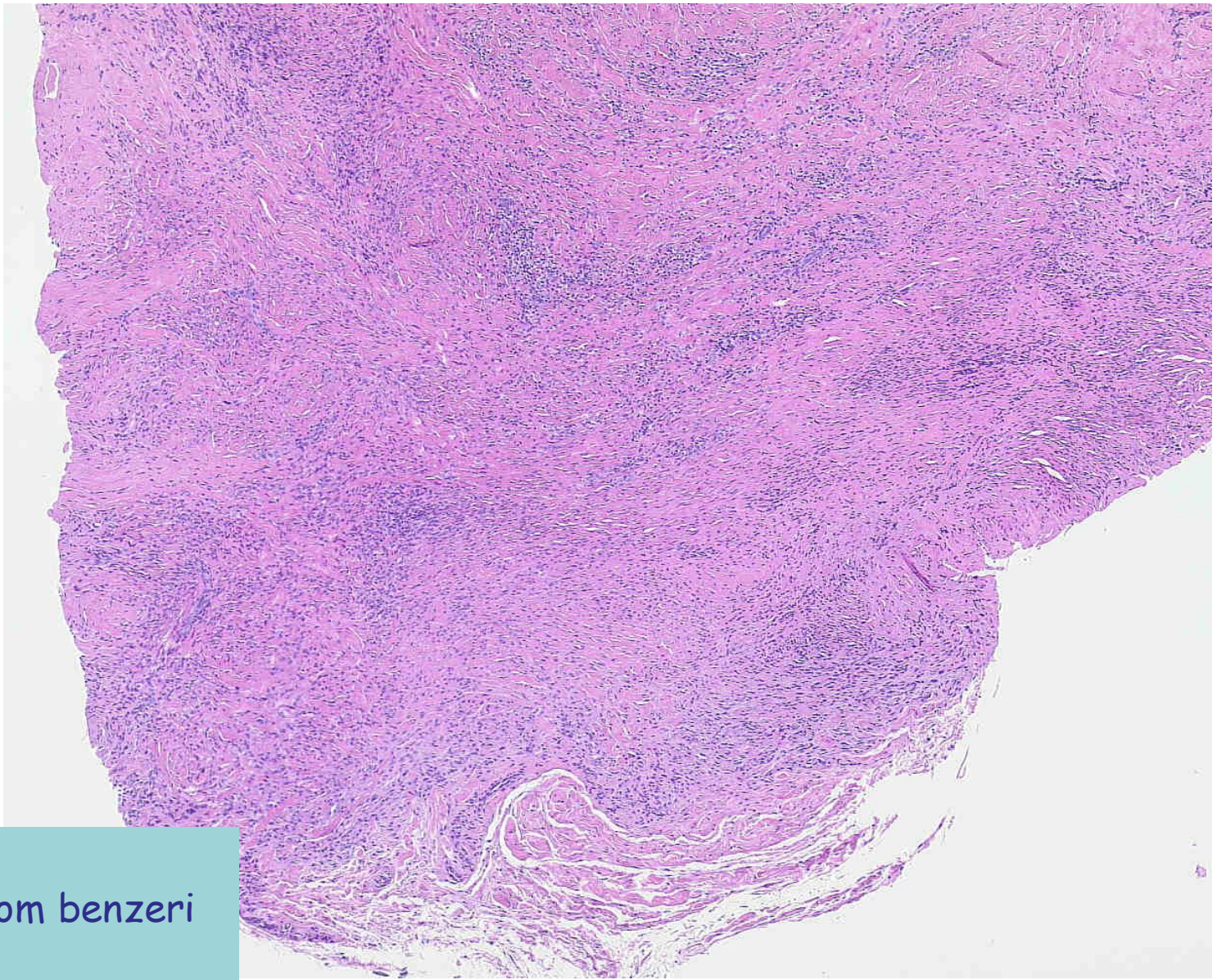
vimentin



- 1970 Enzinger
- Nadir *çok yönlü diferansiasyon
- Genç erişkin distal ekstremitelerde
- Yavaş büyüyen dermal, subkutan derin yerleşimli nodül/plak
- Tendon ve fasya tm fasyal planlar ve nörovasküler yapılar bounca büyür
- Klinik olarak agresif gidişli

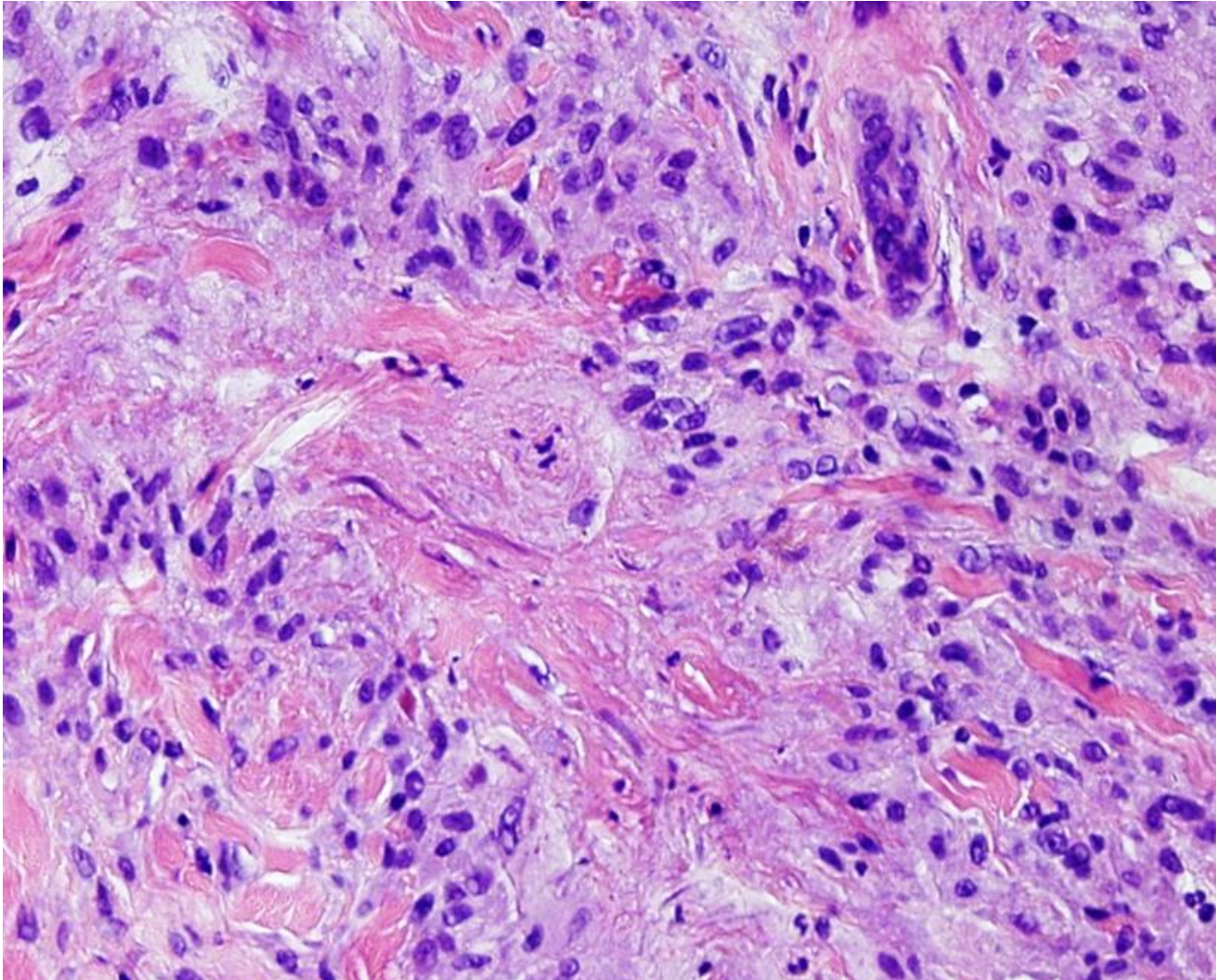
- Lokal nüksler
- % 40- 60 metastaz
- AC, bölgesel lenf düğümleri skalp nadiren kemik ve beyin
- Tümör boyutu ve mitoz metastazsız sağ kalımda prognostik anlamlı
- Proksimal yerleşimi tümör boyutu, proliferatif aktivite ve multifokalite tüm sağkalımda anlamlı

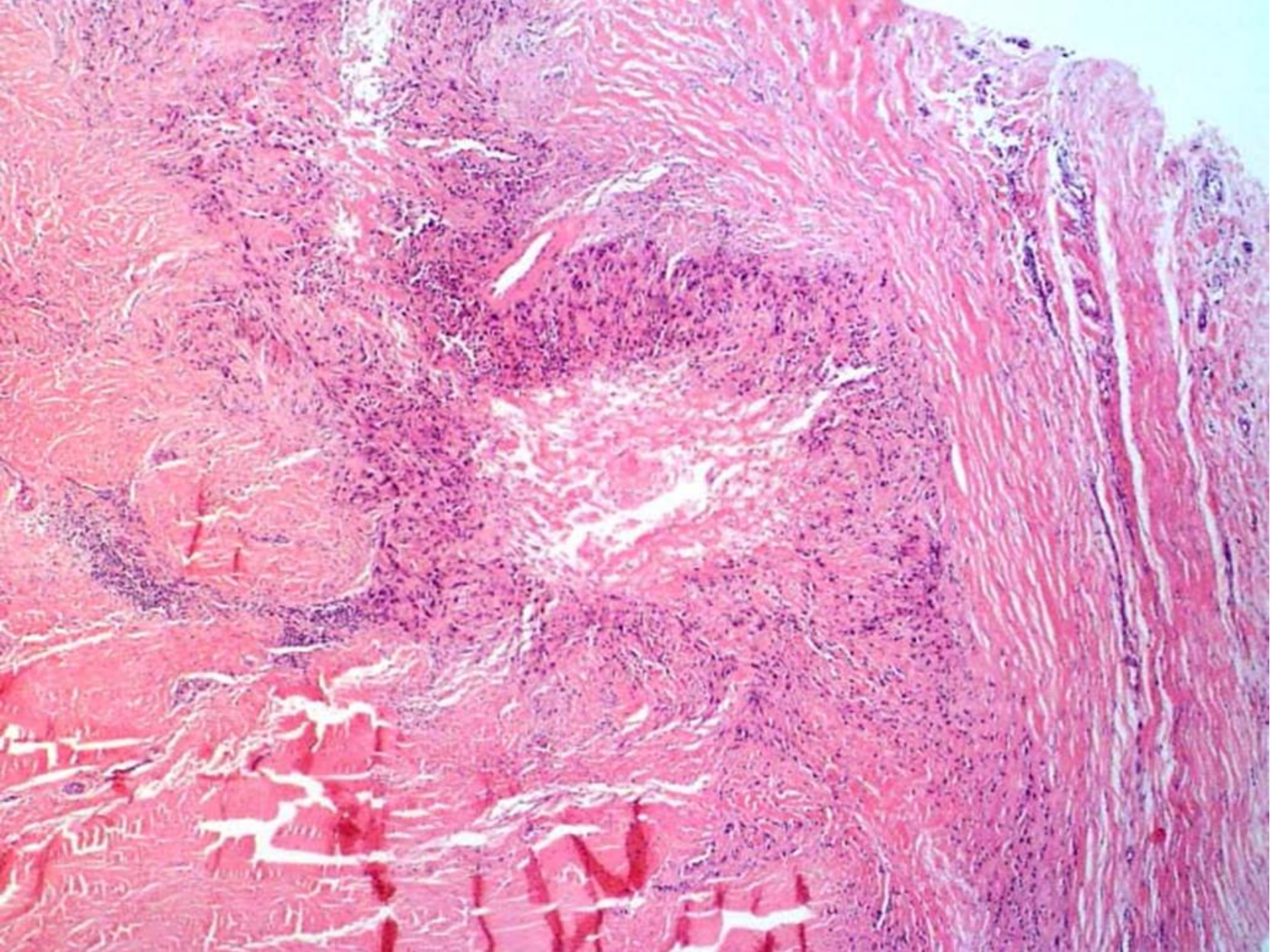


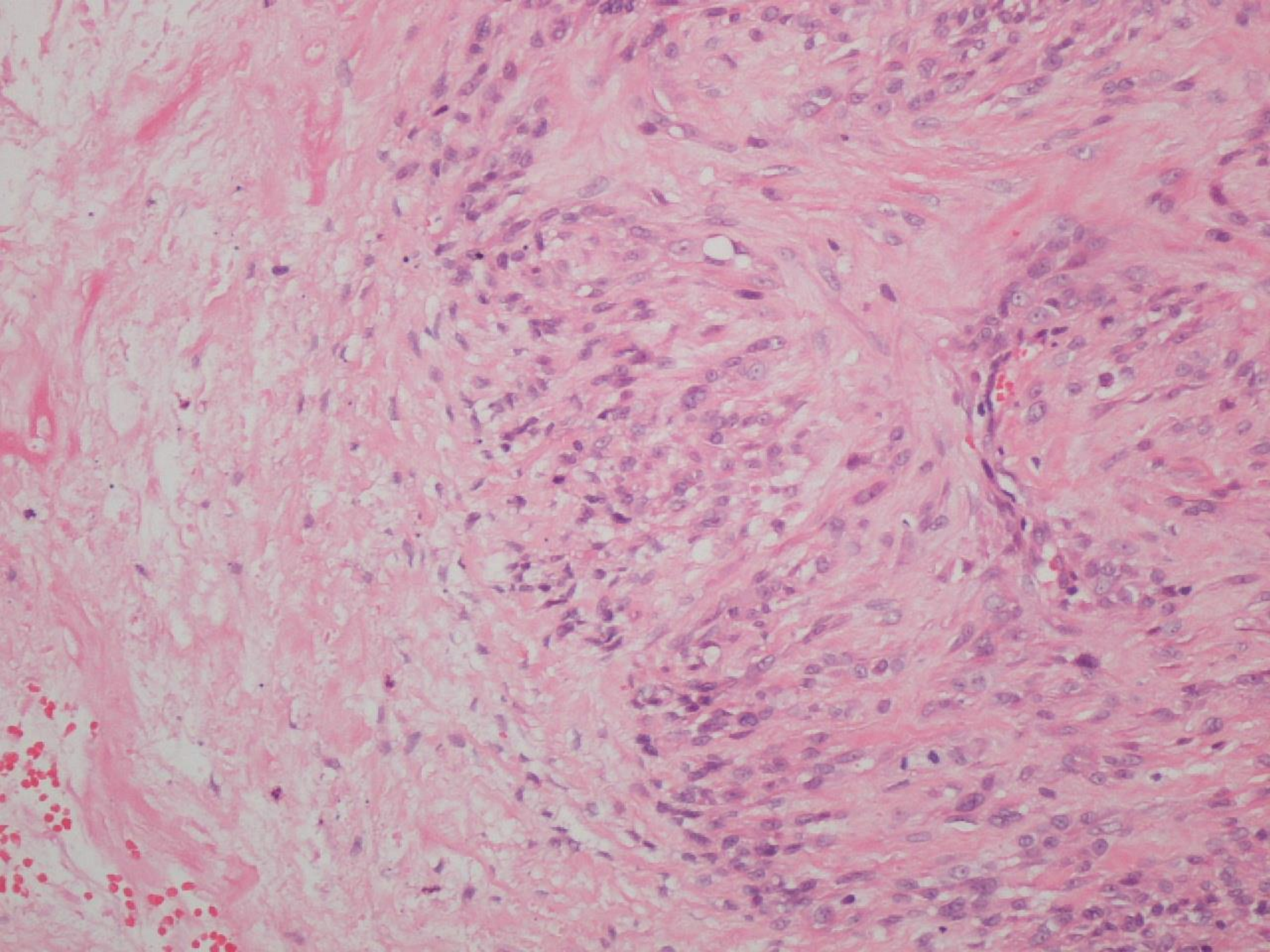


Tm hücreleri
nodüler/granülom benzeri
yapılanma
Santral nekroz
Perivasküler perinöral
yayılım

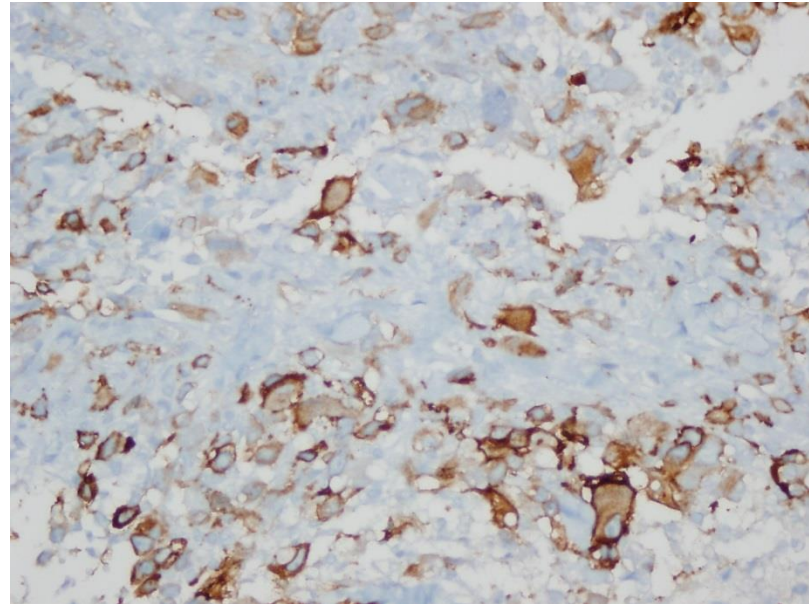
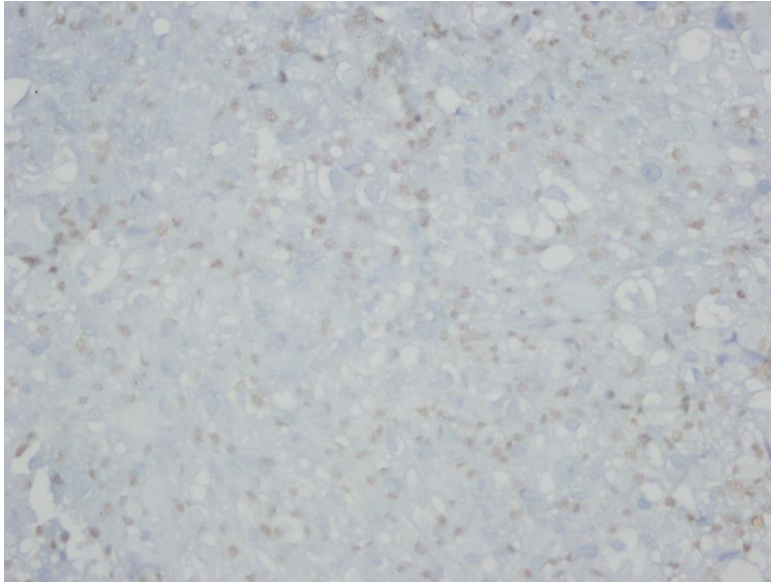
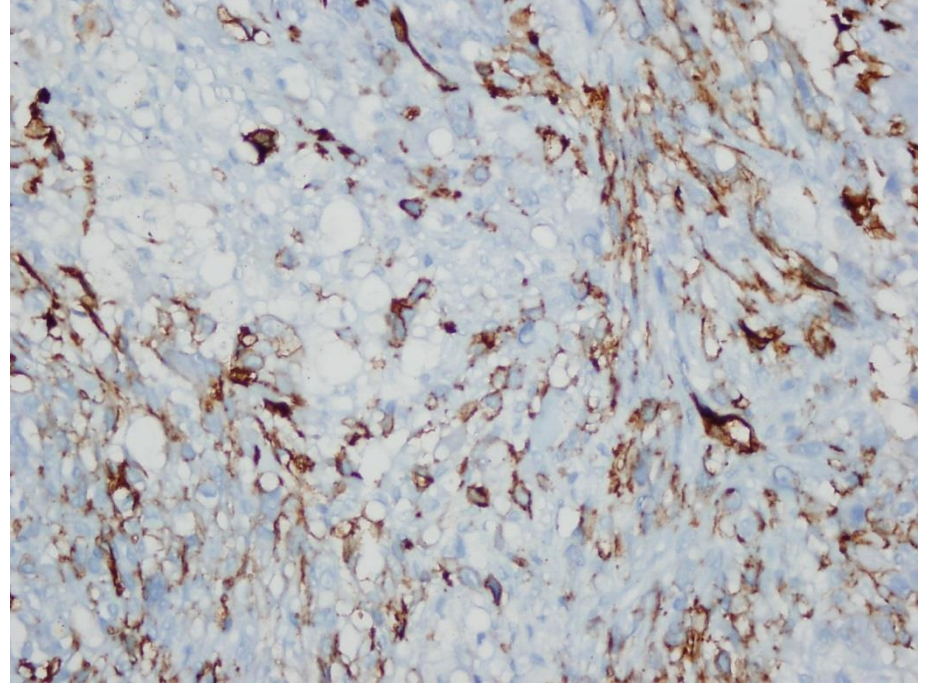
Ne klinisyen ne biz tümöre
benzetemeyebiliriz **DİKKAT**







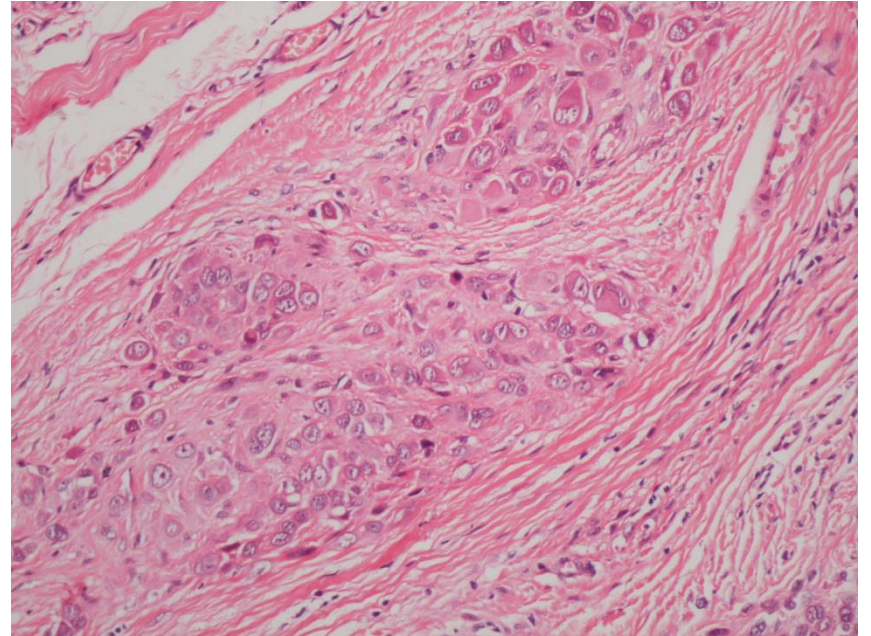
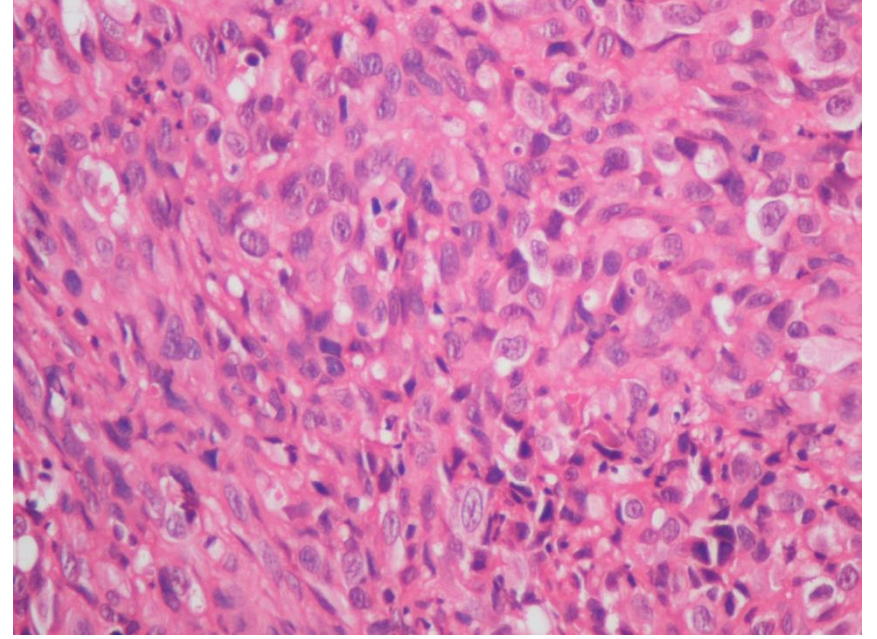
- Vimentin ve epitelial markerlar (pansitokeratin, EMA antigen), ve %50 vakada CD34 ve S-100 protein,
- NSE, desmin, ve aktinler nadiren
- sitokeratin 8, 14, ve 19, nadiren 7, 20, ve 5/6.
- son dönemde karakteristik **INI1 ekspresyon kaybı**
- **Tümör süpressör geni kromozom 22 de**

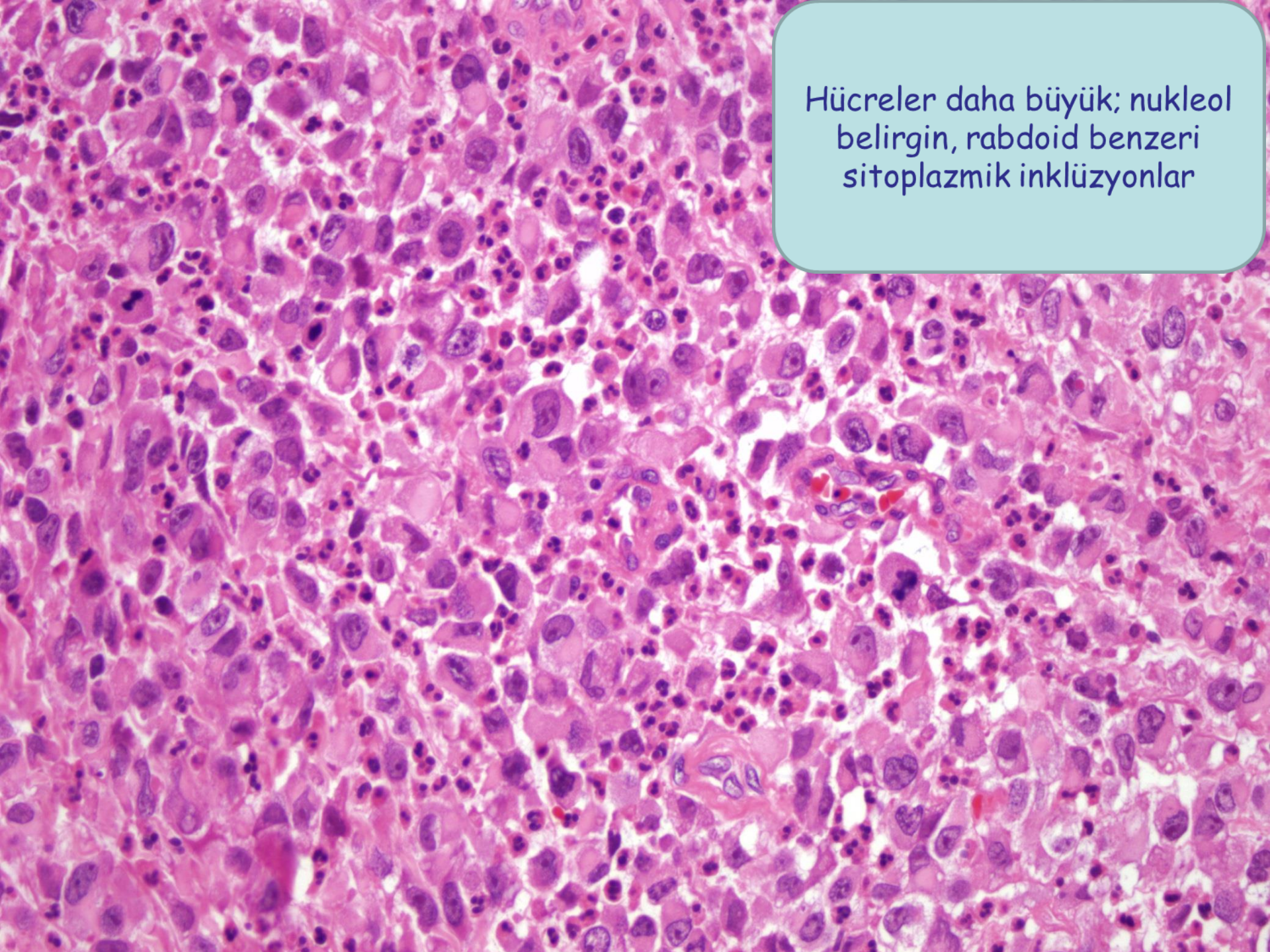


Epiteloid sarkom

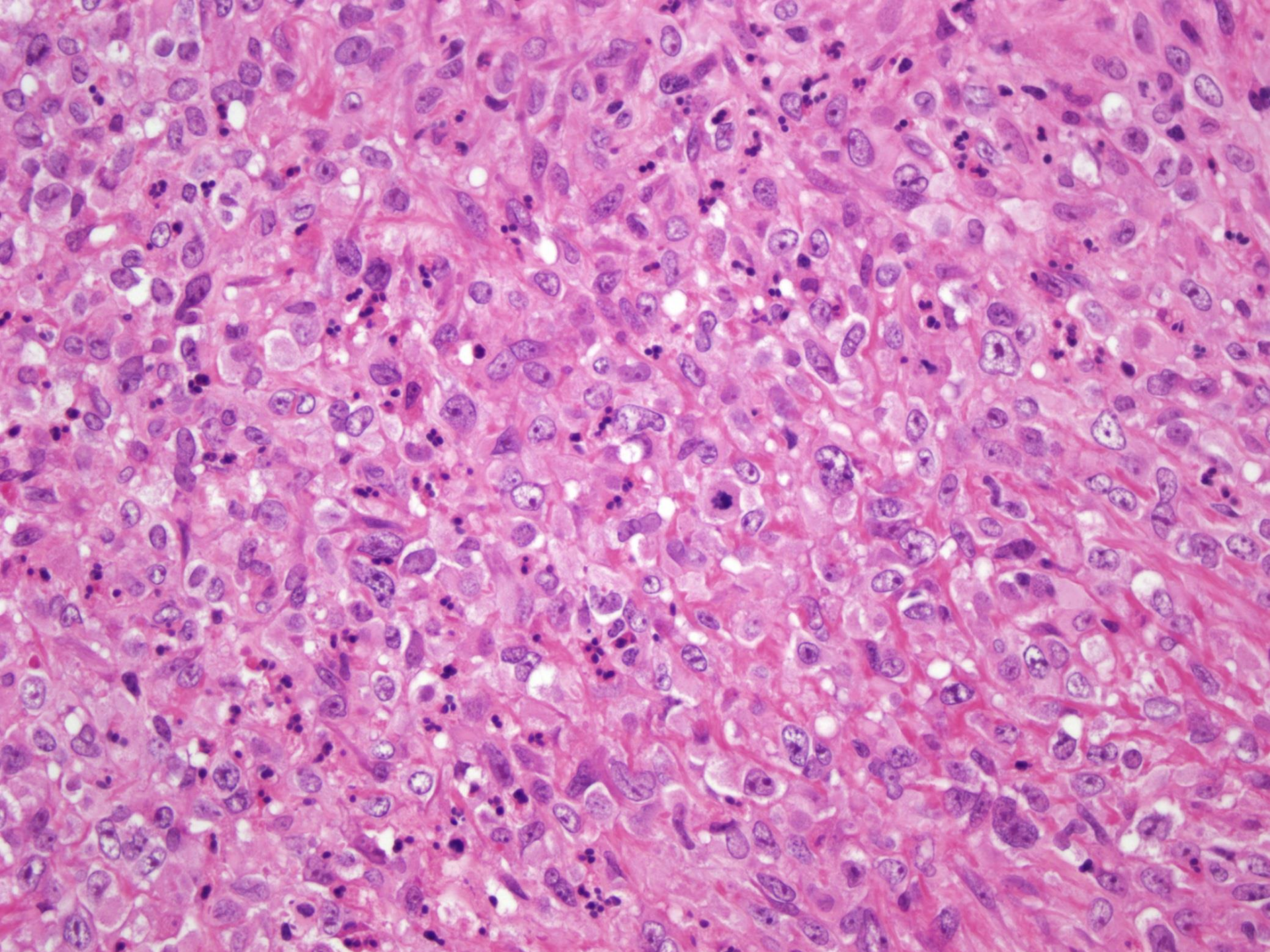
- Proksimal tip
- Anjiomatoid tip
- Fibrom benzeri tip
- Miksoid varyant

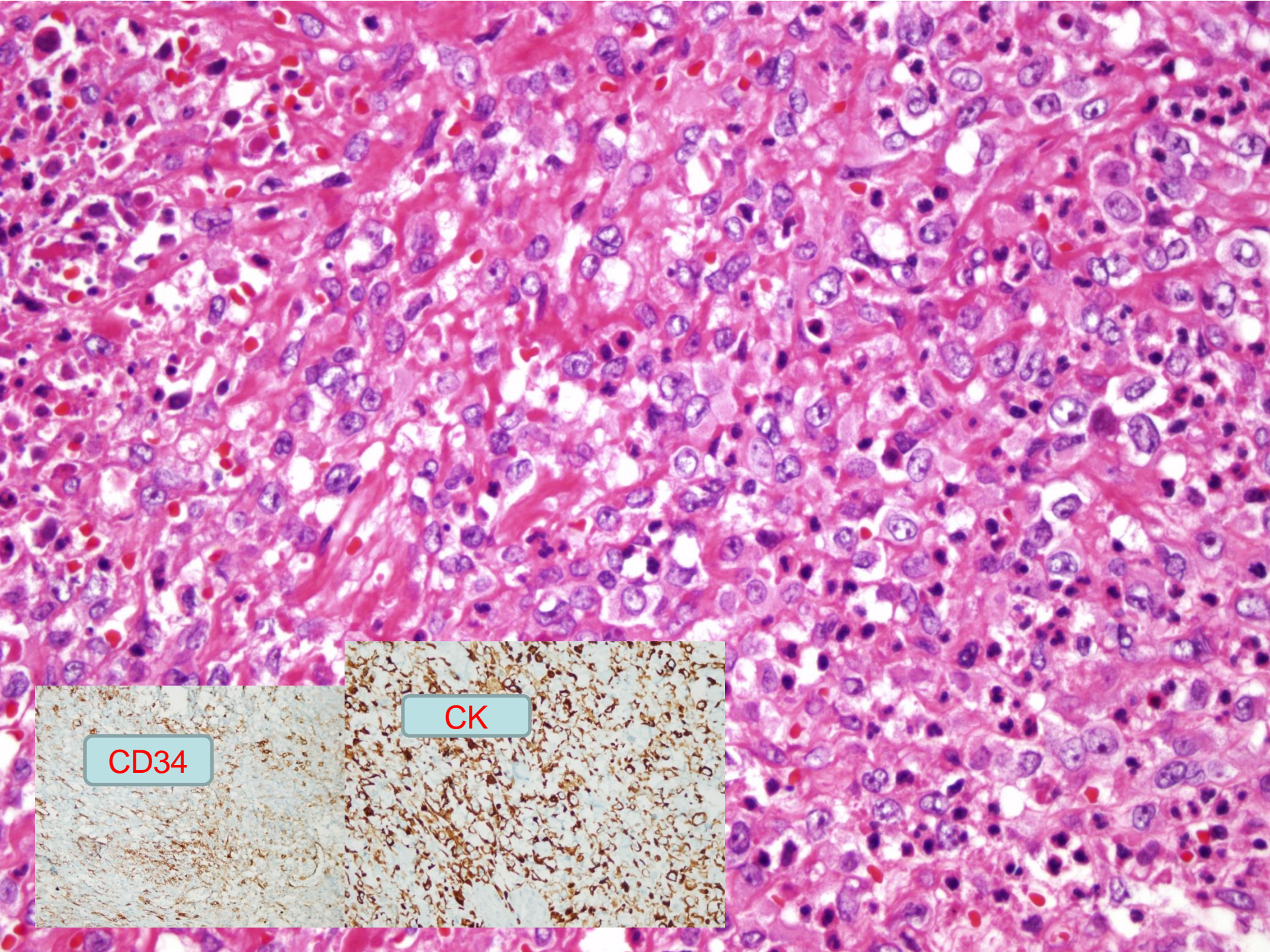
- Proksimal tip büyük poligonal ve yuvarlak tm hücreleri pleomorfik veziküler çekirdekler belirgin nükleol rabdoid morfoloji



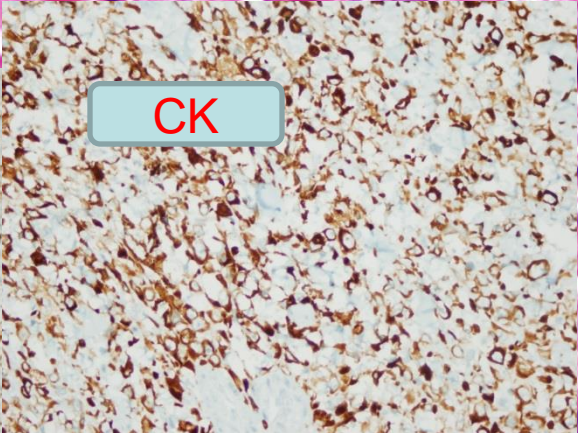


Hücreler daha büyük; nukleol belirgin, rabdoid benzeri sitoplazmik inklüzyonlar



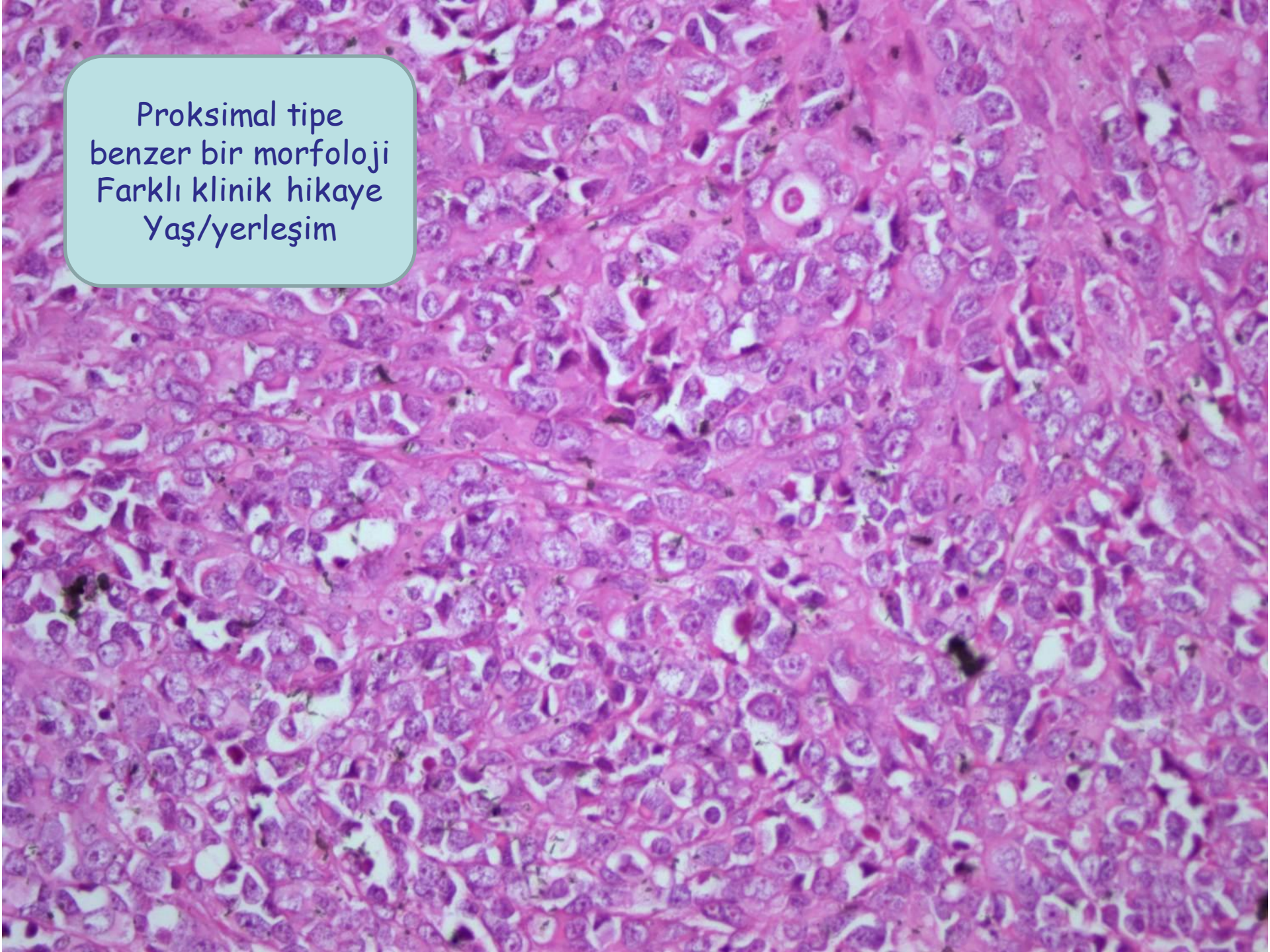


CD34



CK

Proksimal tipe
benzer bir morfoloji
Farklı klinik hikaye
Yaş/yerleşim



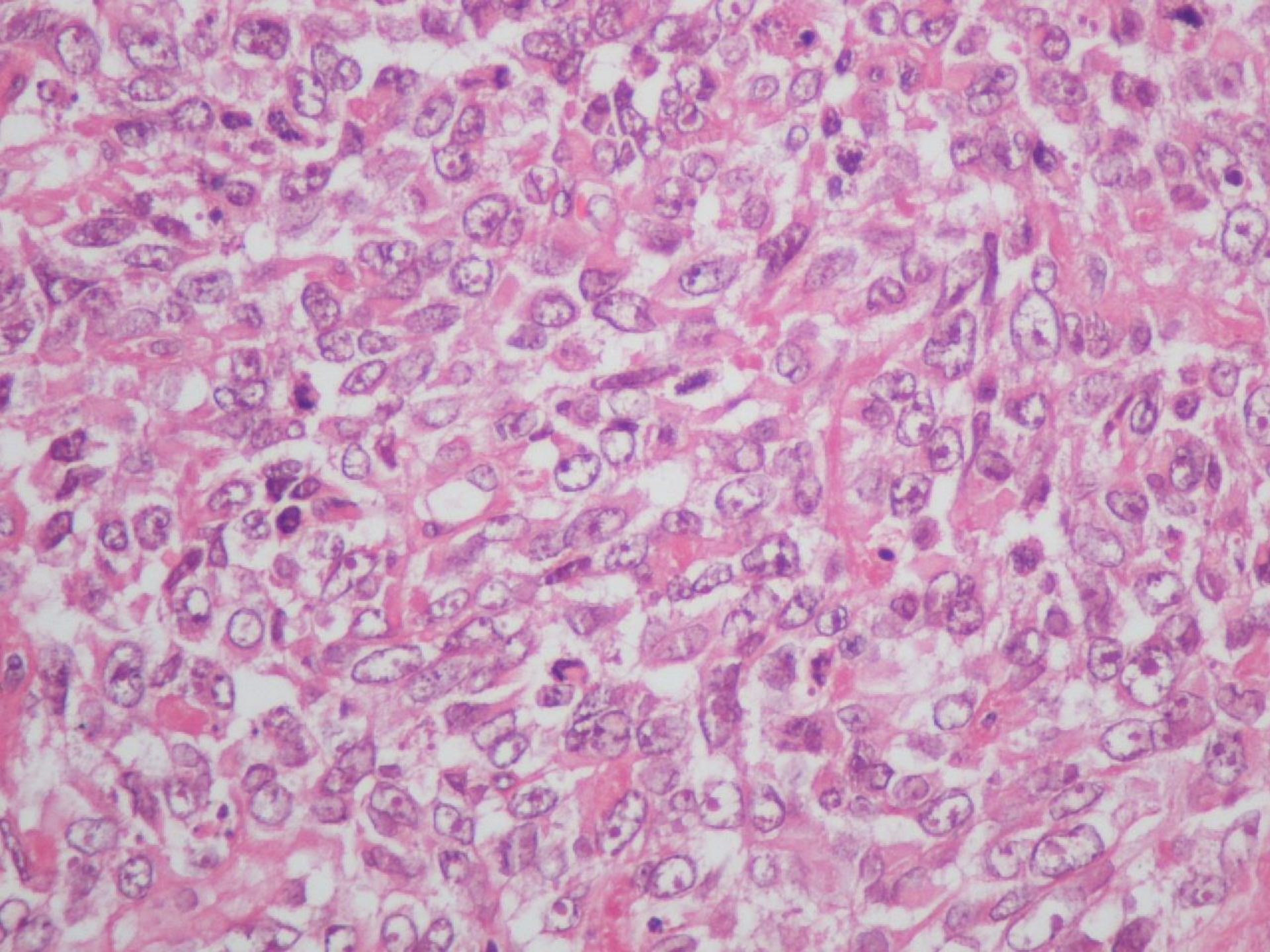
Proksimal tip(büyük hücreli)-Epiteloid sarkom

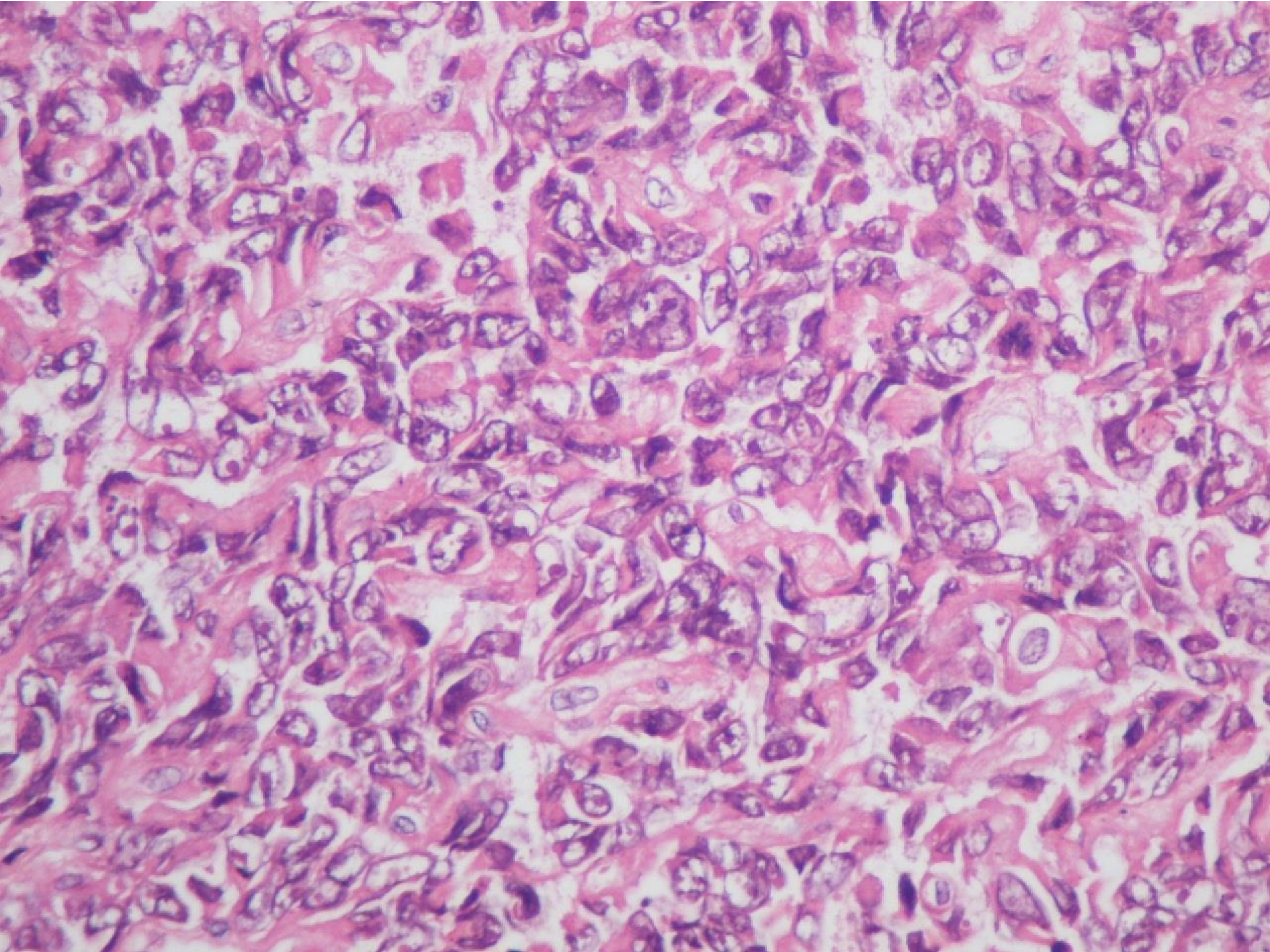
- Yerleşim ve morfoloji farkı
- Proksimal ekstremitelerde; uyluk, göğüs duvarı, aksilla, genitoanal bölge
- İHK benzerliği: PSK, EMA, CD34 (+)
INI-1 (-)
- İHK farkı: %50 desmin (+) (Yanlış RMS tanısına DİKKAT!)

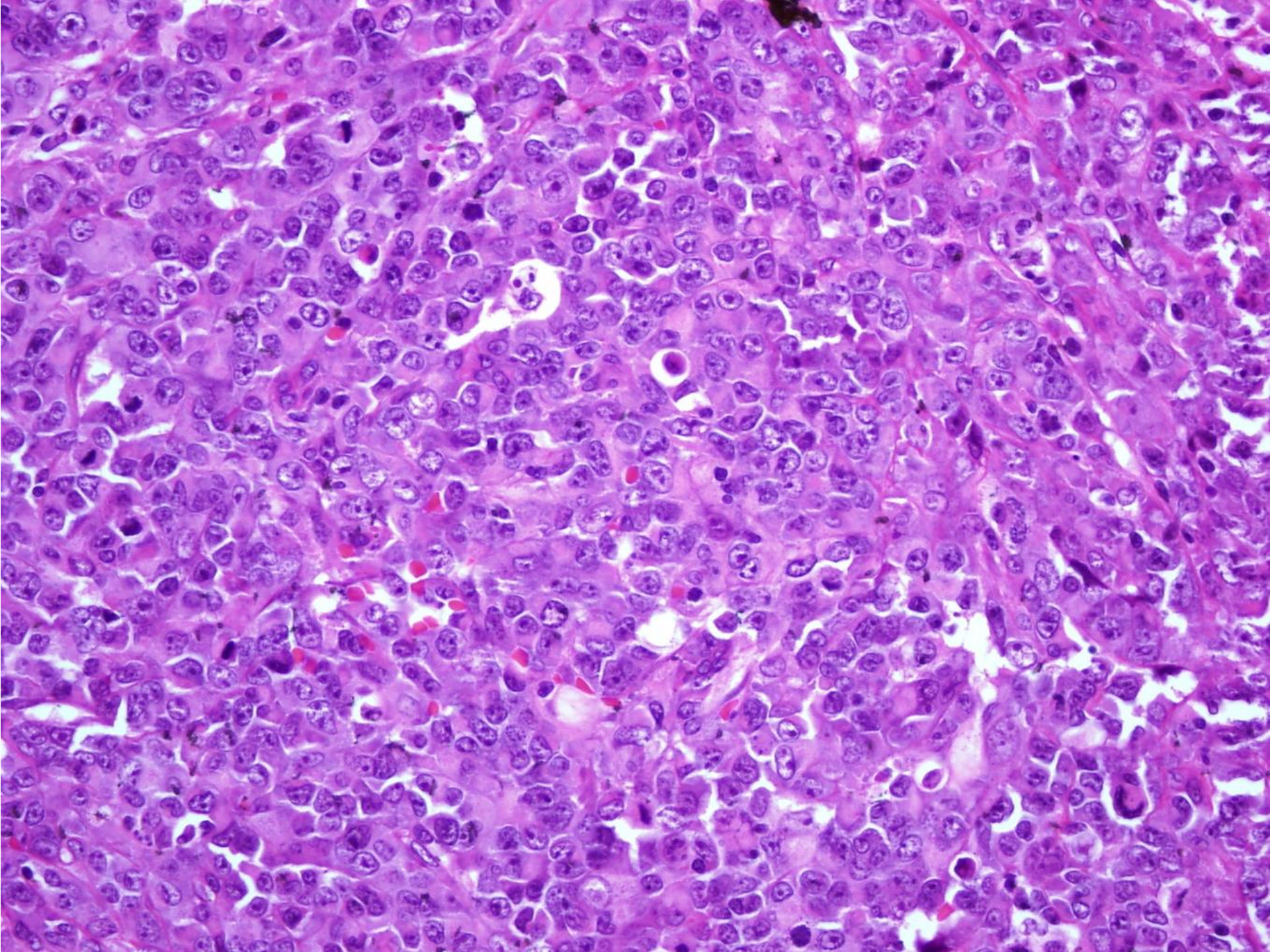
2 aylık bebek
Sağ uylukta şişlik

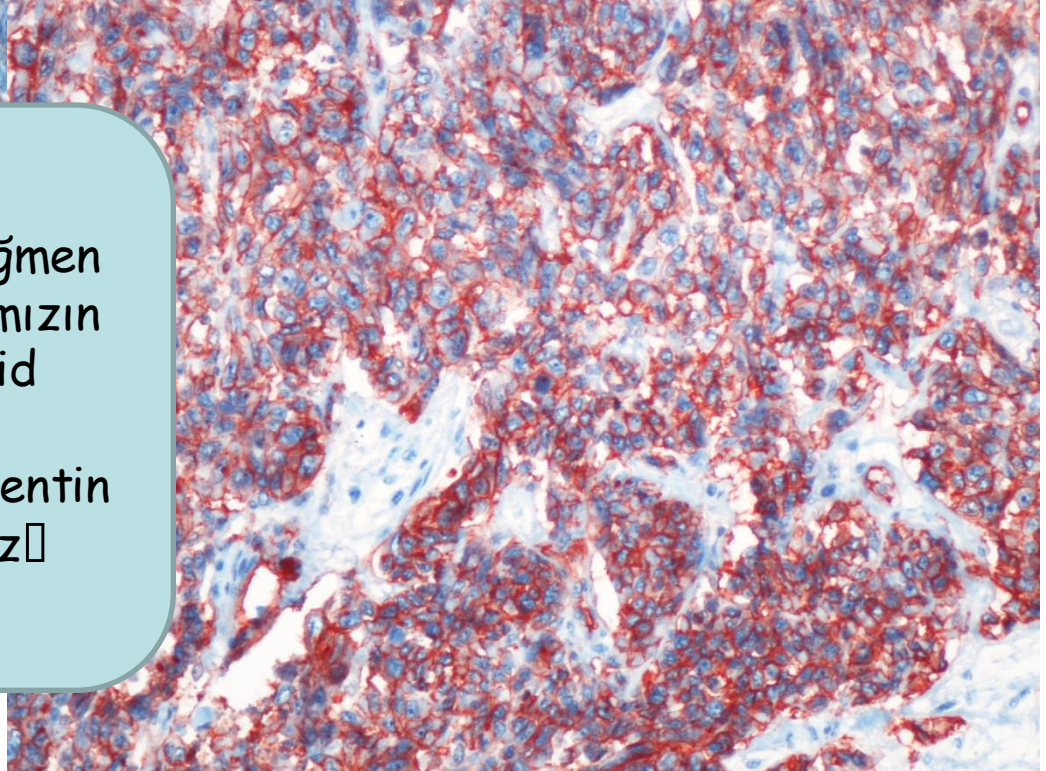
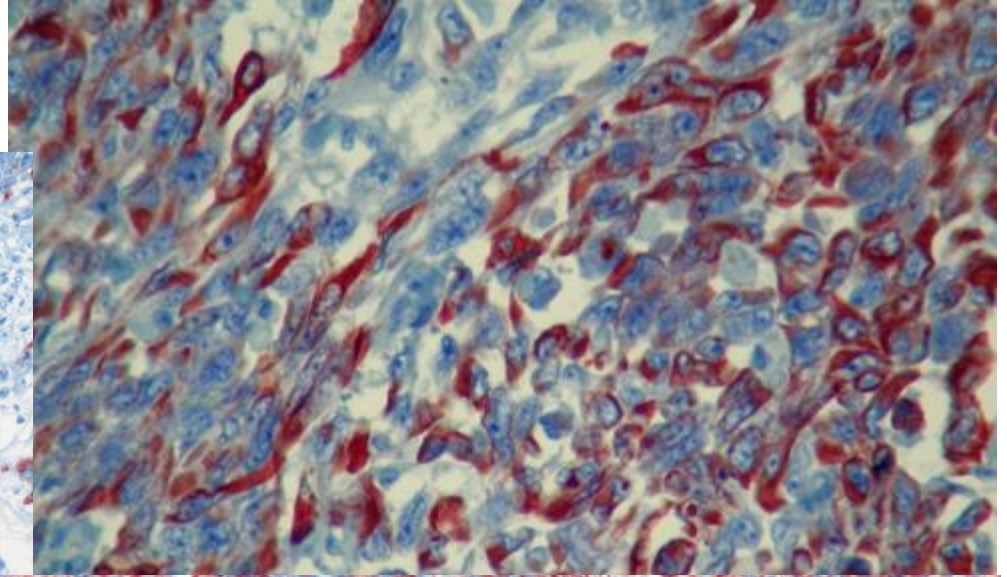
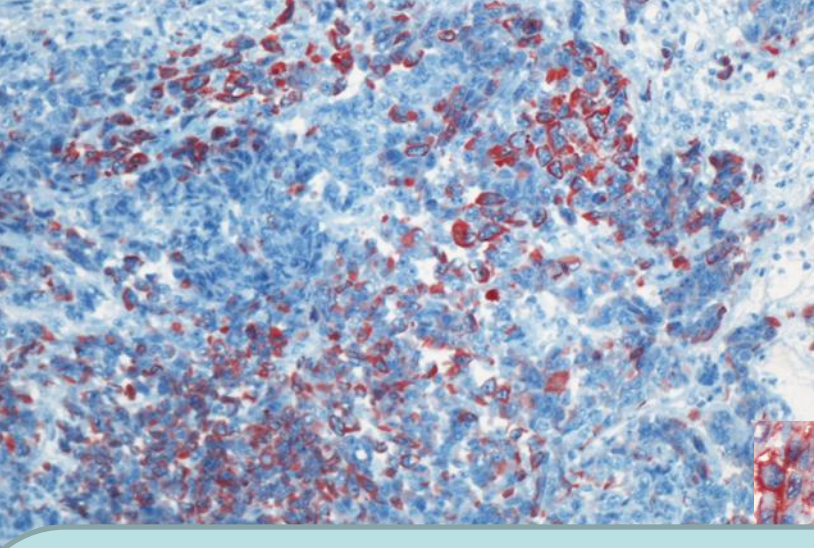
Ön tanılar: infantil
fibrosarkom











Çok nadir bir tümör olmasına rağmen yaşının küçüklüğü nedeniyle aklımızın bir köşesinde bulunan "rabdoid tümör"ü, bulunduğu yerden çıkarabilirsek keratin, EMA, vimentin pozitifliği ile tanıya gidebiliriz□

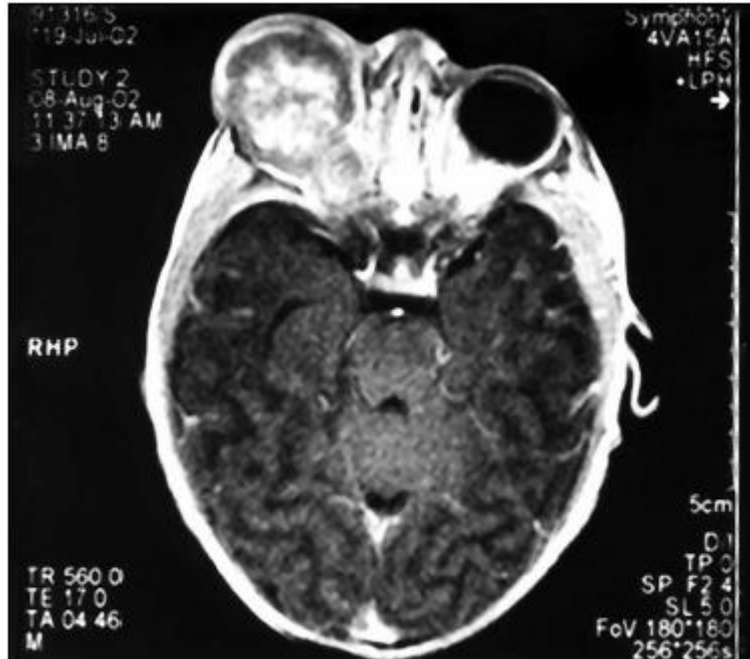
[Turk Patoloji Derg.](#) 2013;29(1):69-72. doi: 10.5146/tjpath.2013.01152.

[**Malignant rhabdoid tumor of the orbit**].

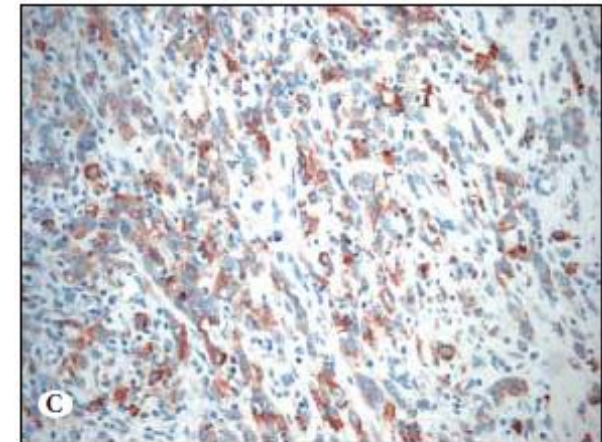
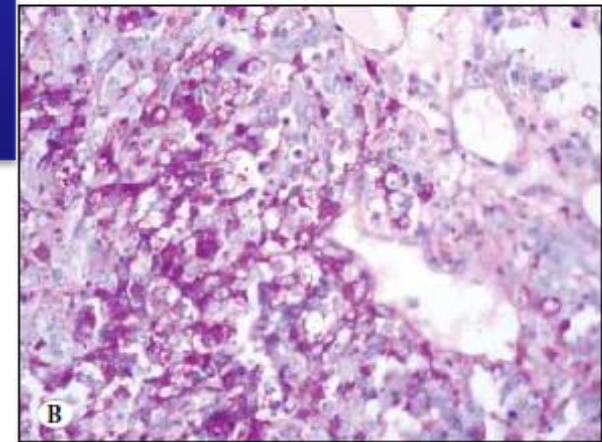
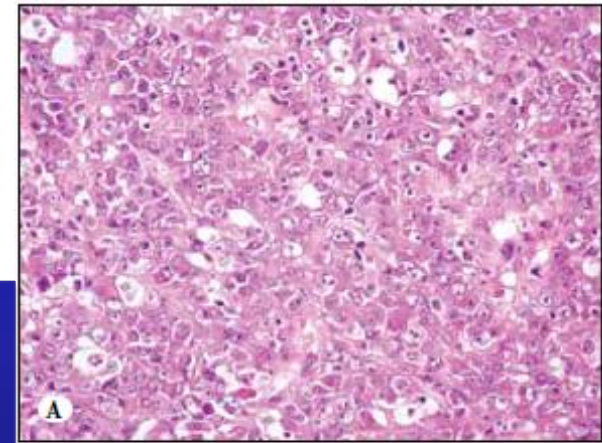
[Article in Turkish]

[Karaman S](#), [Celkan T](#), [Dervişoğlu S](#), [Ergen M](#), [Özkan A](#), [Apak H](#), [Yıldız I](#), [Pazarlı H](#).

Malign rhabdoid tümör,
En sık yerleşim yeri santral sinir sistemi ve böbrek
Nadiren orbita
perinatal dönemde orbital malign rhabdoid tümör
8 haftalık kız
doğumdan itibaren fark edilen sağ göz kapağında şişlik
şikayeti
Sağ proptozisi



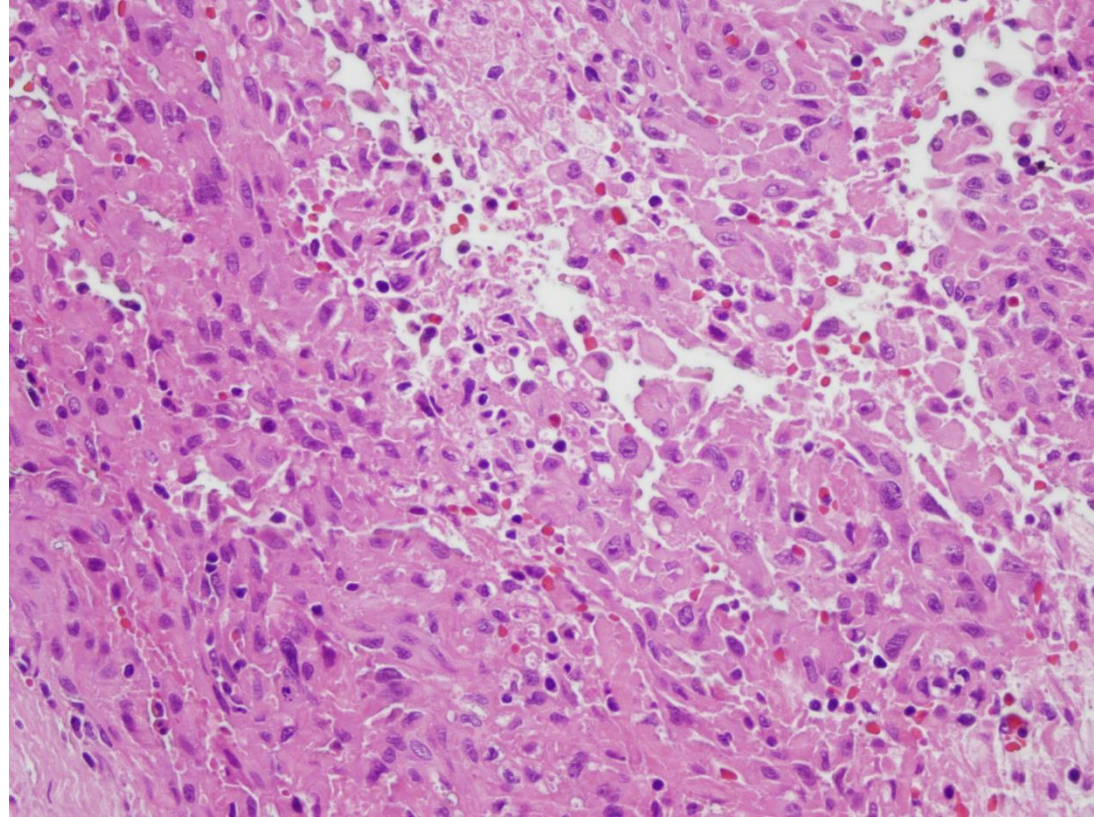
Şekil 1: Olgunun manyetik rezonans görüntülemesinde gözün sağ üst dış bölgesinde yerleşen kitle görüntüsü.

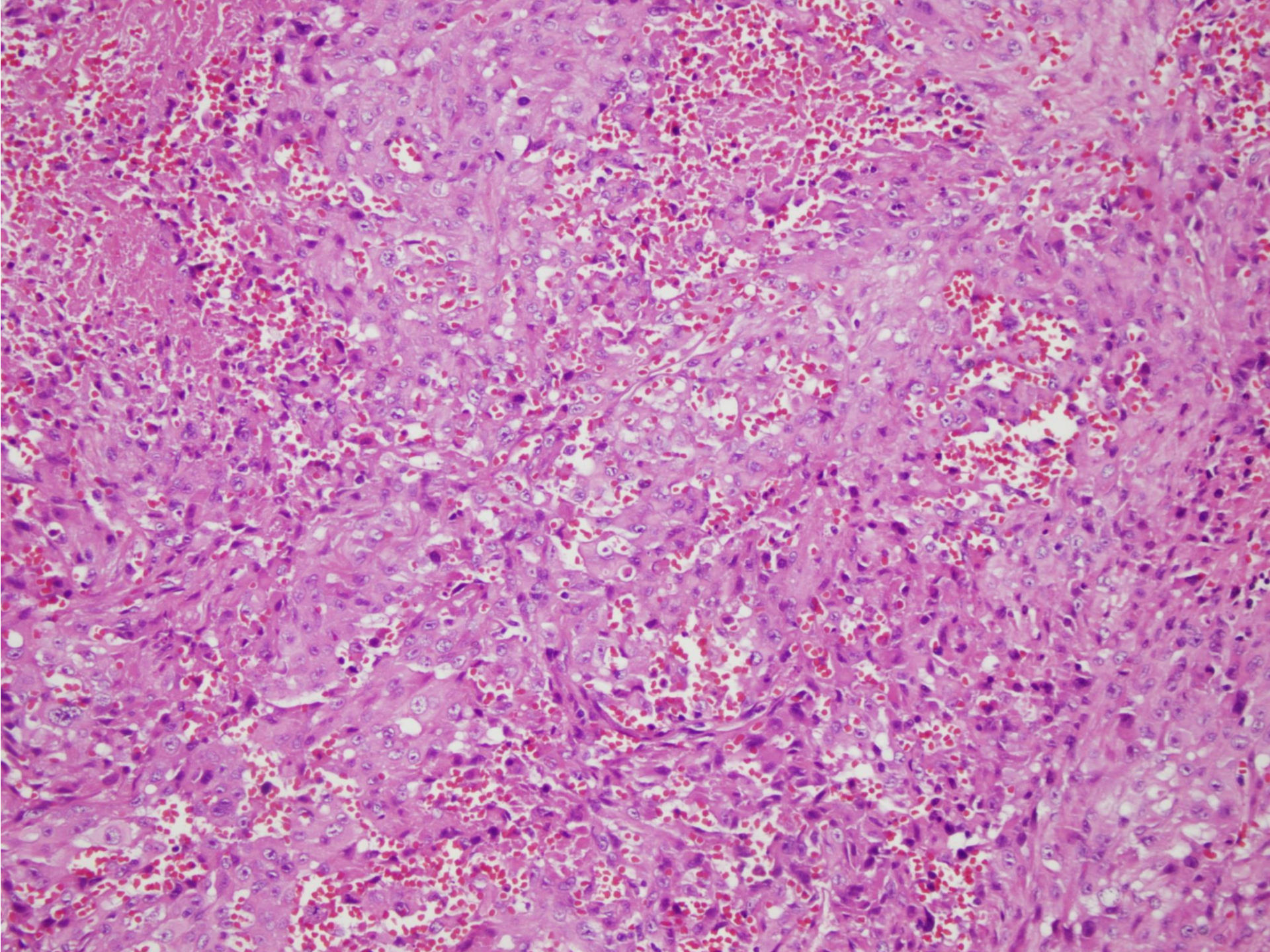


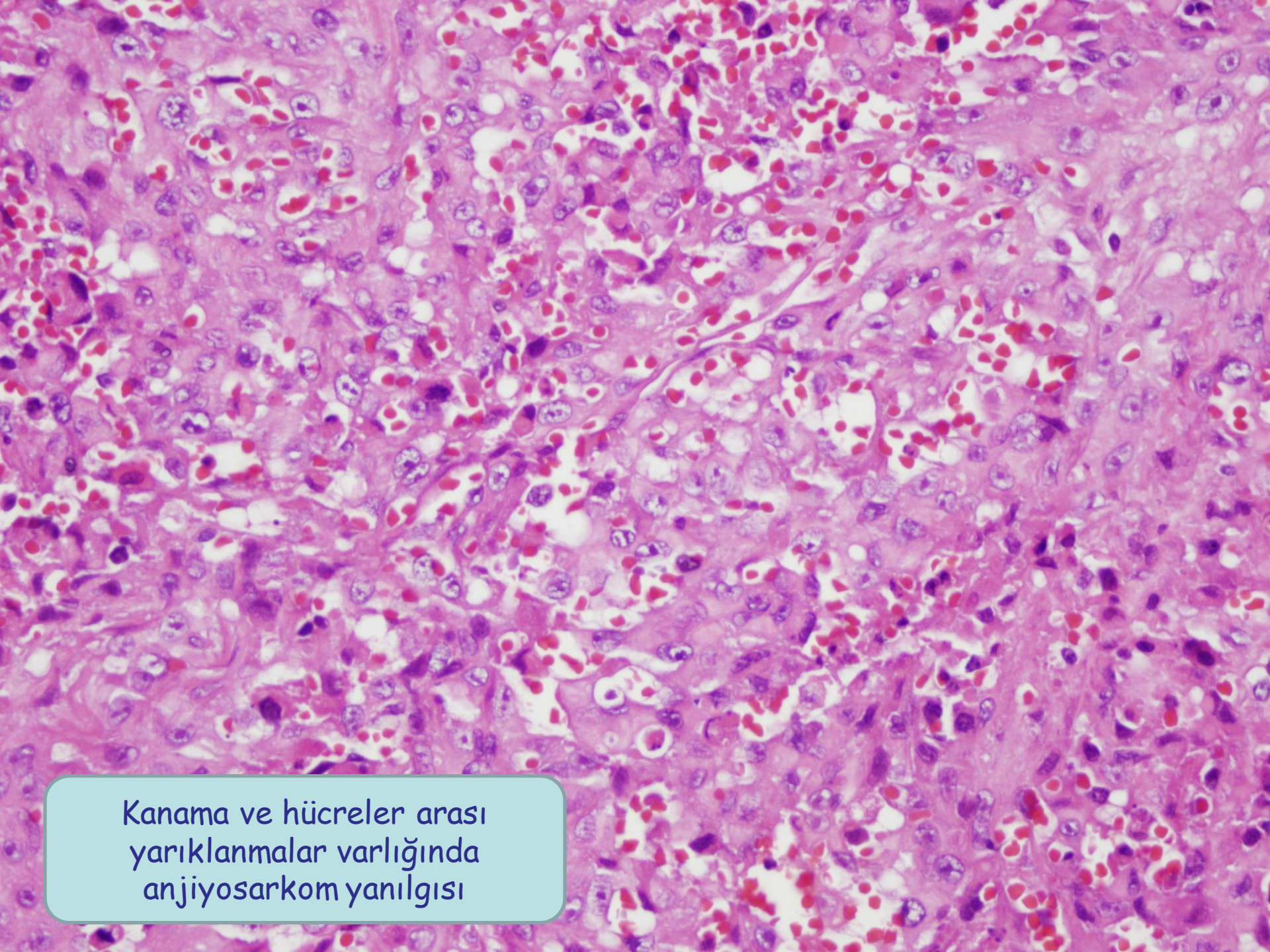
Şekil 2: A) Eksantrik nükleuslu geniş poligonal tümöral hücreler (H&E, x40), B) PAS (+) tümöral hücreler (x40), C) Sitoplazmik keratin pozitifliği, (x20).

Olağan
yer

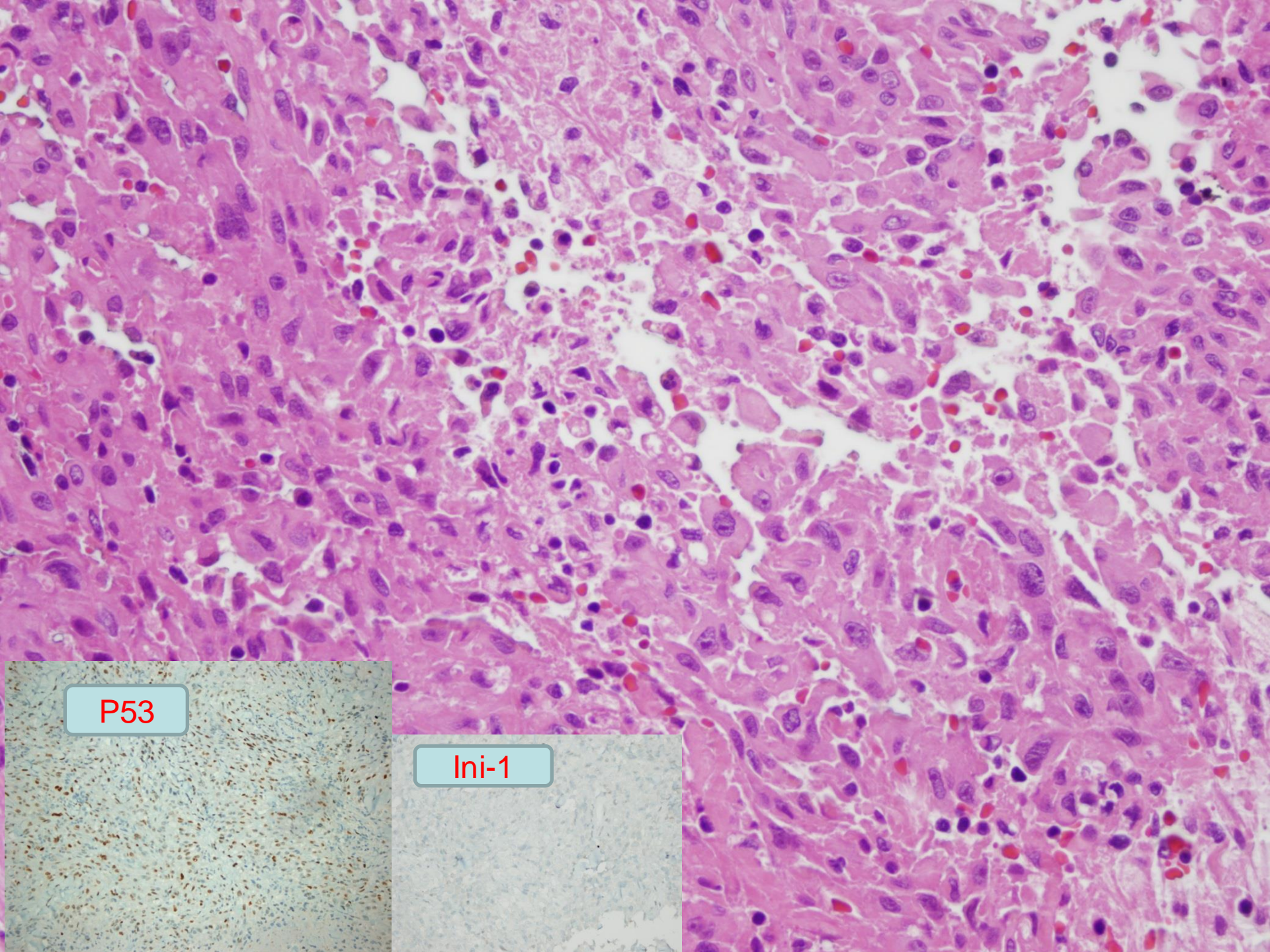
- Anjiomatoid Kistik,
kanla dolu
anjioektatik
boşluklar içsi
epiteloid tm
hücreleri ile döşeli





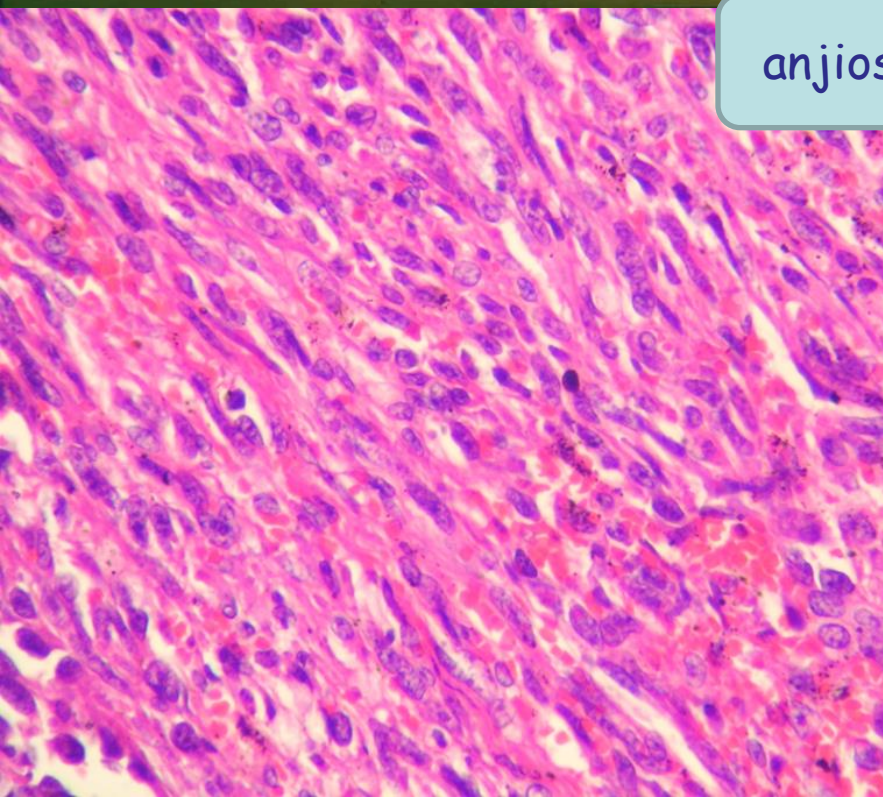
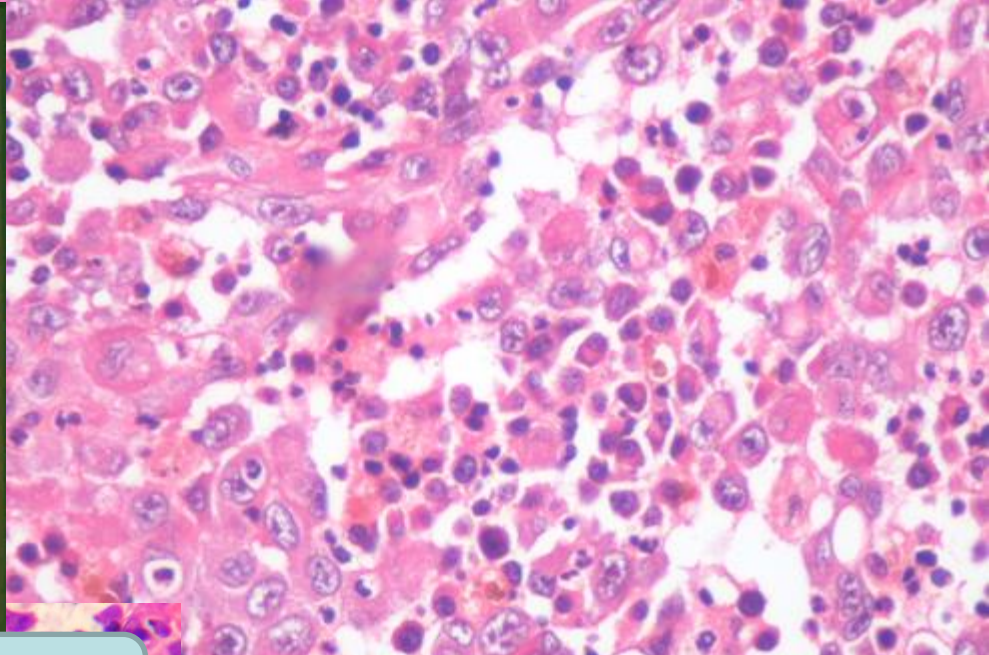
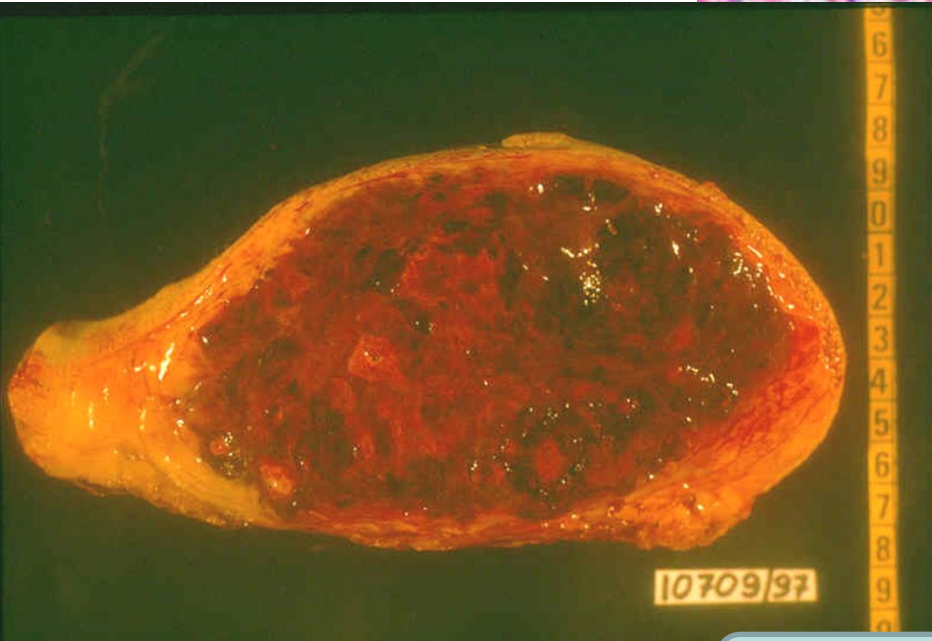


Kanama ve hücreler arası
yarıklanmalar varlığında
anjiosarkom yanılığı

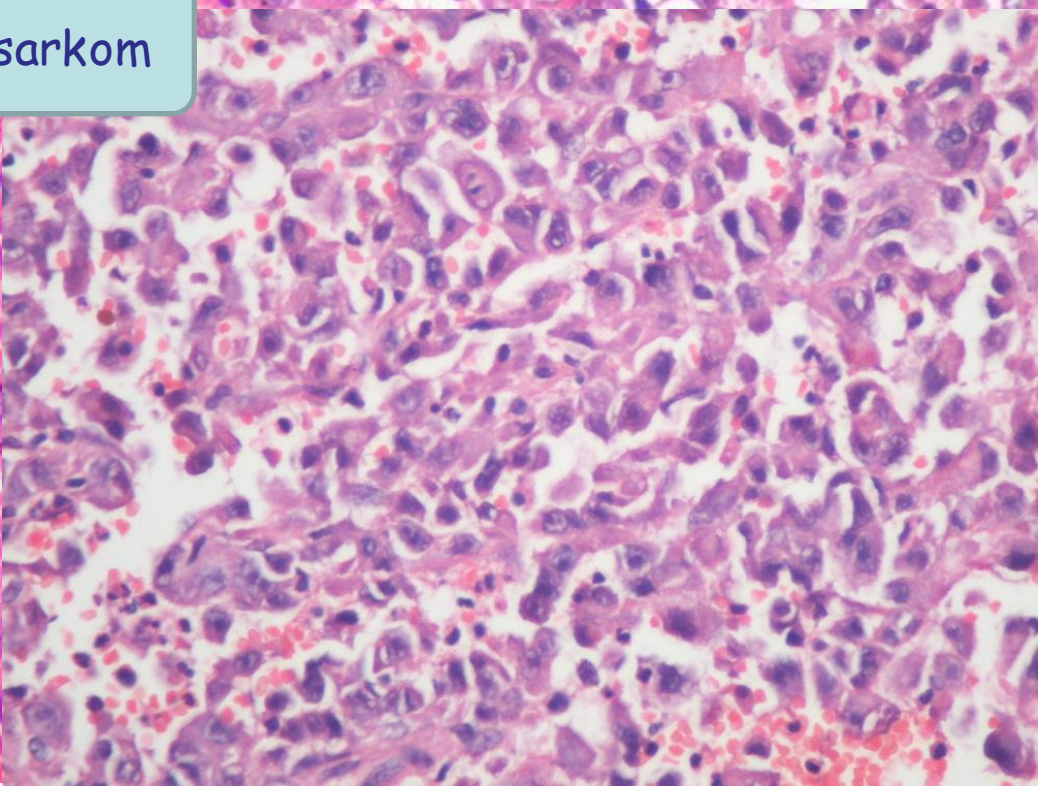


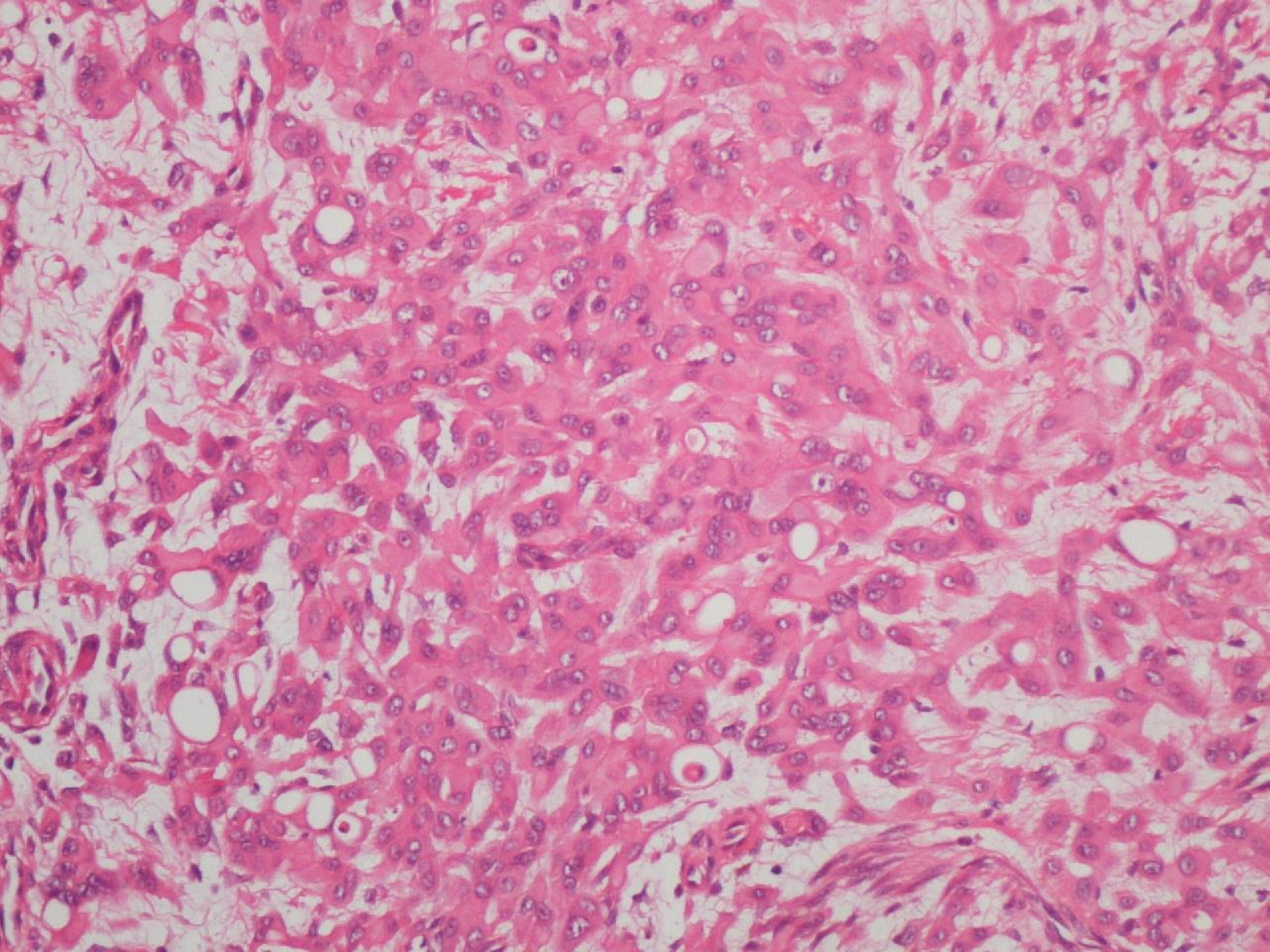
P53

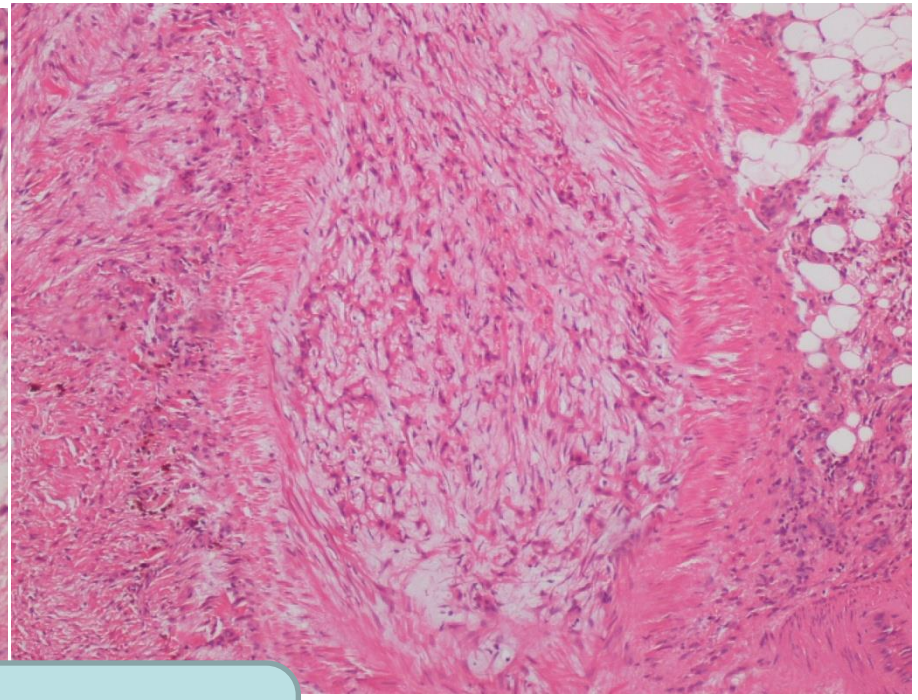
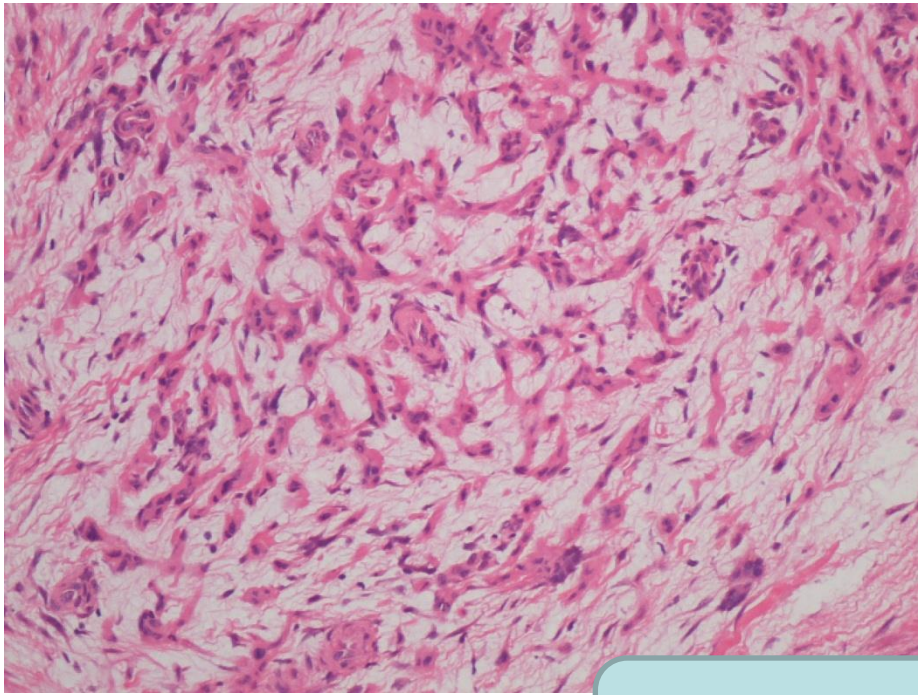
Ini-1



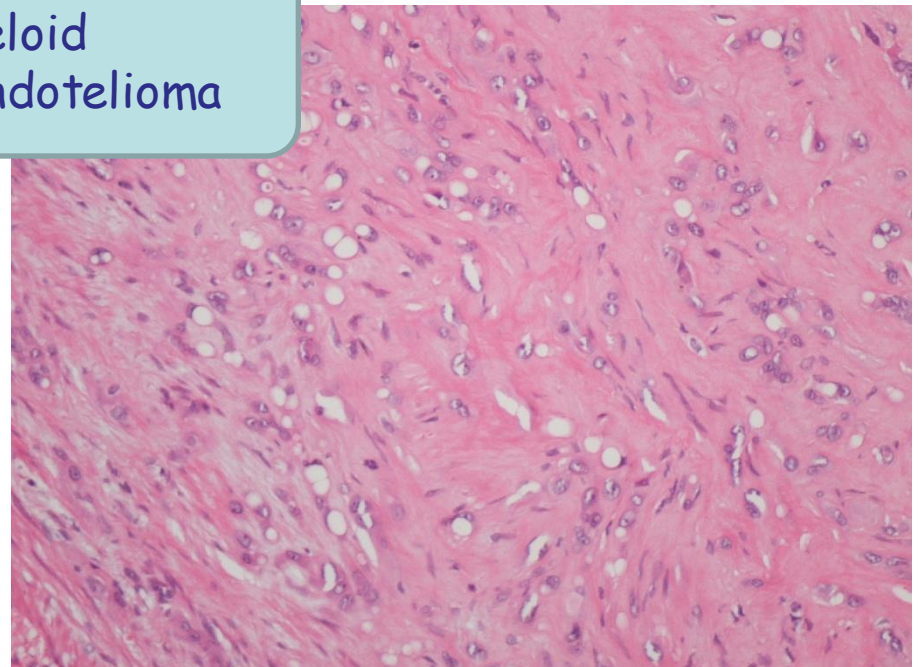
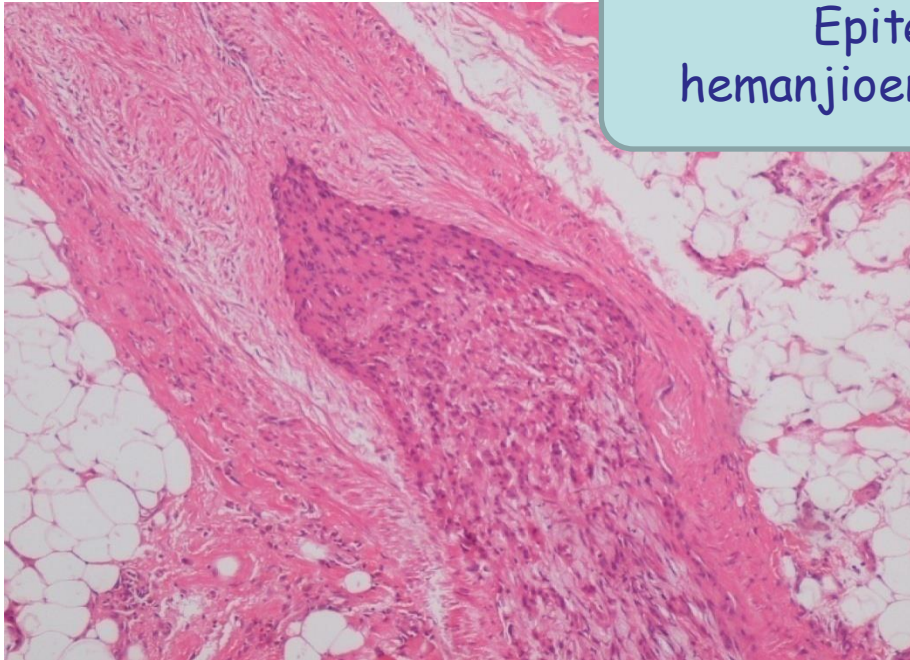
anjiosarkom





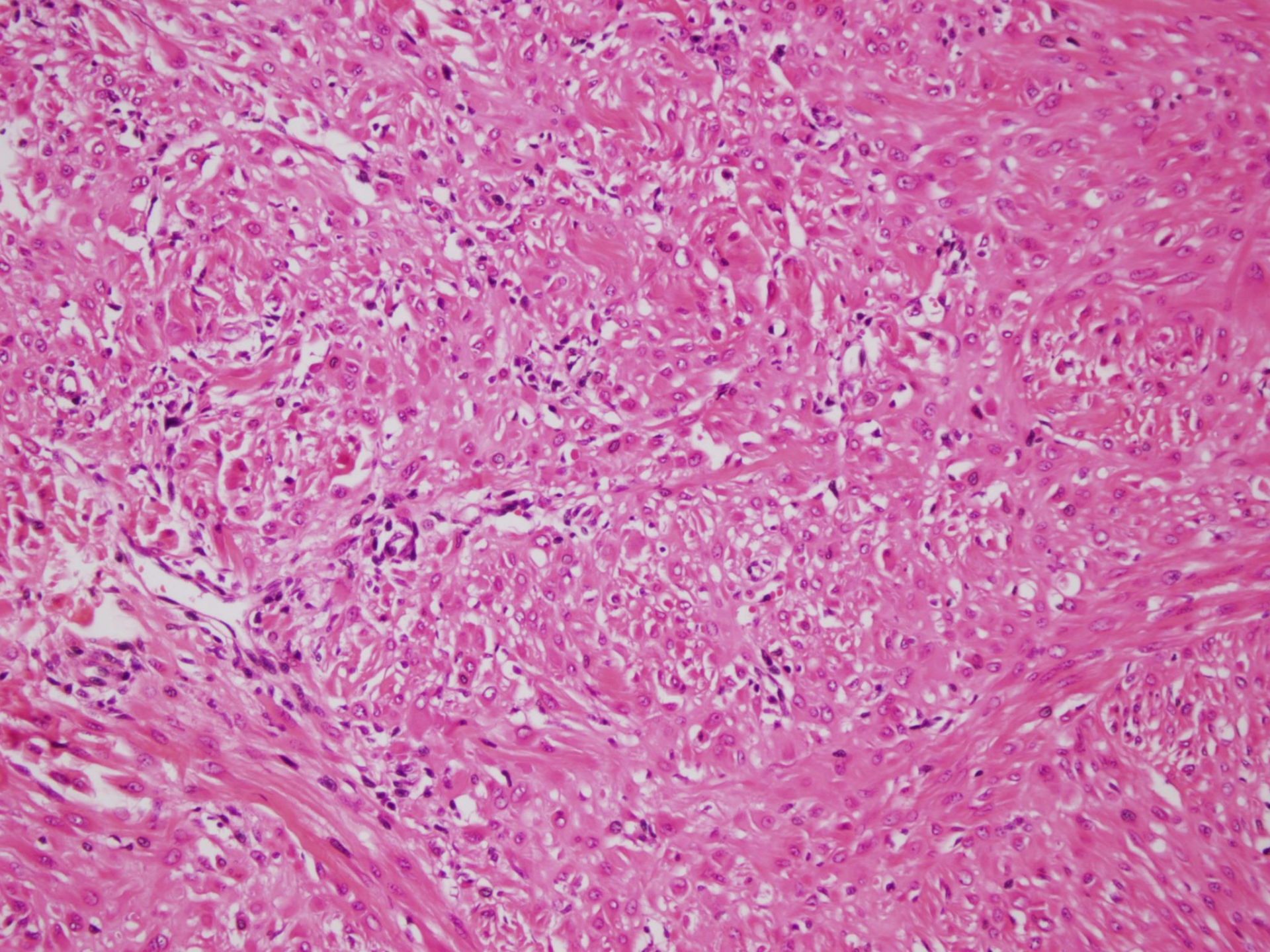


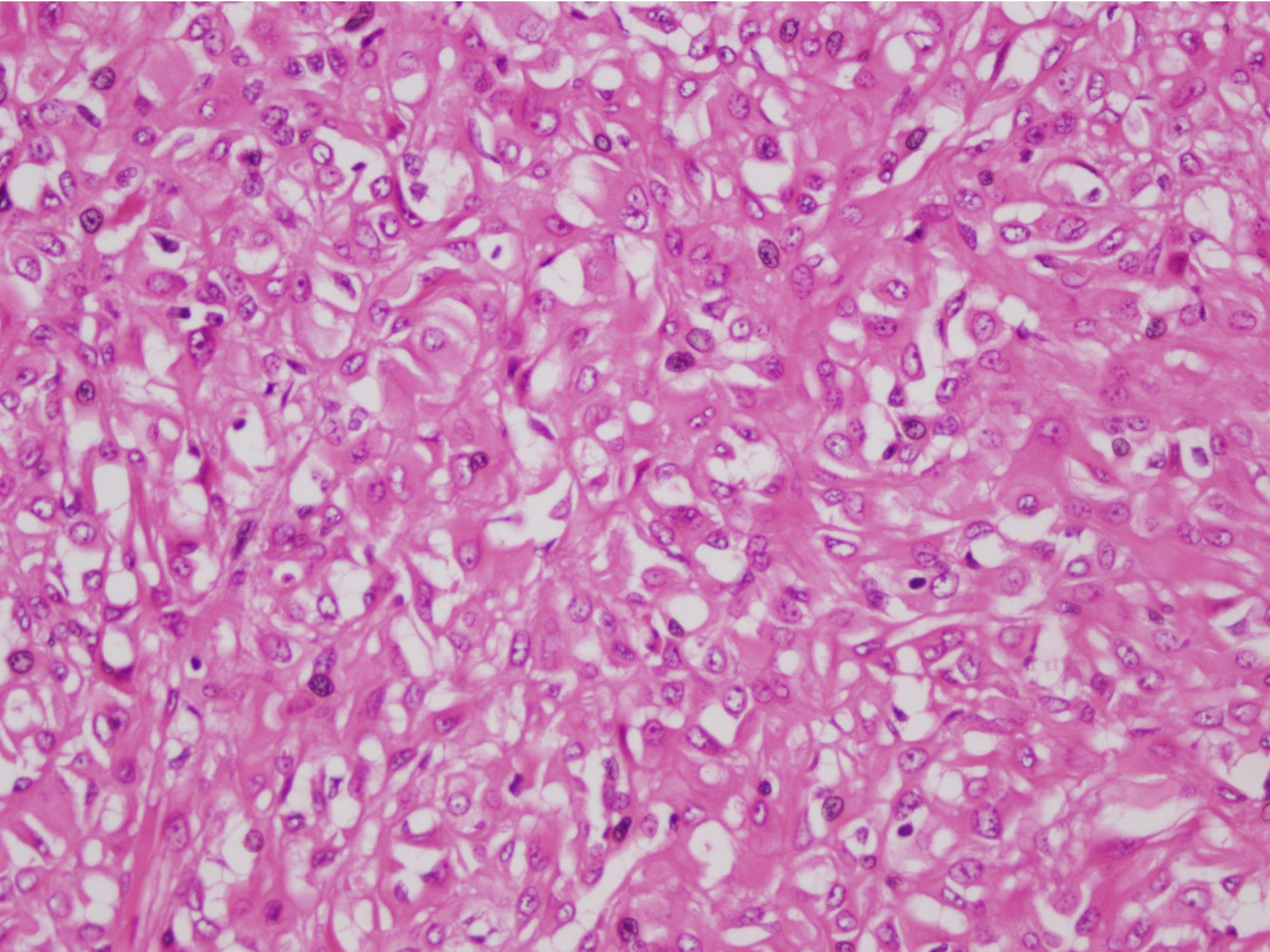
Epiteloid
hemanjioendotelioma

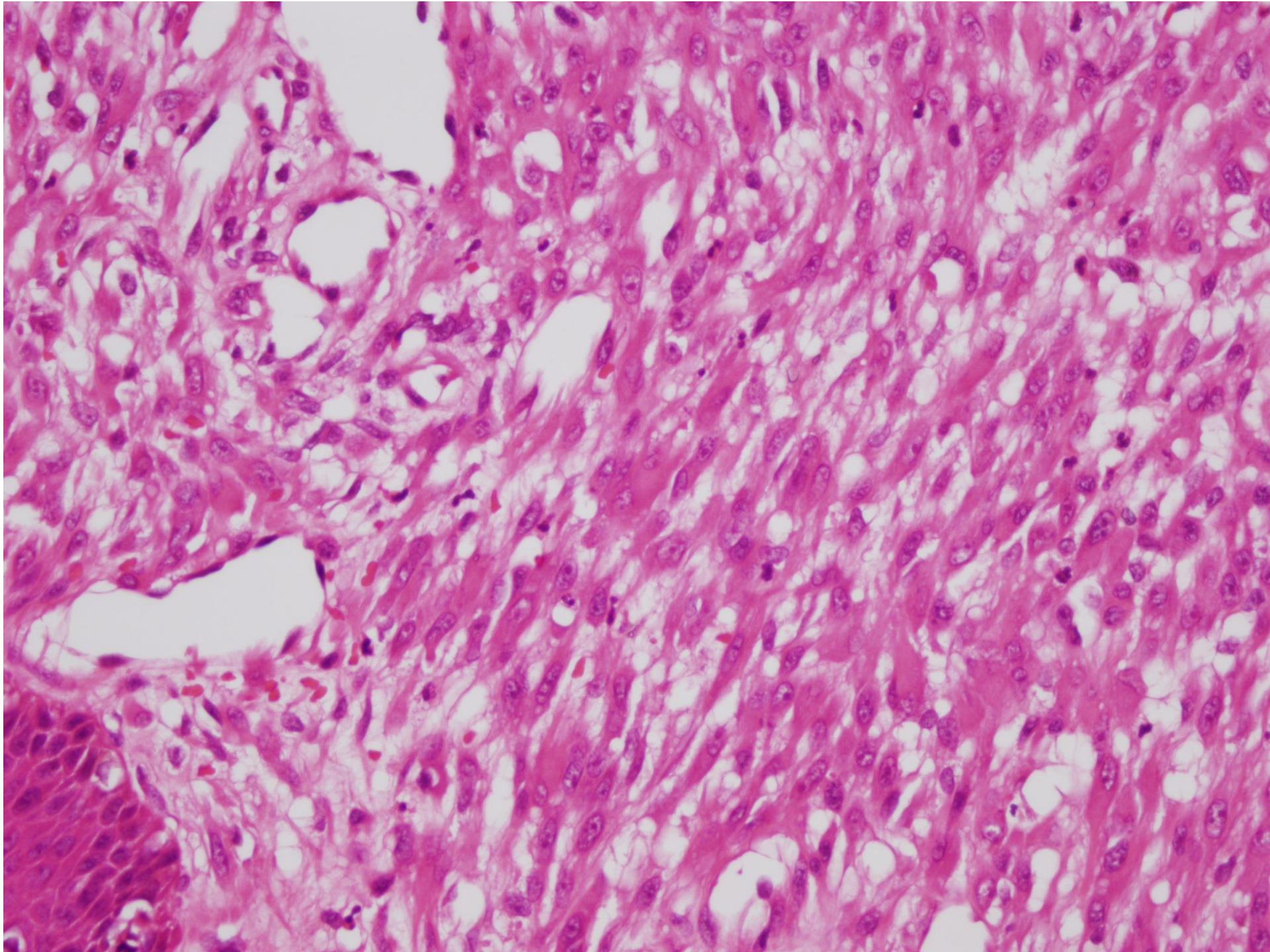


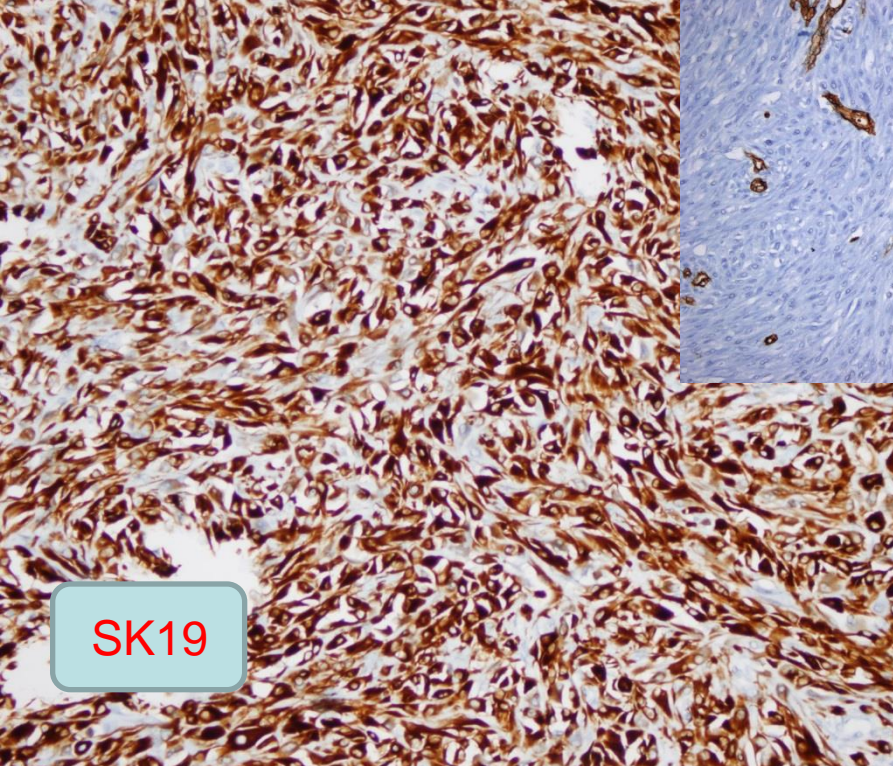
26 Yaş erkek hasta
Multifokal lezyonlar biopsi
ile epiteloid sarkom tanılı



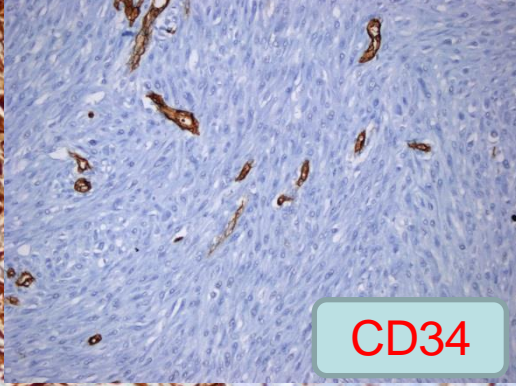








SKI-19

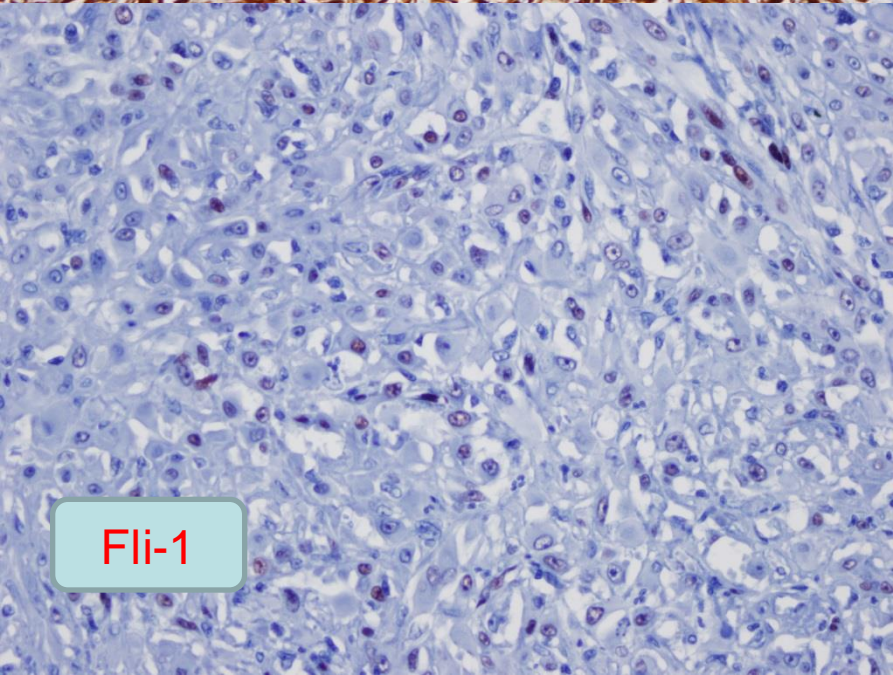


CD34

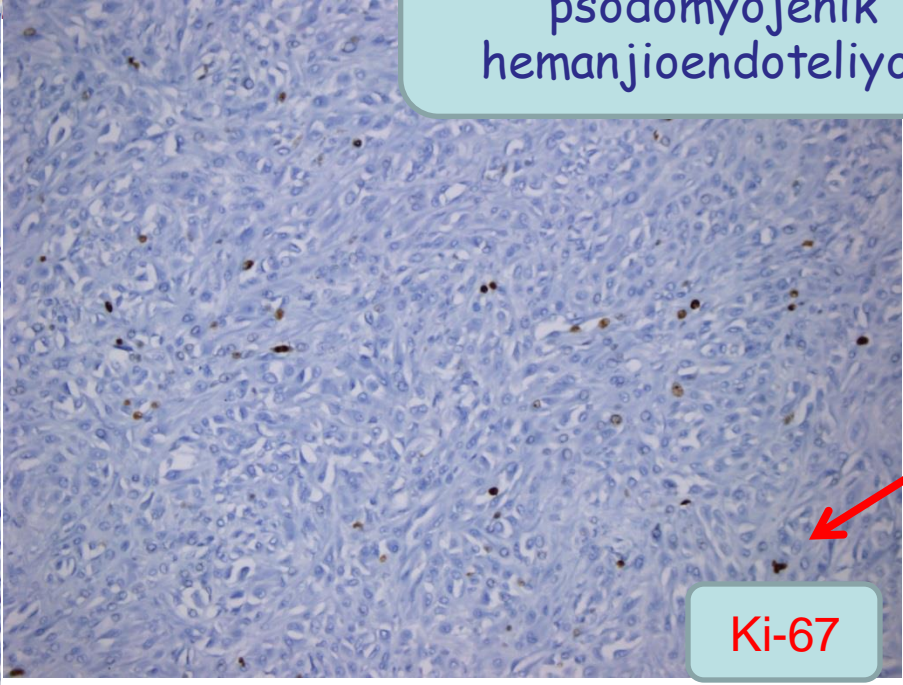


CD31

Epiteloid sarkom benzeri psödomyojenik hemanjioendoteliyom

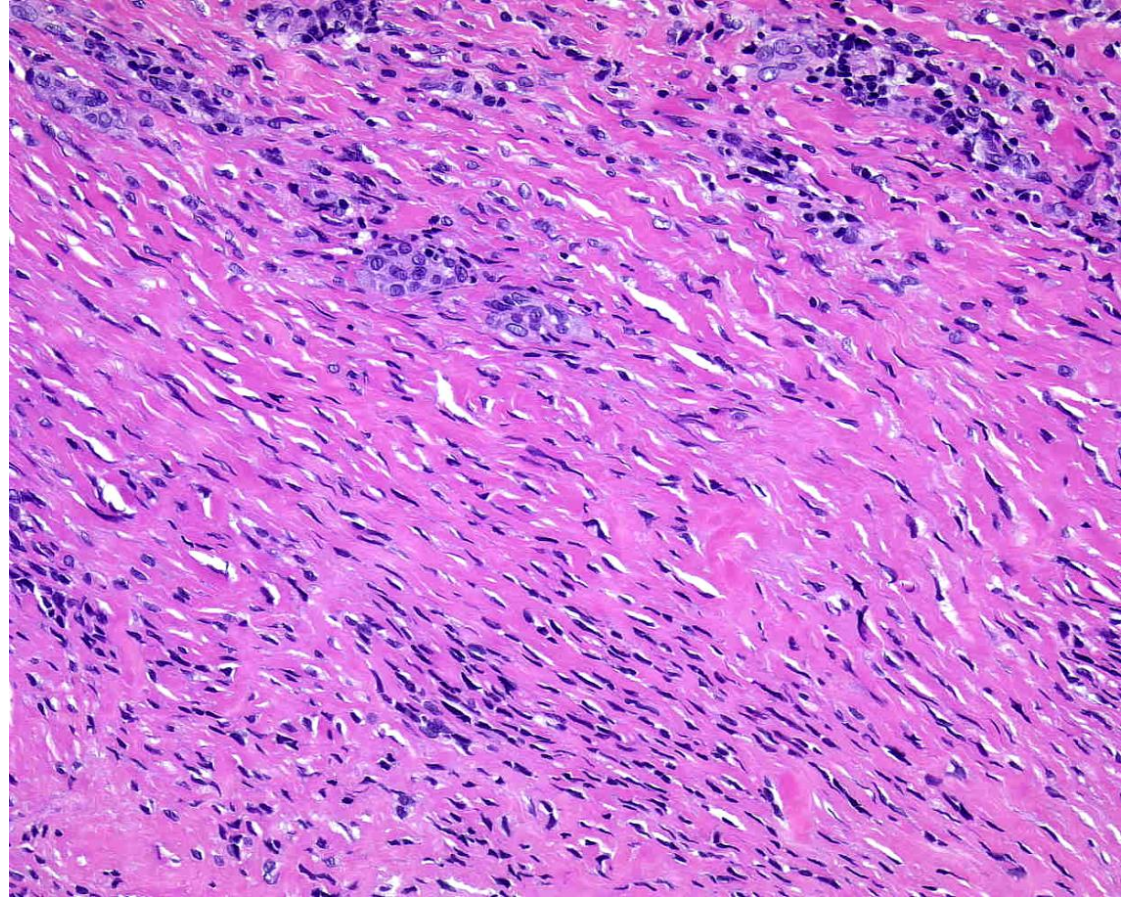


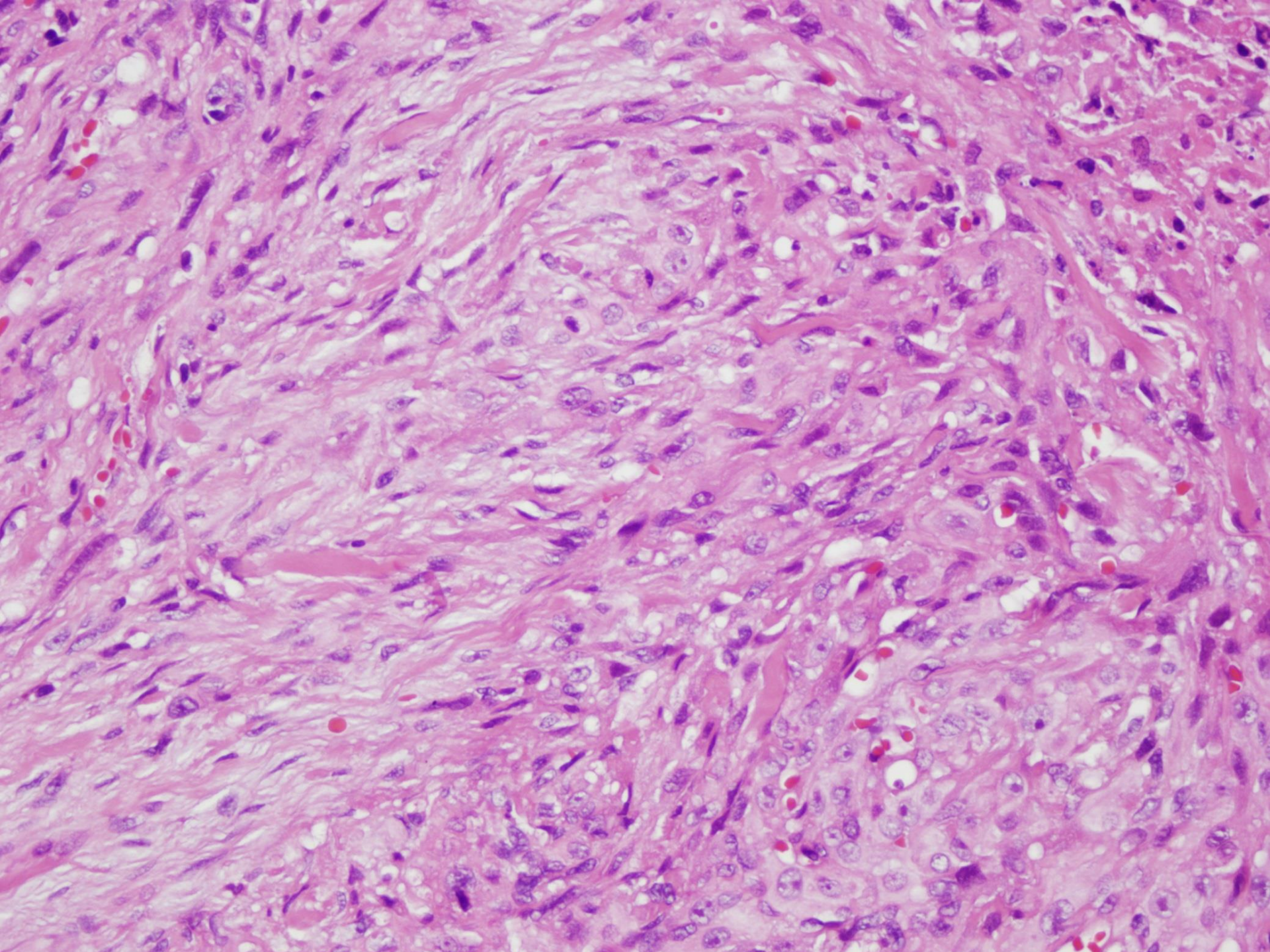
Fli-1

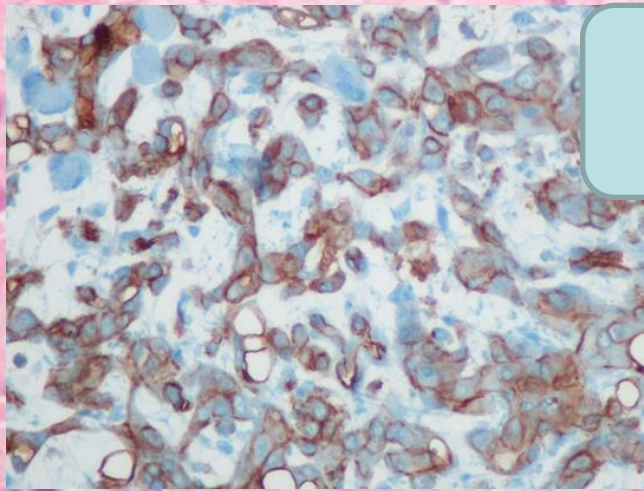
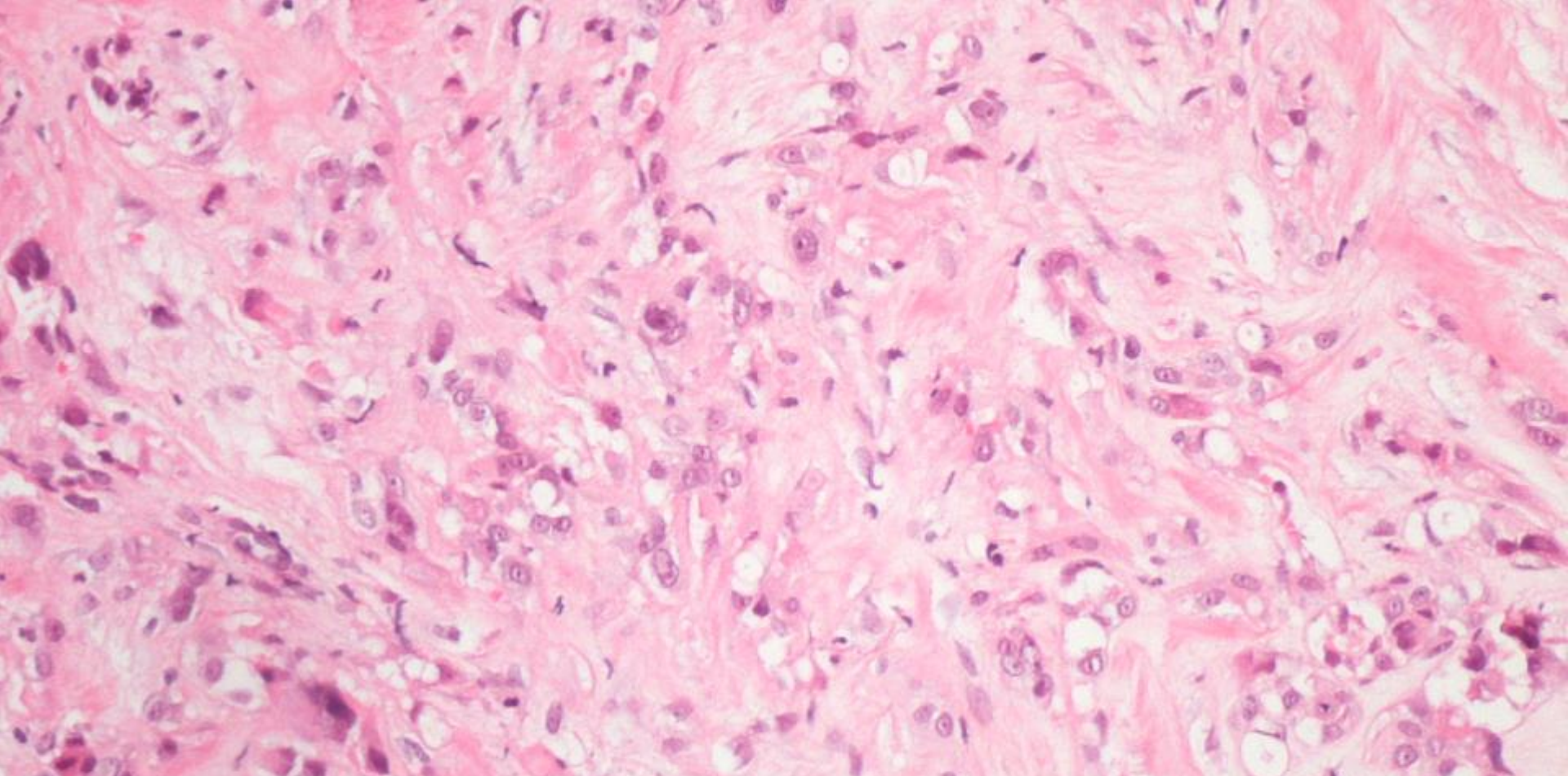


Ki-67

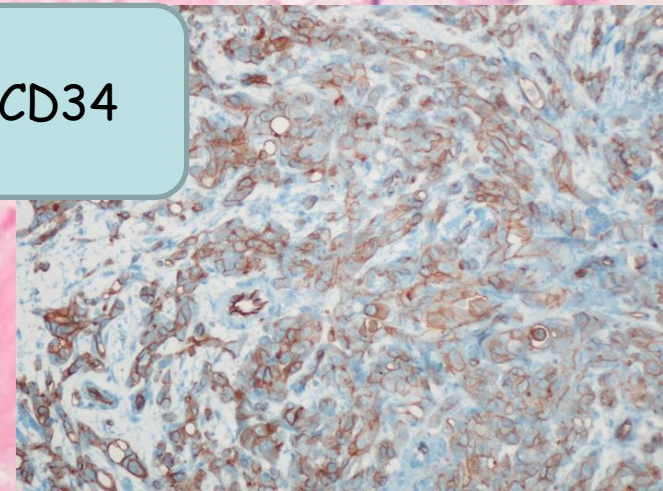
- Fibrom benzeri tip ięsi hücreler baskın
- Hücresel dermatofibroma benzer

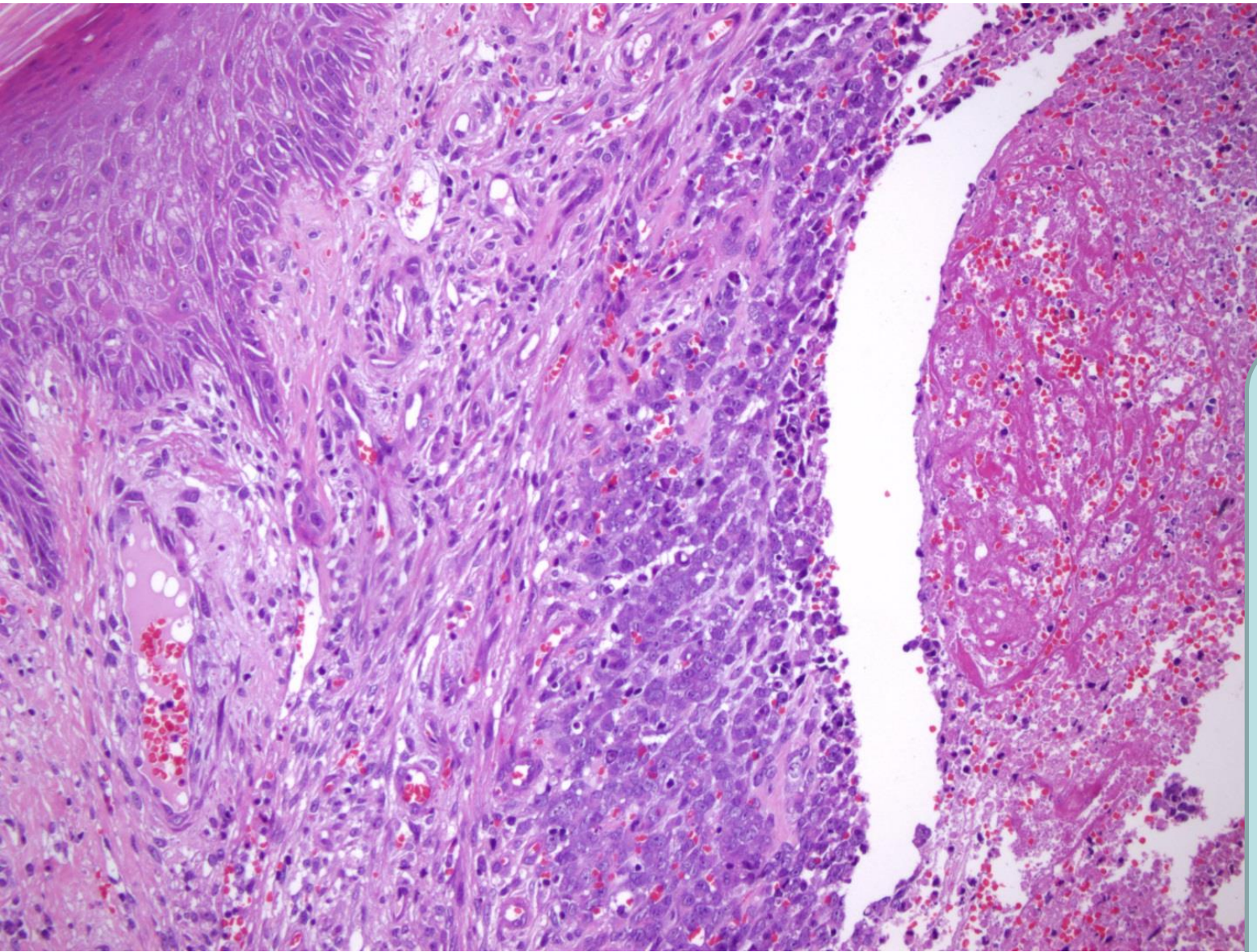






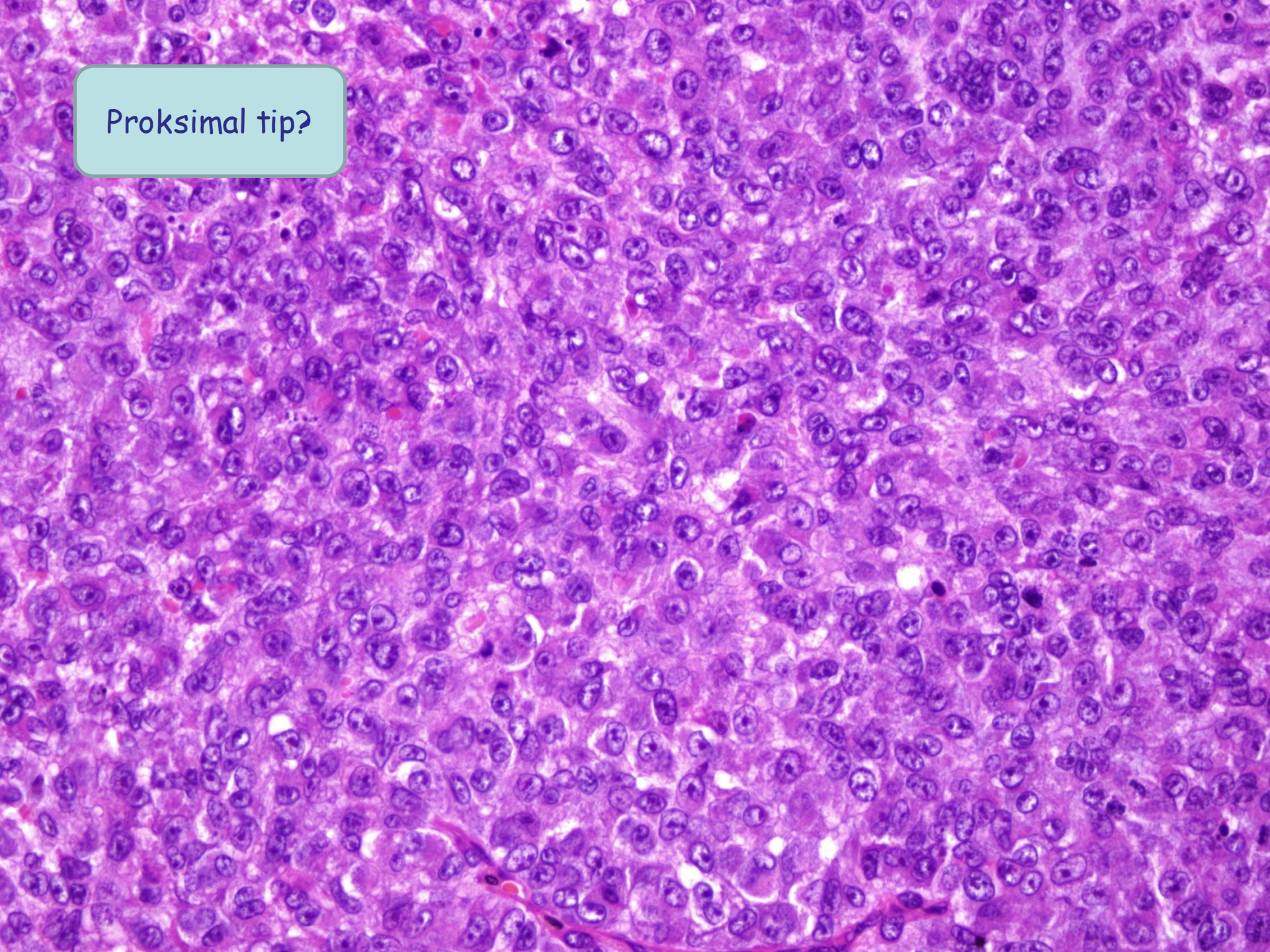
CD34





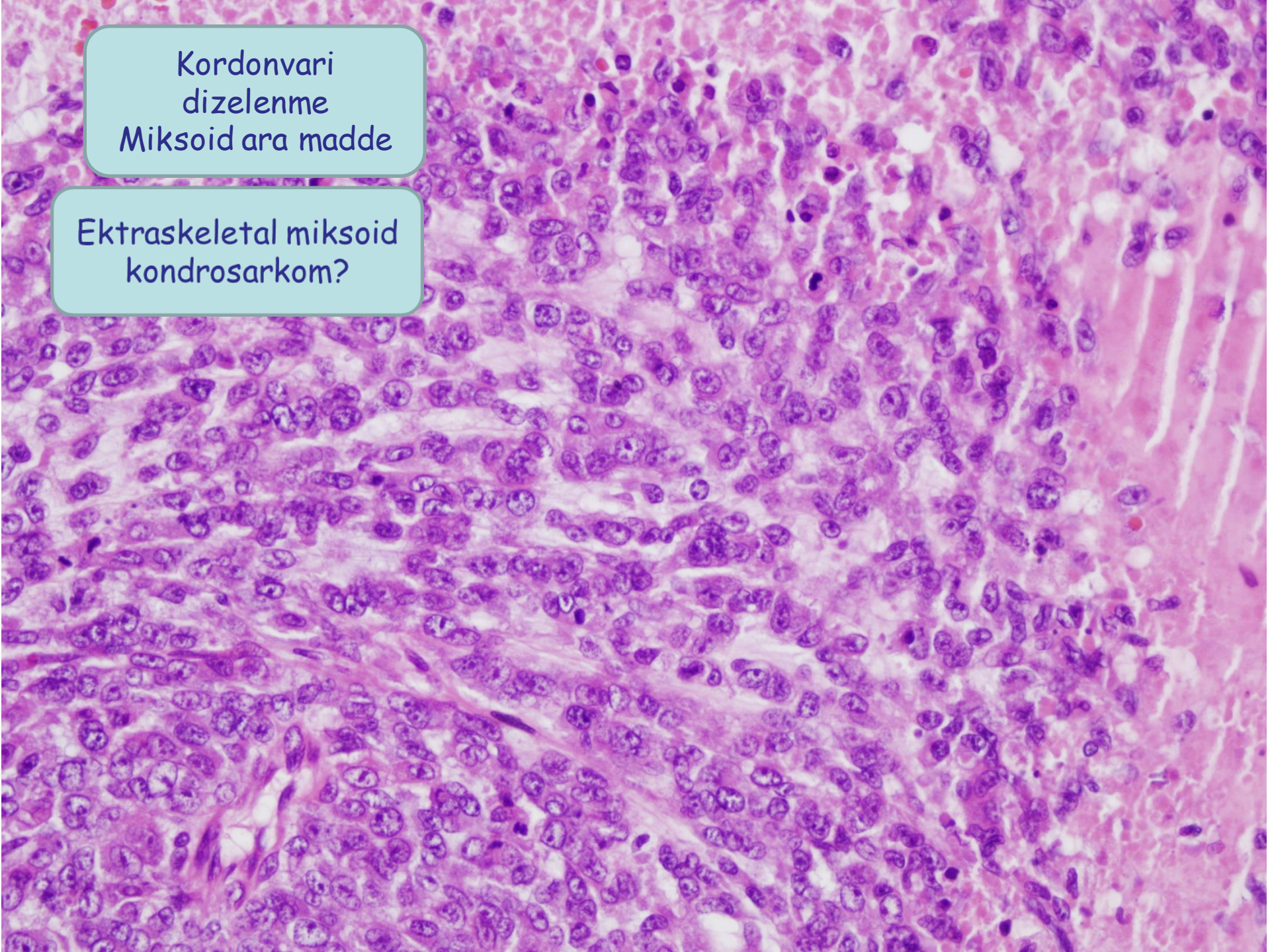
45E
Tibia üzeri
cilt ciltaltı
yumuşak
doku kitlesi
Epiteloid
Sarkom tanılı

Proksimal tip?

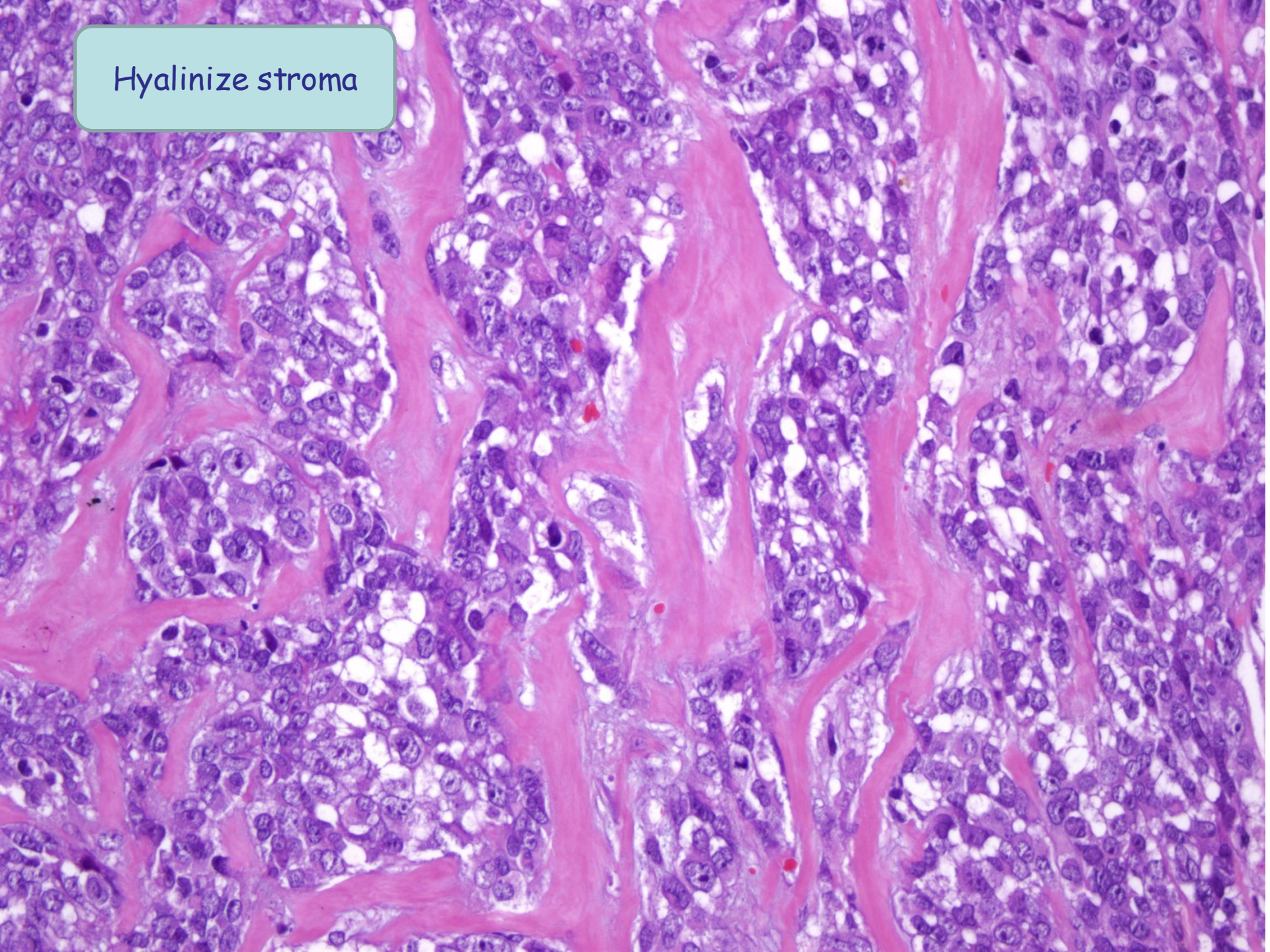


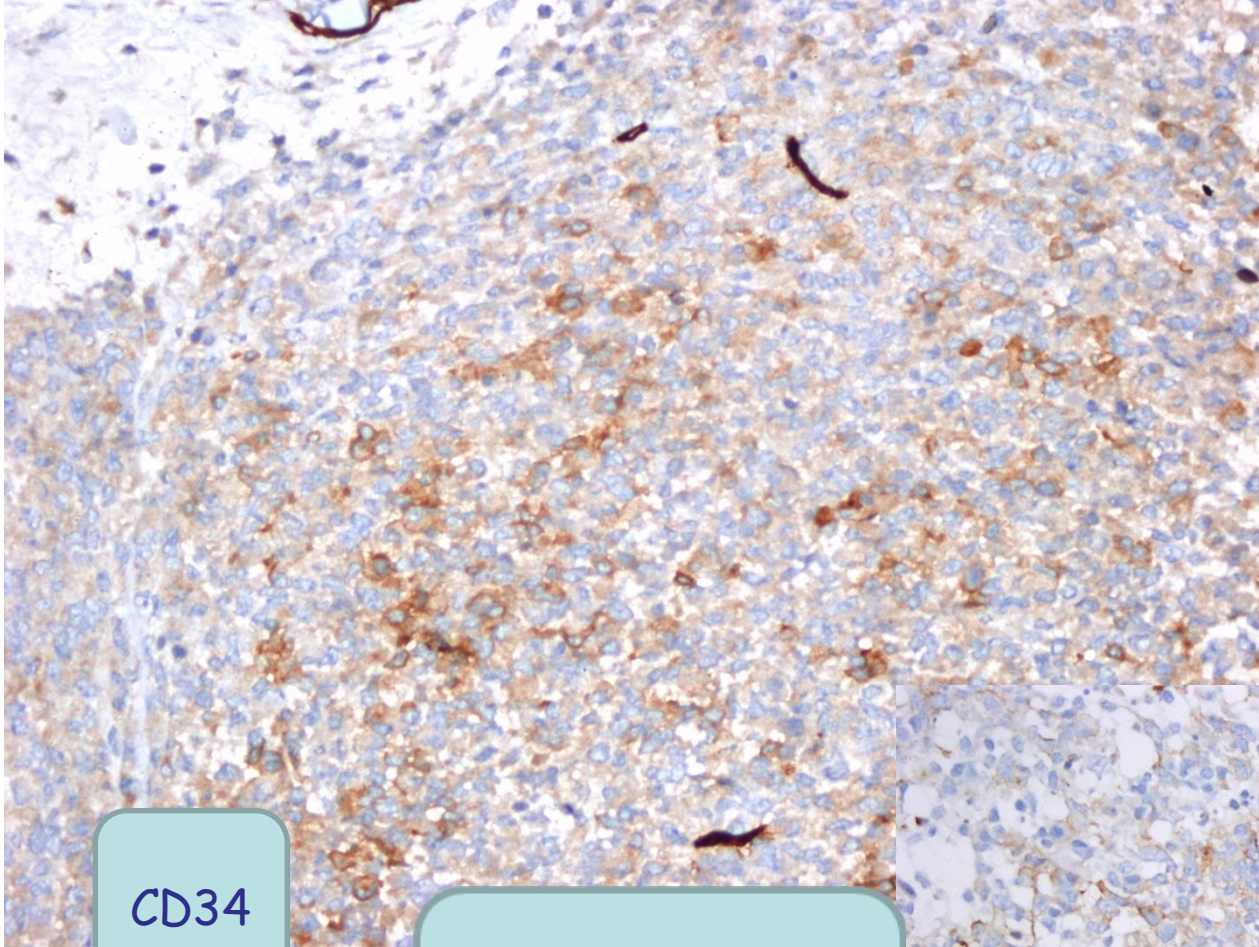
Kordonvari
dizelenme
Miksoid ara madde

Ekstraskeletal miksoid
kondrosarkom?



Hyalinize stroma



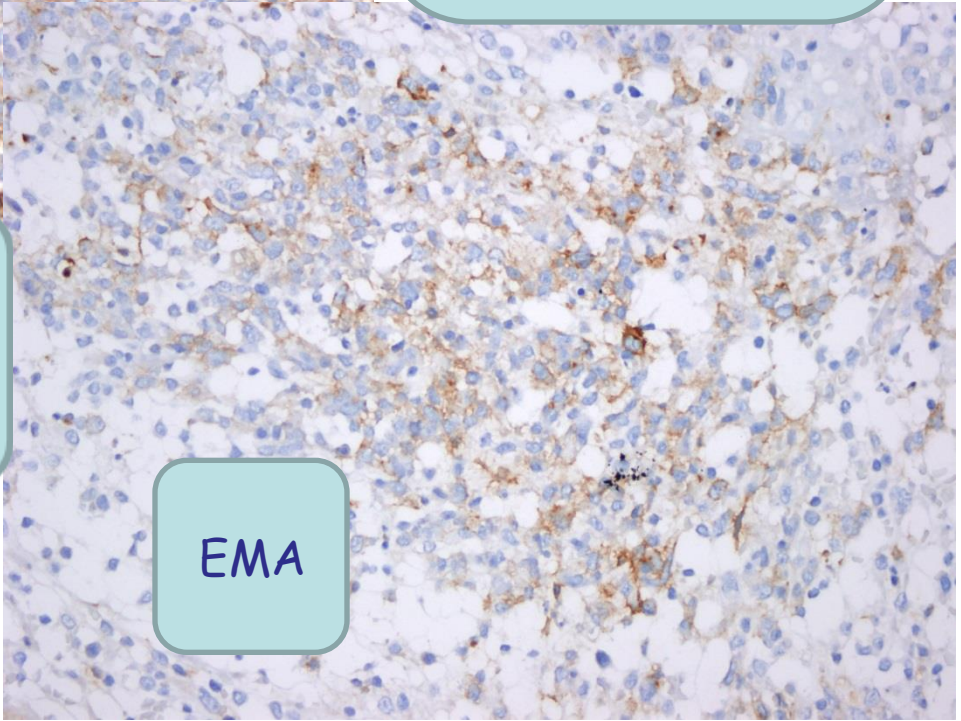


- HMB45
- Desmin
- PanCK
- S100
- CK20
- P63
- CD30
- CyclinD1
- SMA

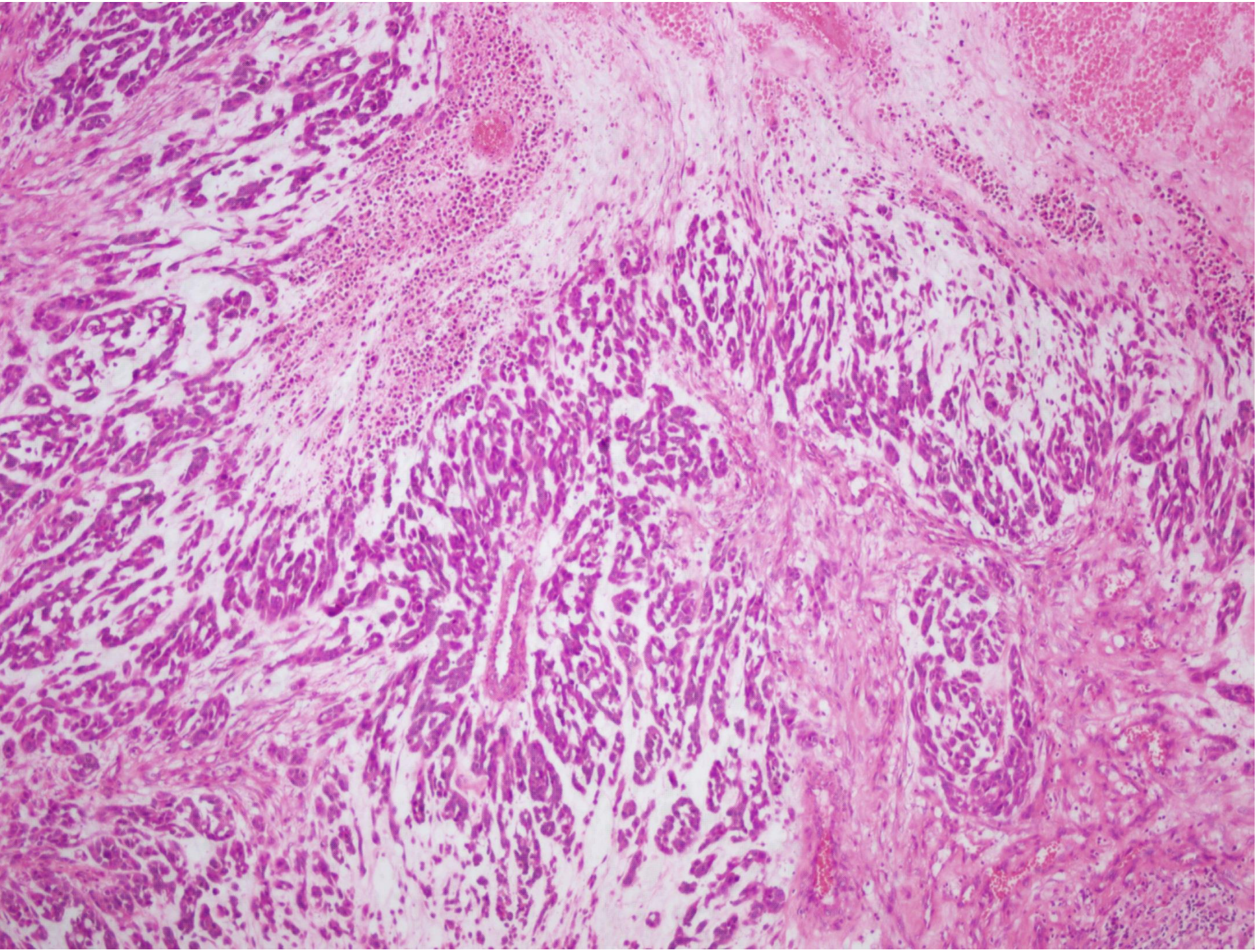
CD34

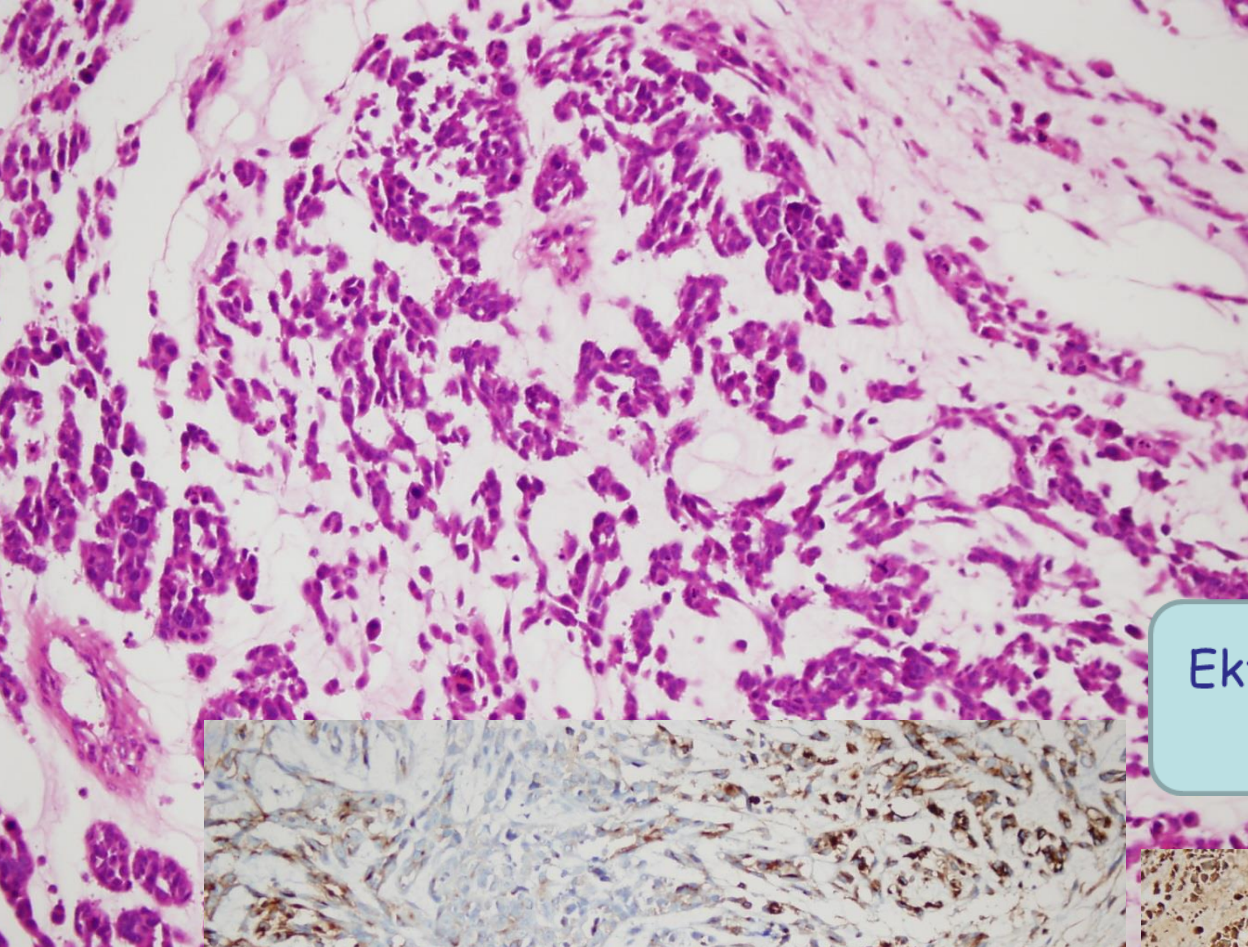
Miksoid varyant??

İHK epiteloid sarkom yönünde

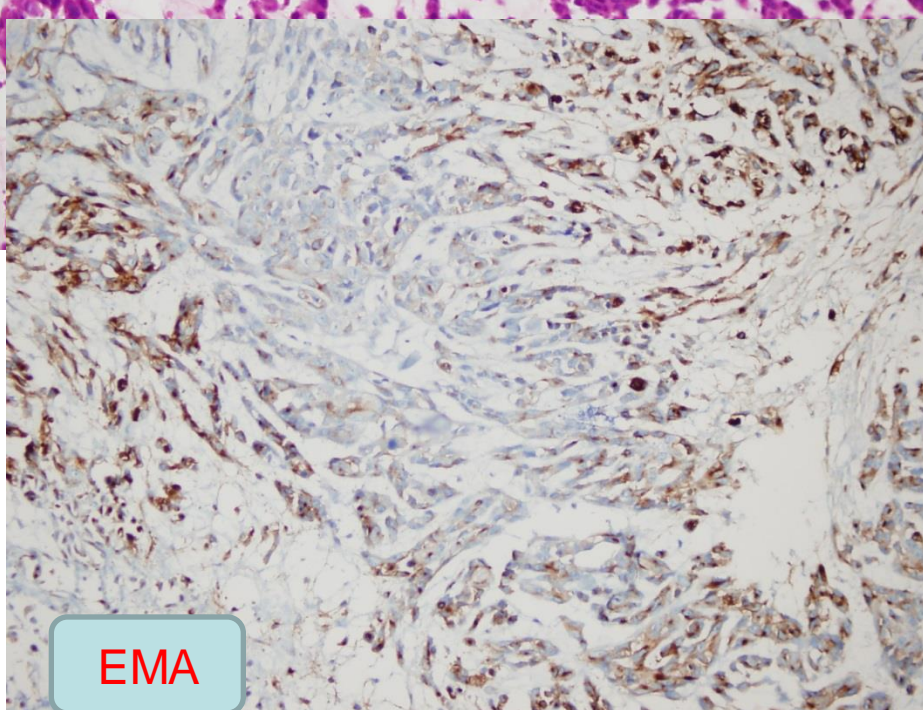


EMA

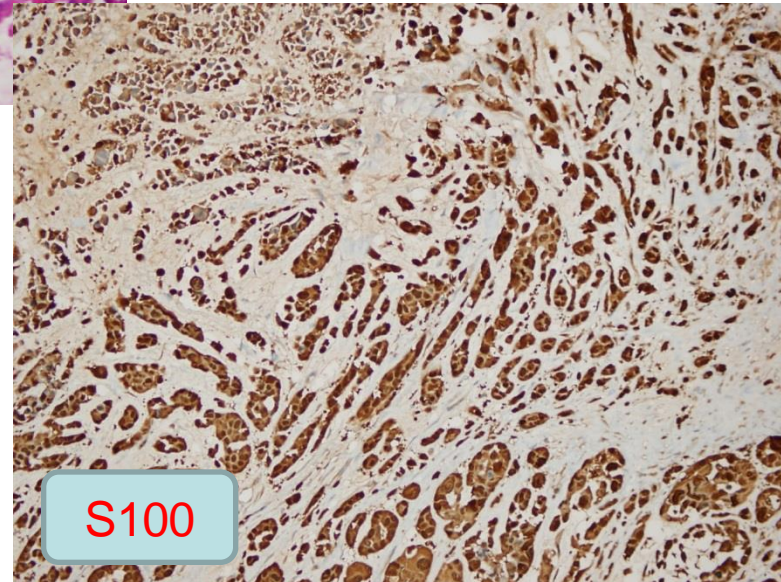




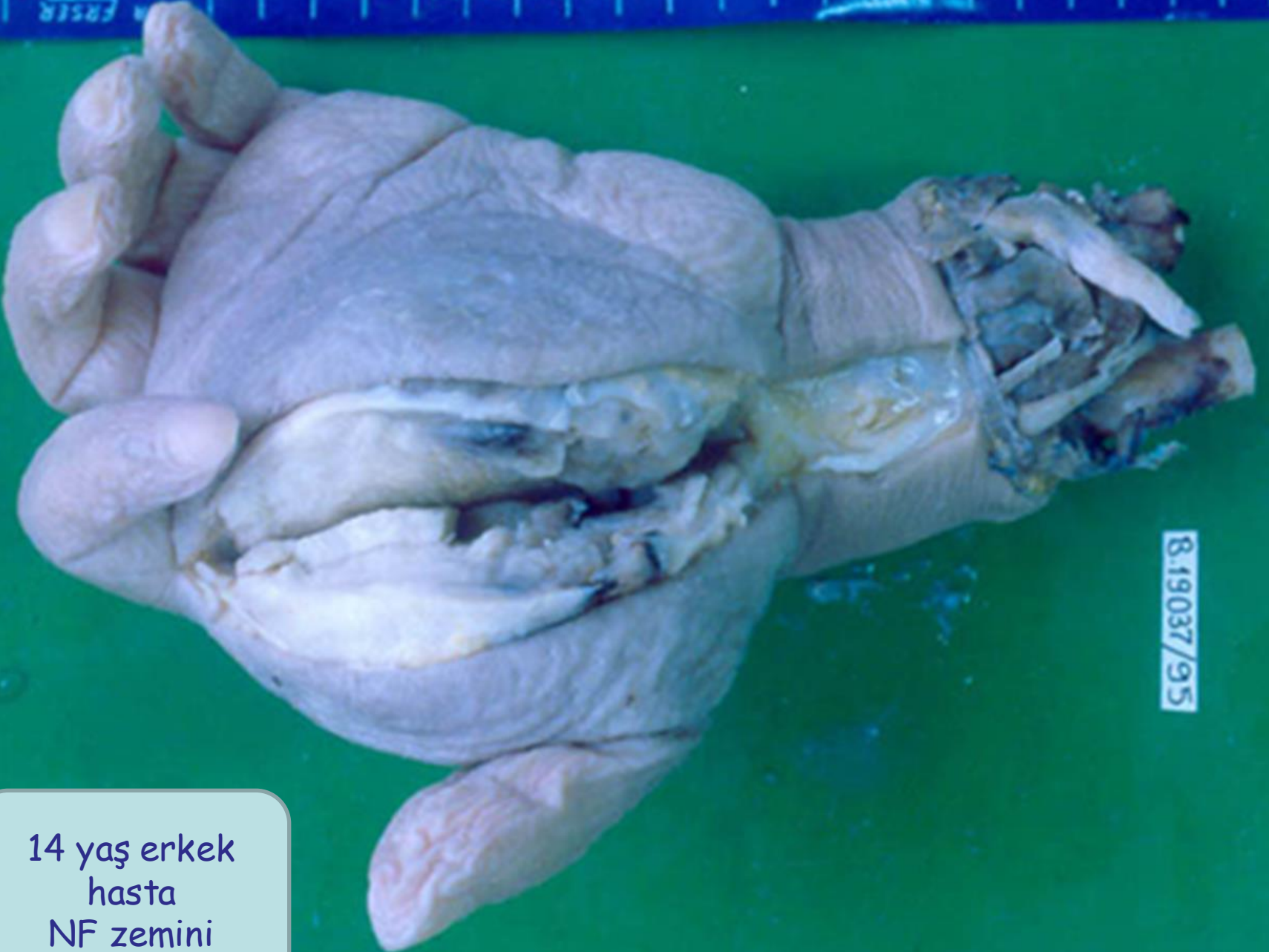
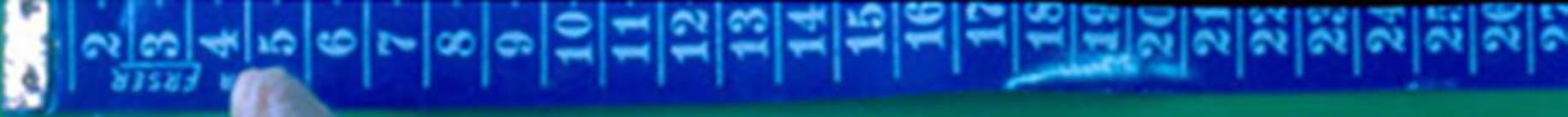
Ekstraskeletal miksoid kondrosarkom?



EMA

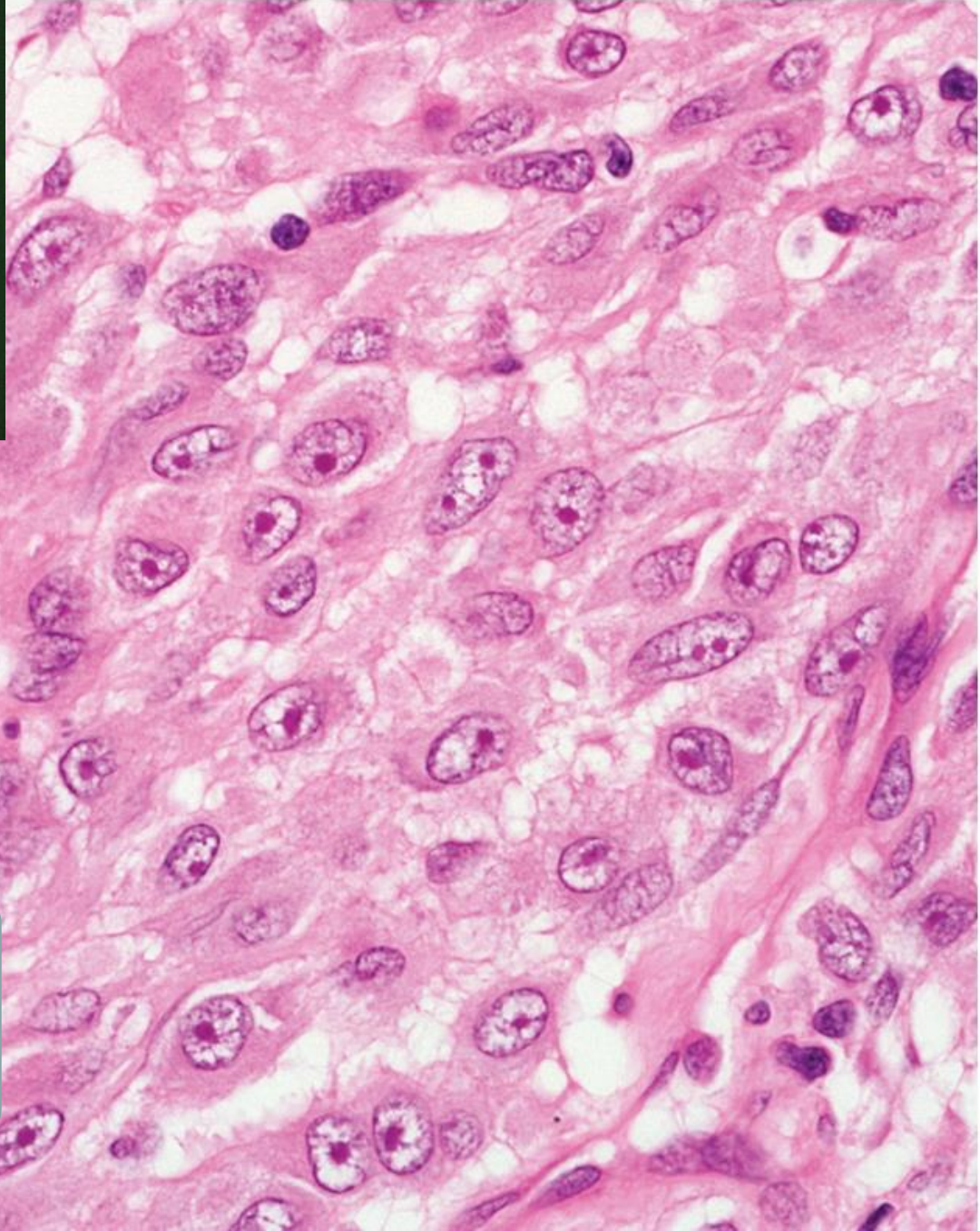


S100



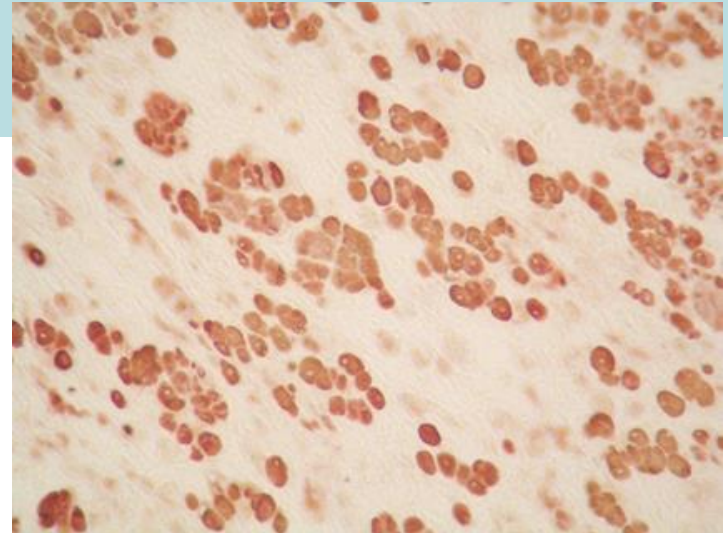
8.19037/95

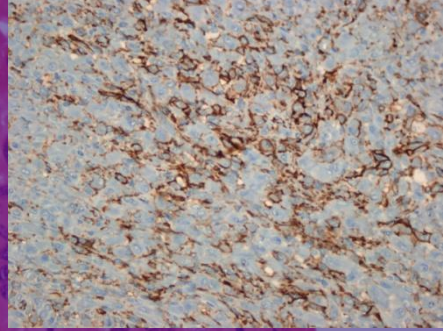
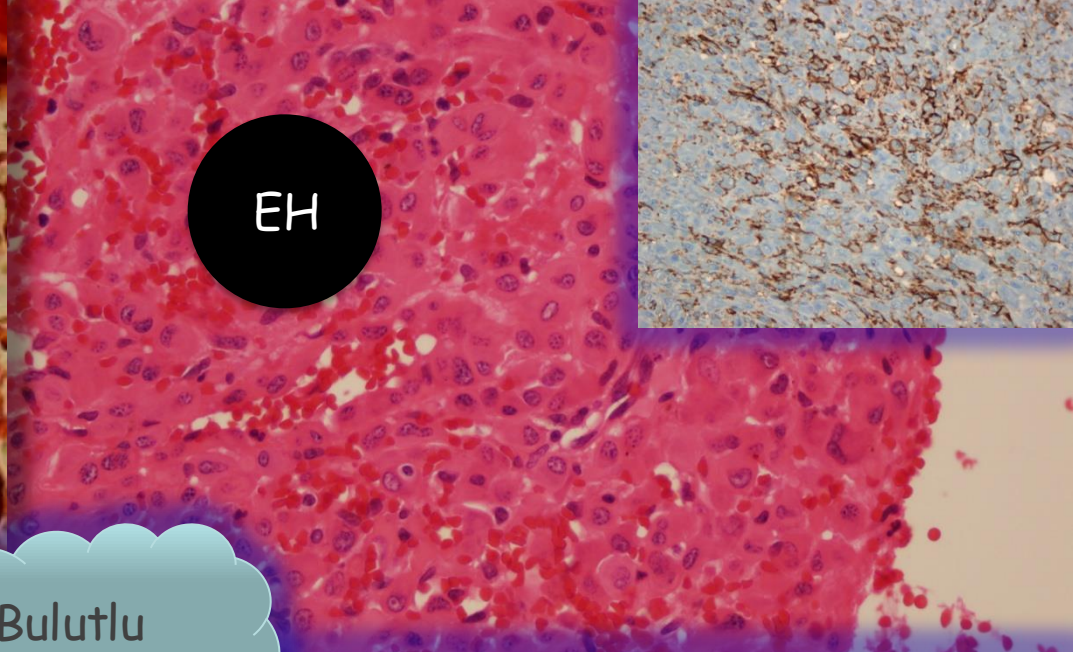
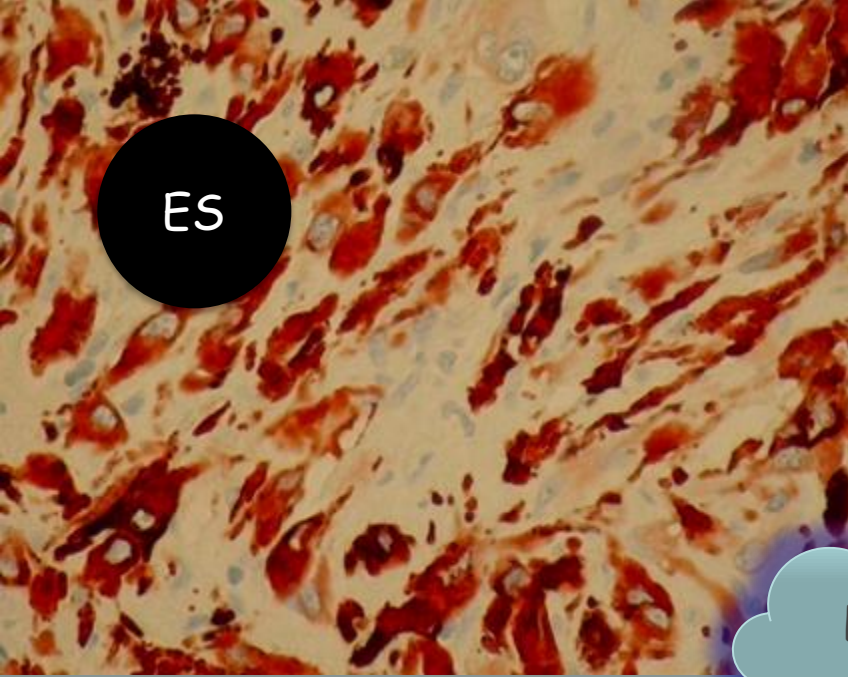
14 yaş erkek
hasta
NF zemini



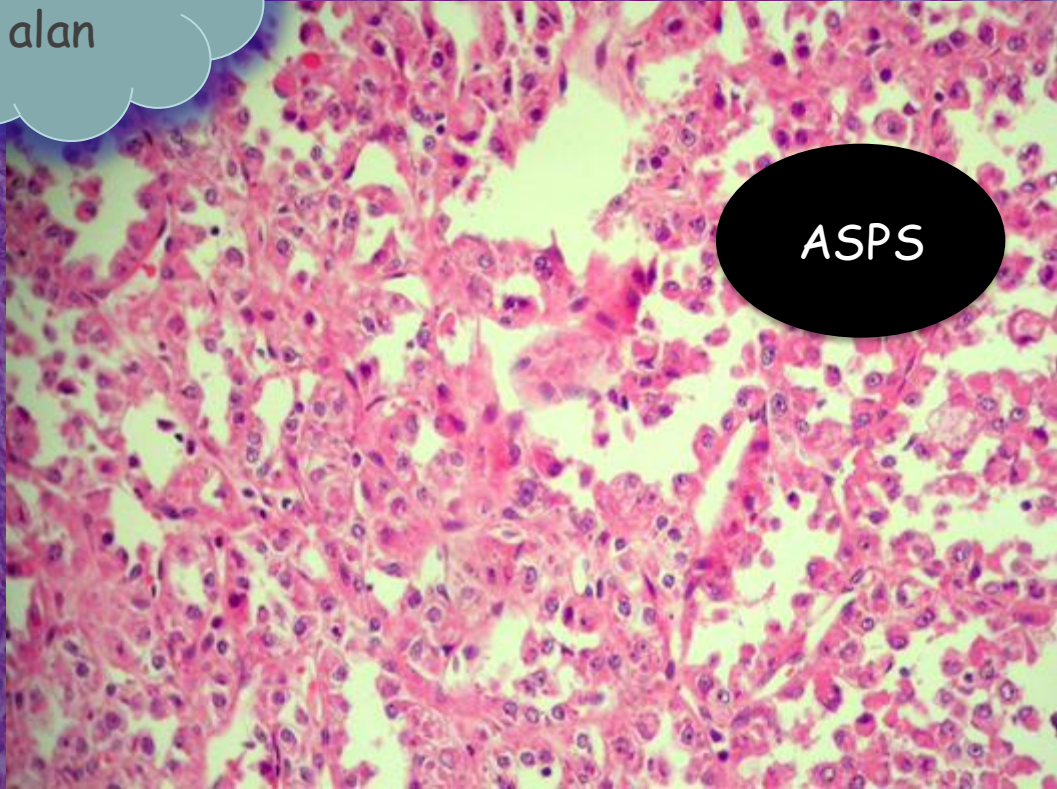
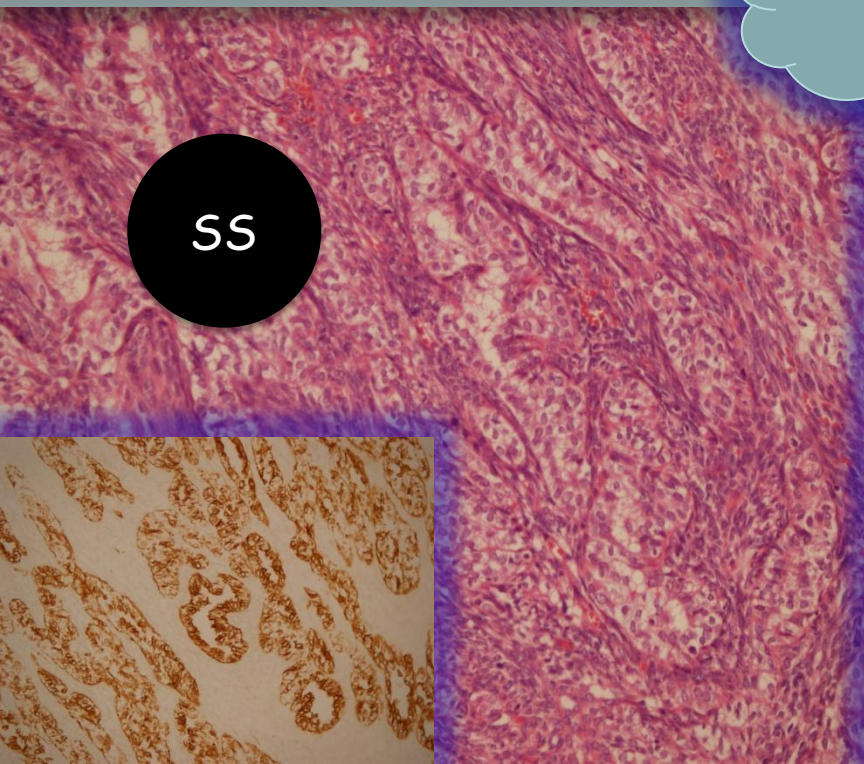
Malign Periferik Sinir Kılıf
Tümörü/Epiteloid Tip
Epiteloid sarkomla karışabilir!!

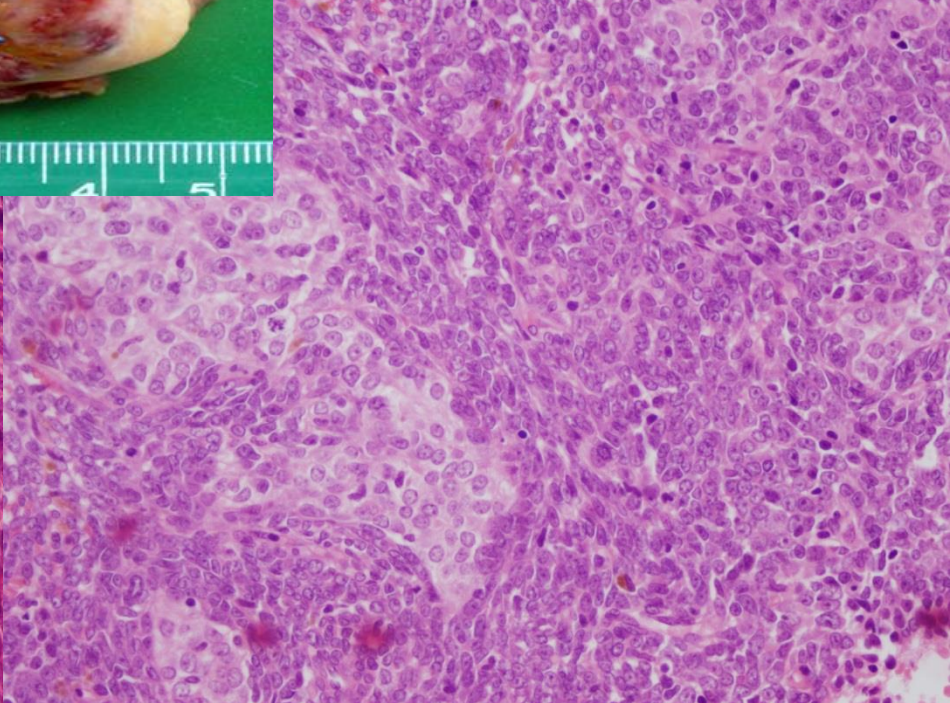
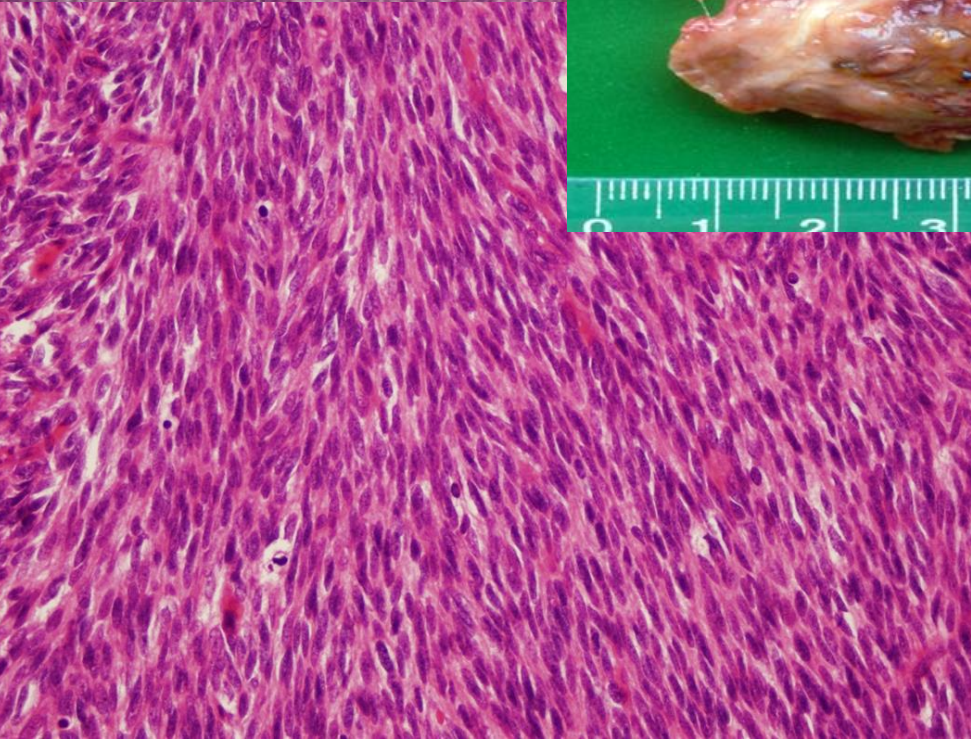
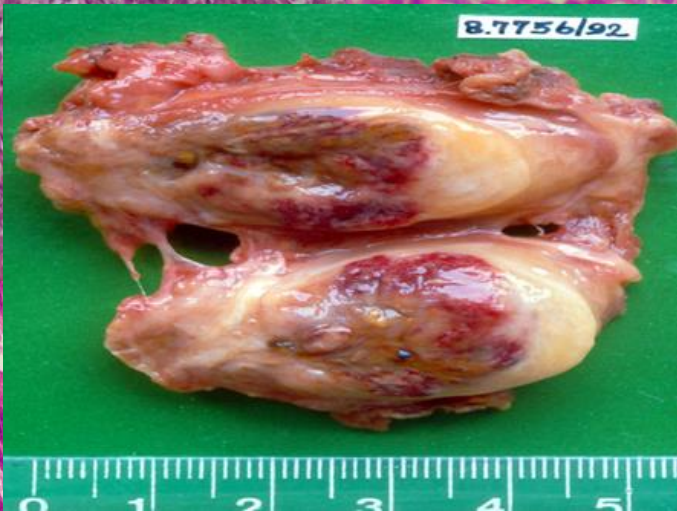
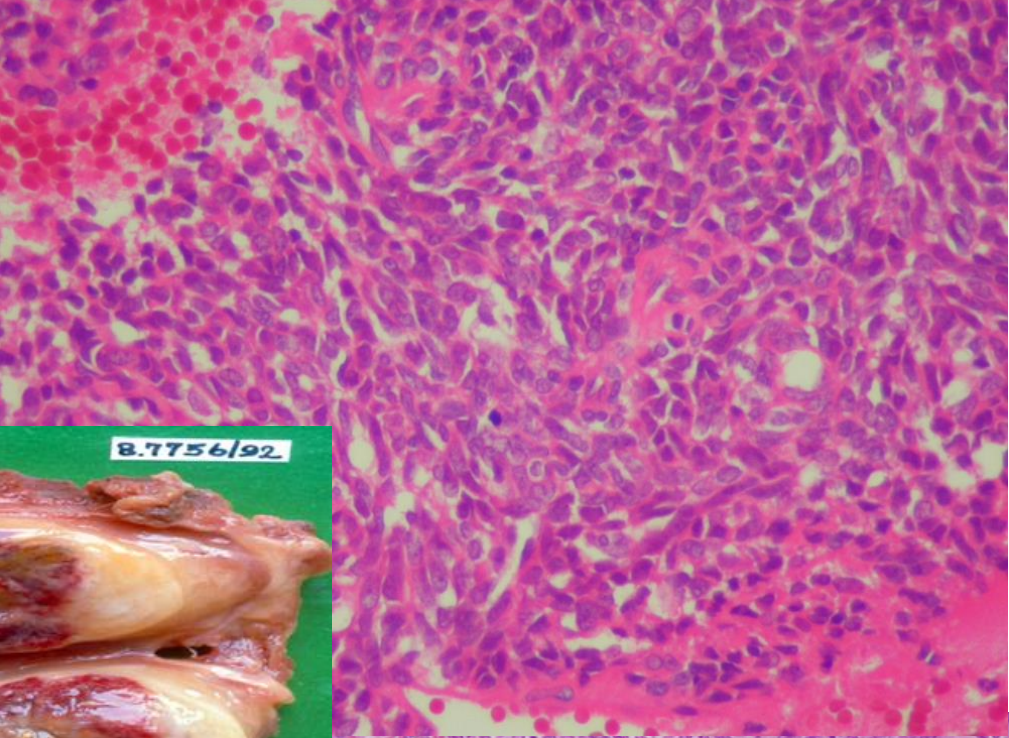
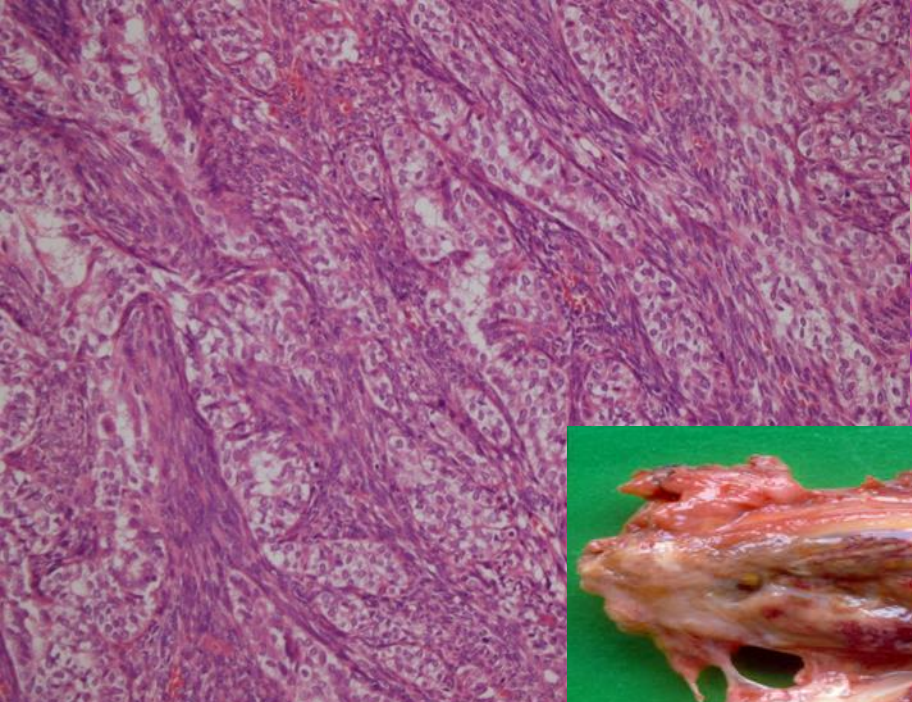
- Genç erişkin, uyluk ve üst ekstremitelerde yerleşimli, sıklıkla periferik bir sinirle ilişkili
- NF-1 ile ilişki zayıf
- Yüzeyel tümörlerin prognozu daha iyi
- S-100 proteini, epiteloid olmayan MPNST'ün aksine kuvvetli pozitif

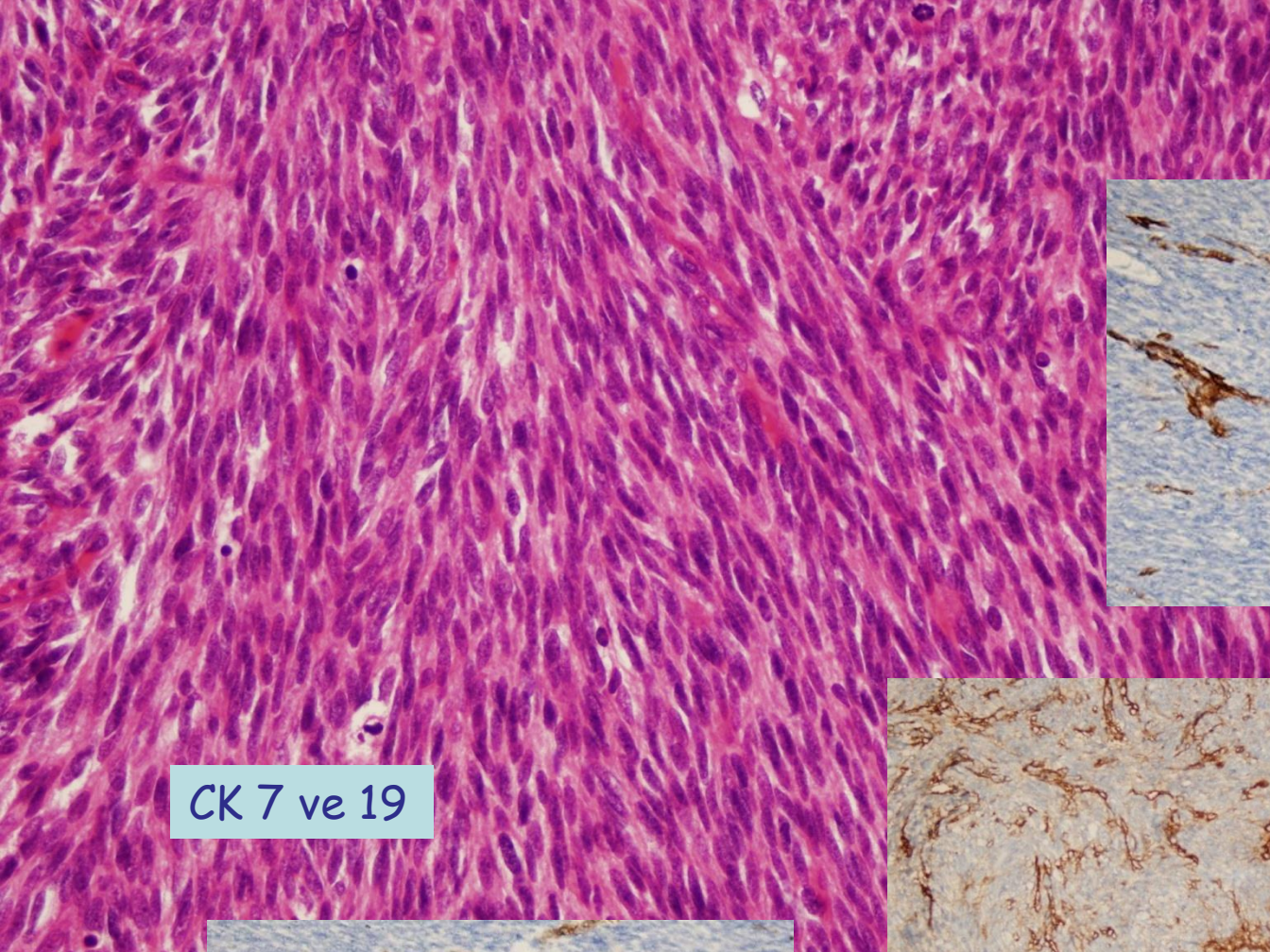




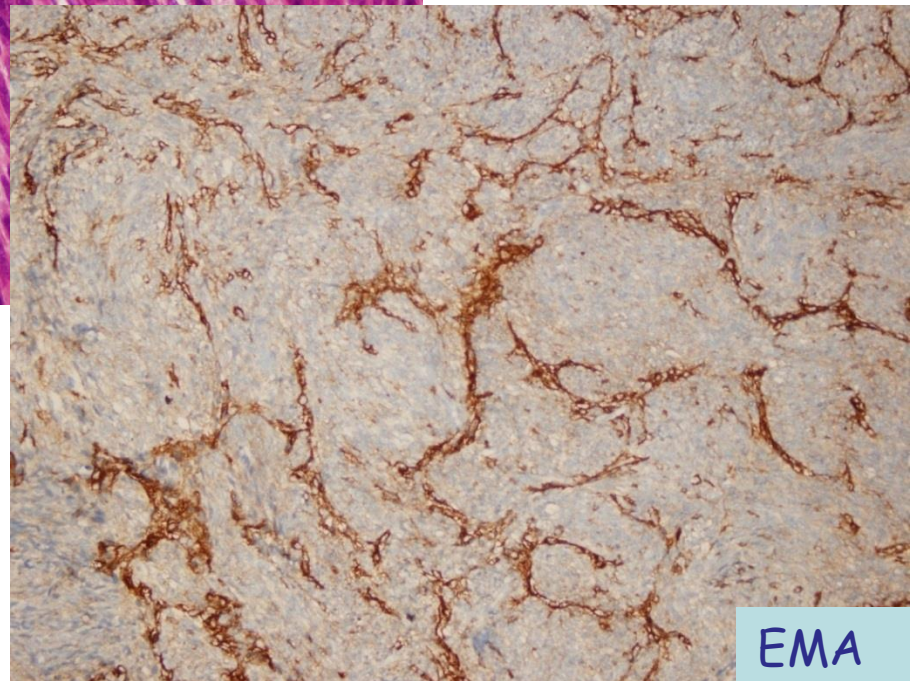
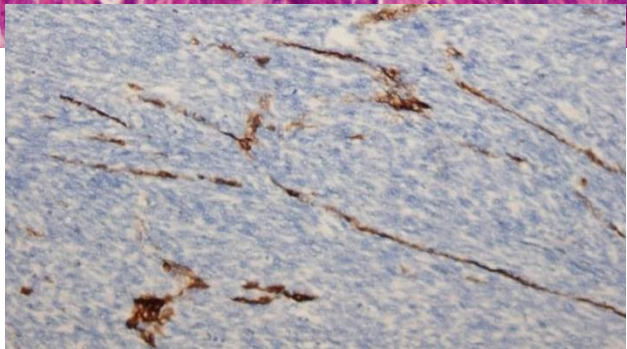
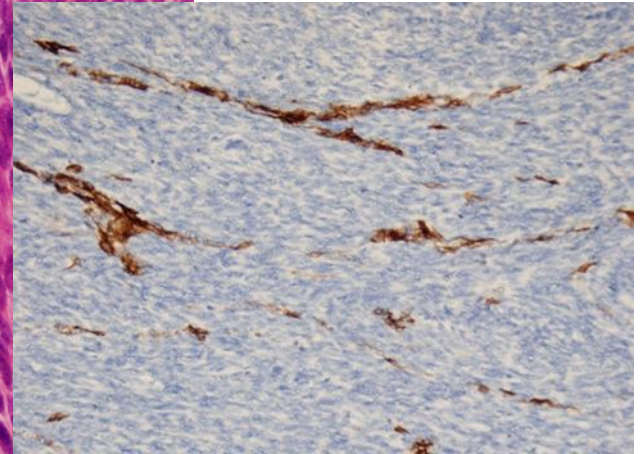
Bulutlu alan







CK 7 ve 19



EMA

Epiteloid yumuřak doku tmrleri

- Alveoler soft part sarkom
- Epiteloid sarkom
- Epiteloid anjiosarkom
- Epiteloid hemangioendoteliom
- Epiteloid hemangiom
- Ekstragastrointestinal stromal tmr
- MPNST epiteloid tip
- Epitelioid schwannom
- Malign rabdoid tmr
- Malign mezotelyoma
- Sinovyal sarkom/bifazik ve epiteloid baskın tip

ncelikle
metastatik
karsinom , melanom
ve byk hcreli
lenfoma dıřlanmalı
İmmunhistokimya

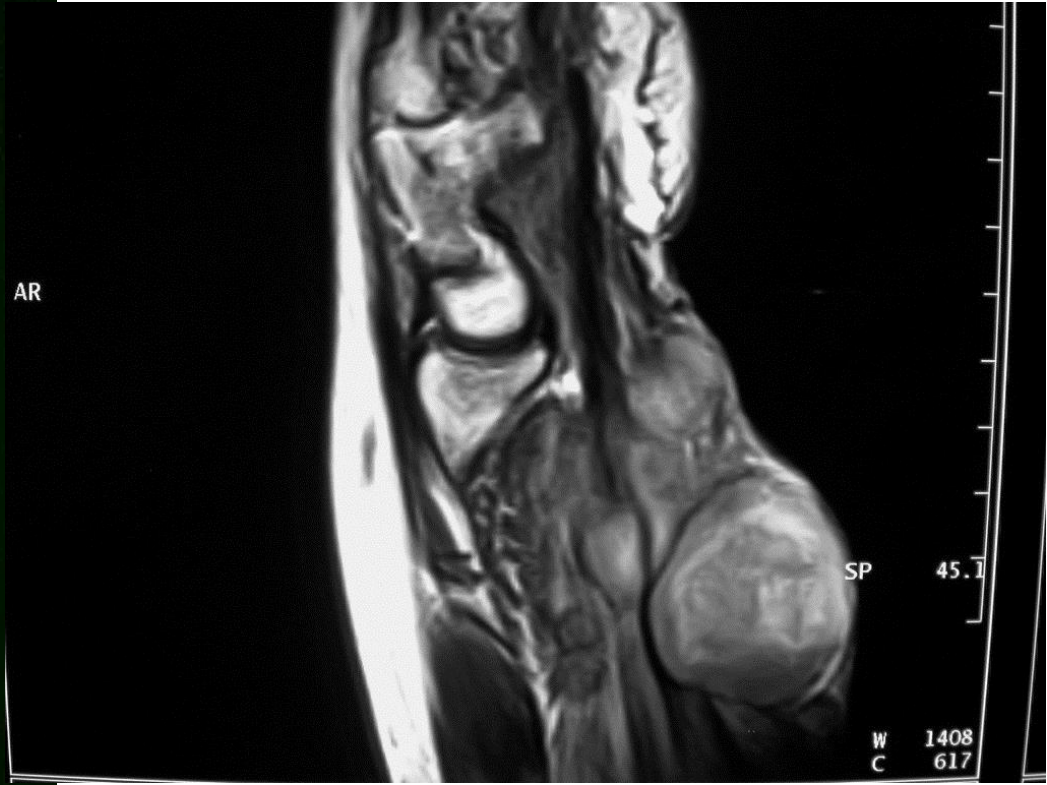
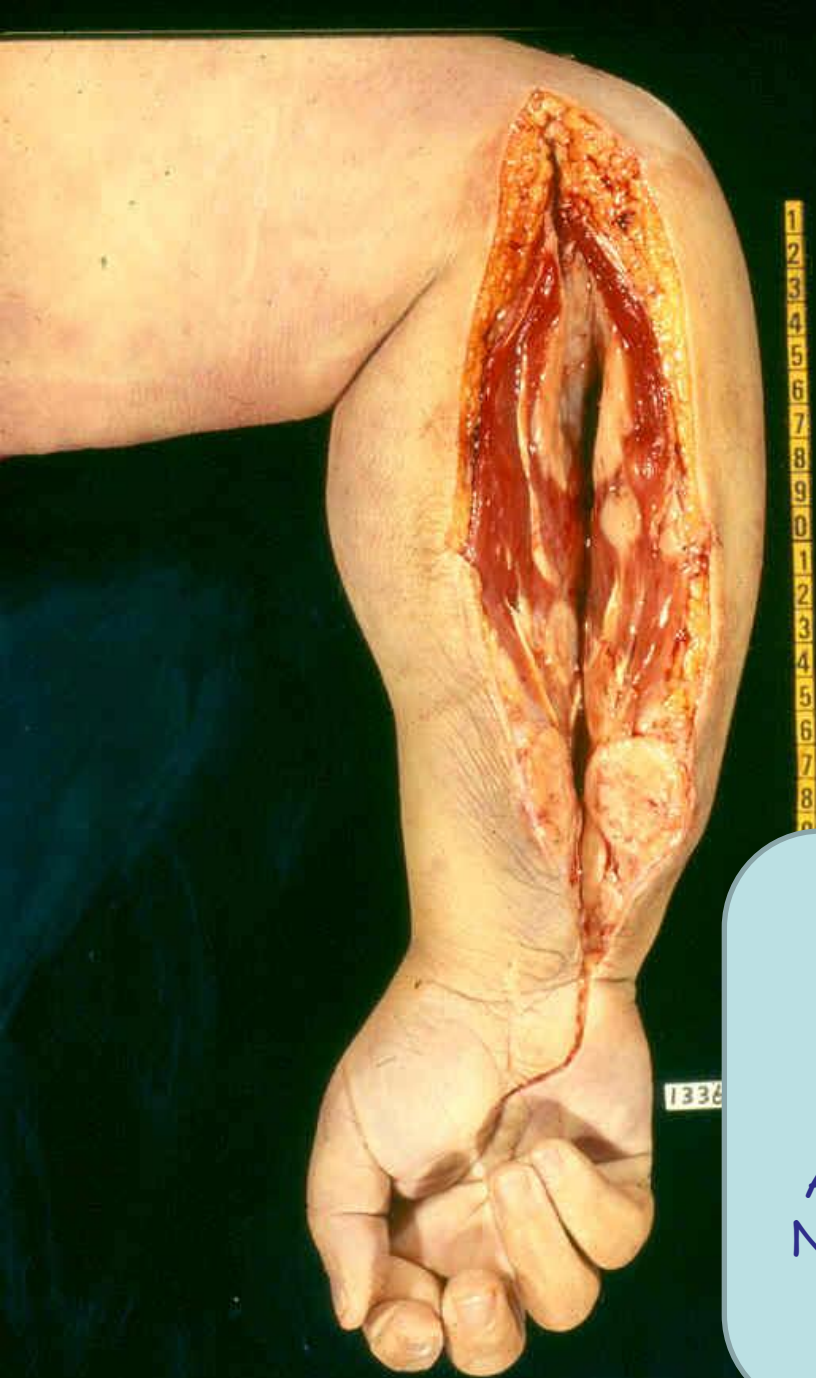
EPİTELOİD SARKOMDA AYIRICI TANIYA GİREN BAŞLICA ANTİTELER

| ANTİTE | KLİNİK | MİKROSKOPI | İMMUNHİSTOKİMYA | MOLEKÜLER |
|--|--|---|---|---|
| Sinoviyal sarkom | Genç erişkin, derin yerleşim, eklemler çevresi ekstremitelerde, iç organlar dahil her yerden çıkabilir | Bifazik patern , epitelial ve fibröz içsi ve az diferansiye alanlar, mast hücreleri | SK7,19+, EMA+, Tle1+, Bcl2 +, CD99+/- | t(X;18)(p11.2;q11.2) SS18-SSX1 SS18-SSX2 SS18-SSX4 füzyonları |
| Epiteloid anjiosarkom | Erişkin, derin yerleşim | Poligonal hücreler, solid üreme, fokal intrasitoplazmik lümen oluşumu | CD31+, CD34+, FLI1+, fokal SK+ ve EMA+, D2 40 +/-, nükleer INI1 kaybı yok | |
| Epiteloid hemanjioendoteliyom | Erişkin, derin yerleşim, karaciğer ve akciğer | Kondromiksoid zemin, kordon ya da yuvalar halinde hücreler, intrasitoplazmik lümen | CD31+, CD34+, FLI1+, bazen SK+, nükleer INI1 kaybı yok | t(,3)(p36.3;q25) WWTR1- CAMTA1 füzyonu %85 |
| Epiteloid sarkom benzeri hemanjioendoteliyom | Genç erişkin, gövde ve ekstremitelerde, subkutan, yavaş gidişli | Desmoplastik stroma, eozinofilik hücre demetleri, vasküler formasyon yok. | SK+, CD31+, FLI1+, CD34-, nükleer INI1 kaybı yok | t(7;9)(q22;q13) |
| Karsinom/ skuamöz hücreli/ deri eki tümörleri | Yerleşim iç organ ya da deri, metastatik odaklar | Epiteloid hücre kordon ve tabakaları, bez halkaları | SK5-6, P63, nükleer INI1 kaybı yok Organ kökenine özgü belirteçler+ | |
| Melanom | Primer ya da metastatik | İri epiteloid, pleomorfik hücre tabakaları, belirgin nükleol, rabdoid eozinofilik sitoplazma, | S100+, melanA+, HMB45+, INI1+, nadir SK pzitifliği | |
| Malign rabdoid tümör | Çocuk, genç erişkin, | Multinodüler, poligonal hücre | SK+, EMA+, CD34; INI1- | <i>SMARCB1</i> (<i>INI1</i>) kaybı |





Ön kol
Multifokal
Sivilce gibi

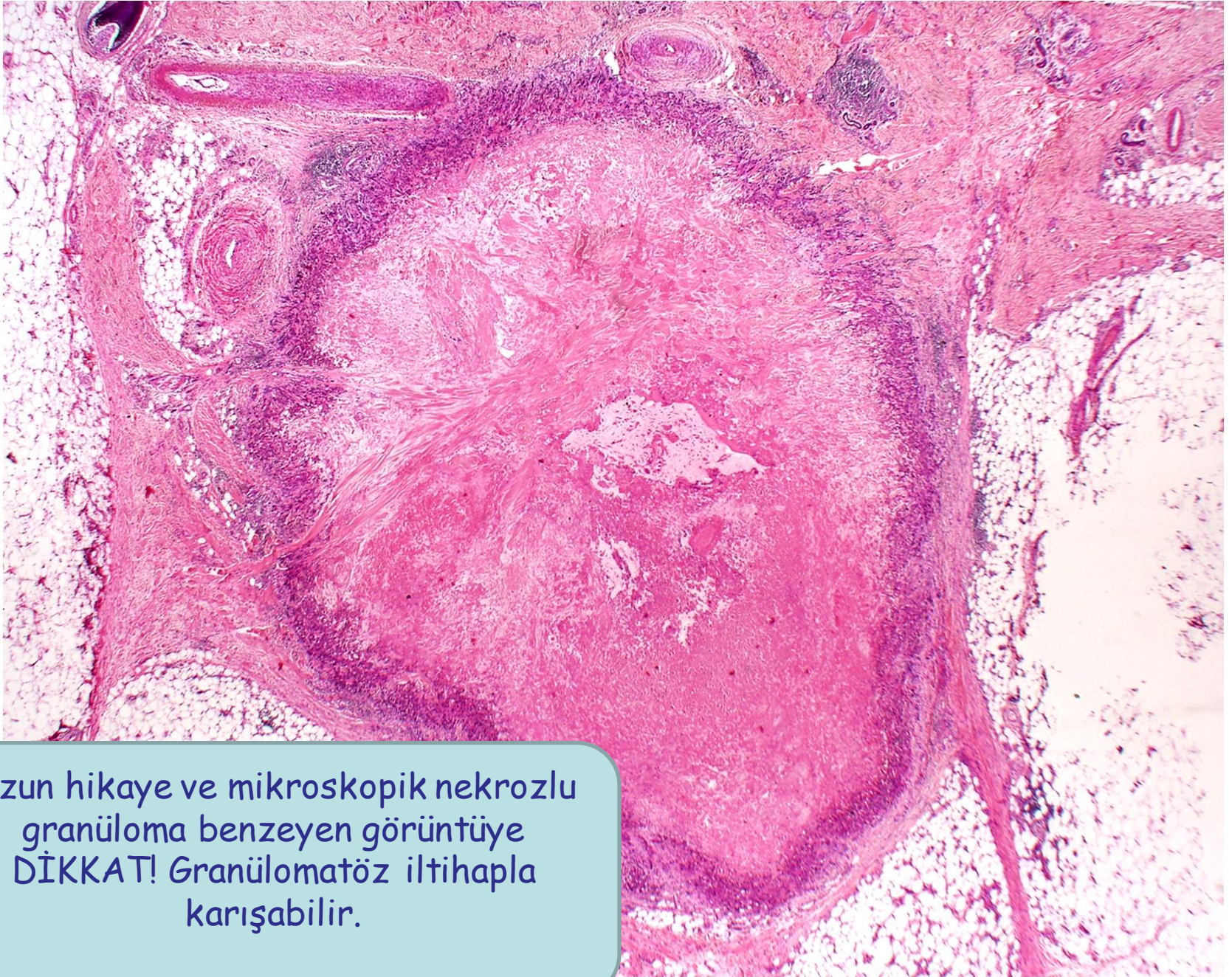


En sık yerleşim el ve önkolda şişlik
MULTINODÜLER
SINIRLAR BELİRSİZ

Ortalama 2 senedir devam eden semptomlar
Alt ekstremitede ayak, bilek ve pretibial bölge
Nadiren perine, vulva, göğüs duvarı, aksilla, baş-
saçlı deri

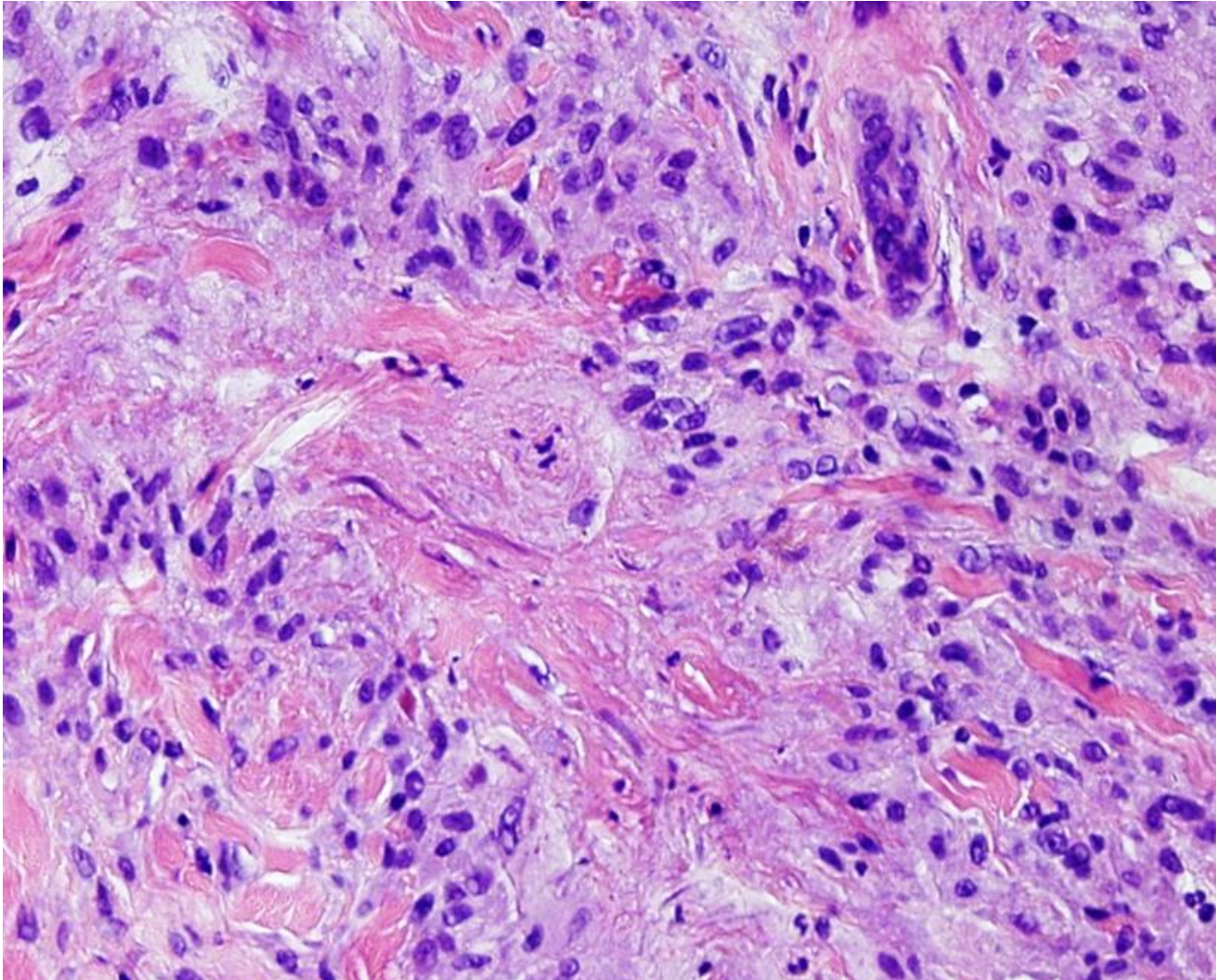
Akılda tutulması gerekenler

- Genç erişkin hasta
- Parmak, önkol yerleşimli kitle (uzun hikaye)
- Eozinofilik epiteloid hücreler, iğsi hücreler, nekroz
- PSK, EMA, CD34



Uzun hikaye ve mikroskopik nekrozlu
granüloma benzeyen görüntüye
DİKKAT! Granülomatöz iltihapla
karışabilir.

Ne klinisyen ne biz tümöre
benzetemeyebiliriz **DİKKAT**



Proksimal tip(büyük hücreli)-Epiteloid sarkom

- Yerleşim ve morfoloji farkı
- İHK farkı: %50 desmin (+) (Yanlış RMS tanısına DİKKAT!)

Clinicopathological Review of Soft Tissue Sarcomas

