

# 21.Ulusal Patoloji Kongresi Yumuşak Doku Kursu Lipomatöz Tümörler



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DEÜTF Patoloji AD  
16-20 Kasım 2011-İZMİR

# Sunum planı

Lipomatöz tümör nedir? ve sınıflama

Sorun yaratan lipomlar

Liposarkomlarda tanı ve ayırıcı tanı

Tanısal zorluk nedenleri

# Lipomatöz tümör

Adipositlerden gelişen  
mezenkimal tümör

(YDT'nin %50'si)

En kompleks,  
en büyük tek grup

Lipom ya da  
liposarkom  
Her yaş, tüm  
anatomik  
lokalizasyon

DSÖ sınıflamasında  
12 benign  
1 intermediate  
5 malign

# Histolojik sınıflama

## Benign tümörler

Lipom (yüzeyel-  
derin)

Anjiolipom

İğsi h/Pleomorfik h  
lipom

Myolipom

AML

Myelolipom

Kondroid lipom

Hibernom

Lipoblastom/  
lipoblastomatozis

Lipomatozis

Sinir lipomatozisi

# Intermediate malignansi

Atipik lipomatöz tümör/

İyi diferansiye LS

\*lipoma-benzeri LS

\*sklerozan LS

\* İnflamatuvar LS

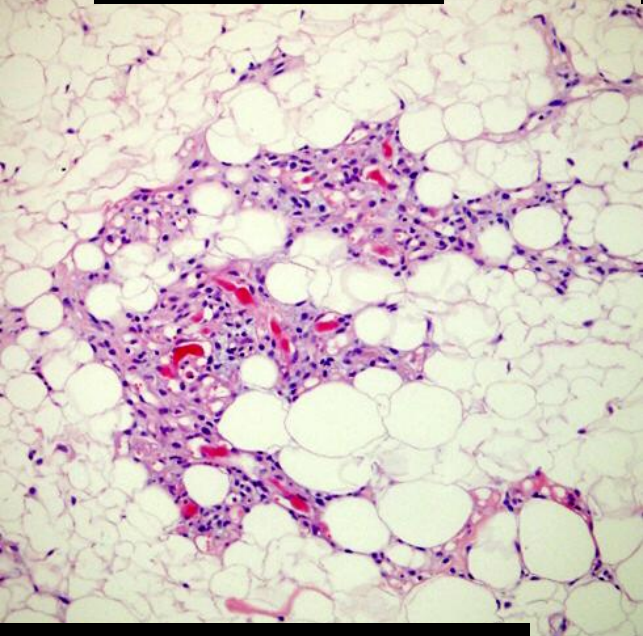
\*İğsi hücreli LS?

(bu ailede mi yer almalı??)

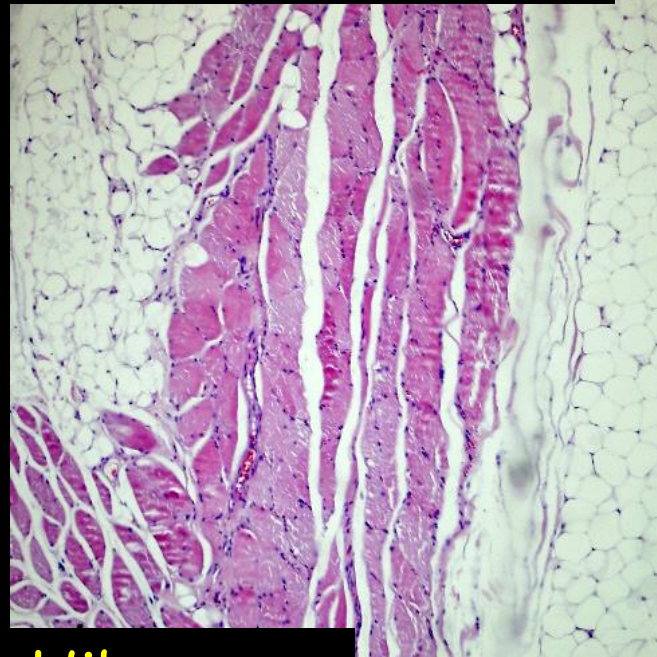
# Malign tümörler

- a. Miksoid LS
- b. Yuvarlak hücreli ( az diferansiye miksoid ) liposarcoma
- c. Pleomorfik LS
- d. Dediferansiye LS
- e. Mikst tip LS
- f. Liposarkom, NOS

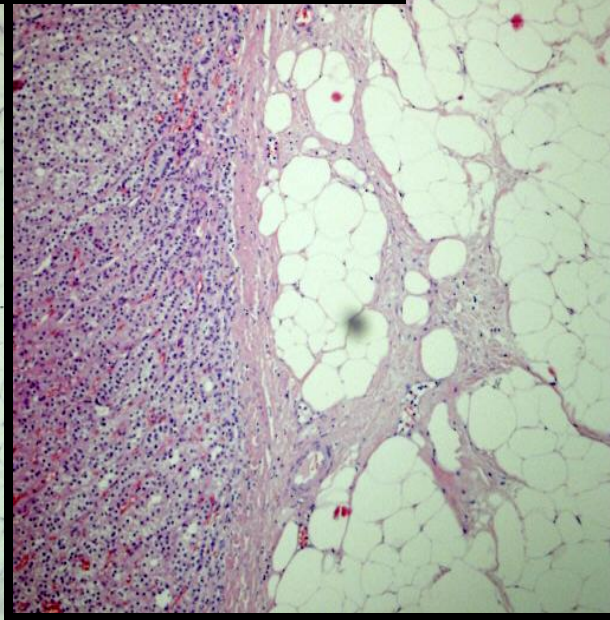
Anjiolipom



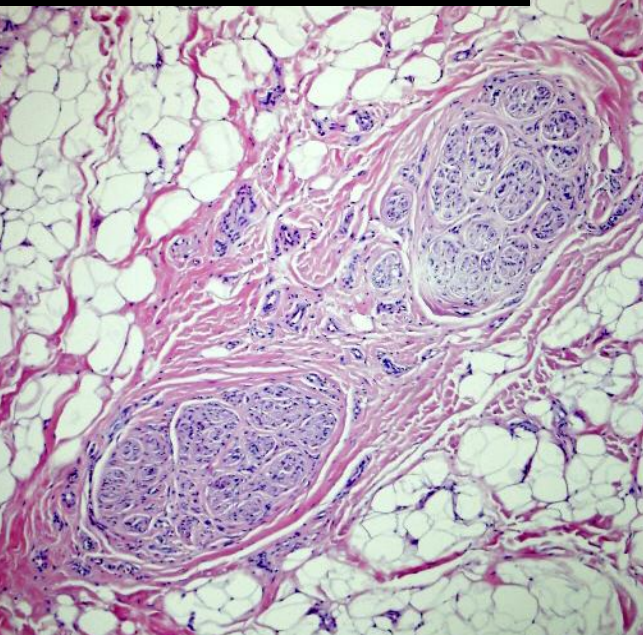
İntramuskuler lipom



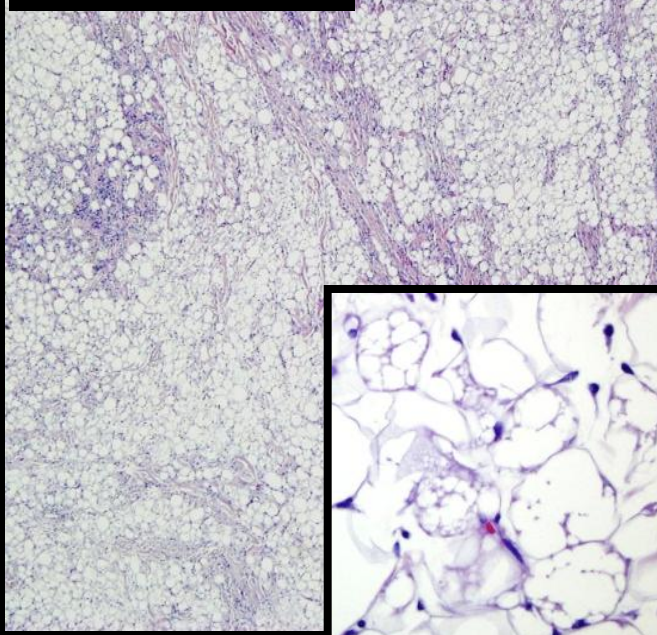
Myelolipom



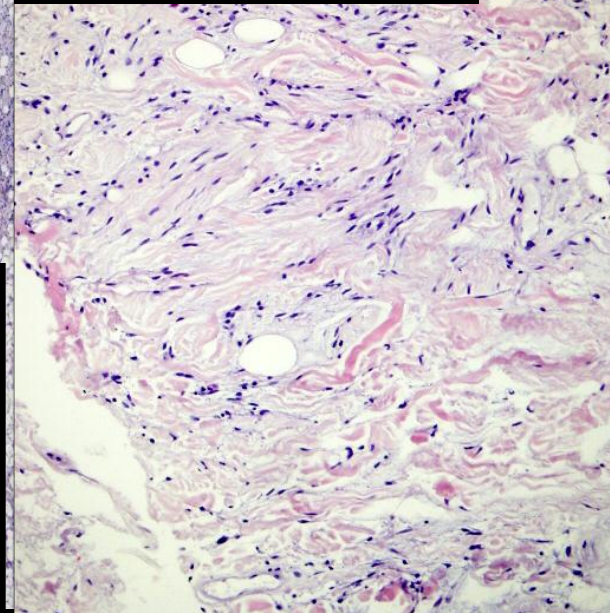
Nöral fibrolipom

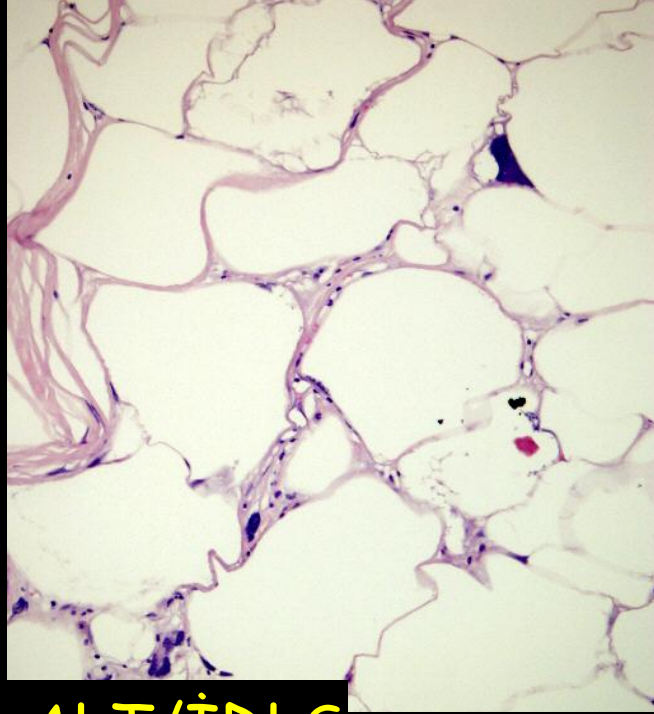


Hibernom

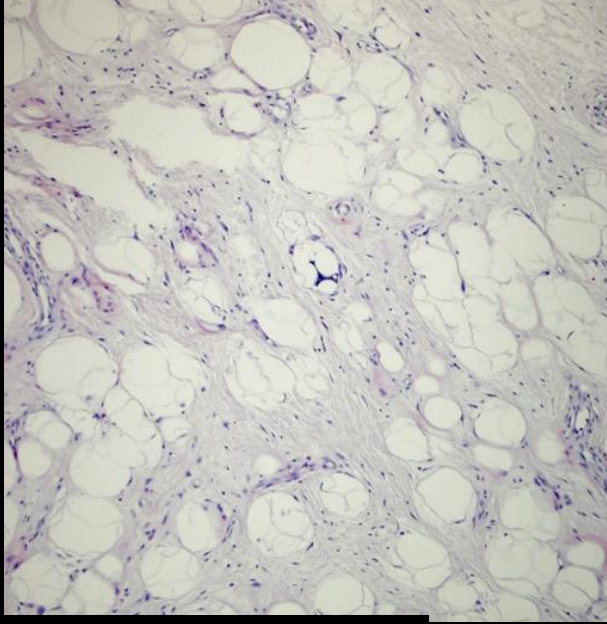


İğsi hücreli/PL

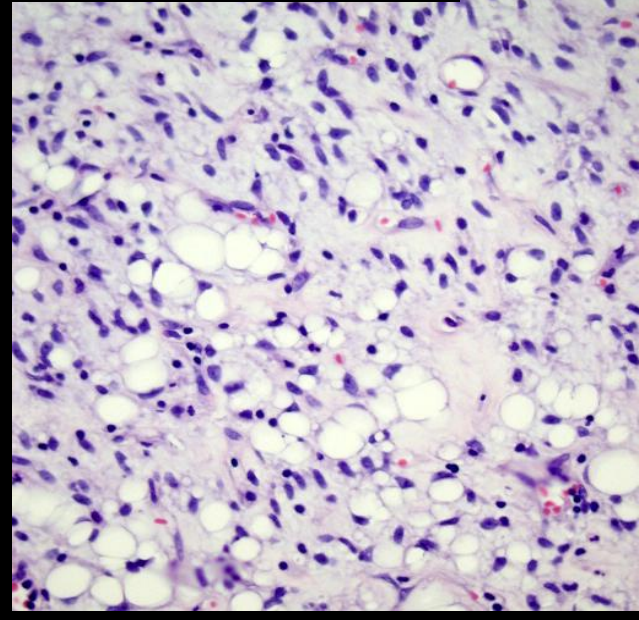




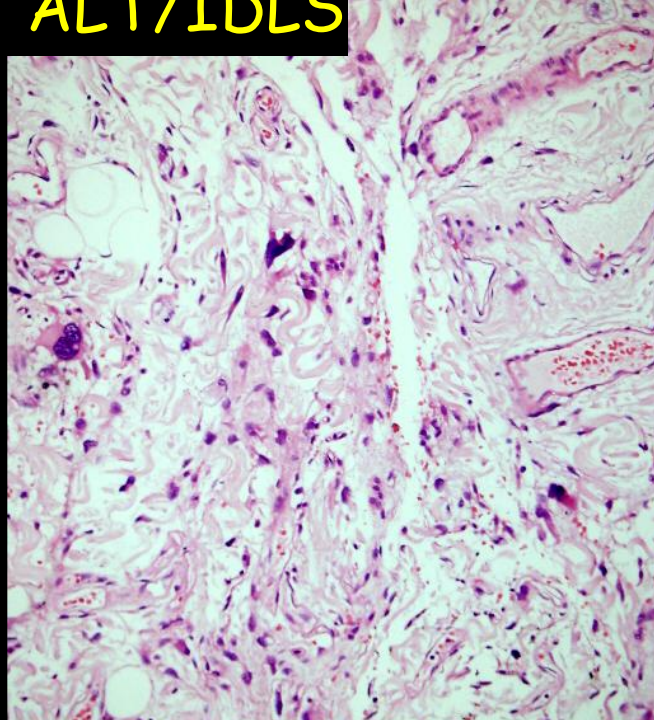
Lipom benzeri LS



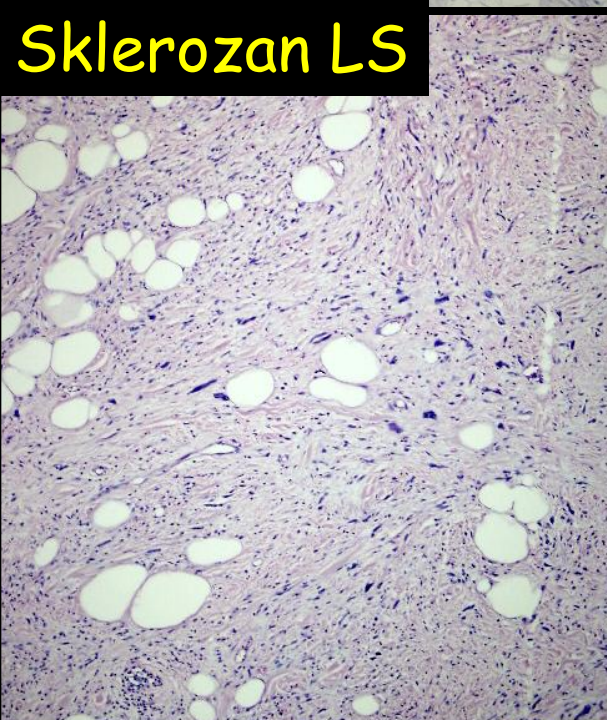
İğsi hücreli LS



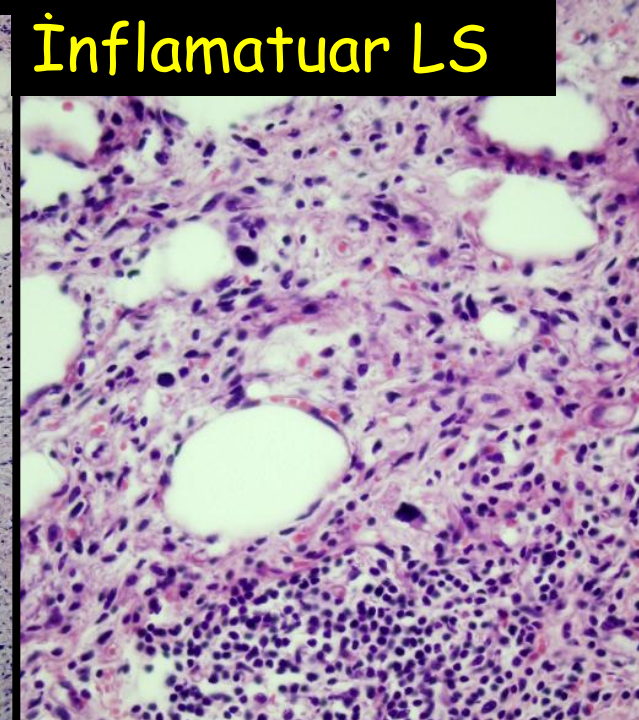
ALT/İDLS



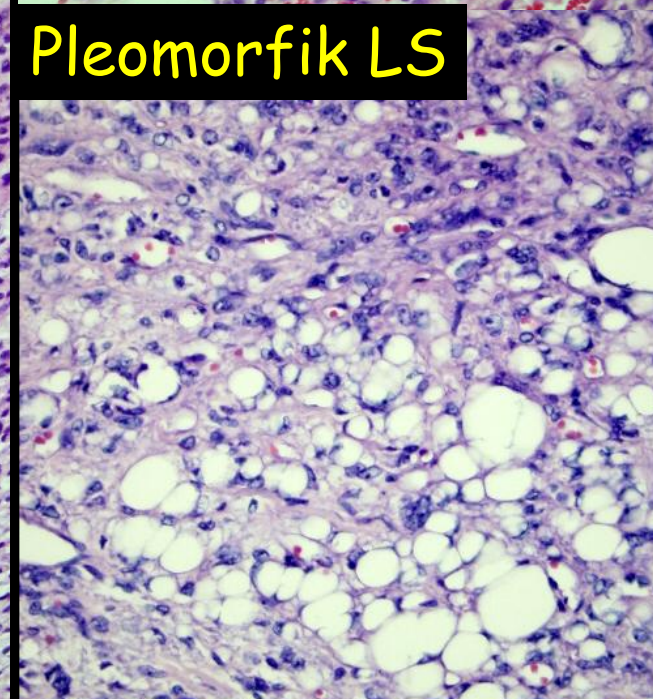
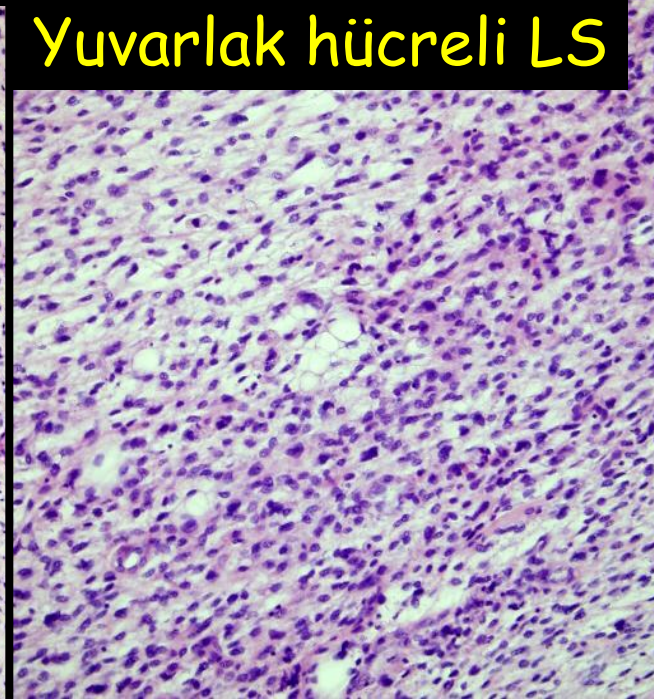
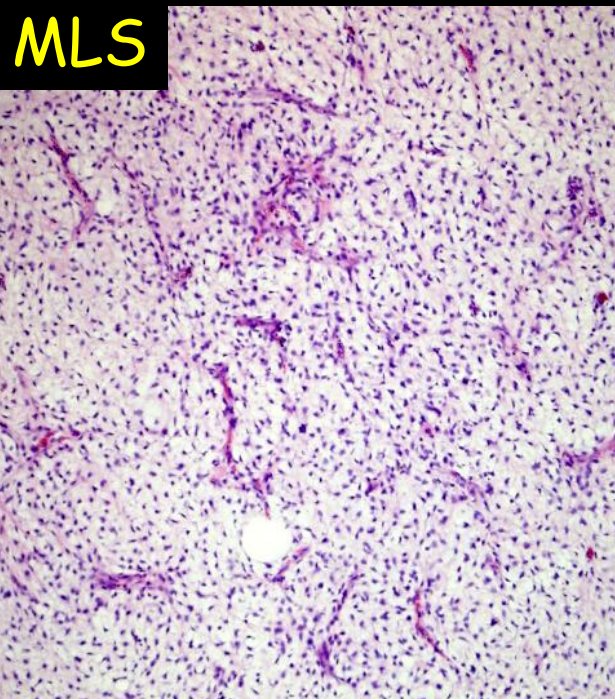
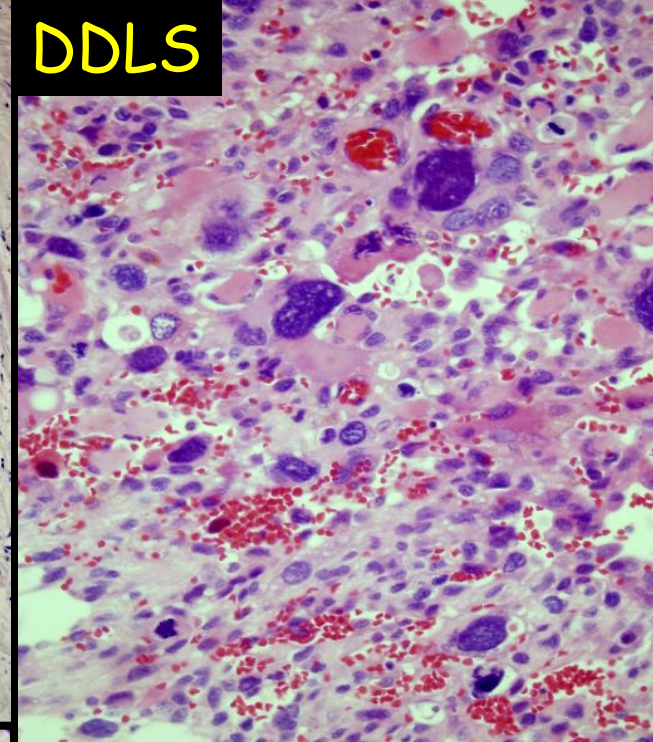
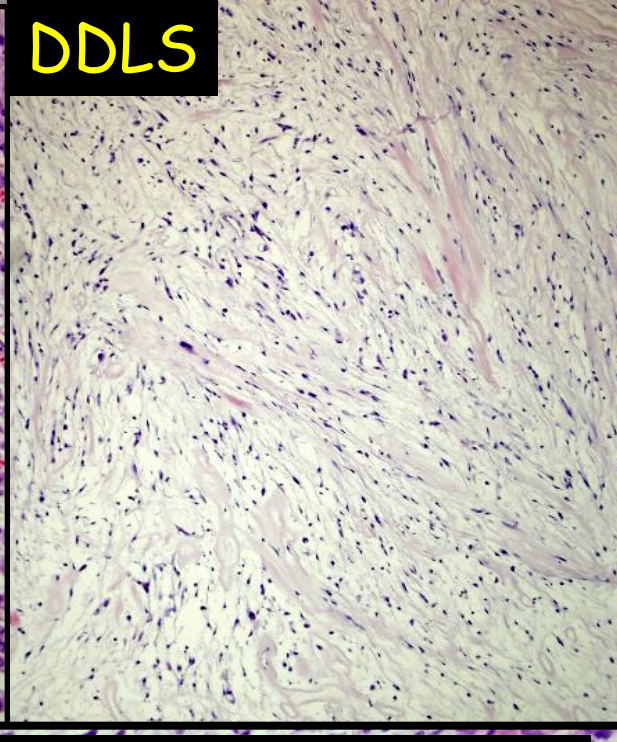
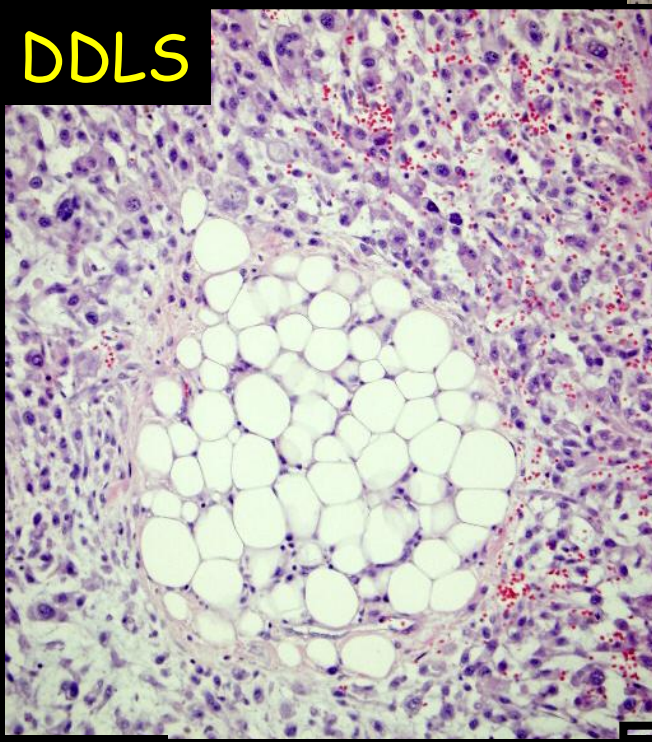
Sklerozan LS



İnflamatuvar LS







# Taniya ulařırken ?

## Klinik bilgi

Yař , Cinsiyet

Lokalizasyon (ekstremiteler, retroperiton, göğüs duvarı, mediasten vs)

Subkutan /Derin

Boyut

Klinik tanı?

Radyolojik bulgular (varsa)

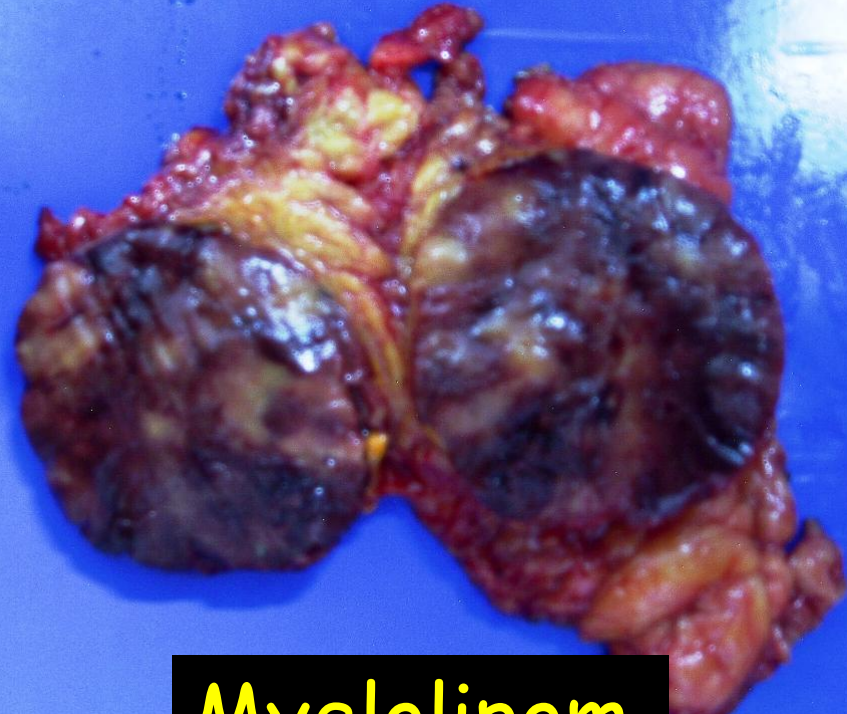
Lipom



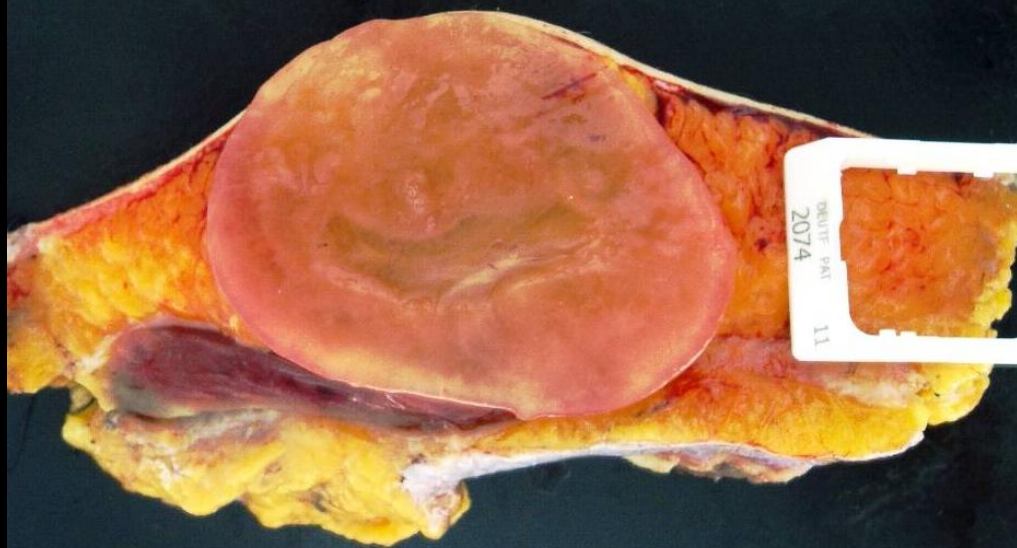
Hibernom



Myelolipom



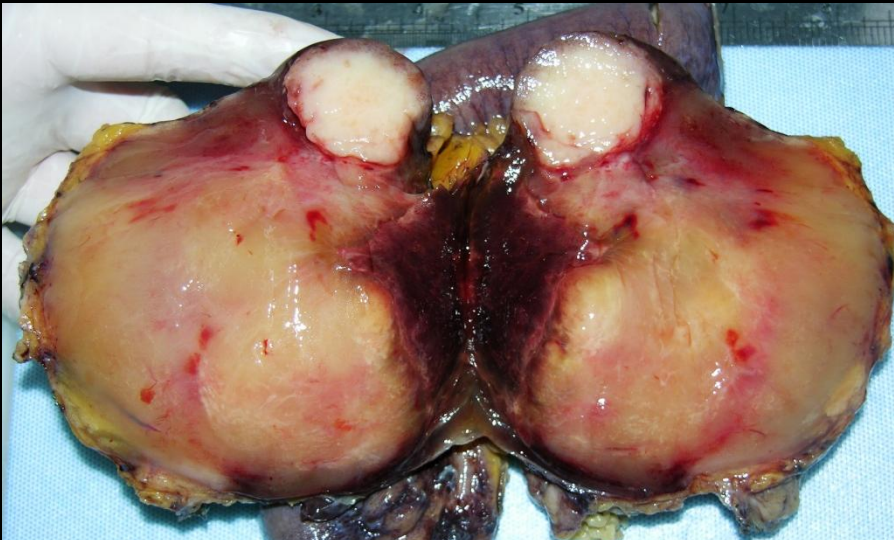
Mikroid LS



DDLS

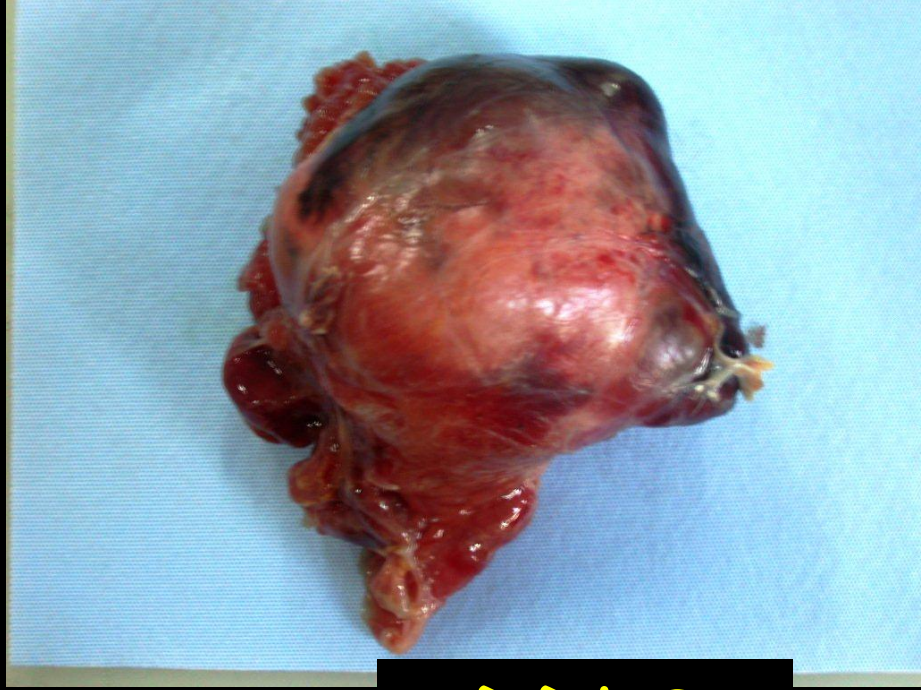


Liposarkom, NOS

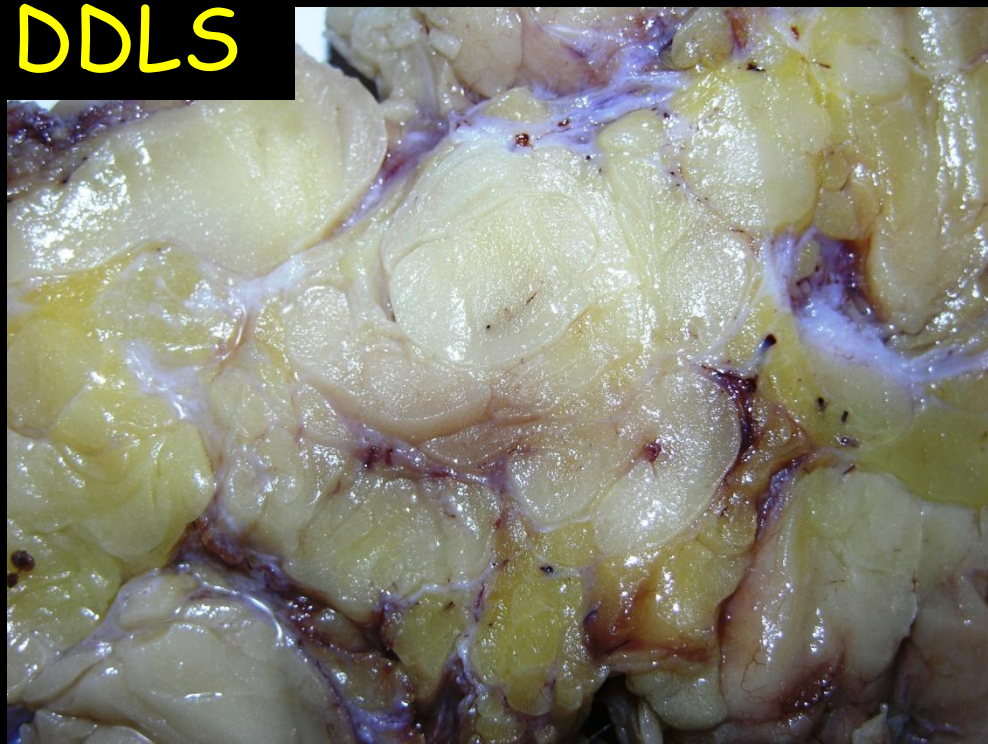


Mezenterik DDLS

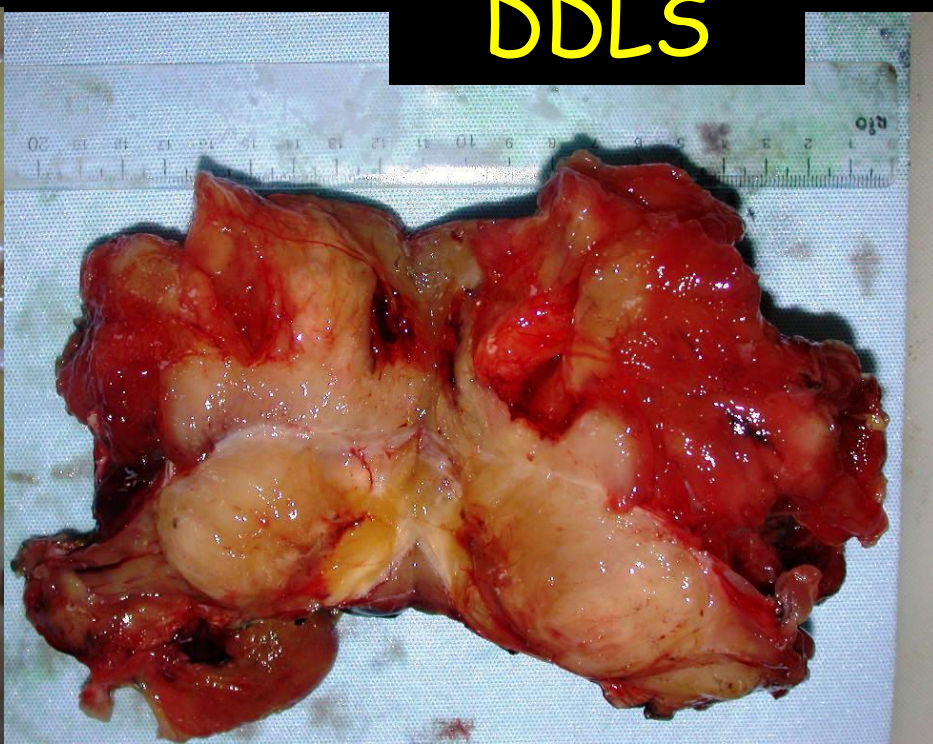
Intramuskulärer IDLS



DDLs



DDLs



# Benign Lipomatöz tümörler

## Lipom:

Etyoloji ?

40-60 y, Ç seyrek, %5 multipl

En sık g.

Matür beyaz yağ hücrelerinden oluşur

Subkutan, intramuskuler, intermuskuler

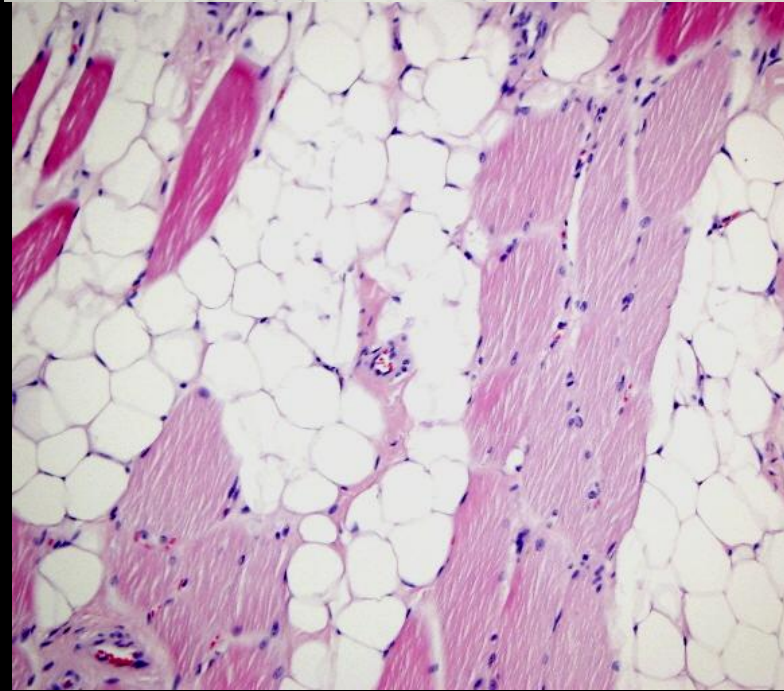
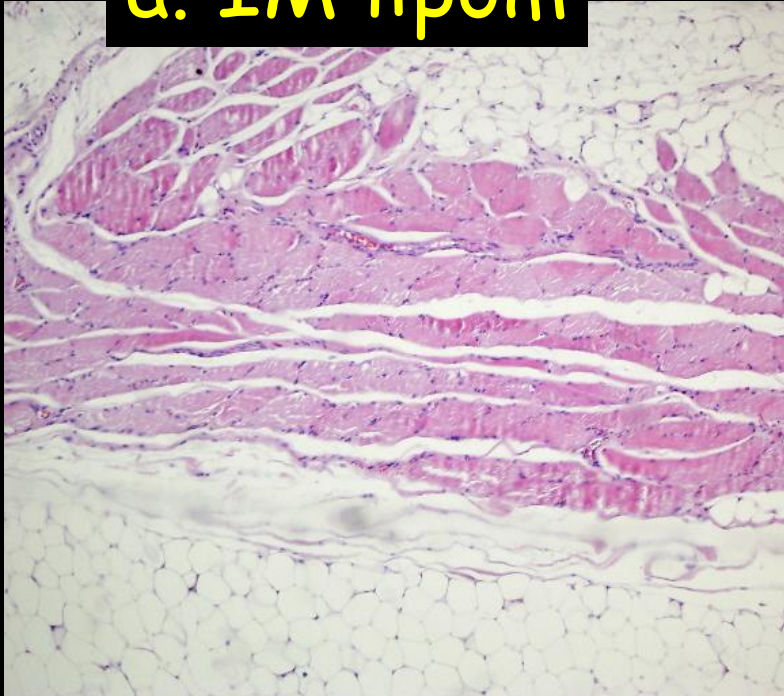
Yüzeyel lipomlar < 5cm

Derin lipomlar > 5cm

## Problemlı Lipomlar

- a. İntı-muskular lipom
- b. Hibernomatöz deęişiklik gösteren Lipom
- c. Yaę nekrozu ięeren Lipom
- d. Sellülar anjiolipoma
- e. İęsi hücreli / pleomorfik lipom
- f. Kondroid lipom

## a. IM lipom

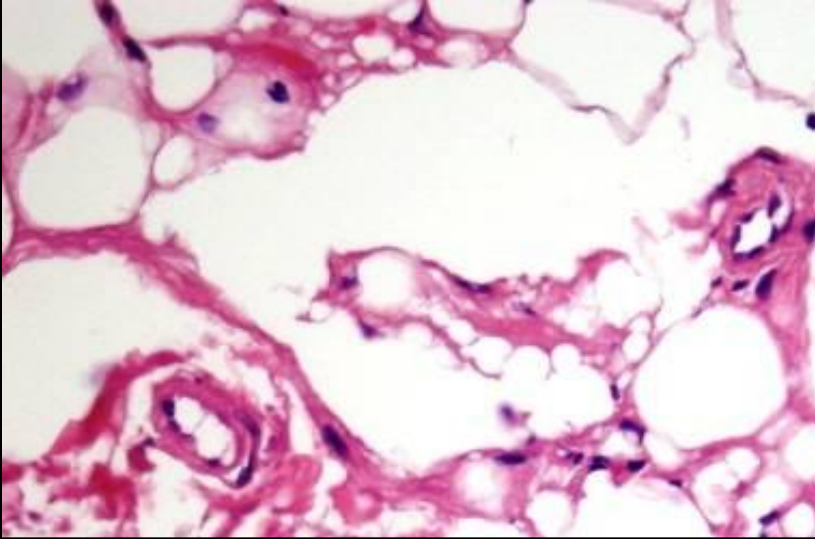
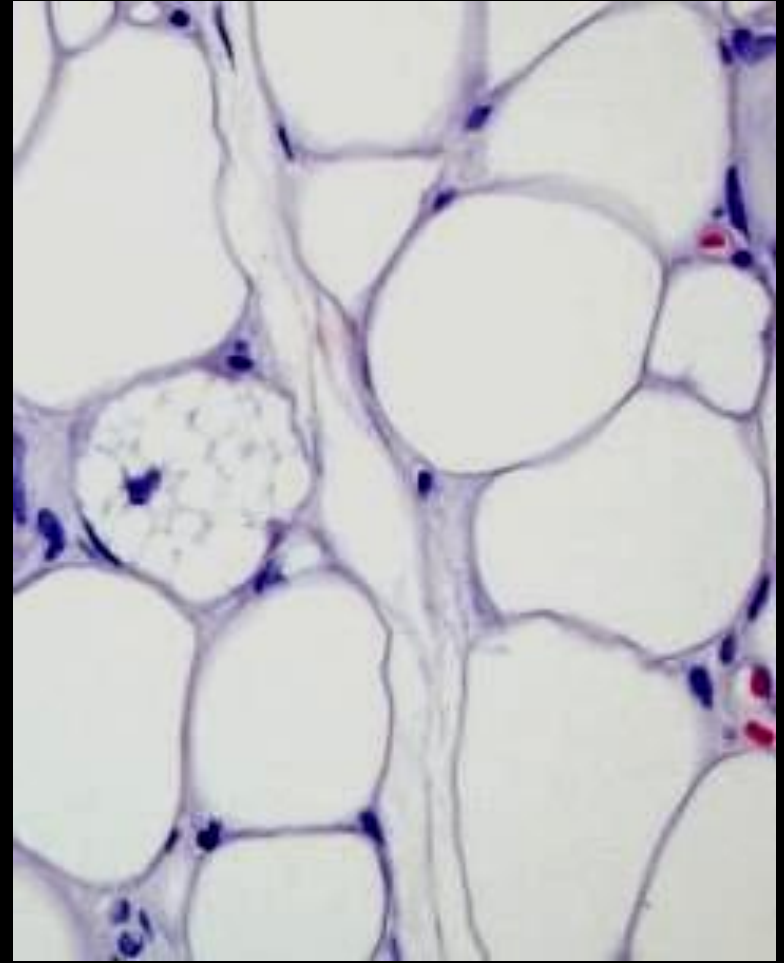
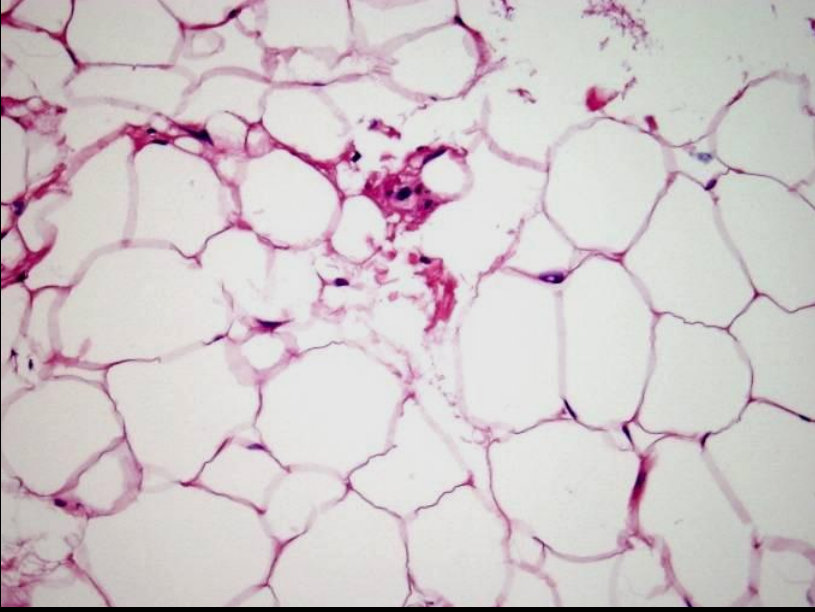


Büyük boyut, derin lokalizasyon,  
İnfiltratif büyüme. 30-60 y, Ç seyrek  
cm başına 1 parça  
Lipositler matür, lipoblast ve atipik  
hücre yok.

Lipoblastomatozis ve  
diffüz lipomatozisten  
ayırım güç ob.

(ç ve infant. Subkutis ve kas doku  
Daha lobüle , değişen  
kalınlıkta bağ doku septaları, değişik  
evrelerdeki lipoblastlar





**b. Hibernomatöz deęişiklik gösteren lipom**

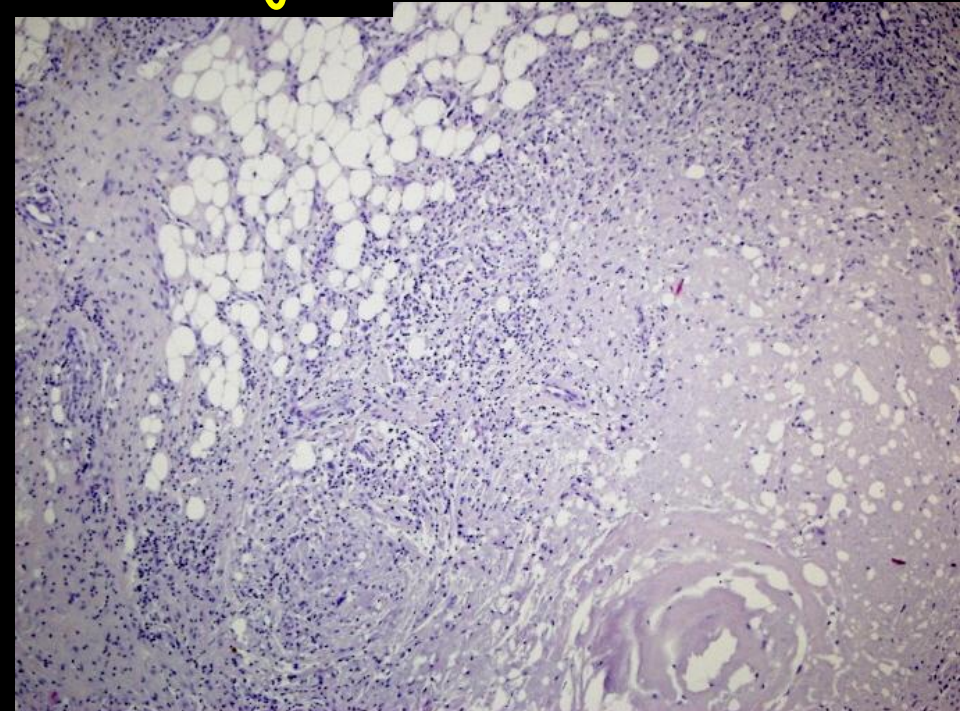
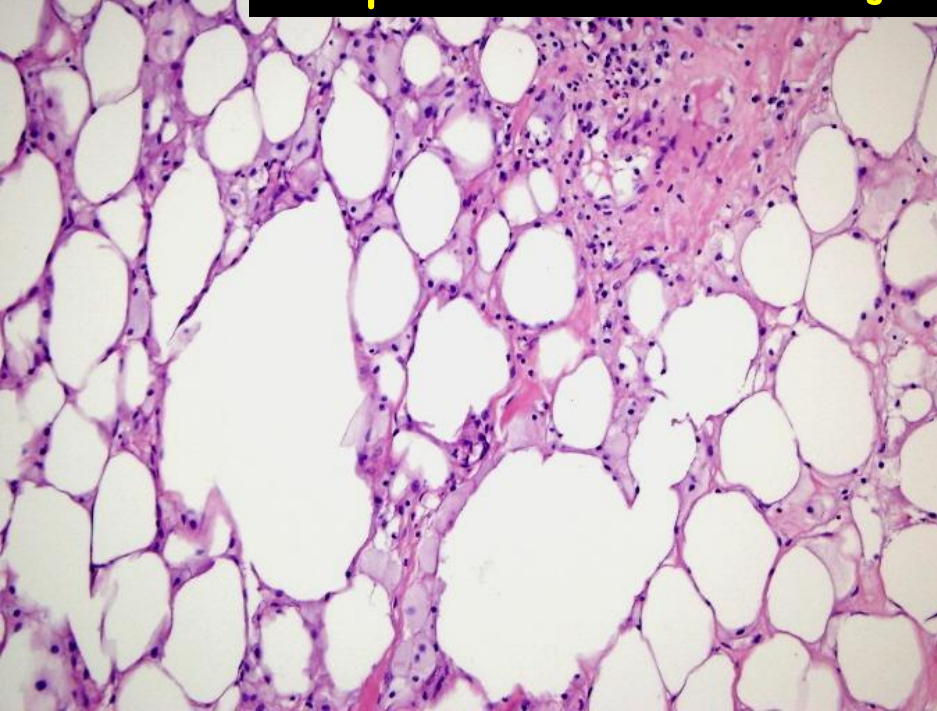
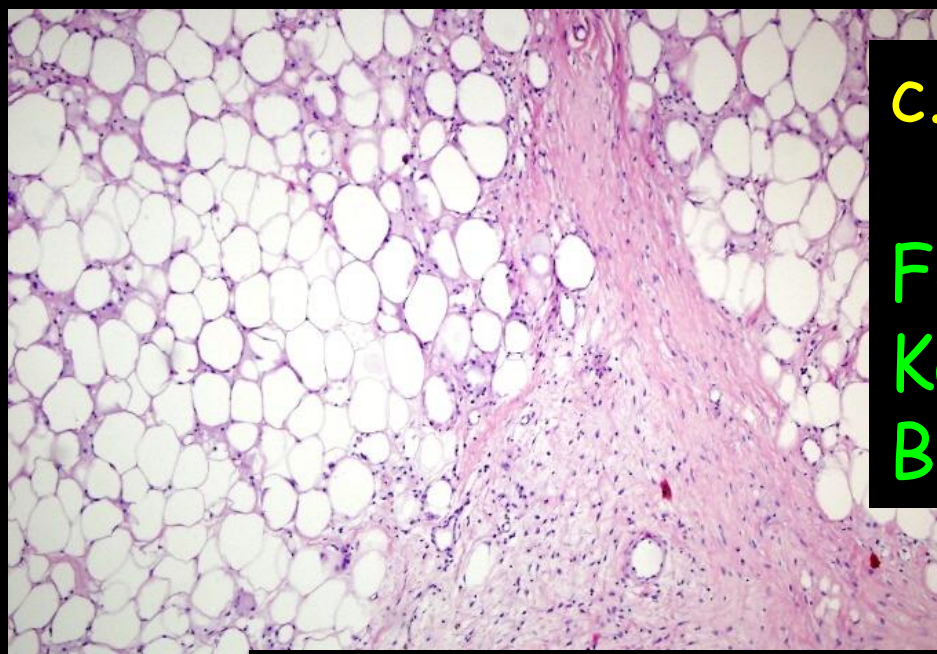
## c. Yağ nekrozu içeren lipom

Fokal/yaygın

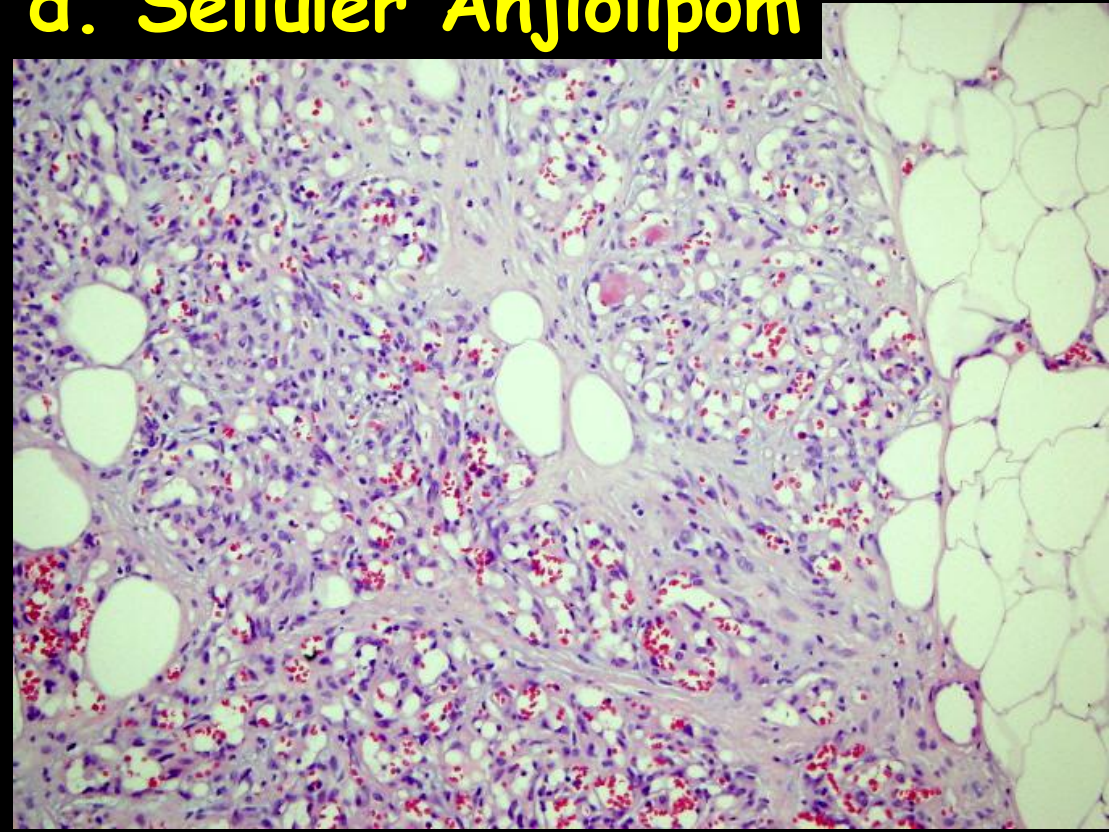
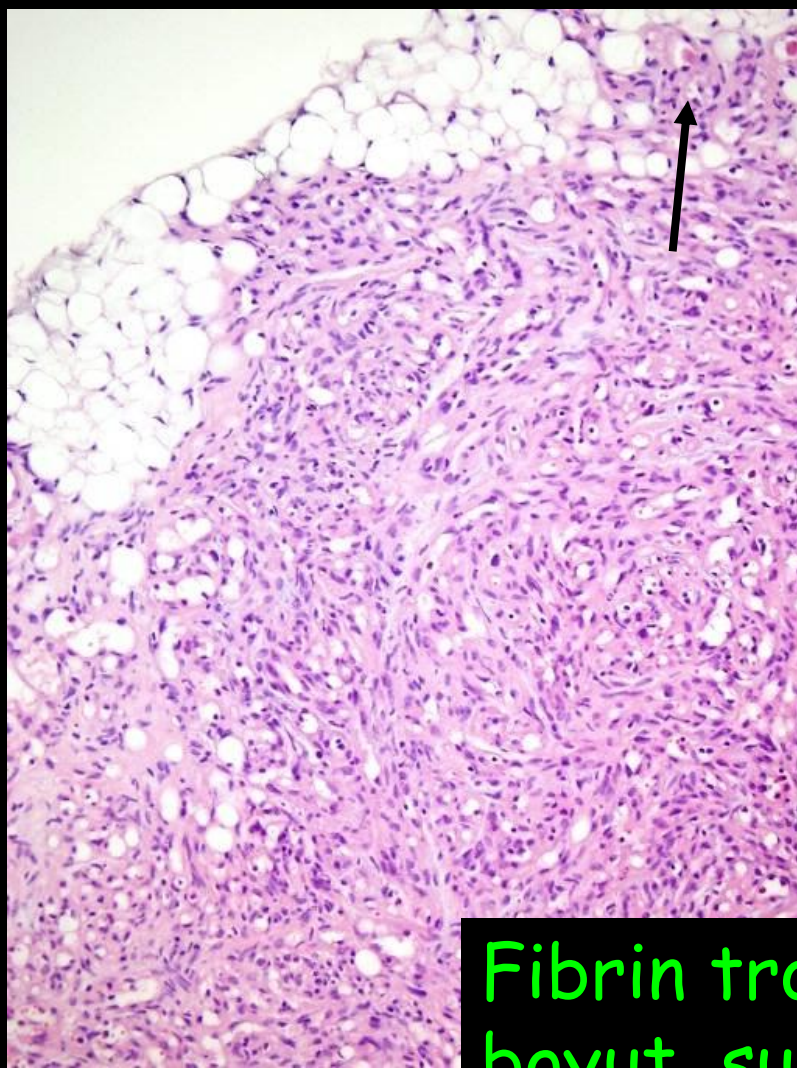
Kan akımı bozulması/travma

Bazen likefaktif nekroz

Atipik hücre ile karışan makrofajlar



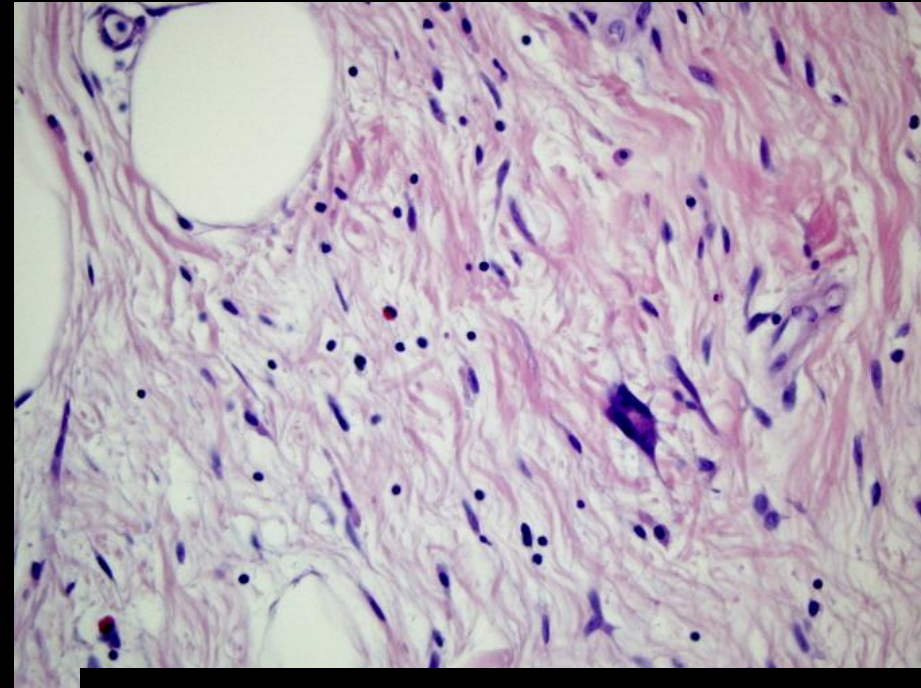
## d. Sellüler Anjiolipom



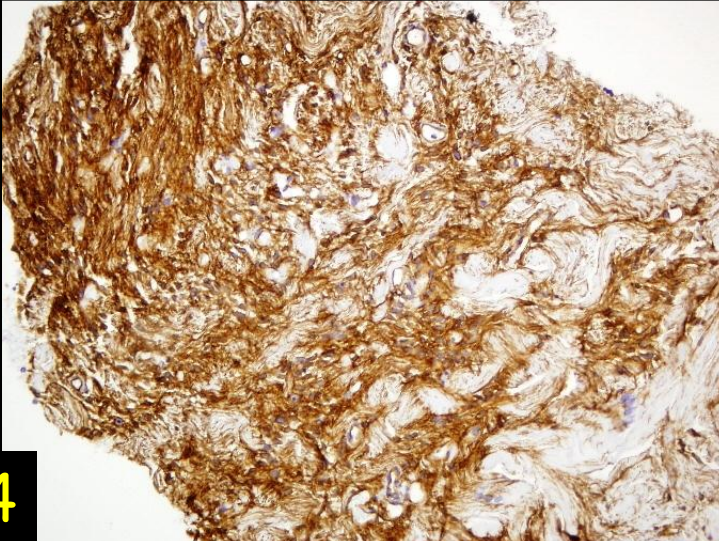
Fibrin trombları ve mast hücre +, küçük boyut, subkutan (kolda), kapsül+ Kaposi sarkom ve içsi hücreli AS ile karışır. Kaposi yarık benzeri vasküler yapılar +, fibrin trombu içermezler

## e. İğsi hücreli/ Pleomorfik Lipoma

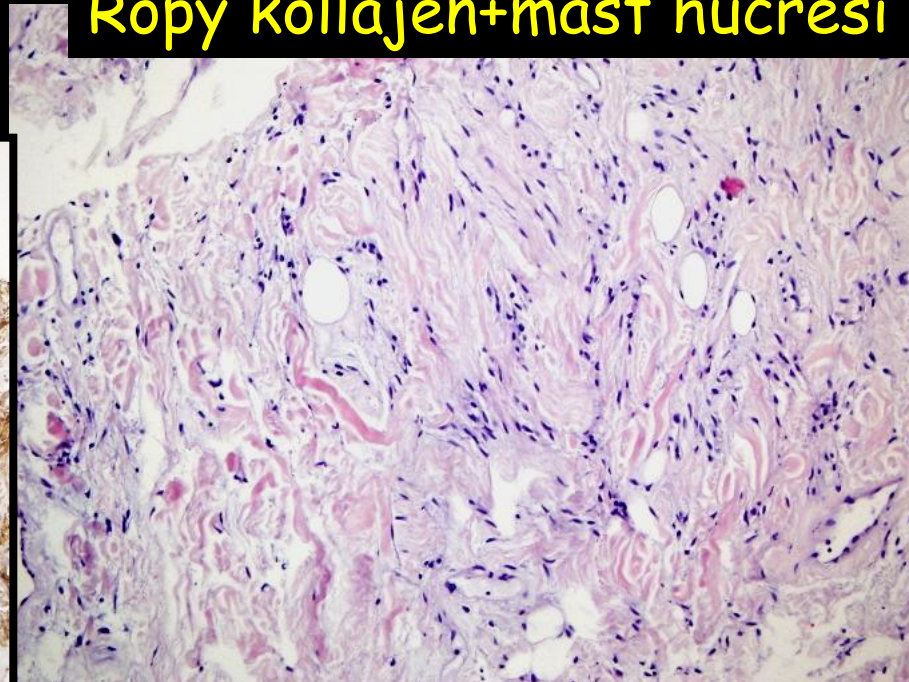
- İyi sınırlı subkutan kitle
- Omuz, boyun ve sırt
- E sık
- Nüks/metastaz yok
- CD34-pozitif
- sitogenetik (16q8, 13q8)

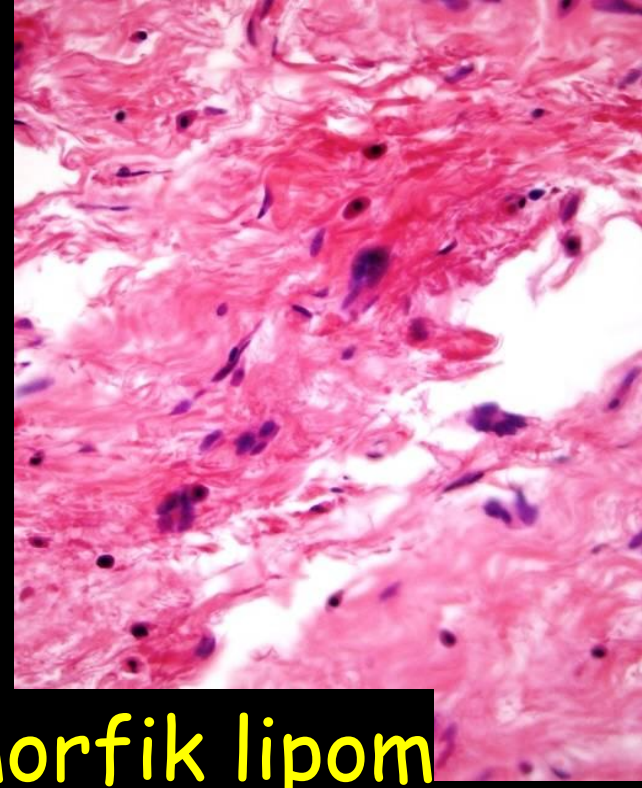
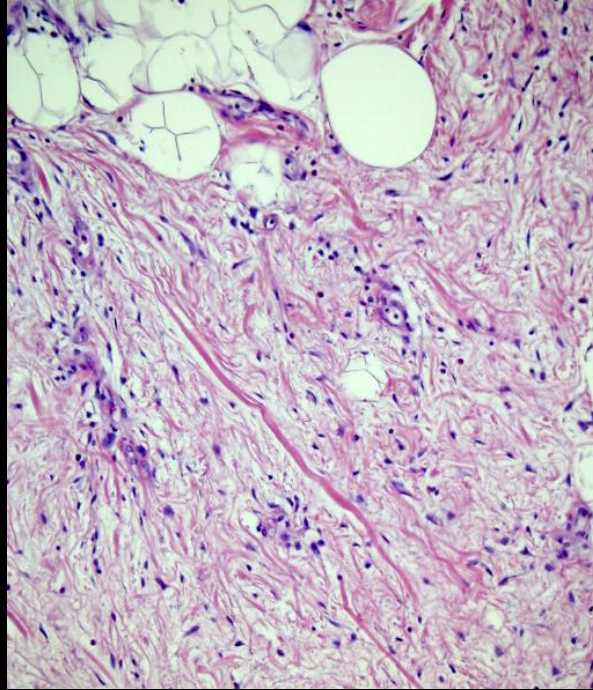
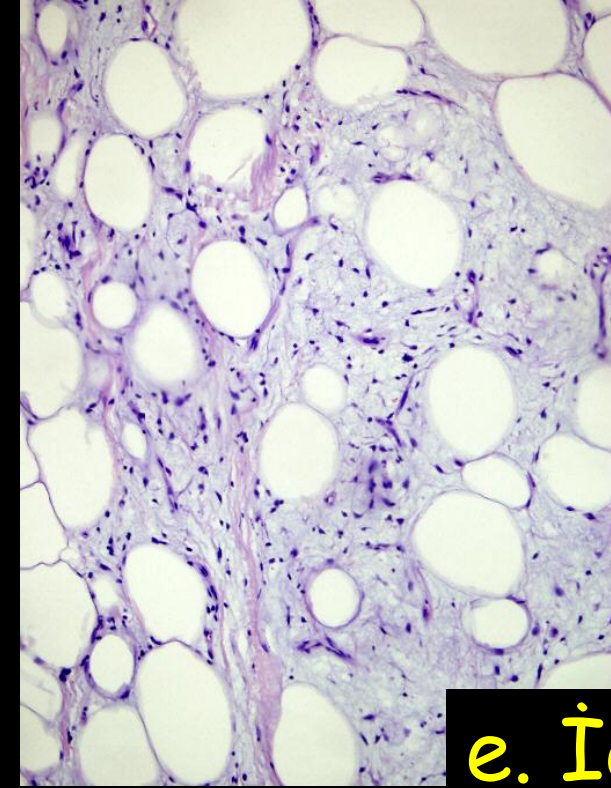


Ropy kollajen+mast hücresi

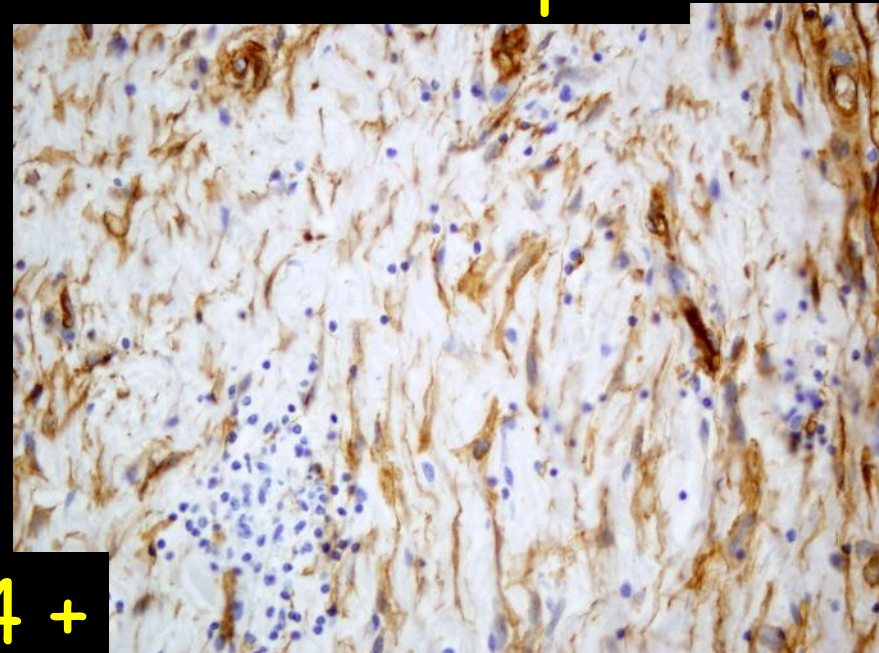
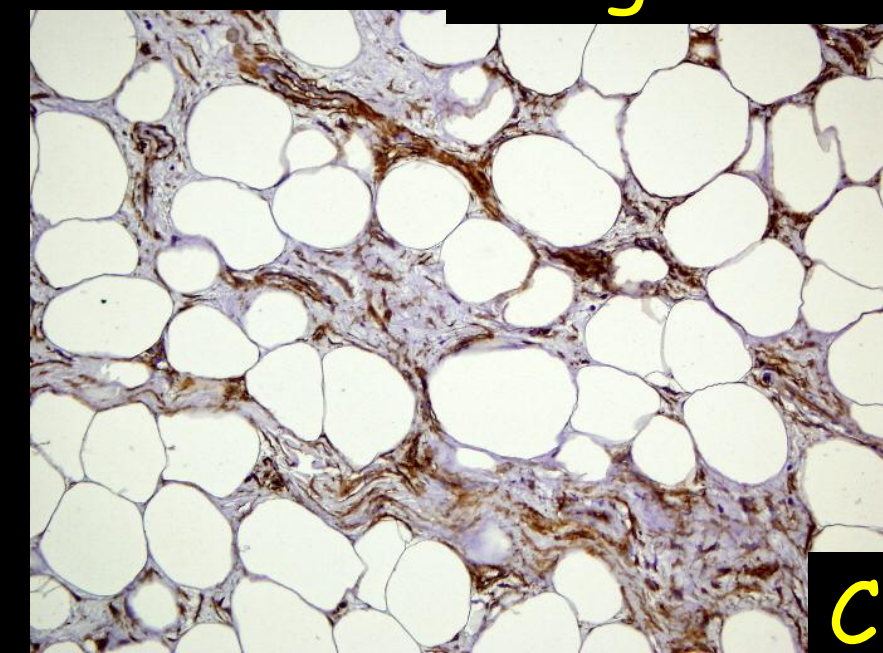


CD34





e. İğsi hücreli/Pleomorfik lipom

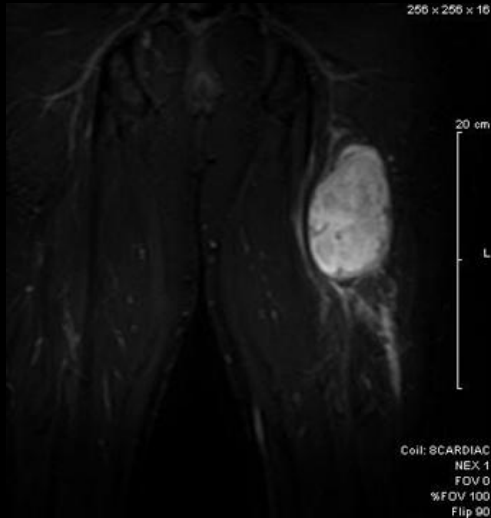


CD34 +

# Kondroid lipom;

iyi sınırlı kapsüllü, subkutan  
KY; sarı-beyaz, pembe-ten  
renkli, 1-11 cm

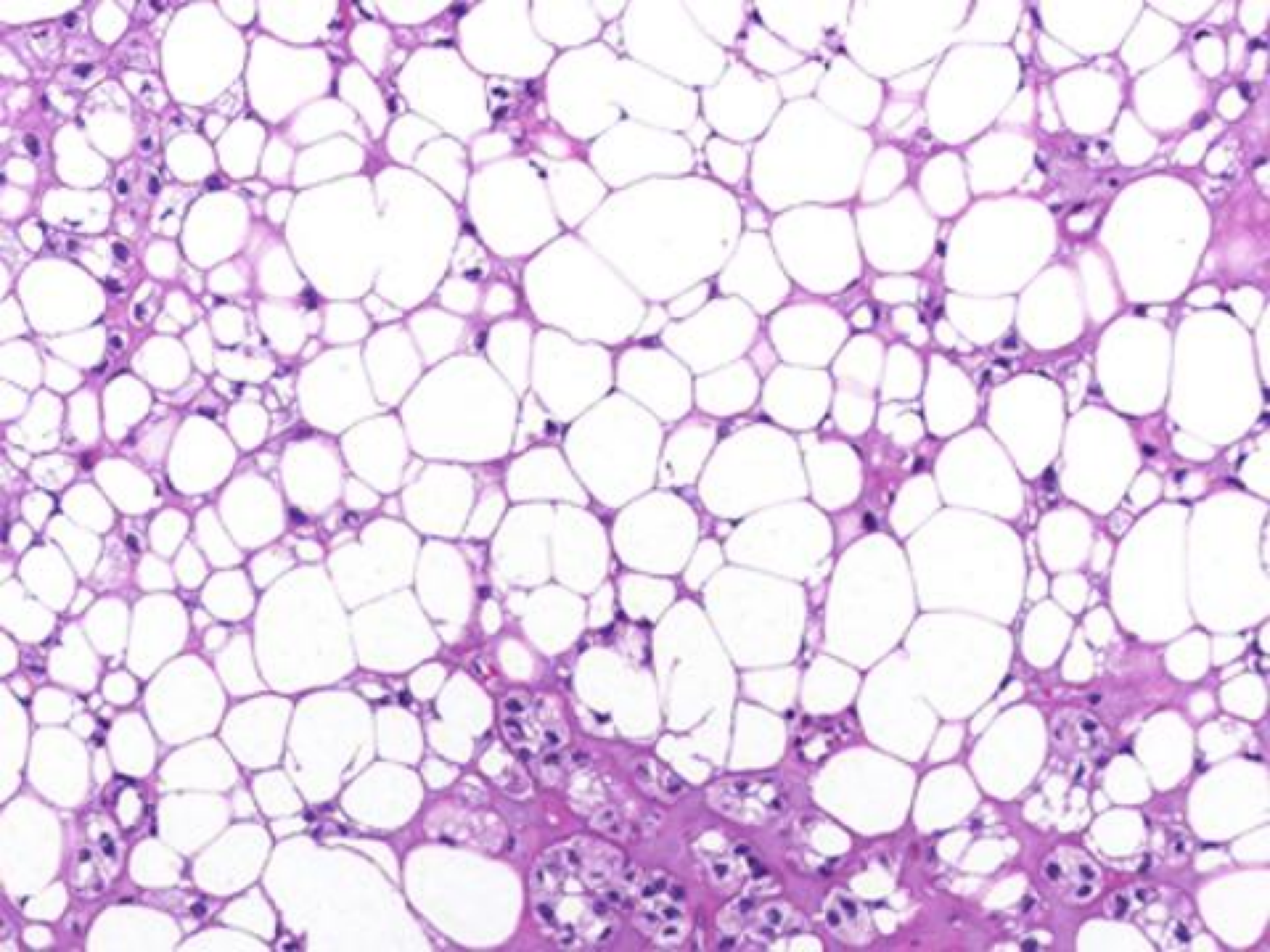
Lipoblastik  
diferansiyasyon hem de  
primitif kıkırdak  
gösteren **bifenotipe sahip**



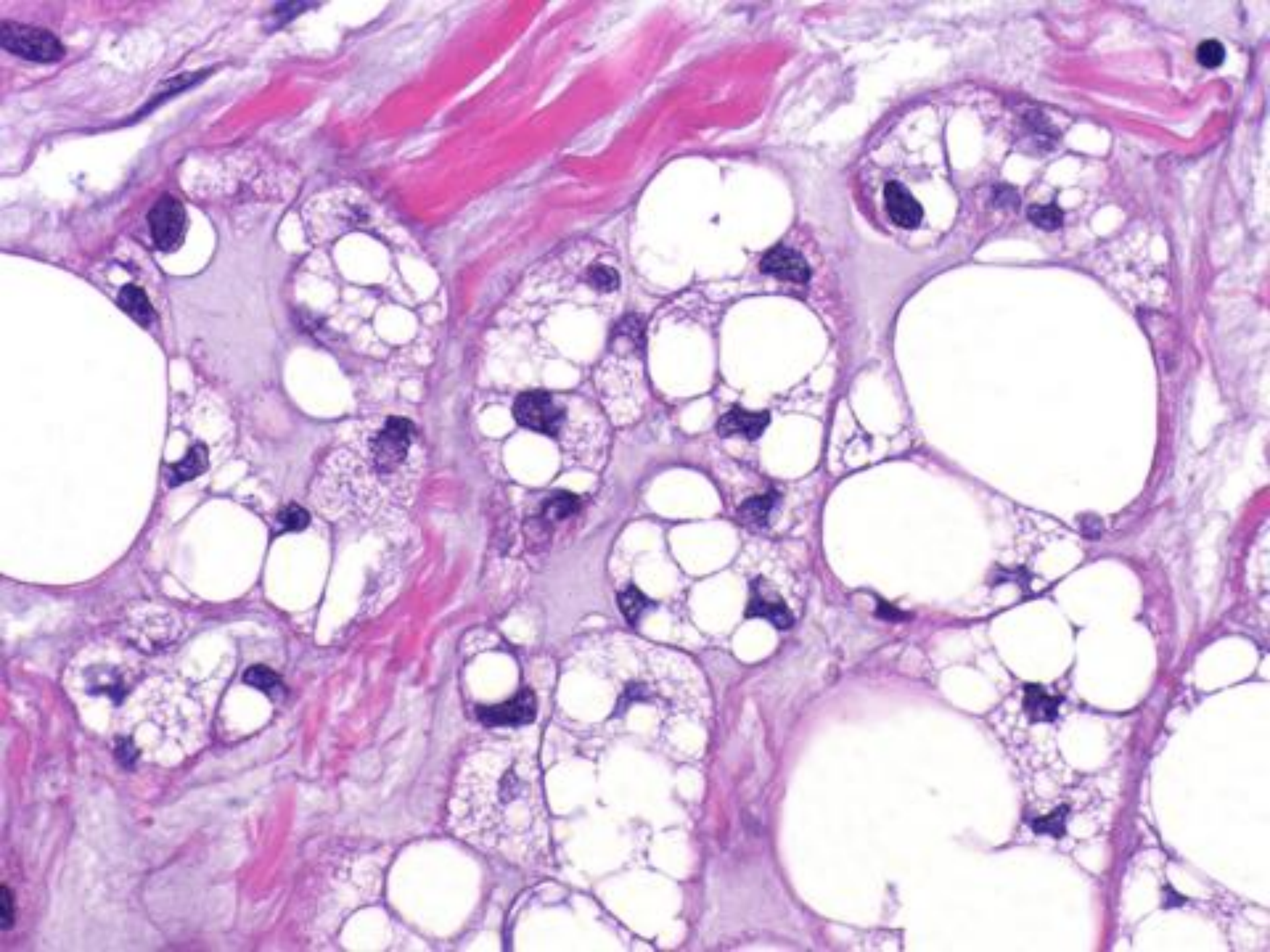


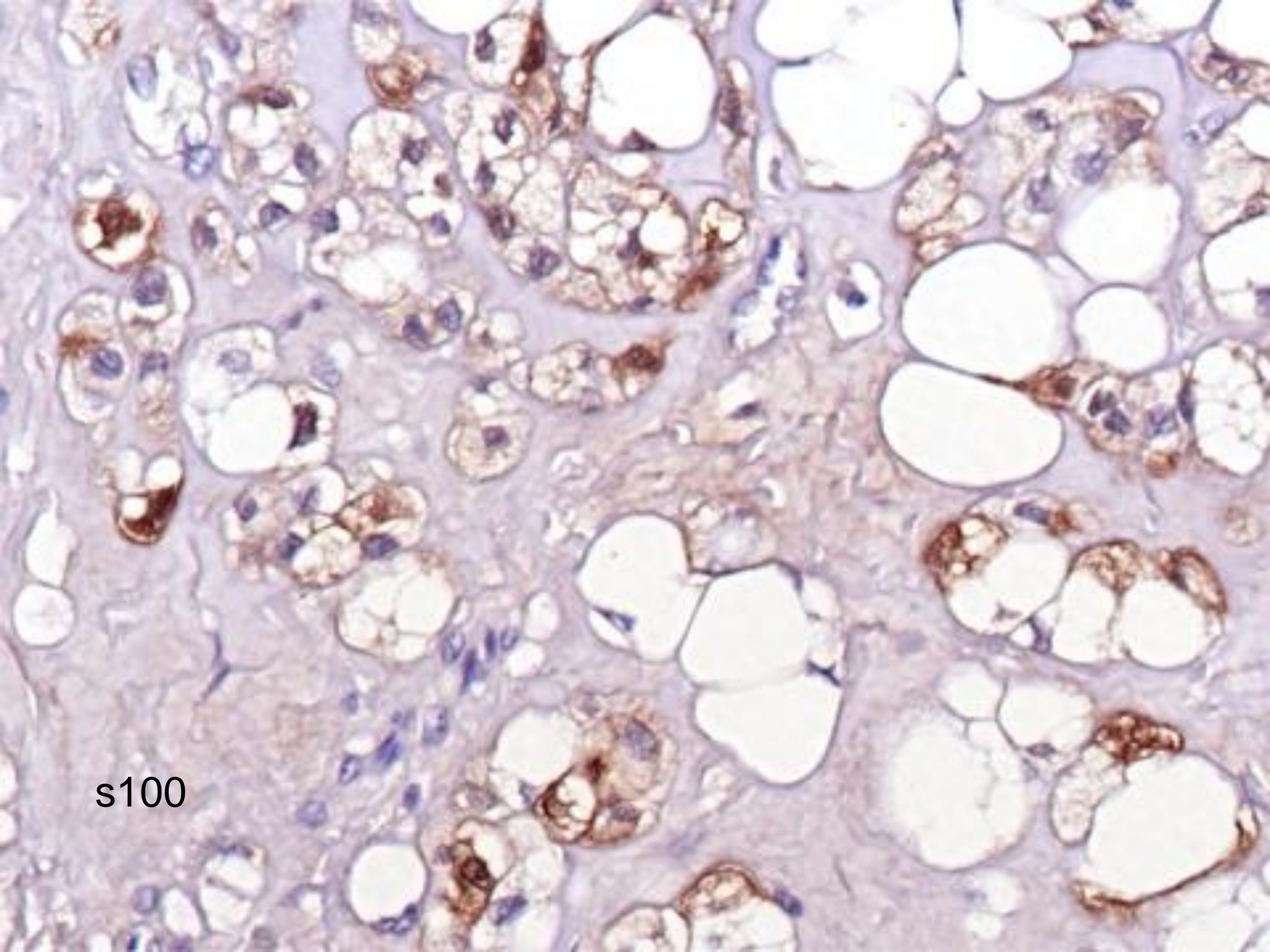
AT; M-KS, MLS

miksokondroid/hyalinize  
fibröz zeminde  
Matür yağ, lipoblast









s100



# Liposarkomlar; YDS'nin %25

Klinikopatolojik ve moleküler genetik özellik

- a. ALT-IDLS / DDLS\*\* (en sık)
  - İğsi hücreli IDLS farklı antite?
- b. Miksoid ve yuvarlak hücreli LS (2.sıklıkta)
- c. Pleomorfik LS (en seyrek)

# Ekstremitte, gövde yerleşimli İDLS

(geniş rezeksiyon kür)

Dediferansiye komponent içermedikçe  
metastaz potansiyeli yok

**ALT-İDLS sinonim terim**

(biyolojik potansiyeline göre, genetik ve  
morfolojik olarak identik)

Terminoloji tercihi; cerrah-patolog

Yetersiz-gereksiz tedavi

# ALT-İDLS;

ekstremiteler,  
retroperiton,  
paratestiküler bölge,  
kasık

Ağrısız yavaş büyüyen  
kitle.

MR ve BT

(fibröz septa,  
Sklerotik-inflamatuvar)



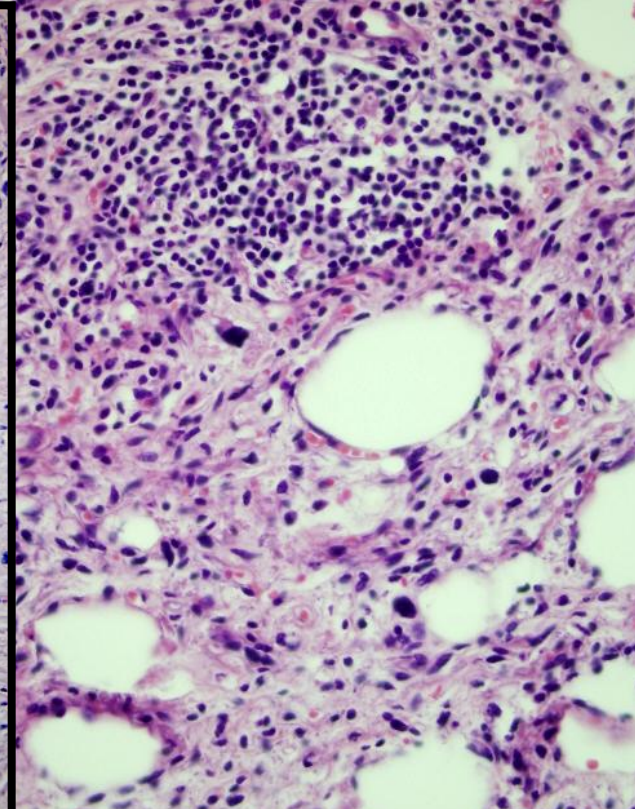
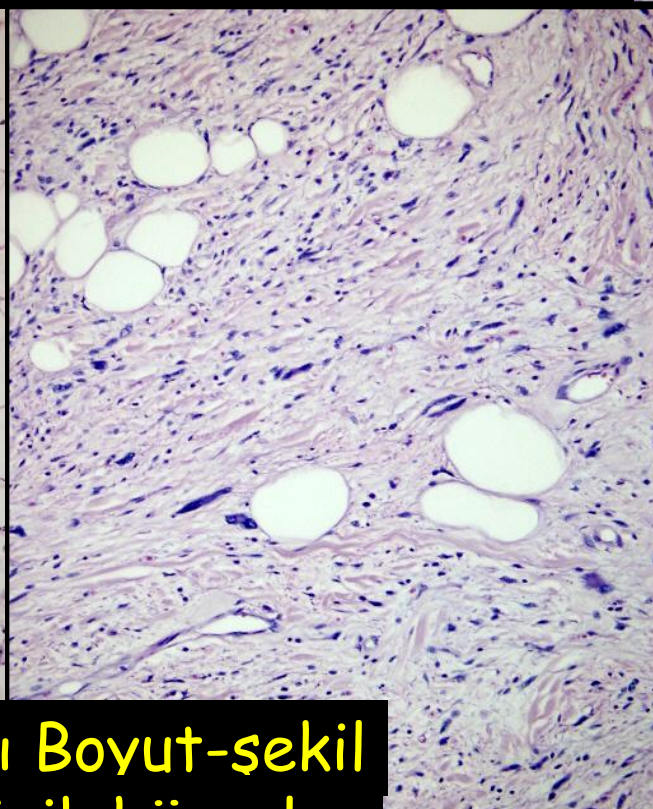
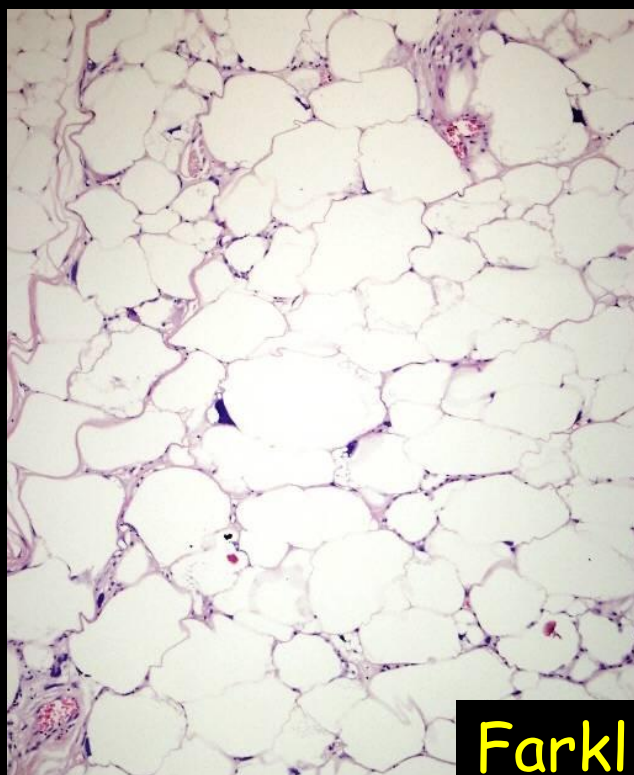
# ALT/İDLS

- a. Lipom benzeri (ekstremiteler)
- b. Sklerozan; retroperiton, paratestiküler
- c. İnflamatuvar; retroperiton  
(a+b+inflamatuvar hücreler)

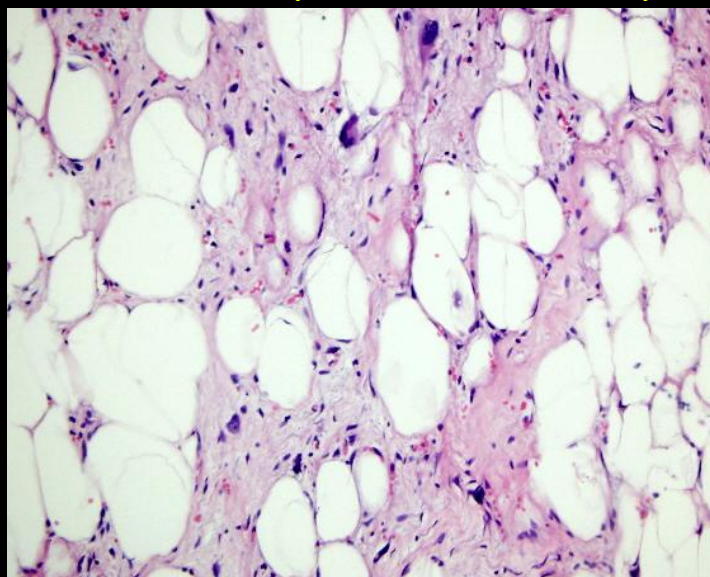
AT: Lenfoma/enflamatuvar süreçler

Retroperitonda İDLS/DDLS'u her zaman  
akılda tutalım!

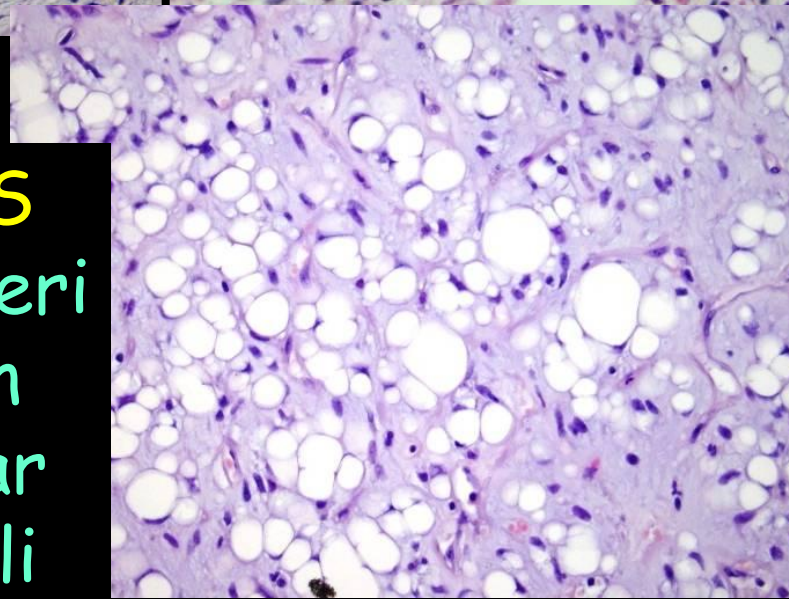
Alt türlerin ayırımı subjektif ve bilinen bir  
prognostik önemi yok



Farklı Boyut-sekil  
Fibröz septalarda atipik hücreler



ALT/İDLS  
Lipom benzeri  
Sklerozan  
İnflamatuvar  
İğsi hücreli





## d. İğsi hücreli İDLS;

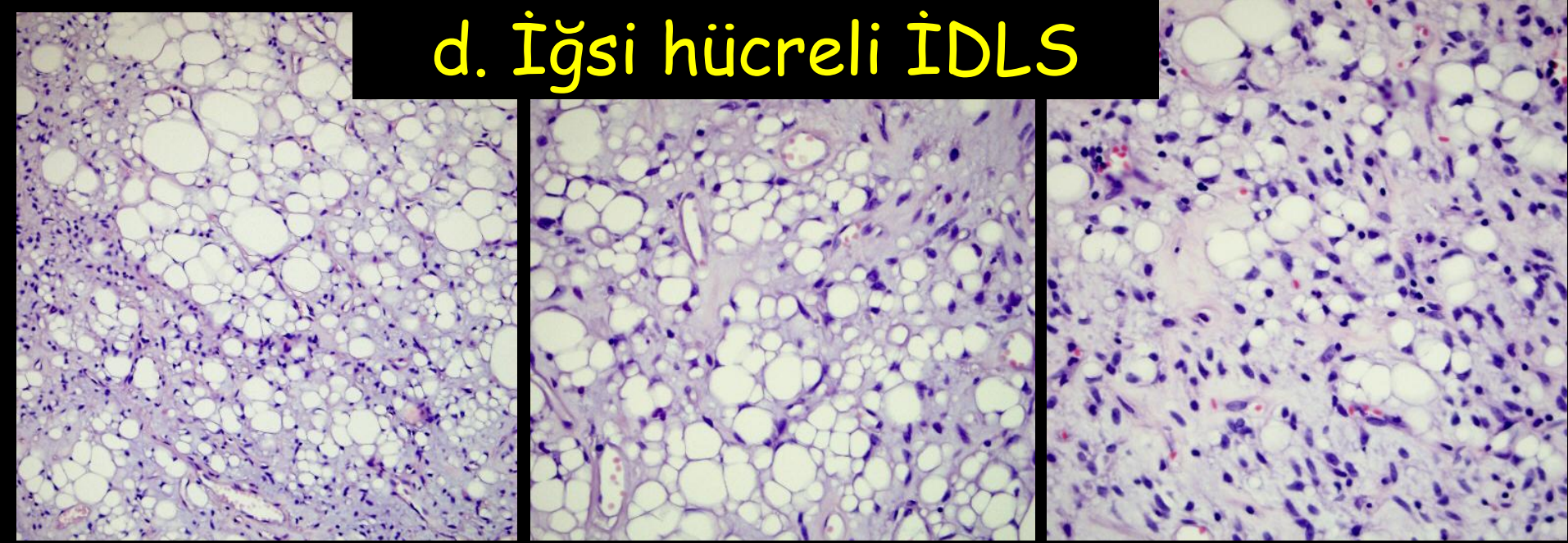
Subkutan yerleşimli, gövde (omuz), ekstremitelerde  
baş boyun da sık,  
Retroperiton ve abdomen seyrek,

ALT/İDLS dan farklı sitogenetik ayrı bir antite

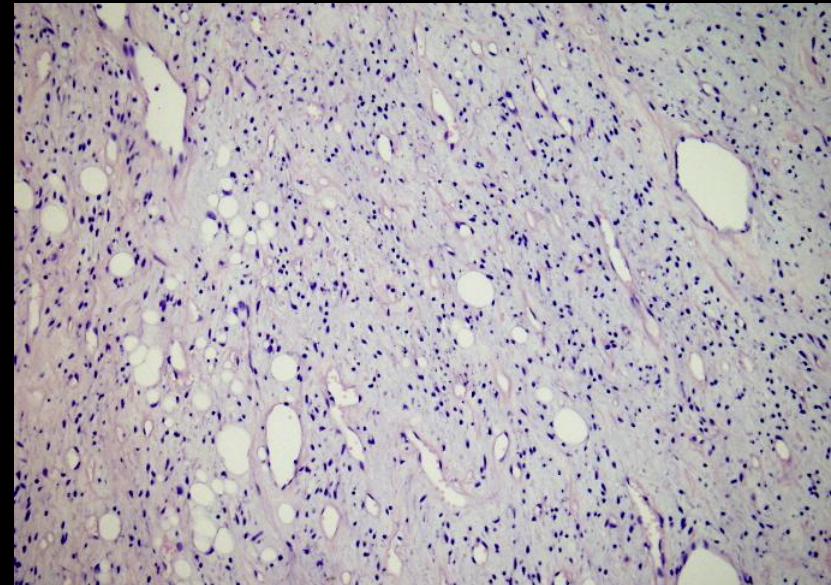
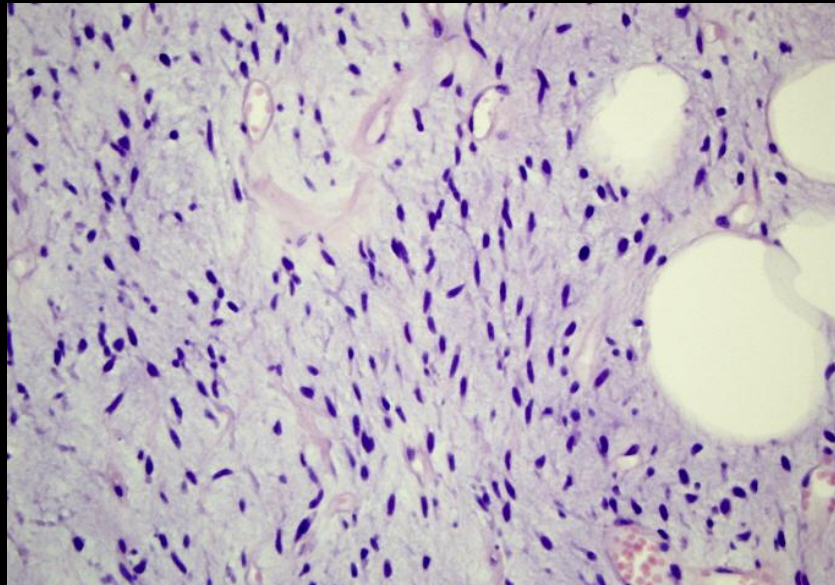
İğsi hücreli lipomun malign karşılığı,  
Miksoid LS un iğsi hücreli alt türü  
Düşük dereceli DDLS

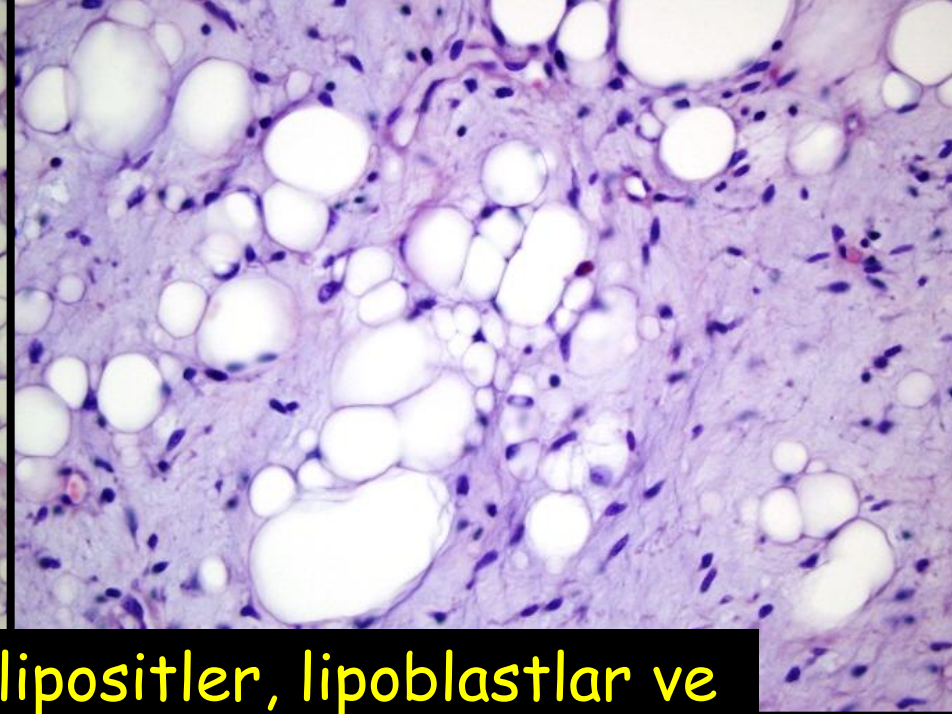
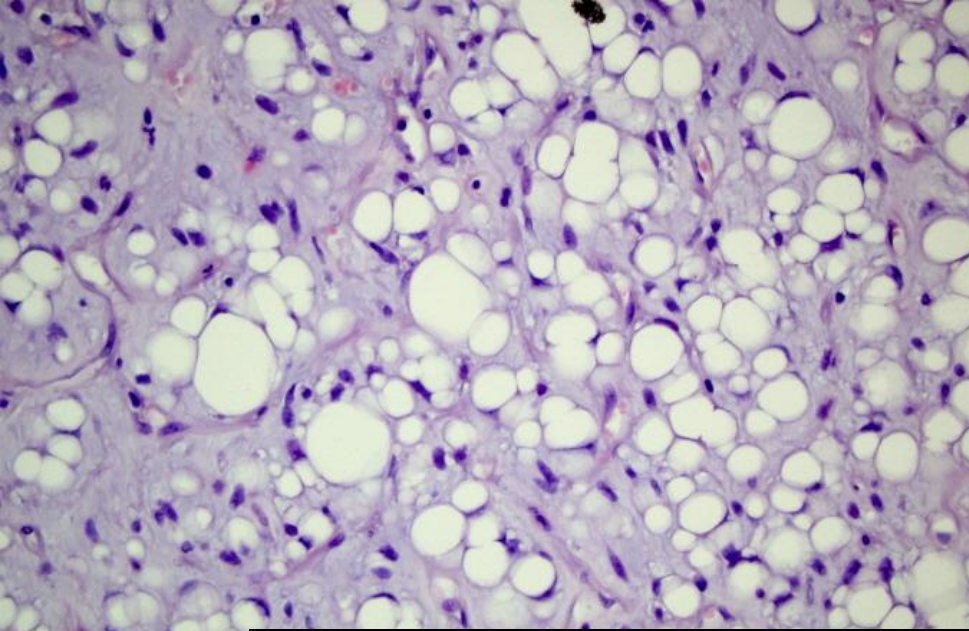
(dediferansiye komponent non lipojenik değil)

## d. İğsi hücreli İDLS

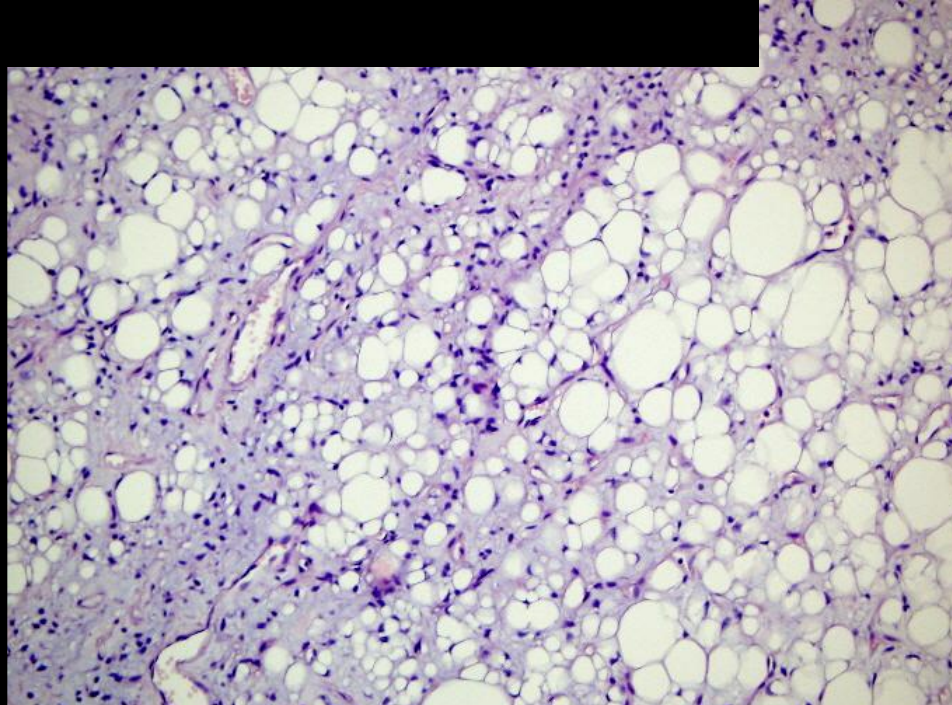
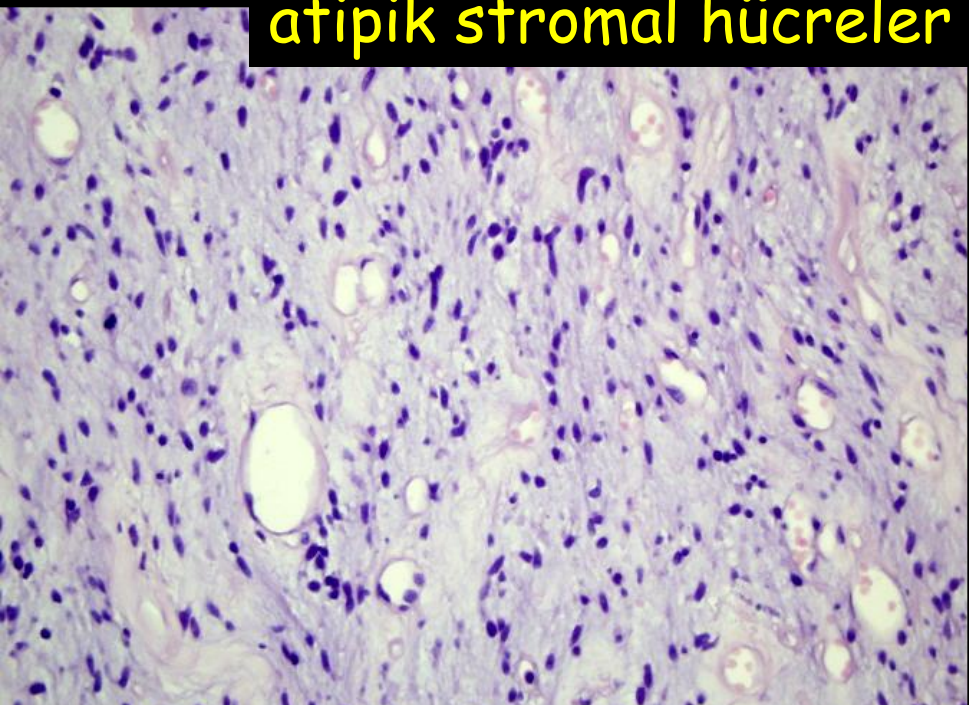


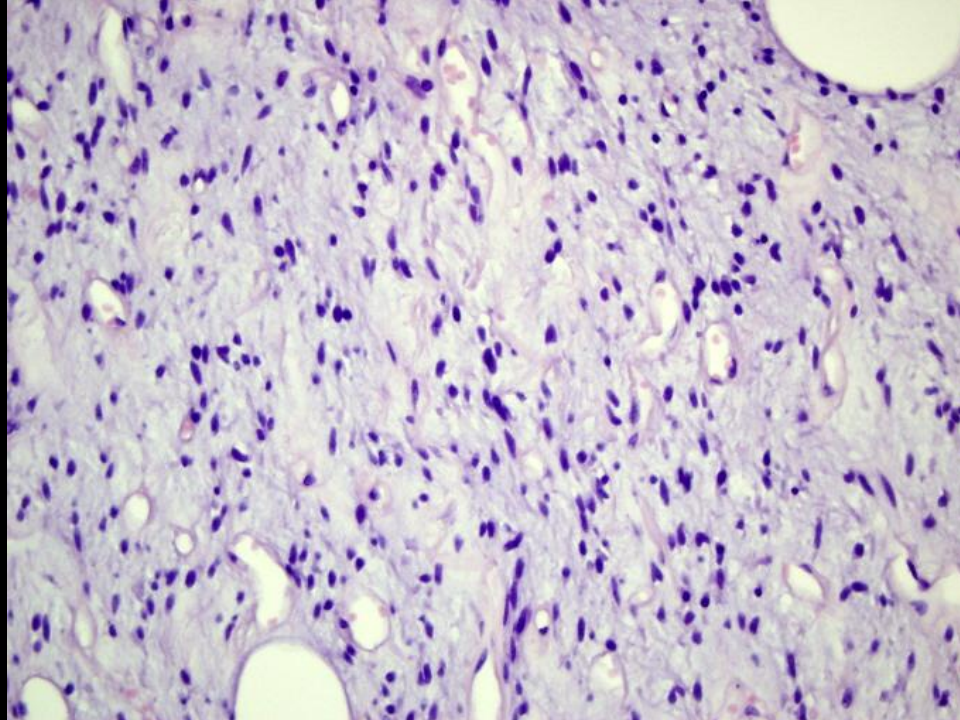
Fibröz ve miksoid zeminde nöral tipte iğsi hücreler ile Lipoblastları içeren atipik lipomatöz komponent



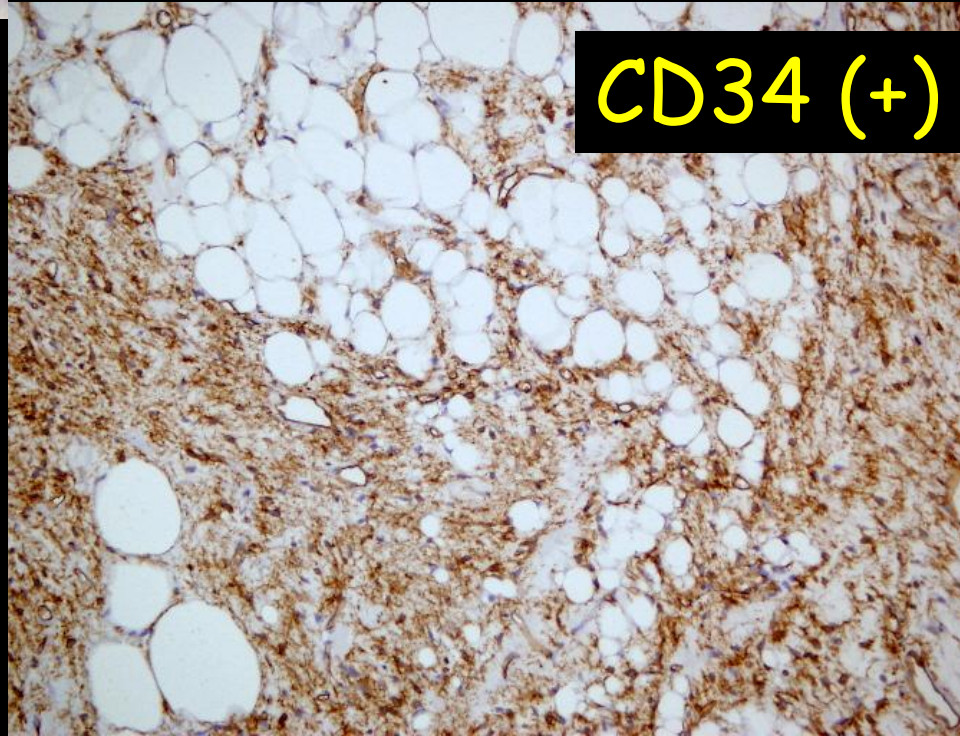
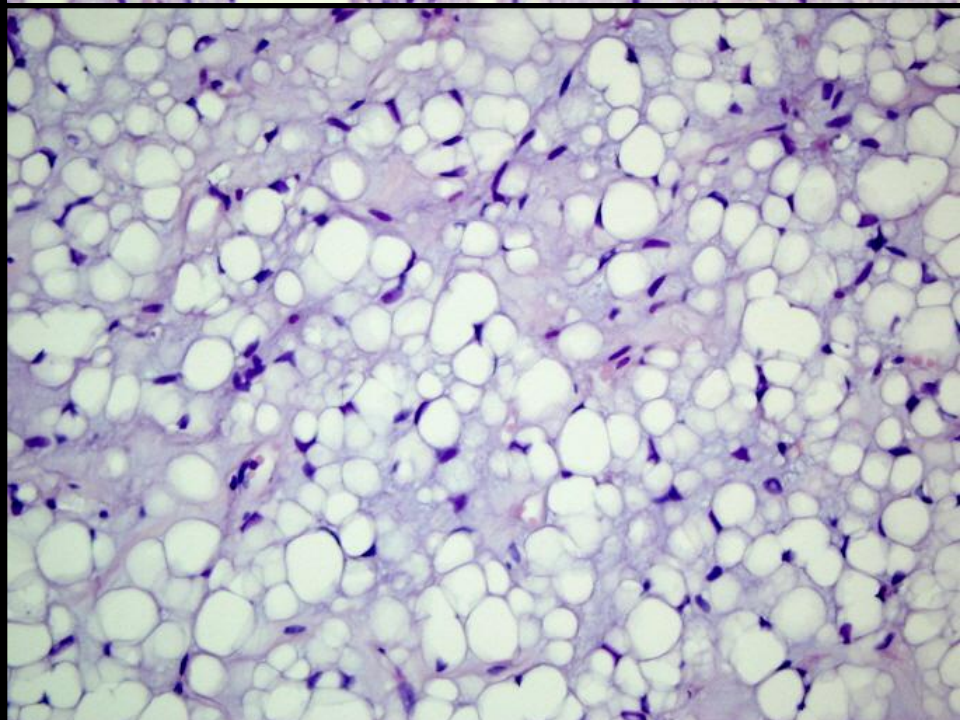


Değişen boyut-biçimde lipositler, lipoblastlar ve atipik stromal hücreler



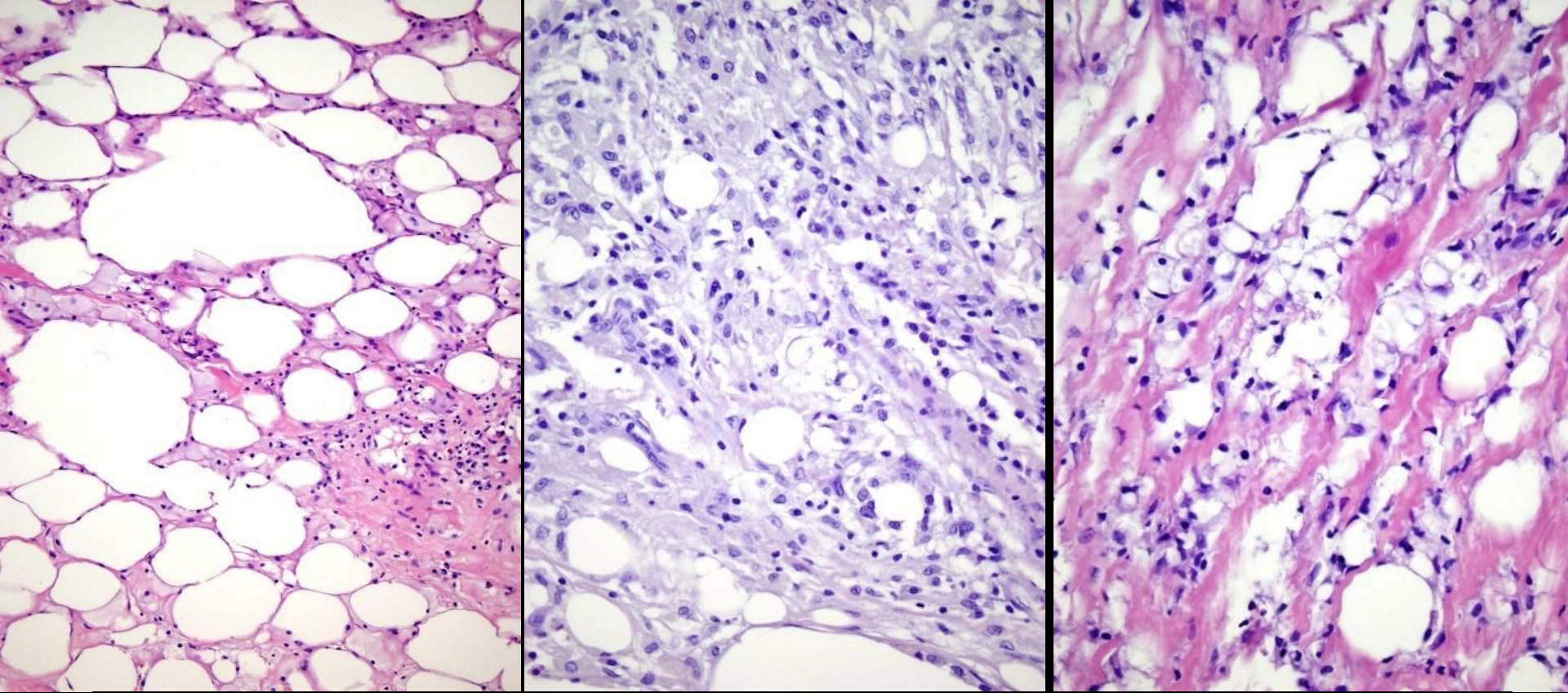


**İH-İDLPS**  
Şekil-boyut farklılığı  
Lipoblastlar ve  
Atipik stromal  
hücreler  
İHL dan ayrılır\*

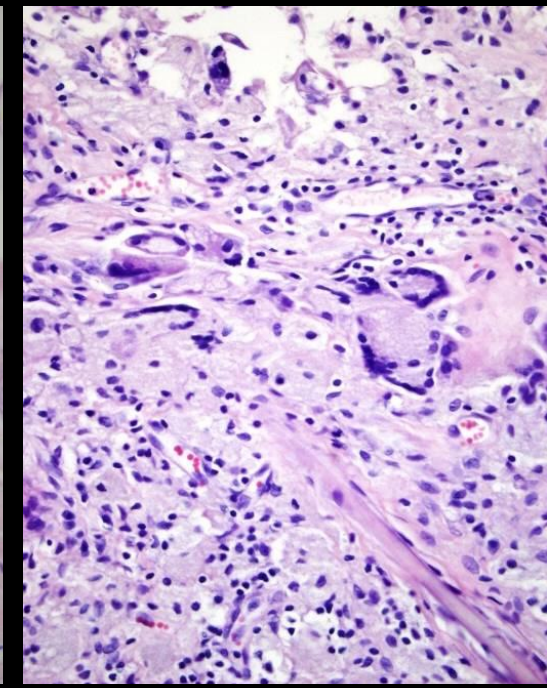
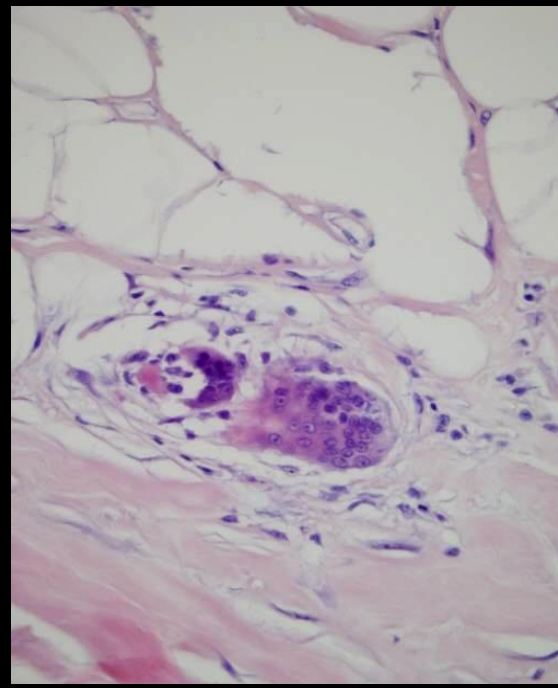
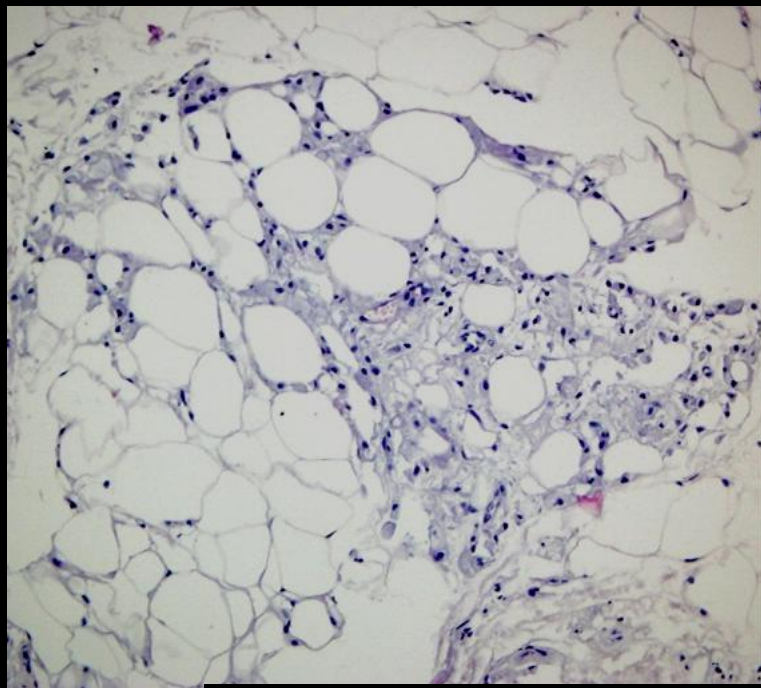


**CD34 (+)**

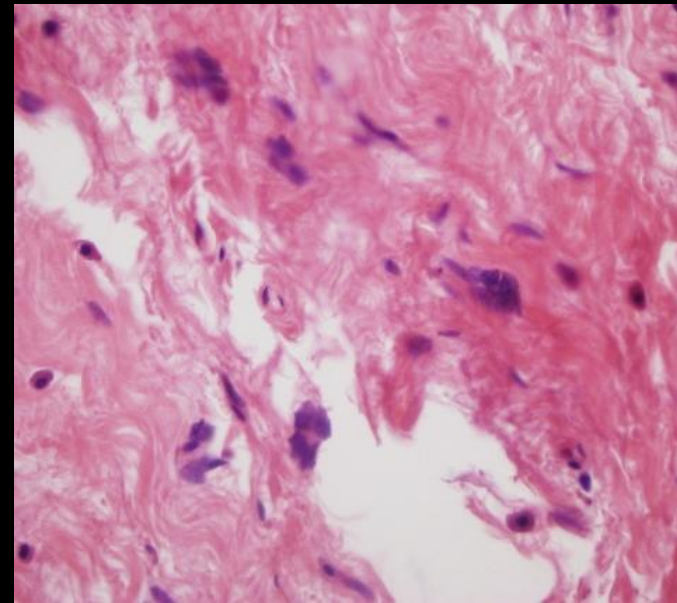
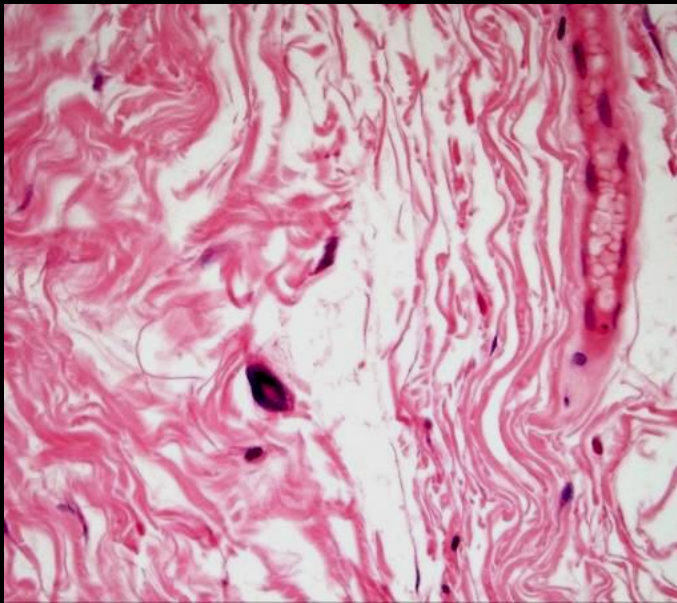
# İDLS da AT; en sık benign lipomatöz tümörler



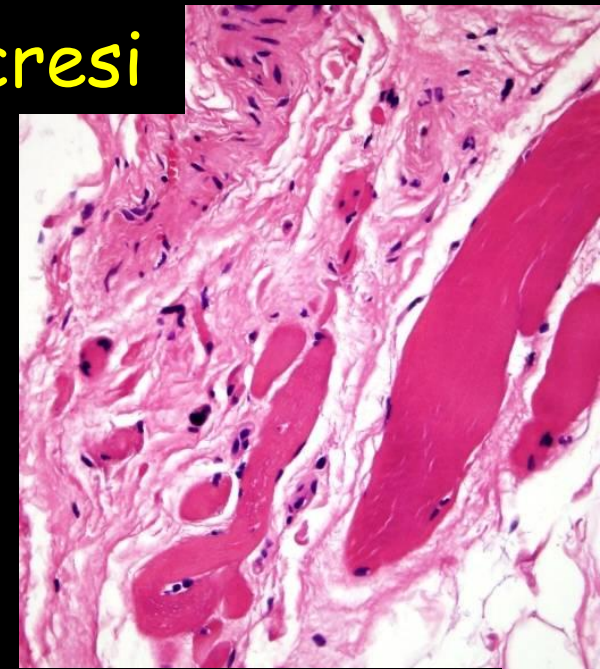
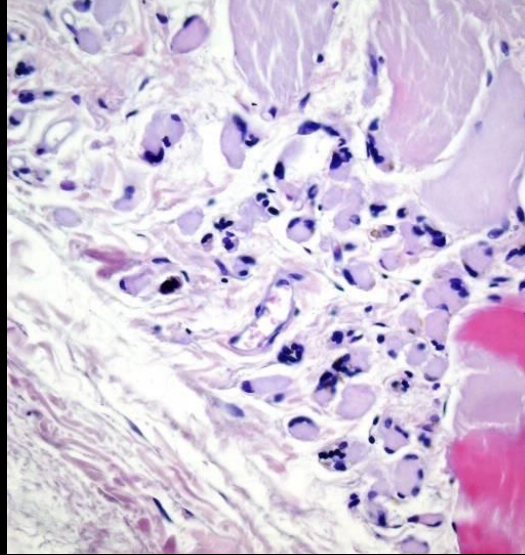
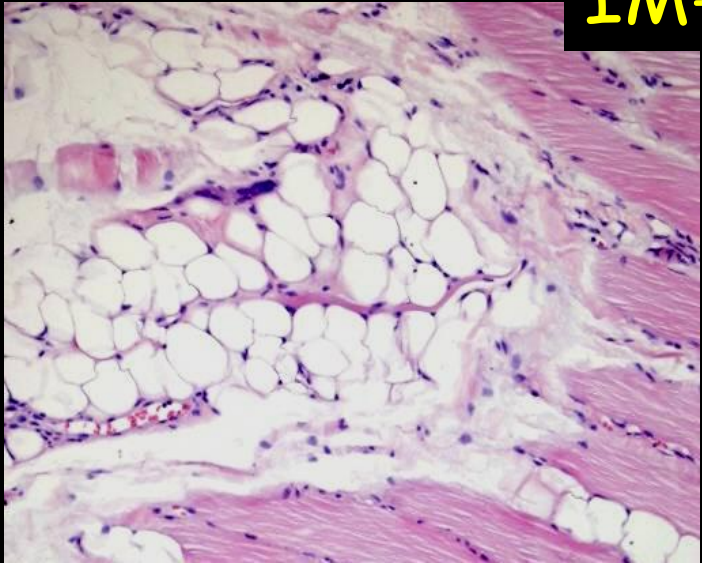
Derin lipomlardaki sekonder deęişiklikler, atipik hücre ve lipoblast benzeri hücre



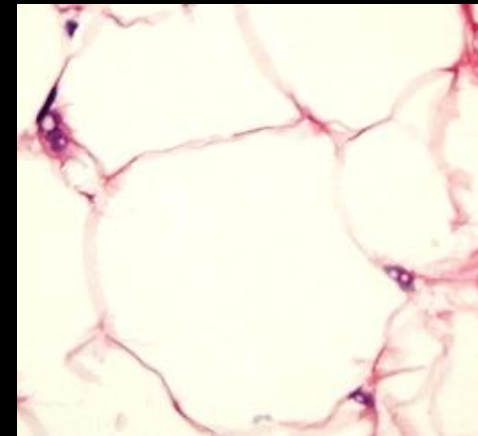
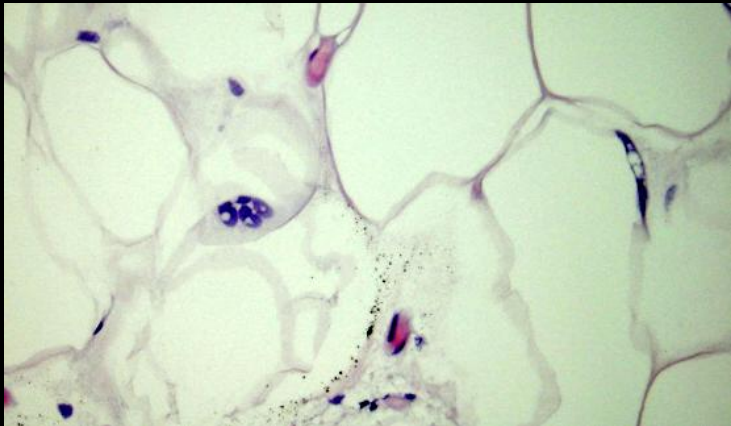
Travma, silikon, İHL/PL lipoblast- atipik hücre



# İM-L atrofik kas hücresi



Dikkatli örnekleme, MDM2, CDK4 (FISH ve İHK)



Lochkern (nükleer vakuol)

ALT/İDLS prognoz;

Metastaz (-)

Lokalizasyona bağlı nüks

(Ekstremitte nüksü < retroperiton nüksü )

Dediferansiyasyon riski

(%5' ten az - %20' den fazla)

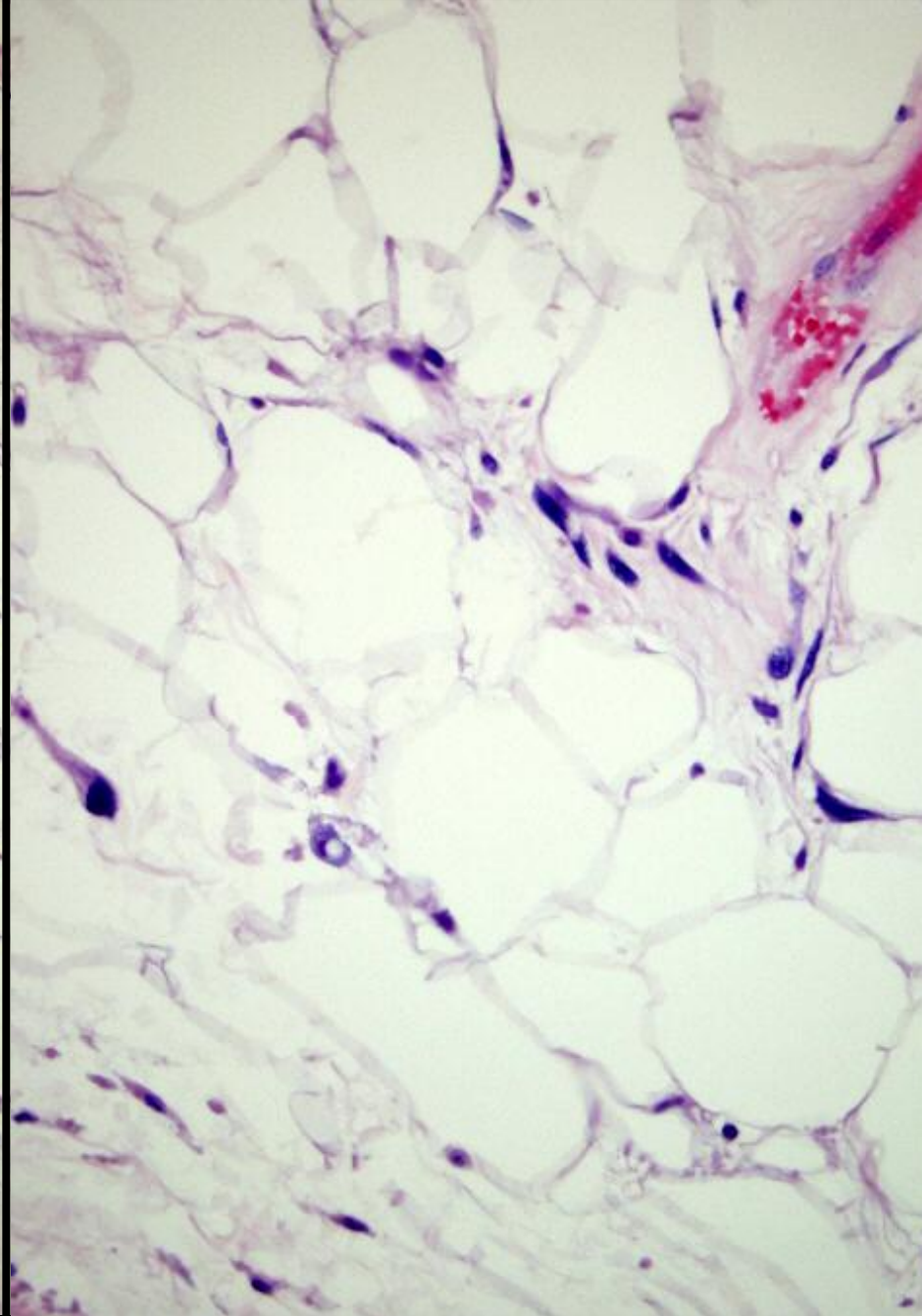
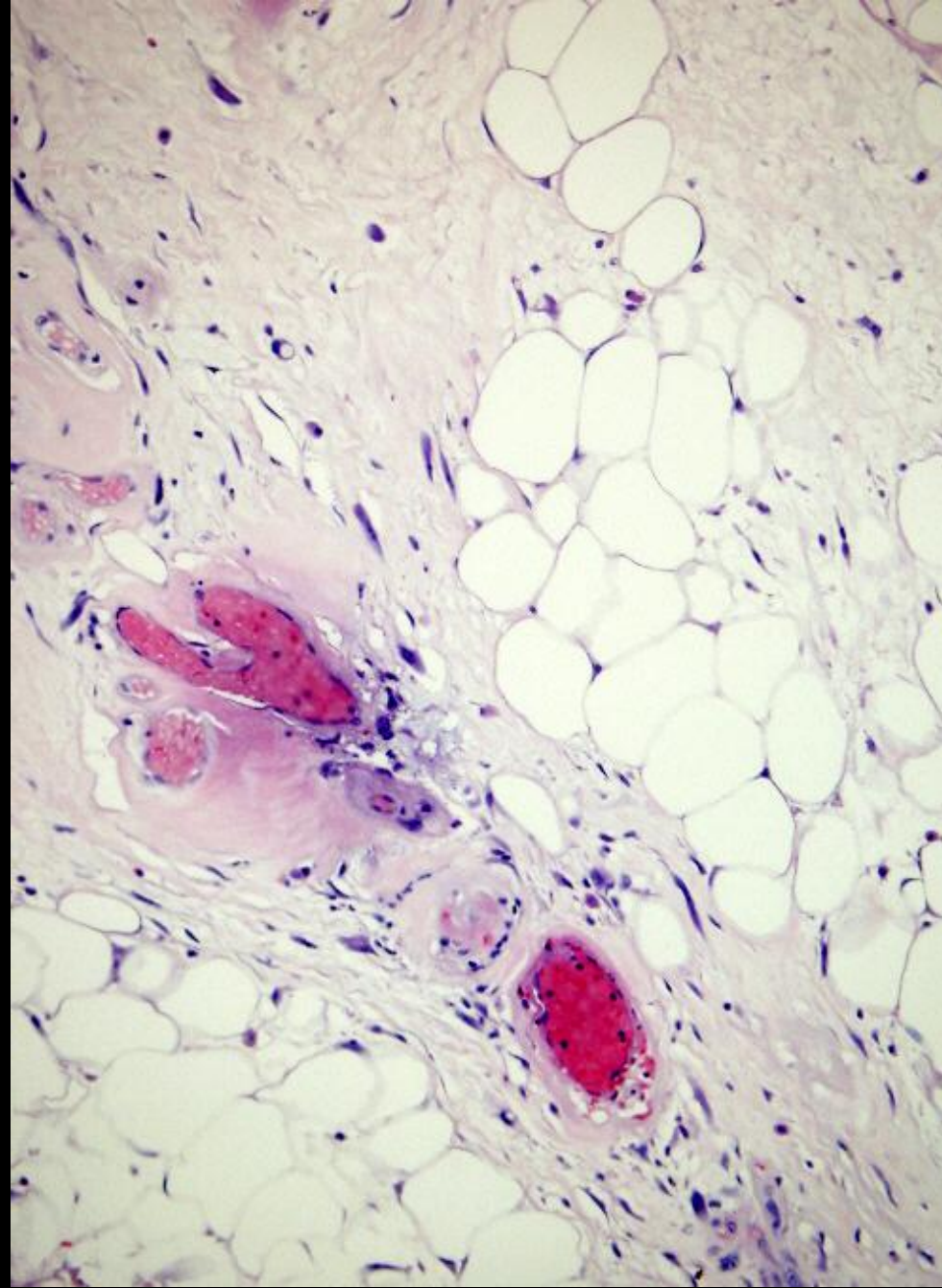
Mortalite

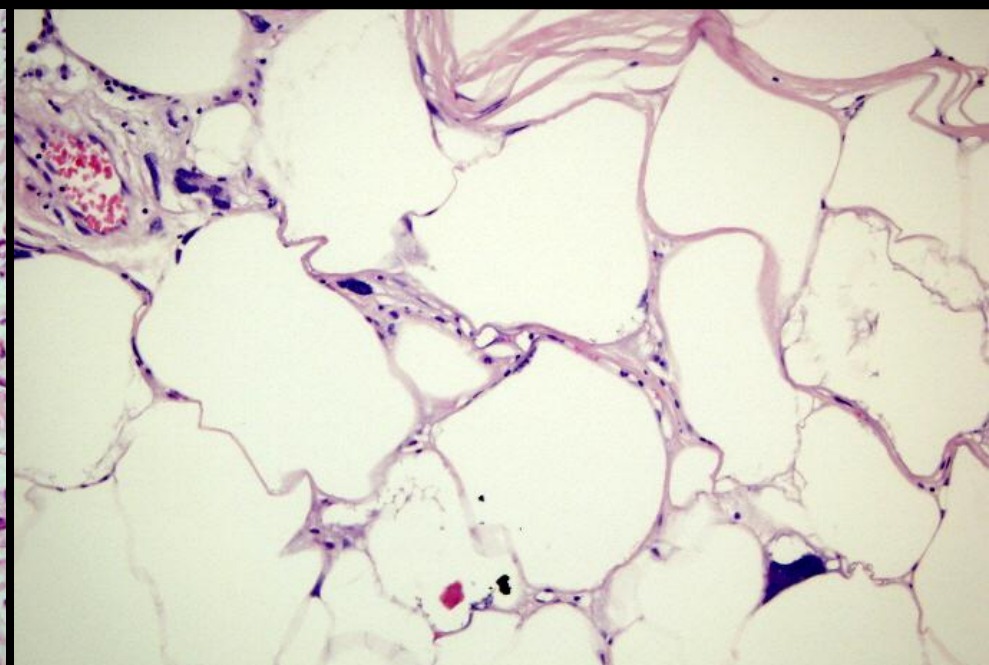
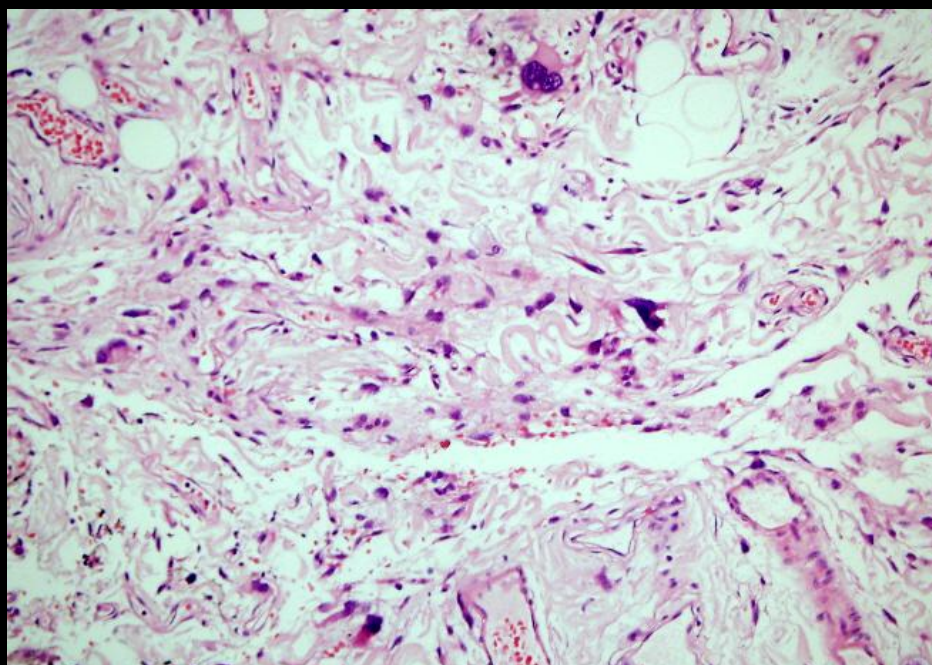
(%0 - %80'den fazla )



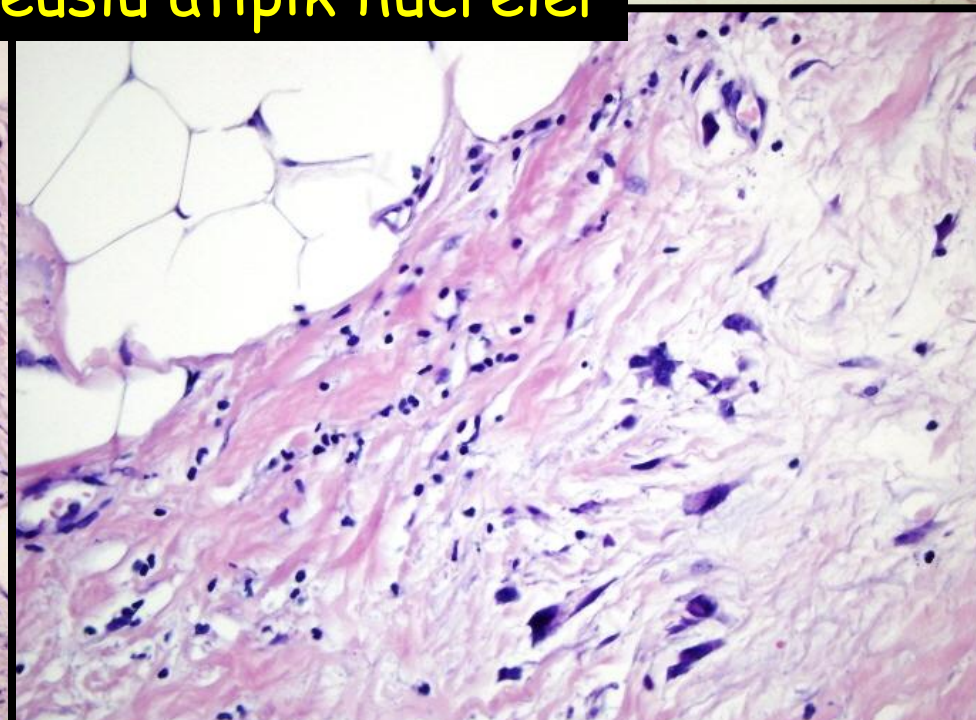
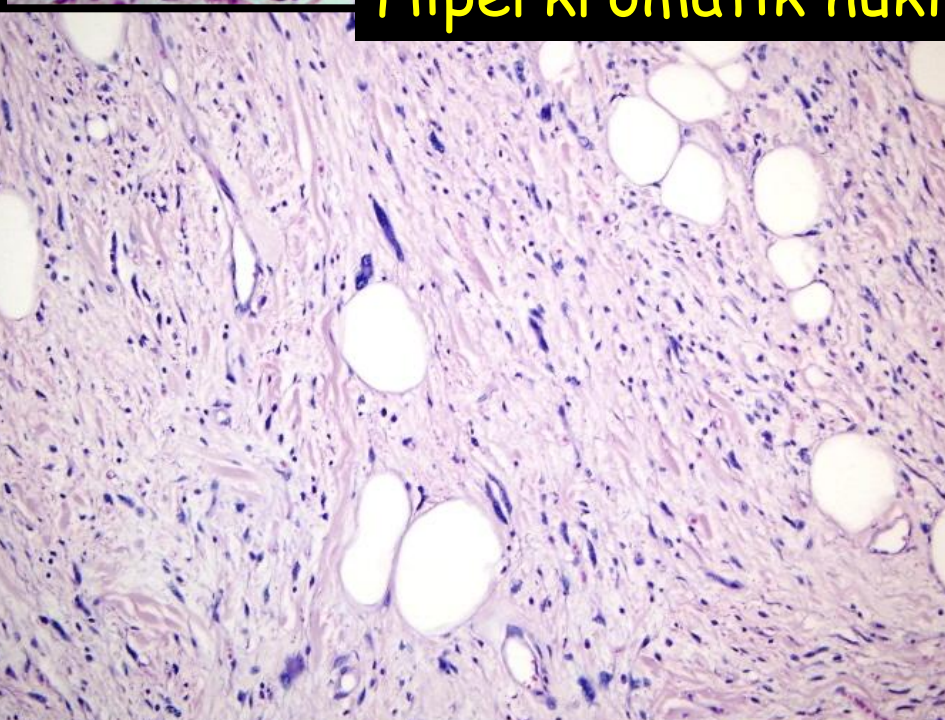
# ALT/İDLS için ne zaman kuşkulalanmalıyız?

- \*Büyük ve derin yerleşimli yağlı tümör
  - \*Retroperitoneal yağlı bir tümör (seyrek istisnalar dışında)
- \*MR da heterogenite (cerrah kuşkulu)
- \*Fibröz bantlar
- \*Küçük büyütmede atipi (hiperkromazi)
  - \*Lipoblast arama!!





**Hiperkromatik nükleuslu atipik hücreler**



**Tanı için lipoblast gerekli değil!!**



## Dediferansiye LS

ALT+ sellüler nonlipojenik sarkom (10 BBA da en az 5 mitoz)

İDLS içeren YD non-  
lipojenik sarkom  
İDLS dan yıllar sonra  
gelişir.

(Non lipojenik sarkom DD  
ob)

İDLS alanları (-) (%18)  
(yetersiz örnekleme,  
ortadan

kalkması, hiç olmaması)

Genomik kanıtlı

DDLs %90 de novo, %10  
nükslerde



DD riski derin yerleşimli tümörlerde  
yüksek (retroperiton)

Retroperiton (%80), ekstremiteler,  
spermatik kord, gövde.

Baş-boyunda gb, subkutan dokuda seyrek

Retroperitoneal sarkomların %60'ı LS  
(2/3'ü DDLS)

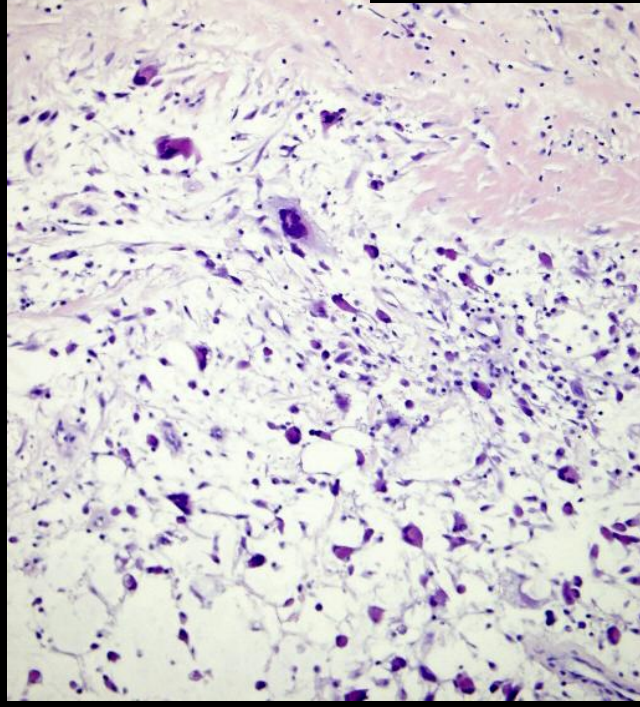
Radyolojik olarak tanınabilir



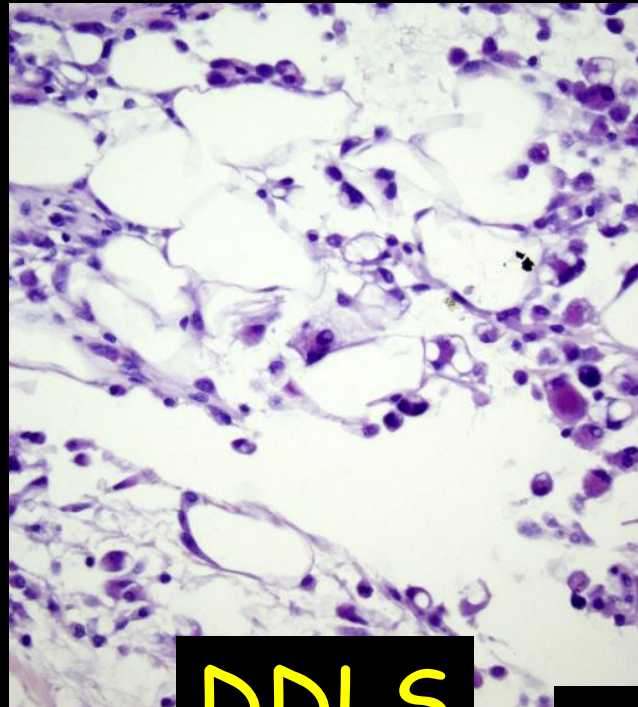
**Makroskopik GBR solid alanlar içeren multinodüler sarı renkli kitleler**



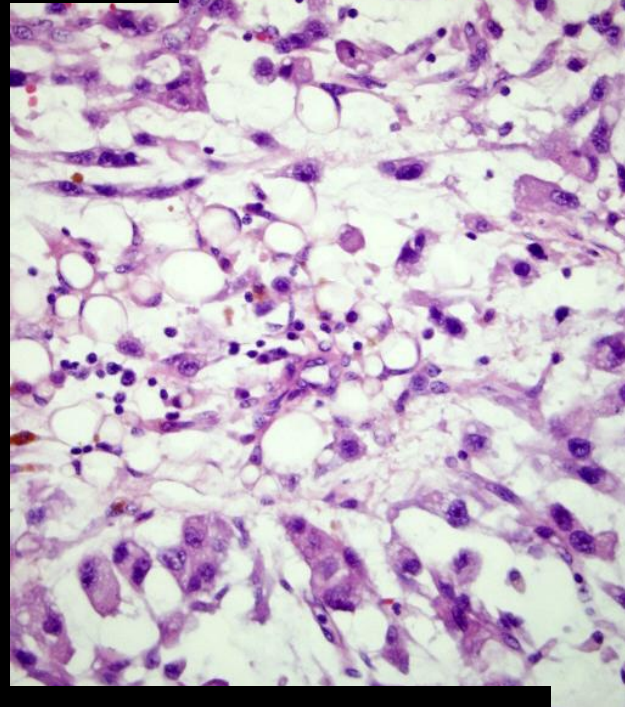
# Myojenik diferansiyasyon



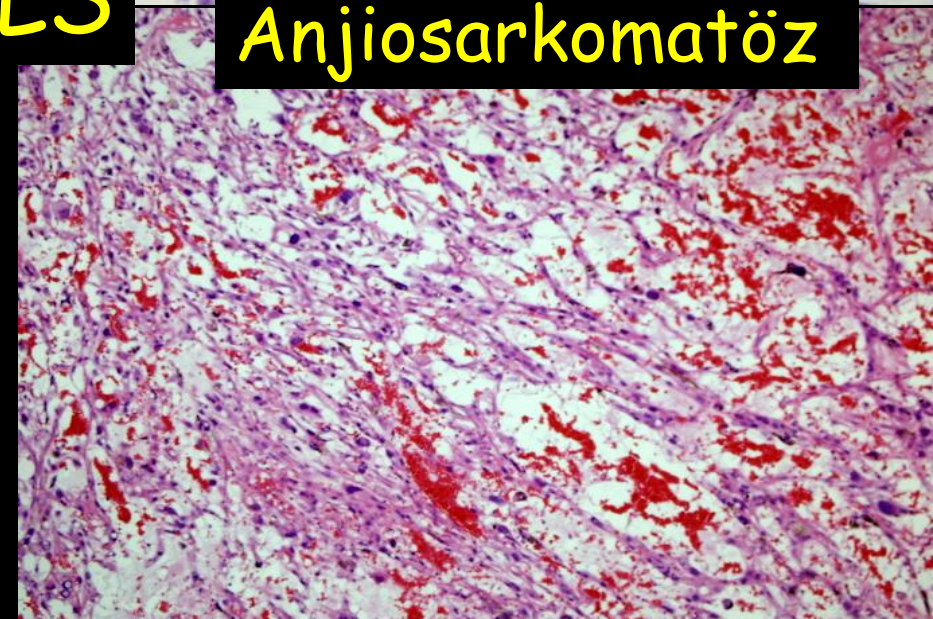
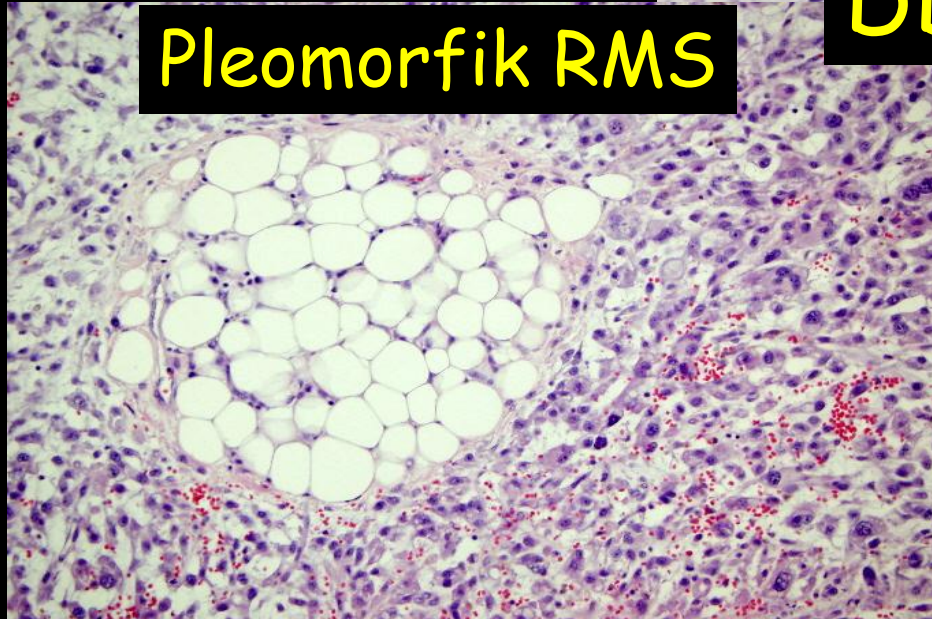
Pleomorfik RMS



DDLs

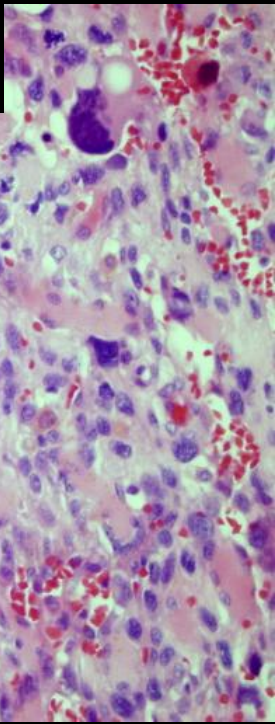


Anjiosarkomatöz

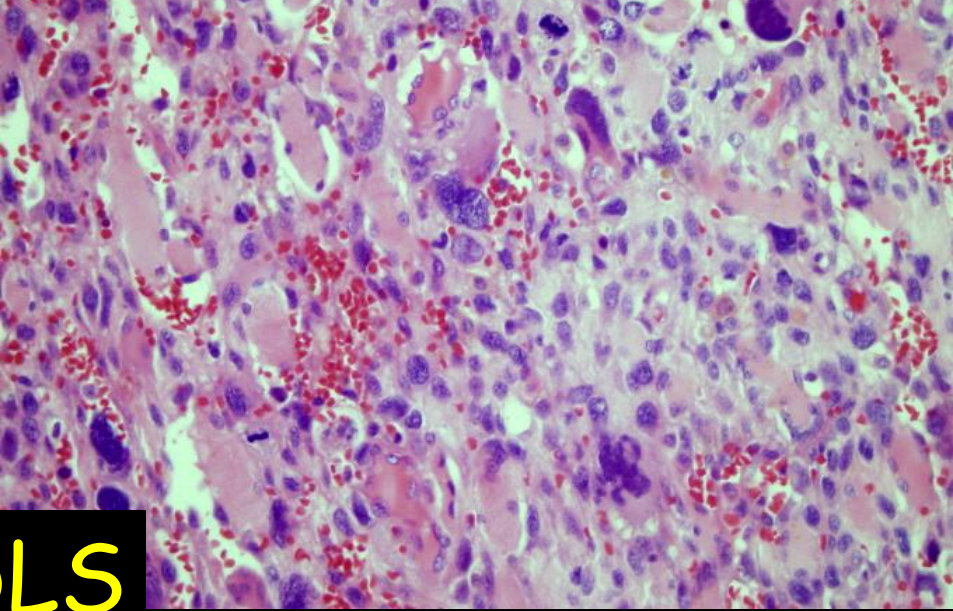




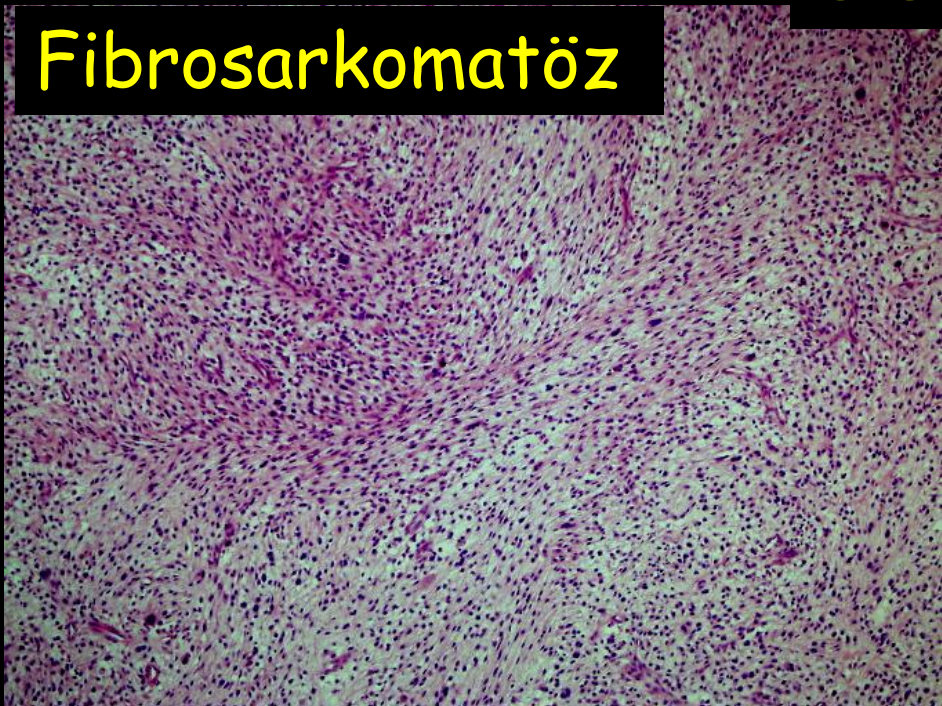
Pleomorfik sarkom



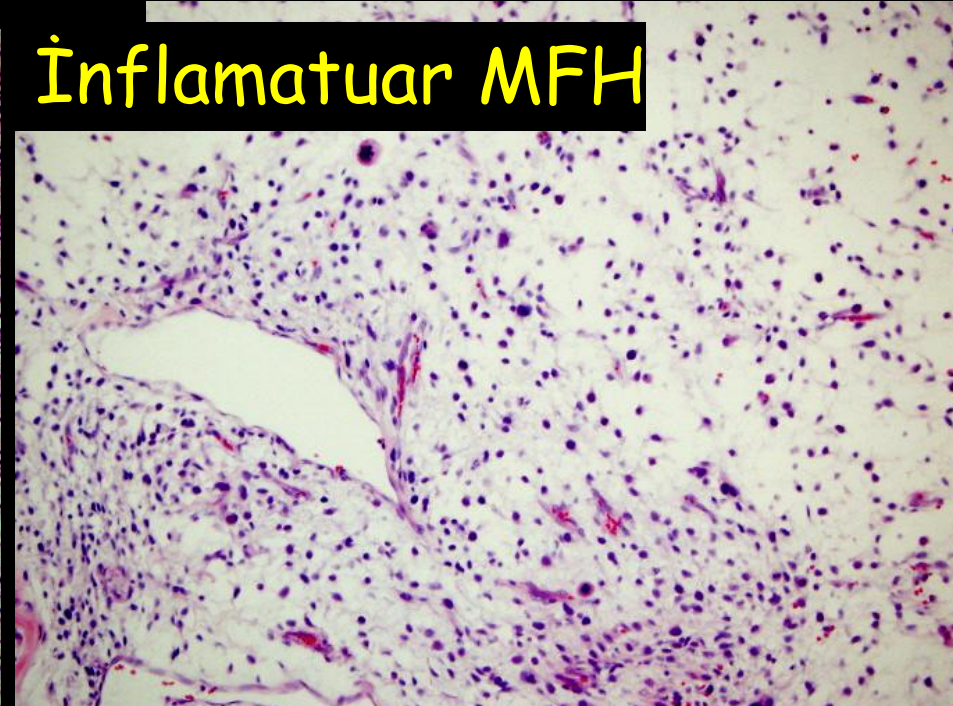
DDLS



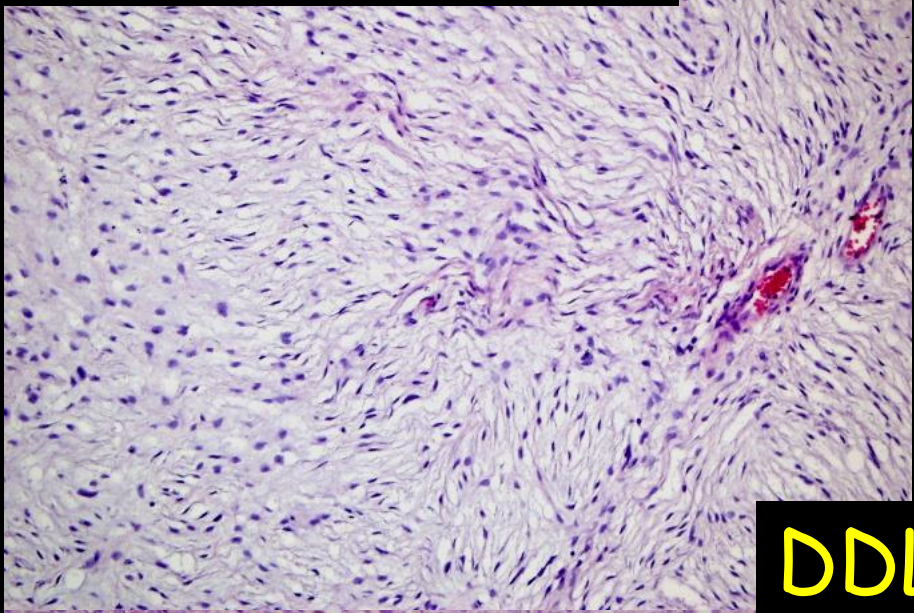
Fibrosarkomatöz



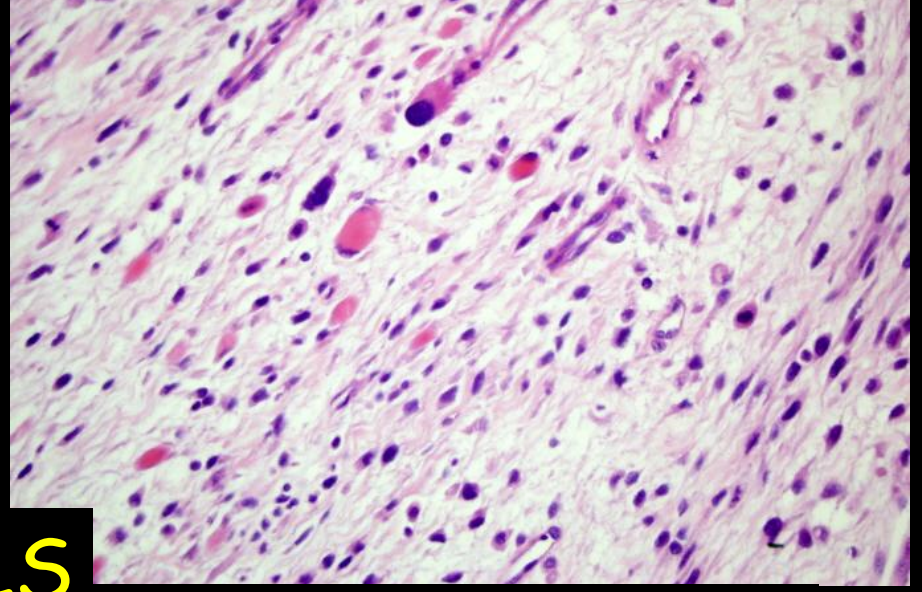
Inflammatuar MFH



Düşük dereeli DDLS

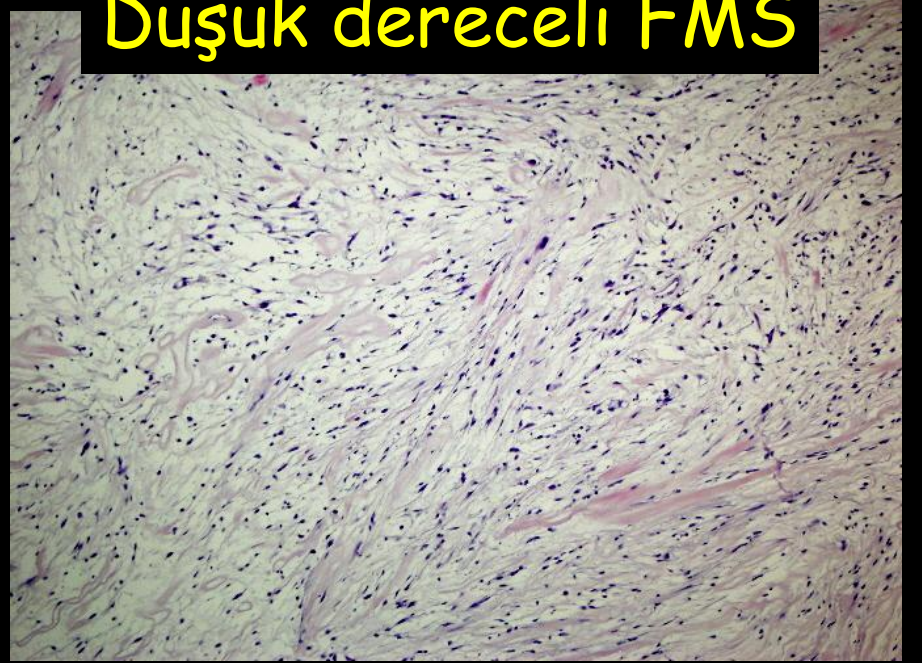
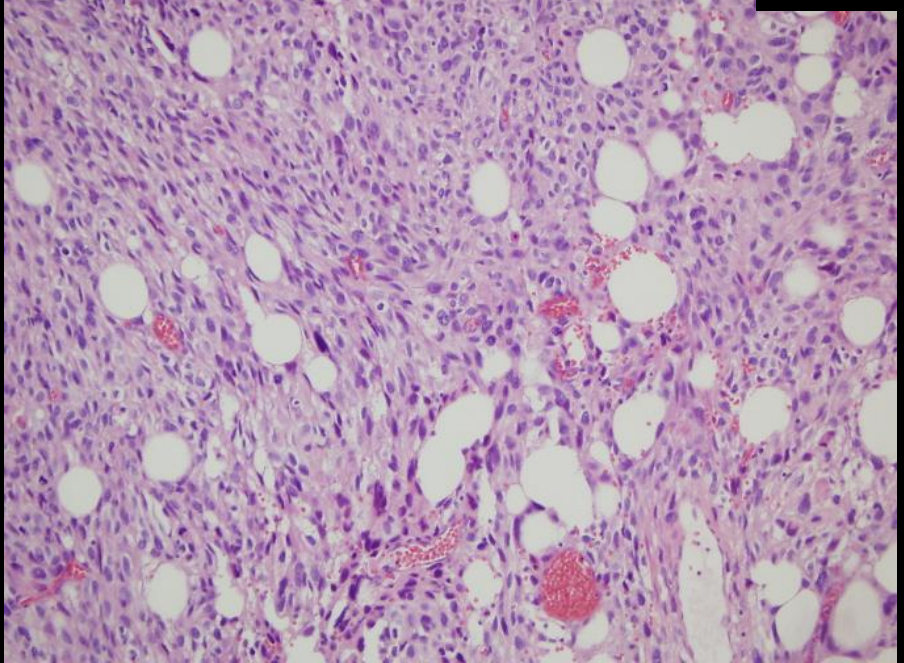


Rabdomyoplastik dif



DDLS

Düşük dereceli FMS

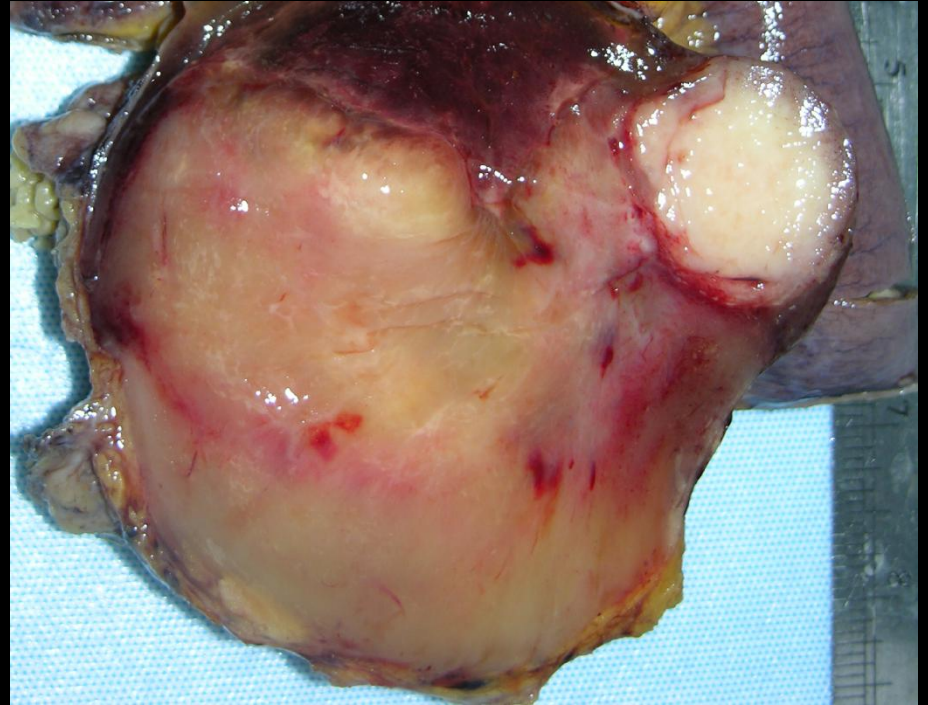


## DDLS AT;

MFH, FS, M-HMP,  
MiksoFS, P- RMS  
Gövde lokalizasyonunda  
bu tanılardan önce,  
DDLS ekarte edilmeli!!  
yaygın örnekleme  
MDM2 ve CDK4

IDLS da minimal DD

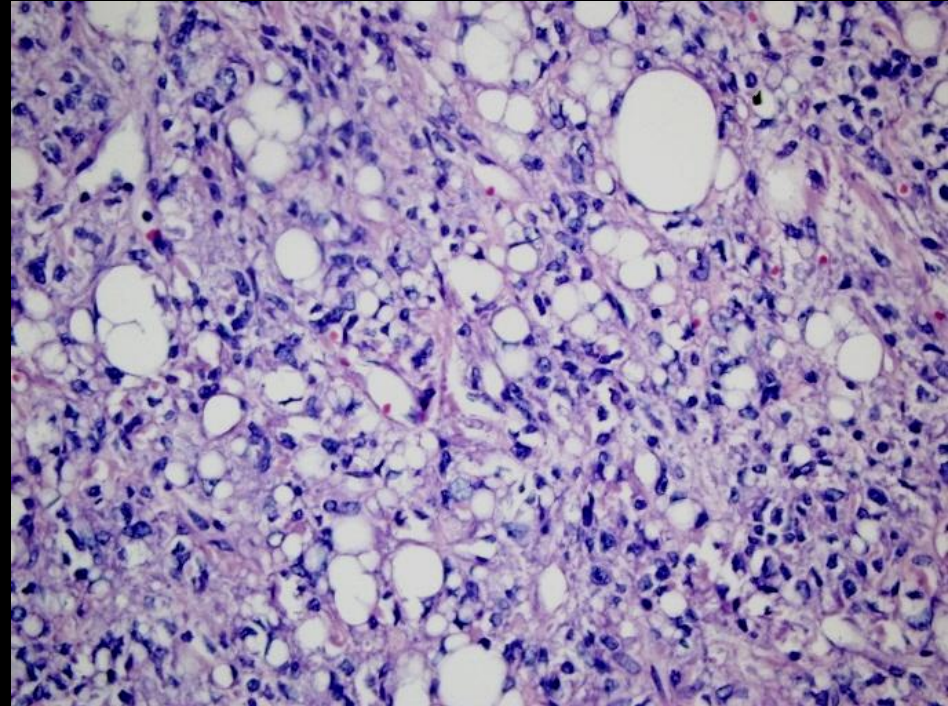
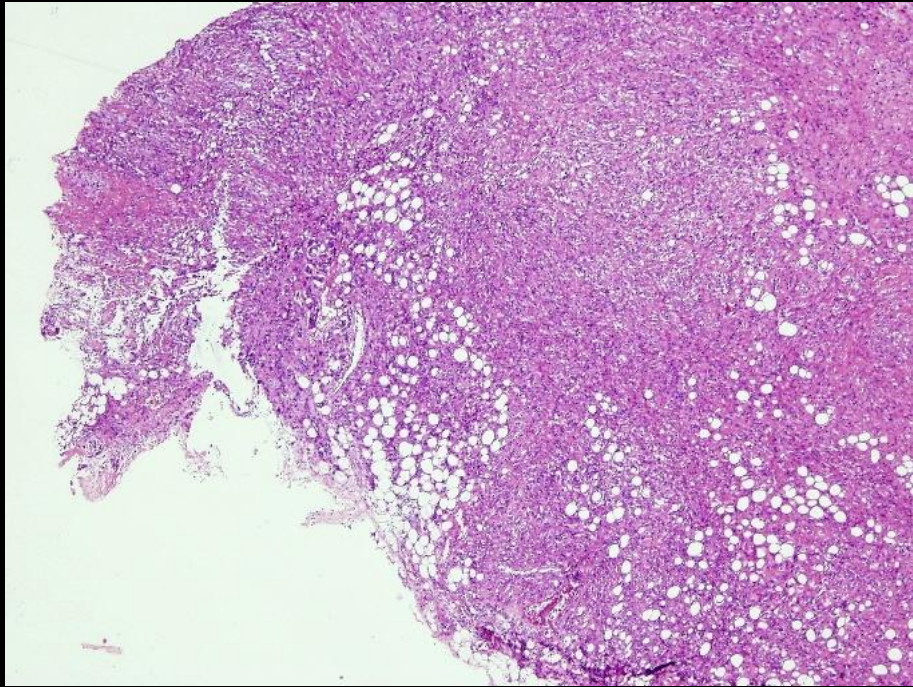
Makroskopik >1 cm nodül



DDLS da AT da diđer bir sorun

Yađı infiltrate eden sarkom;

Yađı infiltrate eden Pleomorfik sarkom ile DDLS ayırımında ,dediferansiye alanlarından uzakta ALT/İDLS alanlarını aramaktır.



Prognozda lokal nüks önemli  
(retroperiton %40-60)

Uzun süreli takipte %100 nüks

Metastaz potansiyeli düşük %15-20

Mortalite %30-40 (5 yıllık)

En önemli prognostik faktör

Ne histolojik derece ne de DD yaygınlığı  
anatomik lokalizasyon!!

# Miksoid -yuvarlak hücreli LS

2. en sık varyant

(LS'ların 1/3'ü, tüm sarkomların %10'u)

Ekstremiteler

Spesifik genetik bulgu+

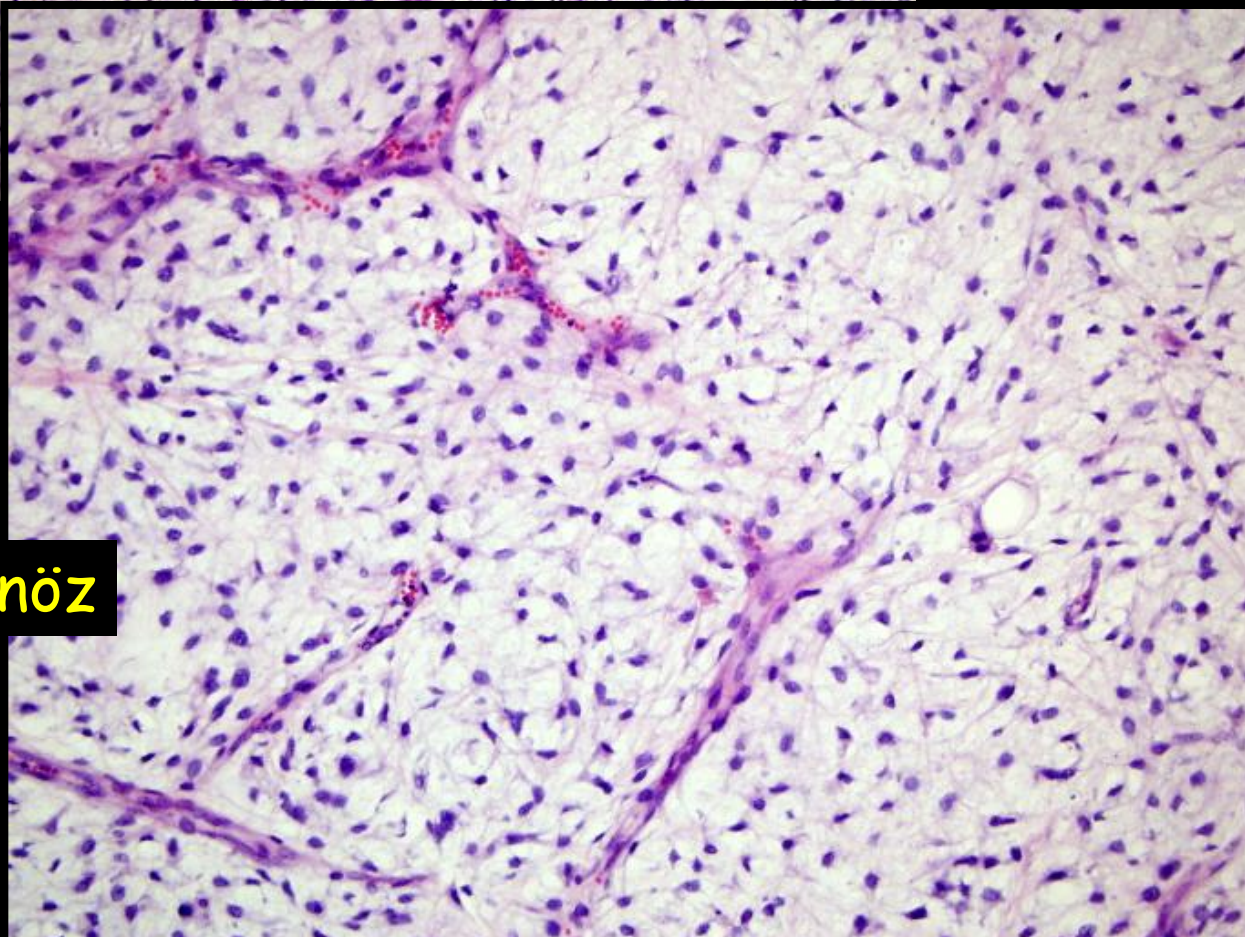
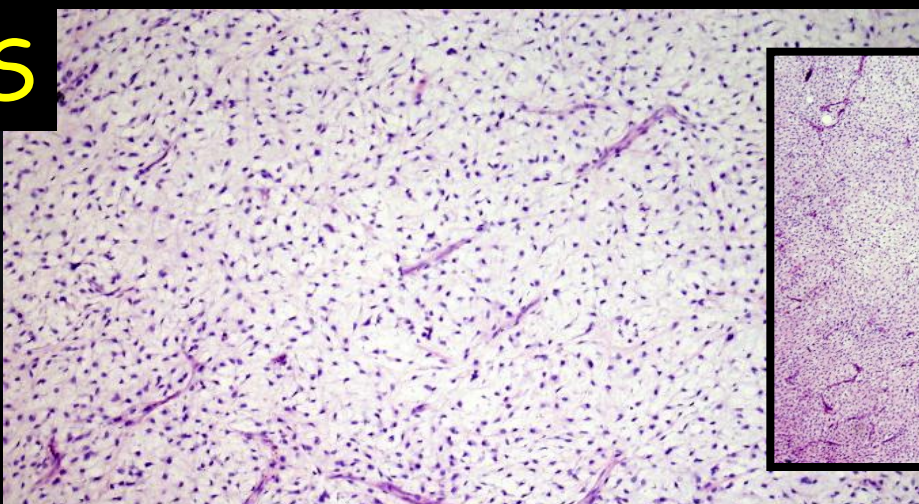
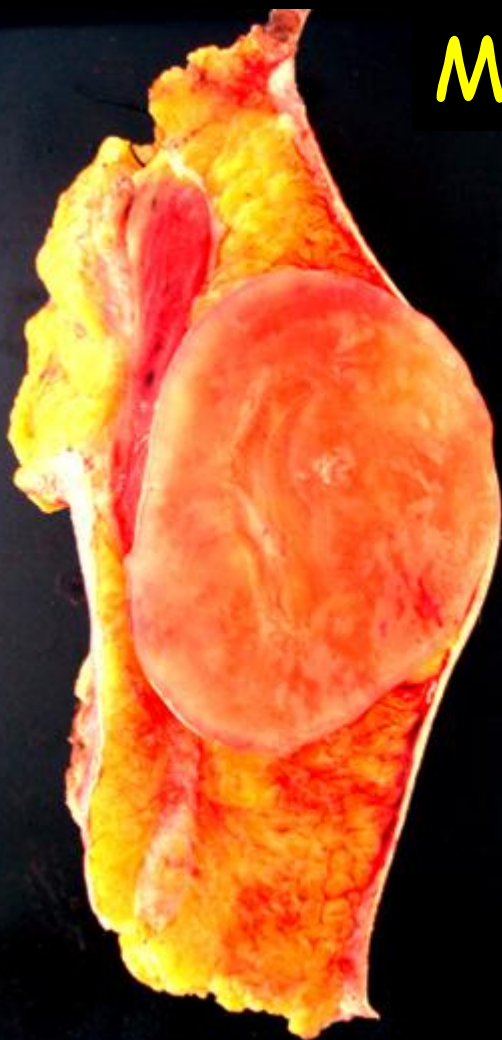
Pür miksoid ve yuvarlak hücreli LS bir spektrumun farklı iki ucu

Klasik MLS

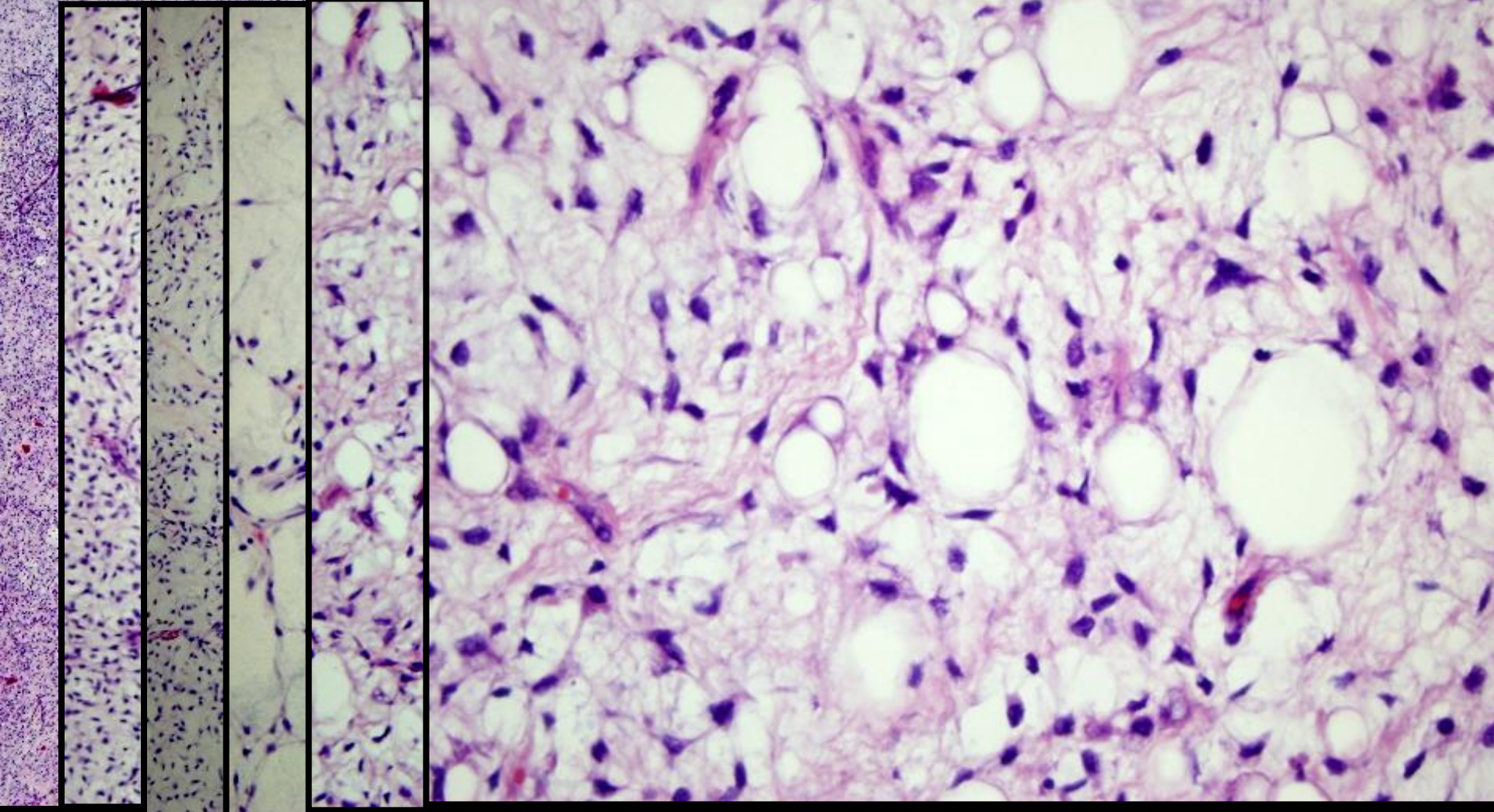
Mikst Miksoid-yuvarlak hücreli

Pür yuvarlak hücreli (çok seyrek)

MLS



Multinodüler, jelatinöz



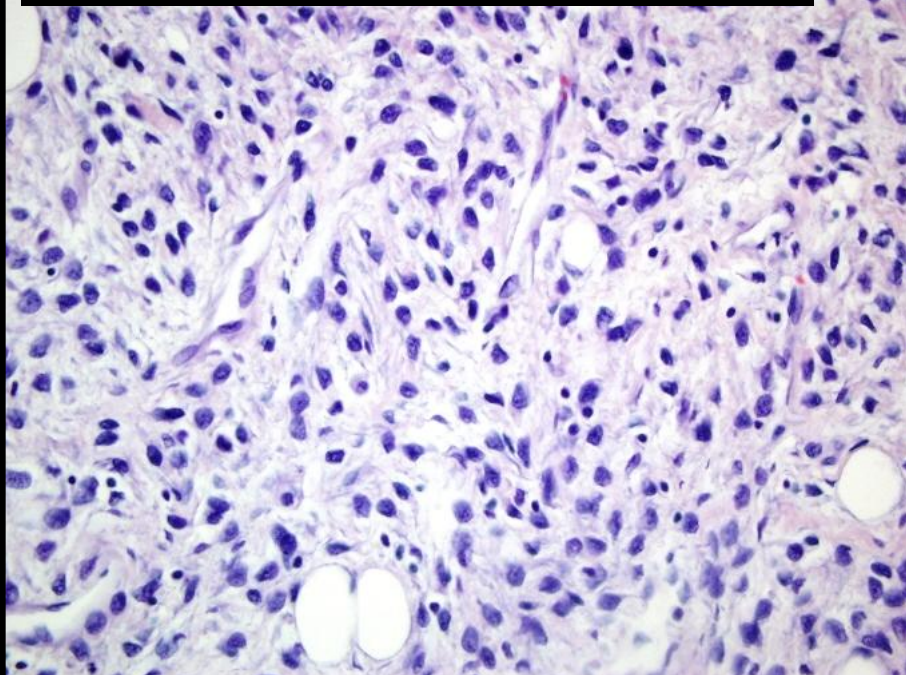
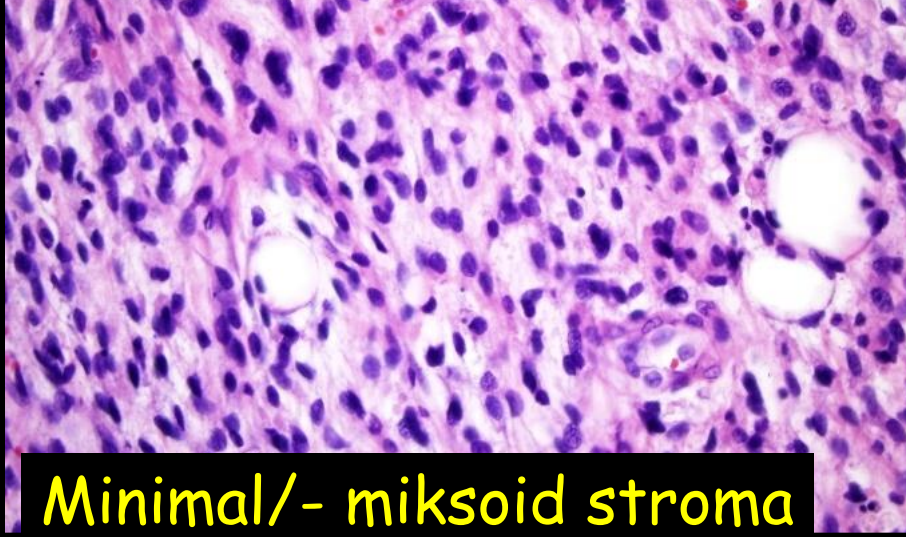
Ekstrasellüler miksoid zemin, uniform oval-yuvarlak primitif mezenkimal hücreler, ince vasküler ağ ve lipoblastlar



**MLS;** 5 yıllık yaşam %90, multifokal prognoz kötü  
Kemik ve yumuşak dokuya metastaz  
%5-25 yuvarlak hücre= yüksek derece ve  
kötü prognoz

AT; miksoma, anjiomiksom, M-DFSP,  
M-MFH\*, (Atipi, psödolipoblast )  
M-KS\* (kordonlar, psödoasiniler oluşturan  
küçük eozinofilik sitoplazmalı hüç,  
H&E zemin soluk mavi yerine  
Açık renklidir  
M-DFSP; yüzeysel, lipoblast yok

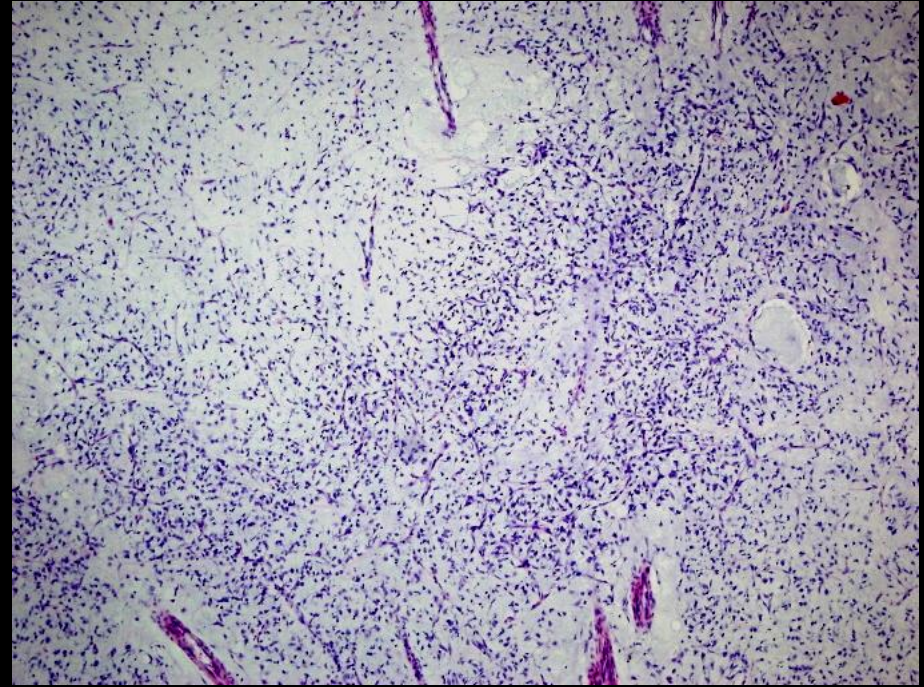
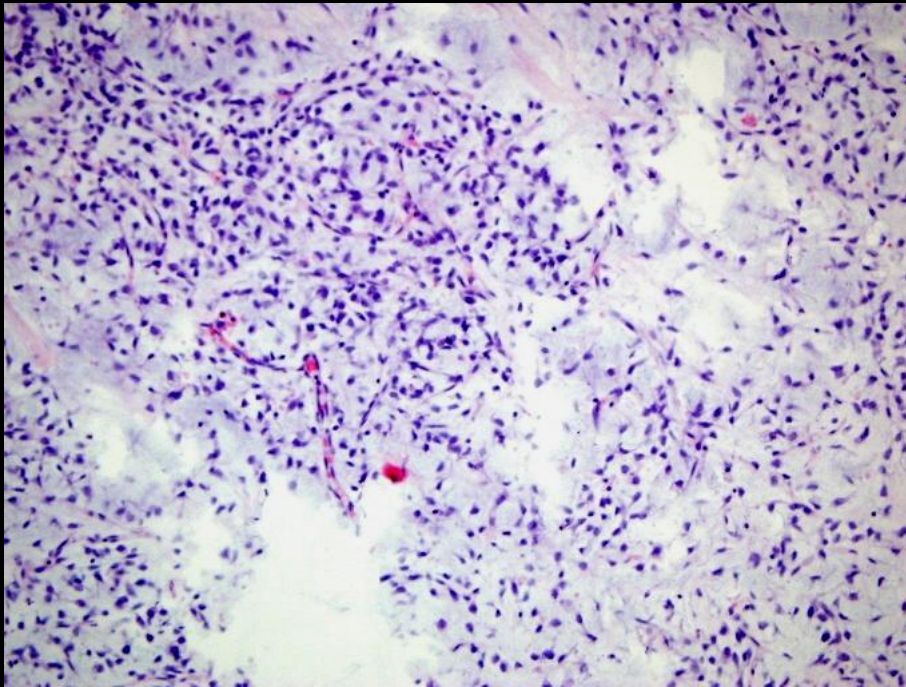
# Yuvarlak hücreli LS için kriter



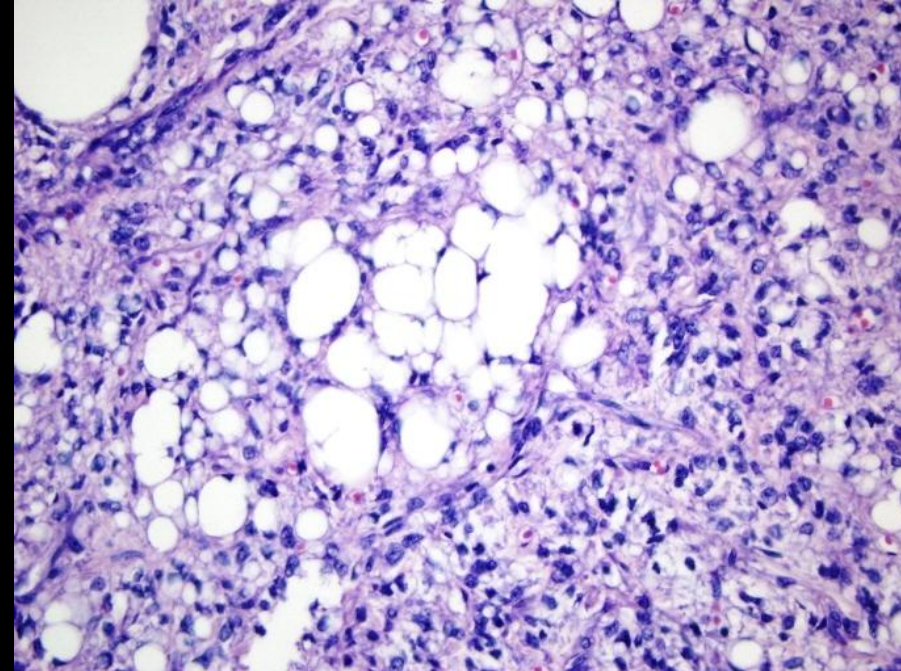
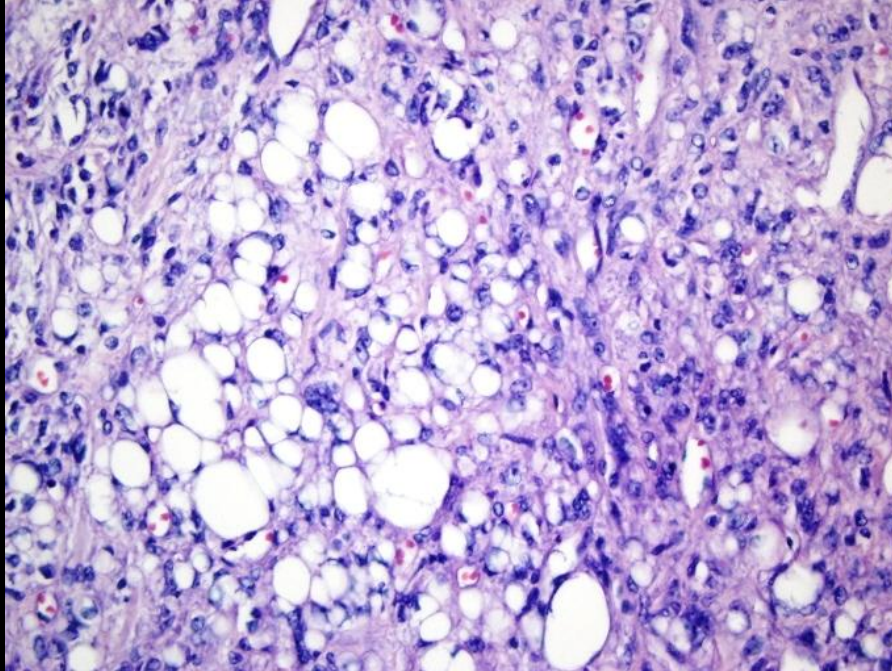
## Hipersellüler/transizyonel alan

MLS dan daha sellüler ancak YHLS kriteri için yetersiz

Bu alanların kanıtlanmış prognostik önemi yok  
Ancak Derece II-III



**Pleomorfik LS;** %5, agresif, ileri yaş  
Alt-üst ekstremiteler, gövde, retroperiton,  
inguinal bölge ve baş-boyun  
Miksoid MFH benzer.



# Niçin hala tanısal sorunlar yaşıyoruz?

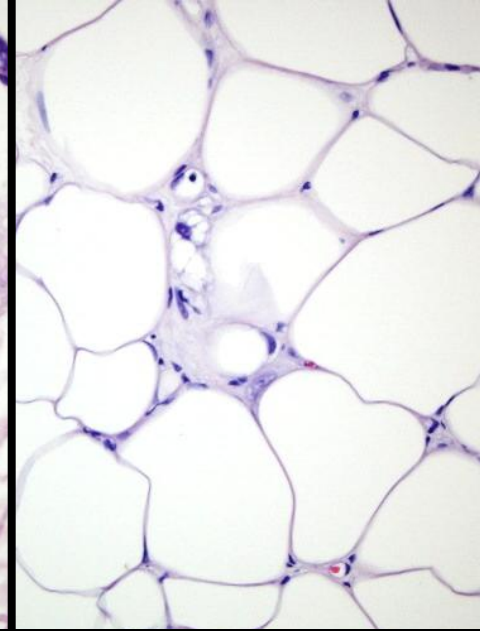
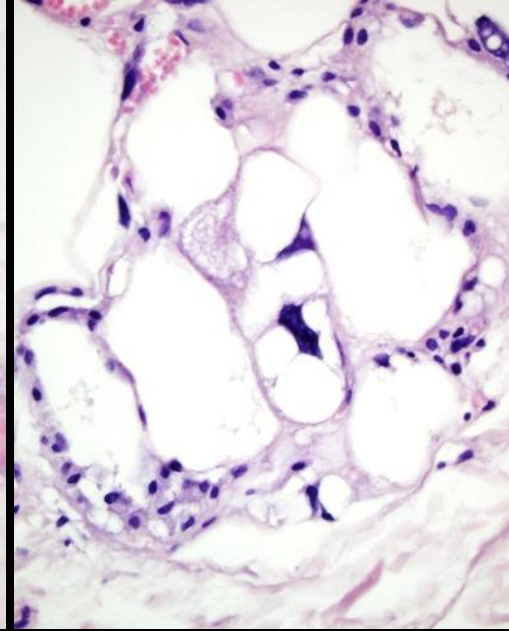
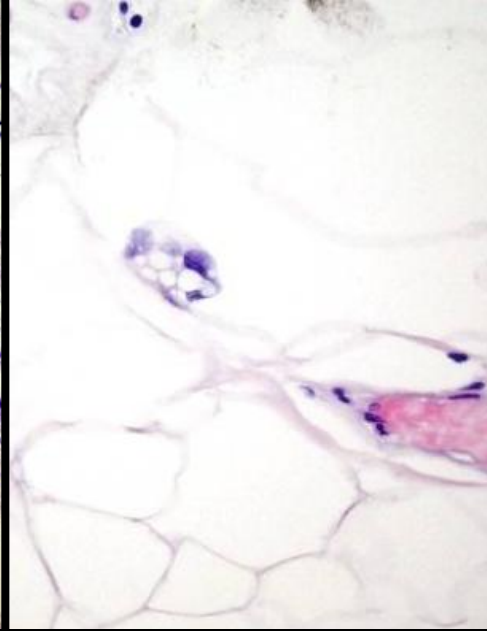
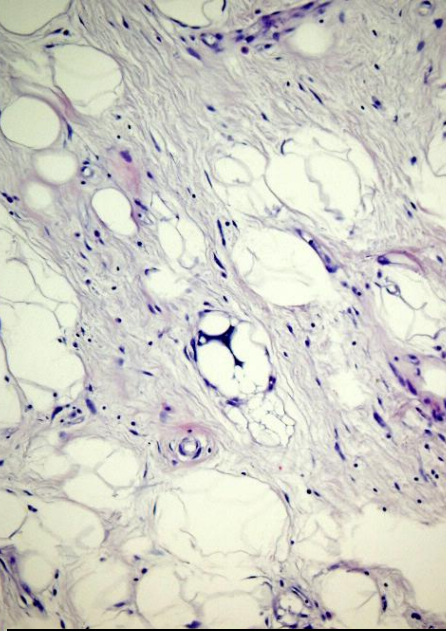
Seyrek ve kompleks lezyonlar

LS tanısı = lipoblast varlığı ve tanıda lipoblastın gösterilmesi gerektiği düşüncesi

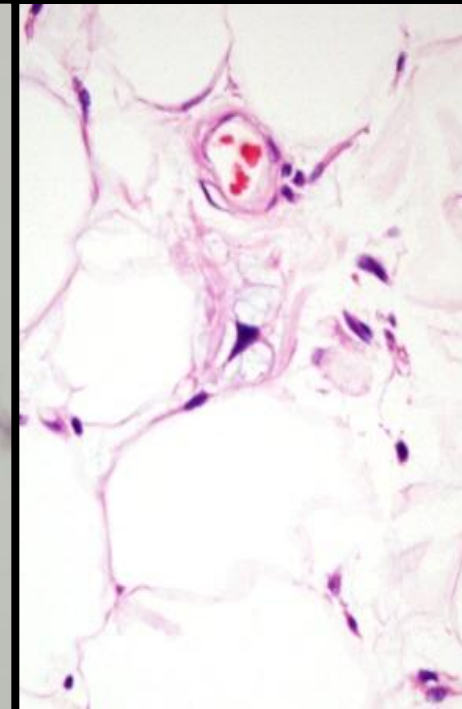
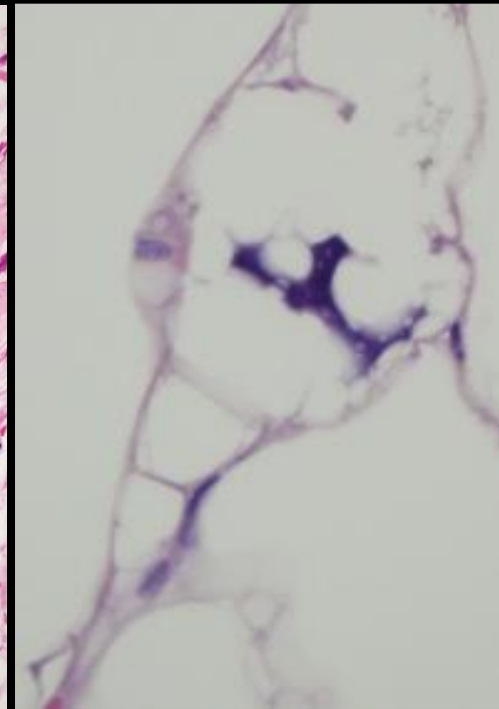
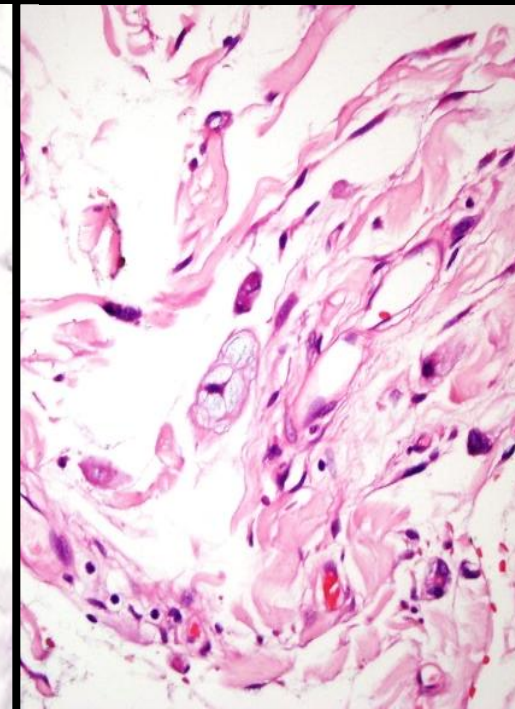
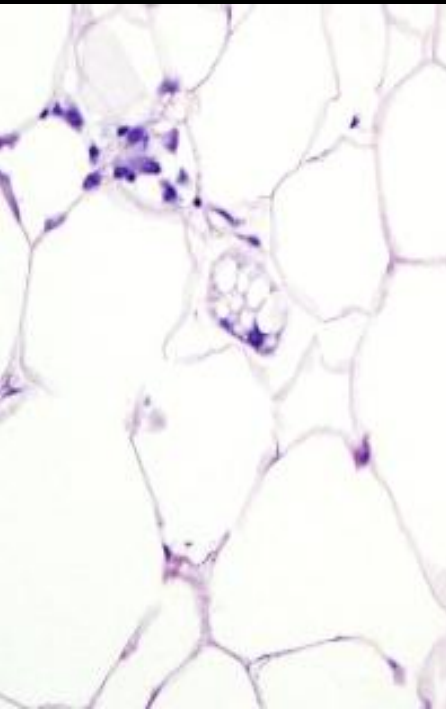
İHL/PHL, kondroid lipom ve lipoblastom gibi benign lezyonlarda lipoblast (+)

İDLS, bazı MLS ve YHLS da lipoblast (-)

MFH benzeri PLS da çok seyrek bizar lipoblast saptanıyor

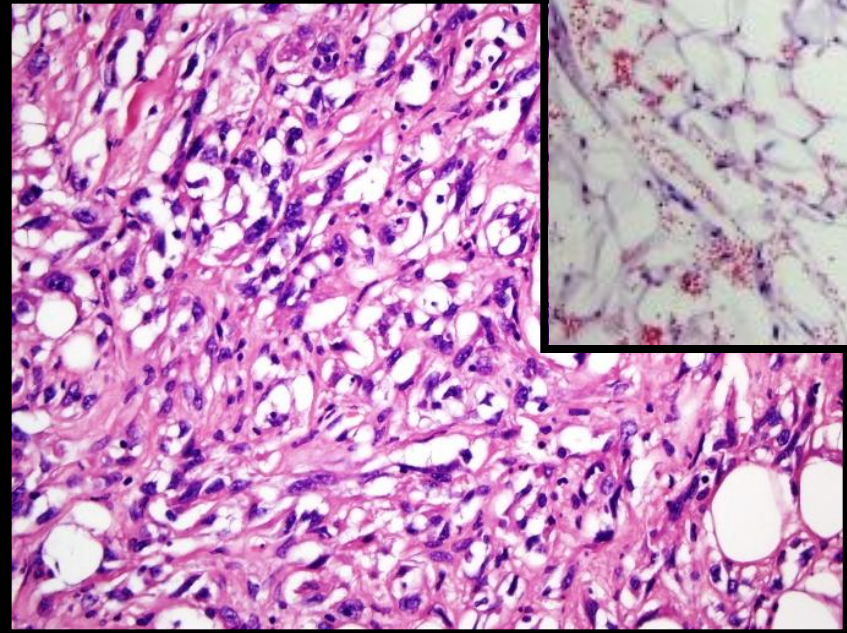
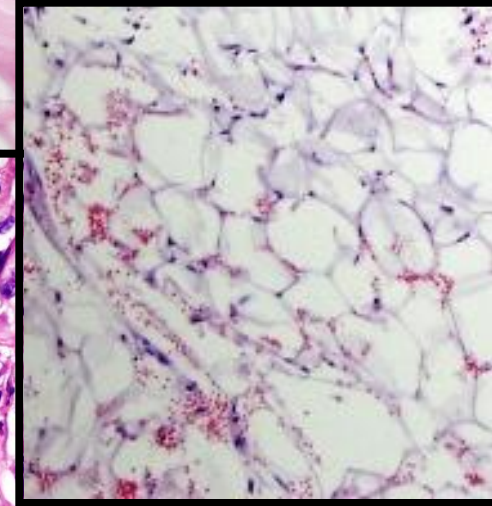
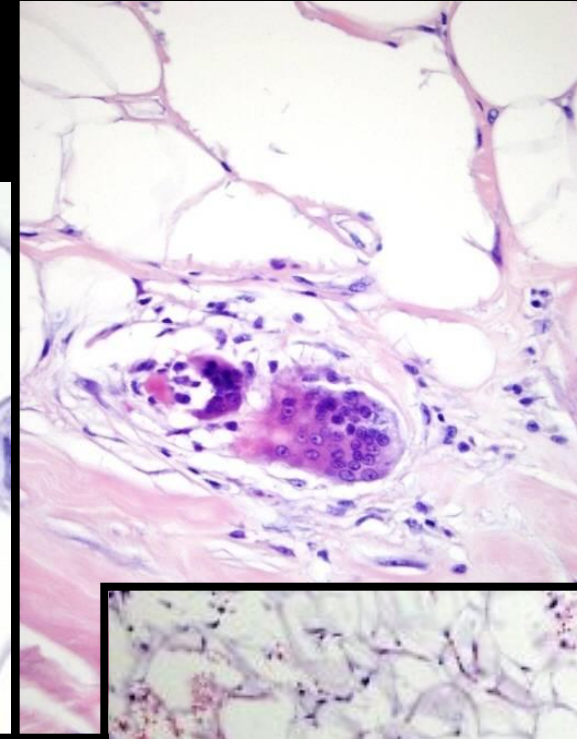
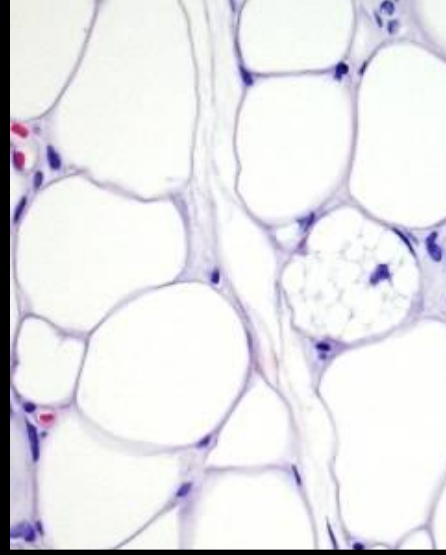


Lipit içeren sitoplazma, hiperkromatik nükleus ve uygun histolojik zemin



# Pseudolipoblasts

Atrofik yağ hücreleri  
Hibernom hücreleri  
Silikon rxn  
Taşlı yüzük Ca,  
Miksofibrosarkom  
Fiksasyon artefaktı  
Neoplastik hücrelerle  
infiltrasyon



Lipomatöz tm leri (iğne bx)  
yeterli klinik-radyolojik bilgi almadan  
yorumlamak son derecede tehlikelidir.

Son olarak.....



Çocukta ve yüzeysel lezyonda LS

tanısından önce,

Retroperiton, paratestiküler bölge ve  
kasık

büyük lipomatöz tümörlere

benign lipom tanısı vermeden önce

2 kez düşünelim.

LS'ların, karsinosarkom ve MMMT lerin  
heterolog komponentlerinden biri olarak  
görülebileceğini de unutmayalım



Sabrınız ve  
katılımınız için  
teşekkürler.....