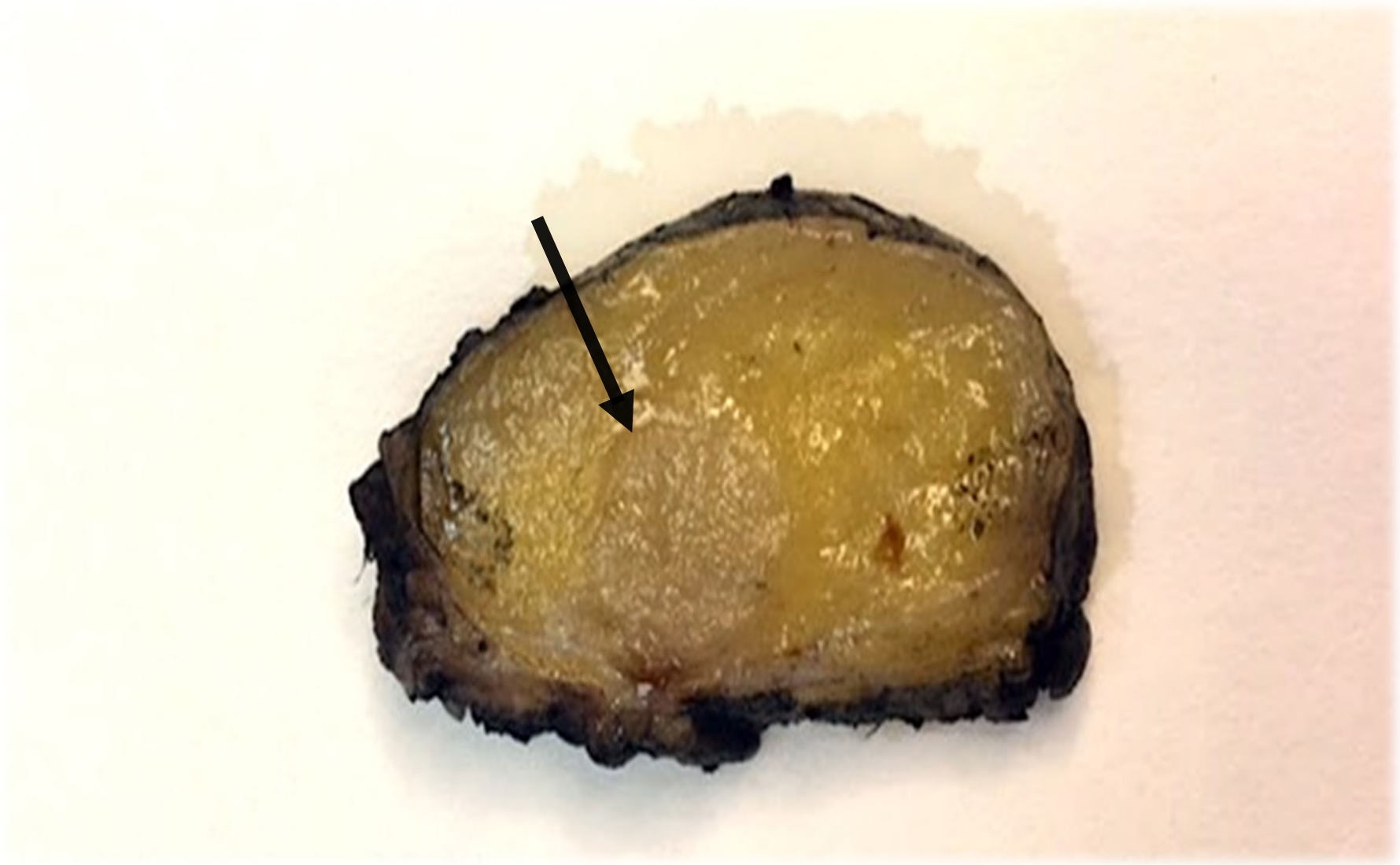
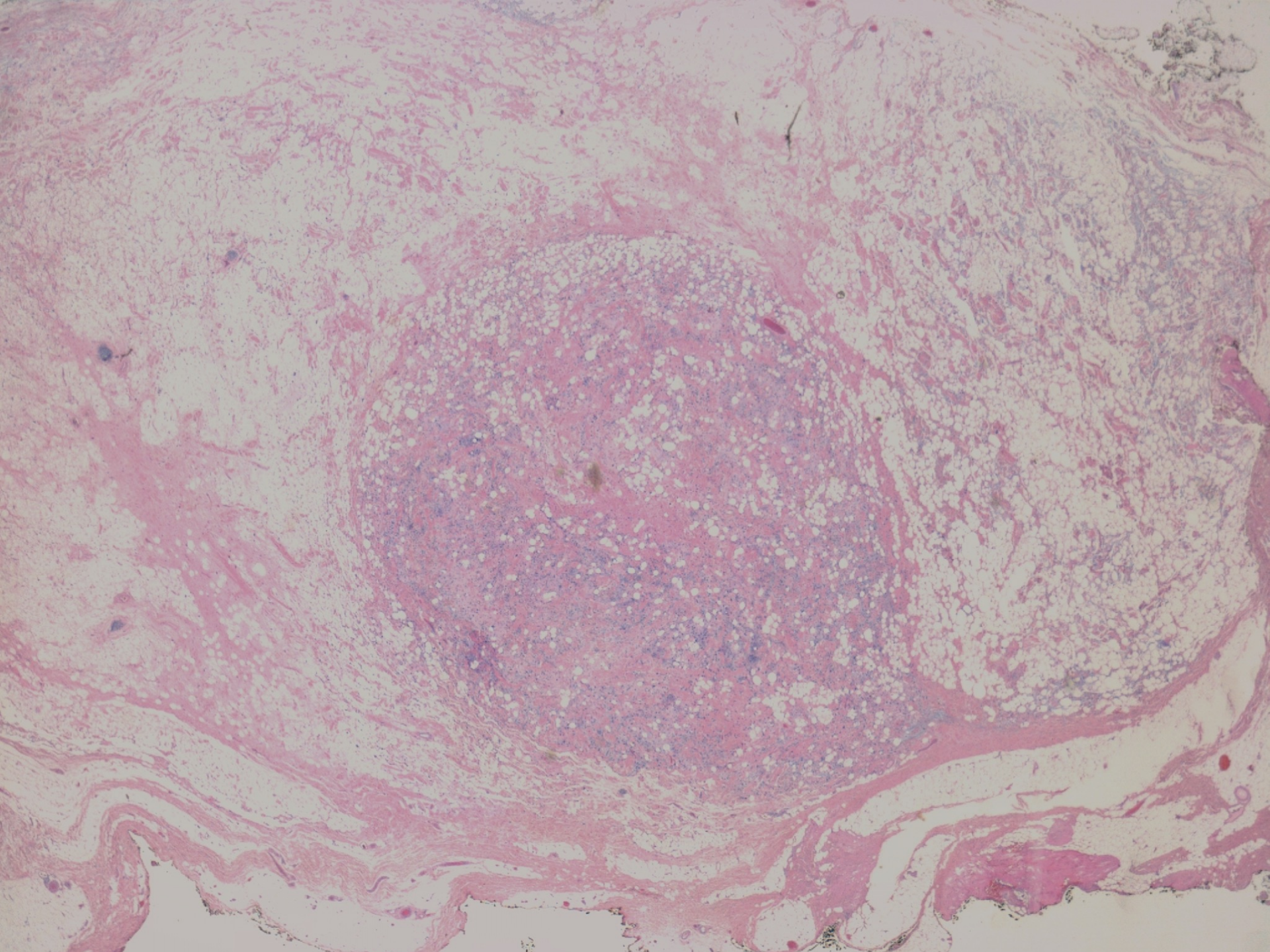
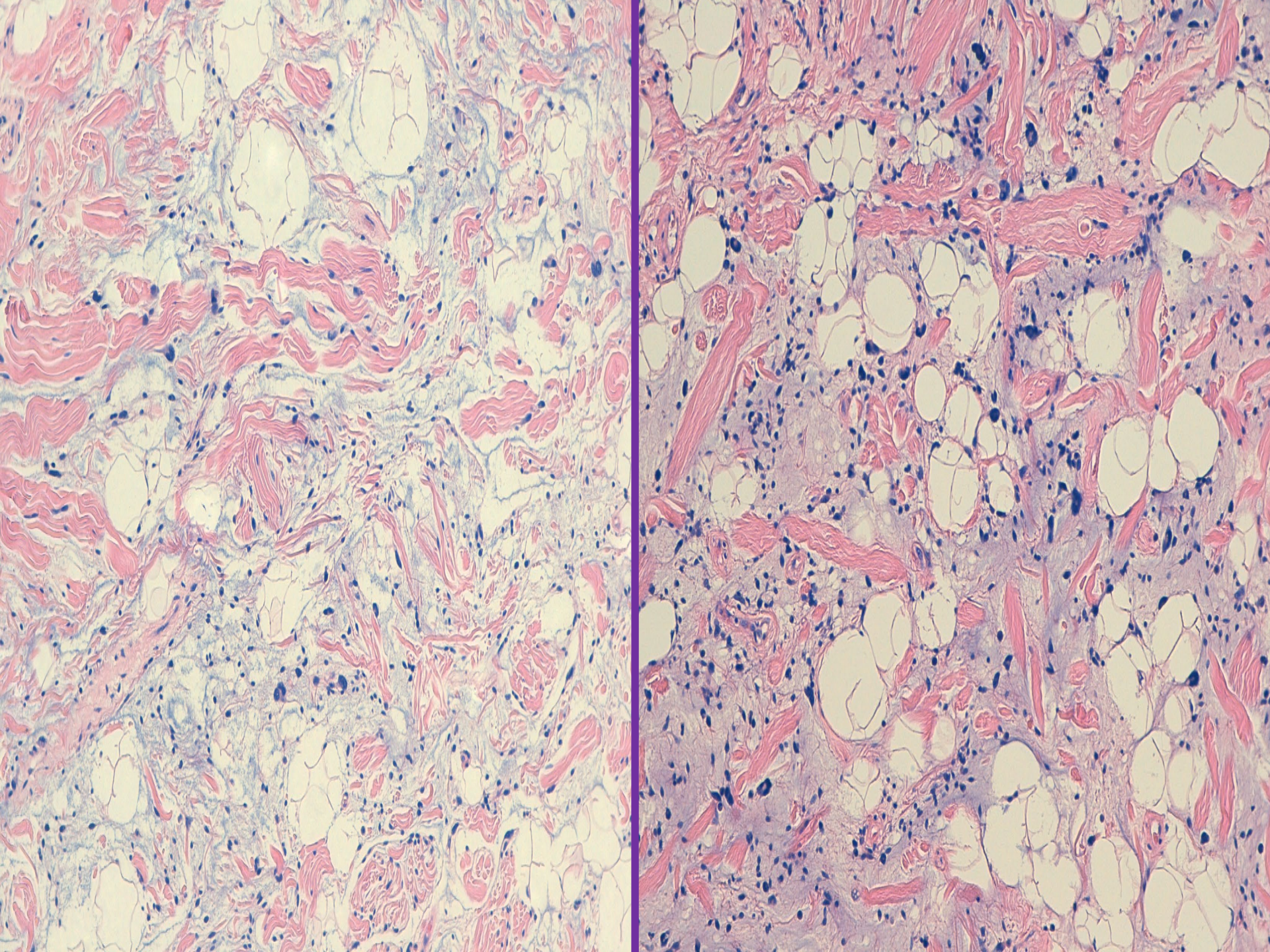
Nódulo preauricular subcutáneo

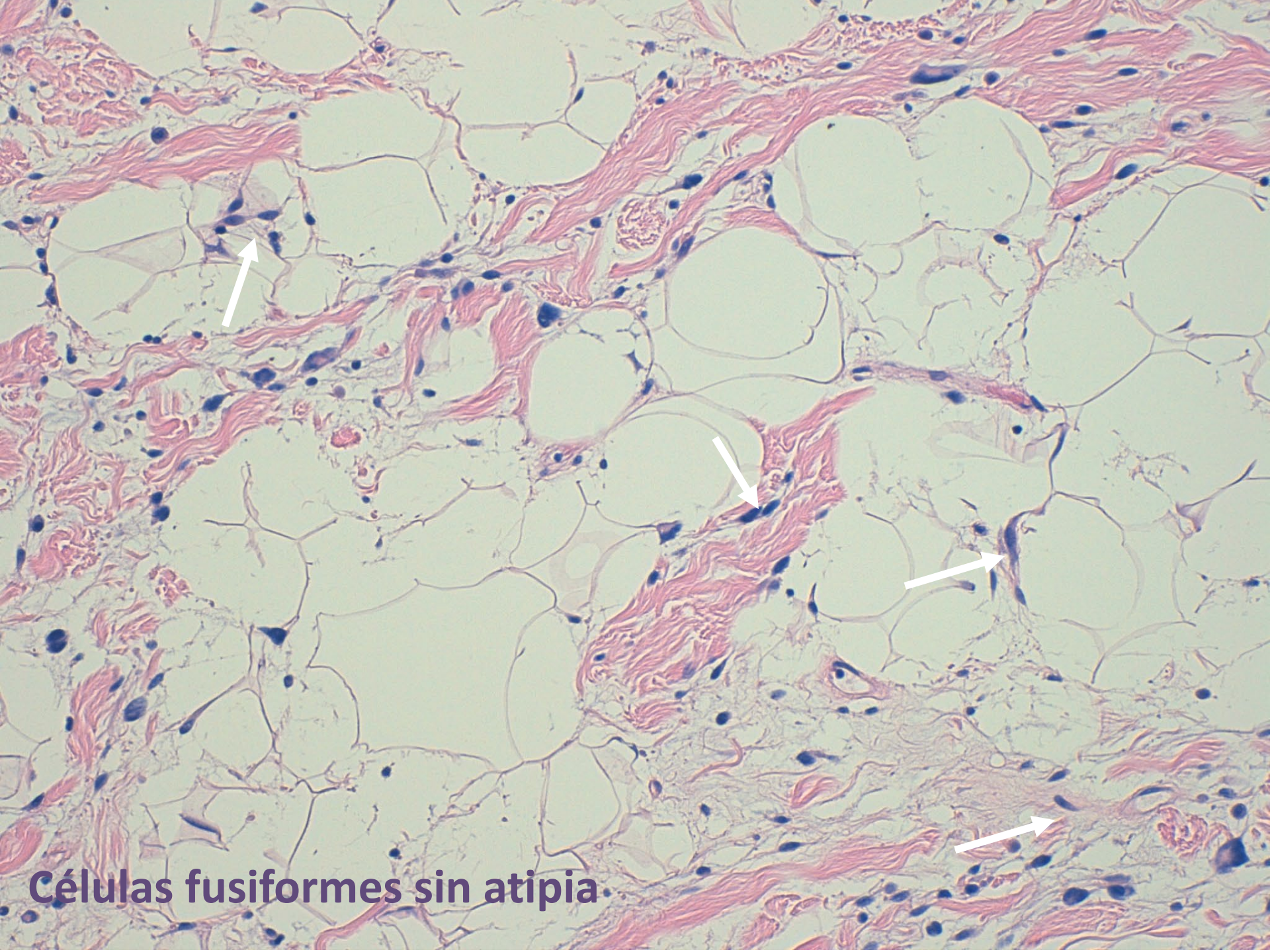
Oviedo Ramírez MI,Hernandez Barcelo JE,López motos D, Ruiz García G, Caballero Illanes A.

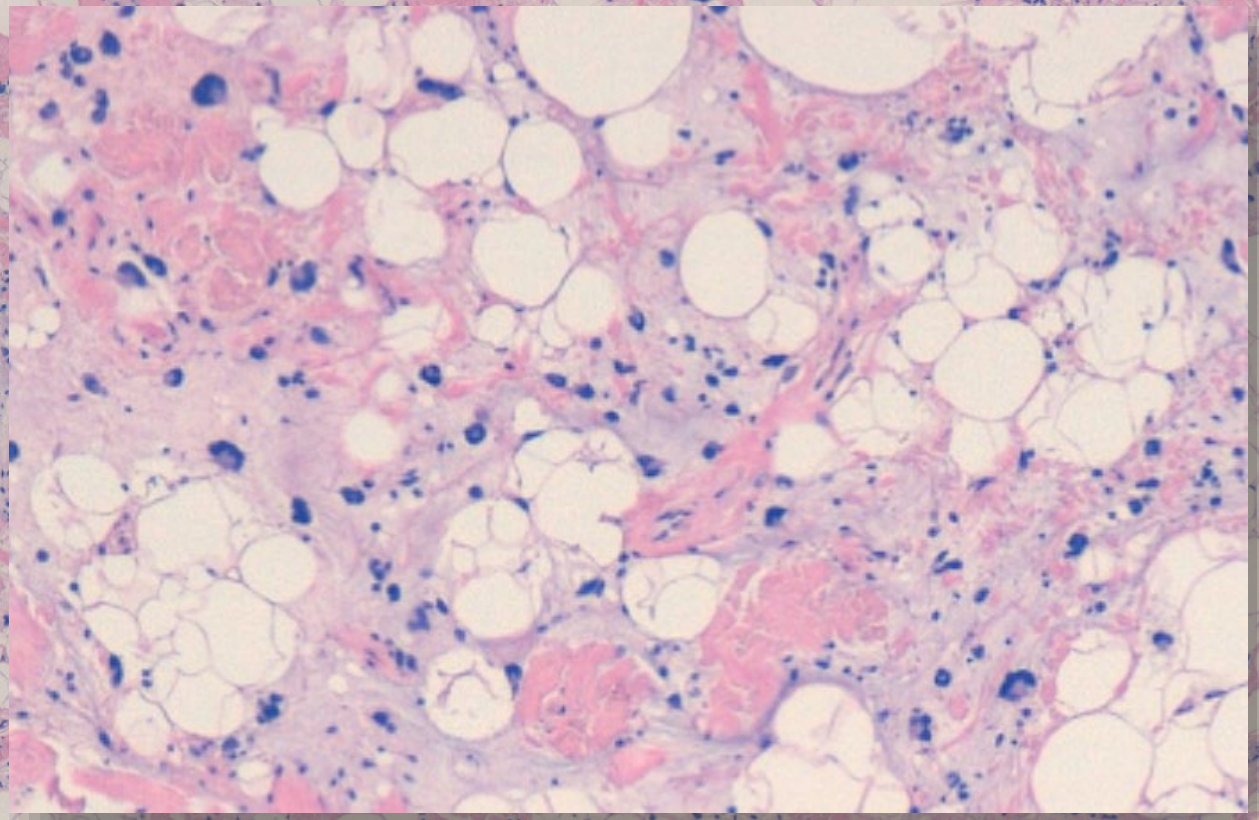
- Hombre de 71 años sin antecedentes de relevancia.
- Nódulo subcutáneo preauricular doloroso
- Ecografía lesión de 2,7 cm bien delimitada de densidad grasa
- PAAF.
- Exéresis.



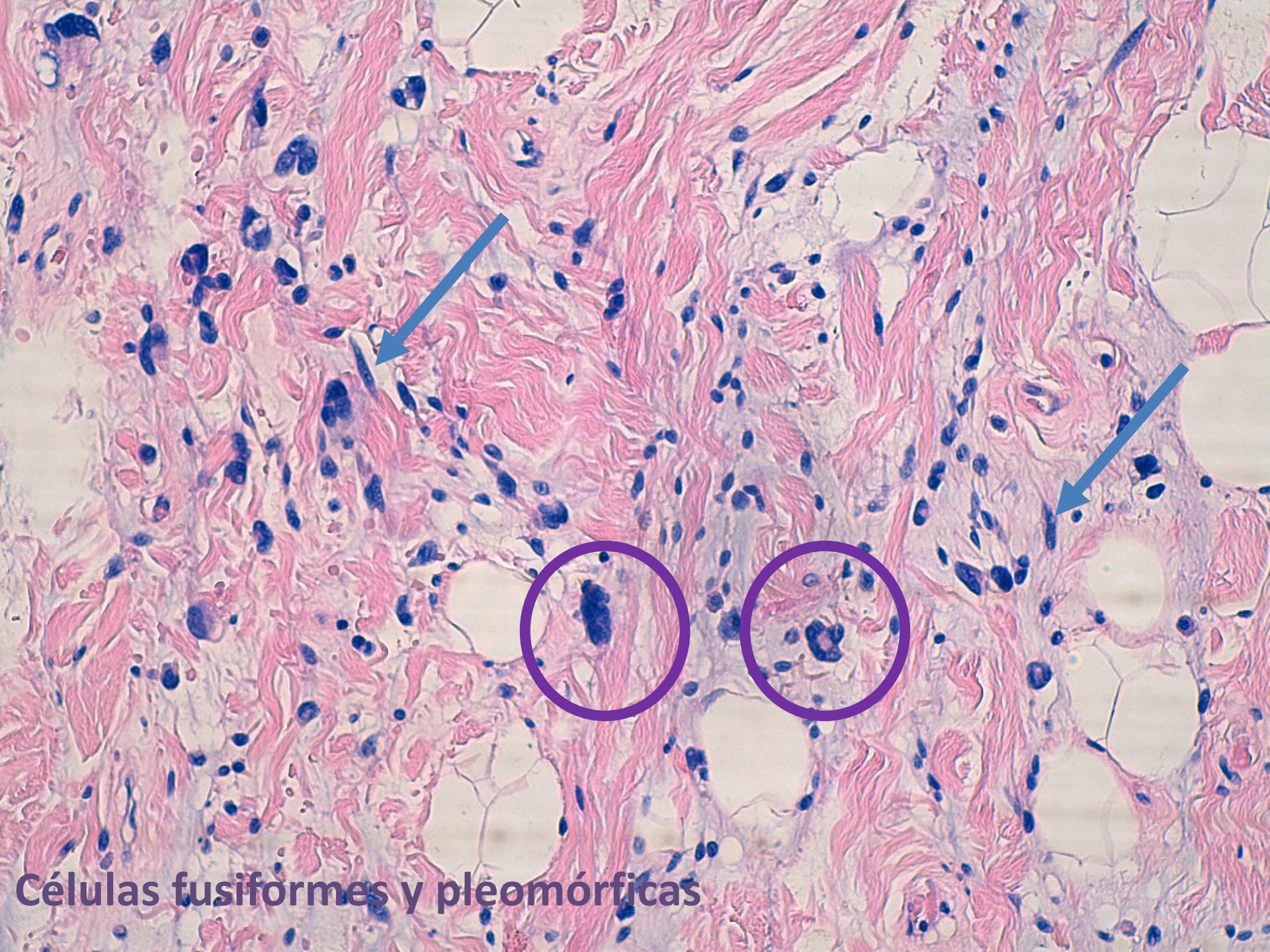


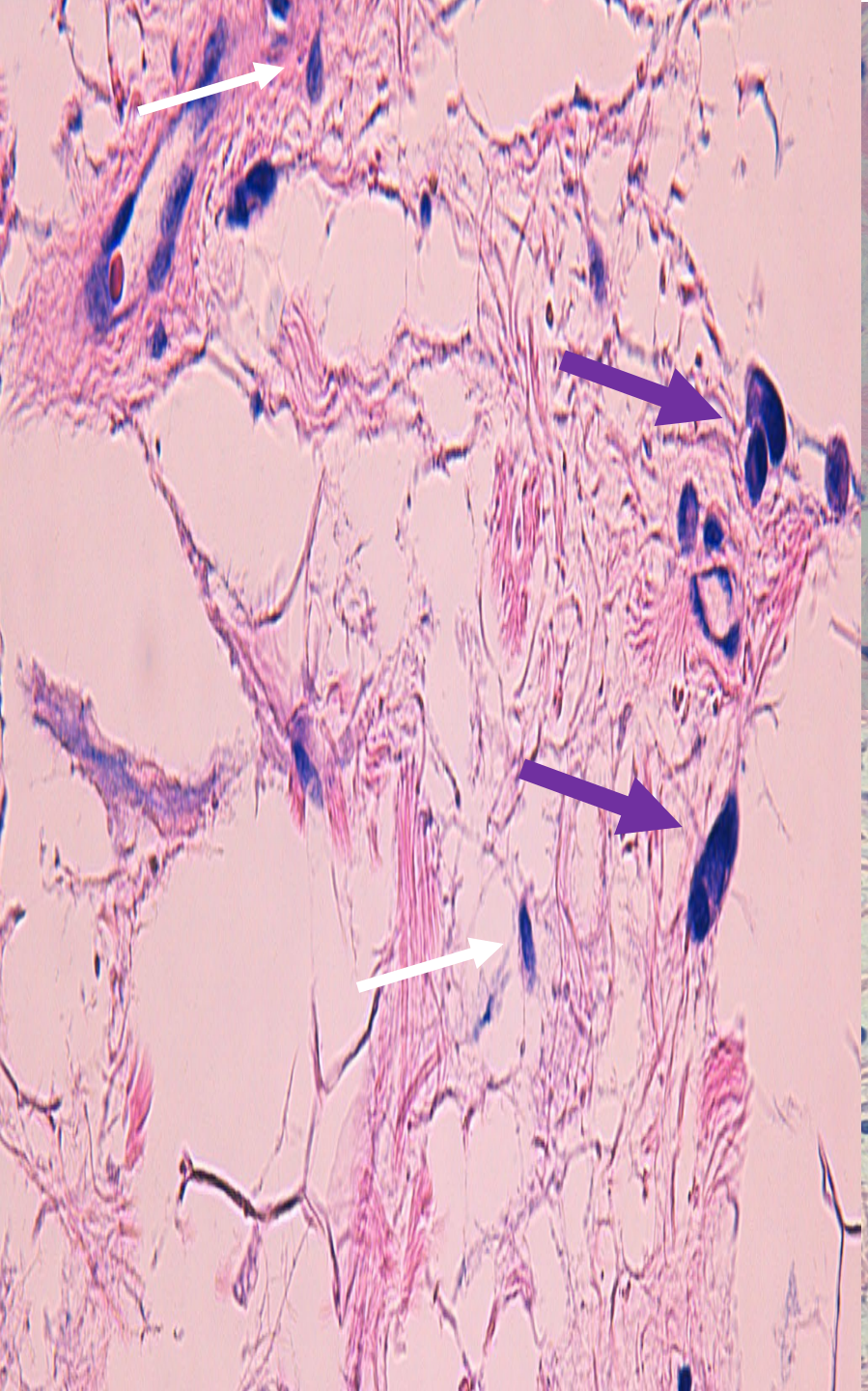


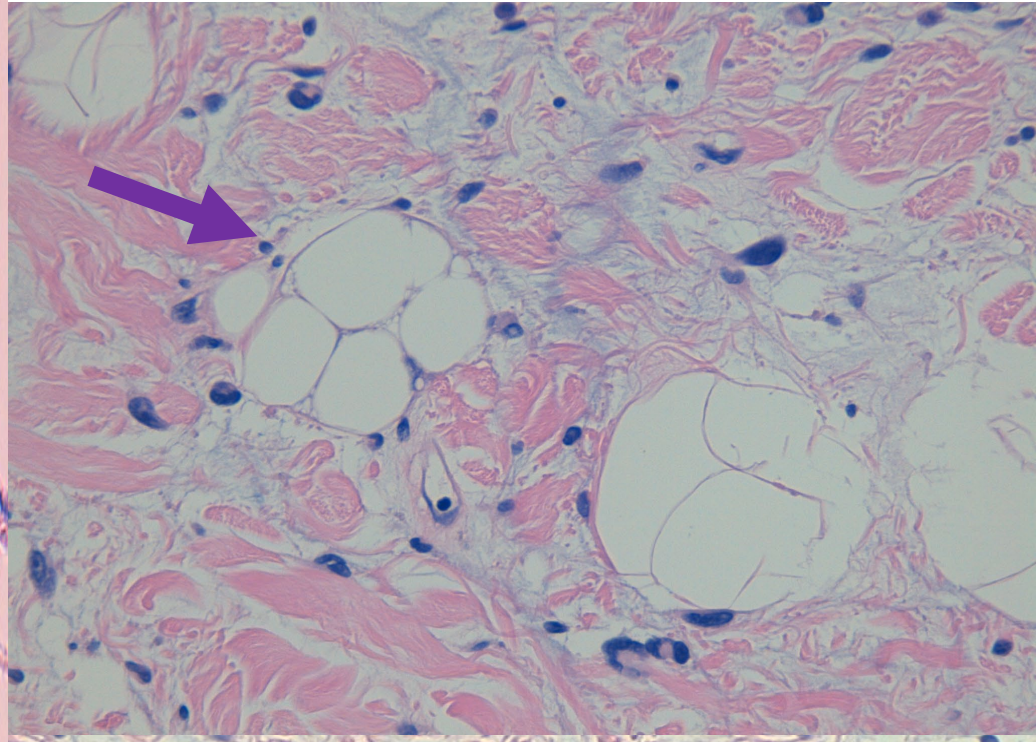

Células fusiformes sin atipia

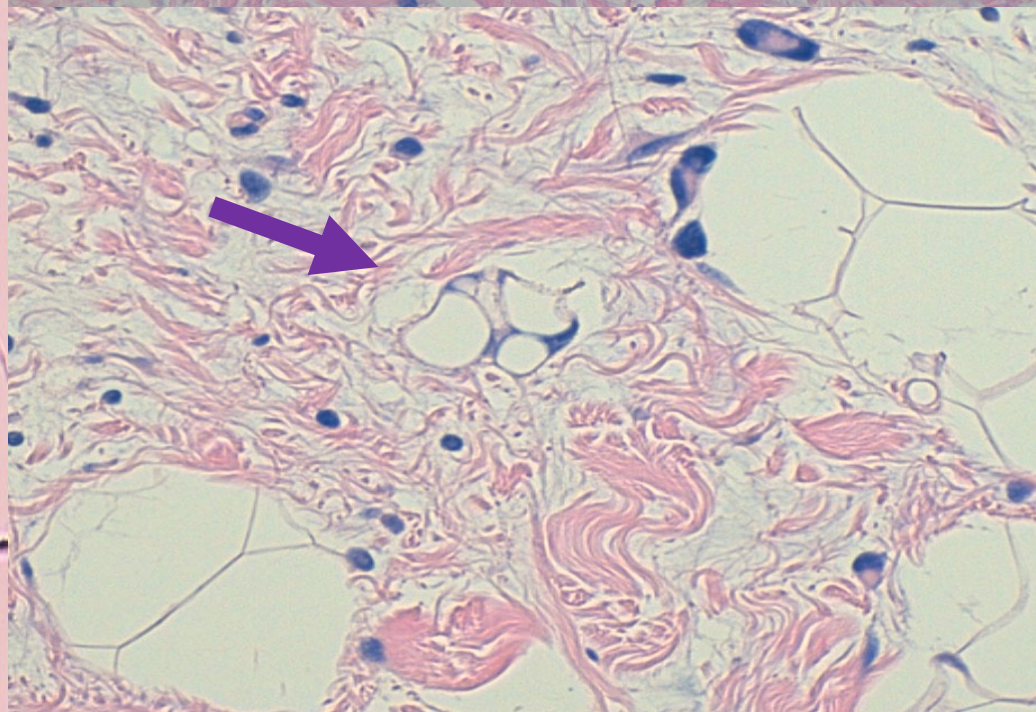


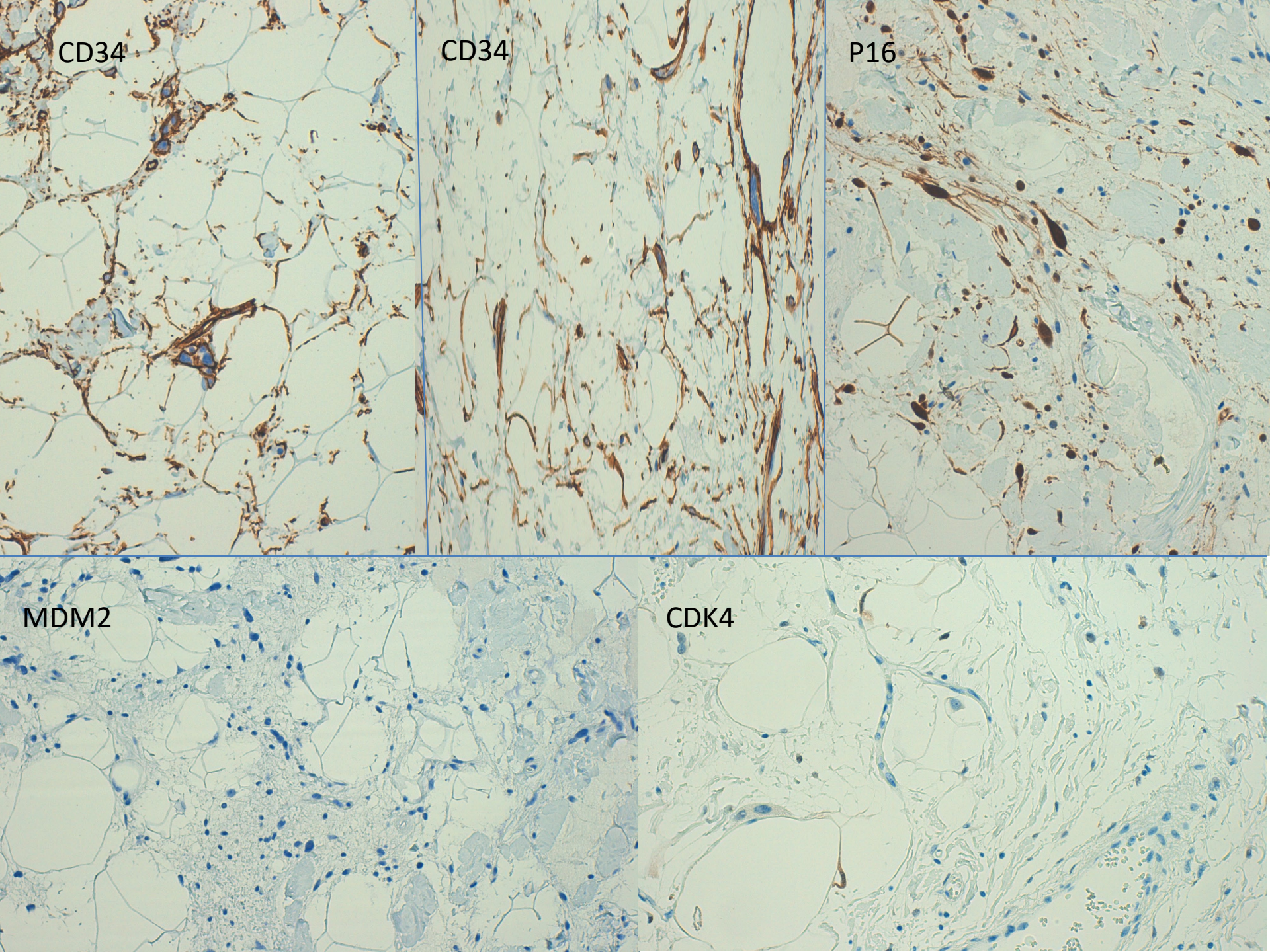
Predominio de células plemórficas y multinucleadas


Células fusiformes y pleomórficas







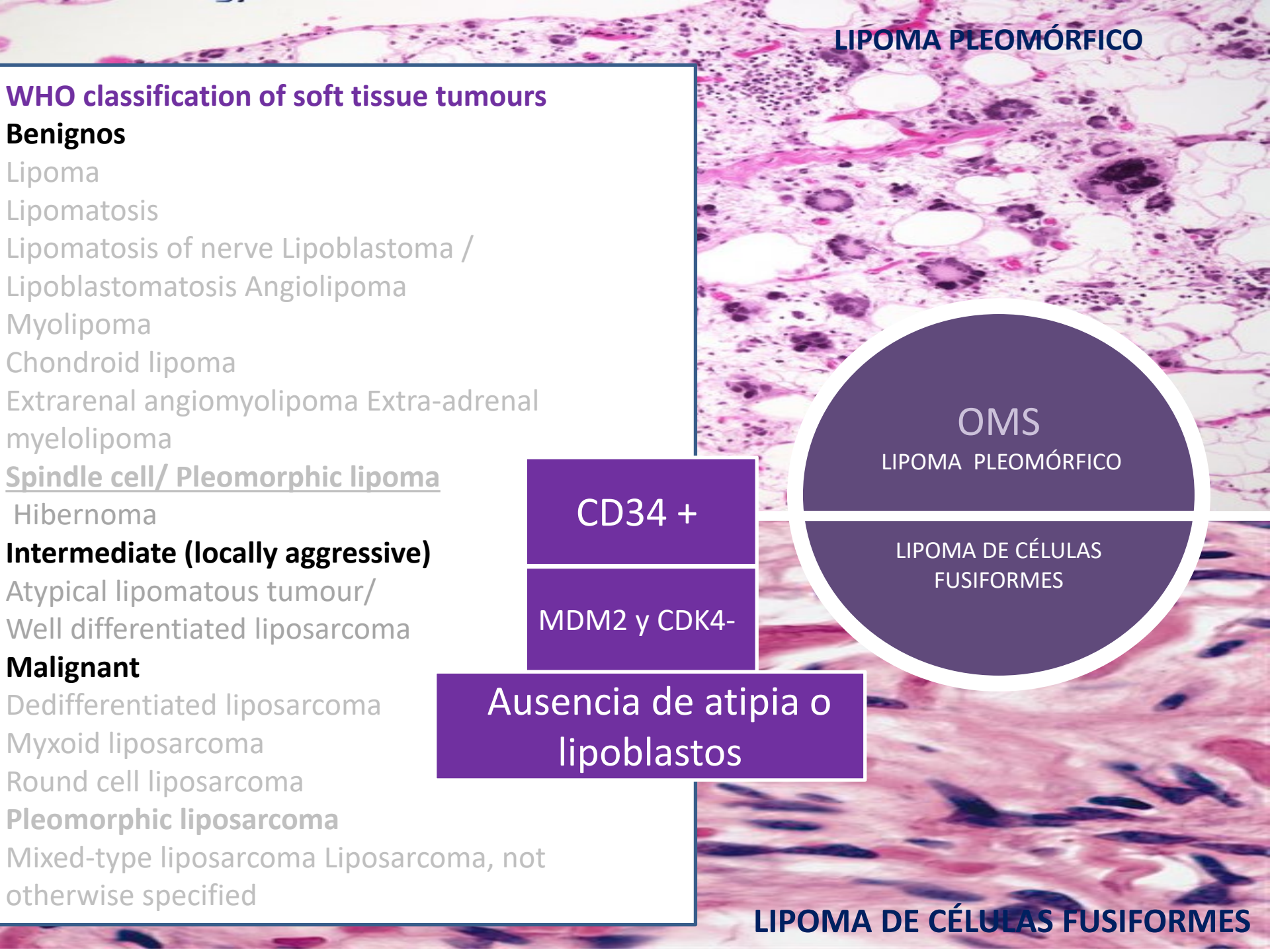

CD34
CD34
P16
MDM2
CDK4

DIAGNÓSTICO DIFERENCIAL

- LIPOMA PLEOMÓRFICO/ LIPOMA CÉLULAS FUSIFORMES (subcutáneos, CD34+,no atipia)

- LIPOSARCOMA BIEN DIFERENCIADO / TUMOR LIPOMATOSO ATIPICO (lipoblastos, Mdm2 positivos, CD34 negativos)

- LIPOSARCOMA PLEOMÓRFICO (otras localizaciones, no encapsulados, atipia citológica marcada, necrosis, mitosis. CD34 +)



LIPOMA PLEOMÓRFICO

WHO classification of soft tissue tumours

Benignos

Lipoma
Lipomatosis
Lipomatosis of nerve Lipoblastoma /
Lipoblastomatosis Angiolipoma
Myolipoma
Chondroid lipoma
Extrarenal angiomyolipoma Extra-adrenal
myelolipoma
Spindle cell/ Pleomorphic lipoma
Hibernoma

Intermediate (locally aggressive)

Atypical lipomatous tumour/
Well differentiated liposarcoma

Malignant

Dedifferentiated liposarcoma
Myxoid liposarcoma
Round cell liposarcoma
Pleomorphic liposarcoma
Mixed-type liposarcoma Liposarcoma, not
otherwise specified

CD34 +

MDM2 y CDK4-

Ausencia de atipia o lipoblastos

OMS

LIPOMA PLEOMÓRFICO

LIPOMA DE CÉLULAS FUSIFORMES

LIPOMA DE CÉLULAS FUSIFORMES

Lipoma de células fusiformes/lipoma pleomórfico

USUAL: Cabeza y cuello y en la cintura escapular (>80%).

INUSUAL: 10% región anterior del cuello, cara, órbita, boca.

Lesiones únicas. Casos múltiples familiares.

Recidivas excepcionales. Debe restringirse este término para las lesiones superficiales situadas en la hipodermis o más raramente, dermis.

Lipoma de células fusiformes/lipoma pleomórfico

MACROSCÓPICO:

Subcutáneos, bien delimitados, usualmente cápsula

HISTOPATOLOGÍA:

LCF

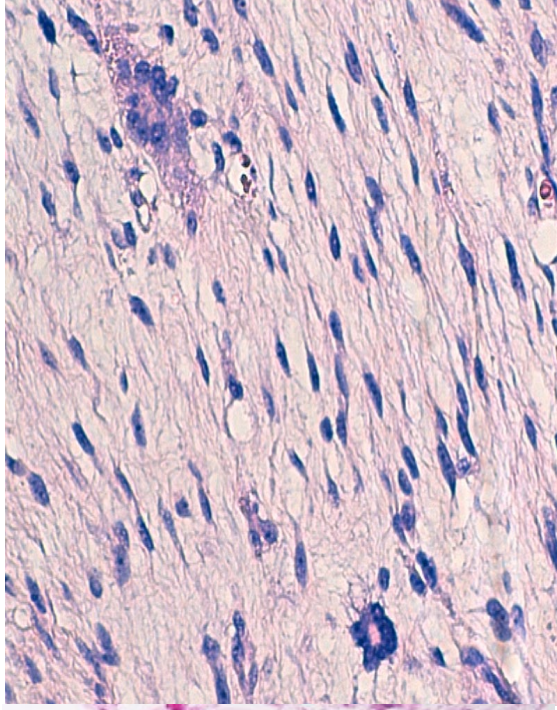
-Variable proporción de adipocitos, células fusiformes, colágeno y matriz mixoide.

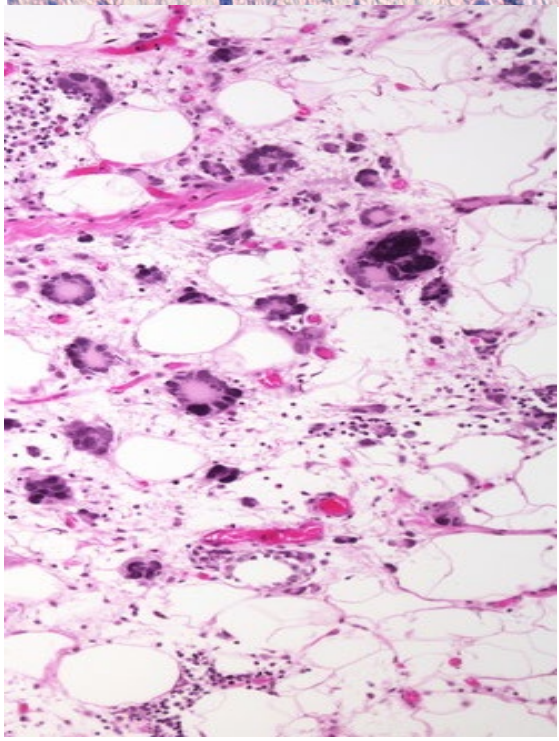
-Las células fusiformes tienden a disponerse en haces o dispersas y contienen escaso citoplasma y núcleo pálido y uniforme.

LP

-Células multinucleadas, células floret, a veces nucleolo.
El estroma es de tipo fibromixoide con mastocitos.

-El grado de vascularización es también muy variable (patrón hemangiopericítico, plexiforme o con frecuentes espacio pseudovasculares)





¿ Lipoma pleomórfico o de células fusiformes con atipia nuclear CD34 + y MDM2 negativo?

ORIGINAL ARTICLE

"Atypical" Pleomorphic Lipomatous Tumor

A Clinicopathologic, Immunohistochemical and Molecular Study of 21 Cases, Emphasizing its Relationship to Atypical Spindle Cell Lipomatous Tumor and Suggesting a Morphologic Spectrum (Atypical Spindle Cell/Pleomorphic Lipomatous Tumor)

David Creytens, MD, PhD,† Thomas Mentzel, MD, PhD,‡ Liesbeth Ferdinande, MD, PhD,*† Evelyne Lecoutere, MD,* Joost van Gorp, MD, PhD,§ Lilit Atanesyan, PhD,‖ Karel de Groot, MSc,‖ Suvi Savola, PhD,‖ Nadine Van Roy, PhD,†¶ Jo Van Dorpe, MD, PhD,*† and Uta Flucke, MD, PhD#*

Abstract: The classification of the until recently poorly explored group of atypical adipocytic neoplasms with spindle cell features, for which recently the term atypical spindle cell lipomatous tumor (ASLT) has been proposed, remains challenging. Recent studies have proposed ASLT as a unique entity with (in at least a significant subset of cases) a specific genetic background, namely deletions/losses of 13q14, including *RB1* and its flanking genes *RCBTB2*, *DLEU1*, and *ITM2B*. Similar genetic aberrations have been reported in pleomorphic liposarcomas (PLSs). This prompted us to investigate a series of 21 low-grade adipocytic neoplasms with a pleomorphic lipoma–like appearance, but with atypical morphologic features (including atypical spindle cells, pleomorphic [multinucleated] cells, pleomorphic lipoblasts and poor circumscription), for which we propose the term "atypical" pleomorphic lipomatous tumor (APLT). Five cases of PLS were also included in this study. We used multiplex ligation-dependent probe amplification to evaluate genetic changes of 13q14. In addition, array-based comparative genomic hybridization was performed on 4 APLTs and all PLSs. Multiplex ligation-dependent probe amplification showed consistent loss of *RB1* and its flanking gene *RCBTB2* in all cases of APLT. This genetic alteration was also present in all PLSs, suggesting genetic overlap, in addition to morphologic overlap, with APLTs. However, array-based comparative genomic hybridization demonstrated more complex genetic alterations with more losses and gains in PLSs compared with APLTs. APLTs arose in the subcutis (67%) more frequently than in the deep (subfascial) soft tissues (33%). With a median follow-up of 42 months, recurrences were documented in 2 of 12 APLTs for which a long follow-up was available. Herein, we also demonstrate that APLTs share obvious overlapping morphologic, immunohistochemical, genetic and clinical characteristics with the recently defined ASLT, suggesting that they are related lesions that form a spectrum (atypical spindle cell/pleomorphic lipomatous tumor).

Key Words: "atypical" pleomorphic lipomatous tumor, atypical spindle cell lipomatous tumor, pleomorphic liposarcoma, *RB1*, MLPA

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Liposarcomas are the most common sarcomas in adults. They are a heterogeneous family of tumors consisting of 4 major well-defined entities. In decreasing frequency, the 4 entities are atypical lipomatous tumor (ALT)/ dedifferentiated liposarcoma (DDLS), myxoid liposarcoma (MLS), and pleomorphic liposarcoma (PLS). The first 3 entities are characterized by diagnostic recurrent genetic changes, more specifically, *MDM2/CDK4* amplifications in ALT and DDLS, and *FUS/EWSR1-DDIT3* gene fusions in MLS. PLSs demonstrate complex genetic

Se propone que el APLT es una entidad con alteraciones genéticas tipo deleción o pérdidas de cromosoma 13q14 incluida proteina Rb1 y otros .

Estas alteraciones genéticas así como la positividada a CD34 se han descrito en liposarcomas pleomórficos y no las presentan los TLA ó LBD.

¿Variante de tumor adiposo de bajo grado?
¿Posible precursor
De liposarcoma pleomórfico?

Am J Surgical pathol 2017;41:1443-1445

ENTIDAD AÚN NO RECONOCIDA POR LA OMS

TABLE 2. Summary of Morphologic, Immunohistochemical and Molecular Characteristics of "Atypical" Pleomorphic Lipomatous Tumor and Pleomorphic Liposarcoma

	n (%)	
	APLT (N = 21)	PLS (N = 5)
Tumor margin		
Infiltration of adjacent tissue	15 (71)	5 (100)
Overall cellularity		
Low	5 (24)	0
Moderate	10 (48)	0
High	6 (29)*	5 (100)
	*Pronounced fascicular growth in 3 cases	
Degree of pleomorphism†		
Mild	0	0
Moderate	17 (81)	0
Severe	4 (19)	5 (100)
Atypical spindle cell component	21 (100)*	0
	*Focal: 16 (75) Diffuse: 5 (25)	
Stroma		
Myxoid		
0%	7 (33)	3 (60)
<10%	10 (48)	0
10%-50%	3 (14)	2 (40)
>50%	1 (4)	0
Ropey collagen	15 (71)	0
Floret-like multinucleated cells	21 (100)	0
Pleomorphic lipoblasts	21 (100)	5 (100)
Mitotic activity	21 (100)*	5 (100)*
	*Low (<5/50 HPFs)	*High (>30/50 HPFs)
Tumor necrosis	0	5 (100)
Immunohistochemistry		
Rb (loss)	21 (100)	5 (100)
CD34	21 (100)	1 (20)
MDM2	0	0
p16	20 (95)	5 (100)
Genetic features		
Loss of 13q	21 (100)	21 (100)
RB1 loss	21 (100)	5 (100)
RCBTB2 loss	21 (100)	5 (100)
DLEU1 loss	21 (100)	2 (40)
MDM2 amplification	0	0

The presence of a spindle cell component, ropey collagen and floret-like mul-

COMPATIBLE CON :

TUMOR LIPOMATOSO

ATÍPICO"PLEOMÓRFICO"

CONCLUSIONES

-Atención a los lipomas subcutáneos con células fusiformes/pleomórficas: valorar atipia citológica en casos de lipomas CD34 positivos.

-Ante un lipoma pleomórfico o lipoma de células fusiformes con ATIPIA, lipoblastos y p16 positiva considerar nueva terminologia propuesta aunque no reconocida por OMS.


GRACIAS POR SU ATENCIÓN