



Sociedade Brasileira de Patologia

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**NÚCLEO DE ESPECIALIDADES – 18º
ENCONTRO - ÁREA DE PATOLOGIA
CIRÚRGICA E IMUNO-HISTOQUÍMICA**

**Carlos Renato A. Melo
Laboratórios Reunidos
Santa Maria - RS**

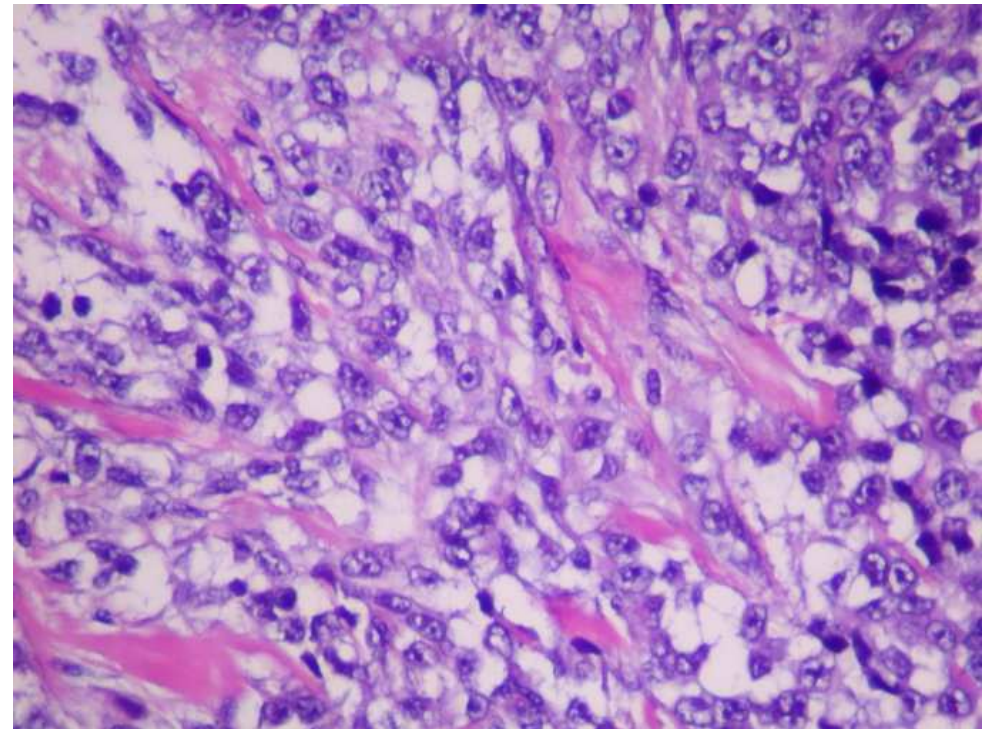
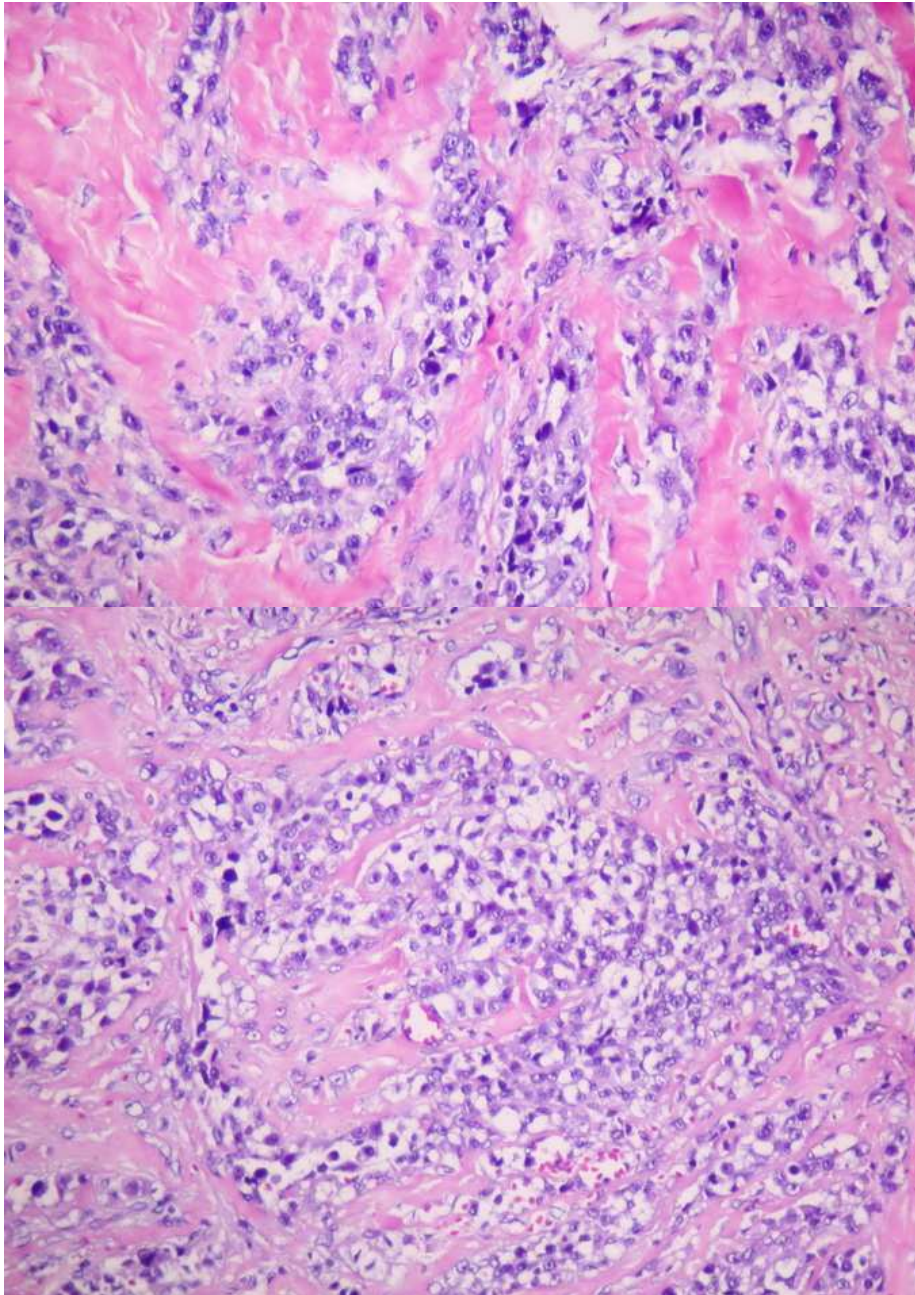
São Paulo, 28 de setembro de 2013

Achados clínicos do caso discutido na presente reunião



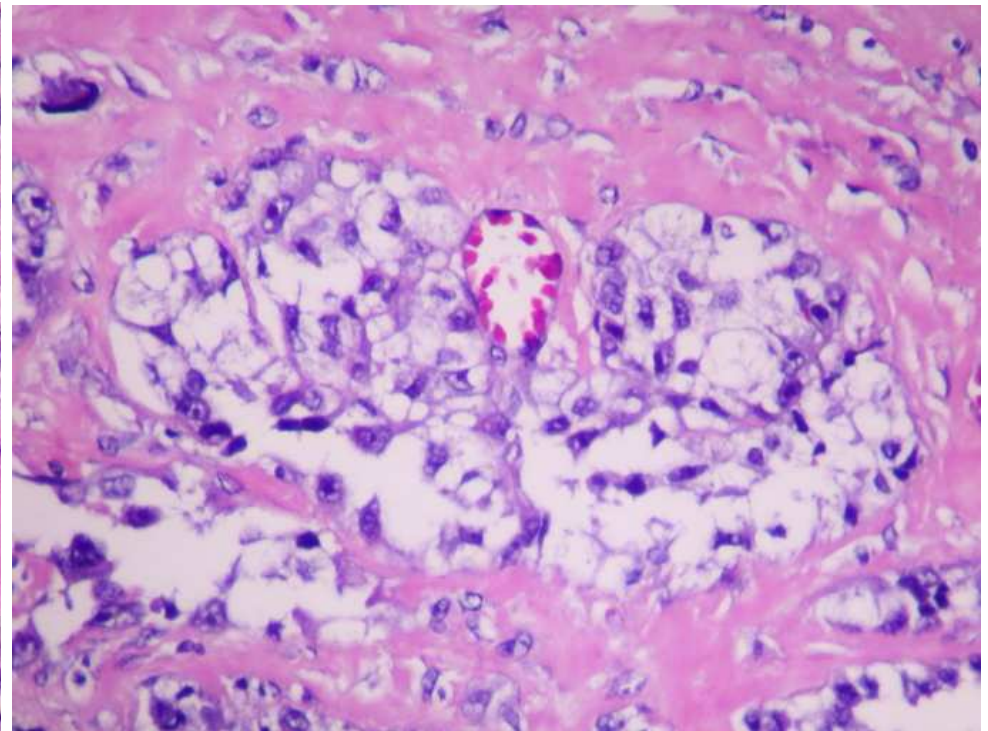
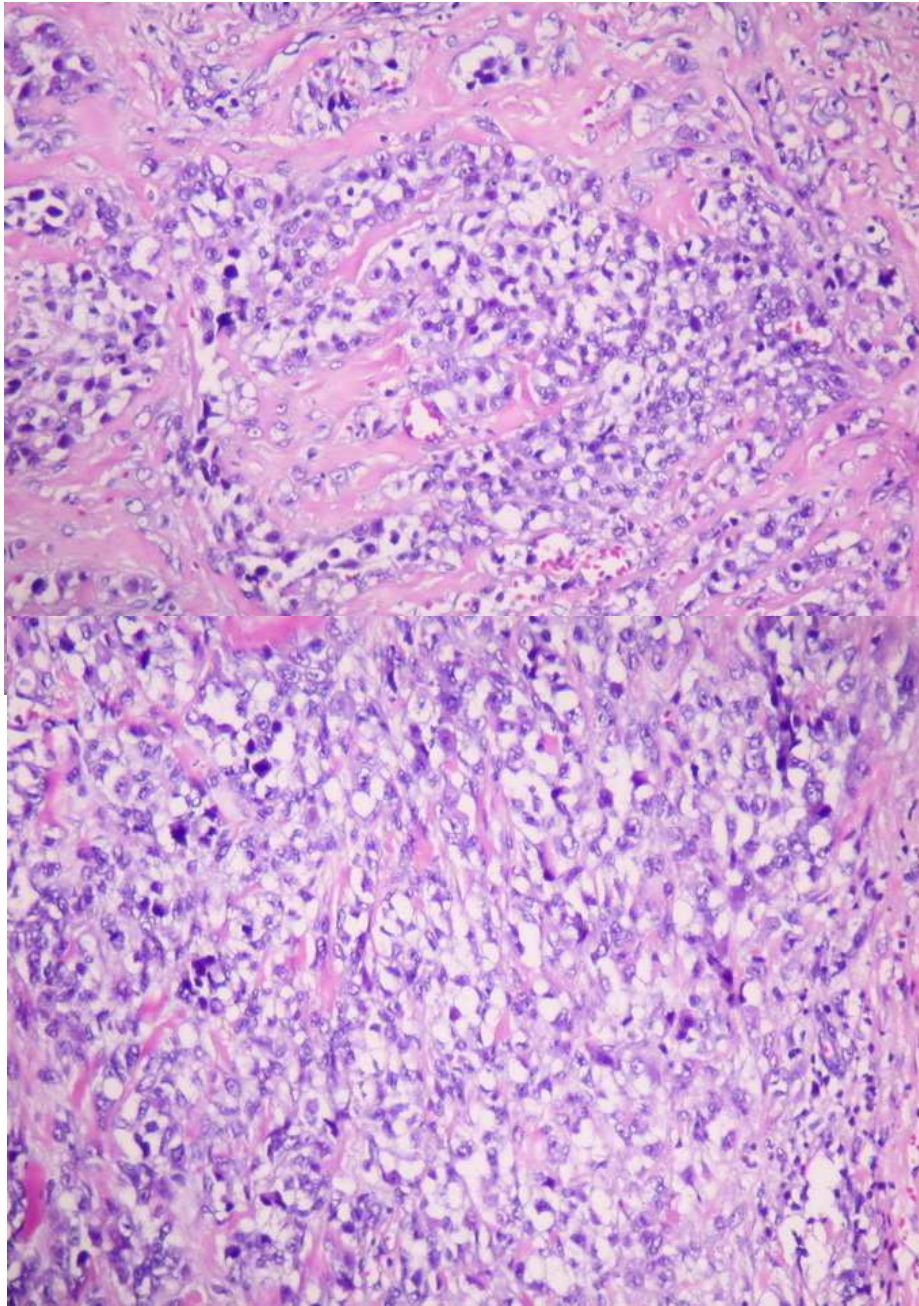
Homem de 40 anos, com lesão
na face medial do pé

ACHADOS HISTOLÓGICOS



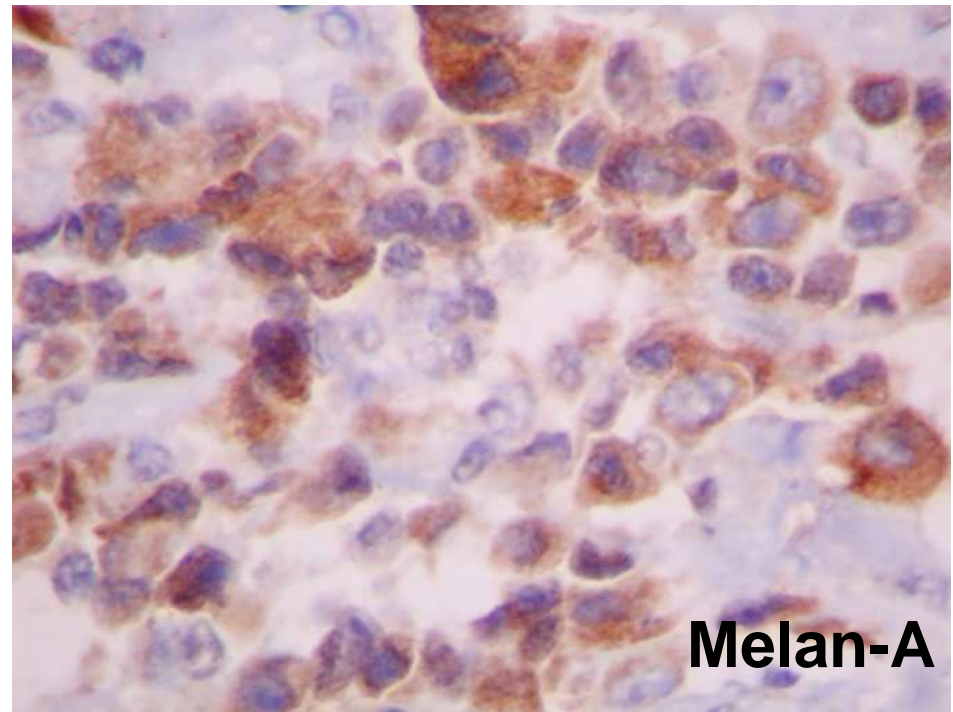
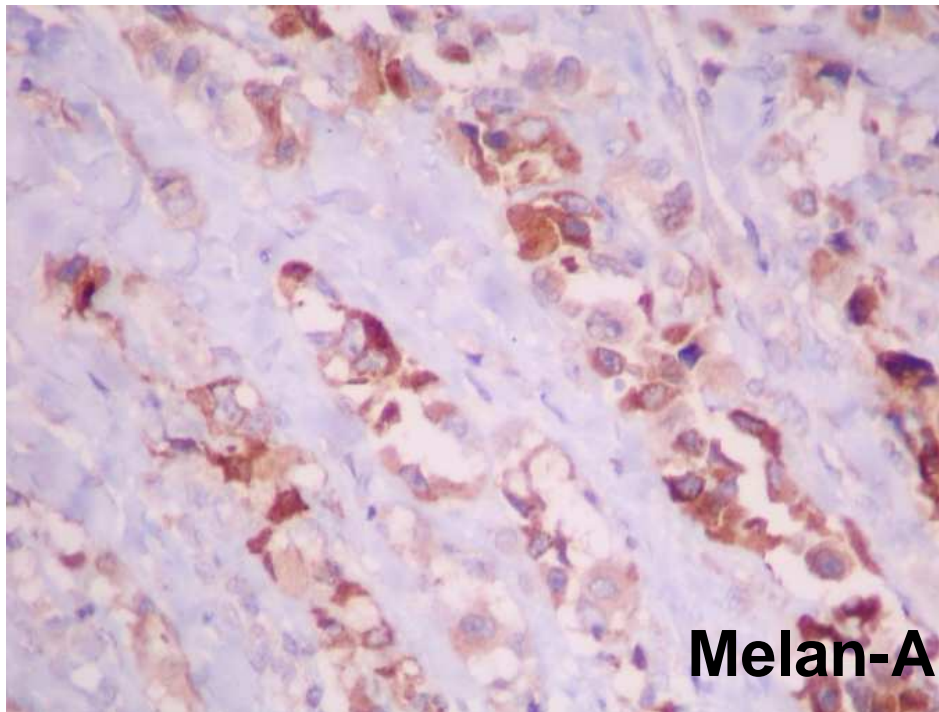
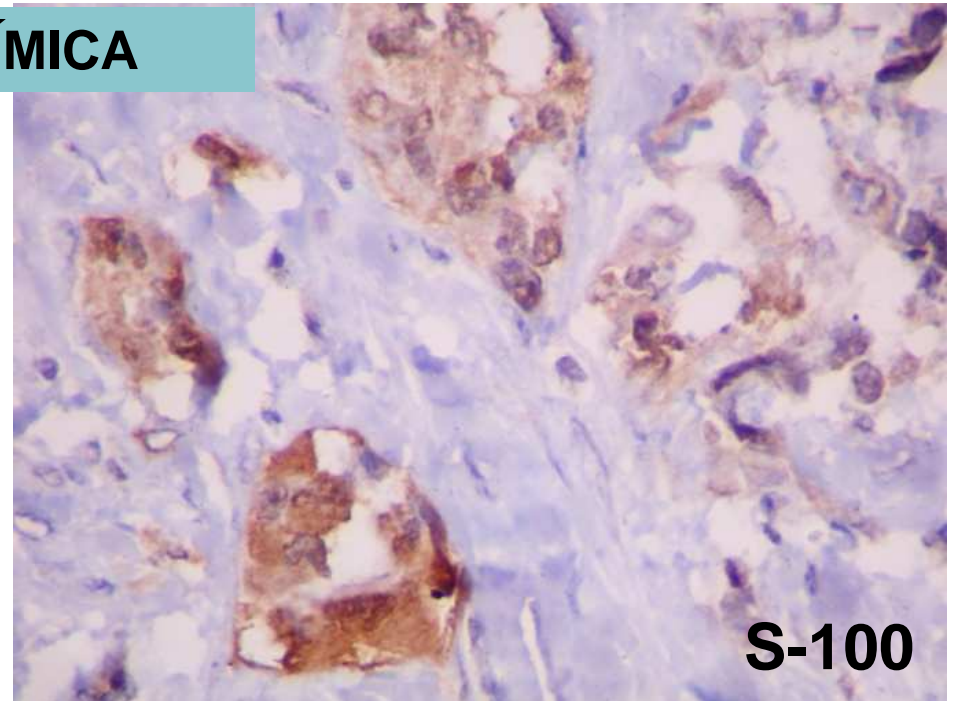
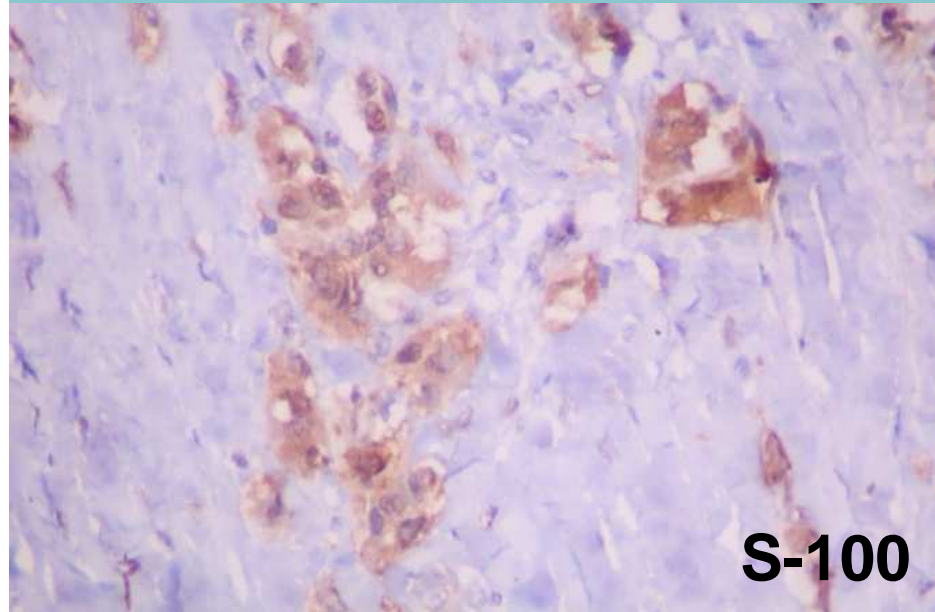
Grupos de células, muitas delas com citoplasma claro, aparecem separadas por colágeno.

ACHADOS HISTOLÓGICOS

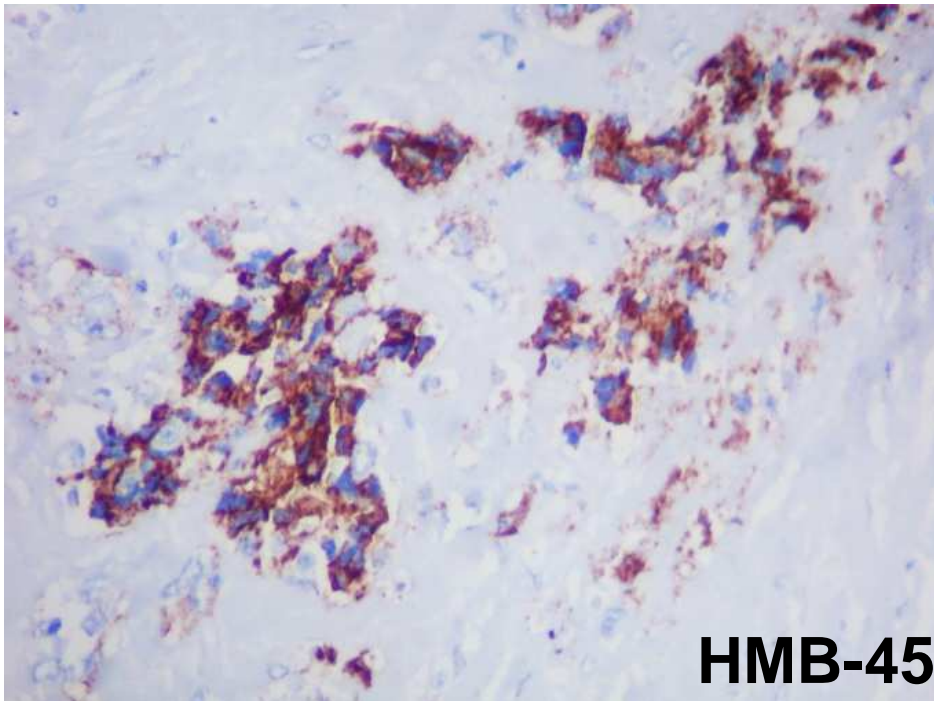
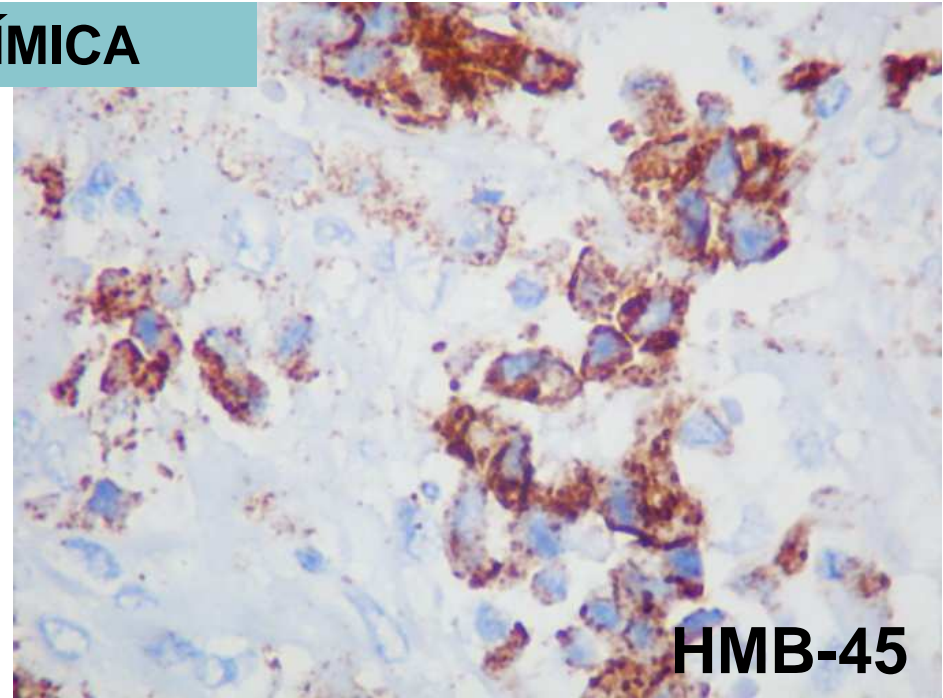
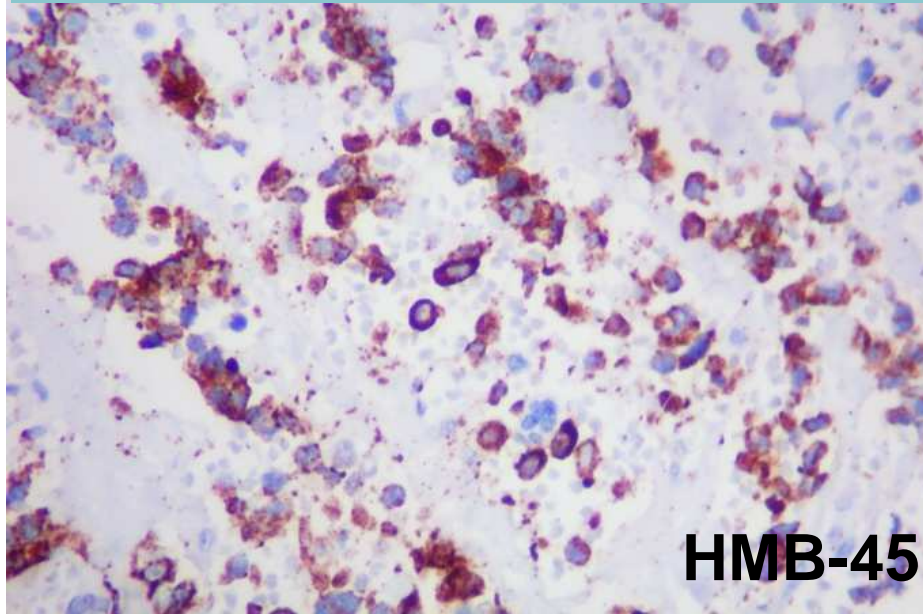


Grupos de células, muitas delas com citoplasma claro, aparecem separadas por colágeno.

ACHADOS DE IMUNO-HISTOQUÍMICA



ACHADOS DE IMUNO-HISTOQUÍMICA

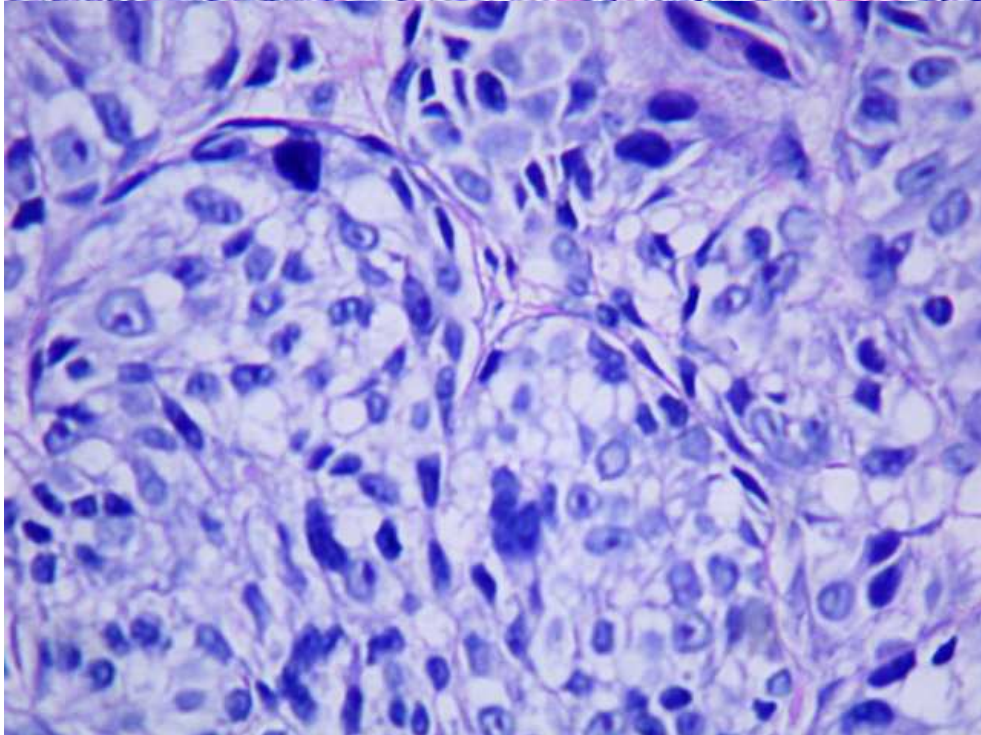
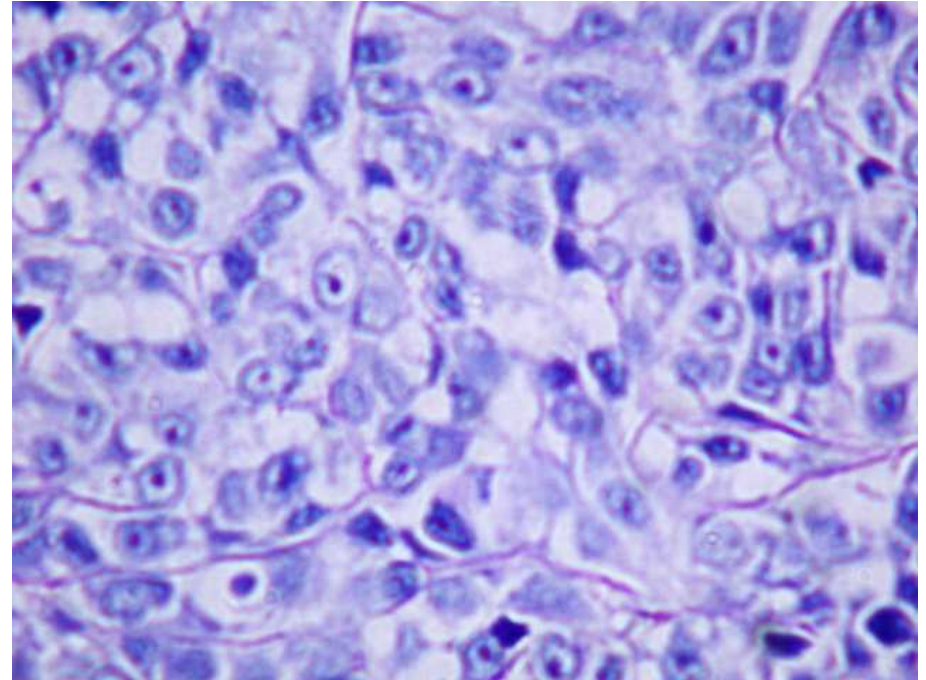
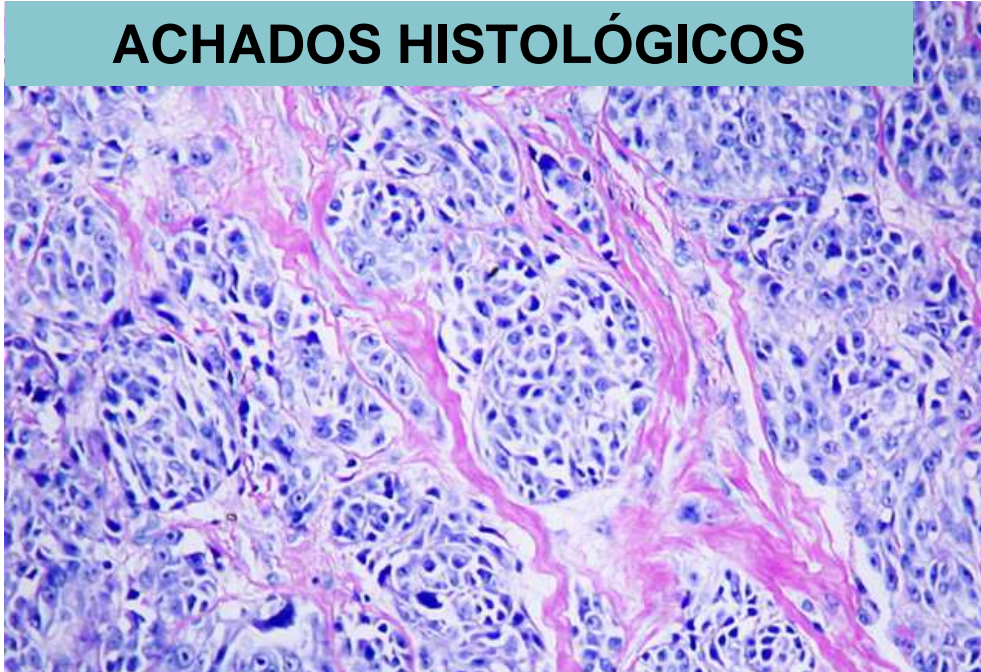


As células neoplásicas mostram positividade para marcadores melanocíticos.

Achados clínicos de um caso similar

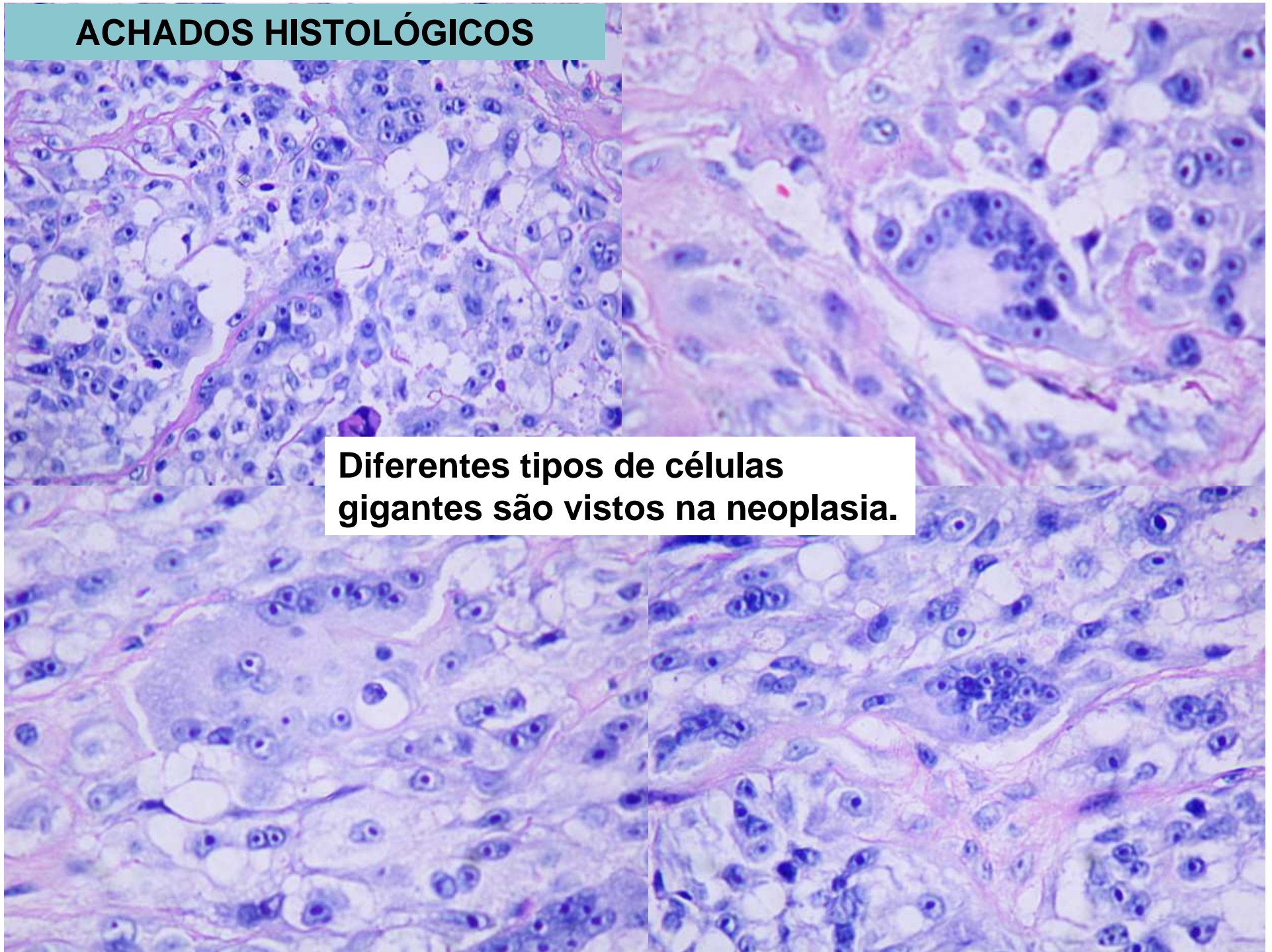
Mulher de 30 anos, com lesão saliente na nádega, que o cirurgião interpretou como lipoma.

ACHADOS HISTOLÓGICOS

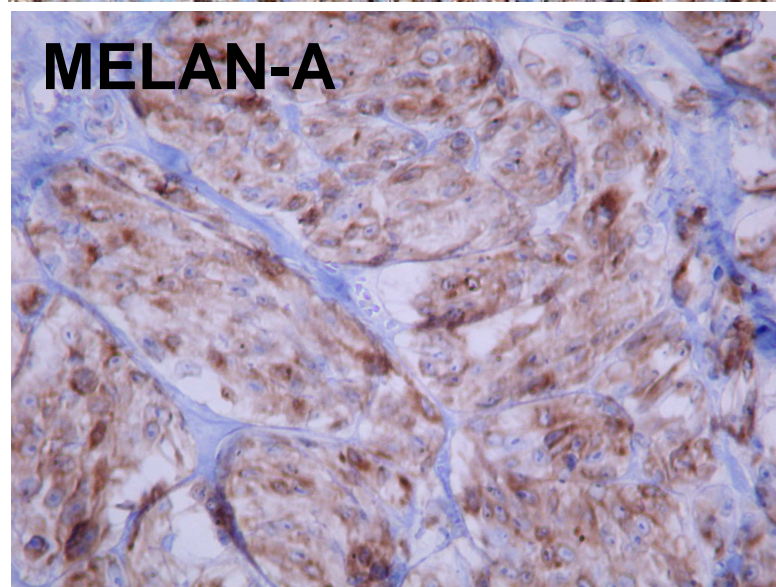
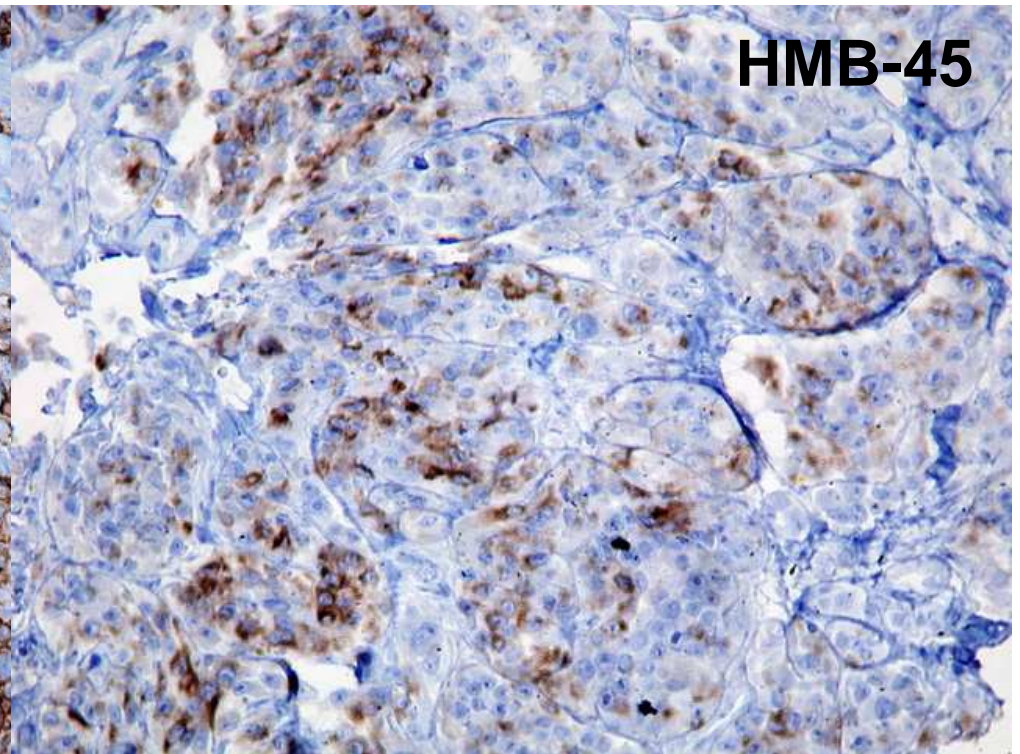
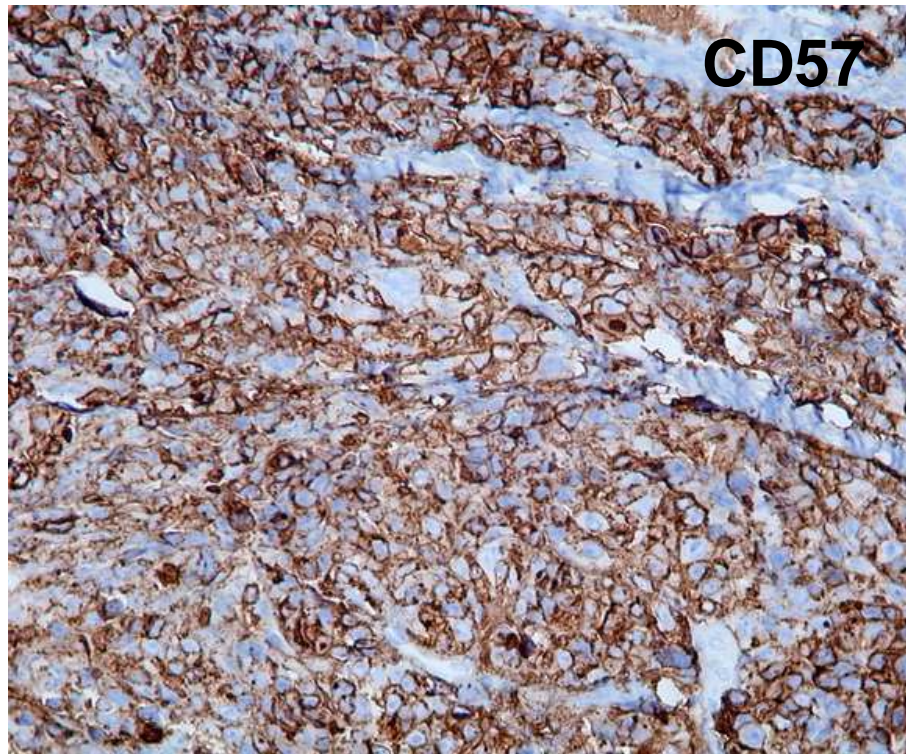


Grupos de células, muitas delas com citoplasma claro, aparecem separadas por colágeno.

ACHADOS HISTOLÓGICOS



ACHADOS DE IMUNO-HISTOQUÍMICA



As células neoplásicas mostram positividade para marcadores melanocíticos e para CD57.

DIAGNÓSTICO
É O MESMO PARA OS DOIS CASOS

**SARCOMA DE CÉLULAS
CLARAS DO TENDÃO
(PERIFÉRICO)**

CLEAR CELL SARCOMA—FACT SHEET

Definition

- ▶ A malignant melanocytic lesion that occurs within soft tissue, frequently in association with a tendon, differing from conventional malignant melanoma

Incidence and Location

- ▶ Lower extremity, relatively rare tumor

Morbidity and Mortality

- ▶ 50% metastatic potential with low long-term survival rate

Clinical Features

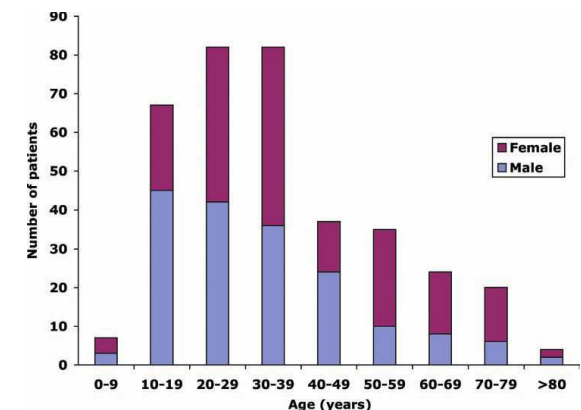
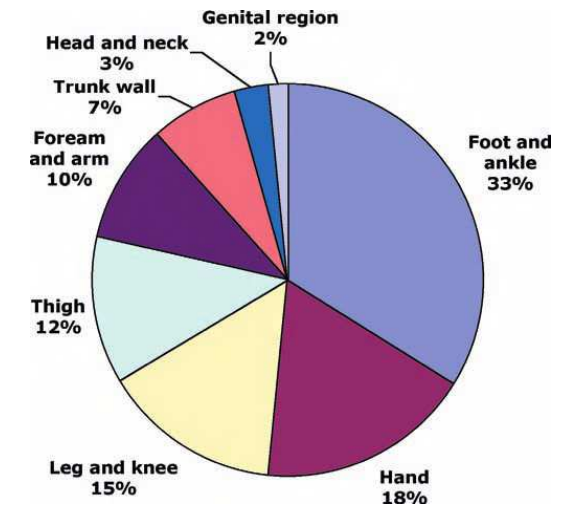
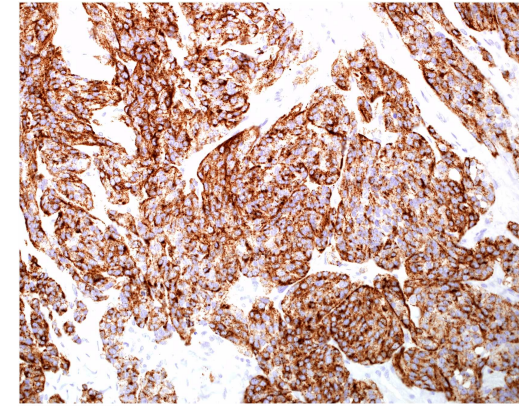
- ▶ 20 to 40 years, lower extremity tendinous painful mass with no overlying ulceration

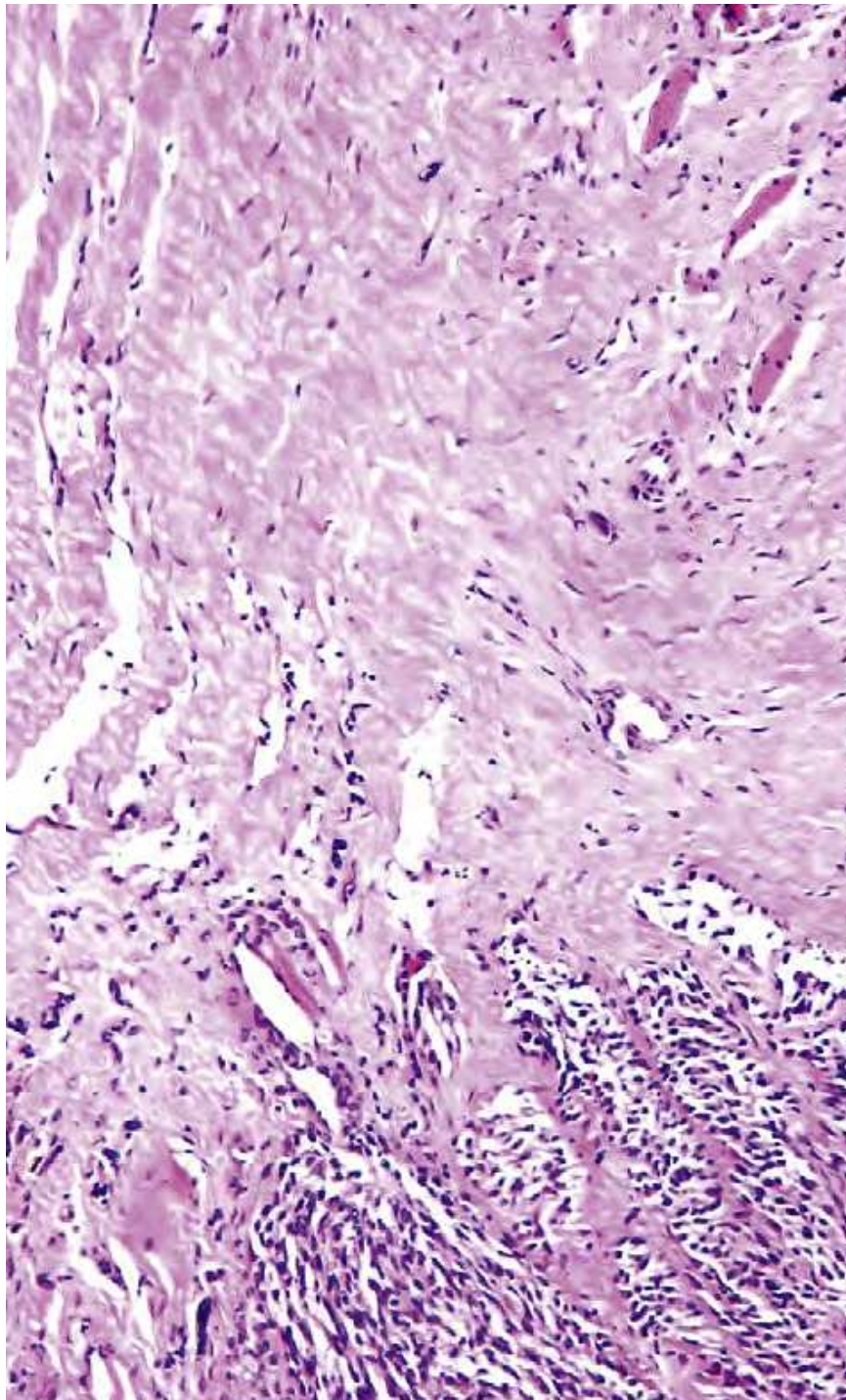
Radiologic Features

- ▶ Intratendinous mass with possible hyperintensity on T1-weighted MRI

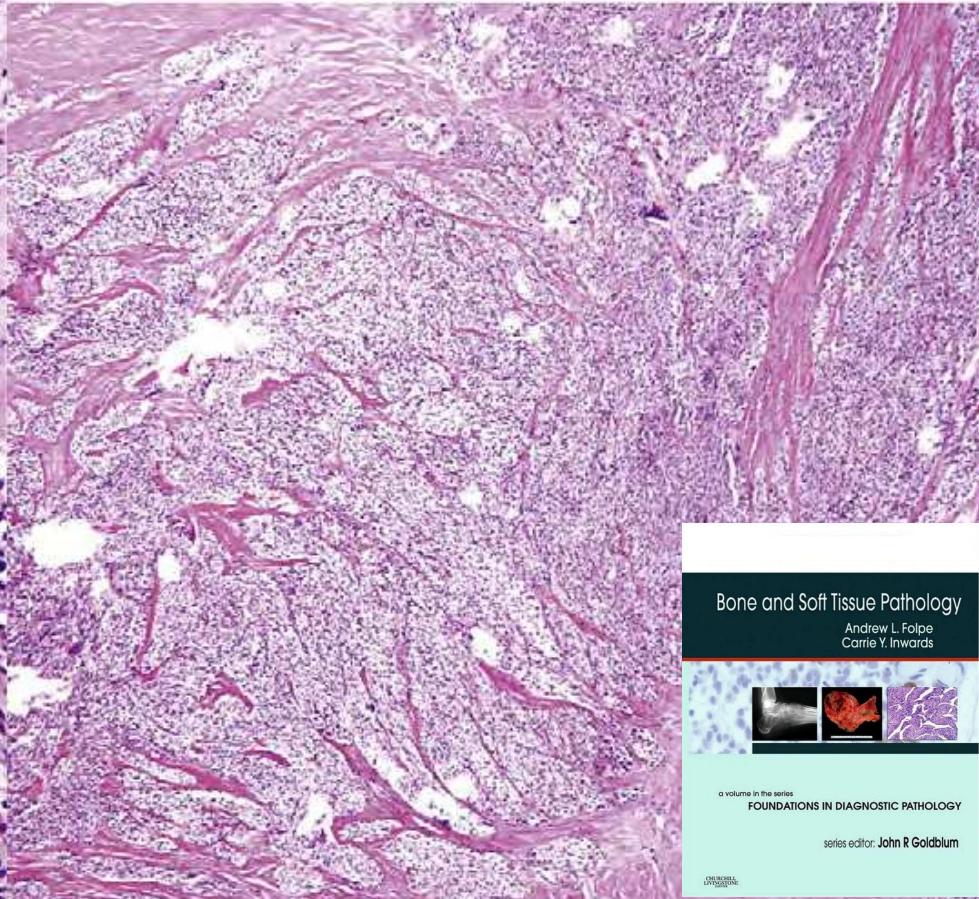
Prognosis and Treatment

- ▶ Poor; adjuvant therapy advocated but largely ineffective

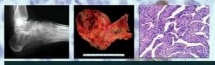





As células neoplásicas infiltram conjuntivo denso (tendão) e se compartimentalizam em pequenos ninhos.

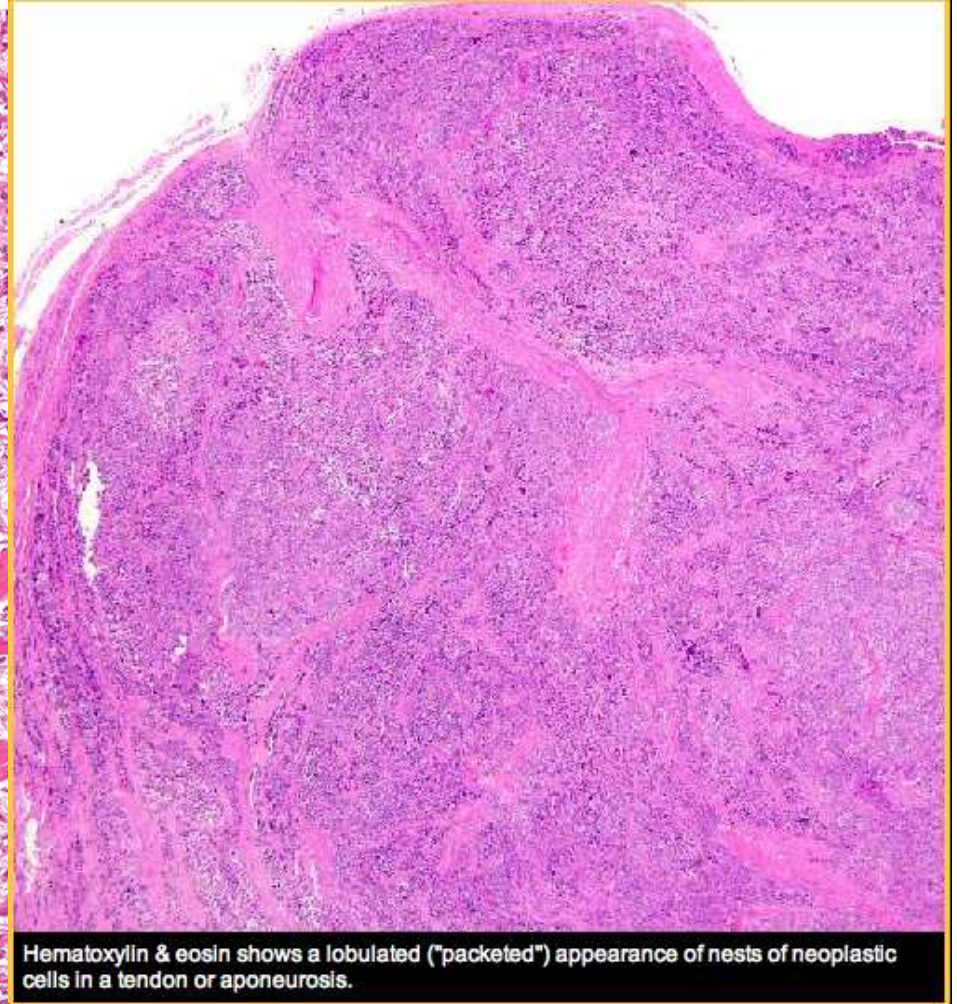
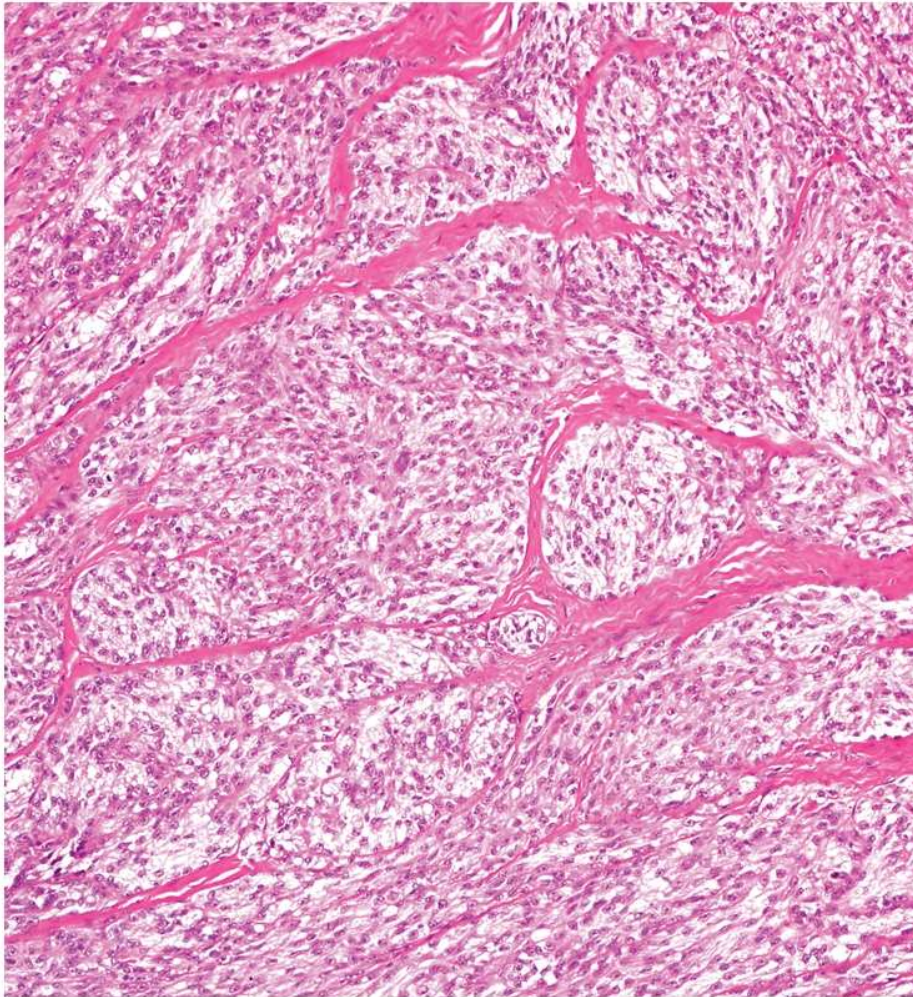


Bone and Soft Tissue Pathology
Andrew L. Folbe
Carrie Y. Inwards



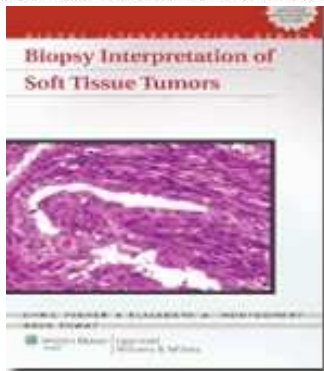
a volume in the series
FOUNDATIONS IN DIAGNOSTIC PATHOLOGY
series editor: John R Goldblum



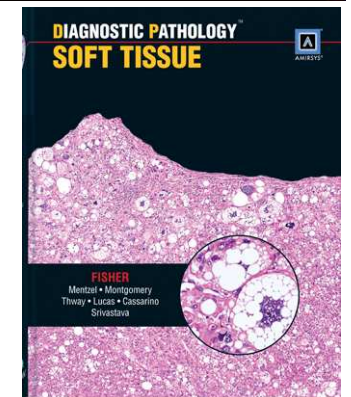


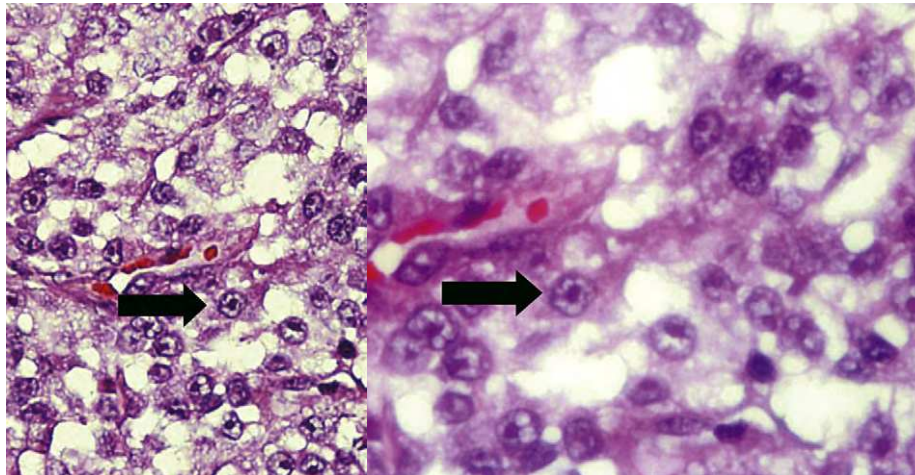
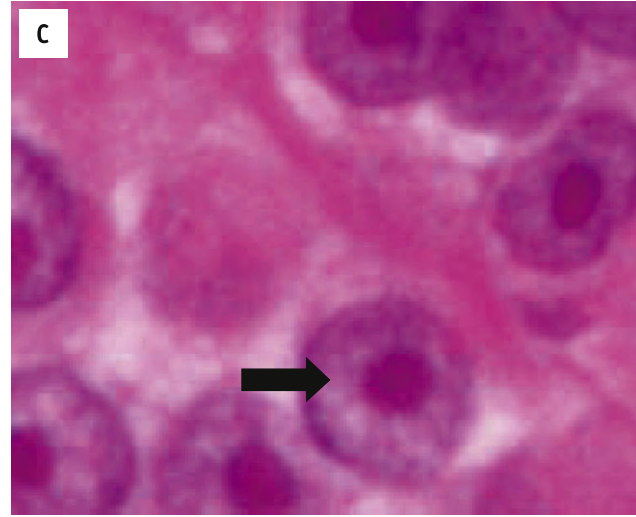
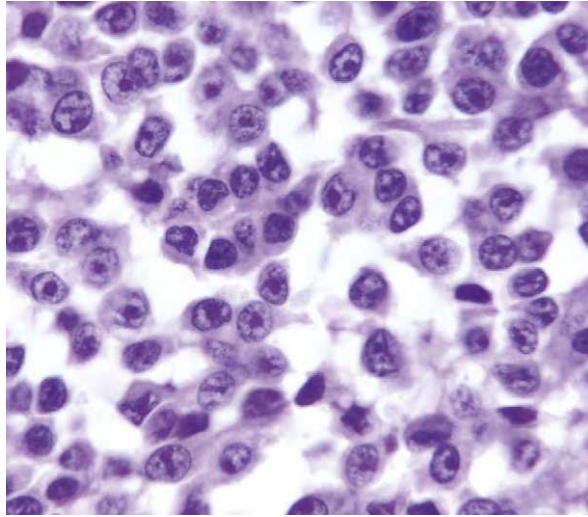
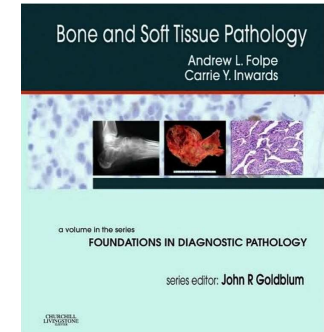
Hematoxylin & eosin shows a lobulated ("packeted") appearance of nests of neoplastic cells in a tendon or aponeurosis.

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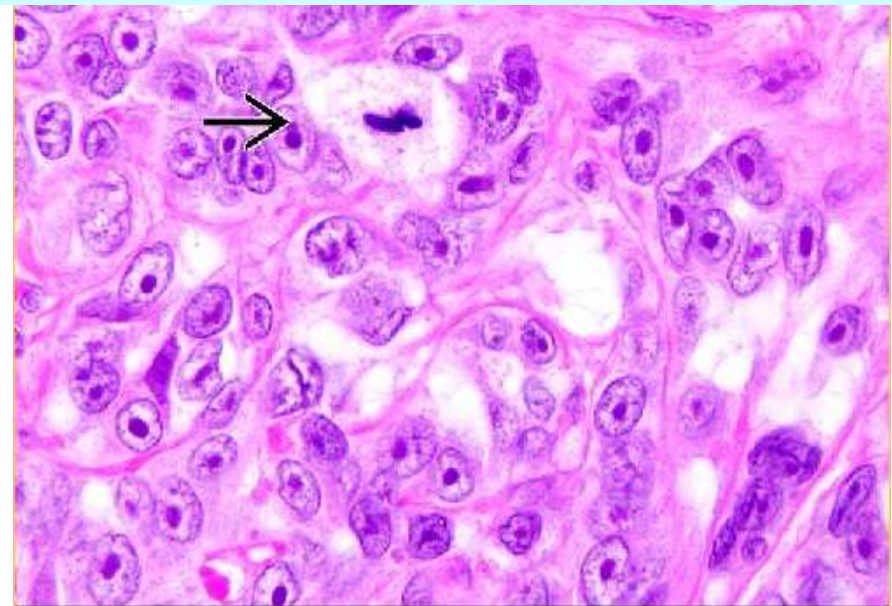
Sarcoma de células claras. Células tumorais uniformes formam ninhos de tamanhos variados, separados por septos fibrosos de espessuras variáveis.

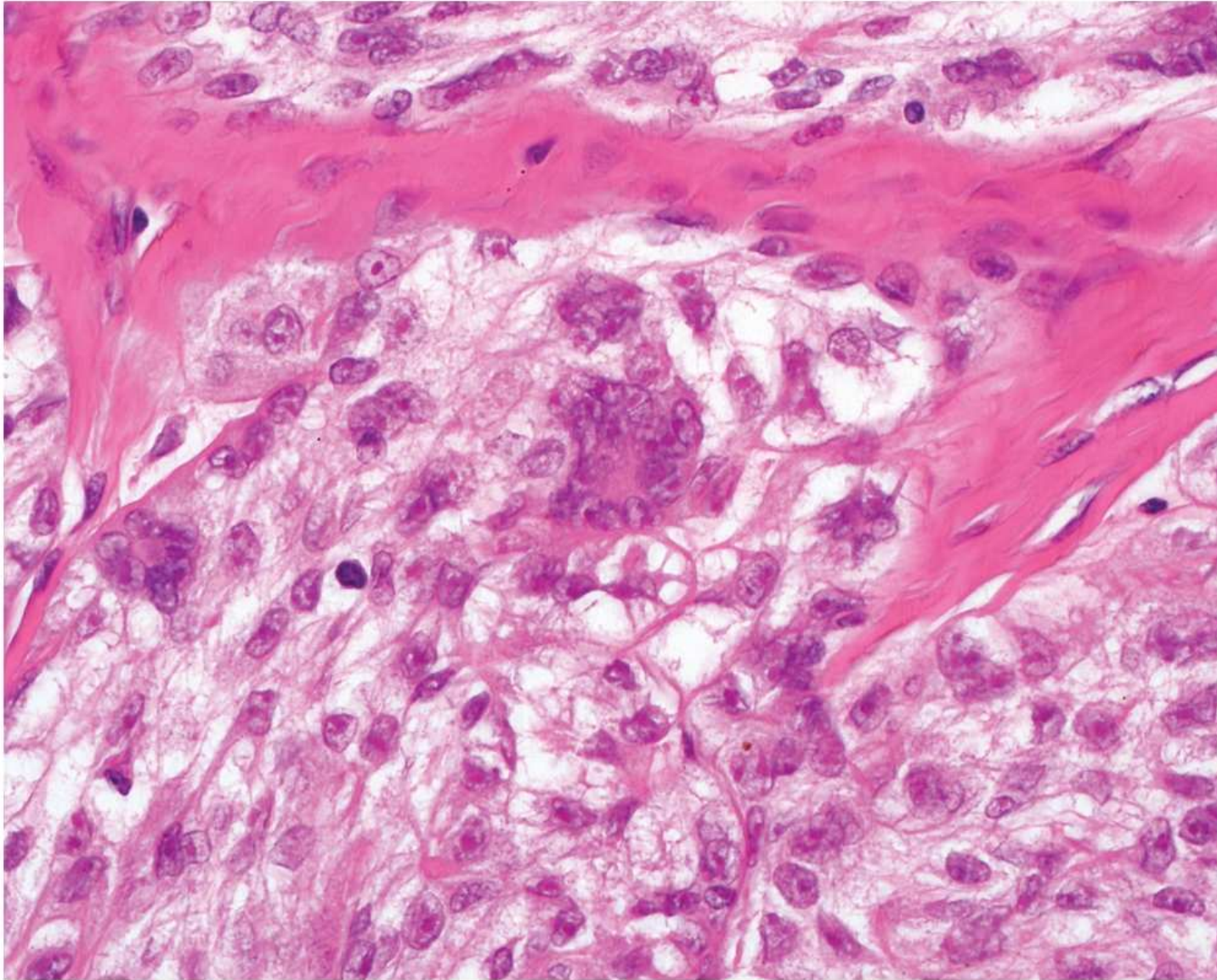




As mitoses não são atípicas porque a neoplasia resulta de translocação e não possui instabilidade cromossômica.

Qualquer que seja a aparência das células neoplásicas, os seus núcleos apresentam nucléolos proeminentes.

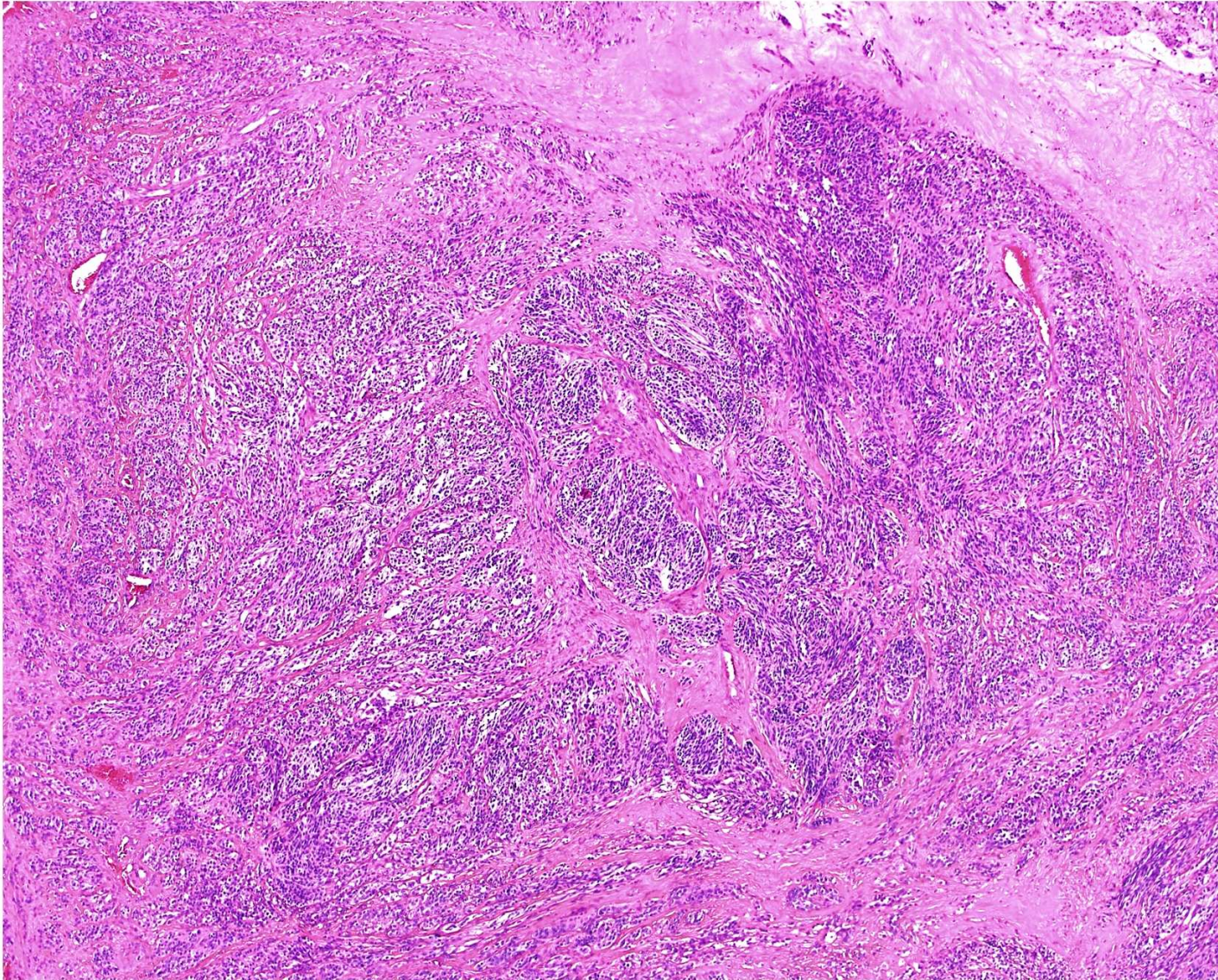




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Sarcoma de células claras. As células têm núcleos redondos, nucléolos distintos e moderada quantidade de citoplasma anfófilico. A célula multinucleada que aparece é um achado frequente nesse tipo de tumor .

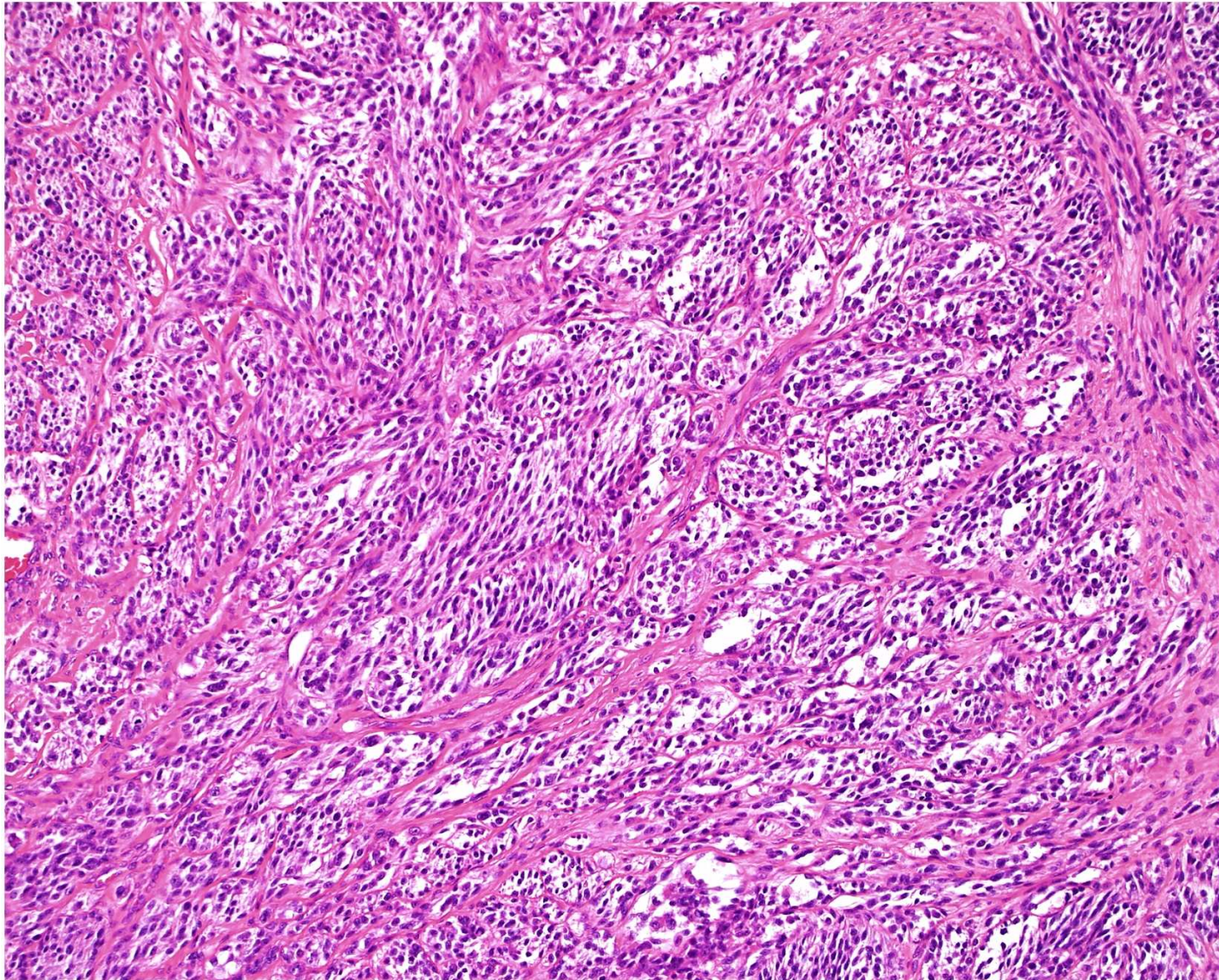




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Clear cell sarcoma. Nodules of tumor are dispersed in dense fibrous tissue.

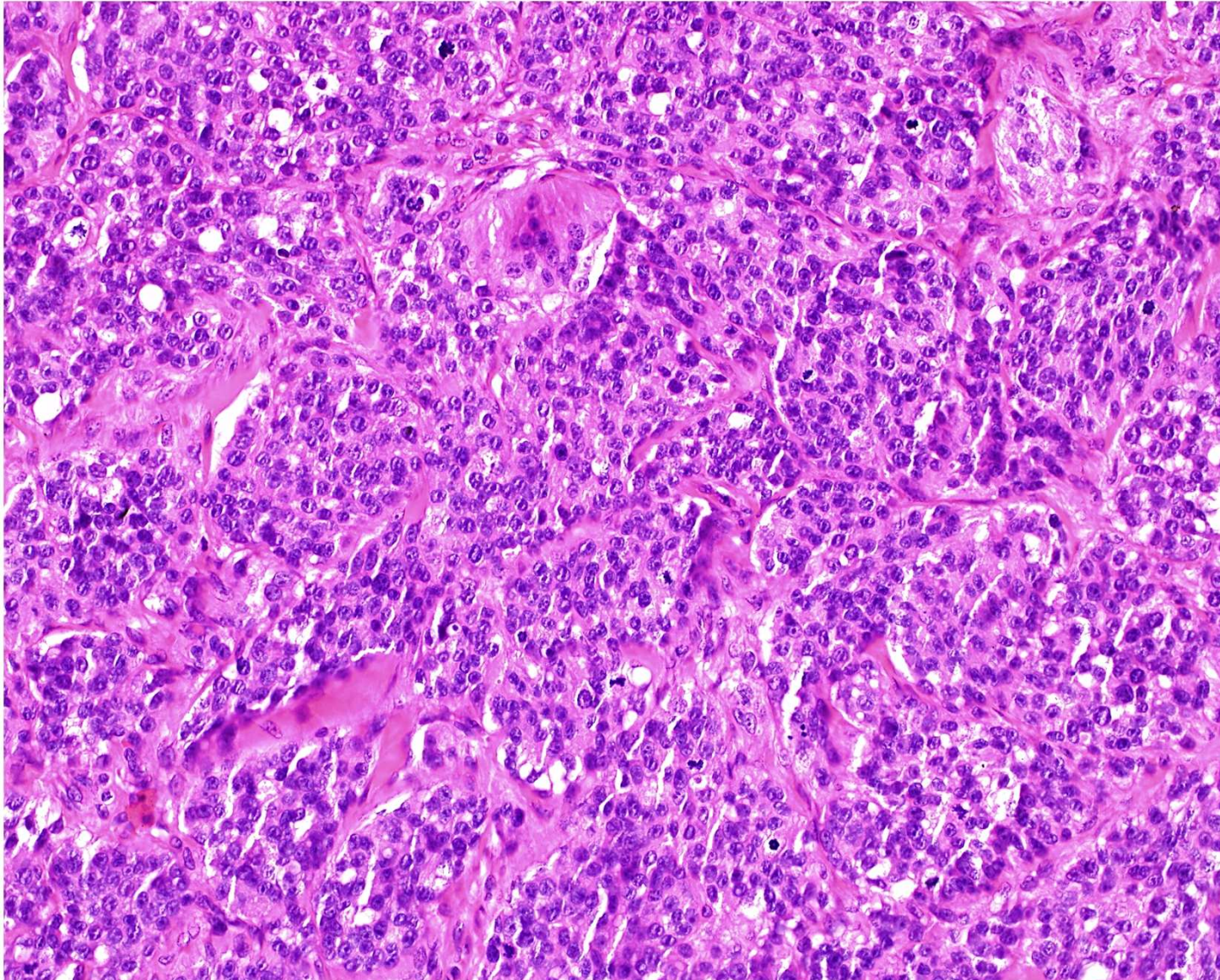




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Clear cell sarcoma. The tumor forms nests and sheaves of polygonal and more spindled cells. Note variation in thickness of fibrous septa.

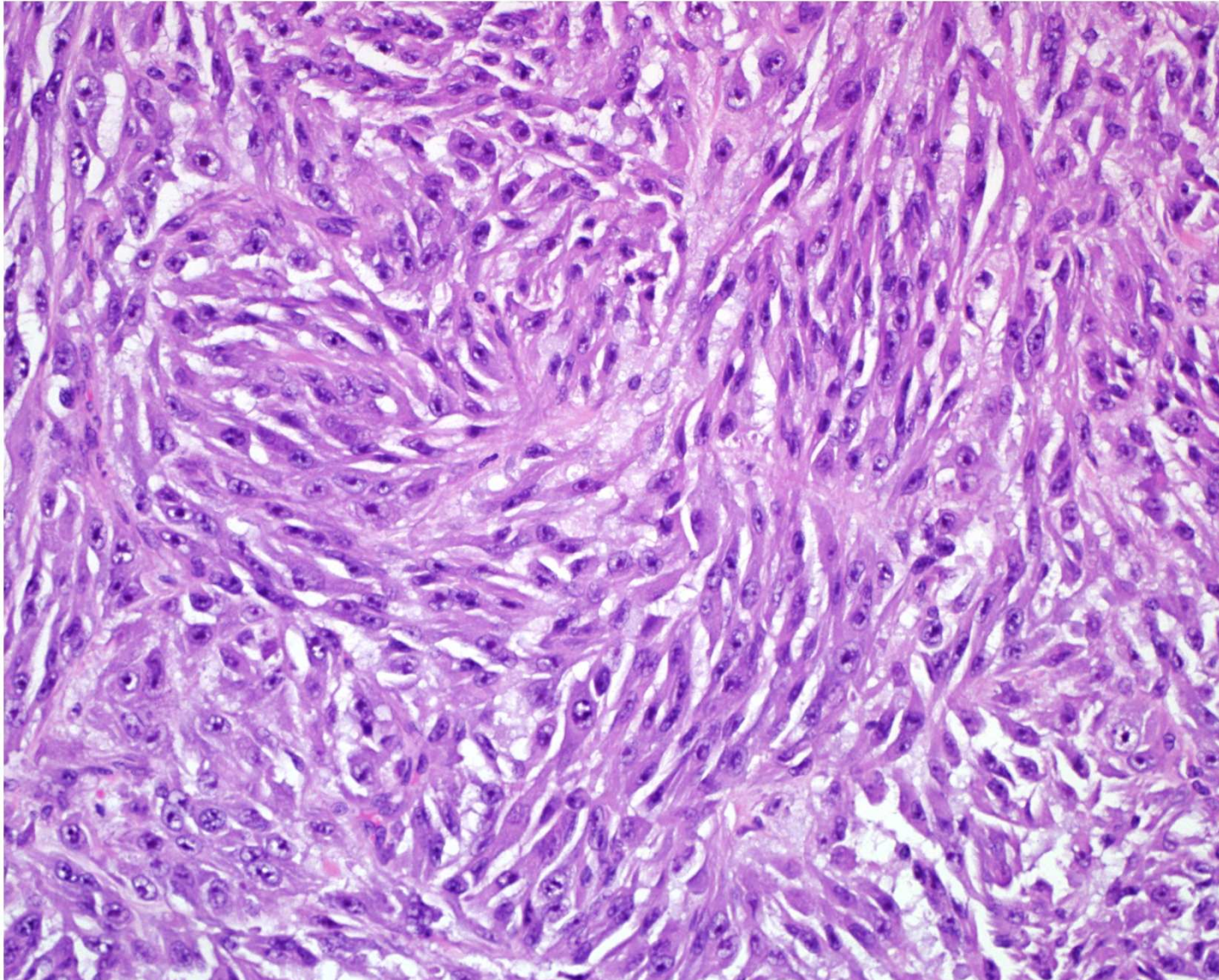




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Clear cell sarcoma. The cells are uniform with rounded central nuclei and small nucleoli. Note the multinucleated giant cell (upper center).

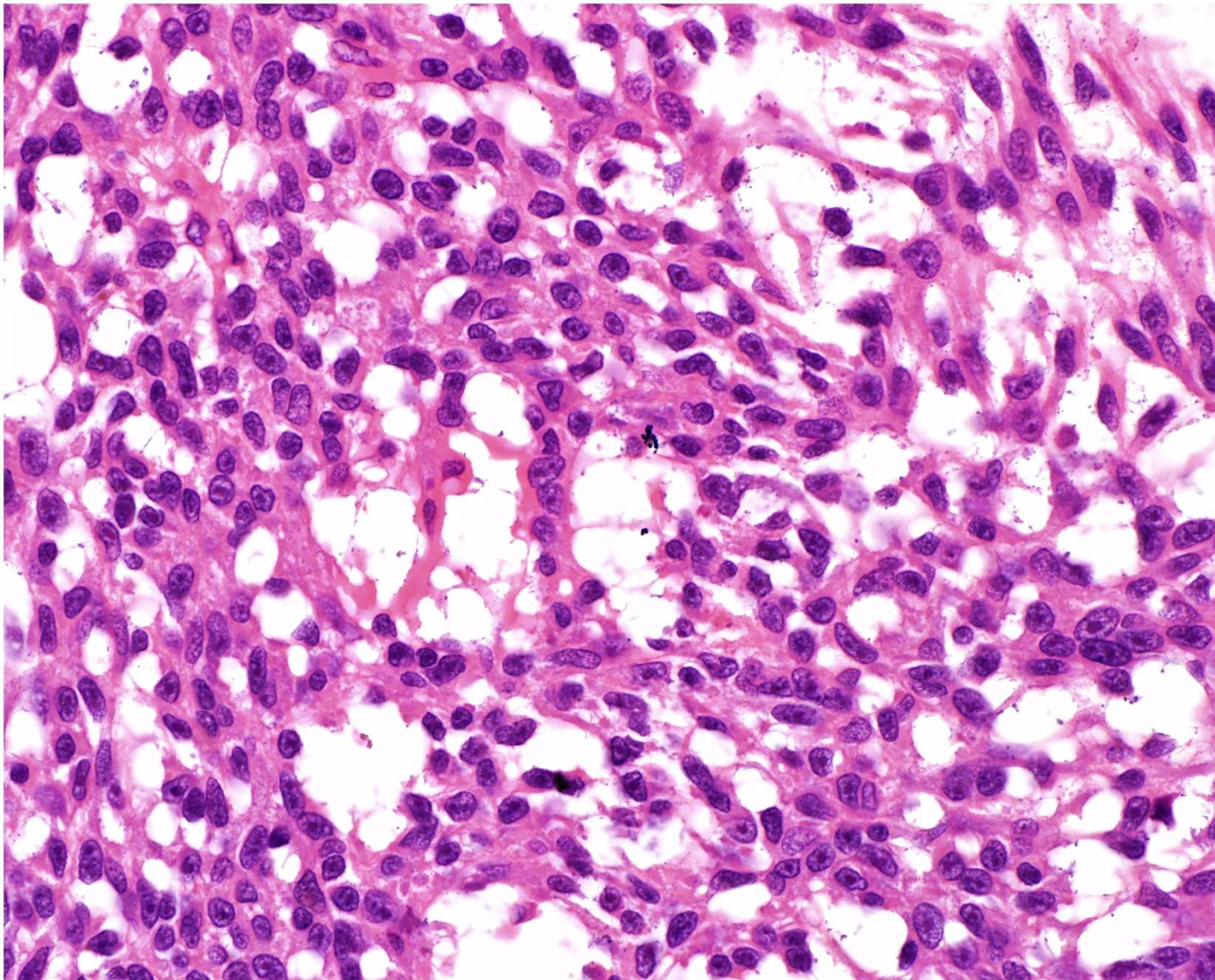




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Clear cell sarcoma. An area with prominent spindling. The nuclear features, with prominent nucleoli, are preserved.

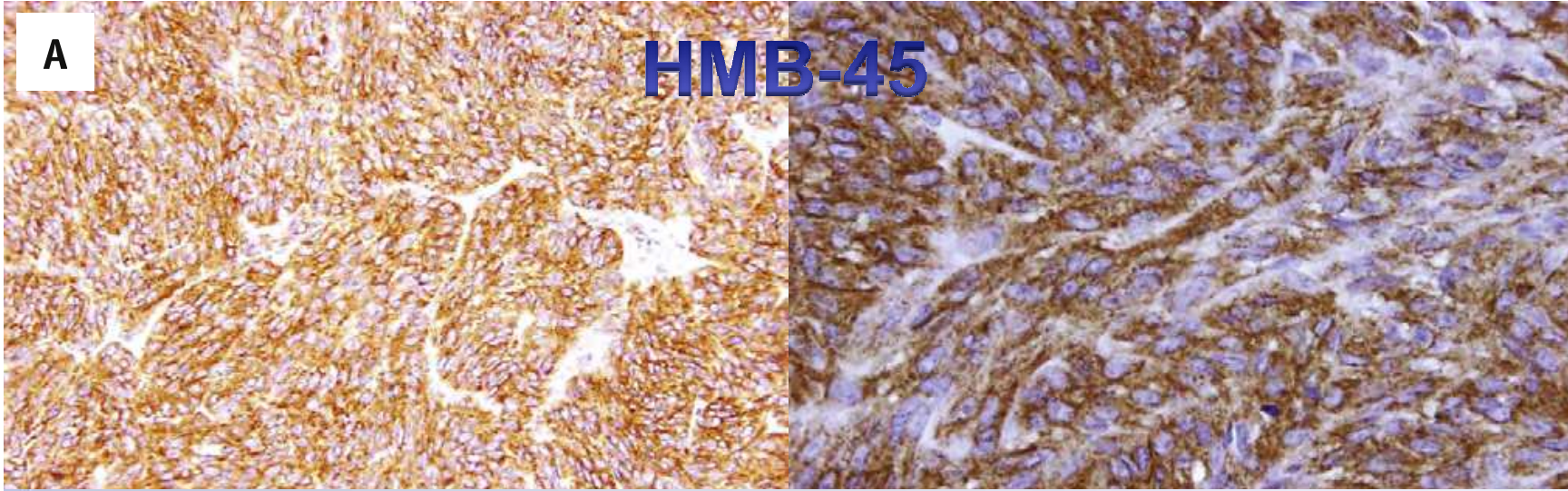




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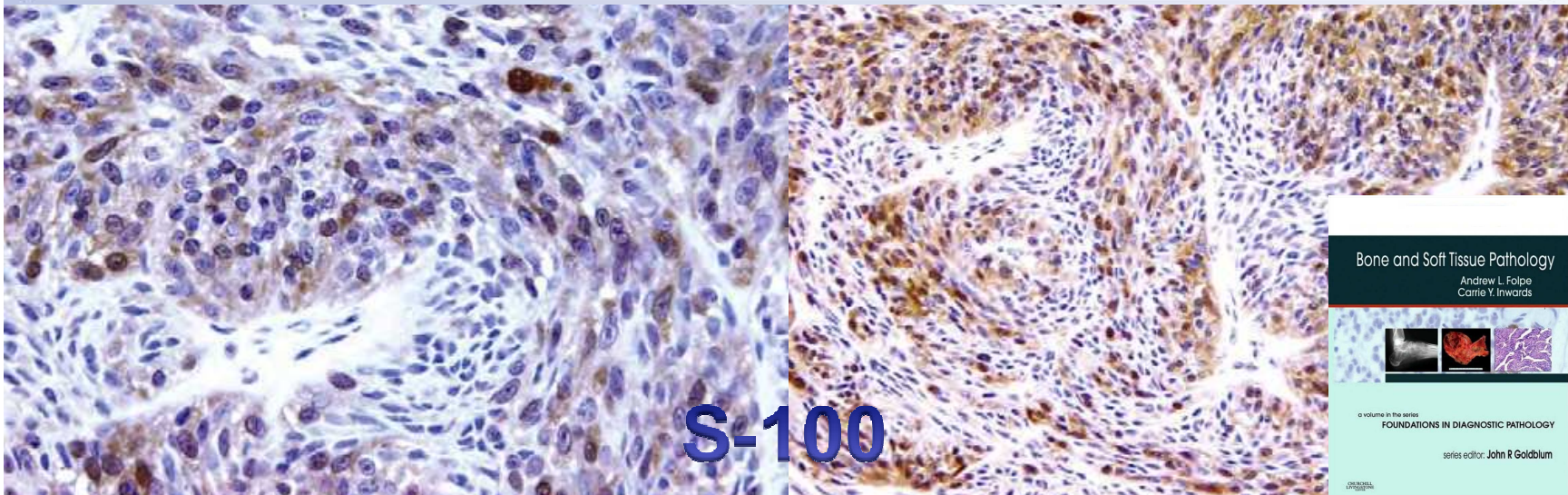
Clear cell sarcoma. Myxoid stroma change is an occasional feature.

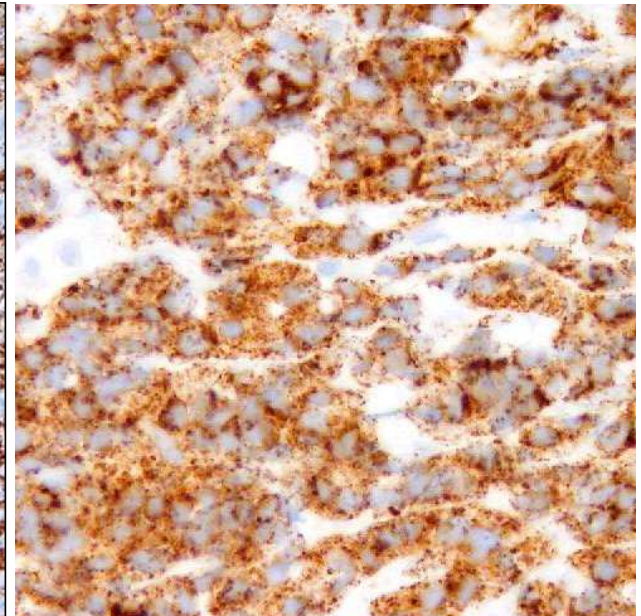
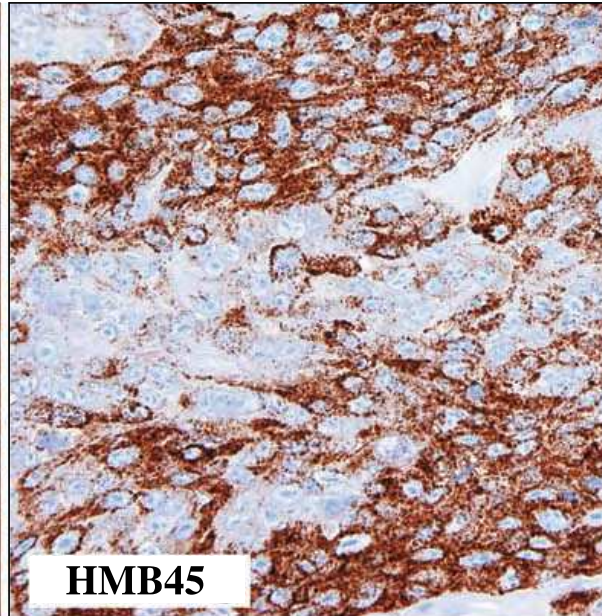
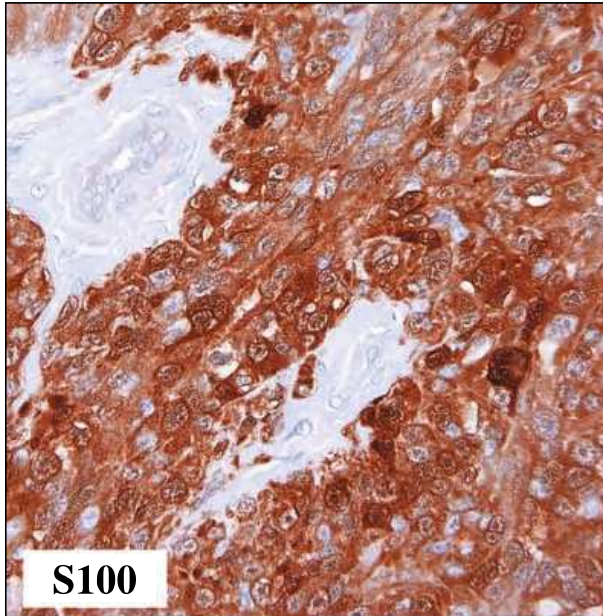




Immunohistochemical Findings

- ▶ Express S-100 protein, melanocytic markers
- ▶ Molecular features: t(12;22) (EWS-ATF1)





HMB-45 shows cytoplasmic labeling in most clear cell sarcomas of soft tissue, although melanocytic markers are often negative in clear cell sarcomas of the gastrointestinal tract.

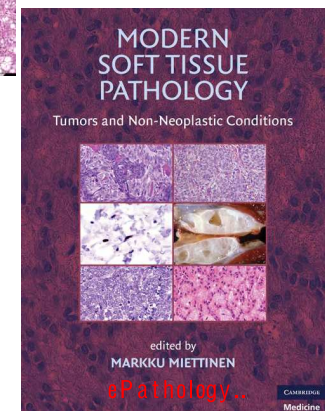
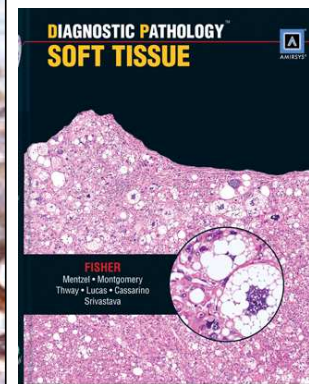
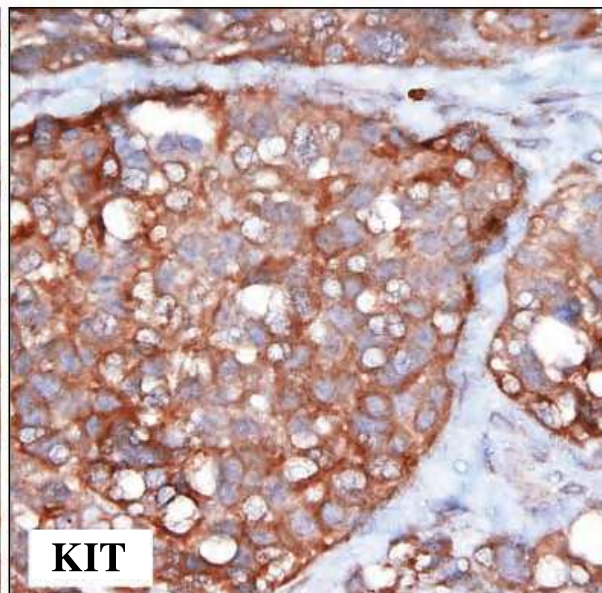
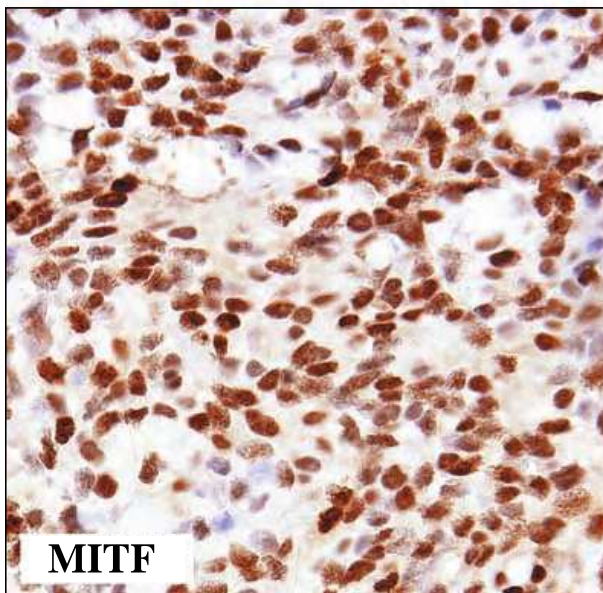
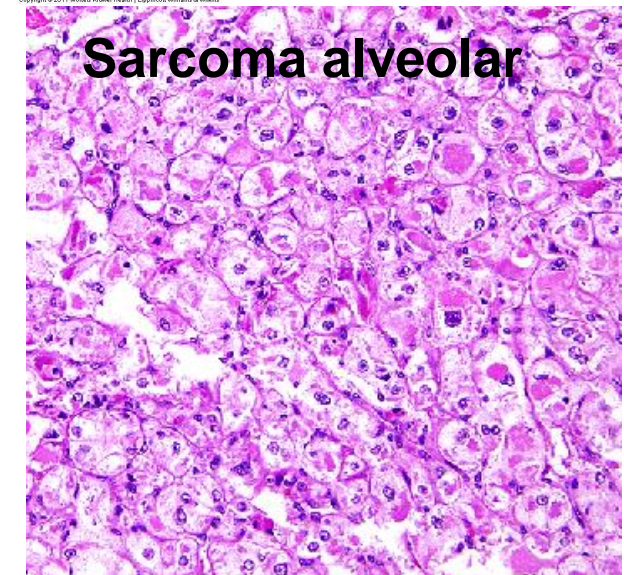
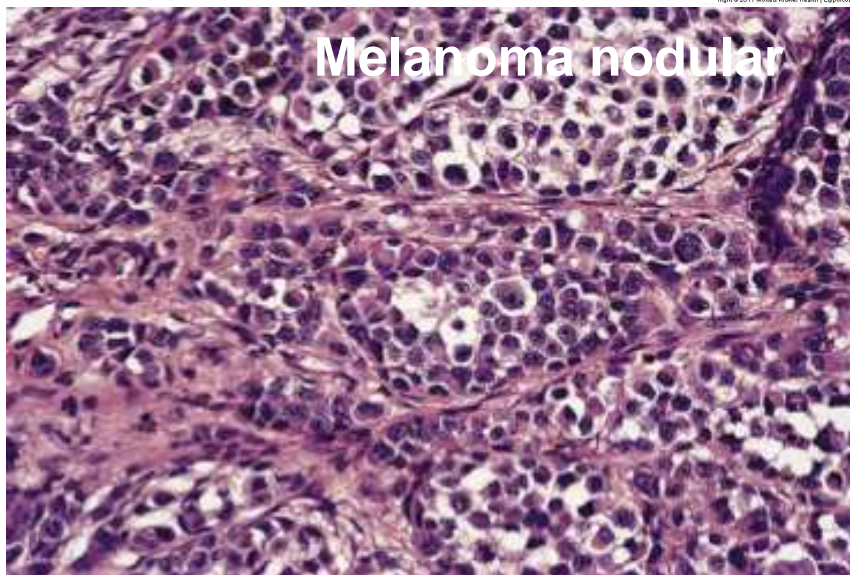
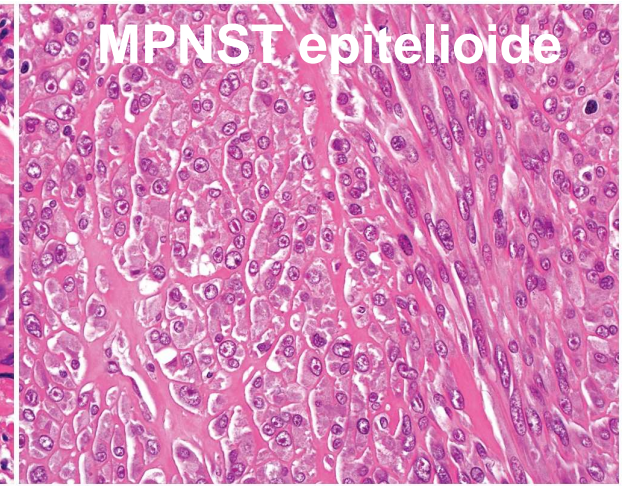
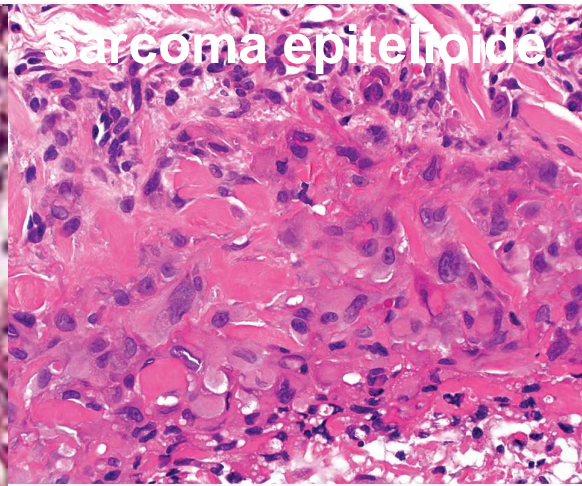
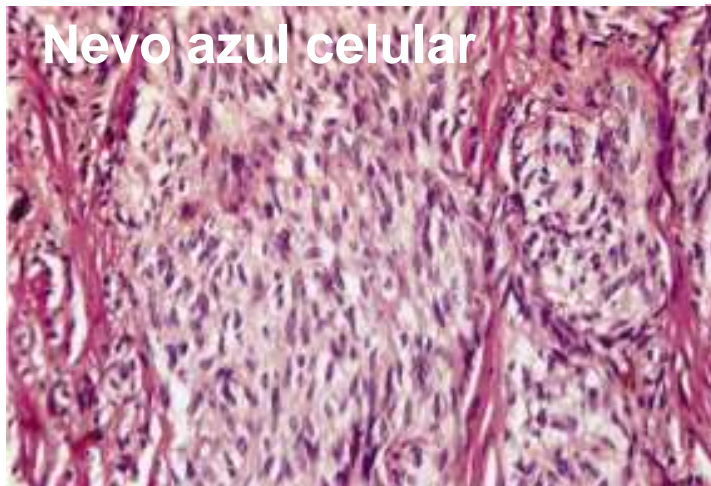


FIGURE 25.22: Immunohistochemical findings in clear cell sarcoma. Melanocytic markers are typically expressed, whereas KIT positivity varies.

Differential Diagnosis

- ▶ Conventional malignant melanoma, epithelioid MPNST, epithelioid sarcoma, cellular blue nevus



▼ KEY FACTS

Terminology expand/collapse.

- Sarcoma showing melanocytic differentiation
- Malignant melanoma of soft parts

Clinical Issues

- Young adults (3rd or 4th decade)
- Usually affects extremities (> 90%)
 - Foot most common site
- Behaves as high-grade sarcoma
- Metastasizes to lymph nodes and lung
- Often attached to tendon or aponeurosis
- Occasional visceral examples
 - Ileum most common GI site

Microscopic Pathology

- Monotonous clear to spindled cells arranged in nests/packets usually infiltrating tendon sheath
- Prominent uniform nucleoli
- Scattered wreath-like tumor giant cells
- Most cases have few mitoses

Ancillary Tests

- Immunohistochemical profile like that of melanoma in most cases
- $t(12;22)(q13;q12)$
 - *EWS-ATF1* fusion (detectable by PCR)
- *EWS-CREB1* fusion: Often in gastrointestinal CCS (lacks melanocytic differentiation)
 - Cases express S100 protein but not melanocytic markers
 - Same transcripts found in angiomatoid fibrous histiocytoma (different morphology, excellent prognosis)

