



Sociedade Brasileira de Patologia

Lilian Elizabeth Giménez González
Departamento de Anatomia Patológica



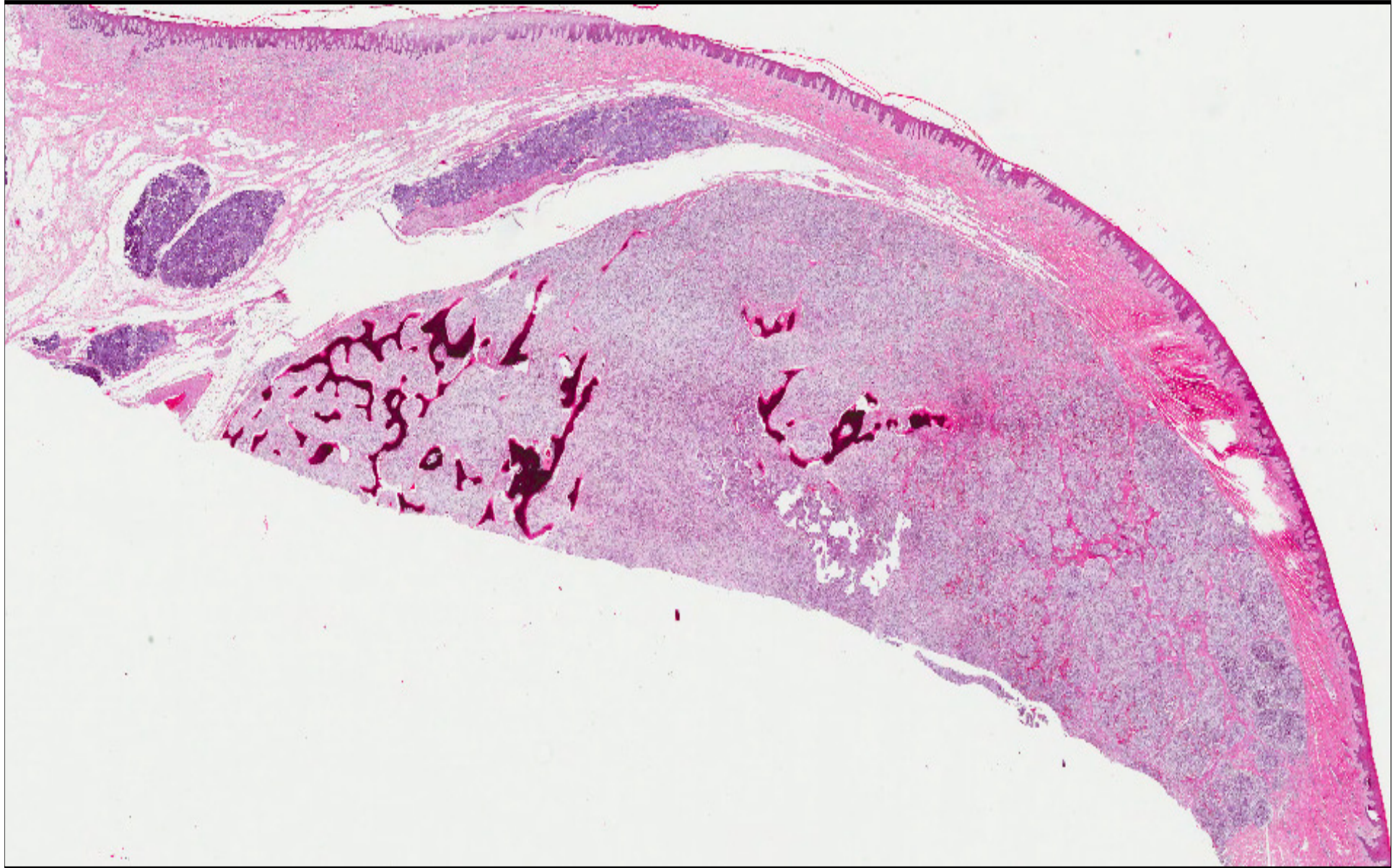
A.C. Camargo
Cancer Center

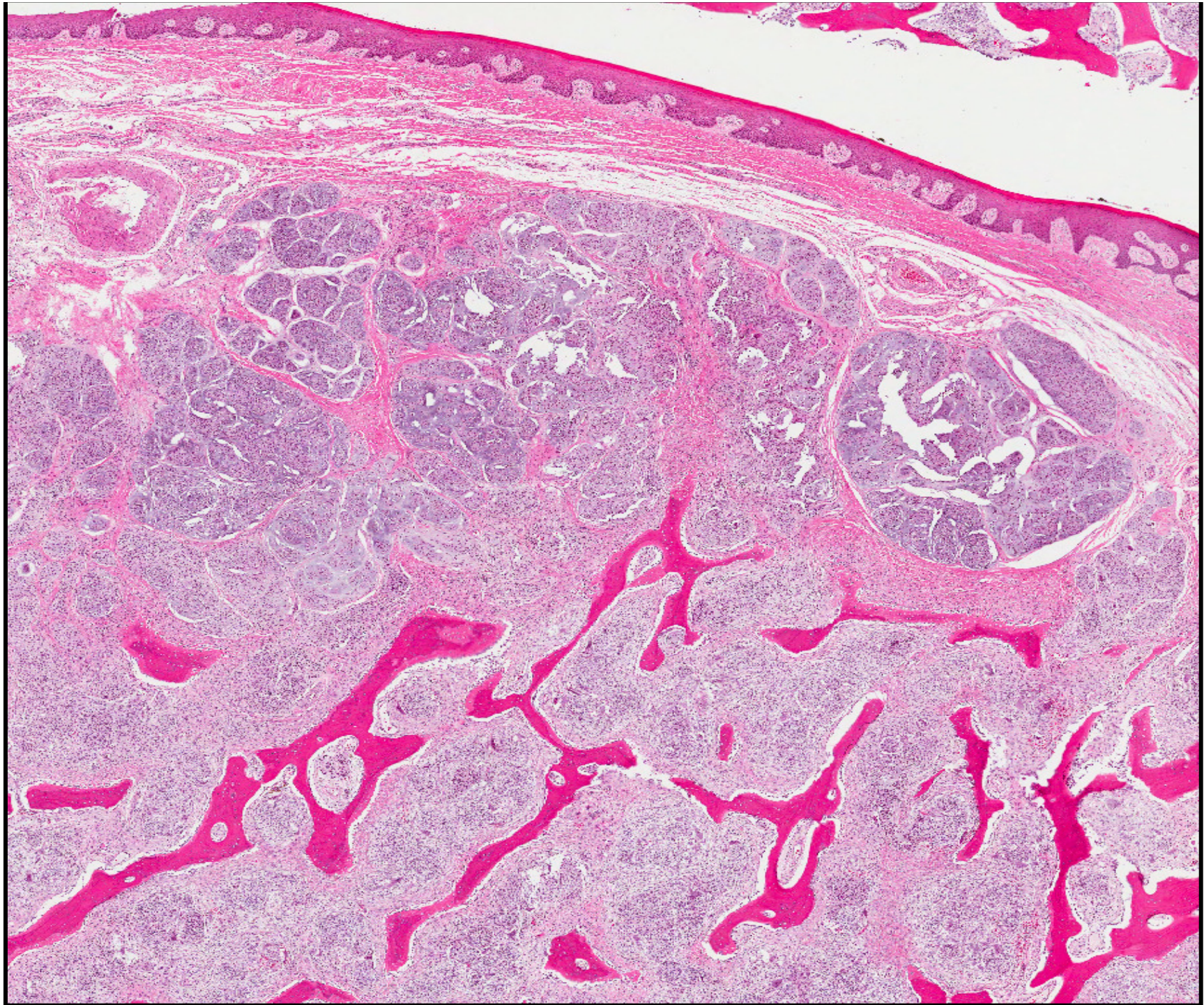
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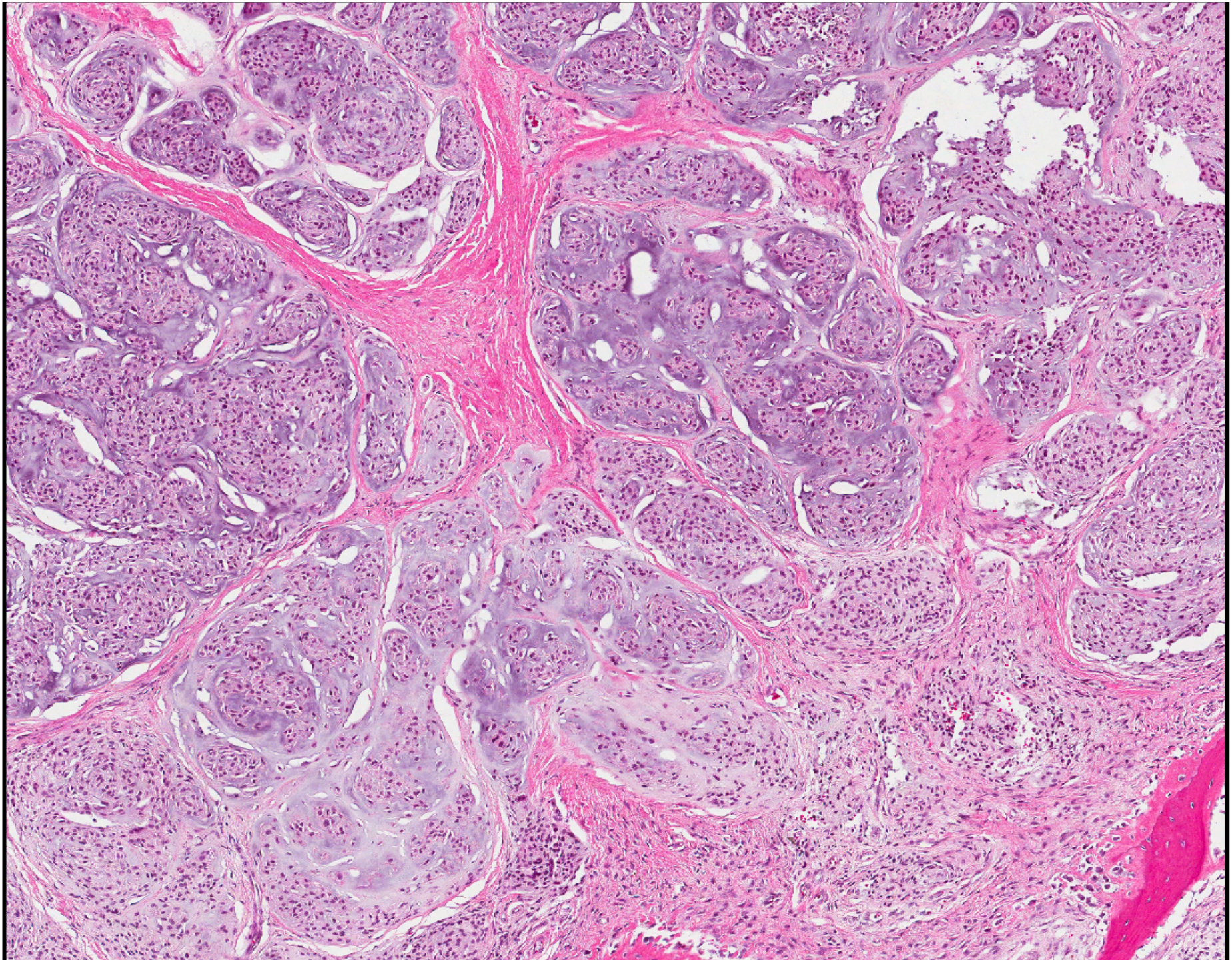


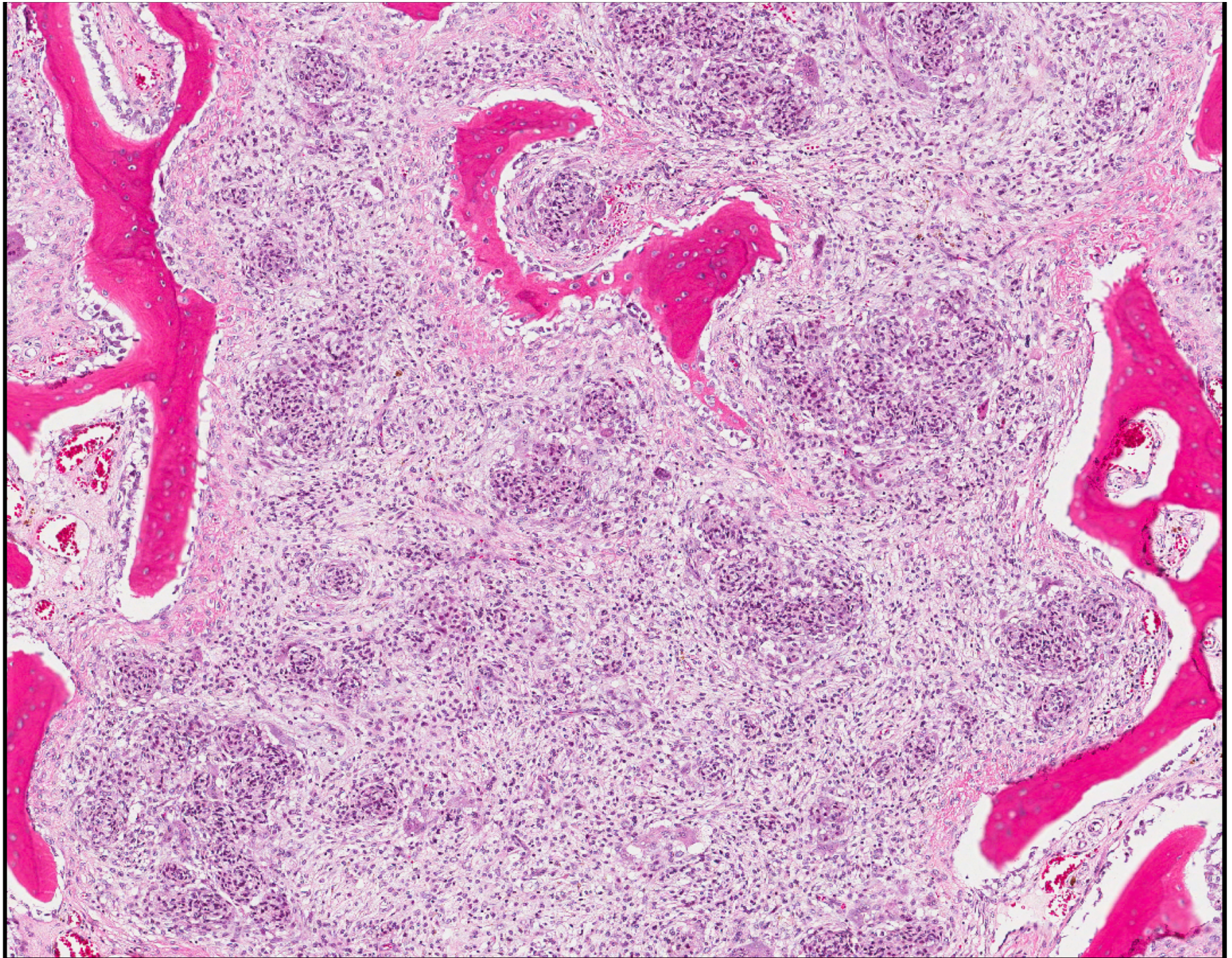
Feminino, 42 anos

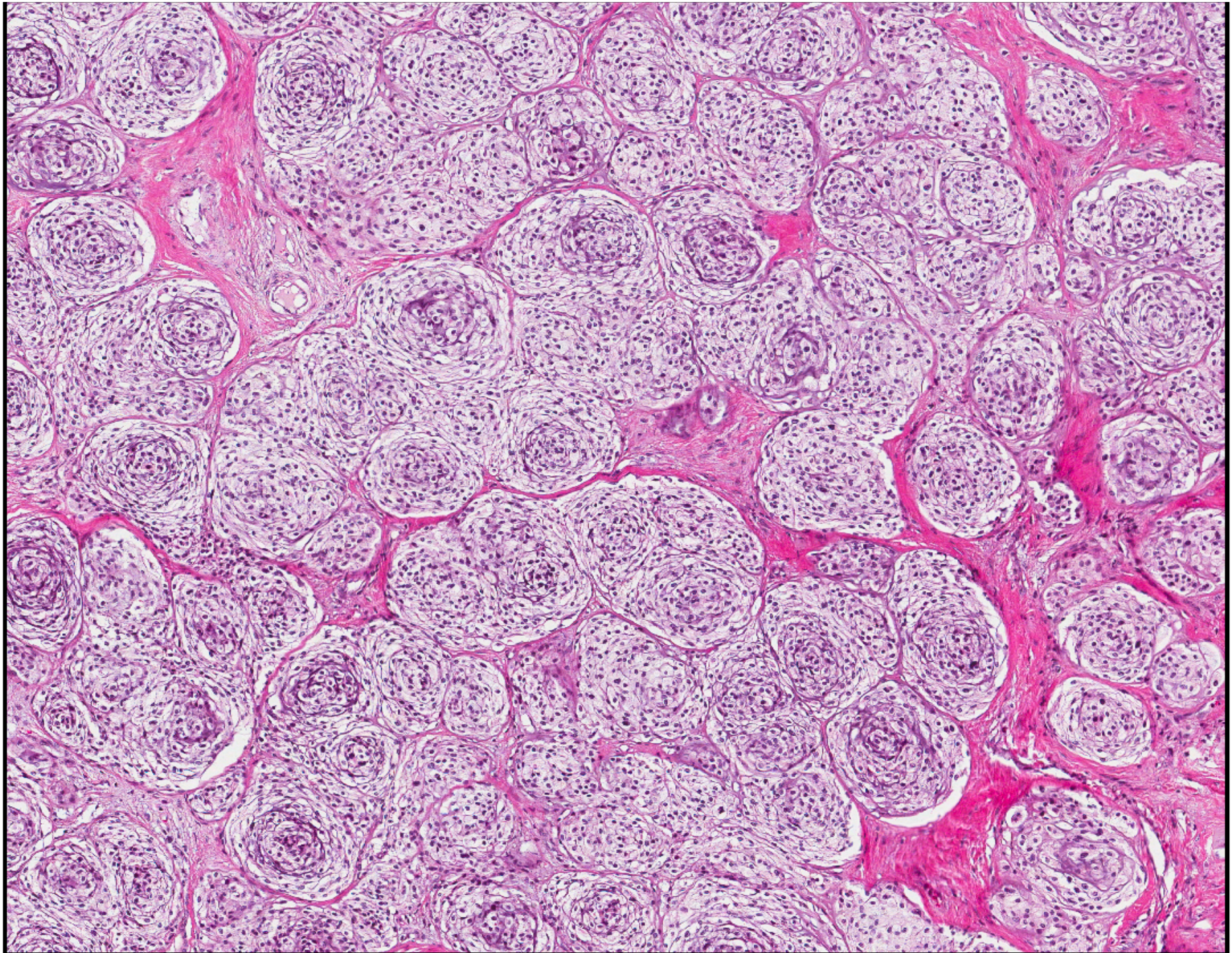
Ressecção de tumoração maxilar

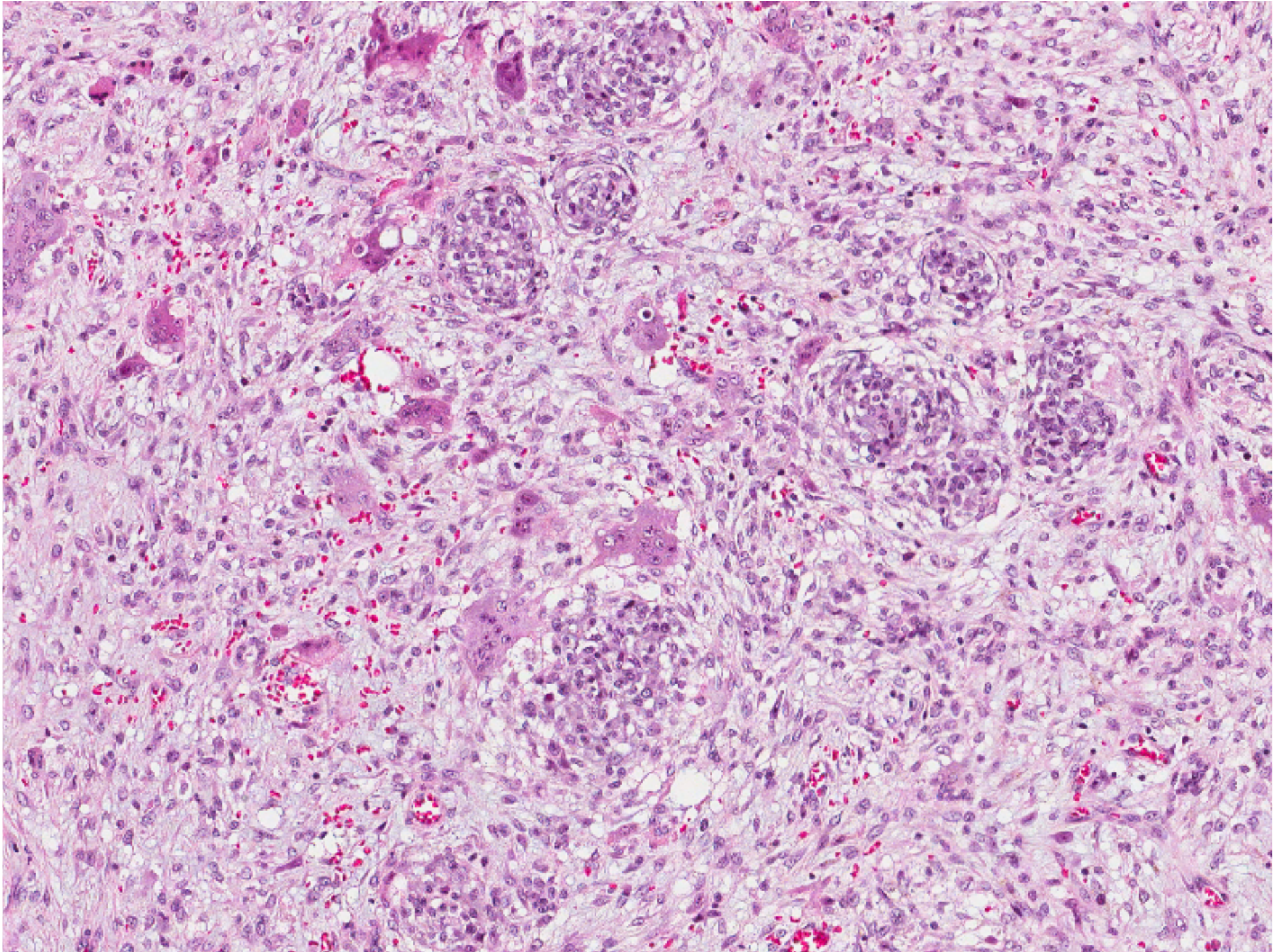


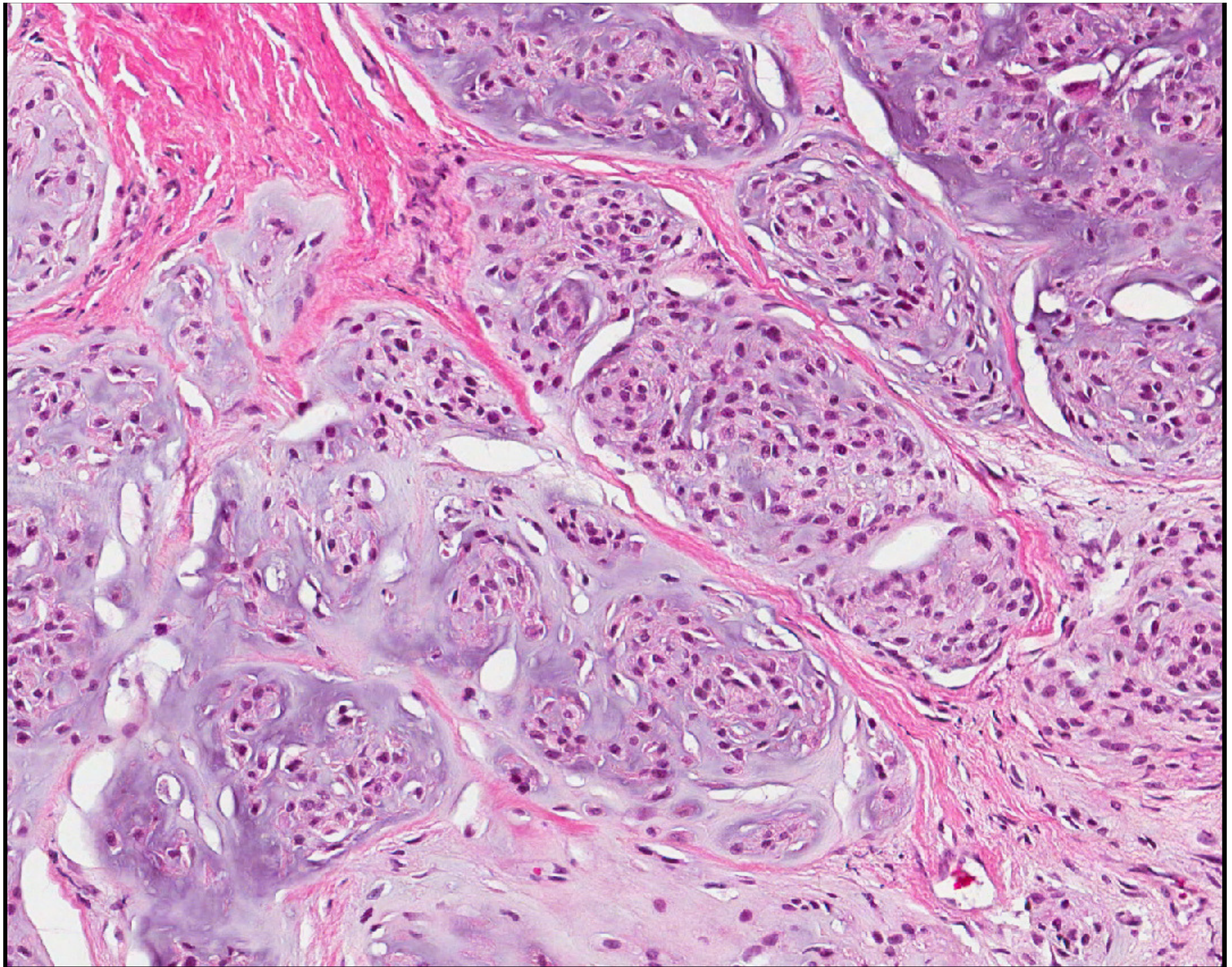


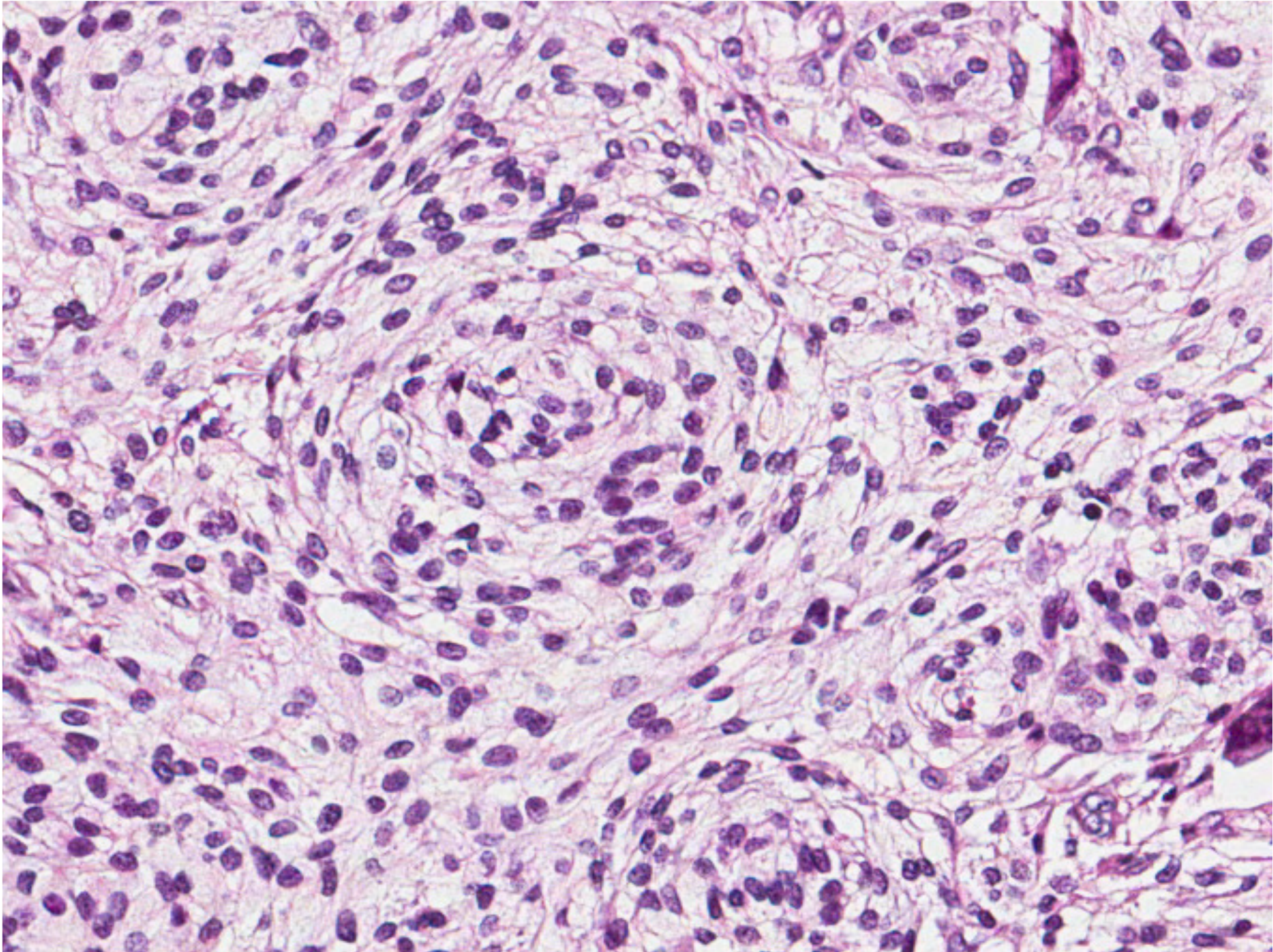










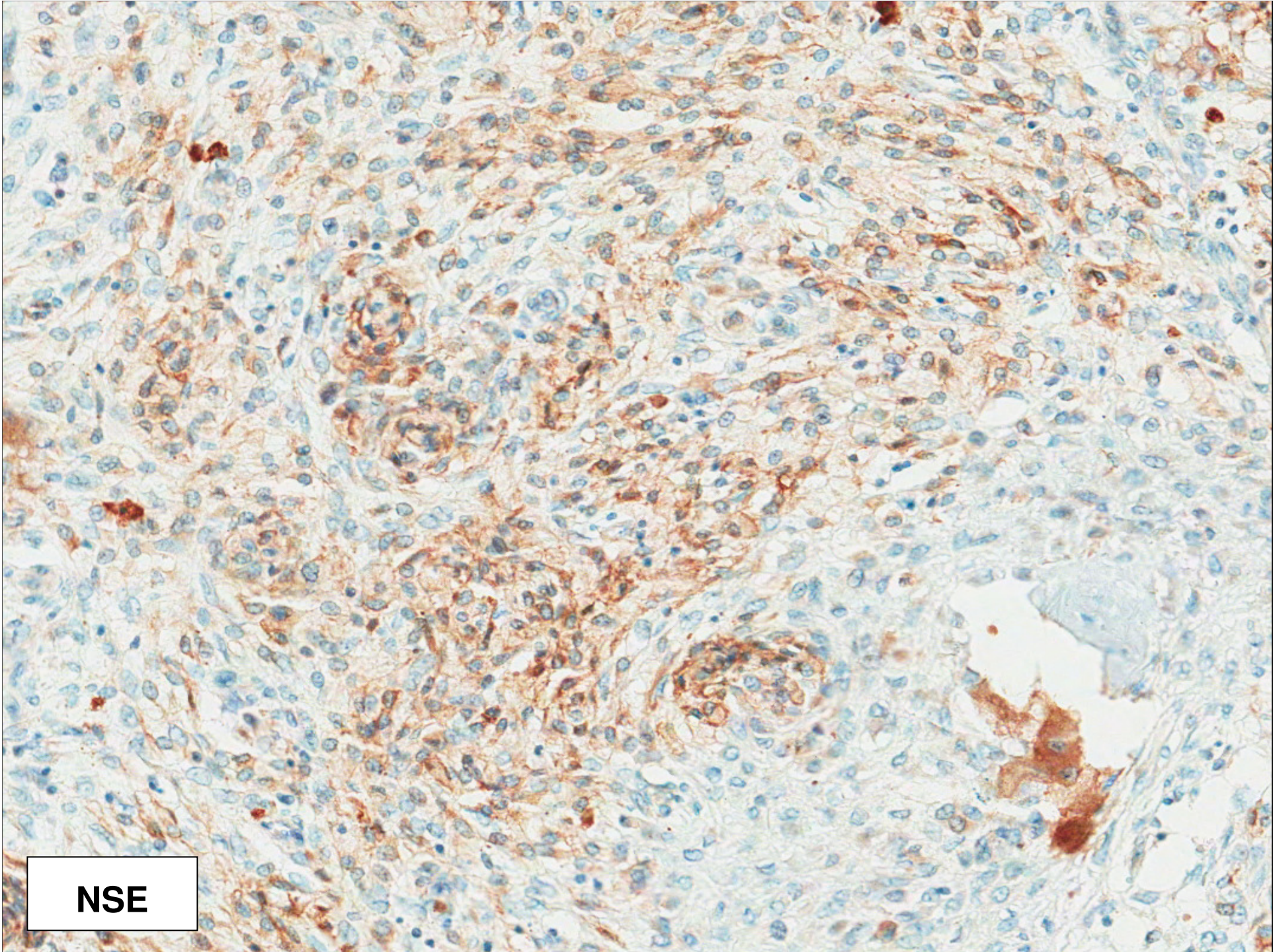




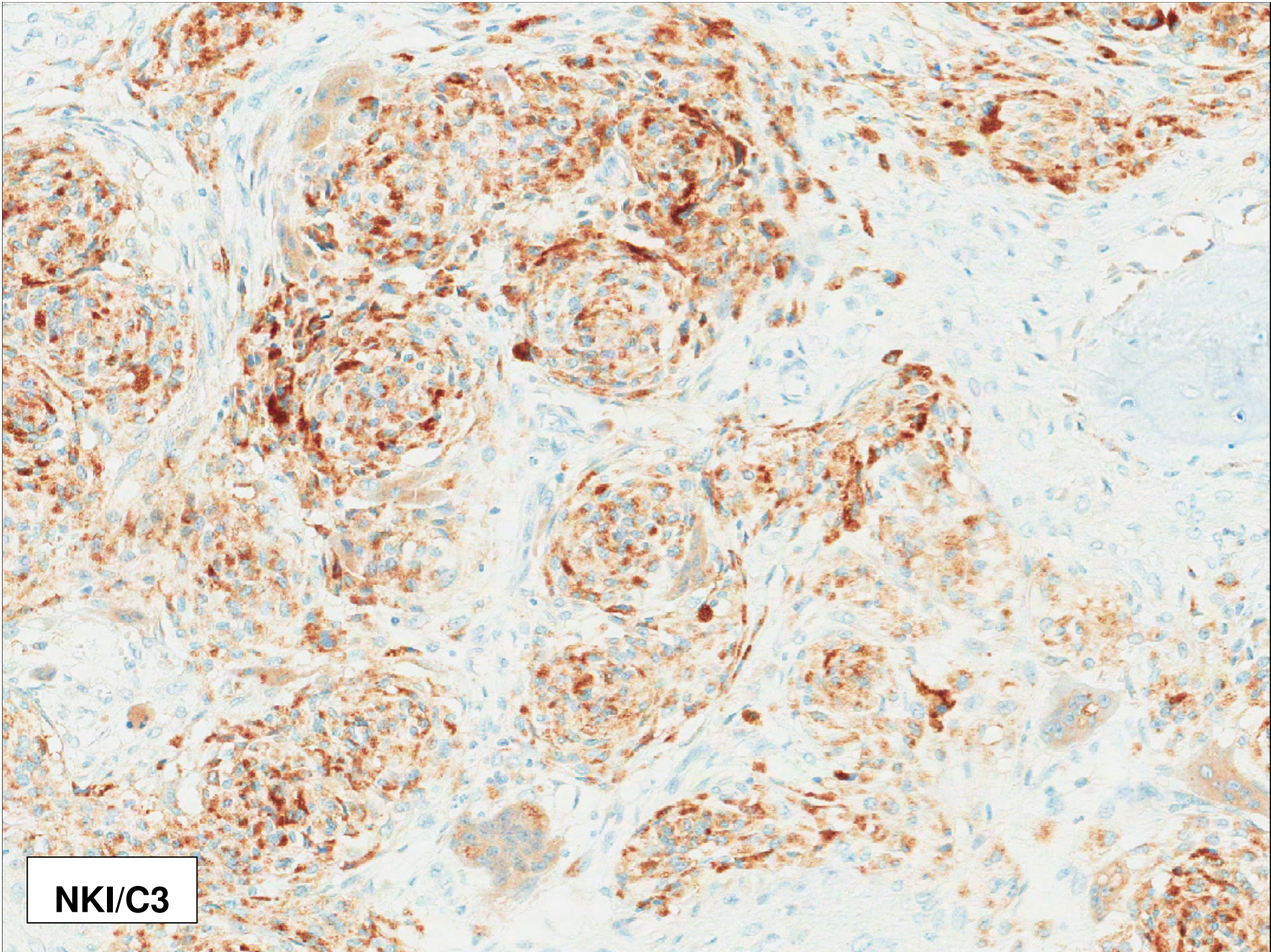


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Cancer Center

Imuno-histoquímica



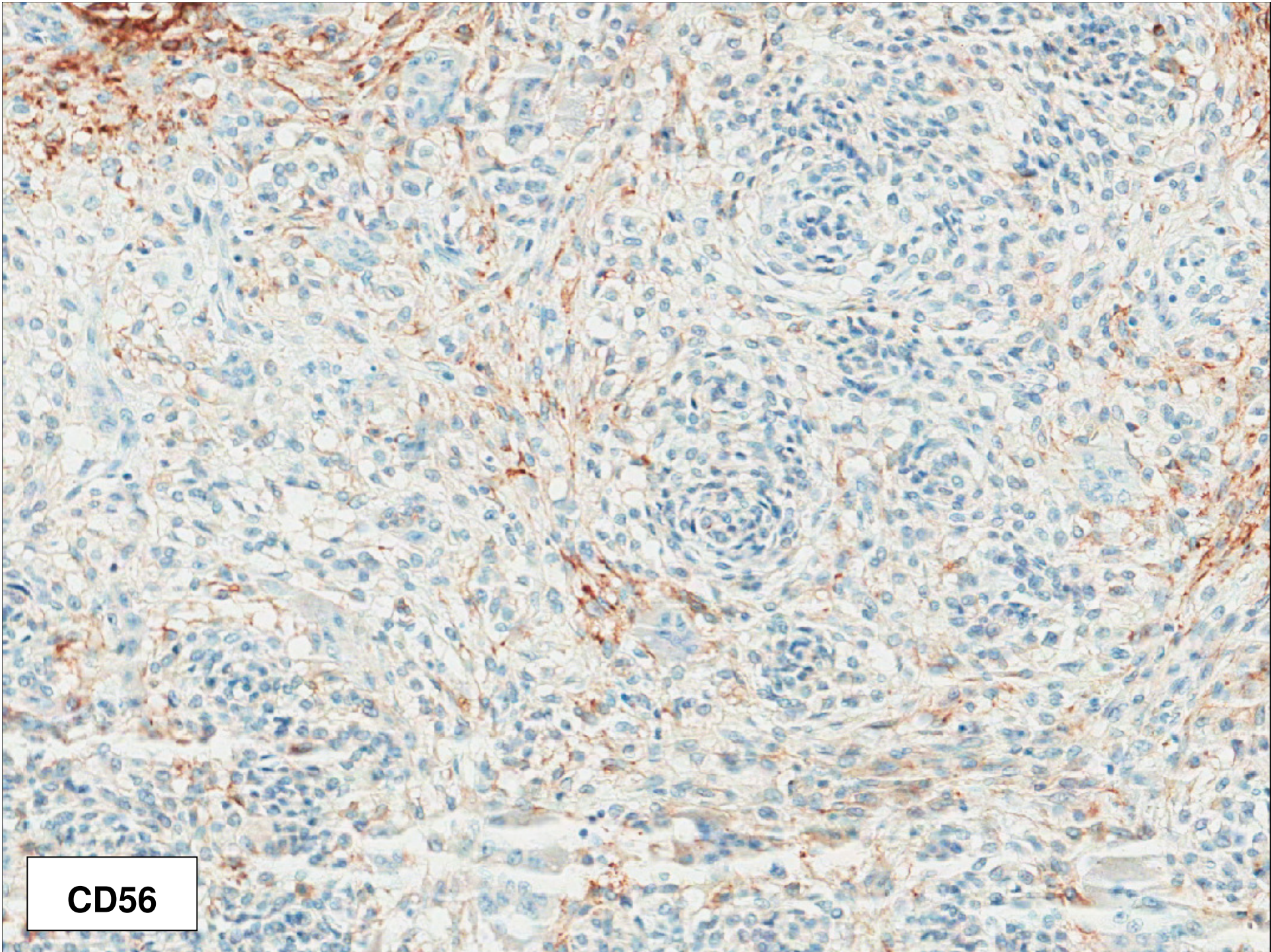
NSE



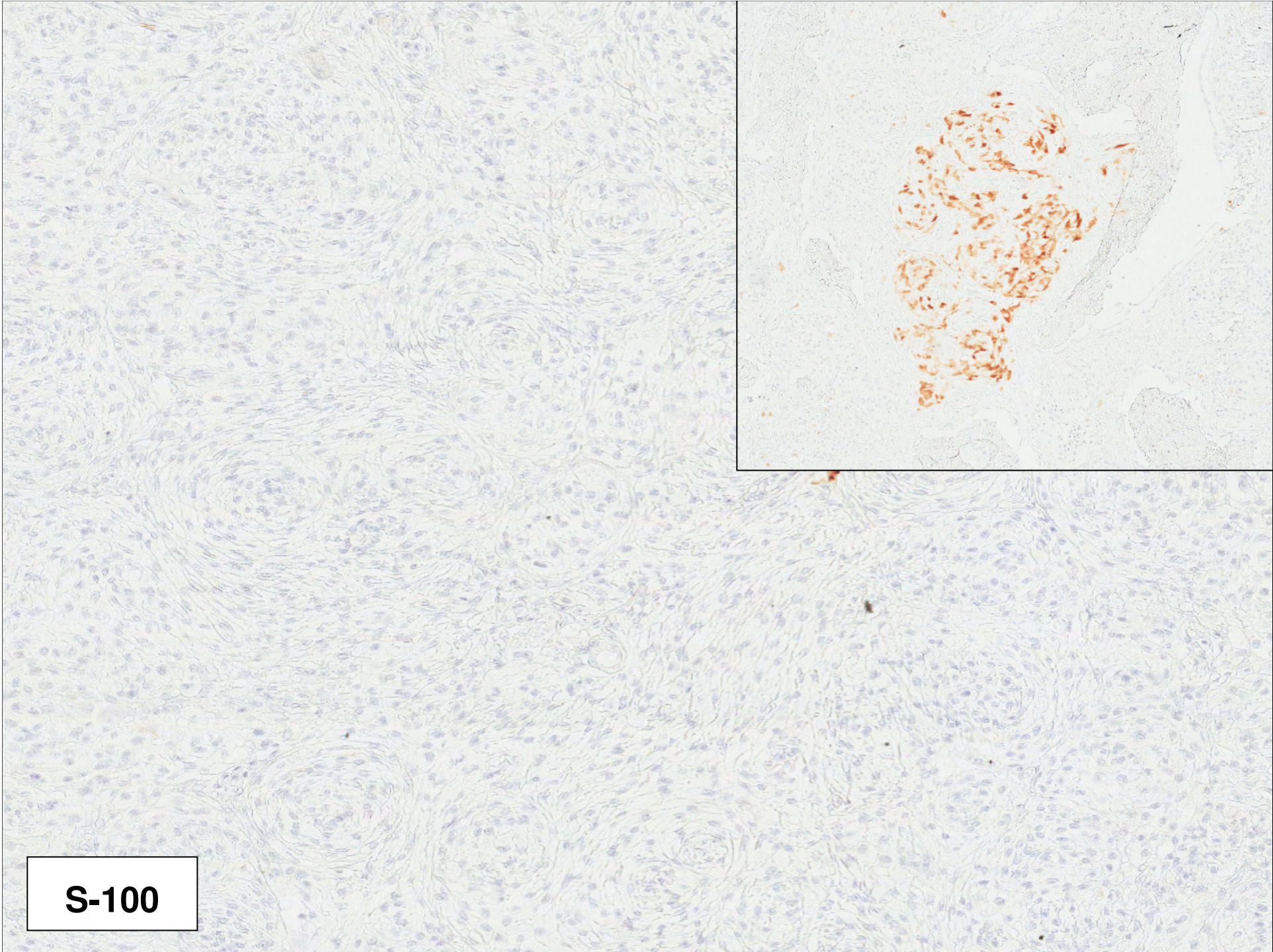
NKI/C3



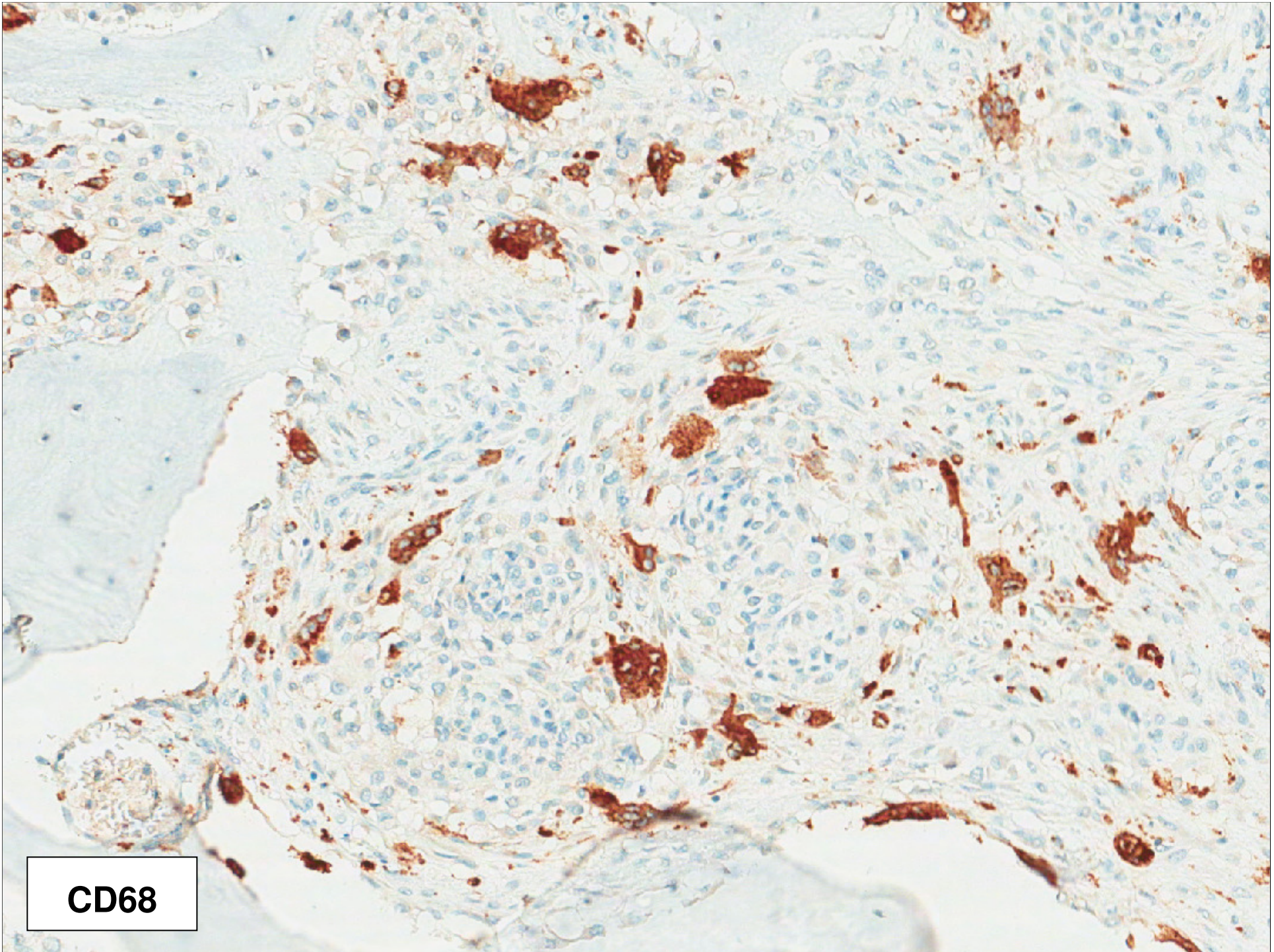
P63



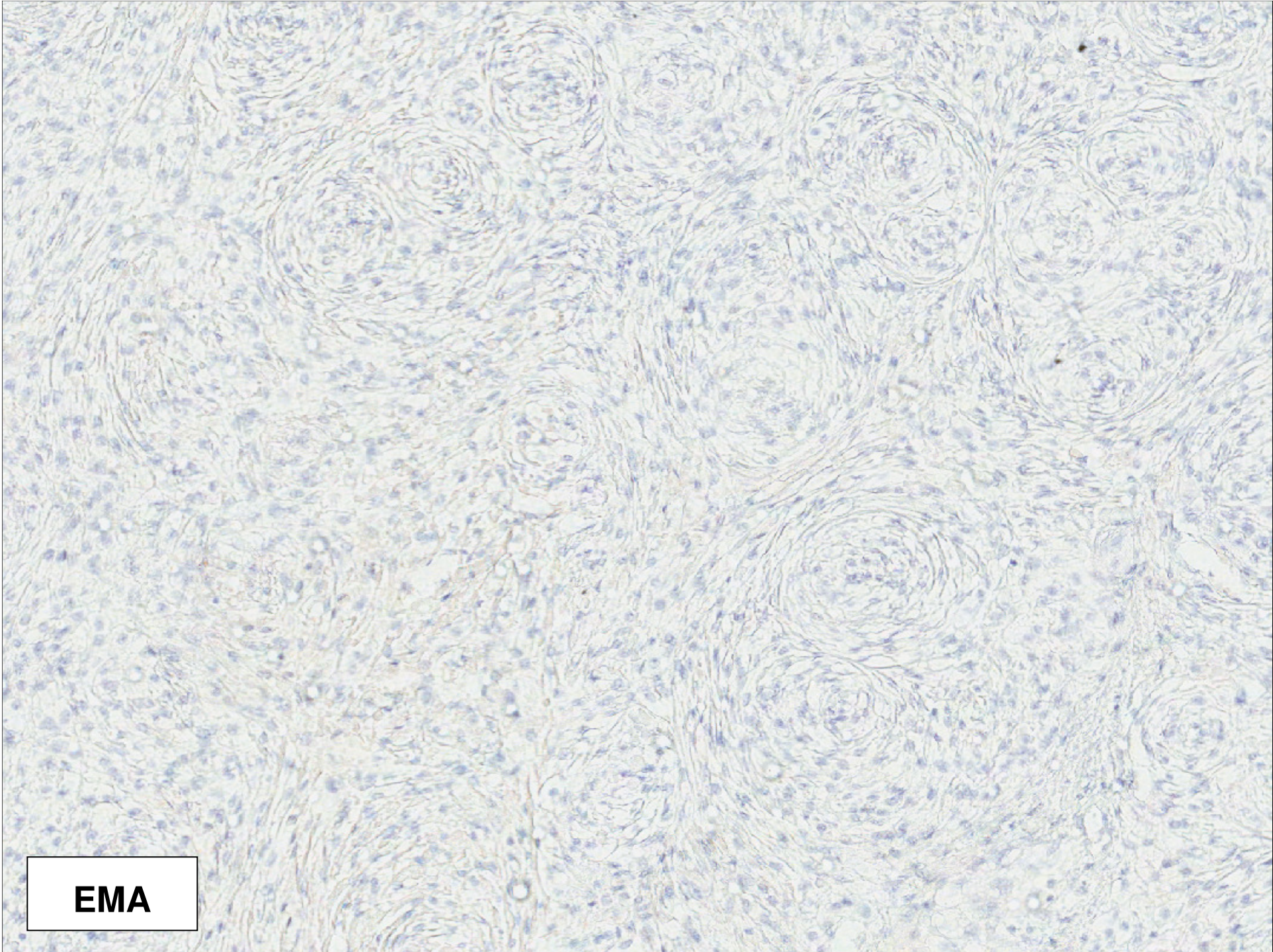
CD56



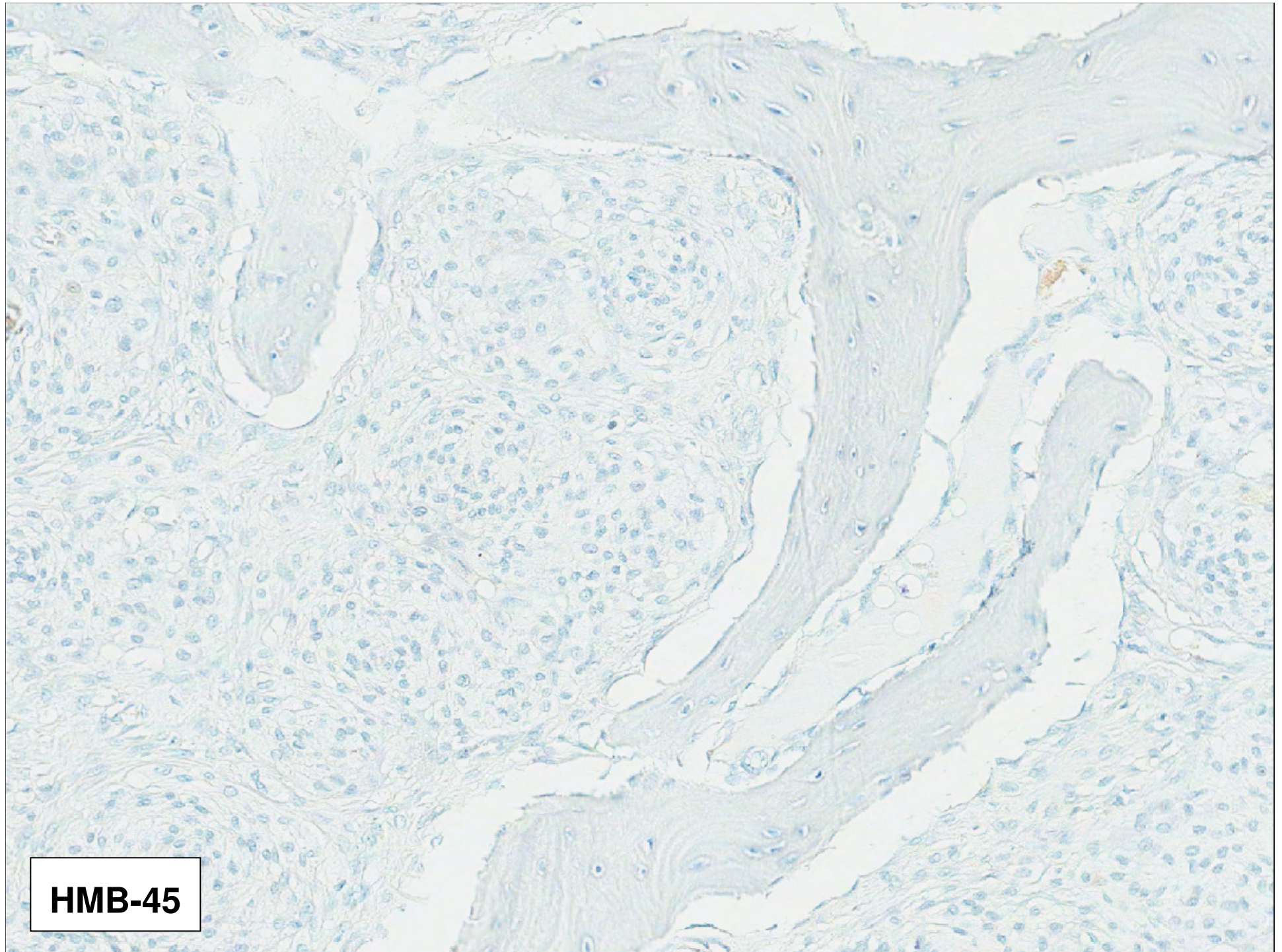
S-100



CD68



EMA



HMB-45





Diagnóstico final

“Neurotequeoma celular”



Neurotequeoma celular

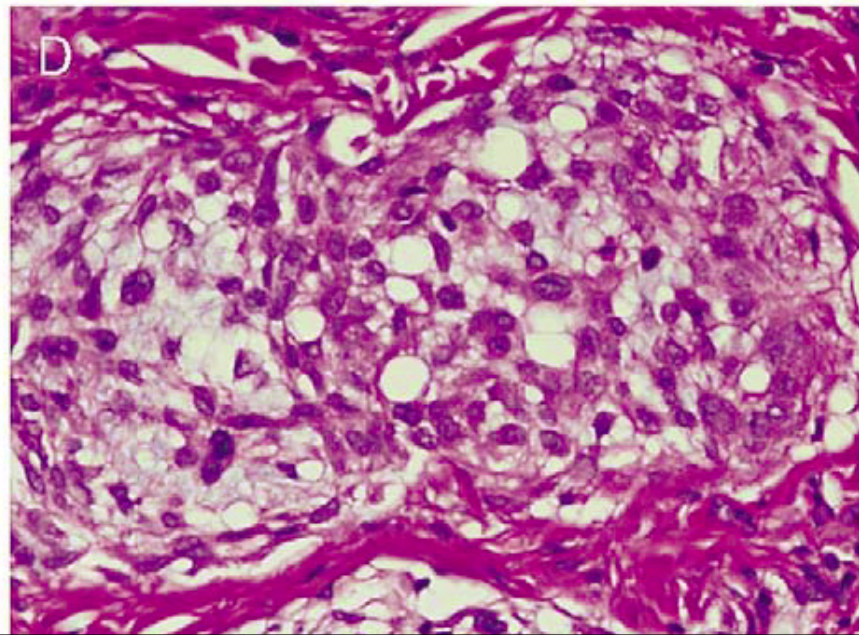
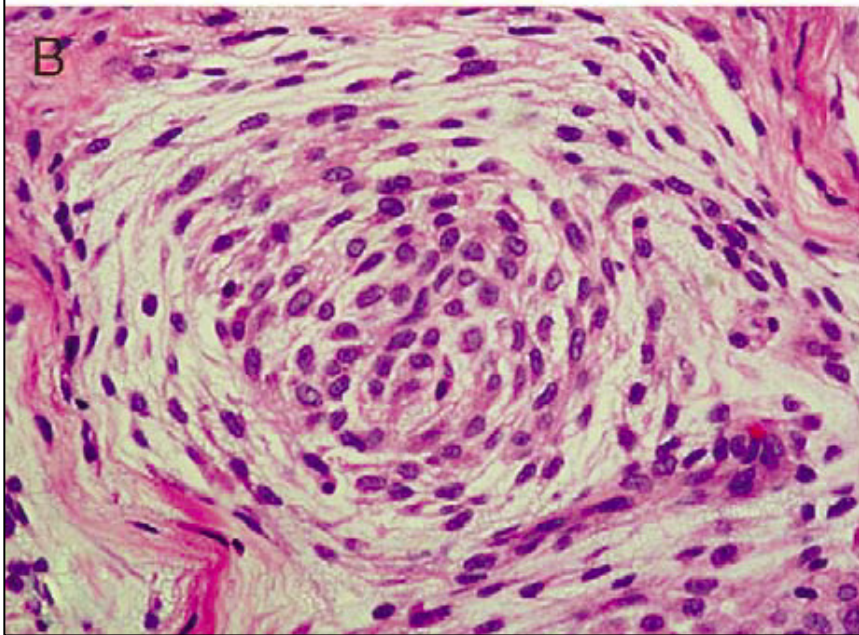
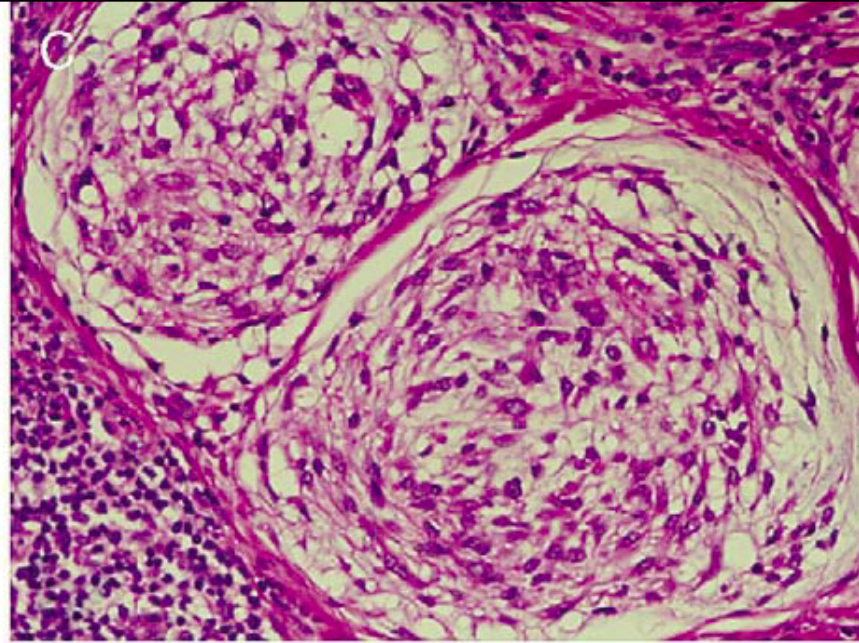
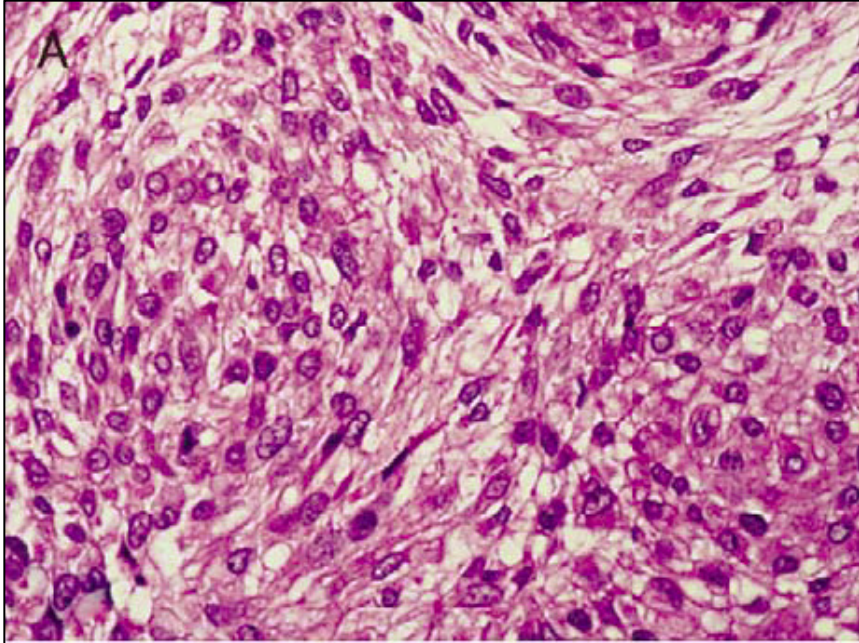
- ❑ “**Neurotequeoma**” (*gr. theke, sheath*): **1980** Gallager e Helwig
 - 53 casos
 - Origem na bainha neural
 - > > “**Mixoma da bainha neural**”: descrito em **1969** por Harkin e Reed → “**Mixoma da bainha neural dérmico**”.

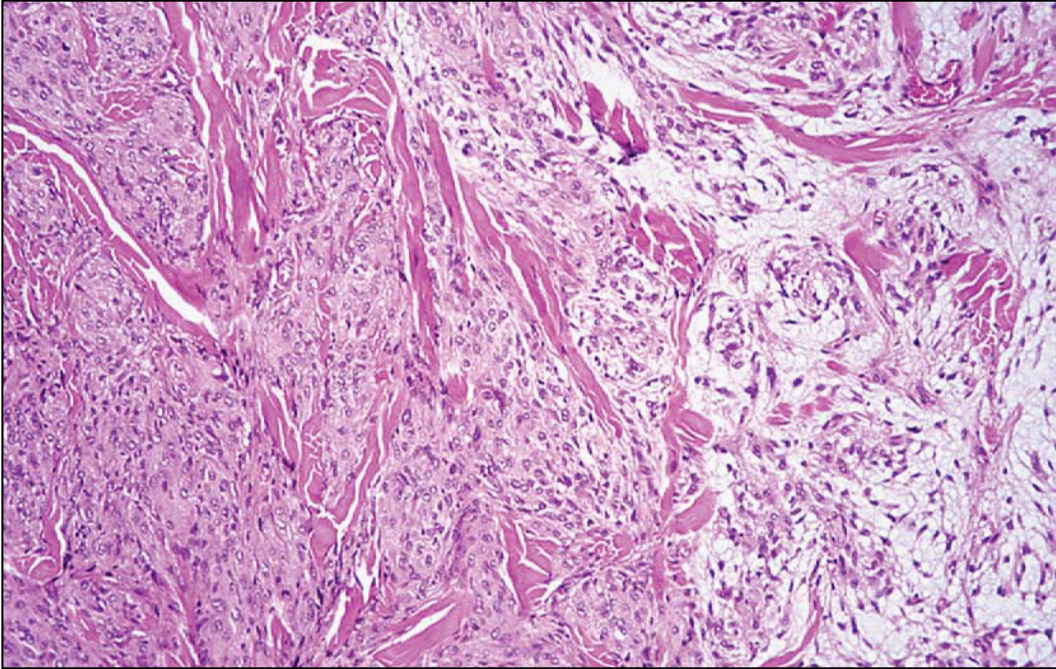
- ❑ “**Neurotequeoma celular**”:
 - Termo dado por 1^a vez em **1986** por Rosati *et al.*
 - variantes: celular e mixoide

Neurotequeoma celular

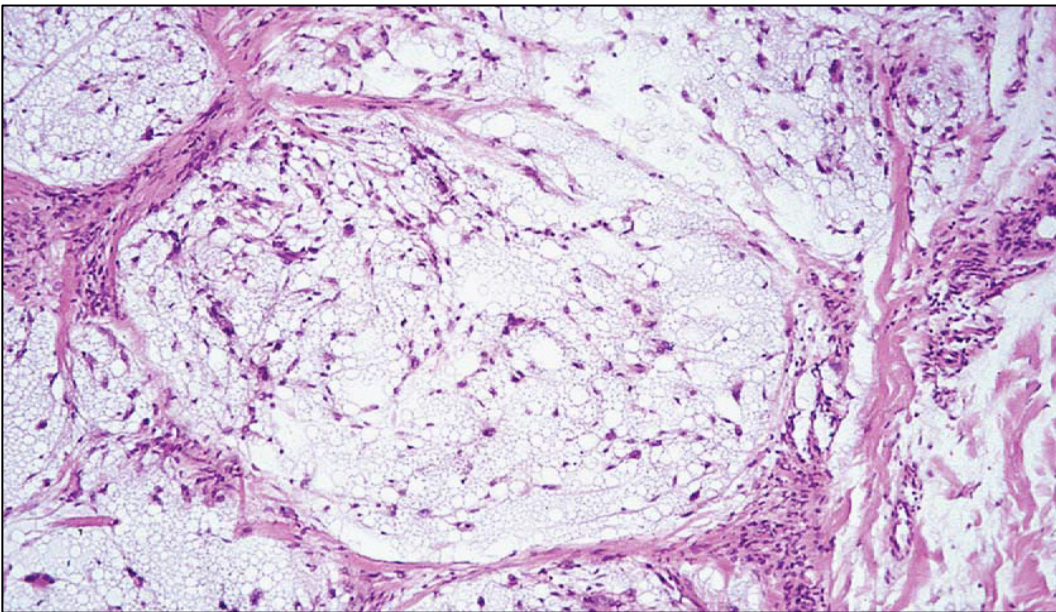
- Jovens, 1^a e 2^a décadas da vida
- Mulheres
- Cabeça e pescoço, extremidade superior e ombro.
- Derme, tecido subcutâneo (50%) e músc. esquelético (raro)
- Tamanho $\leq 2,0$ cm (90%)
- Recidivas: raras (margens +)
- Metástases: não reportados
- **Variantes:**



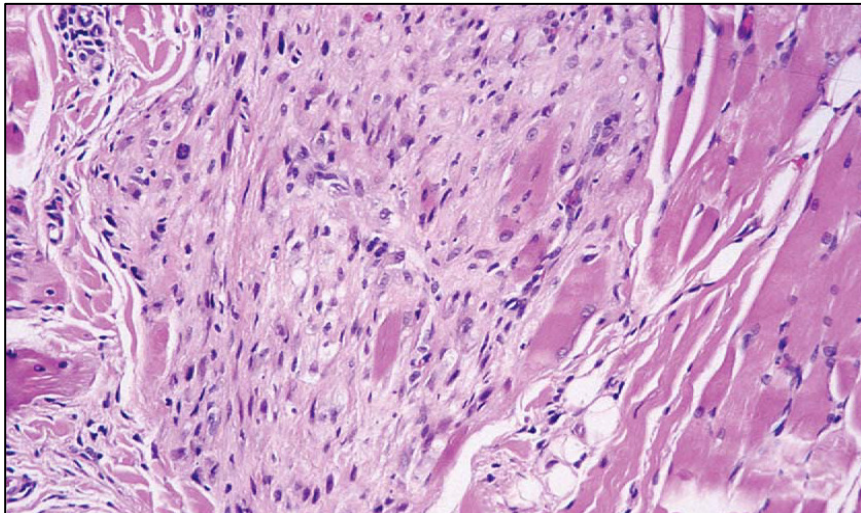
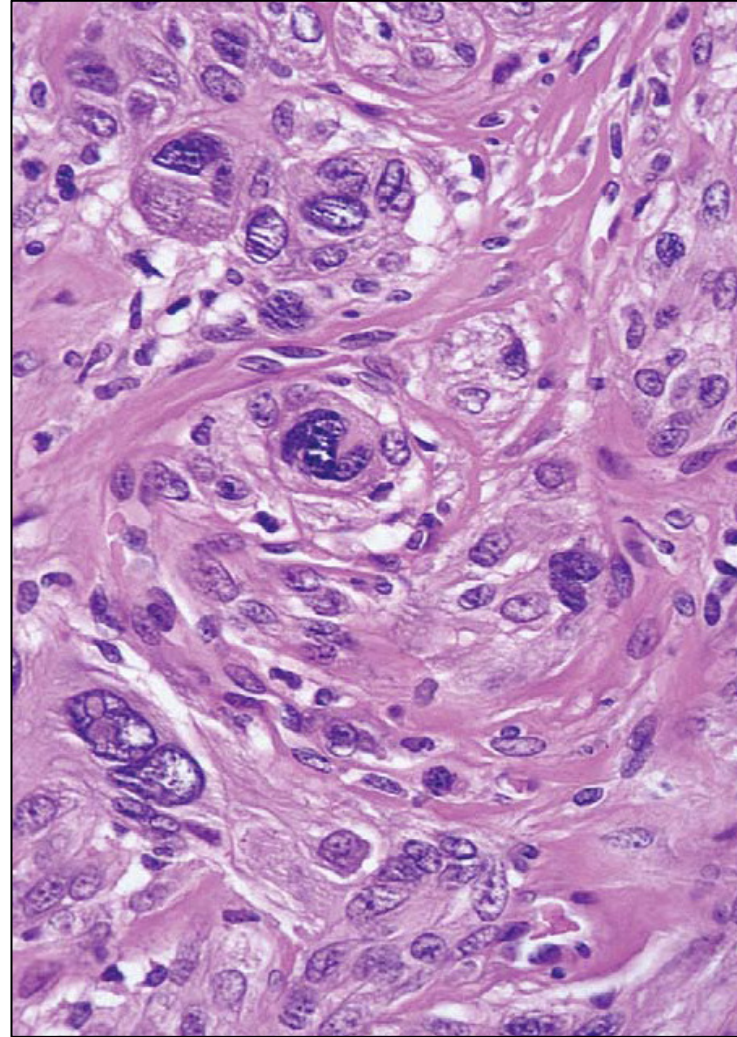
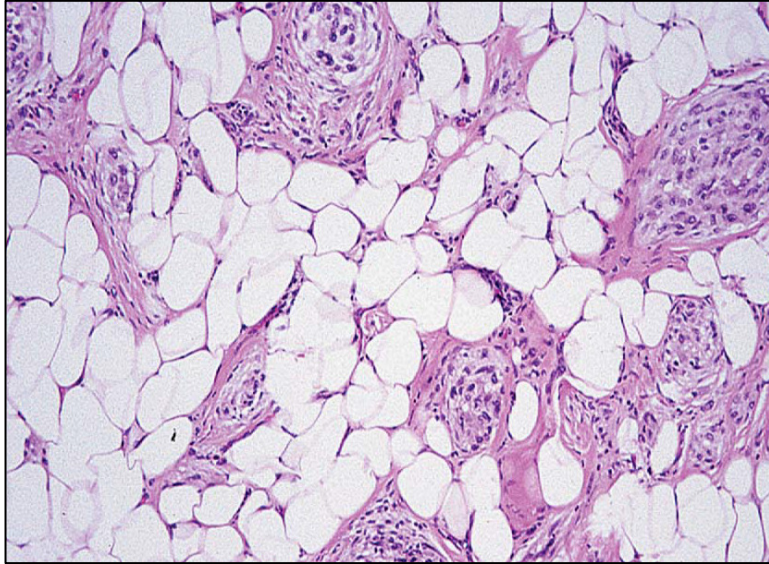




Nearly 30% of cellular NTK have focally myxoid stroma

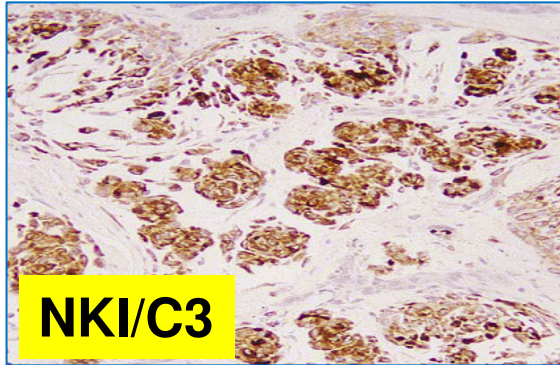


A predominantly myxoid CNTK mimicking DNSM

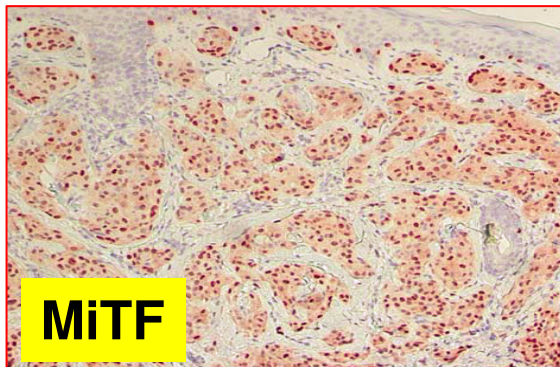




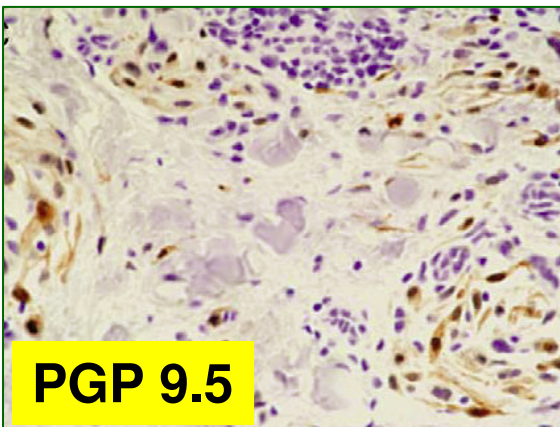
Neurotequeoma celular. IHQ



- **NKI/C3 (CD63)**; melanossomos e lisossomos.
- Neopl melanocíticas, xantogranuloma juvenil, fibroxantoma atípico, histiocitoma fibroso celular, reticulohistiocitoma, xantoma, tumor de células granulares.



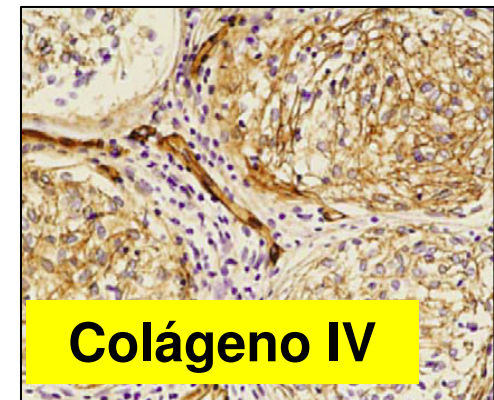
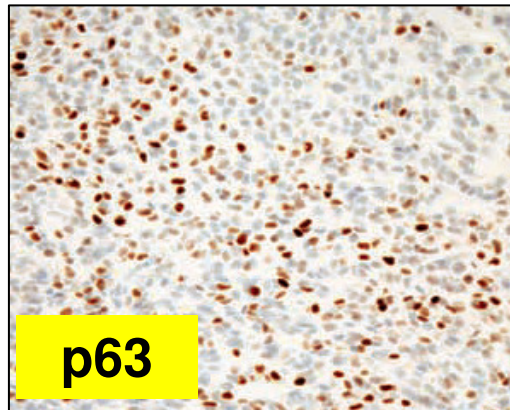
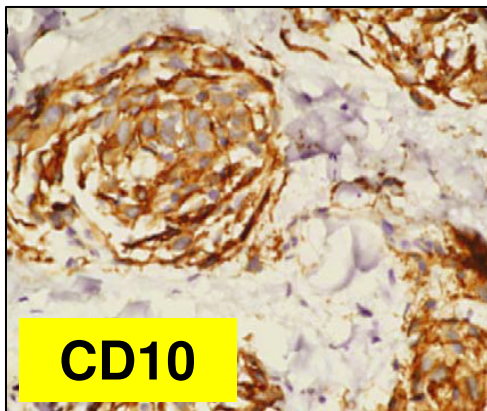
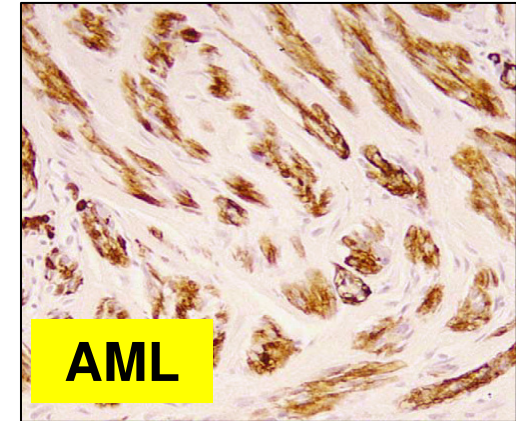
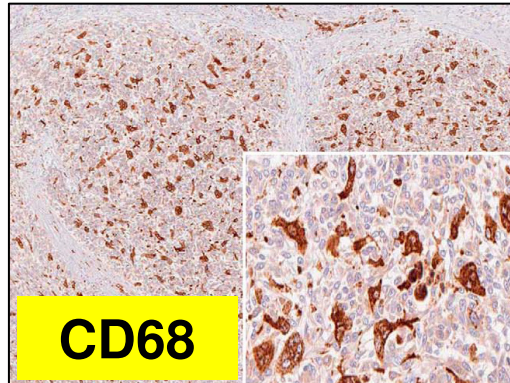
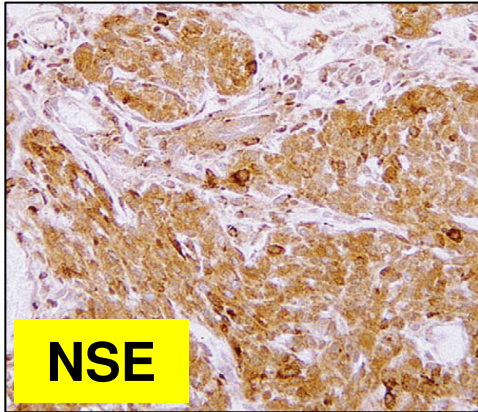
- **MiTF (Fator de transcrição de microftalmia)**
- Neoplasias melanocíticas, PEComas



- **PGP 9.5 (Protein gene product 9.5)**
- Marcador neural, > em tecido cerebral, porem não específico



Neurotequeoma celular. IHQ





Neurotequeoma celular. IHQ

GFAP

NEGATIVO:

Melan
A

S100

EMA

HMB45

Des-
mina

CELLULAR NEUROTHEKEOMA OF THE MAXILLA

NOAM A. COHEN, MD, PhD
BRUCE R. PAWEL, MD

DANIEL S. SAMADI, MD
KEN KAZAHAYA, MD

PHILADELPHIA, PENNSYLVANIA

- Masculino
- 15 meses
- Tumor em maxila direita, 2 meses
- IHQ: vimentina+, NKI/C3+, S100-, HMB45-, Factor XIIIa-
- Tratamento: ressecção local
- Seguimento 2 anos: sem recorrência.

CELLULAR NEUROTHEKEOMA OF THE MAXILLA

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Fig 1. Axial computed tomographic images demonstrate well-defined, locally expansile premaxillary mass. A) Bone window. B) Soft tissue window.

CELLULAR NEUROTHEKEOMA OF THE MAXILLA

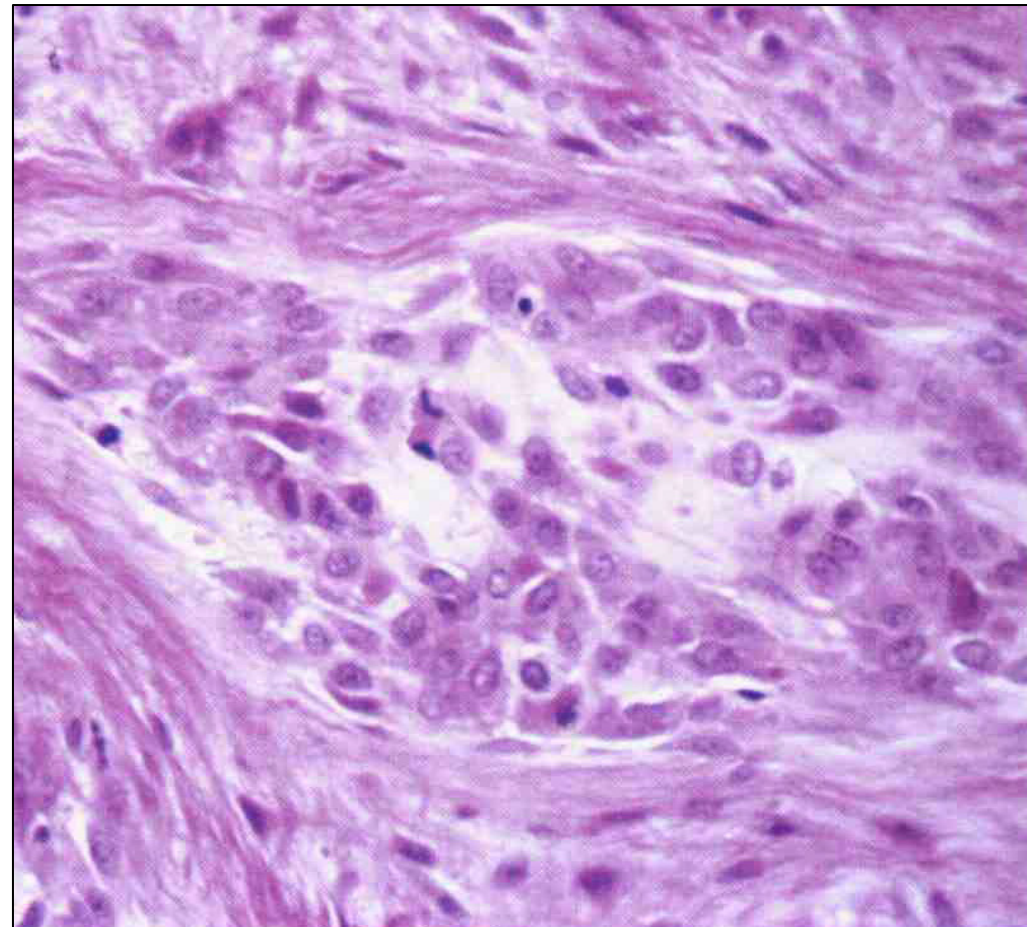
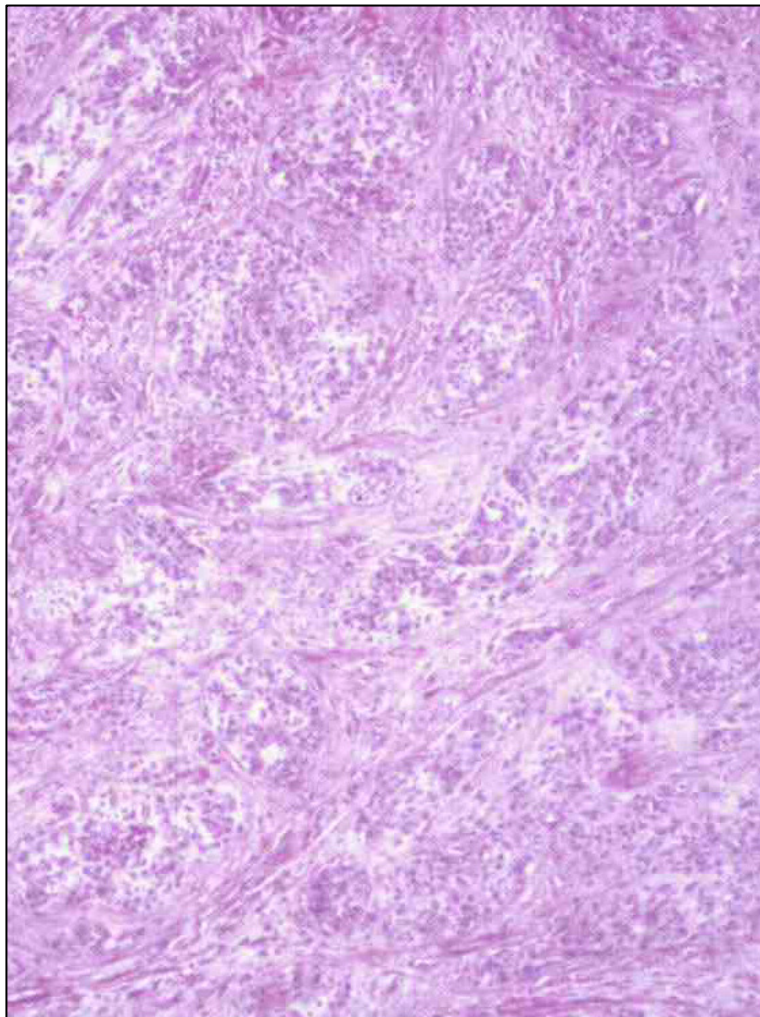
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NTK. Diagnóstico diferencial

- Mixoma da bainha neural dérmico
- Angiomixoma superficial.
- Mixofibrossarcoma



- Neoplasias melanocíticas
- Leiomioma pilar
- Tumores dérmicos fibrohistiocíticos



Cellular Neurothekeoma

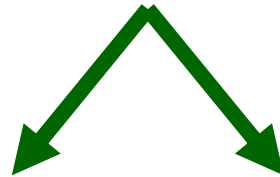
A Distinctive Variant of Neurothekeoma Mimicking Nevomelanocytic Tumors

Raymond L. Barnhill, M.D., and Martin C. Mihm, Jr., M.D.

The American Journal of Surgical Pathology 14(2): 113-120, 1990

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- Células epitelioides em ninhos, fascículos ou enovelados
- Abundante citoplasma eosinofílico
- Atipias ocasionais



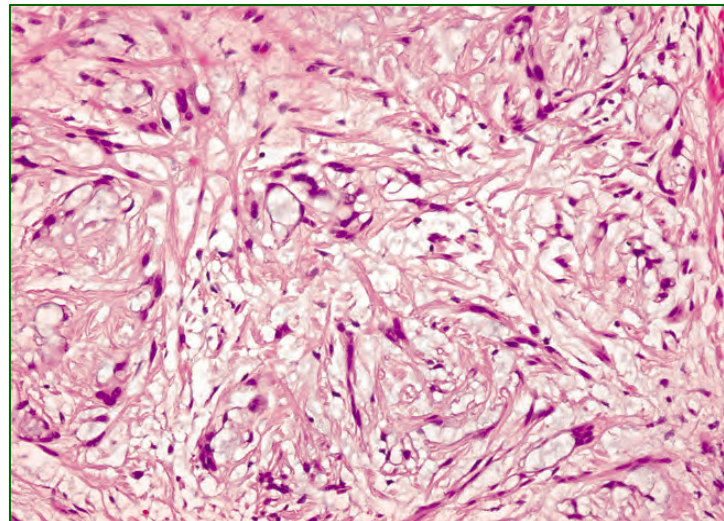
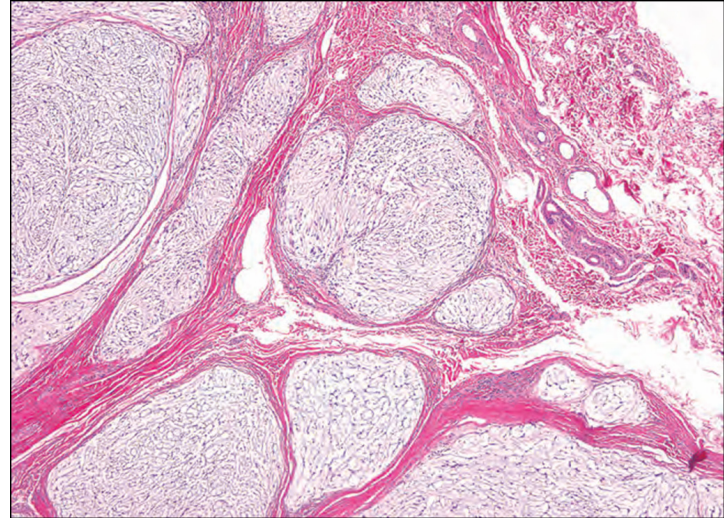
- Nevo de Spitz
- Melanoma
- Nevo azul celular

- Fibrohistiocitoma epitelióide
- Xantogranuloma juvenil.



Mixoma da bainha neural (dérmico)

- Superficial
- Multinodular
- Bem delimitado
- Extremidade distal
- Adulto joven-idade média
- Abundante estroma mixoide
- Células epitelioides pequenas, estreladas, em anel e fusiformes
- S100+



Neurothekeoma and Plexiform Fibrohistiocytic Tumor

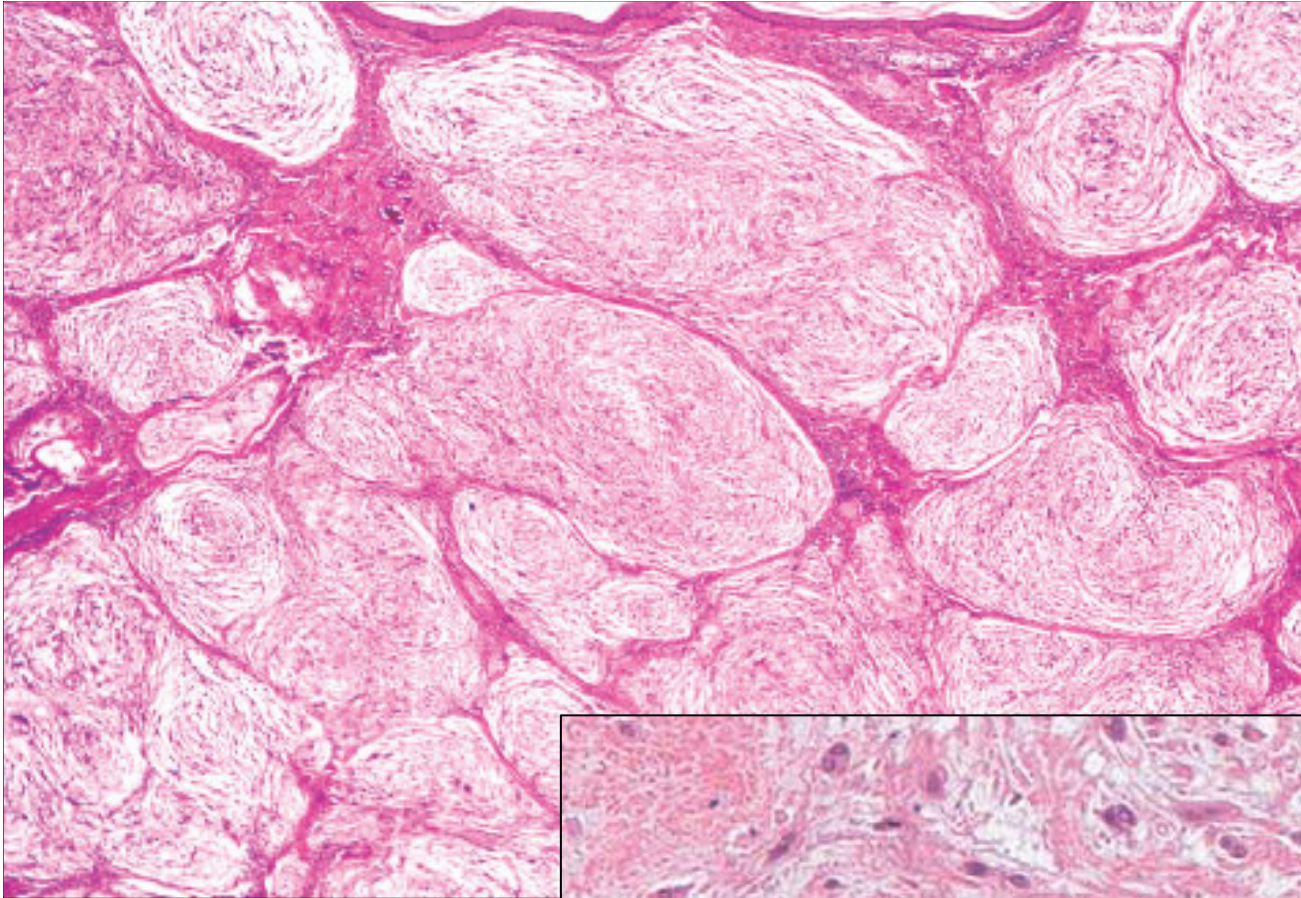
Mere Histologic Resemblance or Histogenetic Relationship?

Shabnam Jaffer, MD, Andrea Ambrosini-Spaltro, MD,† Antonio M. Mancini, MD,†
Vincenzo Eusebi, MD, FRCPath,† and Juan Rosai, MD‡*

□ Neurotequeoma (NTK): 43 casos; *estroma mixoide*

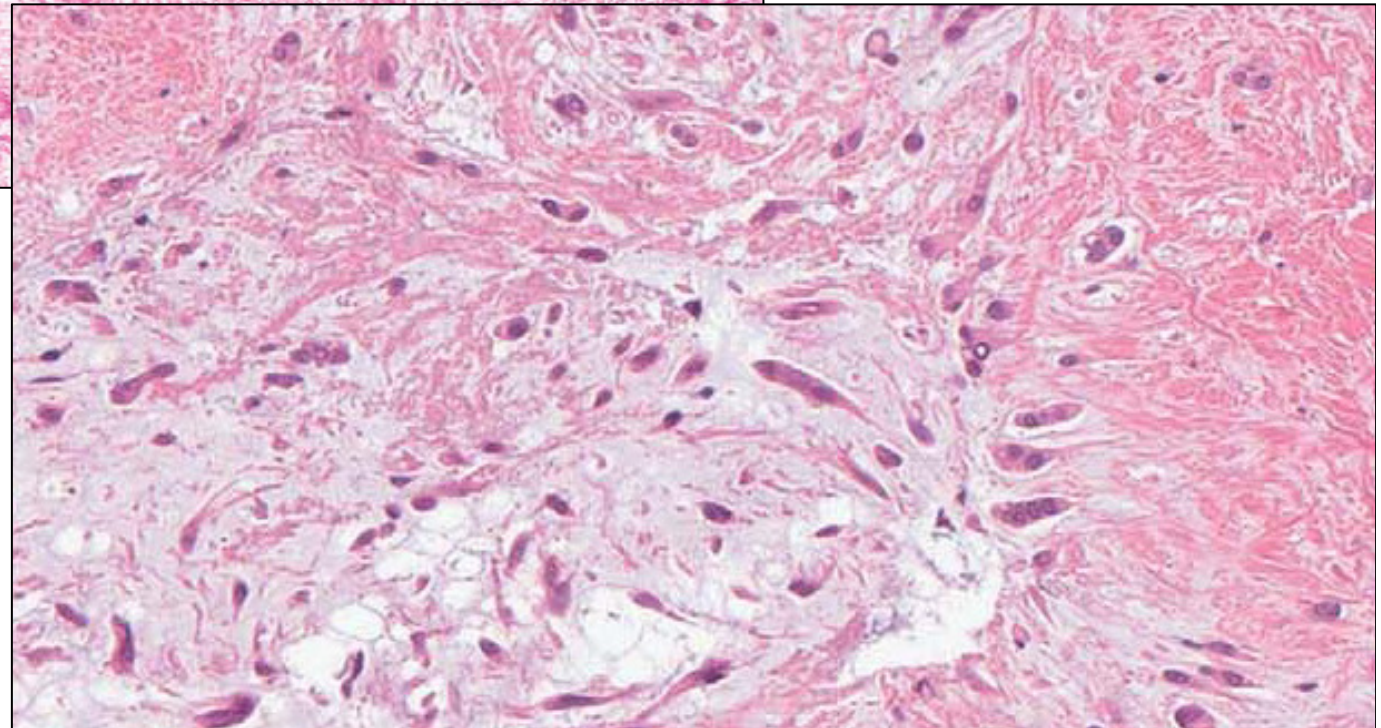
- Mixoide (MyNTK-100%): 8
- Misto (MiNTK- > 10% < 100%): 15
- Celular (CNTK- < 10%): 20

□ Tumor fibrohistiocítico plexiforme (PFHT): 18



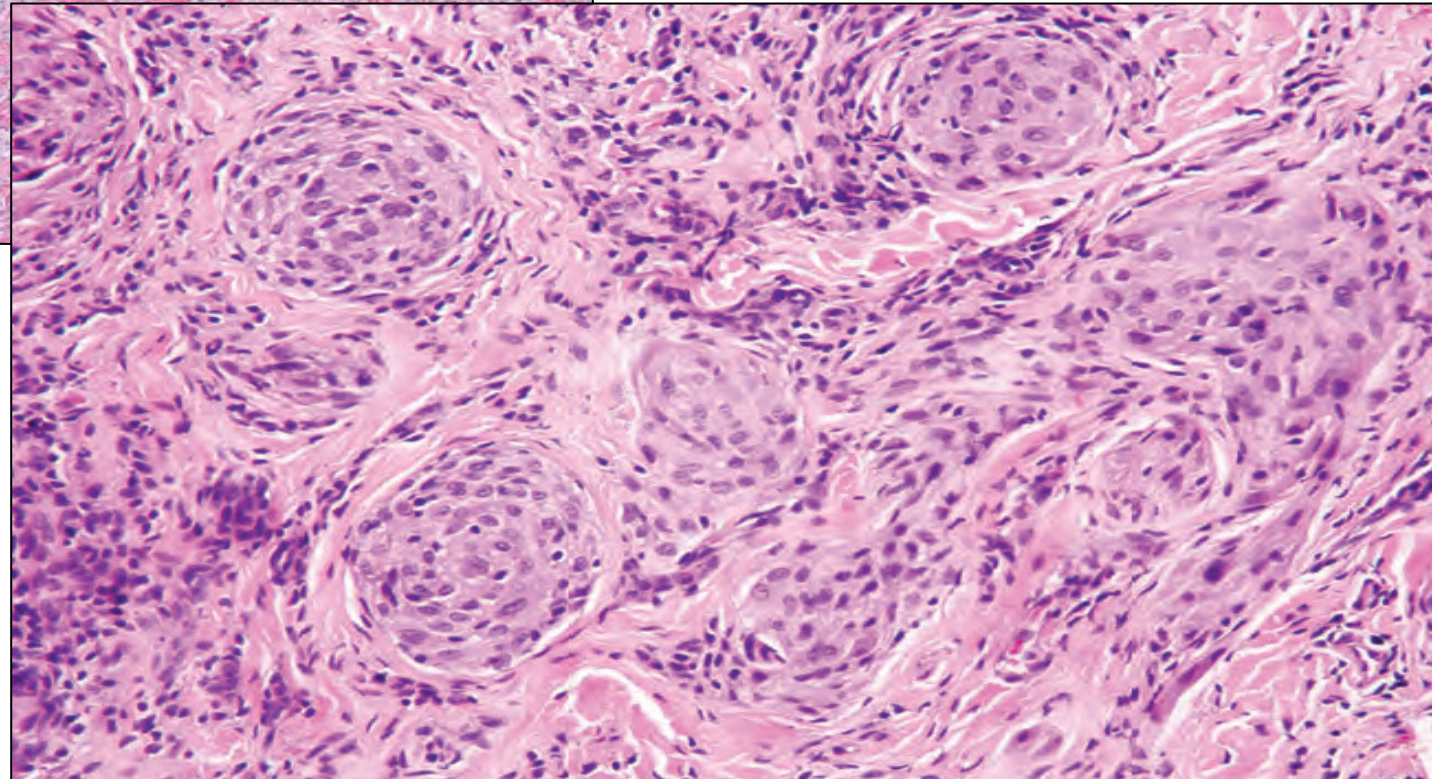
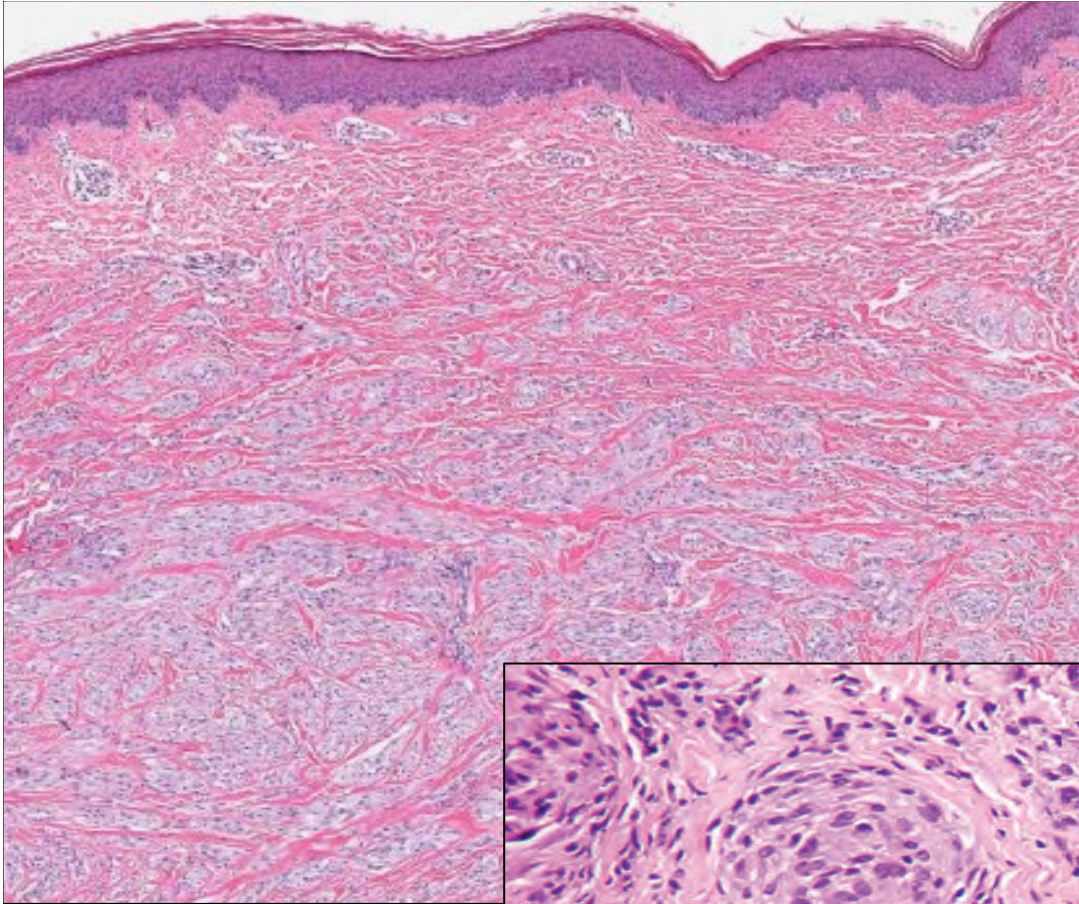
Neurotequeoma ***“mixoide”***

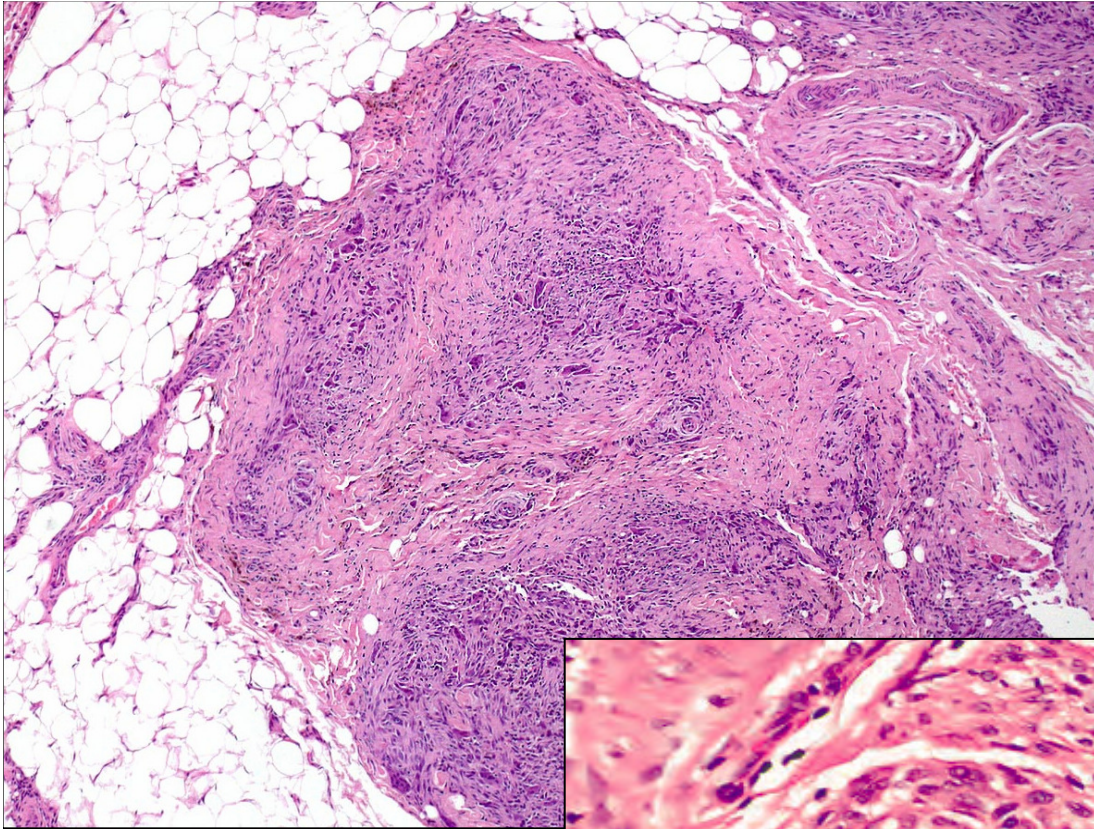
- Multinodular
- Bem circunscritos
- Matriz mixoide difusa
- Células fusiformes e estreladas
- S100+



Neurotequeoma “celular”

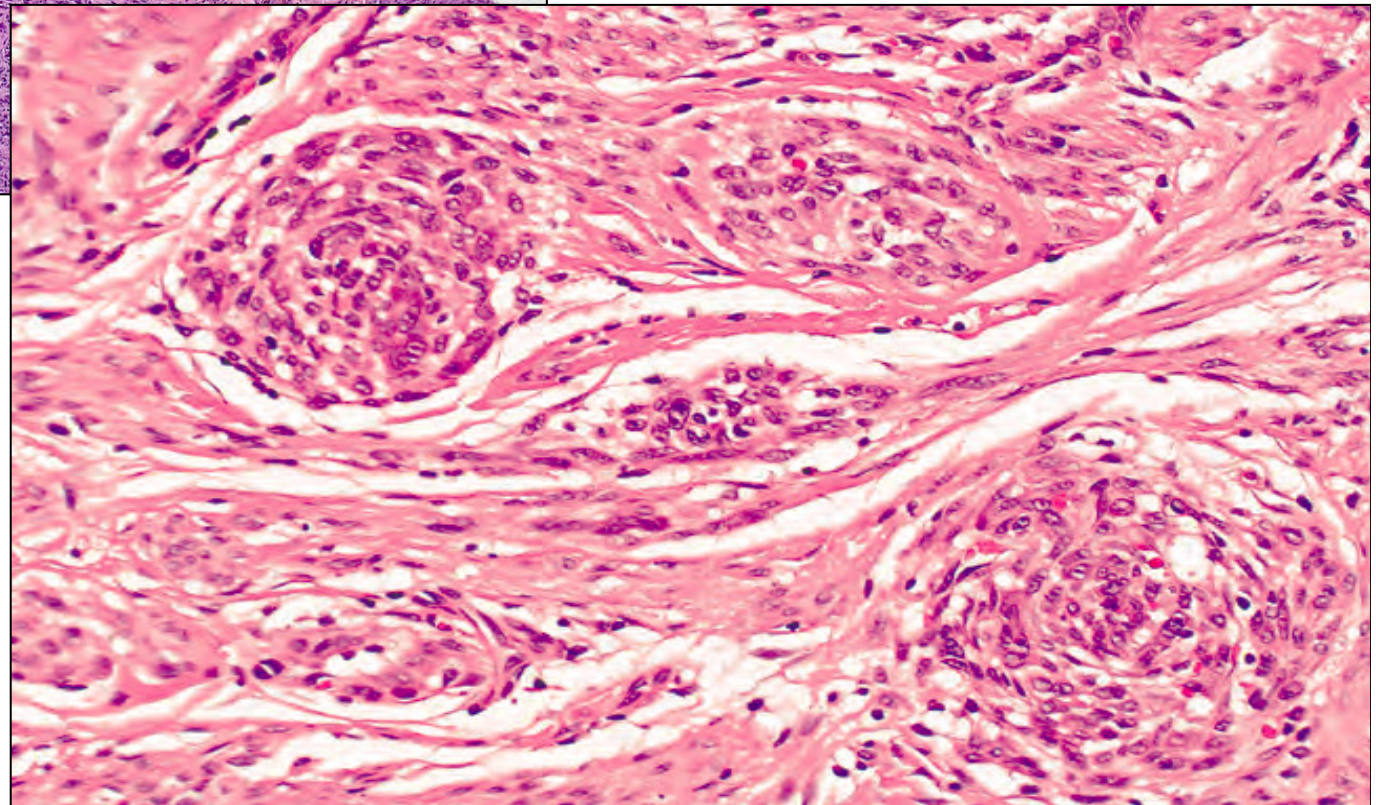
- Multinodular, plexiforme ou difusa
- Infiltrativos
- Matriz mixoide (<10%)
- Células fusiformes e/ou epitelioides, Células gigantes tipo osteoclastos
- S100 -





Tumor fibrohistiocítico plexiforme

- Plexiforme, Infiltrativos
- Matriz mixoide: ausente
- Células epitelioides (histiocitárias), fusiformes (miofibrobásticas) e gigantes multinucleadas tipo osteoclastos



Neurothekeoma and Plexiform Fibrohistiocytic Tumor

Mere Histologic Resemblance or Histogenetic Relationship?

Shabnam Jaffer, MD, Andrea Ambrosini-Spaltro, MD,† Antonio M. Mancini, MD,†
Vincenzo Eusebi, MD, FRCPath,† and Juan Rosai, MD‡*

	Vim	S100	CD68	NSE	CD56
MyNTK	7/7	8/8	0/8	6/6	5/6
MiNTK	11/11	4/15	6/15	7/10	8/10
CNTK	14/15	1/19	9/17	14/14	8/11
PFHT	13/14	0/18	6/11	9/10	8/11

“Existe notável sobreposição nas características histológicas e imuno-histoquímicas entre CNTK e PFHT”



Neurotequeoma celular

Tumor fibrohistiocítico plexiforme

- Crianças e adultos jovens
- Cabeça e pescoço
- Tumores dérmicos infiltrativos que se estendem para o subcutâneo
- Variável padrão de crescimento: nodular, plexiforme e difuso
- Células epitelioides e fusiformes arredondadas
- Células gigantes tipo osteoclastos
- Perfil imunohistoquímico: PGP9.5, NKI/C3, CD68, CD10, CD56

exceto



MiTF

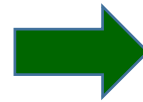
Neurothekeoma: An Analysis of 178 Tumors With Detailed Immunohistochemical Data and Long-term Patient Follow-up Information

John F. Fetsch, MD, William B. Laskin, MD,† James R. Hallman, MD,‡
George P. Lupton, MD,‡ and Markku Miettinen, MD**

• 178 casos

Quantidade de estroma mixoide:

- Celular ($\leq 10\%$) : 63
- Mixoide ($> 50\%$): 48
- Mixto ($> 10\% < 50\%$): 67



Vimentina
NKI/C3
CD10
MITF
NSE
PGP9.5
CD68
AML



S100
GFAP
Melan A

Neurothekeoma: An Analysis of 178 Tumors With Detailed Immunohistochemical Data and Long-term Patient Follow-up Information

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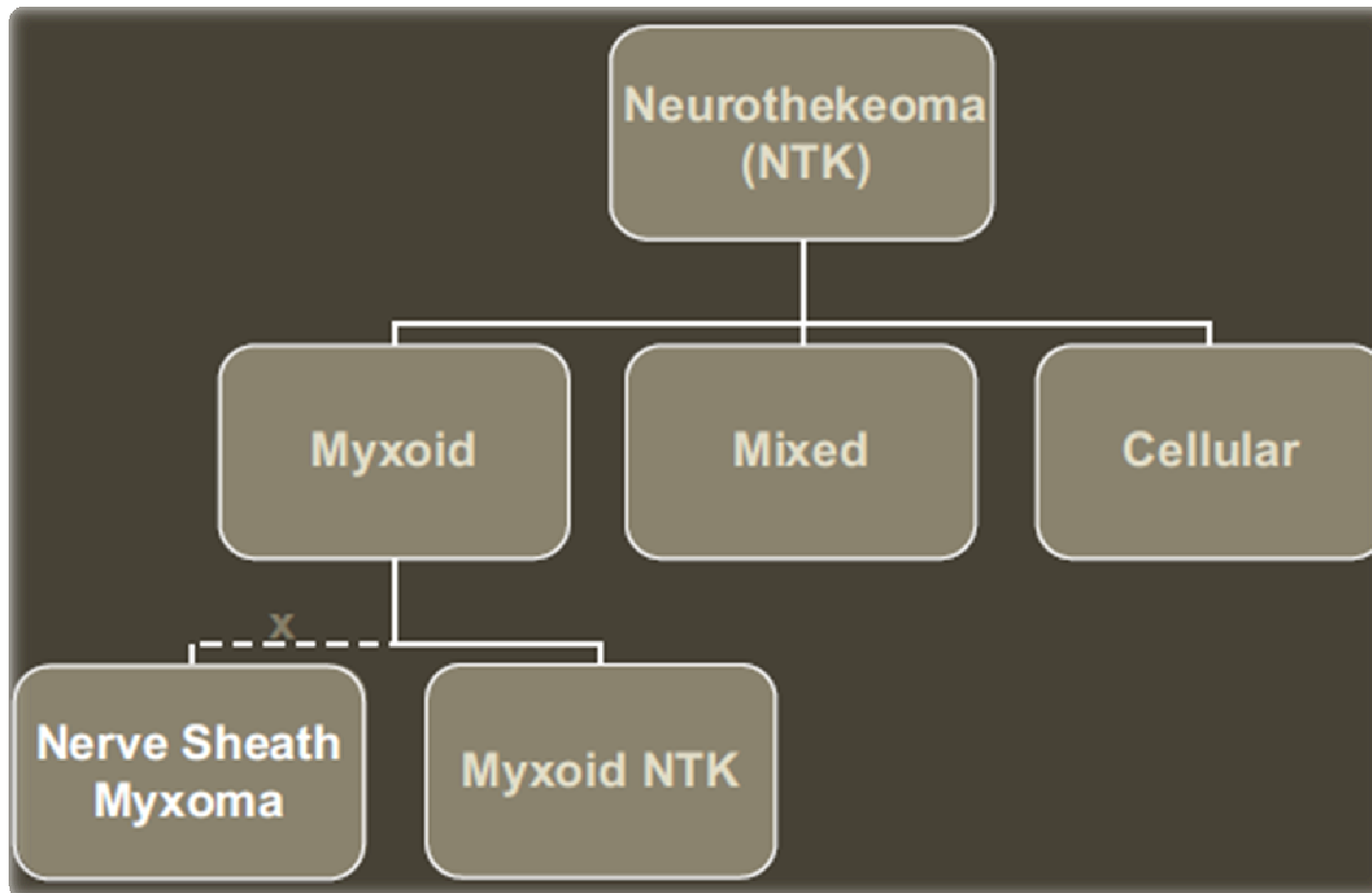
*Porem as diferenças na matriz, todos os tumores
compartilhaban certas características:*



- Células epitelioides e fusiformes com citoplasma eosinofílico relativamente abundante
- Multinodular, com padrão de crescimento em enovelados ou fascicular
- Colágeno esclerótico
- Células gigantes multinucleadas osteoclasto-símile
- Imunohistoquímica similar

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Neurotequeoma celular

- ❑ Linhagem de diferenciação **incerta**

- ❑ **Origens postuladas:**
 - Neural: Morfologia e imunohistoquímica **≠** Mixoma da bainha neural (verdadeiro origem neural)
 - Neuroectodérmico: PGP9.5, MiTF, NKI/C3
 - Família de PEComas (“miomelanocítico”): MiTF, AML
 - Variante epitelióide do leiomioma pilar

- ❑ Atualmente sugere-se origem em células **fibroblásticas** com capacidade de diferenciação para **miofibroblastos** e tendência a recrutar células histiocitárias

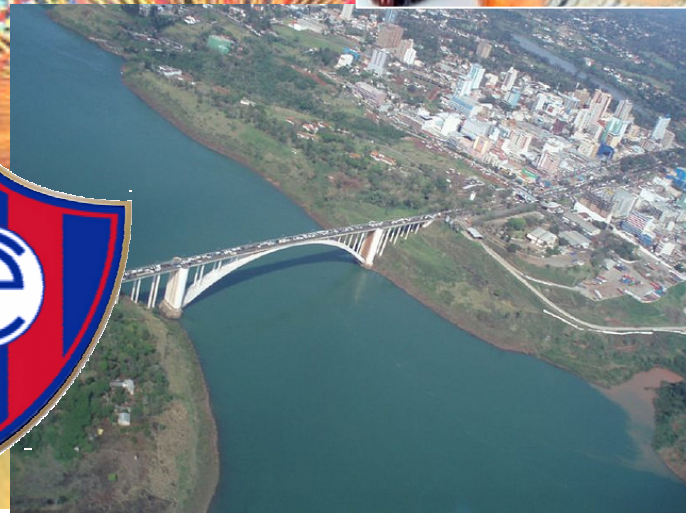


“Espectro Fibrohistiocítico”



- Fletsch, JF, Laskin WB. Neurothekeoma: An analysis of 178 tumors with detailed immunohistochemical data and long-term patient follow-up information. Am j Sur Pathol. July 2007; 33 (6).
- Hornick JL, Fletcher CD. [Cellular neurothekeoma: detailed characterization in a series of 133 cases.](#) Am J Surg Pathol. 2007 Mar;31(3):329-40.
- Cohen NA, Samadi DS, Pawel BR, Kazahaya K. [Cellular neurothekeoma of the maxilla.](#) Ann Otol Rhinol Laryngol. 2004 May;113(5):384-7.
- Barnhill RL, Mihm MC Jr. [Cellular neurothekeoma. A distinctive variant of neurothekeoma mimicking nevomelanocytic tumors.](#) Am J Surg Pathol. 1990 Feb;14(2):113-20.
- Jaffer S, Ambrosini-Spaltro A, Mancini AM, Eusebi V, Rosai J. [Neurothekeoma and plexiform fibrohistiocytic tumor: mere histologic resemblance or histogenetic relationship?](#) Am J Surg Pathol. 2009 Jun;33(6):905-13. doi: 10.1097/PAS.0b013e31819c79f7.

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- Fox MD, Billings SD, Gleason BC, Moore J, Thomas AB, Shea CR, Victor TA, Cibull TL. [Expression of MiTF may be helpful in differentiating cellular neurothekeoma from plexiform fibrohistiocytic tumor \(histiocytoid predominant\) in a partial biopsy specimen.](#) Am J Dermatopathol. 2012 Apr;34(2):157-60. doi: 10.1097/DAD.0b013e3182286a03.
- Page RN, King R, Mihm MC Jr, Googe PB. [Microphthalmia transcription factor and NKI/C3 expression in cellular neurothekeoma.](#) Mod Pathol. 2004 Feb;17(2):230-4.
- Jo VY, Fletcher CD. [p63 immunohistochemical staining is limited in soft tissue tumors.](#) Am J Clin Pathol. 2011 Nov;136(5):762-6. doi: 10.1309/AJCPXNUC7JZSKWEU.
- Goldblum JR, Folpe AL, Weiss SW. Enzinger and Weiss's soft tissue tumors. Cellular neurothekeoma. 6th. Ed. Philadelphia: Elsevier Saunders; 2014. p359-363.
- Goldblum JR, Folpe AL, Weiss SW. Enzinger and Weiss's soft tissue tumors. Nerve sheath mixoma. 6th. Ed. Philadelphia: Elsevier Saunders; 2014. p845-848.





***Muchas gracias por
su atención!***



Lilian Elizabeth Giménez González