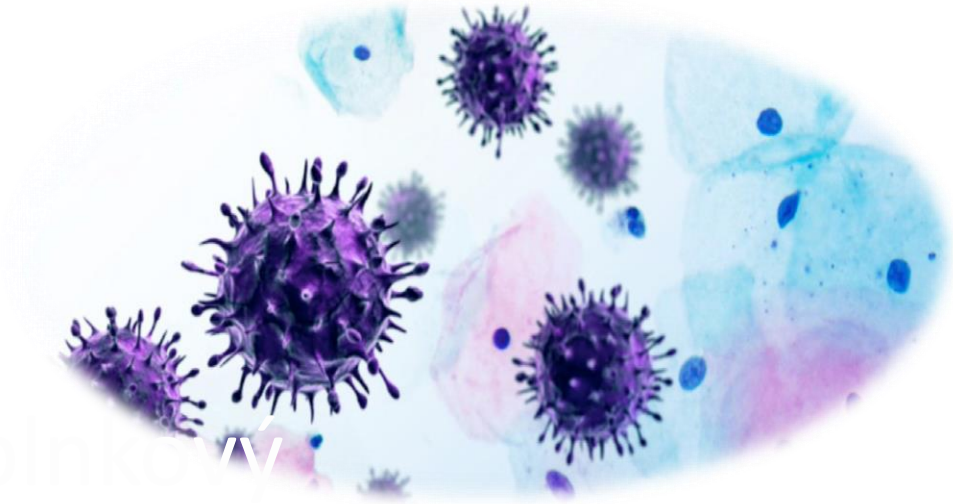


Doplnkový text

8. Východoslovenský BIOPTICKÝ SEMINÁR

27. a 28. apríla 2018, Košice

Golden Royal Boutique Hotel

Prípad SD-IAP 675

Peter Bohuš, Medicyt Košice

70-ročný pacient

Klinická diagnóza:

Exulcerovaný tumor pravej strany vlasatej časti hlavy.

Fotografia poskytnutá s láskavosťou dermatológa – MUDr. Rugoóvá

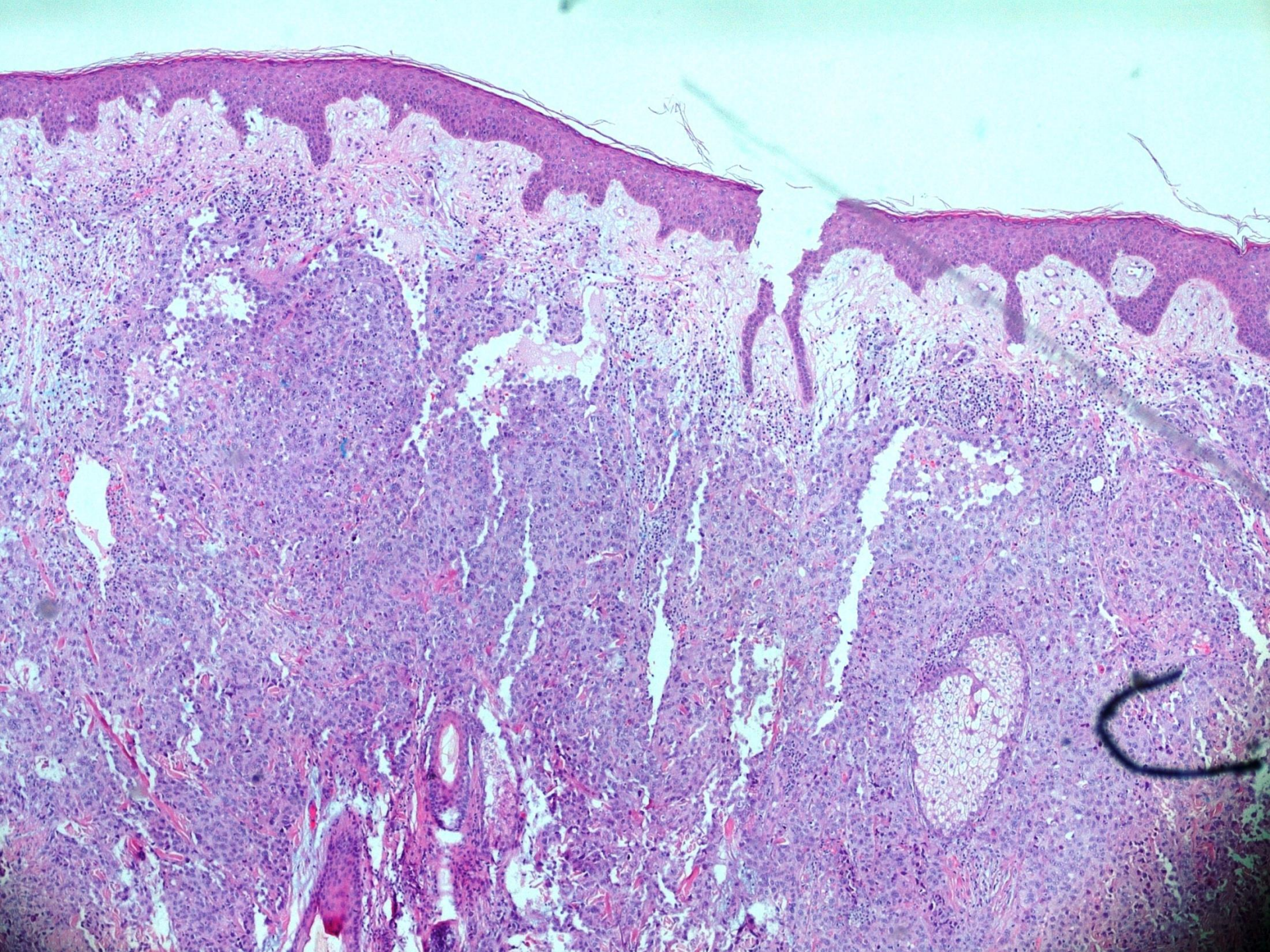
KLINIKA

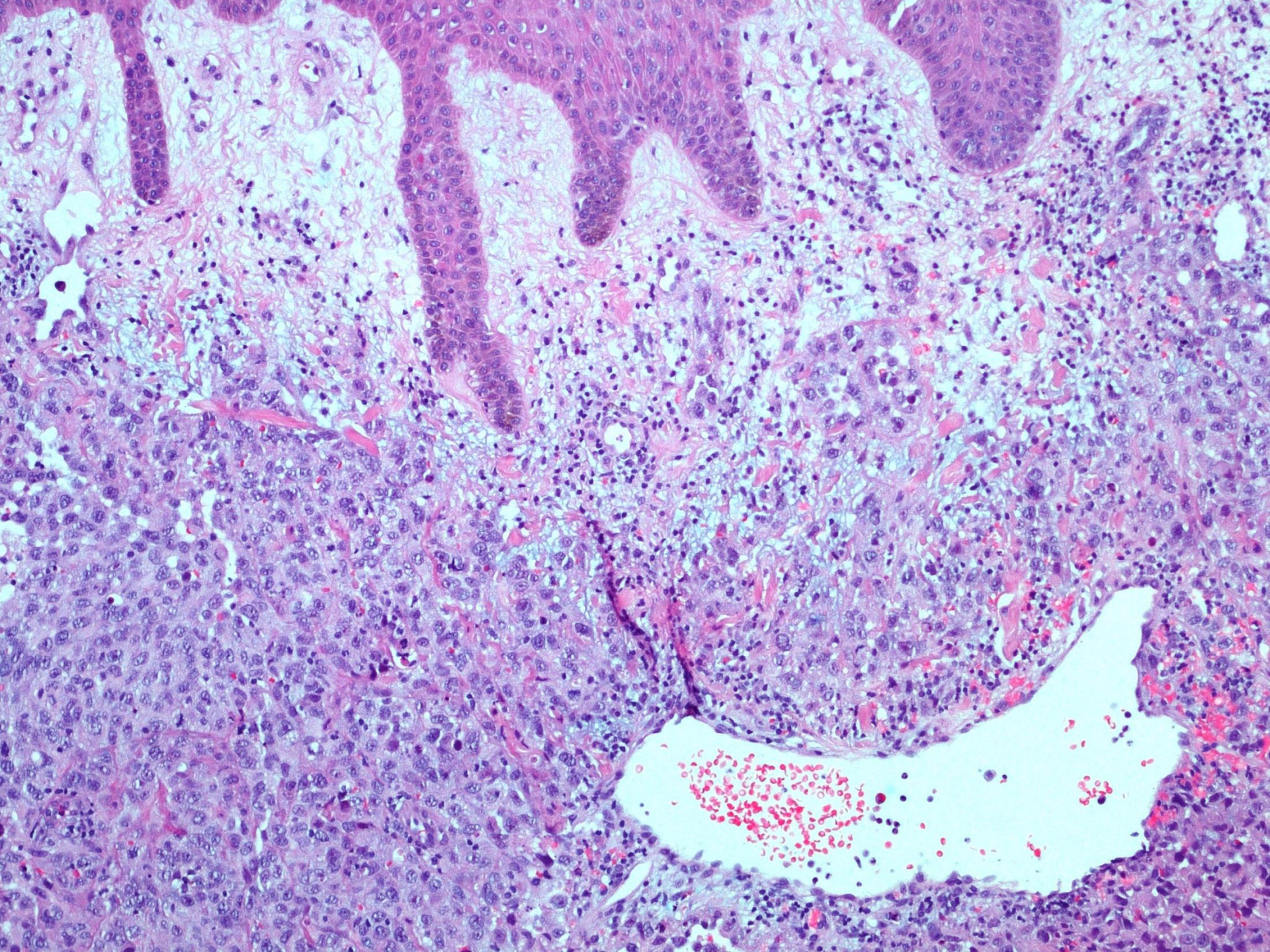
- Imobilný pacient

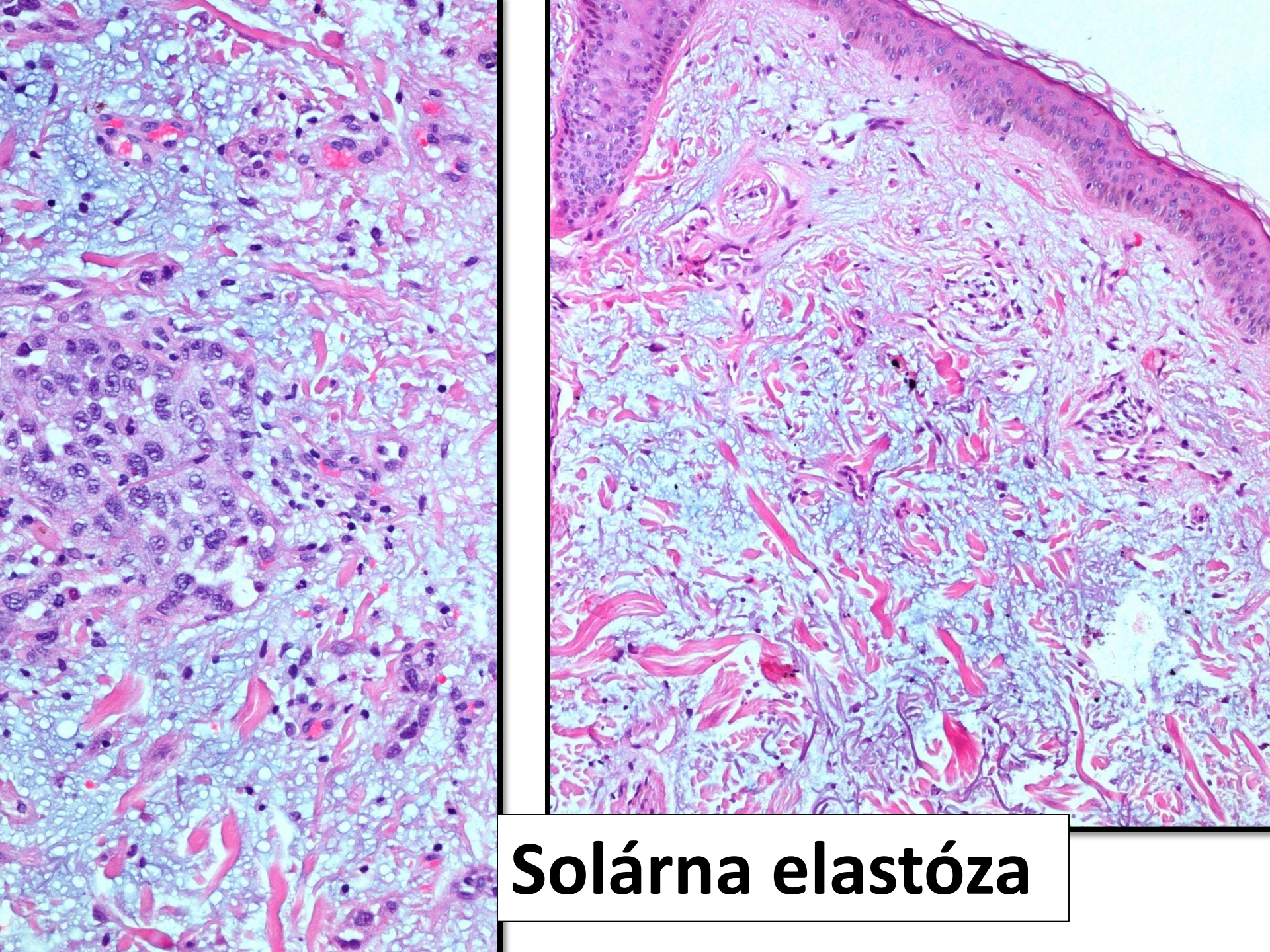
- Anamnesticky:
- Jún 2017 „po páde hematóm“

- Evidovaný a liečený ako posttraumatický infikovaný ulkus, poúrazový defekt

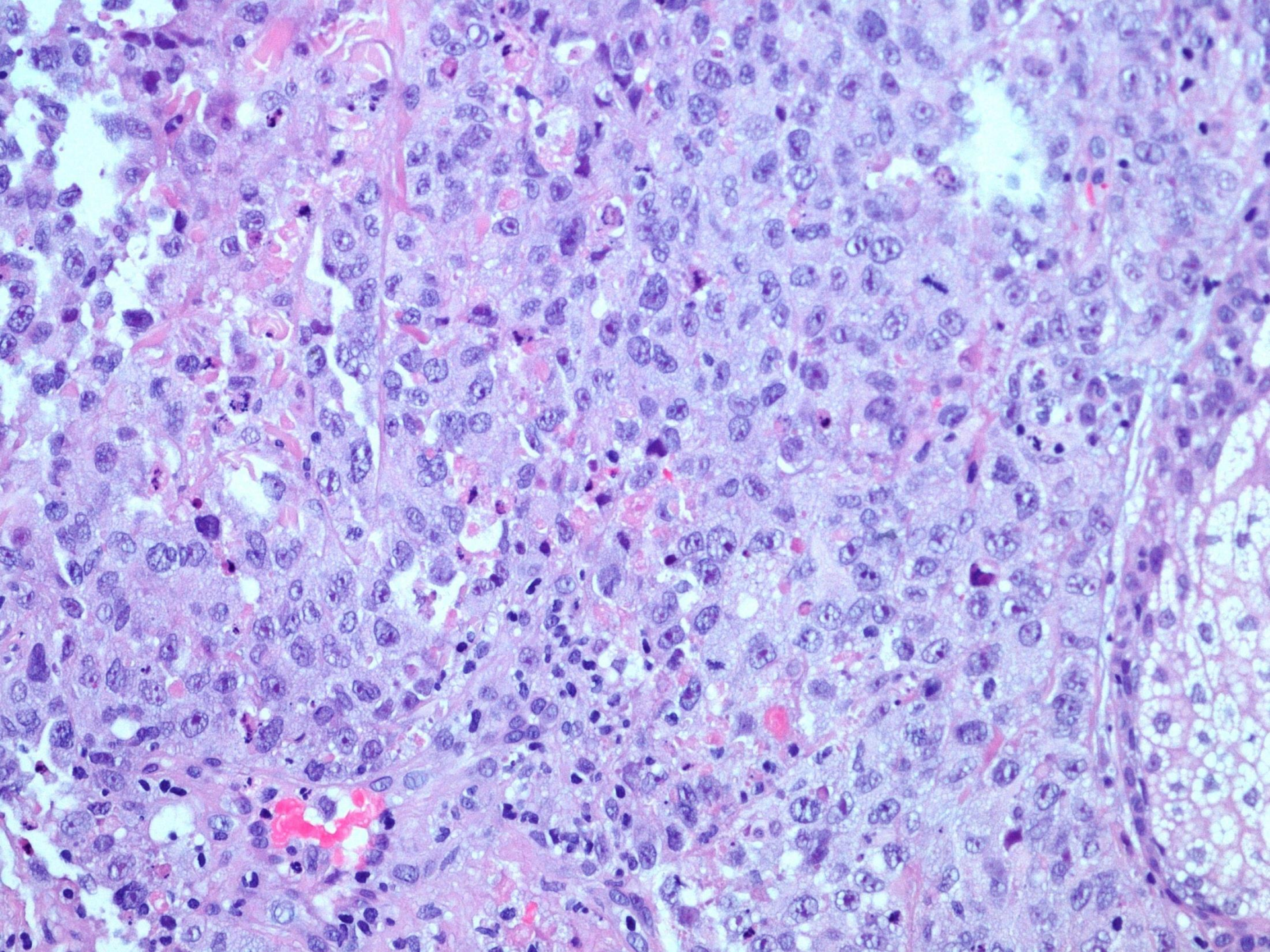
- Po ½ roku (december 2017 – poskytnutá fotografia) ho vidí dermatológ: zvažuje agresívny malígny novotvar , nevylučuje možnú trichofytia profunda, mykologické vyšetrenie negatívne.

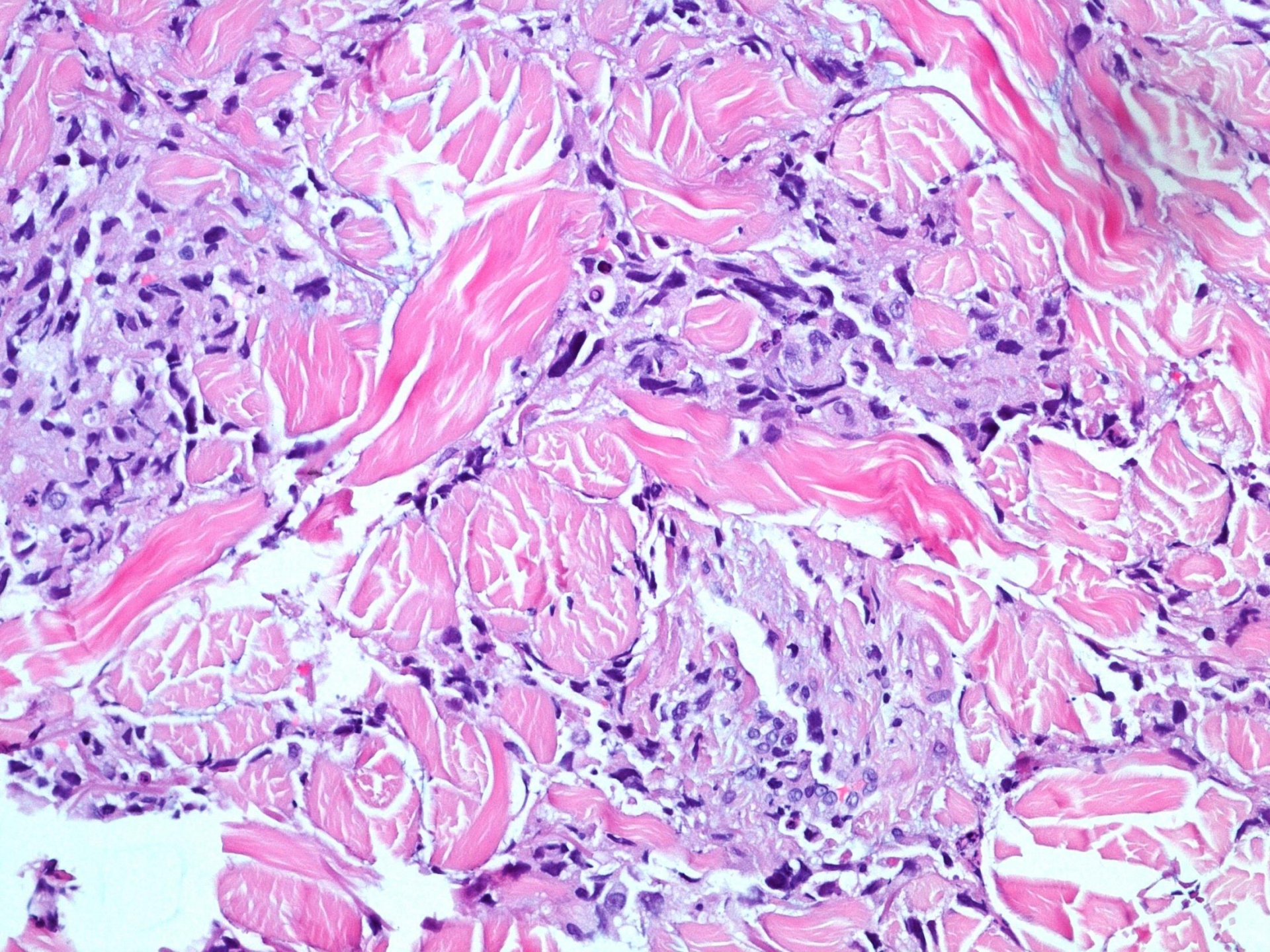


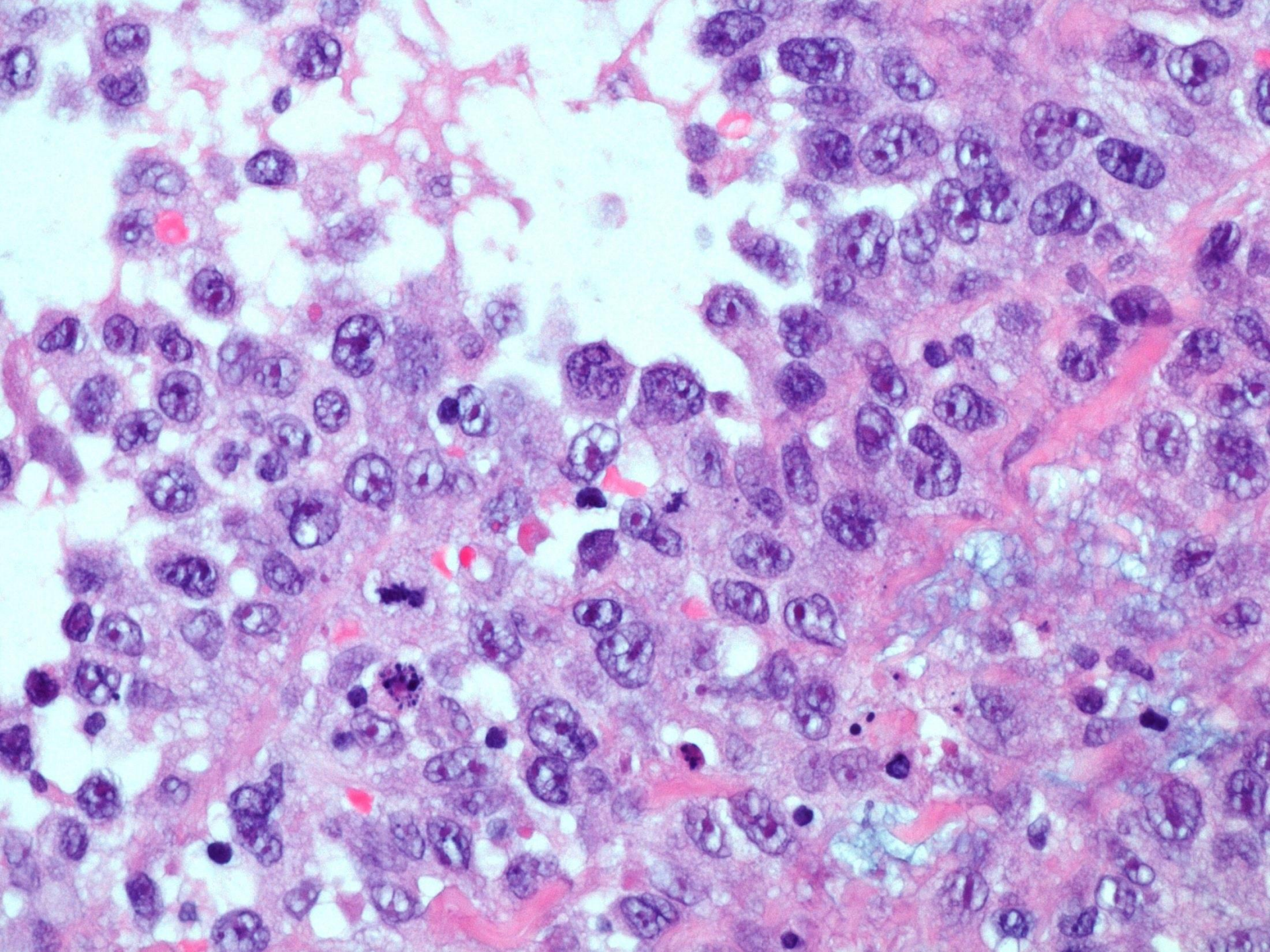


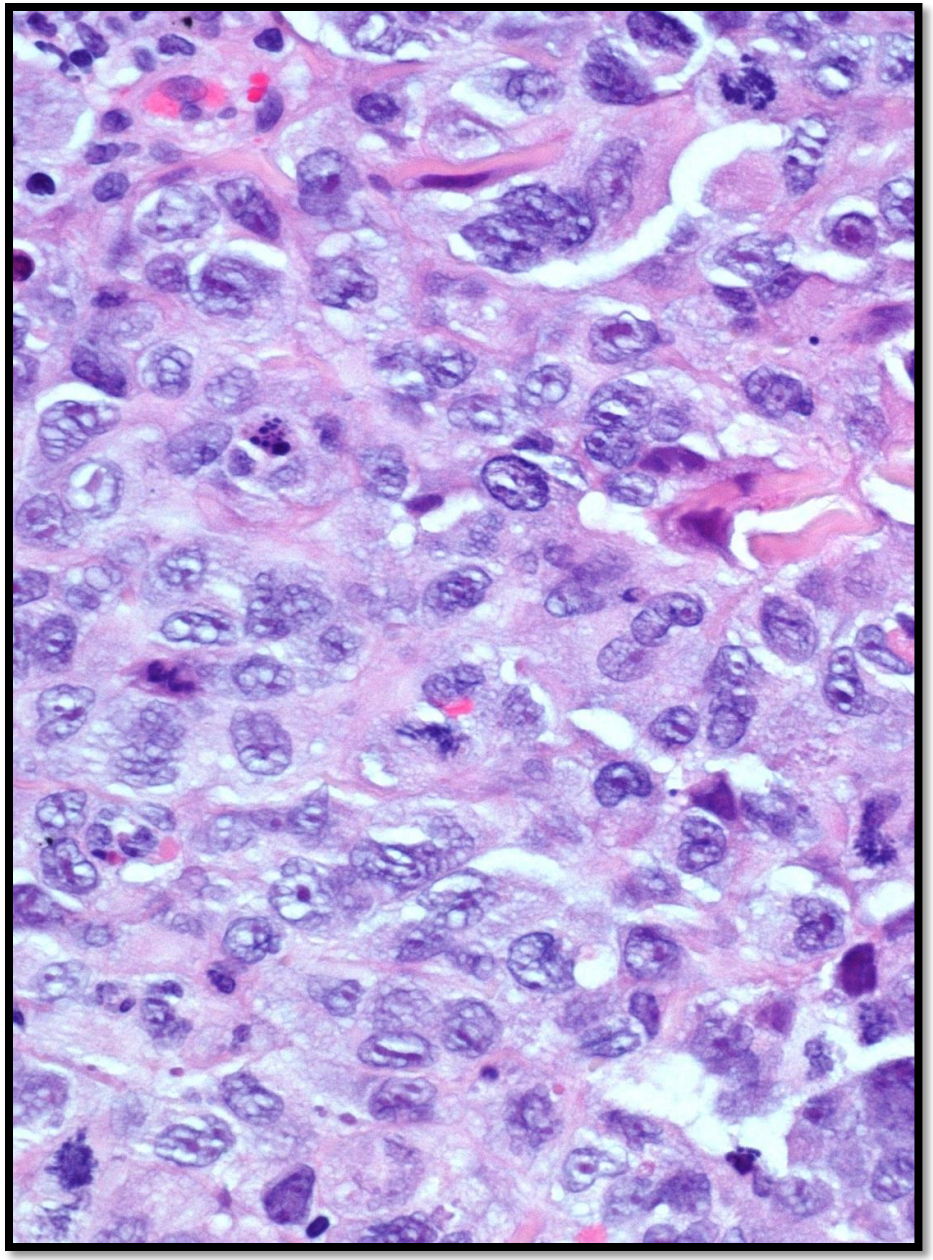


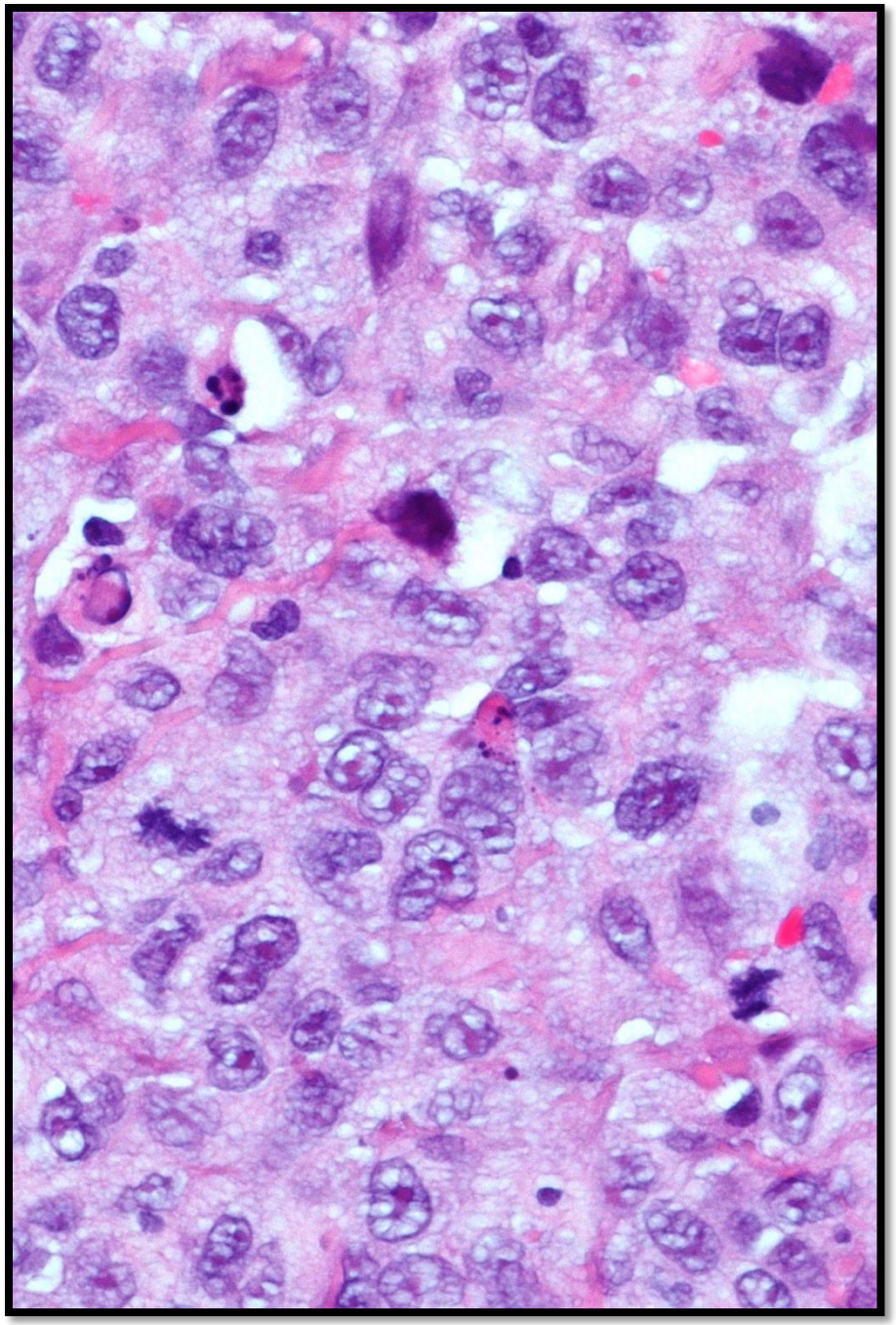
Solárna elastóza

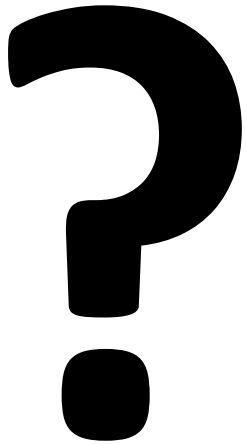


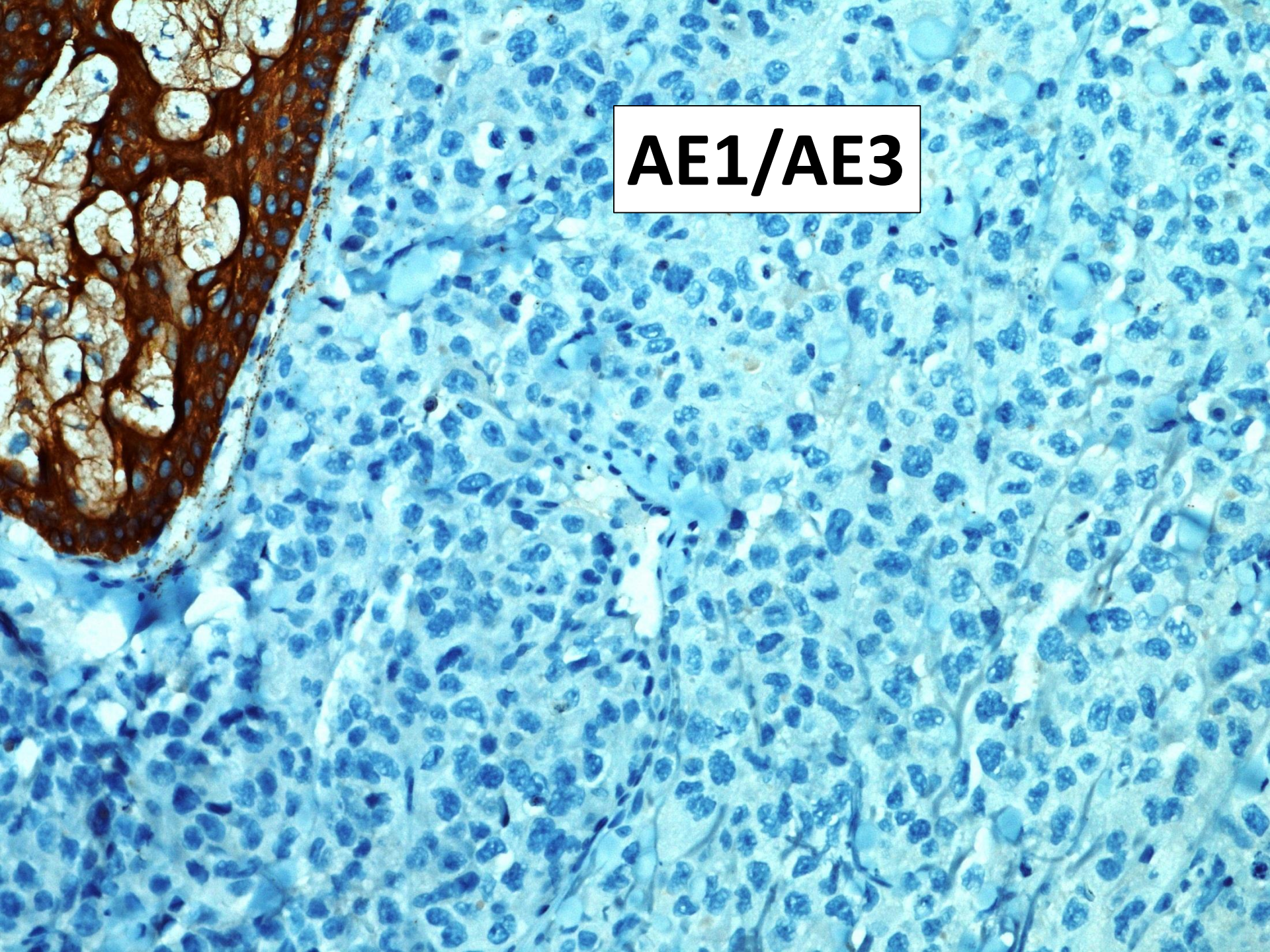








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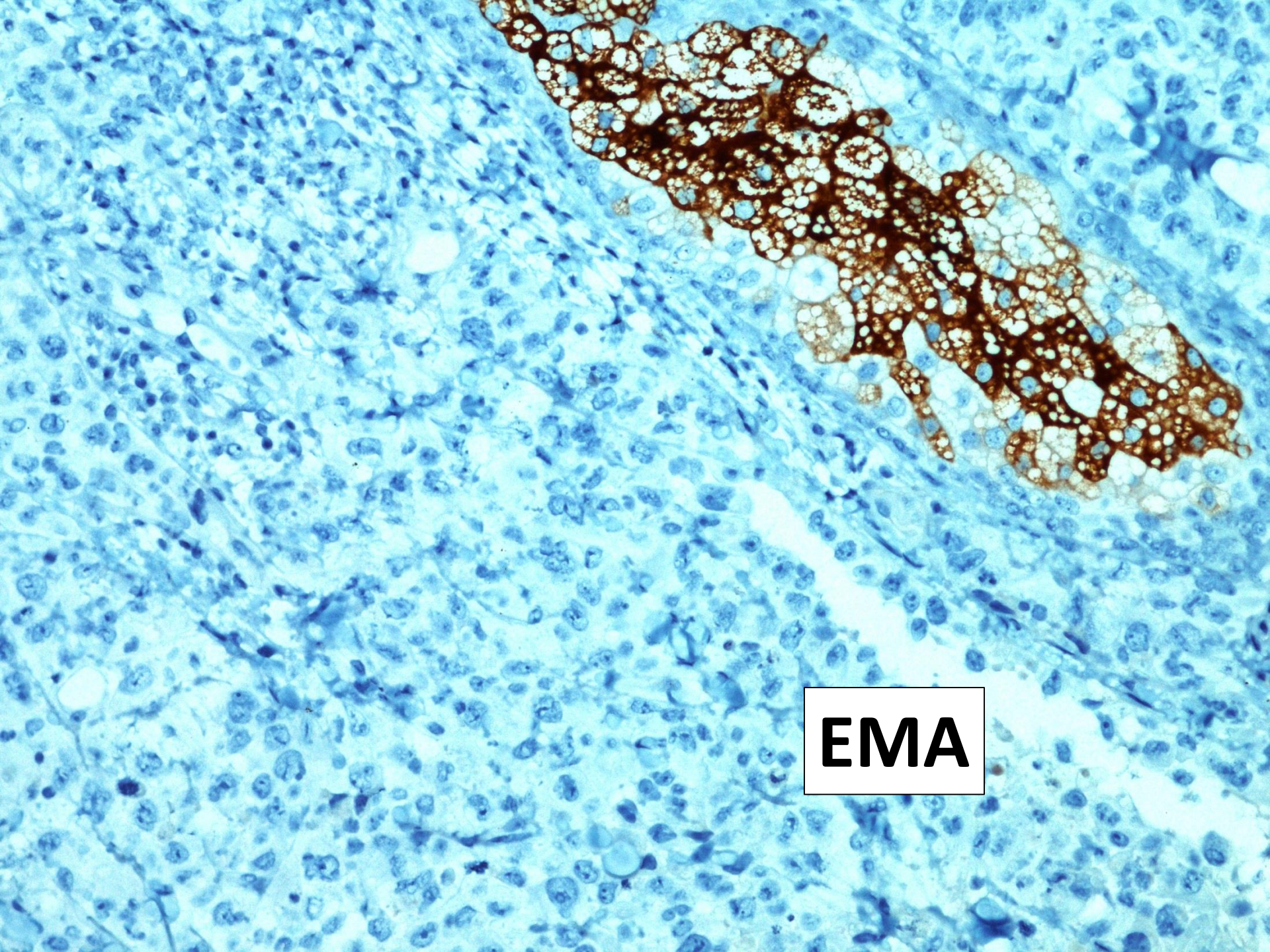
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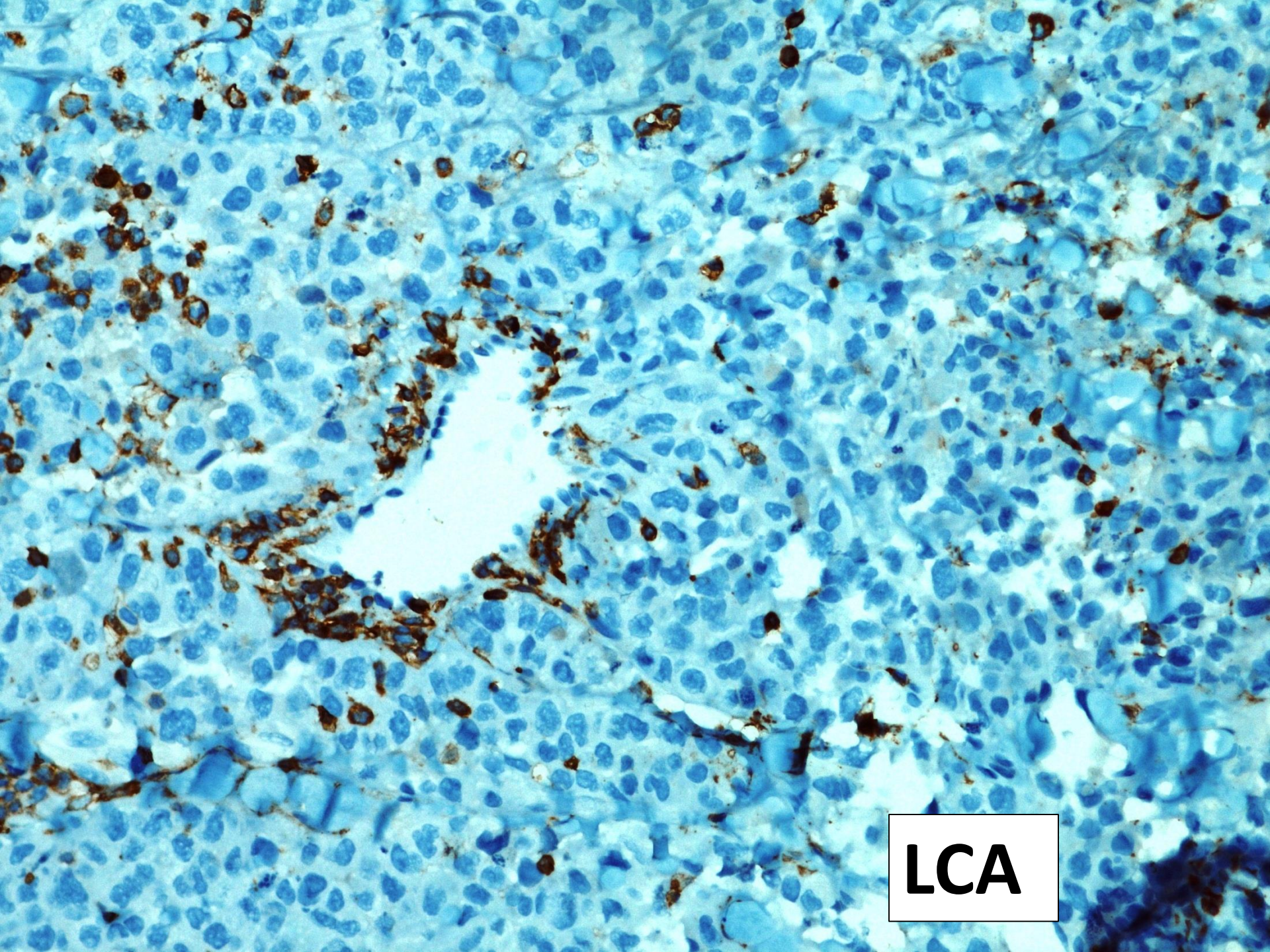
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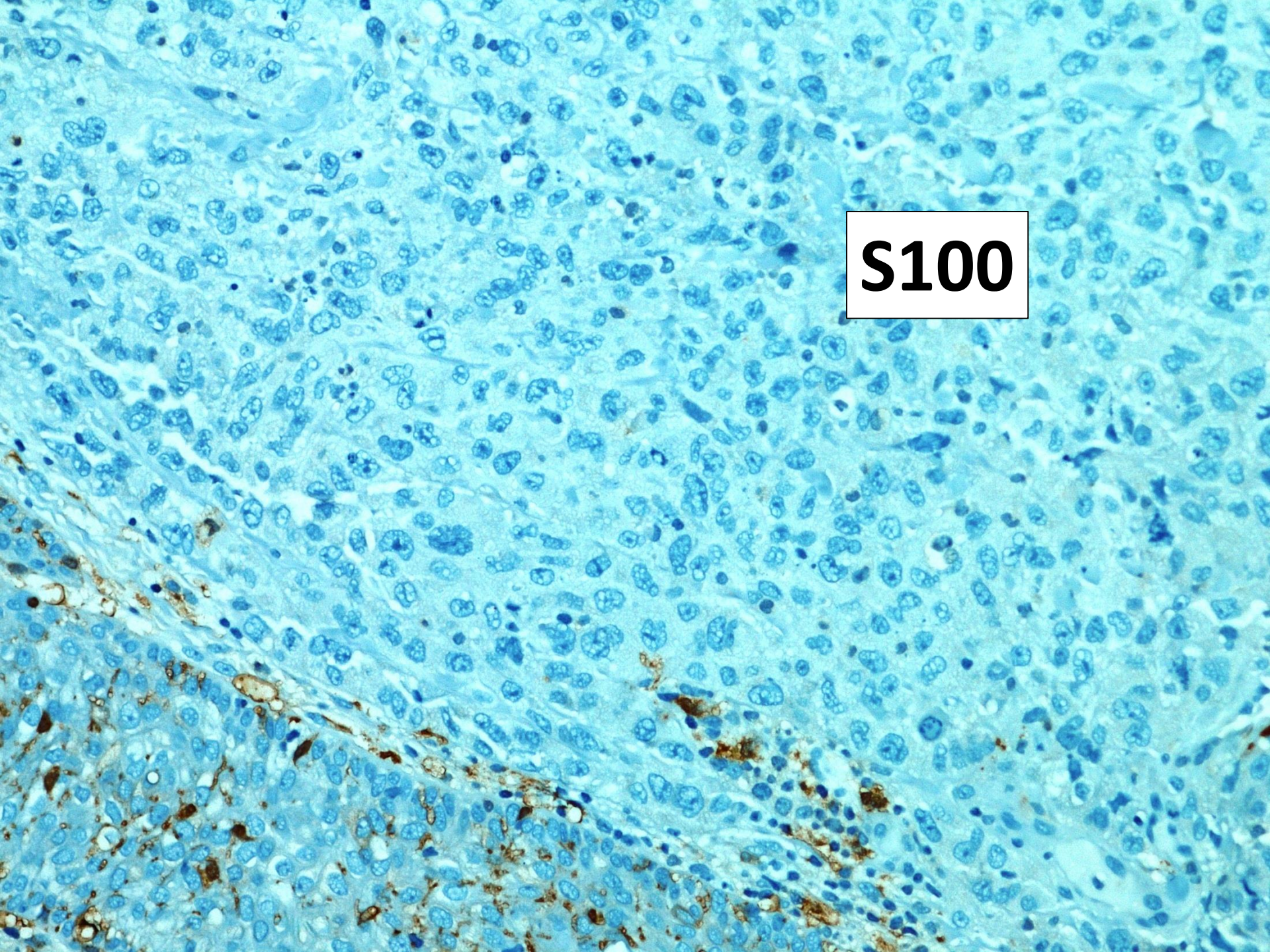
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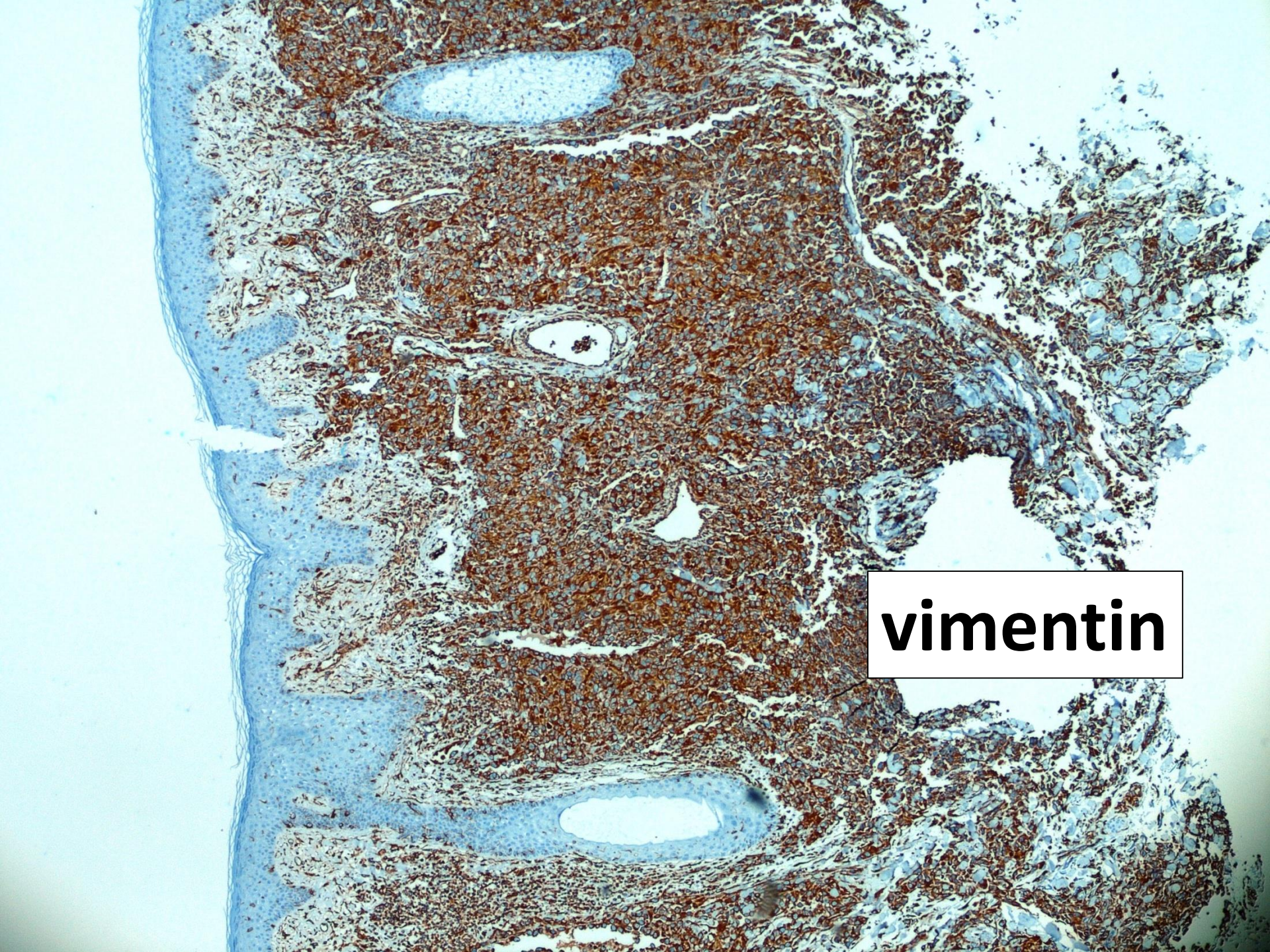
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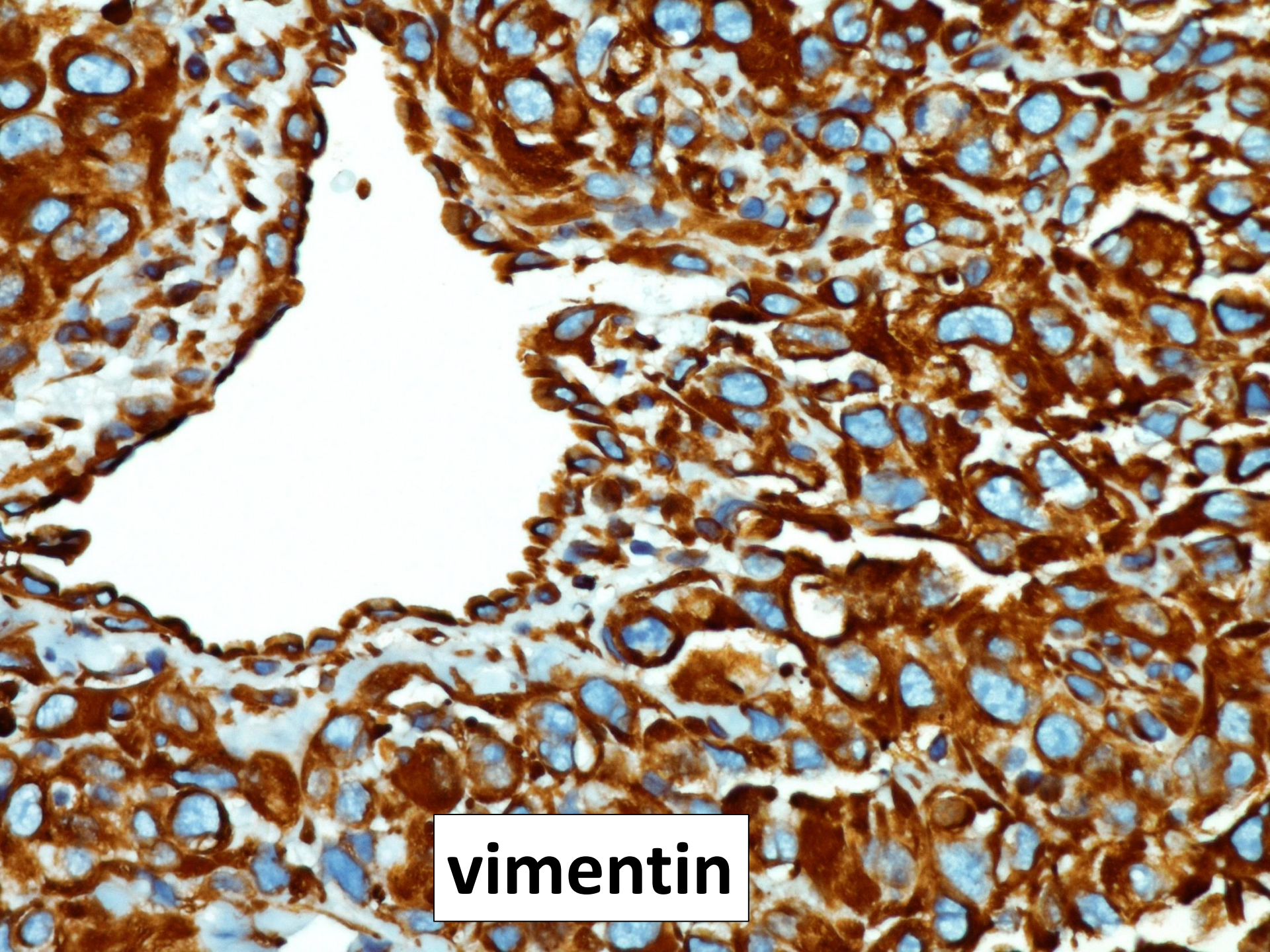
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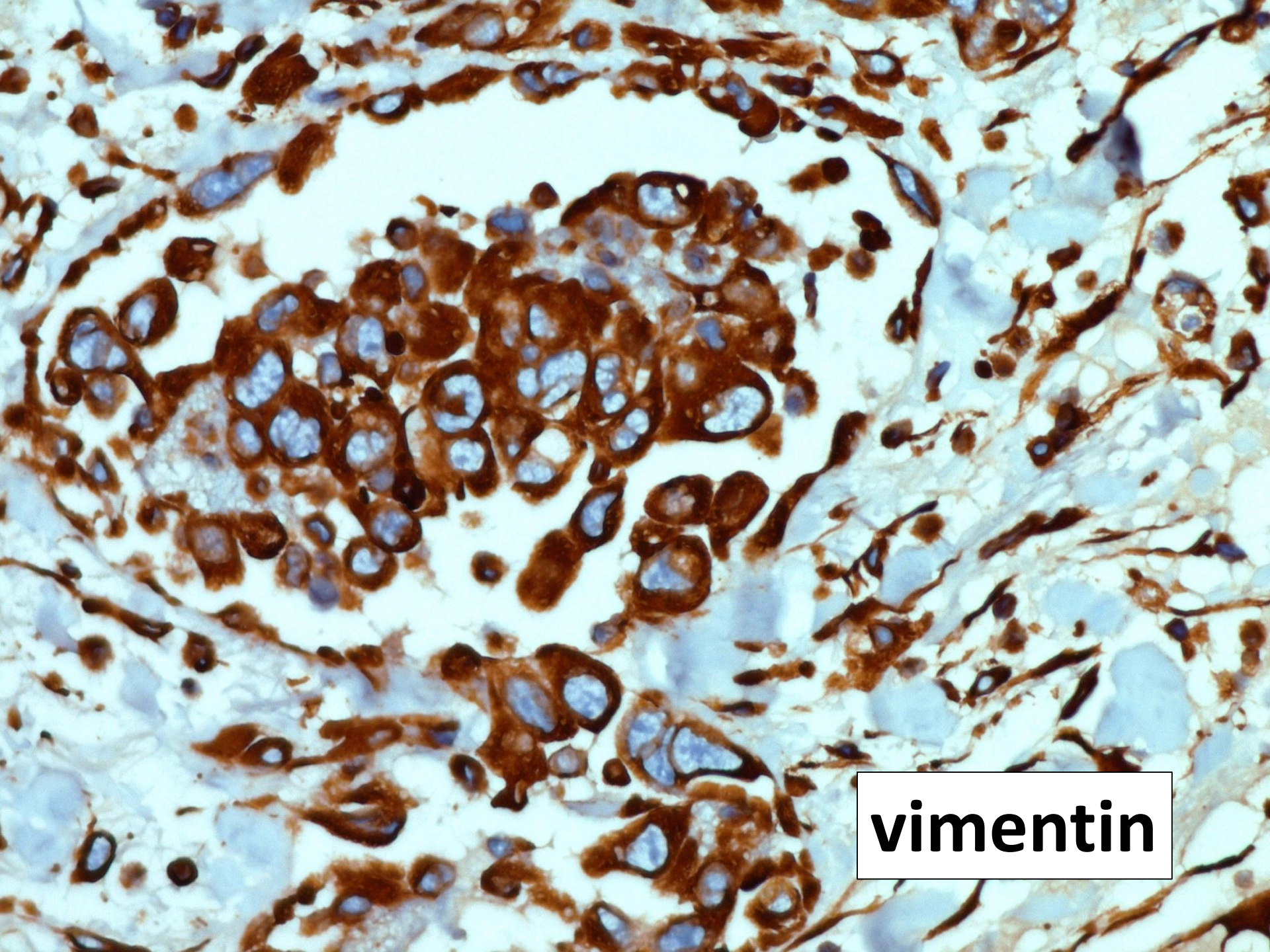
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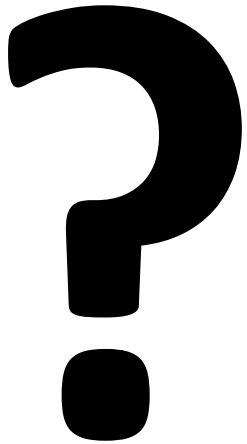

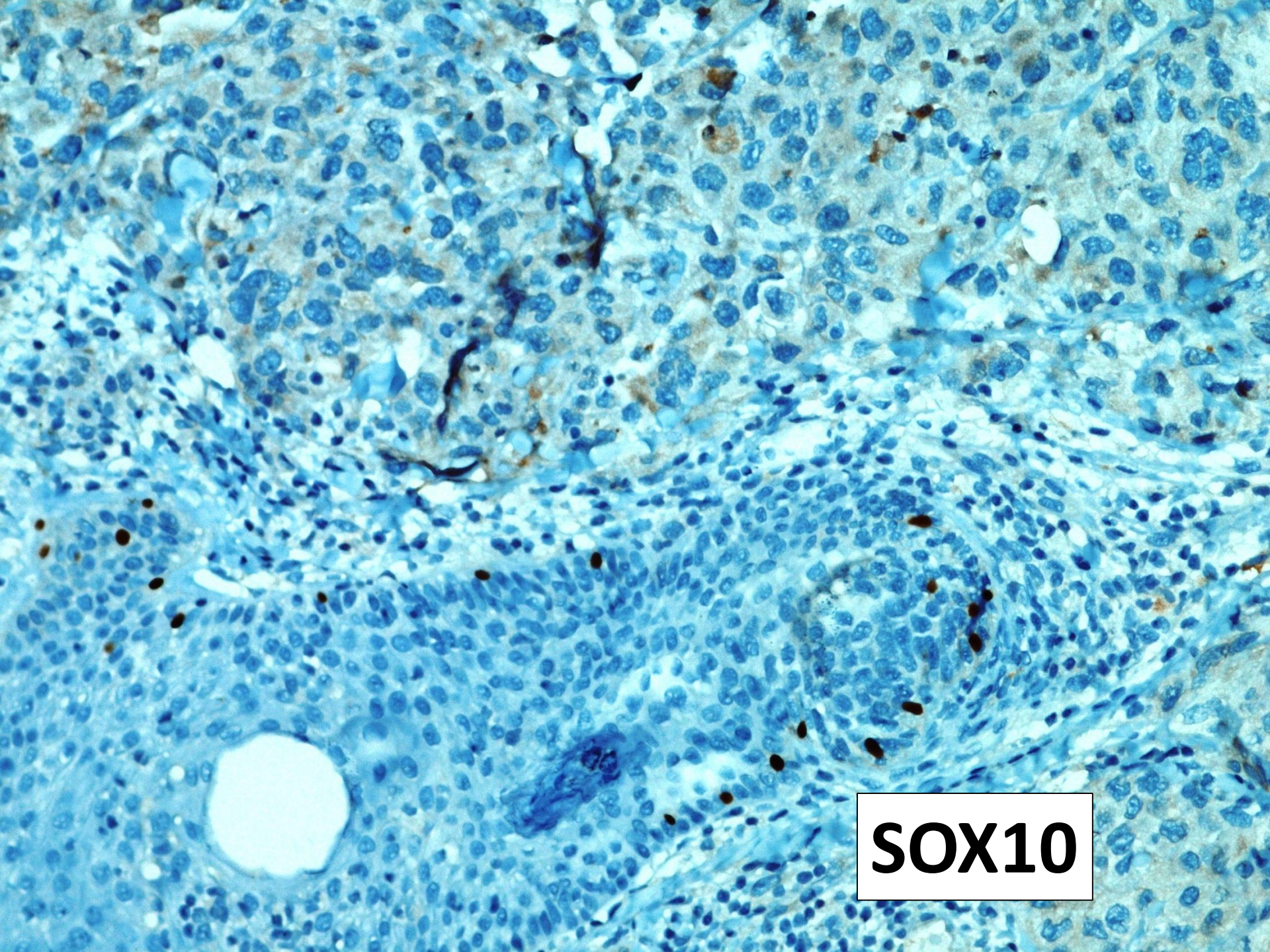
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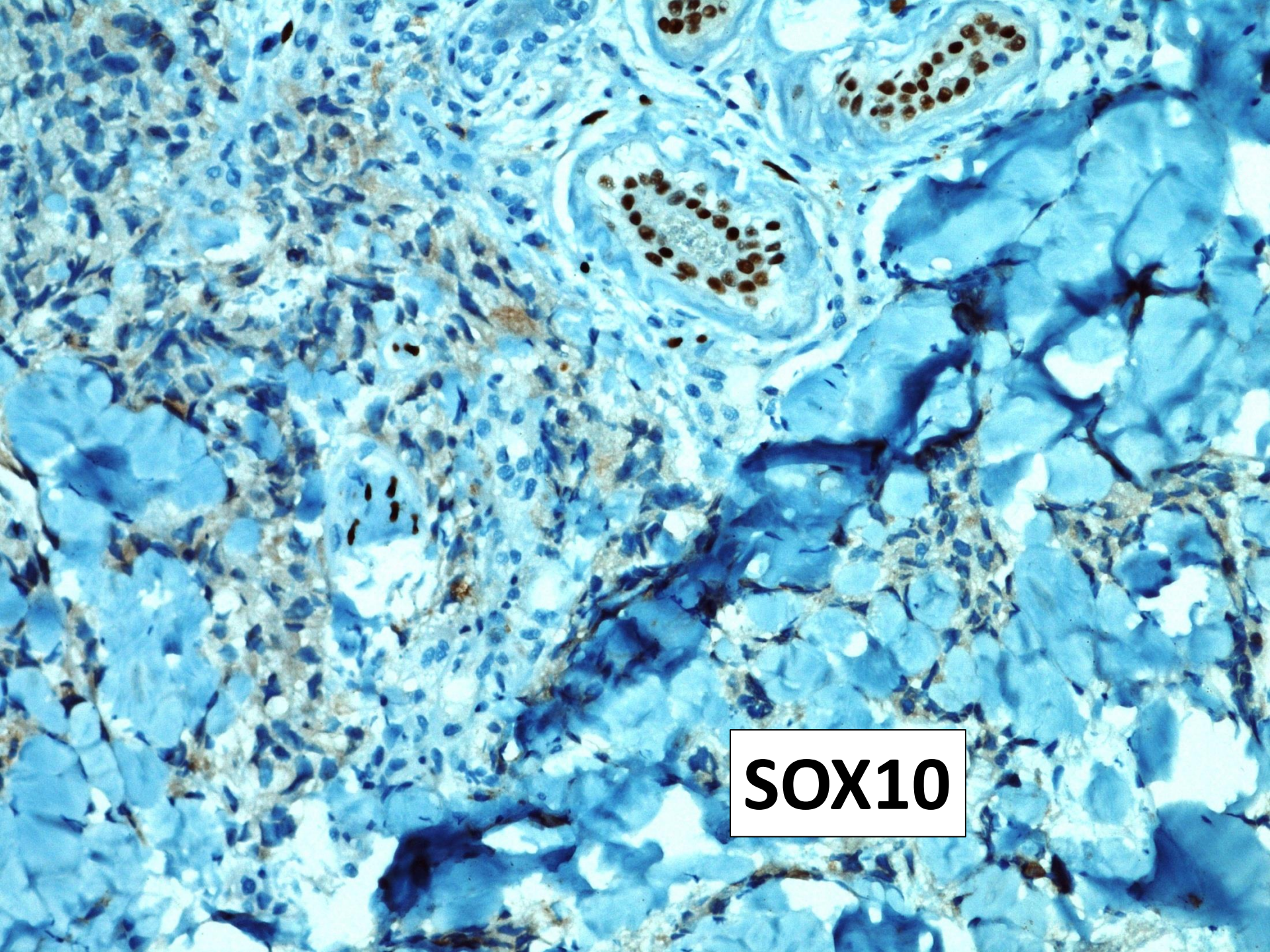
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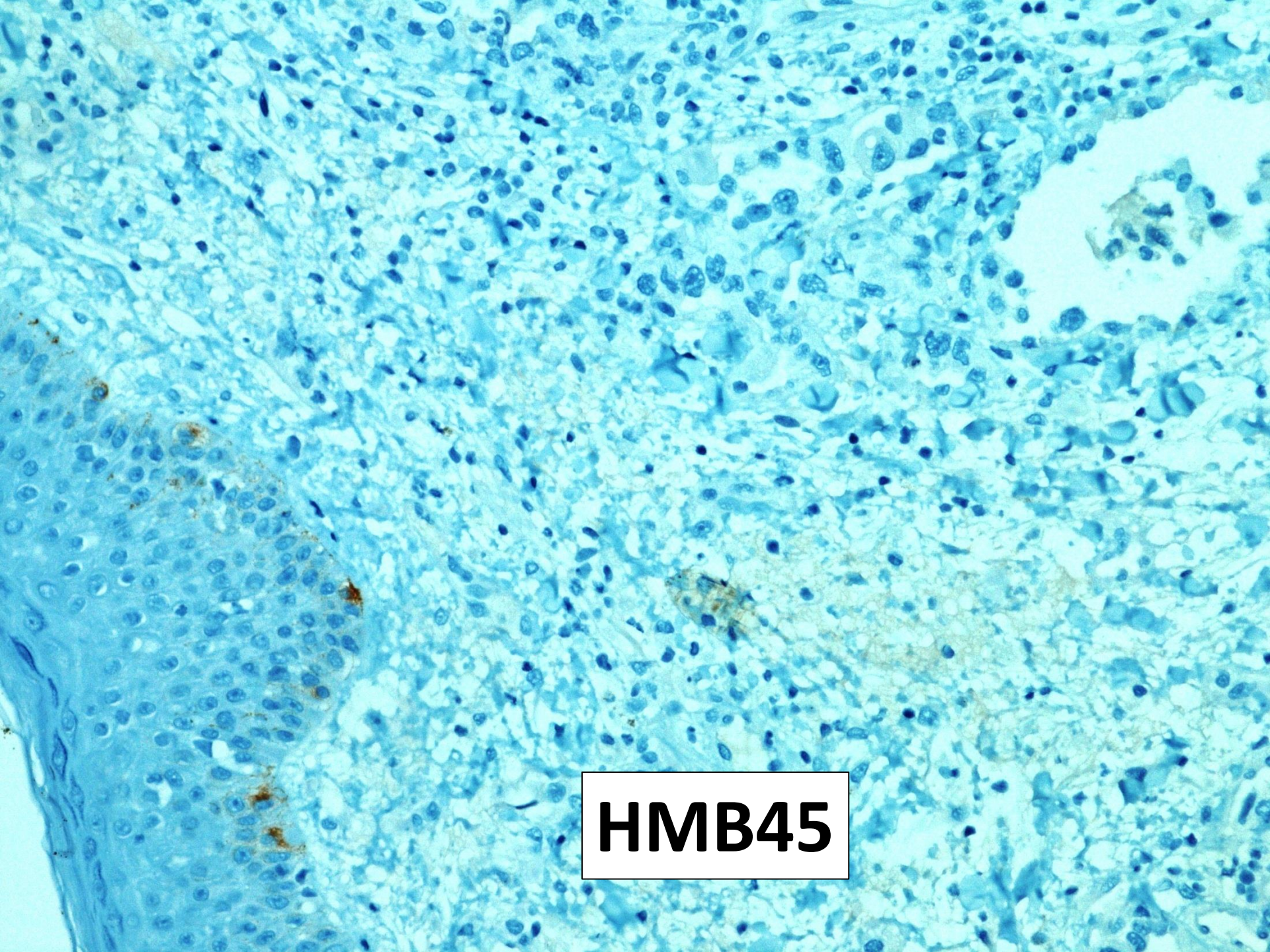
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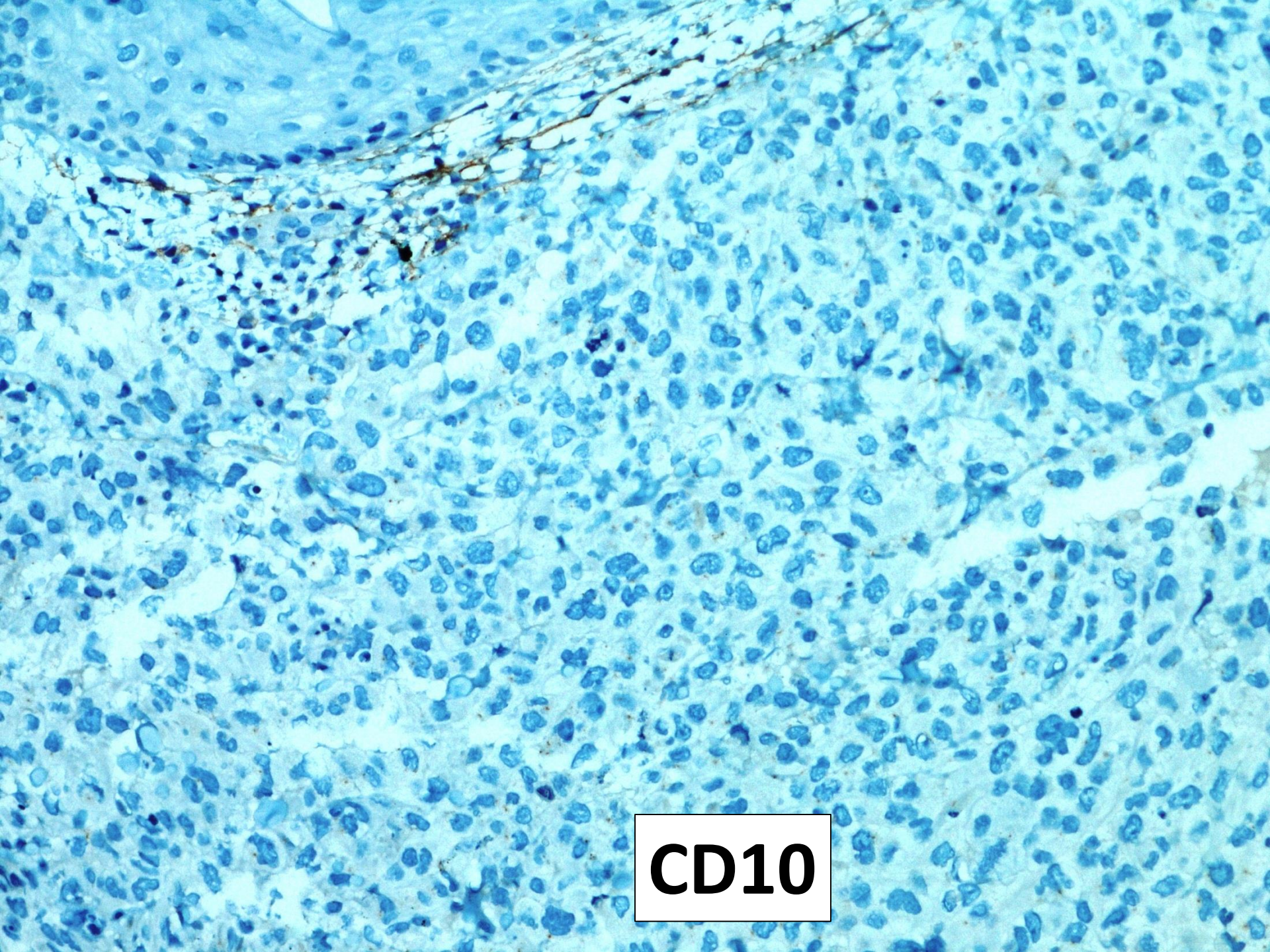
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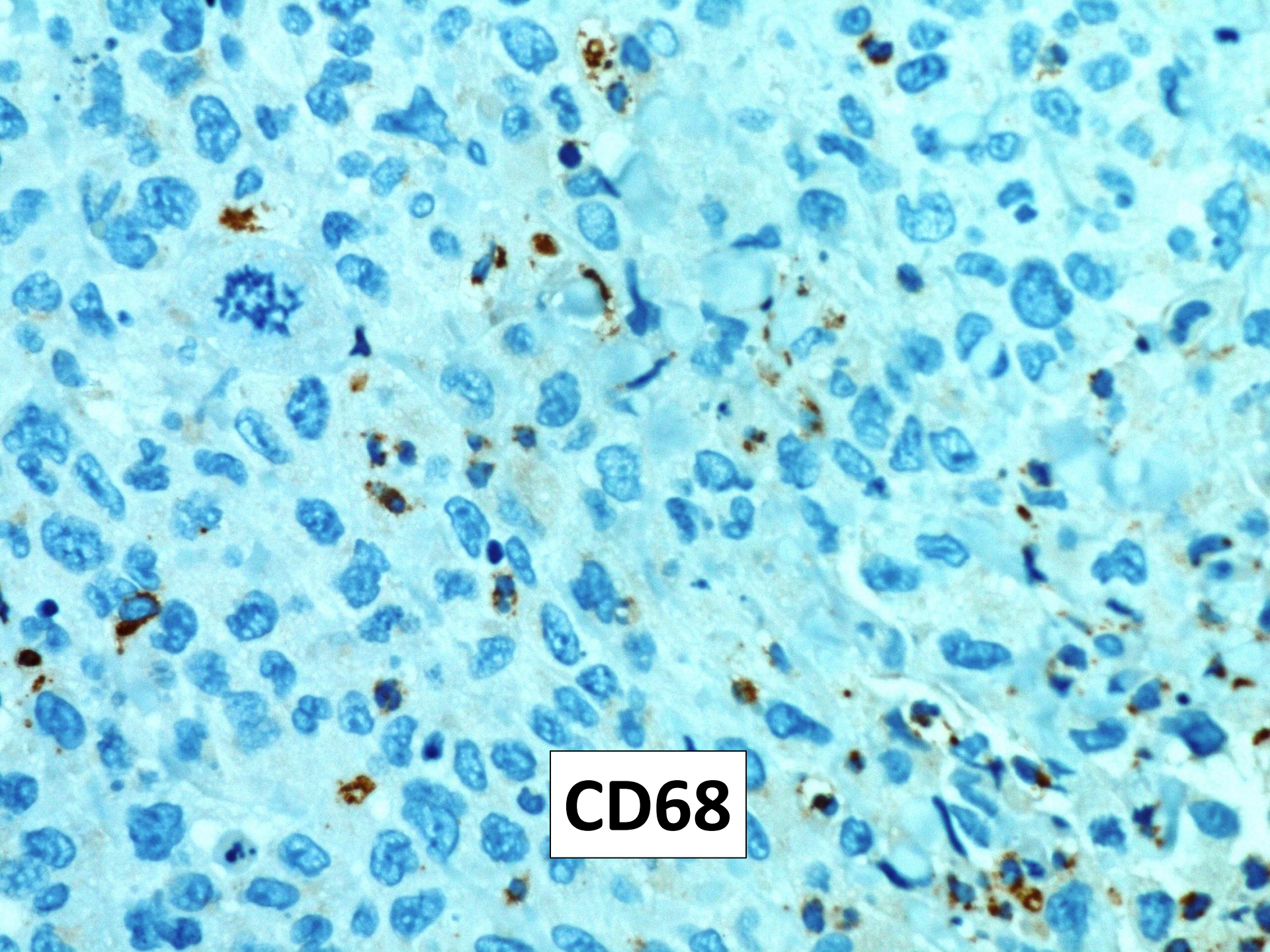
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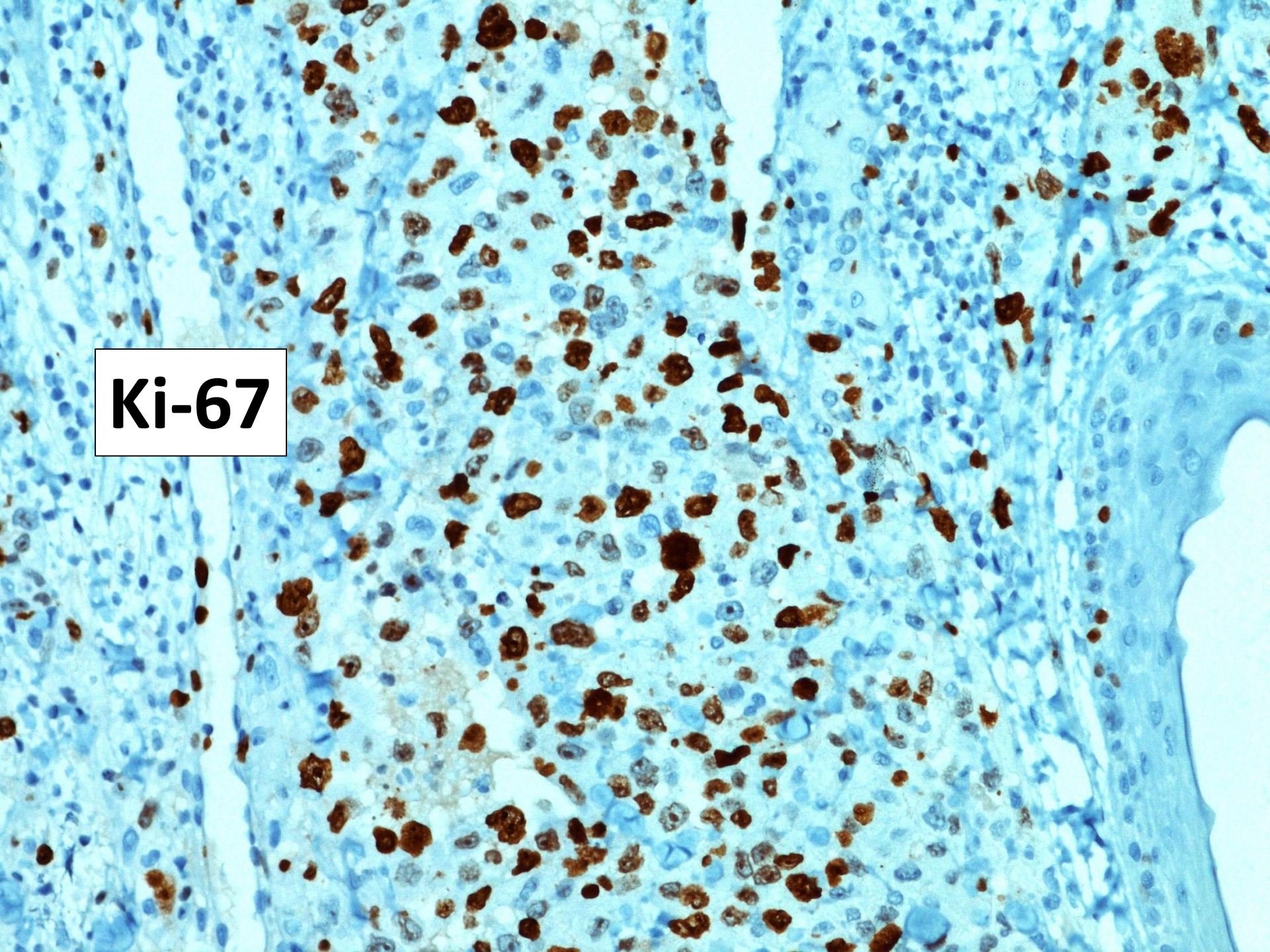
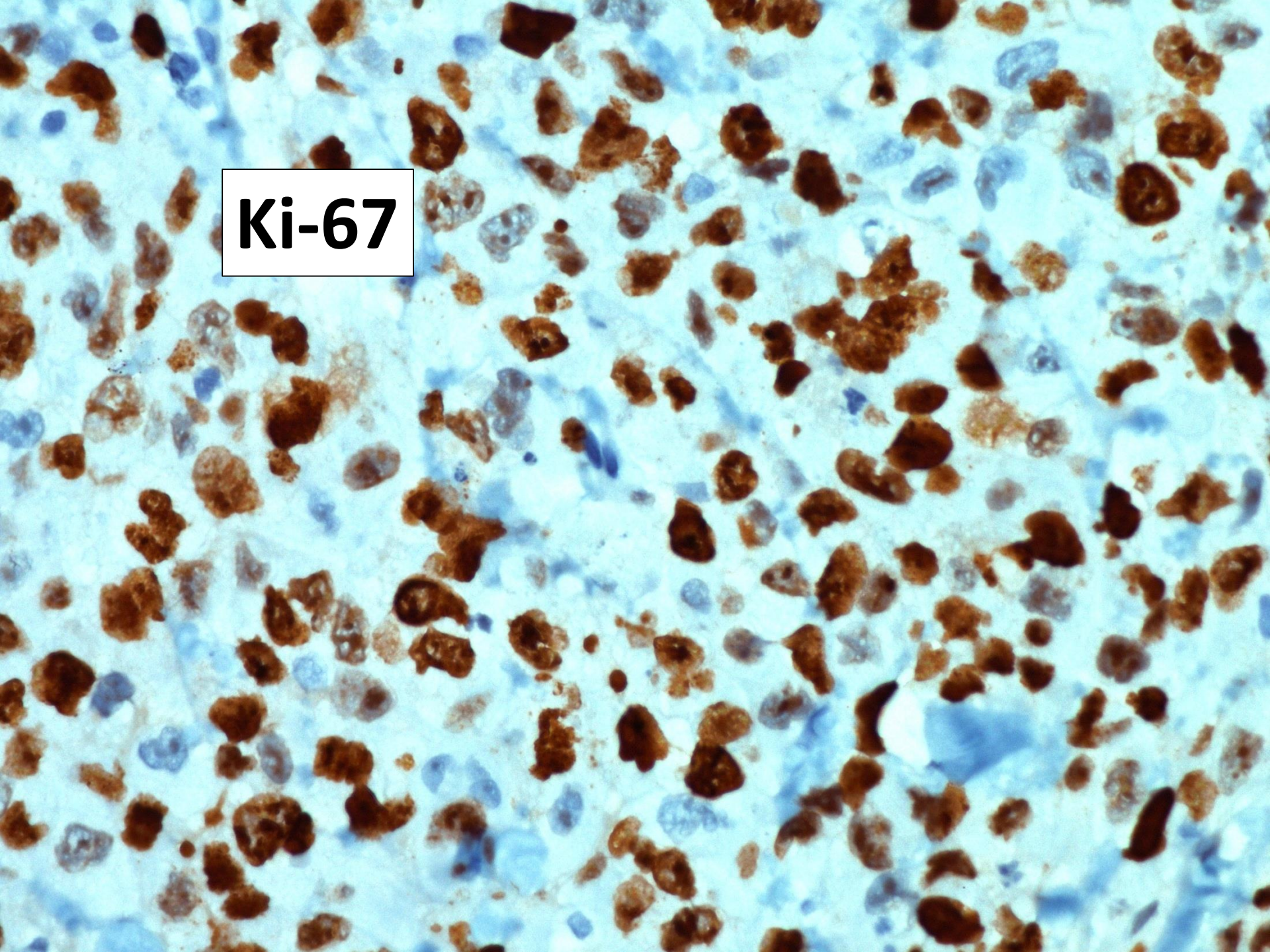

Ki-67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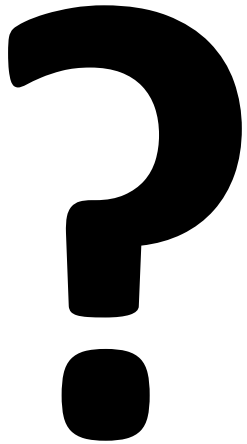

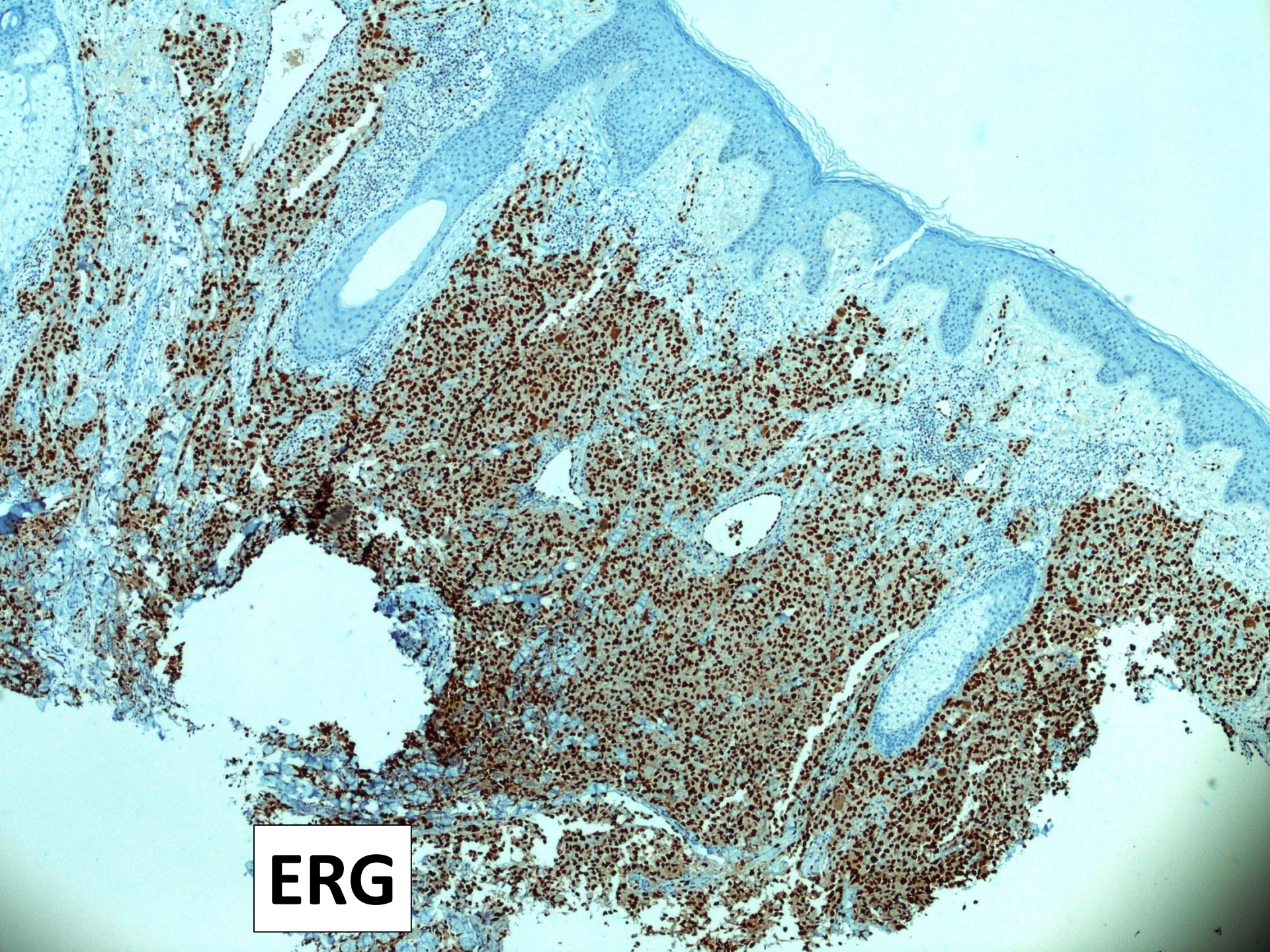
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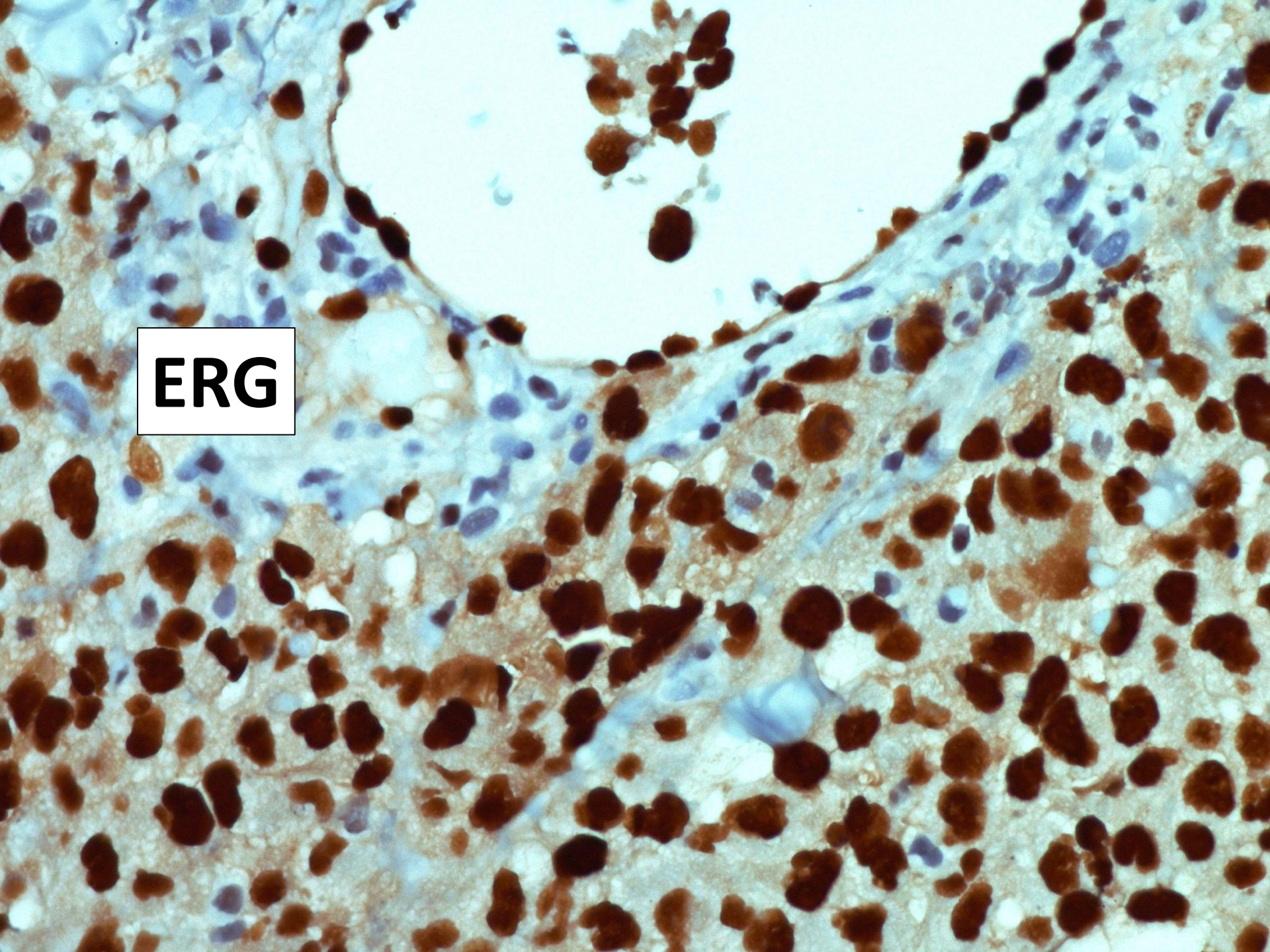
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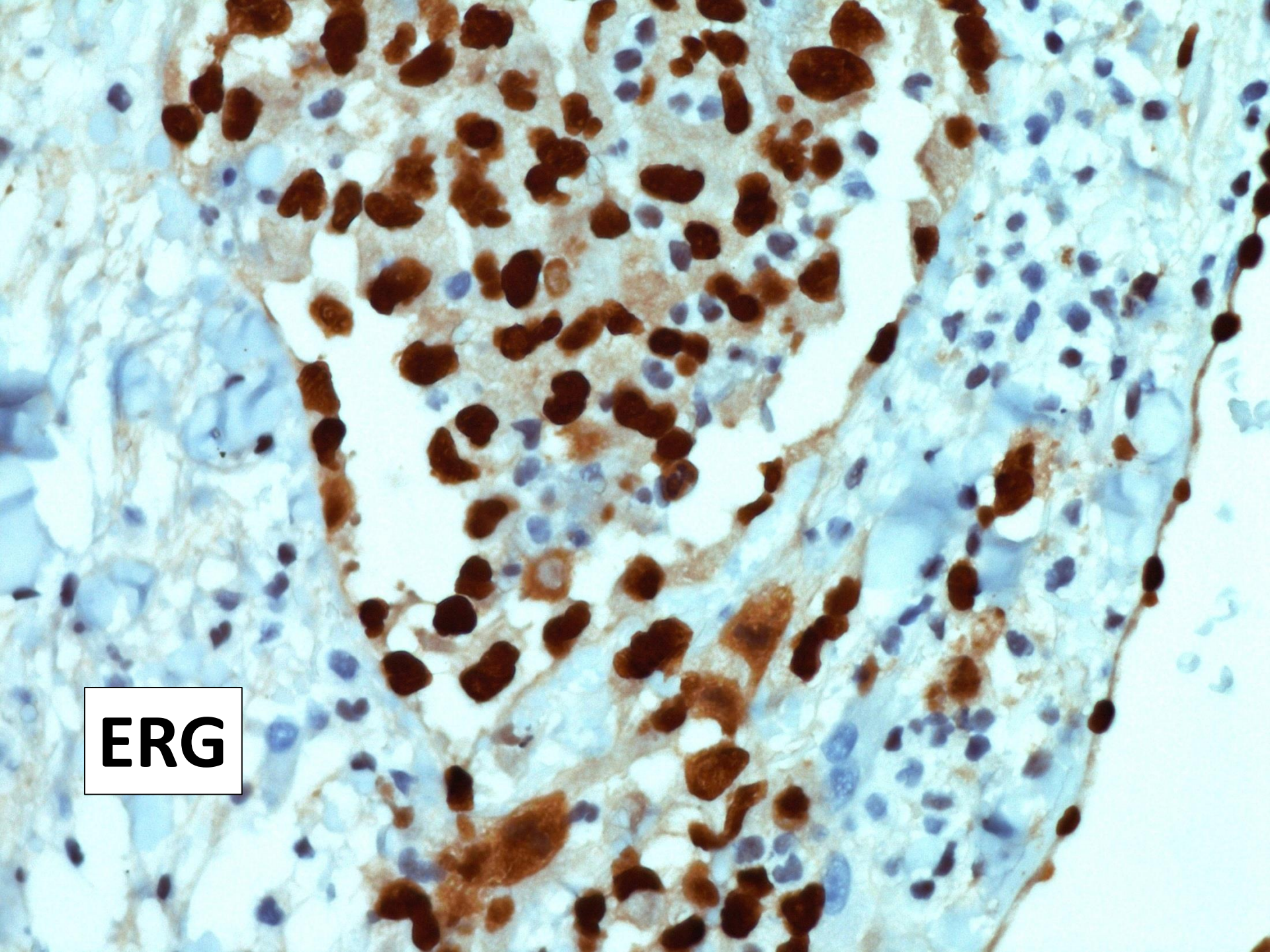
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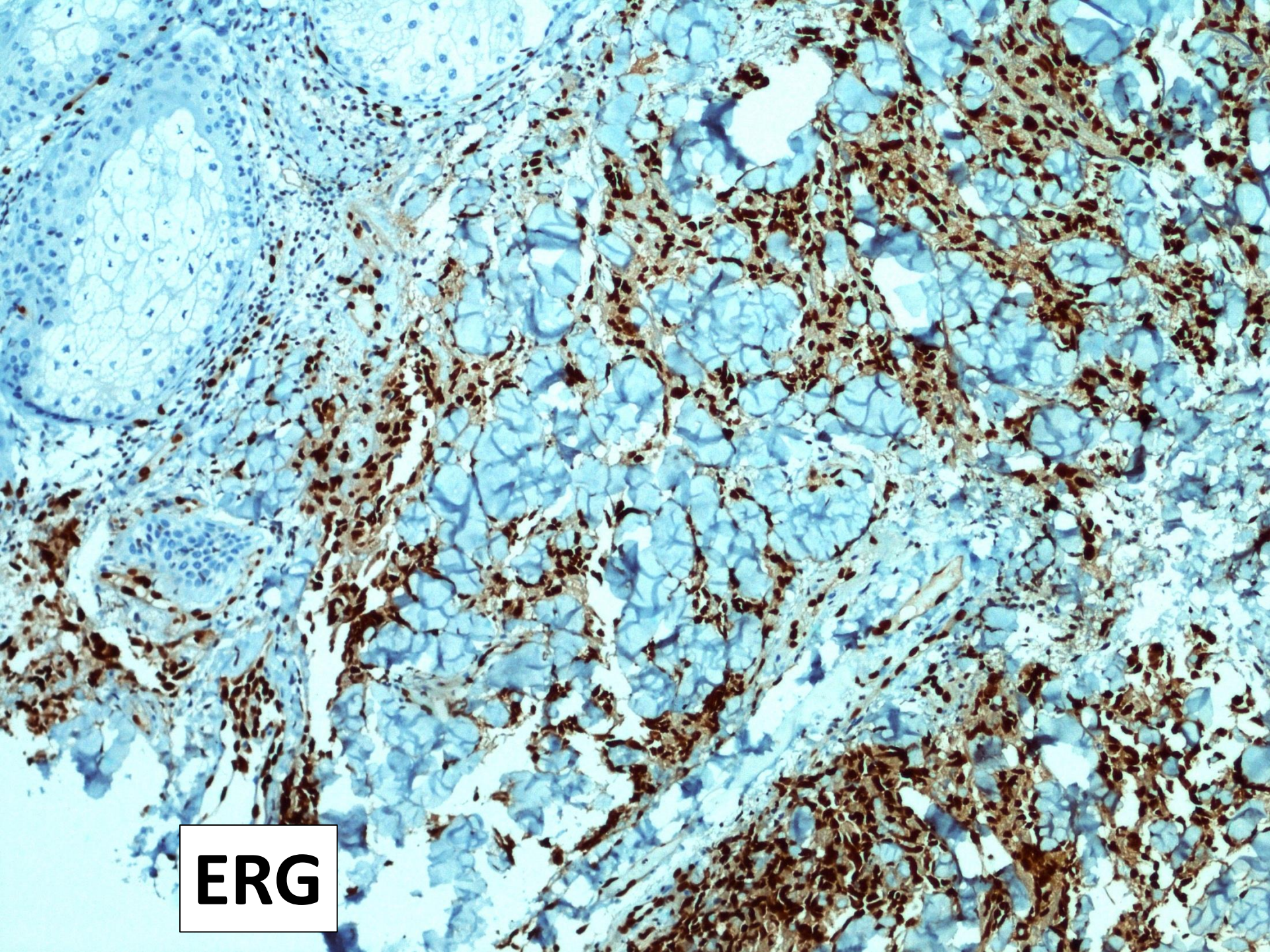
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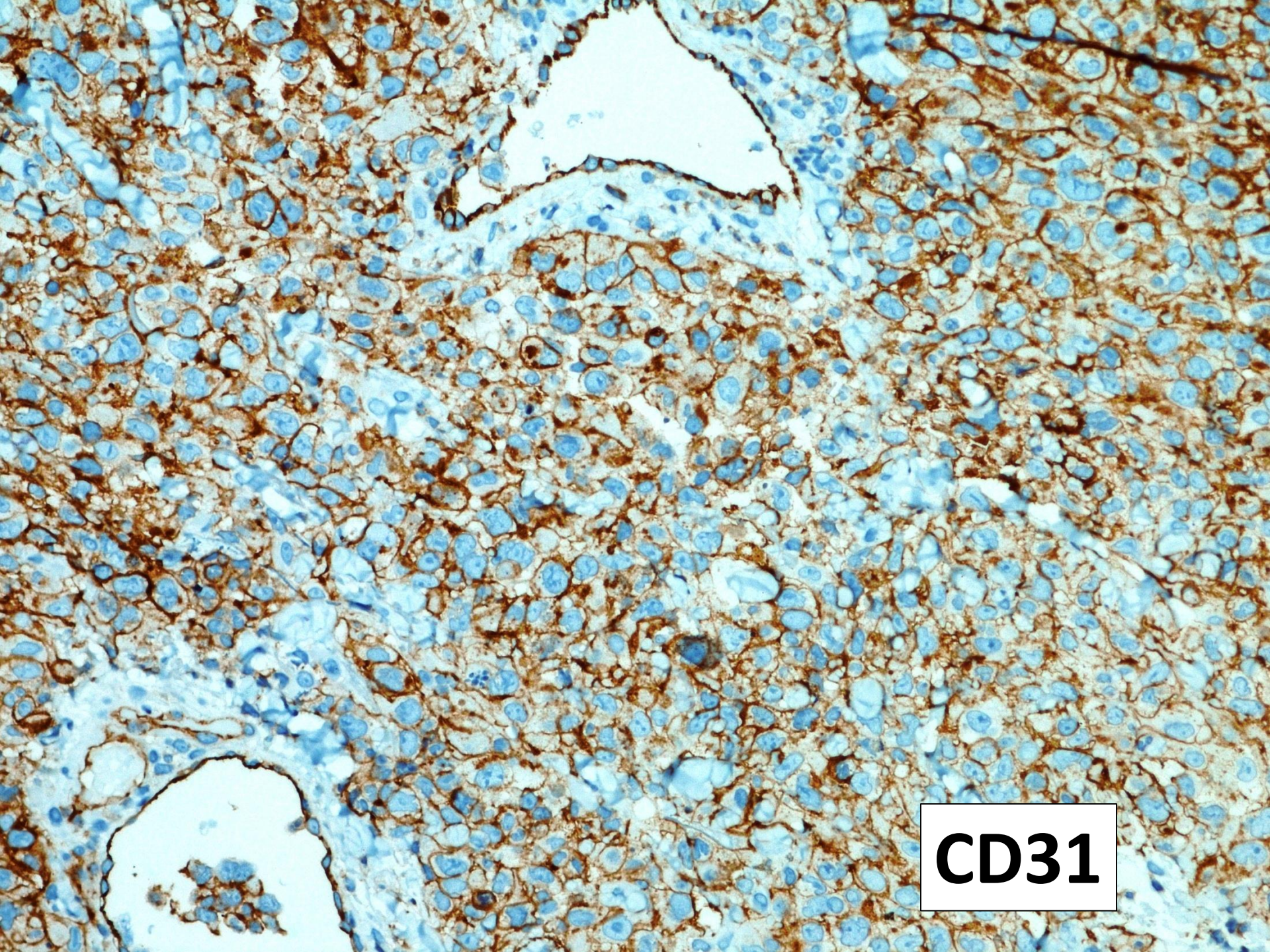
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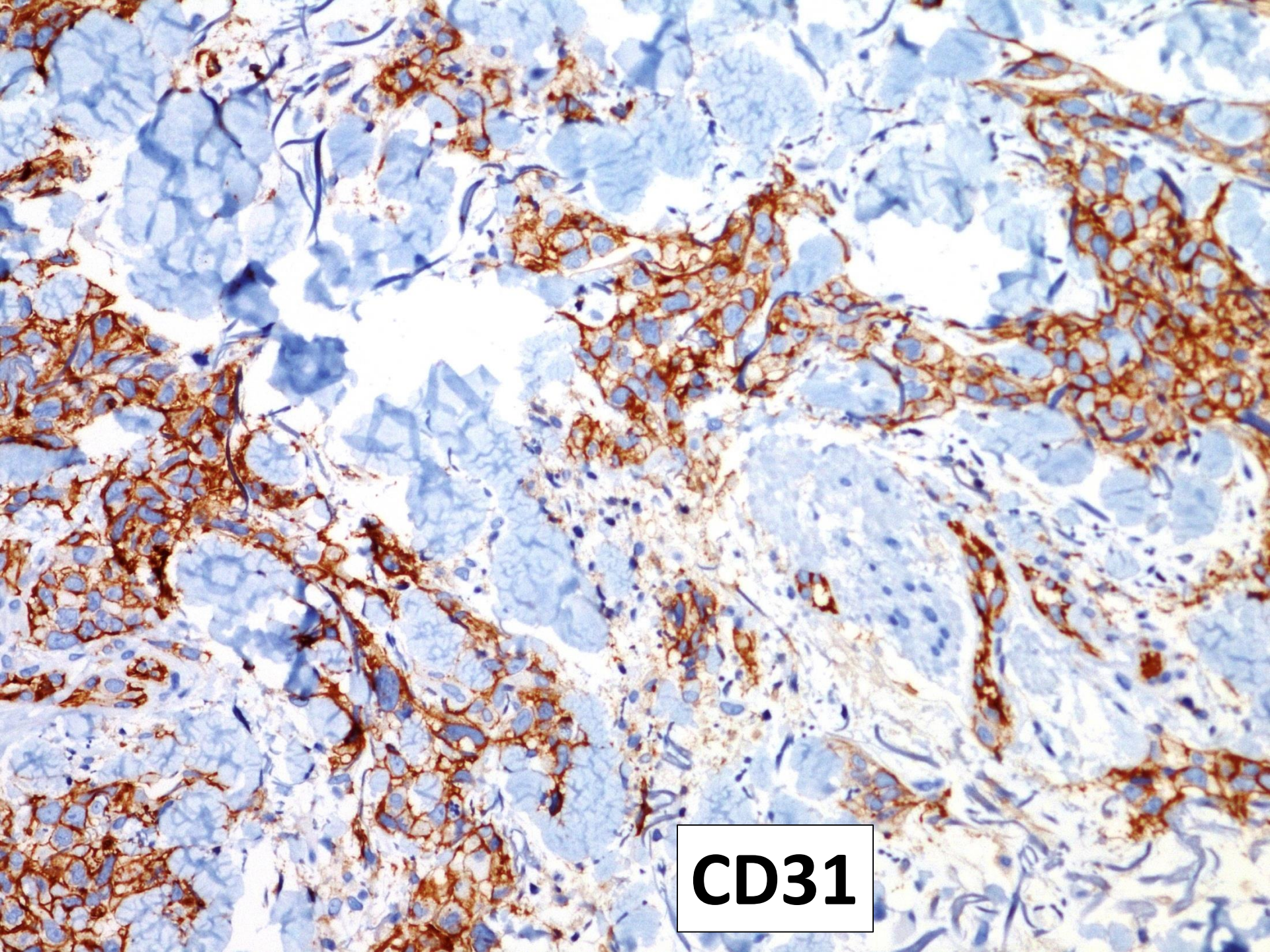
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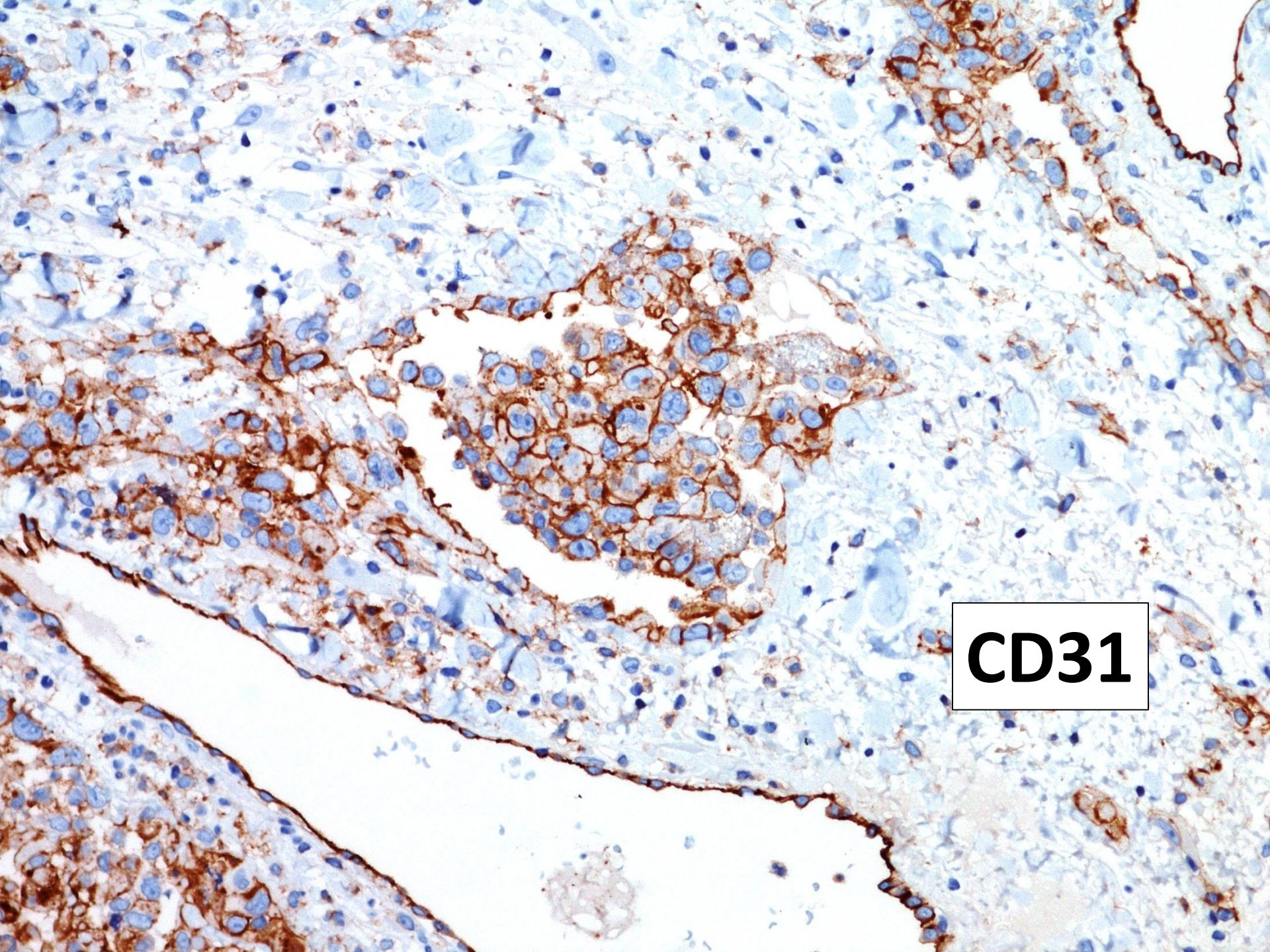
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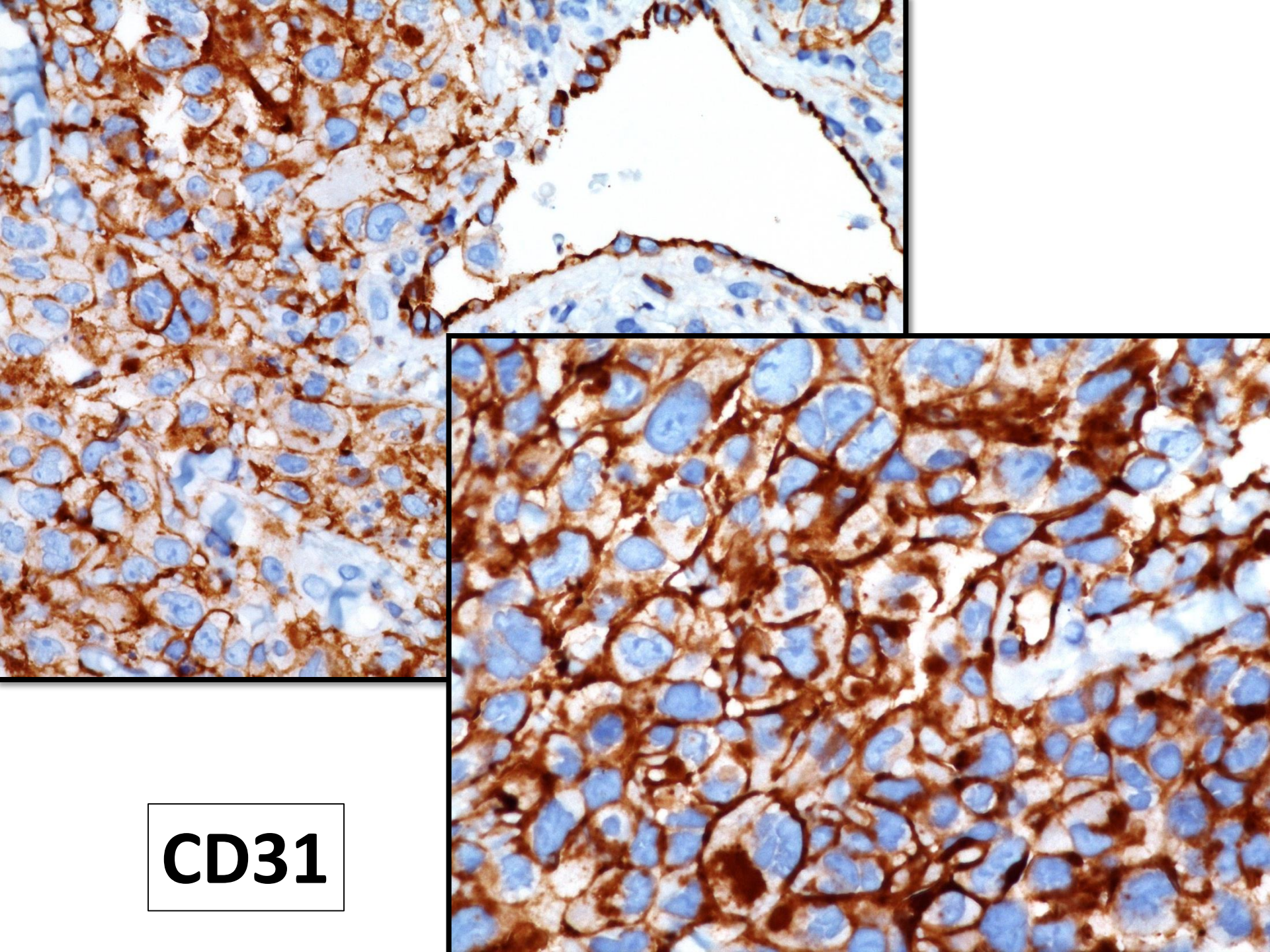
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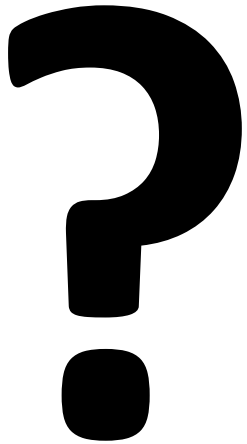


Kutánny angiosarkóm

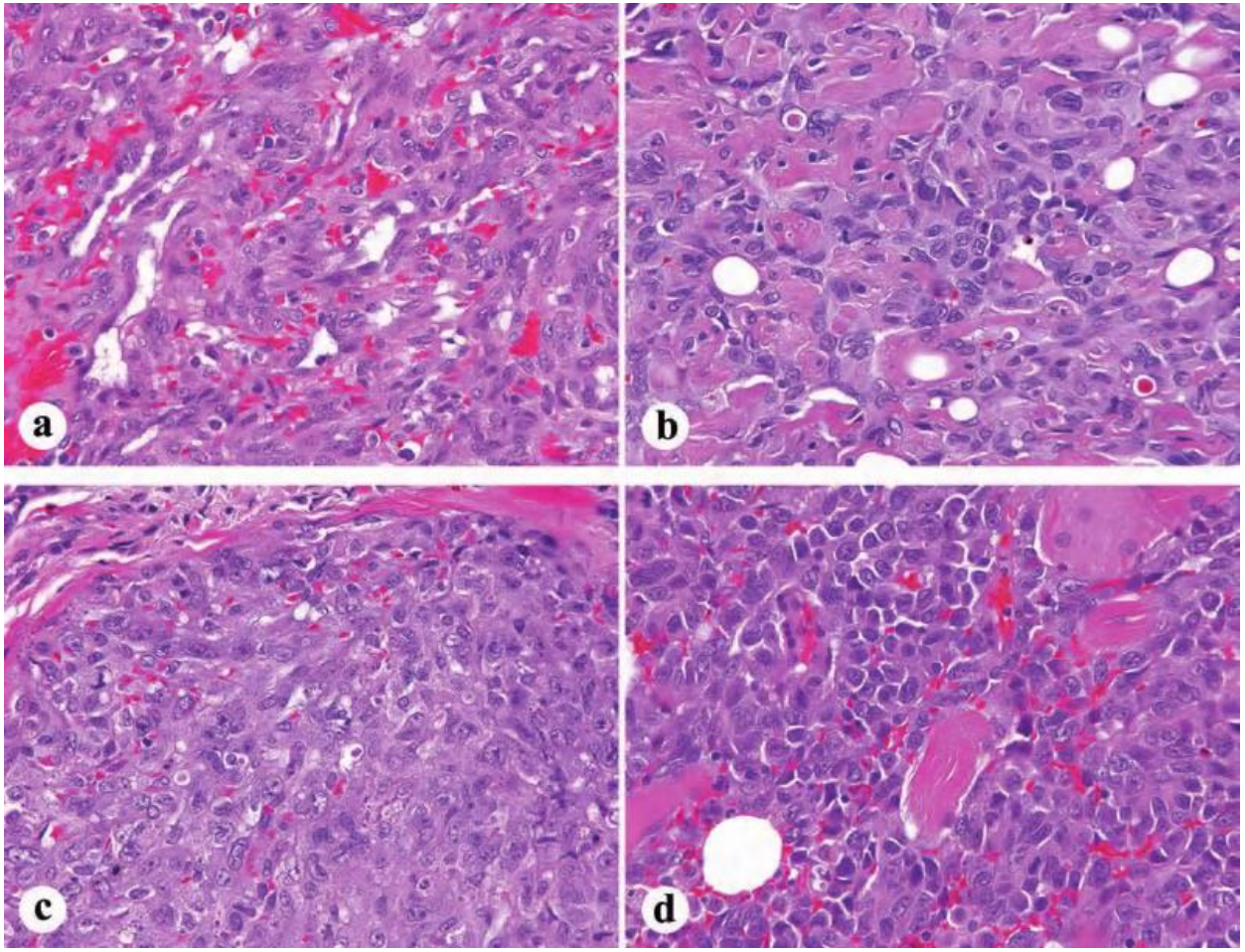
Epiteloidný/nízko diferencovaný

M-9120/3

Poorly differentiated cutaneous AS

Modern Soft Tissue Pathology

Miettinen



ANGIOSARKÓM

- Malígny endotelový novotvar
- < 1 % vaskulárnych novotvarov
- 1 – 2 % všetkých sarkómov

- Tendencia k multifokalite – nie sú zahrnuté v stagingovom systéme sarkómov

Angiosarkóm (AS) – lokalita

(Enzinger 2014; Ann Surg 2010)

- Kutánny (49,6 %)
- Mammárny parenchým (14,4 %)
- Mäkké tkanivá (11,2 %)
- Srdce (6,7 %)
- Kostný (4,1 %)
- Iné lokality (14 %)

Klinické podtypy angiosarkómu

(Enzinger 2014)

- Primárny kutánny AS
- Sekundárny kutánny (asociovaný s lymfedémom) AS
- Poradiačný AS
- AS mammy a iných parenchymatóznych orgánov
- AS mäkkých tkanív

ETIOLÓGIA

- **Chronický lymfedém** *(Stewart Treves syndrome)*
- **Radiácia** *(po časovom odstupe po RAT v miestach bez lymfedému)*
- **Arteriovenózne fistuly** *(transplantované obličky + imunosupresia)*
- **Cudzorodý materiál**
- **Thorotrast** *(cerebrálna angiografia)*
- **Zlúčeniny arzénu** *(pesticídy vo vinohradníctve)*
- Slnečné žiarenie ?

GENETIKA

- Vaskulárne špecifické tyrozín-kinázové receptory: ***TIE1, KDR (VEGFR2), SNRK, TEK, FLT1 (VEGFR1)***
- *Radiačné **(LYN, PRKC0)** a non-radiačné **(FLT1, ACT3)** AS*
- *Aktivačné mutácie **KDR** = možná cielená terapia (Sunitinib)*
- *Amplifikácia **MYC** (poradiačné a s lymfedémom asociované AS; 50 – 100 %)*

PRIMÁRNY KUTÁNNY ANGIOSARKÓM

?

- ½ AS
- M > Ž
- 90 % belosi
- Po 70-tke
- 50 % oblasť kože hlavy a krku (skalp, horné čelo)
- Expozícia slnečnému (UV) žiareniu ???

MAKROSKOPICKÝ OBRAZ

- „Modrina“
- Noduly
- Ulcerácia
- Multifokalita
- Nejasné hranice
- Rezné plochy: hemoragické / špongiovité areály

MIKROSKOPICKÝ OBRAZ

- Väčšina kutánnych AS je dobre a stredne diferencovaných
- *Tvorba vaskulárnych priestorov*
- *Iregularita*
- *Disekcia dermálneho kolagénu a fascie*
- *Papilárne intraluminálne projekcie (papillations, tufting)*

- Minorita kutánnych AS *= high grade (vretenobunkové, epiteloidné) tumory*

- ***Foamy cell variant AS***

Cutaneous Angiosarcoma of the Scalp
A Multidisciplinary Approach
Pawlik et al.
CANCER October 15, 2003 / Volume 98 / Number 8

- **TABLE 1**
- **Patient Characteristics (*n 29 patients)***
- **Characteristic No. (%)**
- Age (yrs)
- Median 71.0
- Range 33–90
- Gender
- Female 11 (37.9)
- Male 18 (62.1)
- Follow-up (mos)
- Median 18.3
- Range 3.2–106.0
- Delay in diagnosis (mos)
- Median 5.1
- Range 0–12
- Total no. of lesions on presentation
- One lesion 17 (58.6)
- One lesion plus satellitosis 4 (13.8)
- Multifocal disease 8 (27.6)
- T classification of disease
- Initial clinical T1 18 (62.1)
- Initial clinical T2 11 (37.9)
- Final pathologic T1 7 (24.1)
- Final pathologic T2 21 (72.5)
- No pathologic T stage available (no surgery) 1 (3.4)
- **Grade of angiosarcoma**
- **Low 6 (20.7 %)**
- **High 19 (65.6 %)**
- Unknown (no surgery) 1 (3.4)
- Not available for review 3 (10.3)

GRADING

STANFORD (AS of the breast)

- **Low grade tumors**
  - **Prominent freely anastomosing vascular channels**
  - Papillary growth and endothelial tufting minimal to absent
  - **Cytologic atypia may be difficult to identify** even after extensive sampling
- **Intermediate grade tumors**
  - Freely anastomosing vascular channels
  - **Papillary growth and endothelial tufting**
  - May have focal solid areas with polygonal or spindle cells
- **High grade tumors**
  - **Prominent solid areas of clearly malignant cells**
    - Polygonal and spindled cells
  - **Blood lakes and necrosis are common**
  - **Vascular channels may be difficult to identify**
    - May require extensive sampling, especially at edge of lesion
  - Demonstration of vascular nature may require immunohistology

IHC

- CD31
- CD34
- FLI1
- ERG (senzitívny vaskulárny marker nezávislý od stupňa diferenciácie / Miettinen)
- Claudin- 5

- CD117 (**50-60 % AS**; expresia ako u fetálnych endotélií; Miettinen – bez KIT mutácie exónov 11 a17, bez efektu liečby TKI)
- CK7, CK8, EMA (!)

Kutánny AS
diferenciálna diagnostika

Non - vaskulárne lézie

- Melanoma
- Lymphoma
- Carcinoma (aj MTS RCC)
- ...sarcoma

Vaskulárne lézie

- Hemangioma *(hobnail, spindle cell, angiomatosis, papillary endothelial hyperplasia - Masson)*
- Kaposi sarcoma
- Hemangioendothelioma

  (epitheloid, Kaposiform, hobnail – Dabska+retiform,epitheloid sarcoma-like / pseudomyogenic, composite)
- AVL

Kutánny AS
Prognóza

- Agresívny novotvar
- Časté vzdialené MTS
- 50 % pacientov umiera do 15 mesiacov
- 15 % pacientov prežíva > 5 rokov

Prognostické faktory AS
(všetky lokality)

FAVORABLE	UNFAVORABLE
< 50 rokov	> 50 rokov
Trup	Hlava a krk
Unifokalita	Multifokalita
Lokalizovaný proces	Šírenie do okolia
< 5 cm	> 5 cm
Povrchová lokalizácia	Hlboké mäkké tkanivá / orgány
Negatívne resekčné okraje	Pozitívne resekčné okraje
Nonepiteloidná histológia	Epiteloidná histológia
Absencia nekrózy	Prezencia nekrózy

FOLLOW UP

- Bioptický nález
- Onkológia – hospitalizácia, nález pľúcnych MTS
- Cca 1 mesiac exitus letalis

ĎAKUJEM ZA POZORNOSŤ