TUMOR DE CÉLULAS GRANULARES MALIGNO FARÍNGEO.

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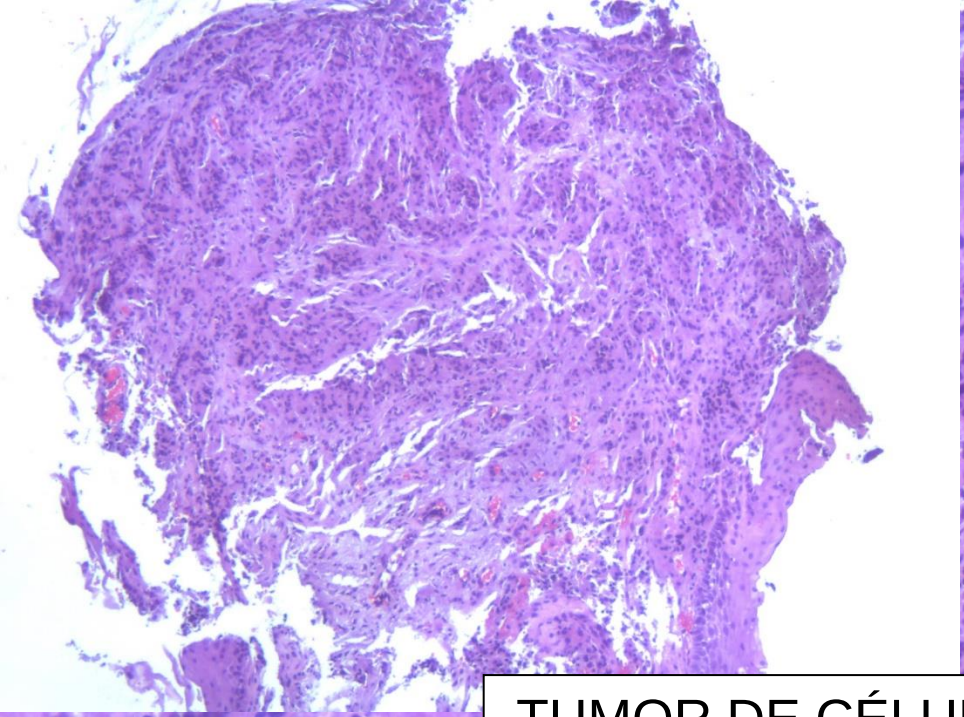
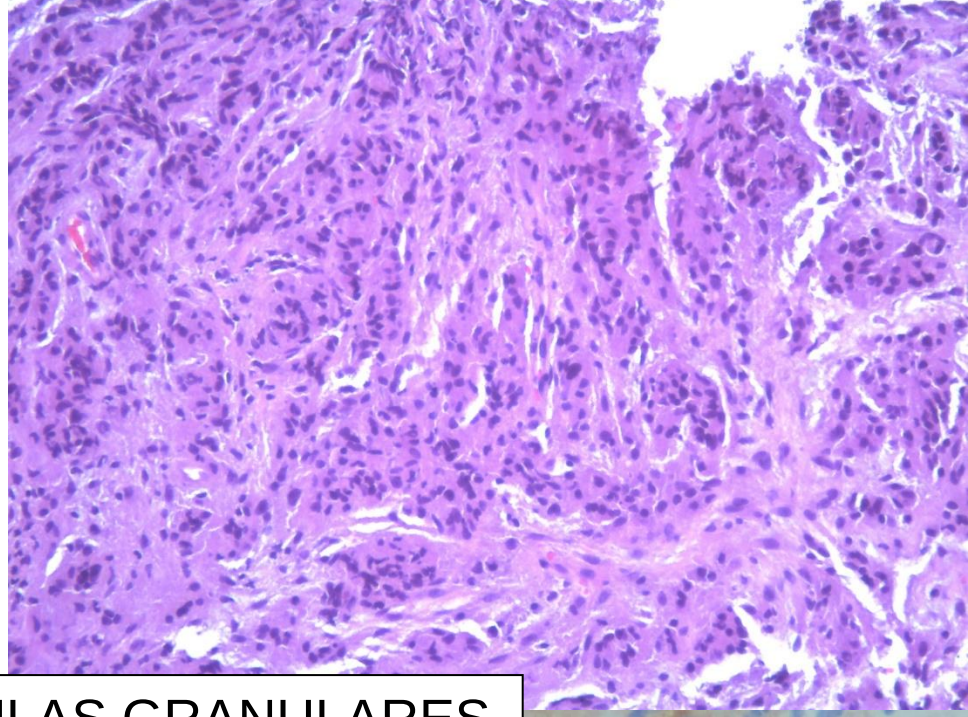
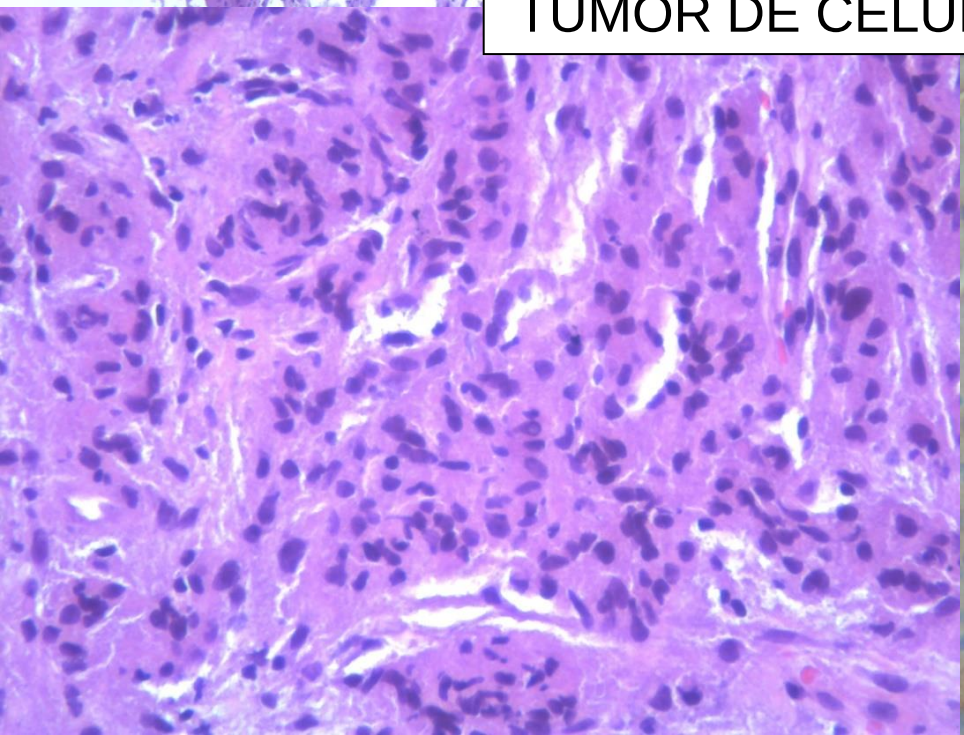
HISTORIA CLÍNICA

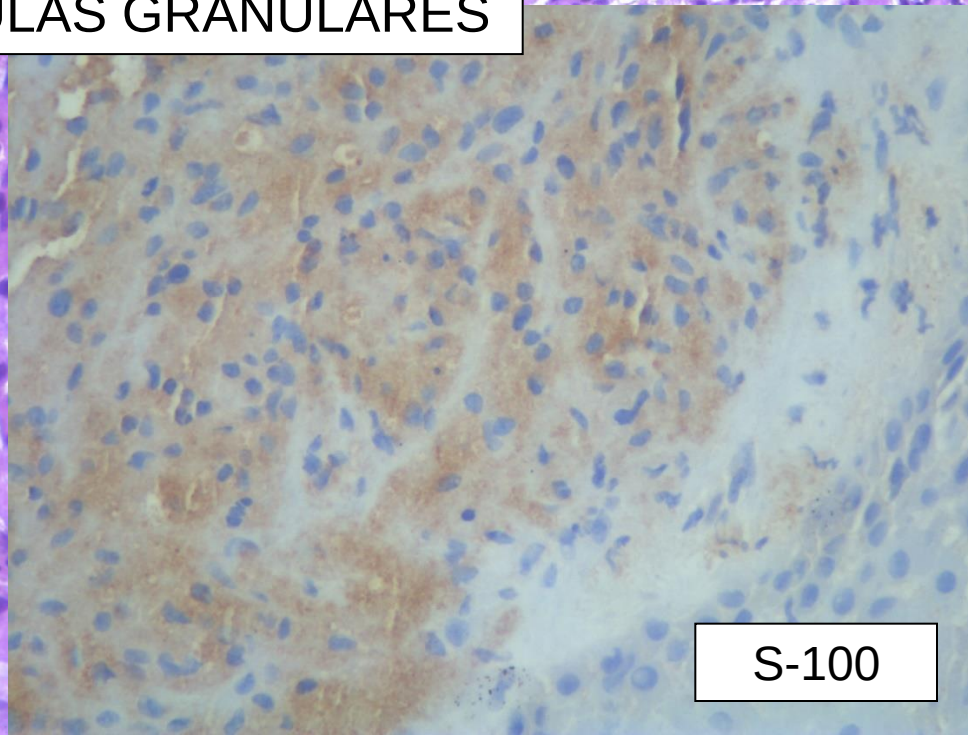
- Mujer de 44 años con **disfagia** a sólidos en estudio por digestivo de un año de evolución que progresa a líquidos, con pérdida de 20 kg de peso y desnutrición calórico-proteica.
- Endoscopia digestiva: **lesión** blanquecina sobreelevada **hipofaríngea** que ocupa seno piriforme derecho, impidiendo el paso del endoscopio hacia el esófago.
- Fibroscopia: **neoformación** excrecente en región **retrocricoidea**. Laringe normal.
- Biopsia de seno piriforme derecho.





TUMOR DE CÉLULAS GRANULARES





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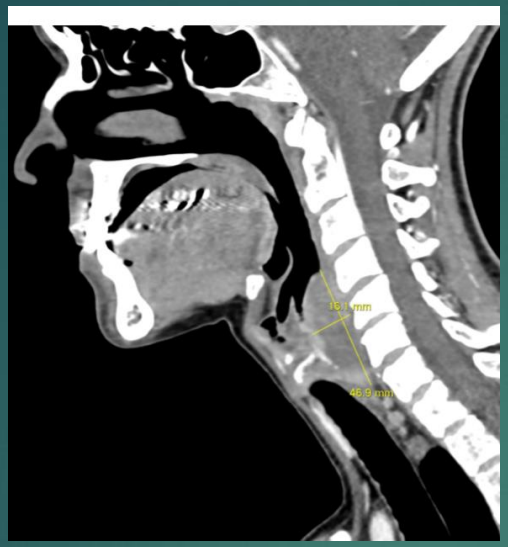

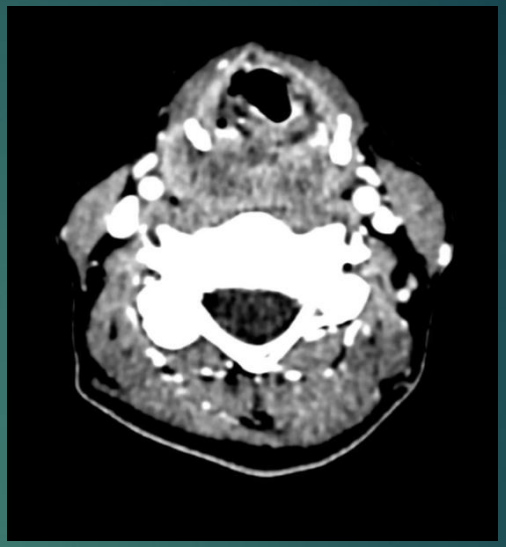

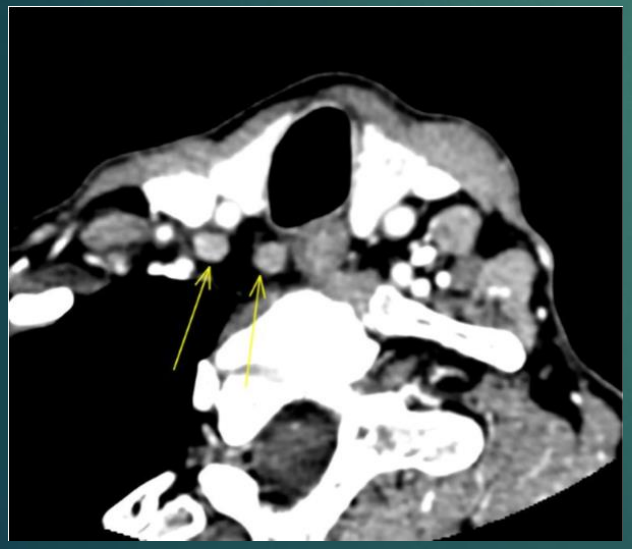

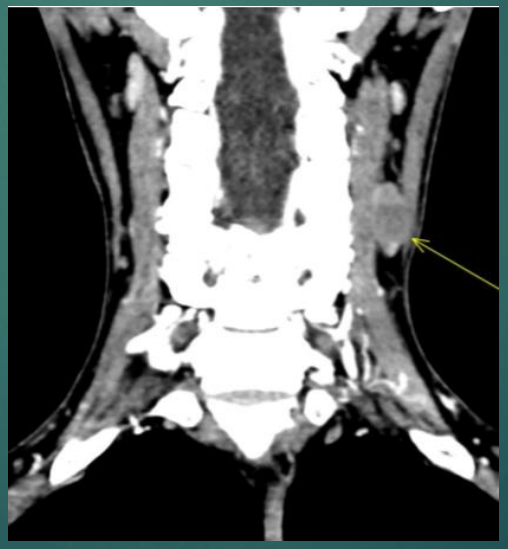

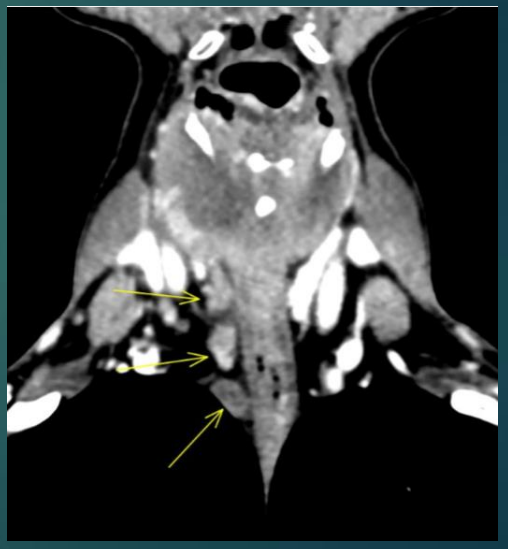

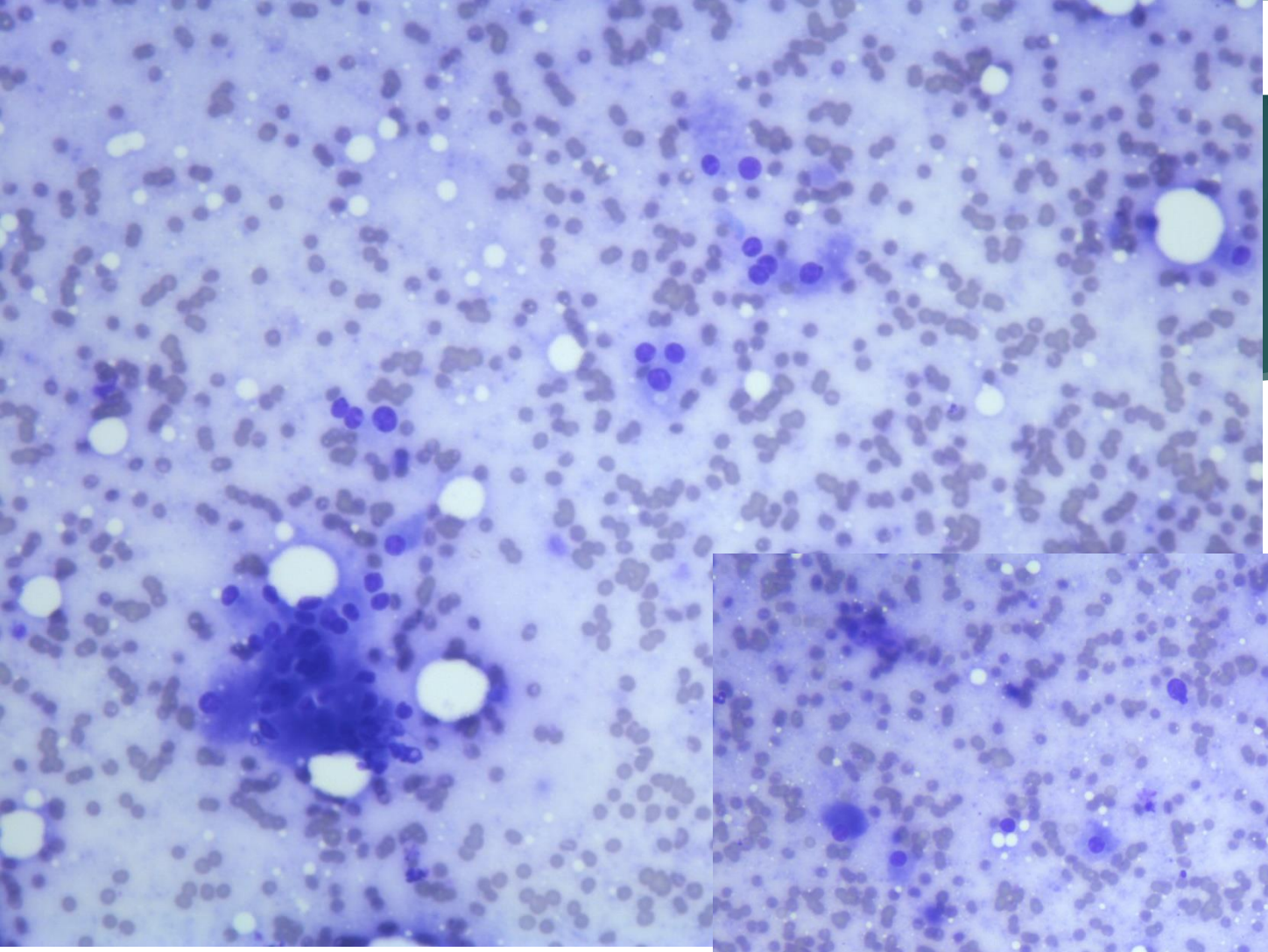

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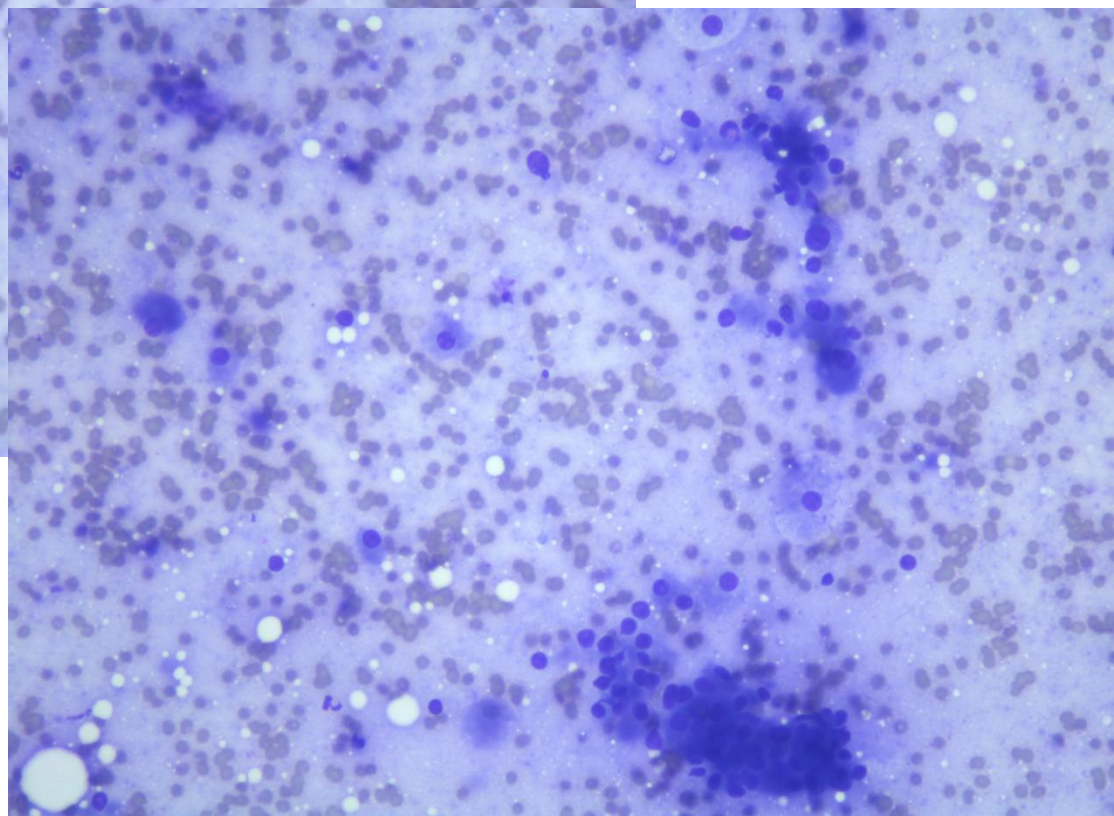
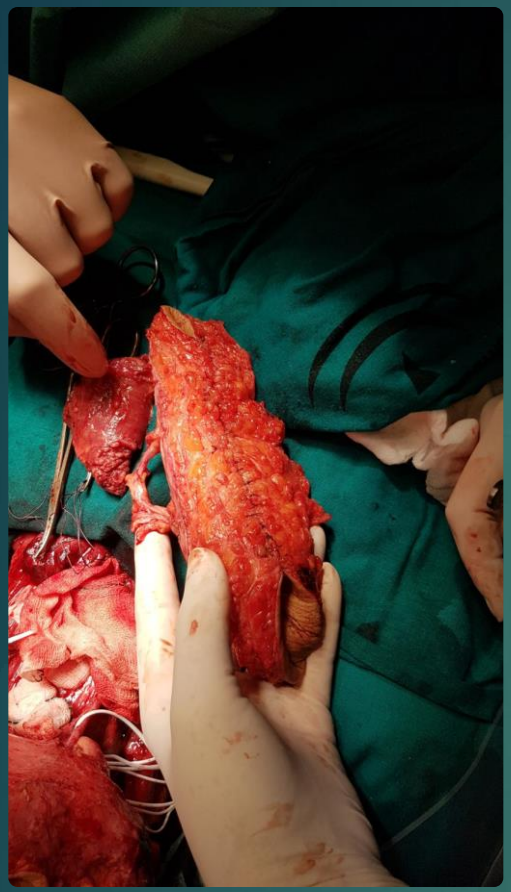
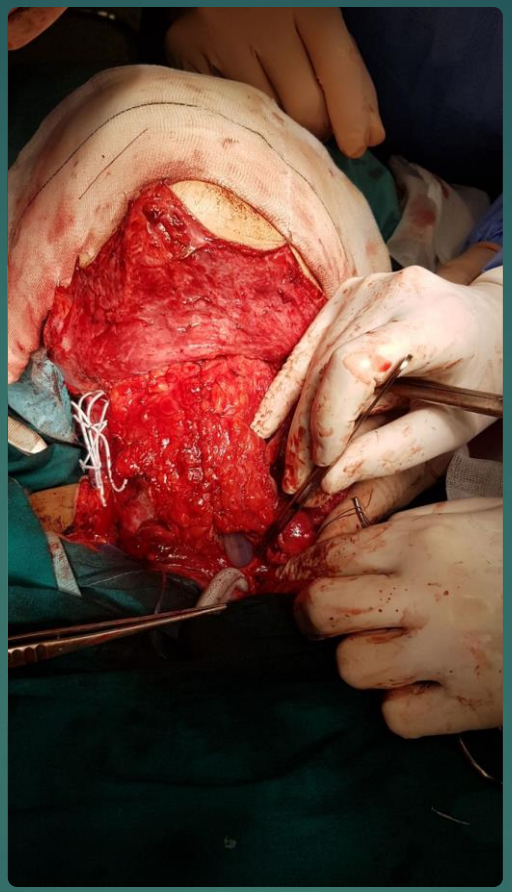
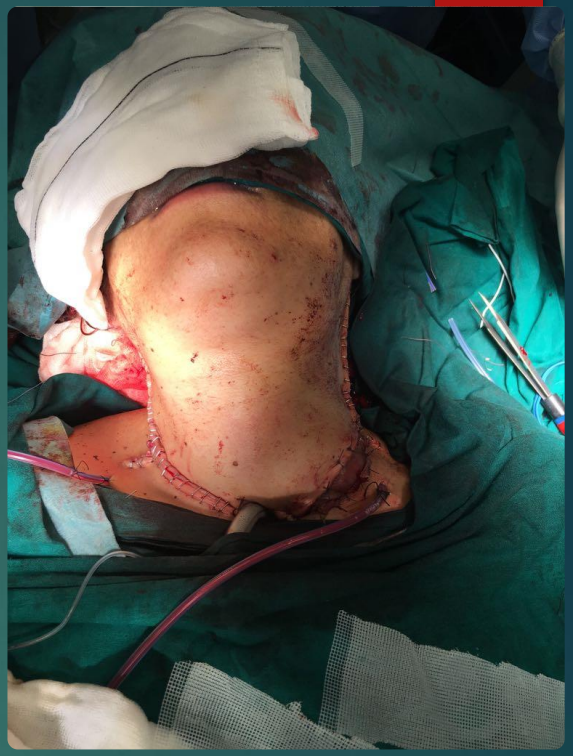


METÁSTASIS DE TUMOR DE CÉLULAS GRANULARES.

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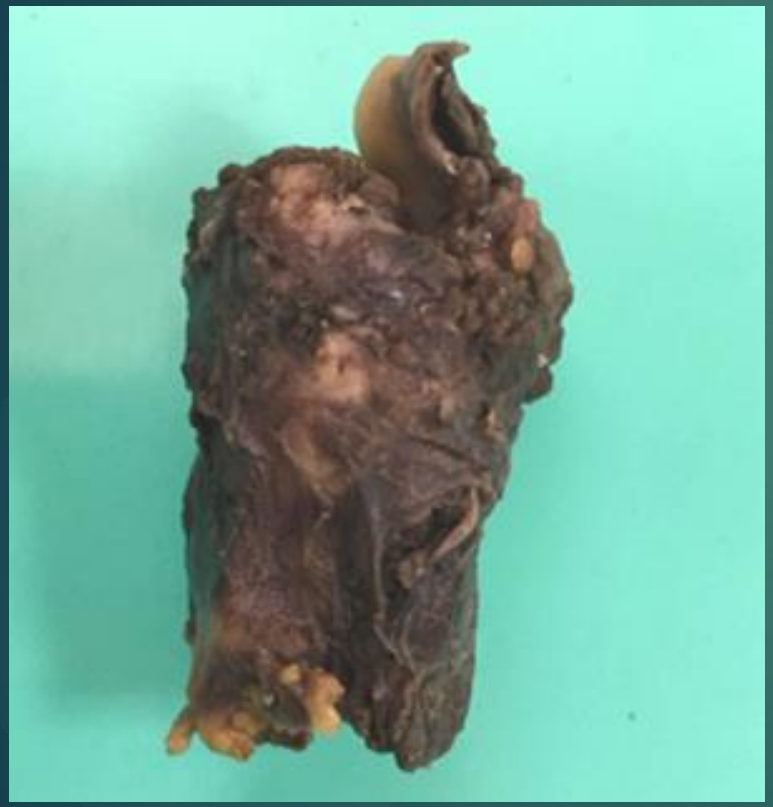
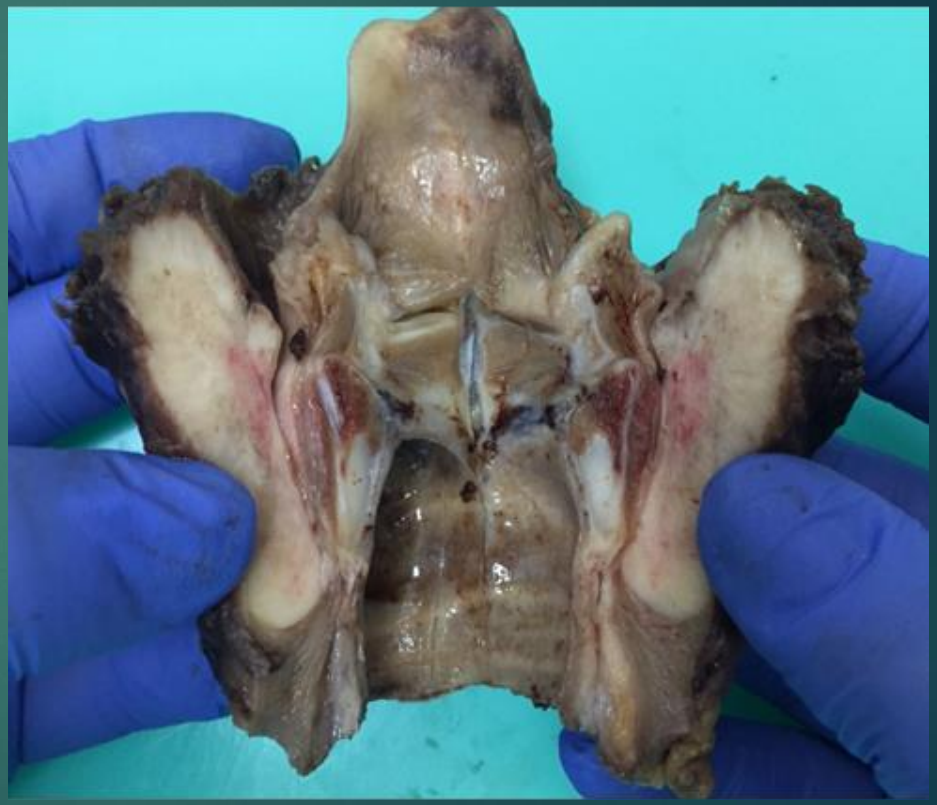
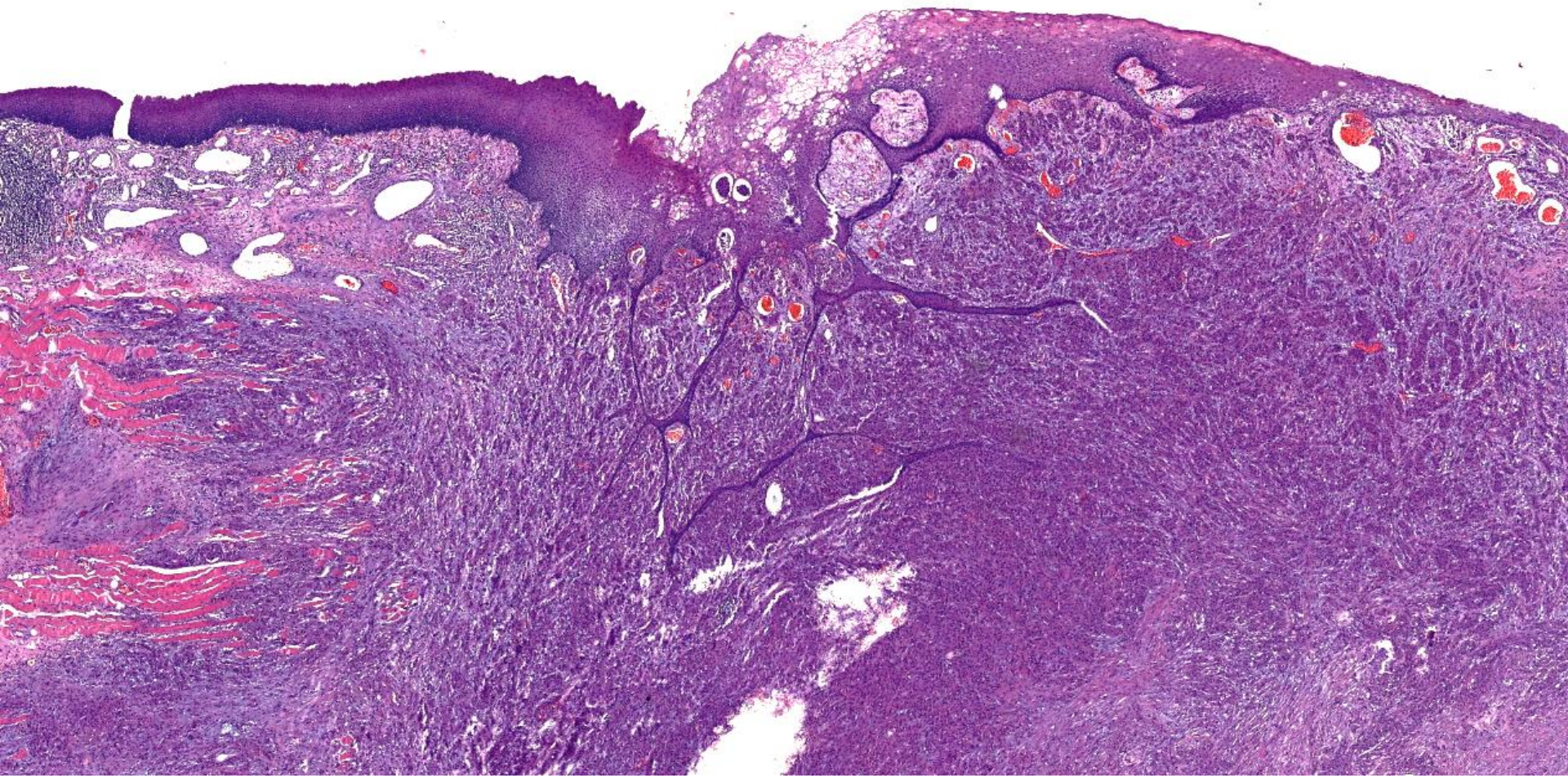
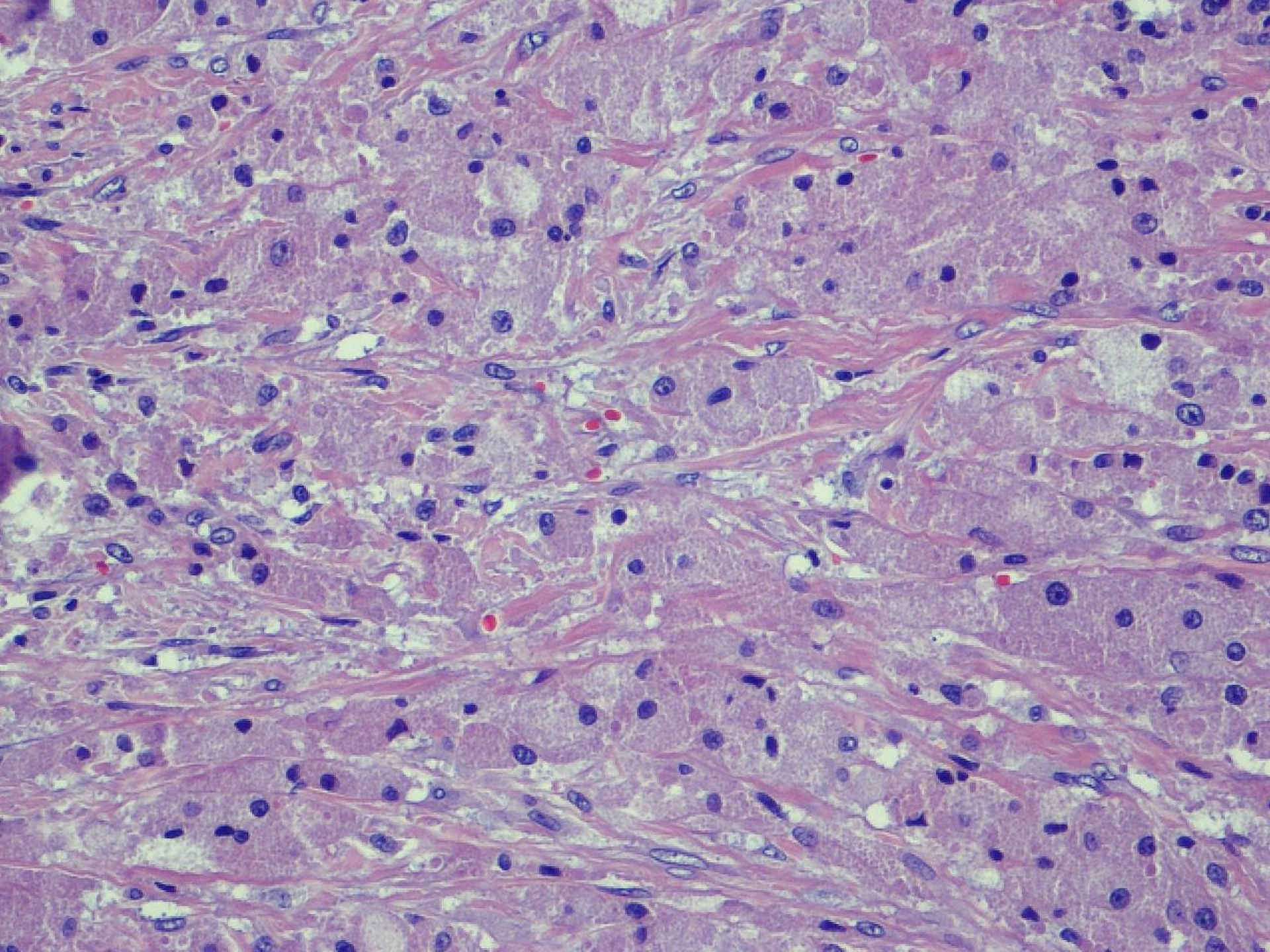
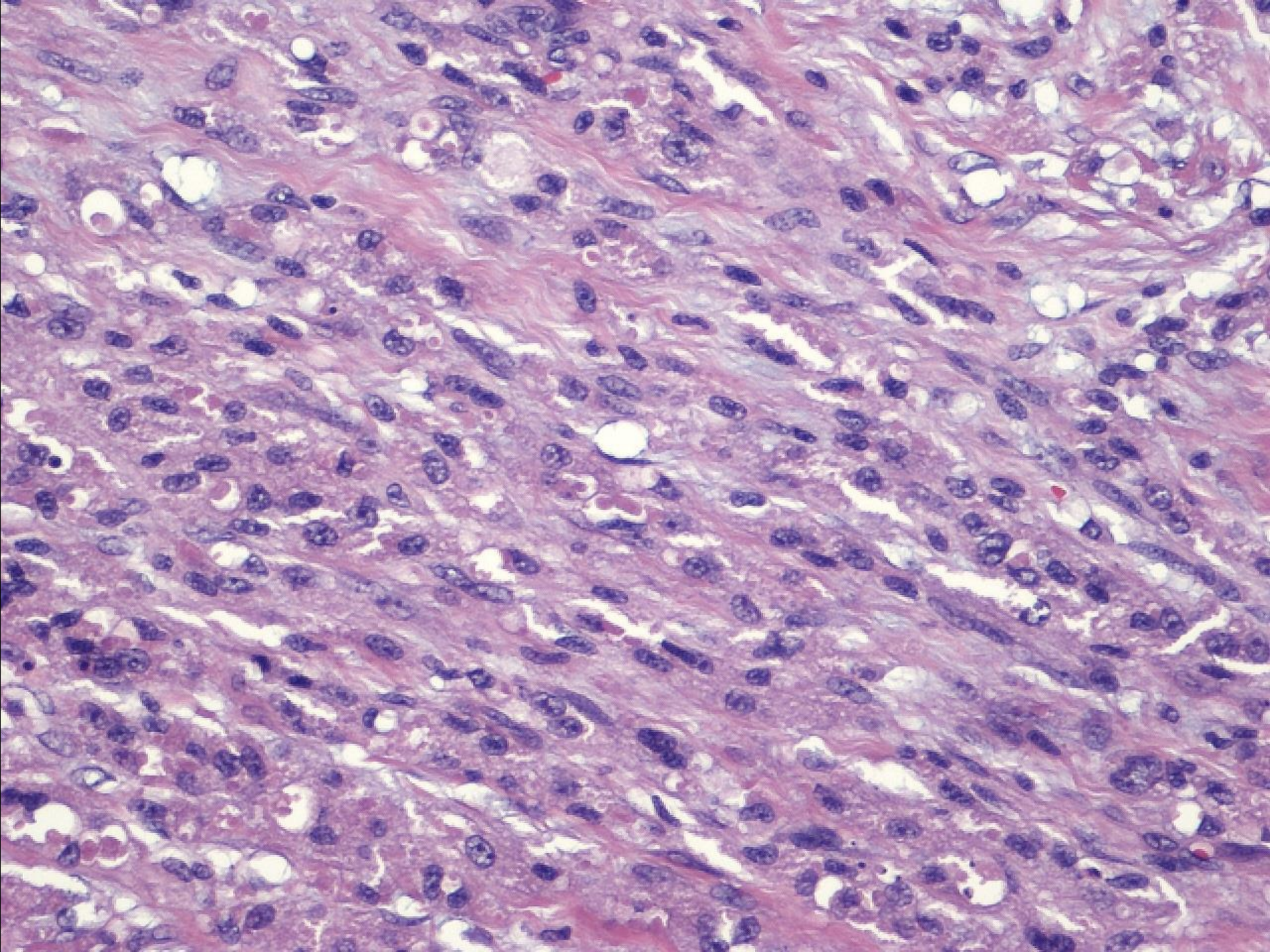
HALLAZGOS MACROSCÓPICOS

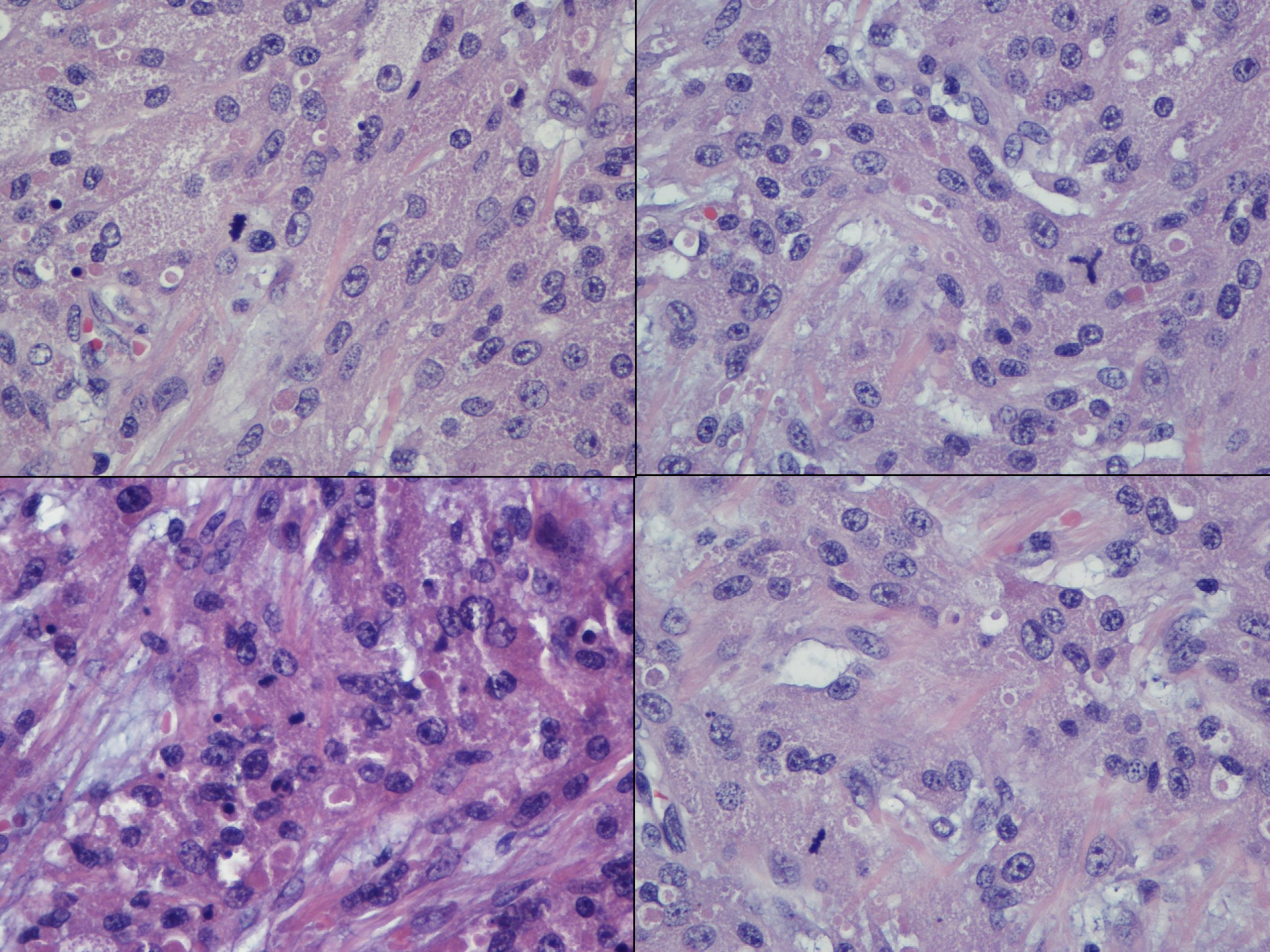


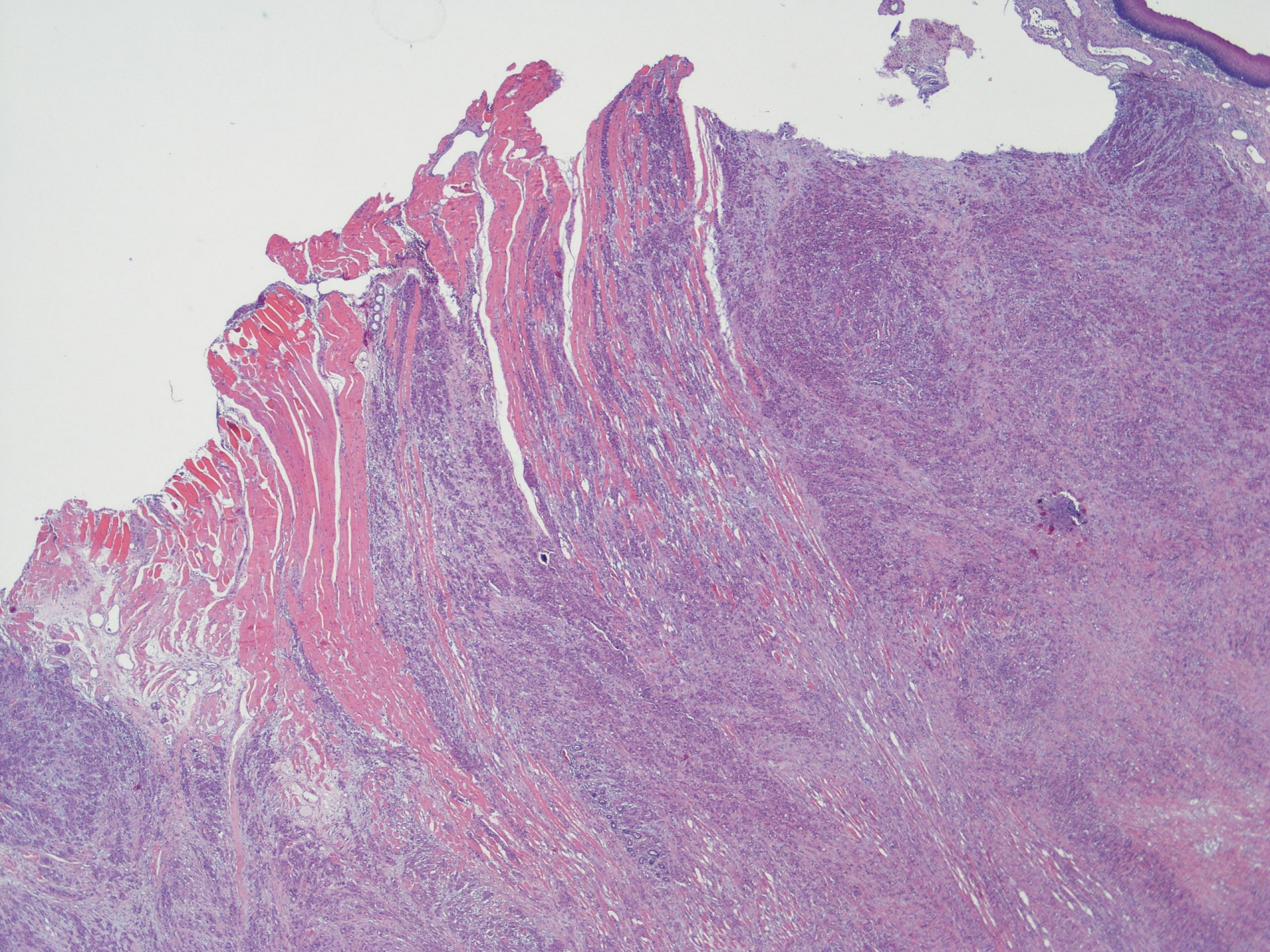


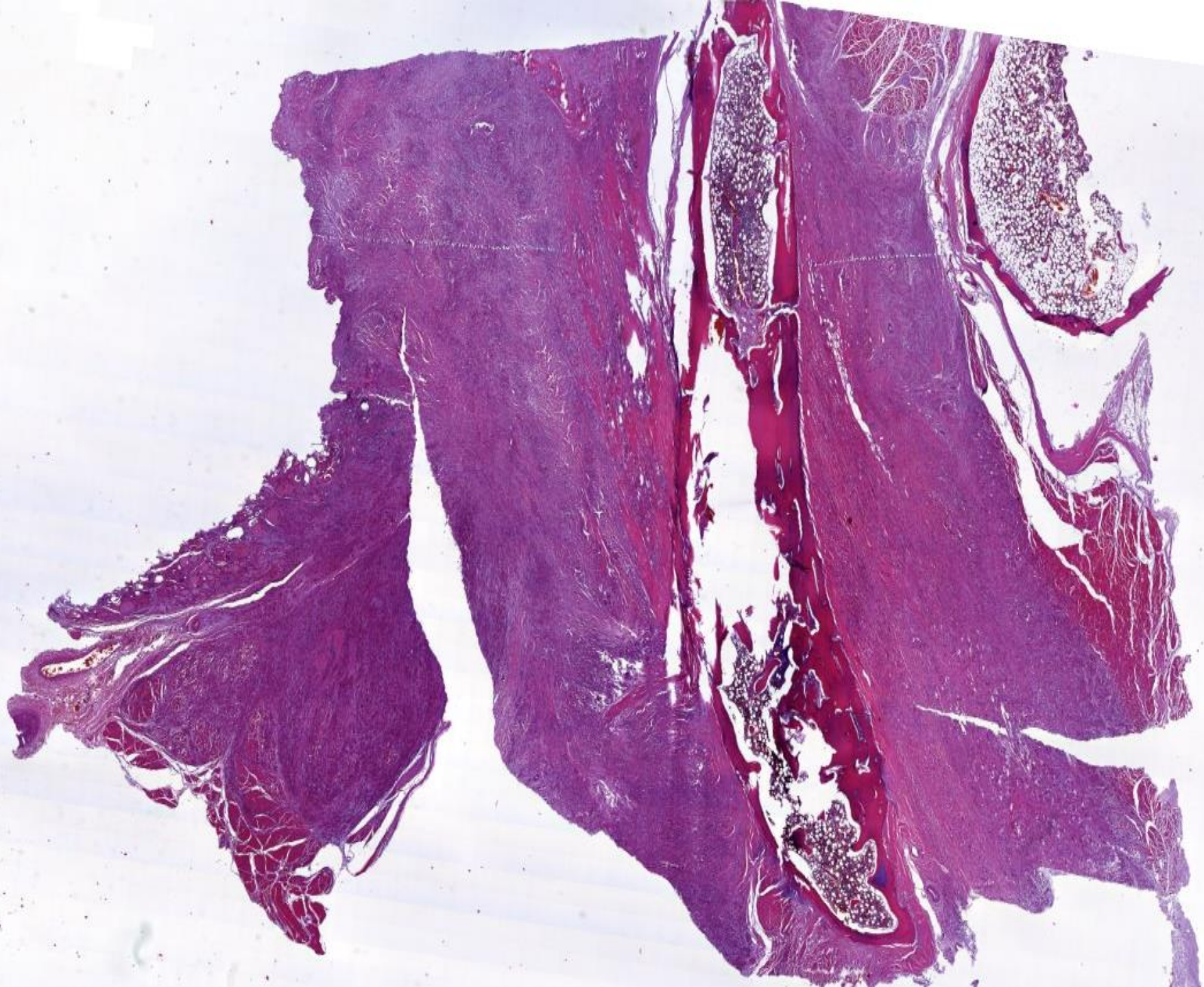


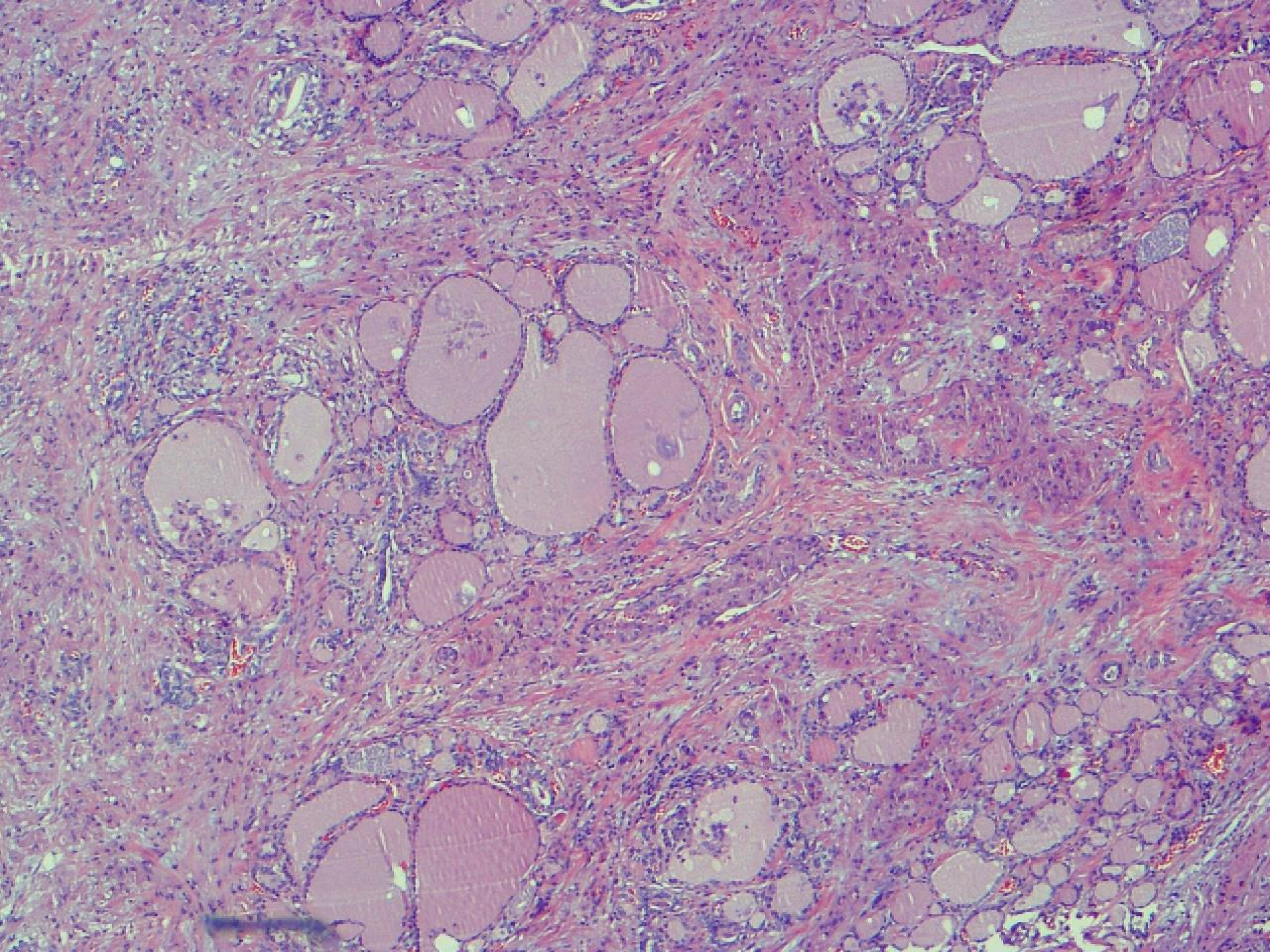


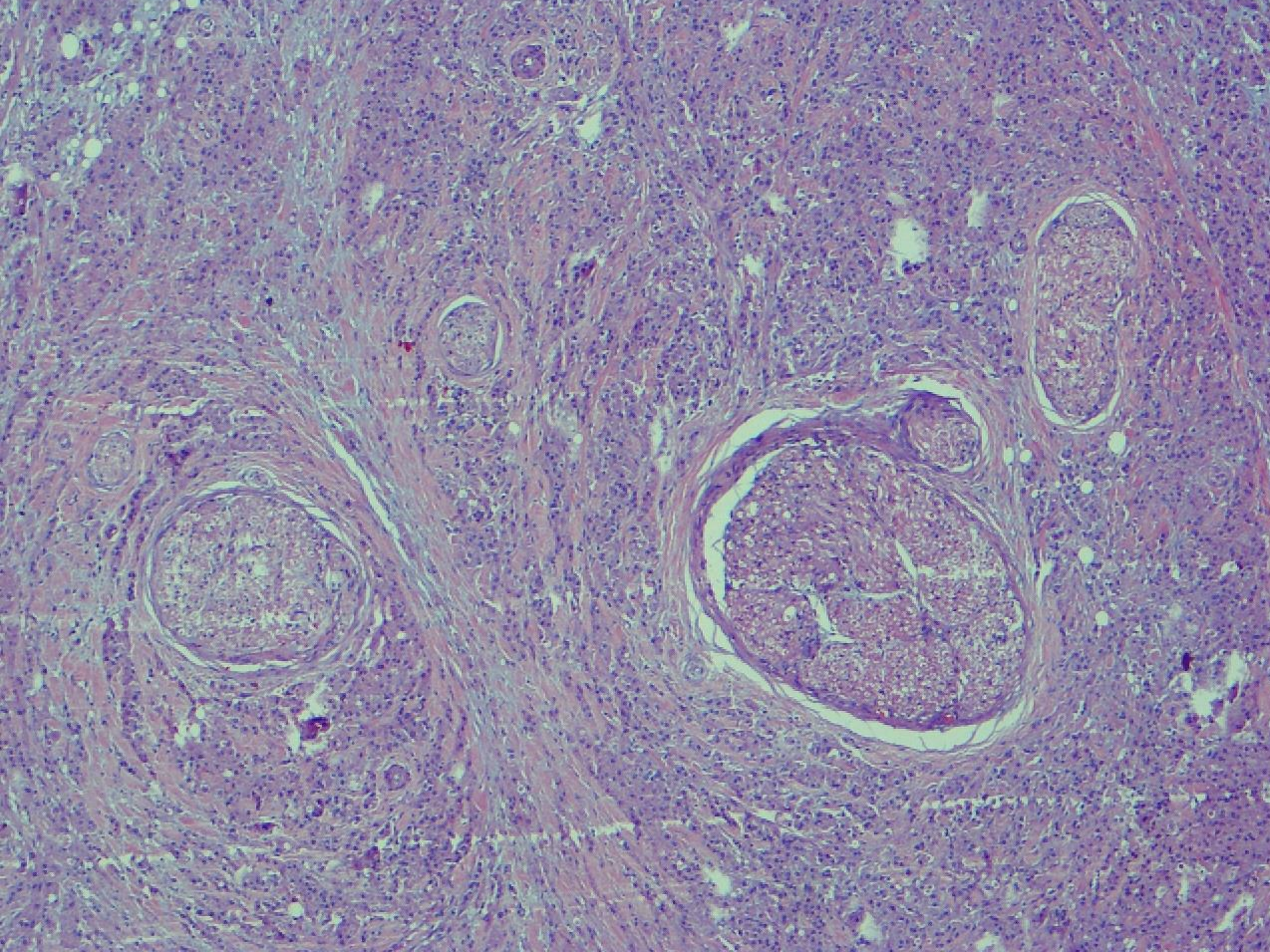


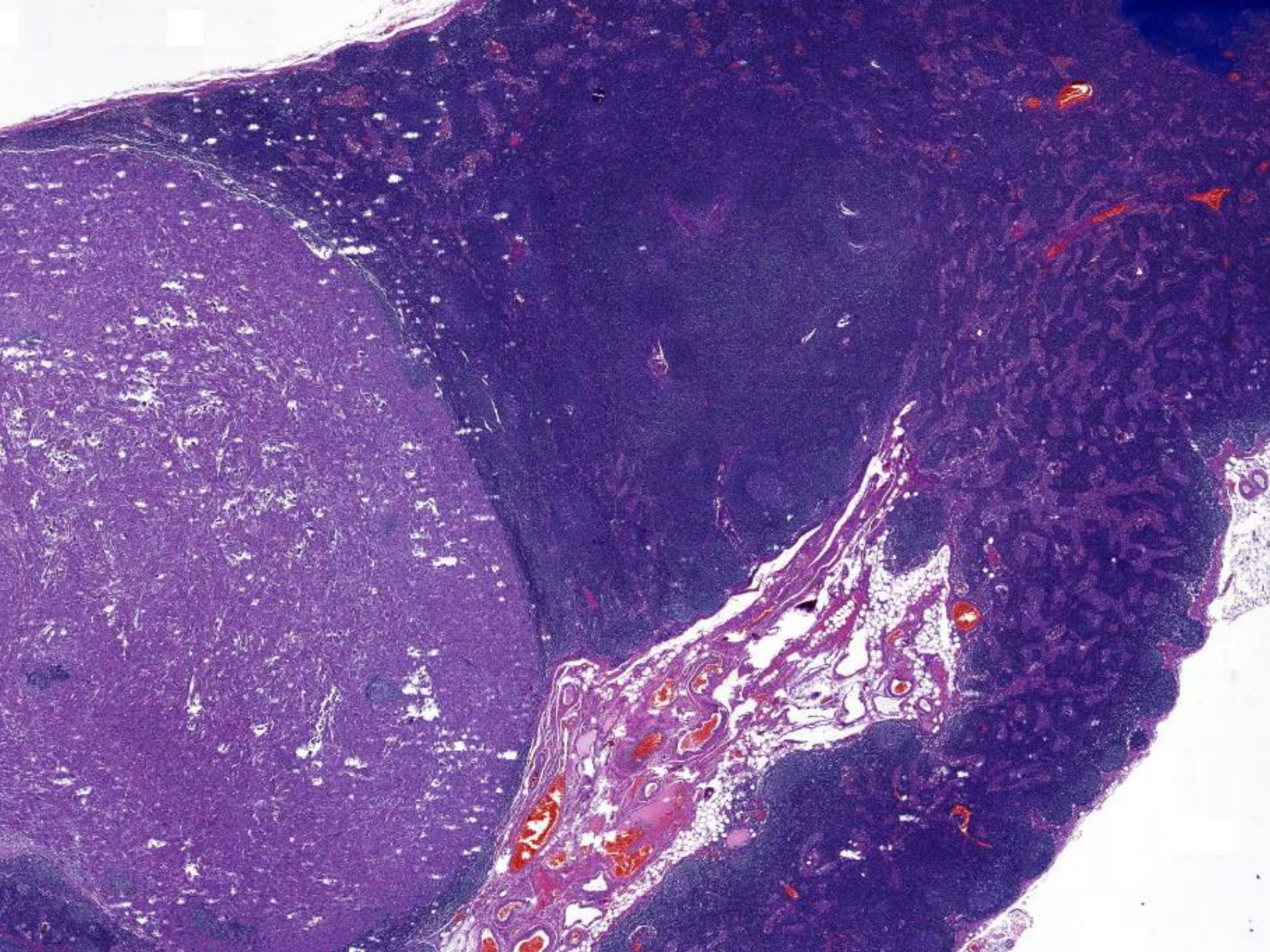


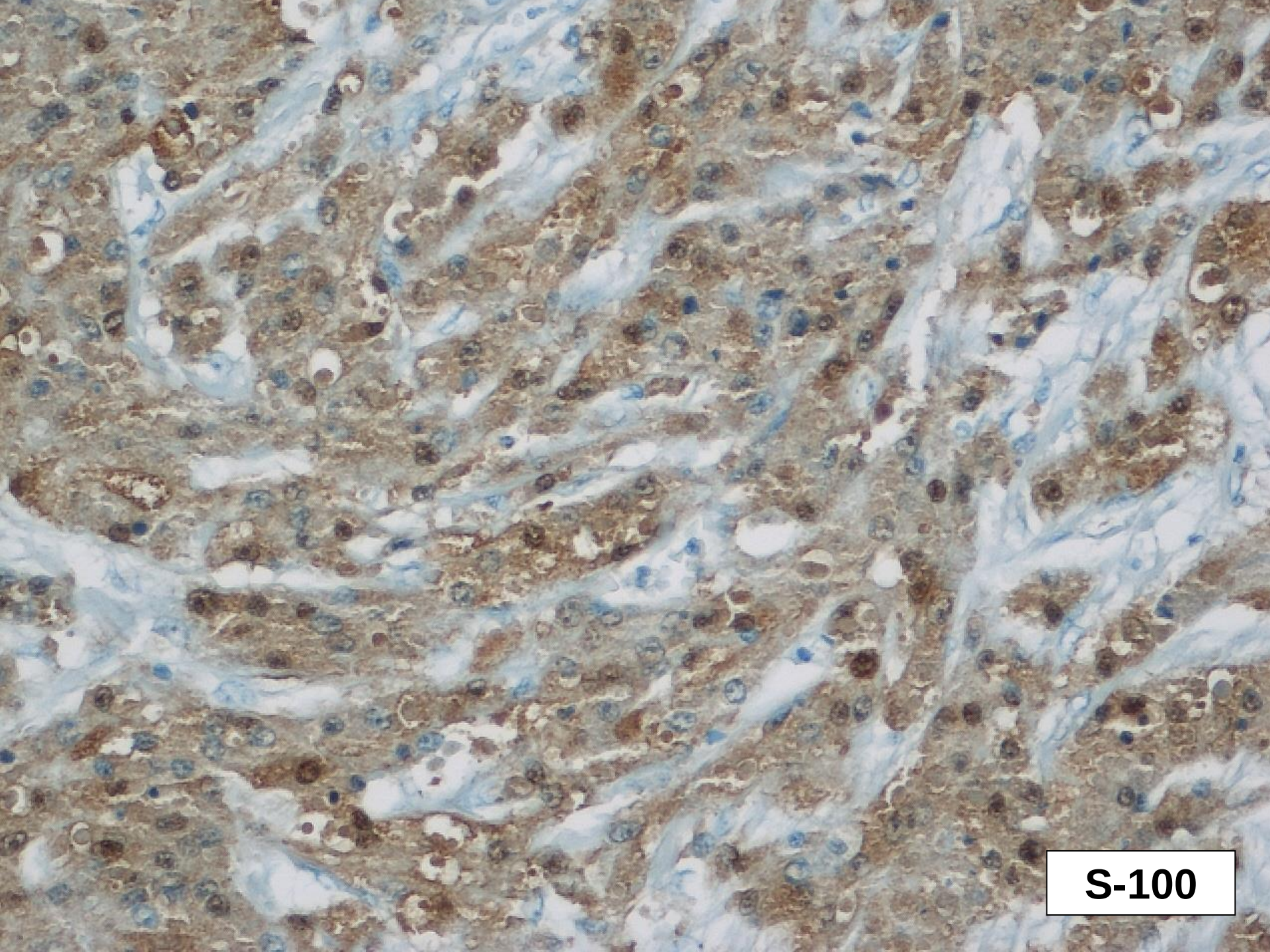








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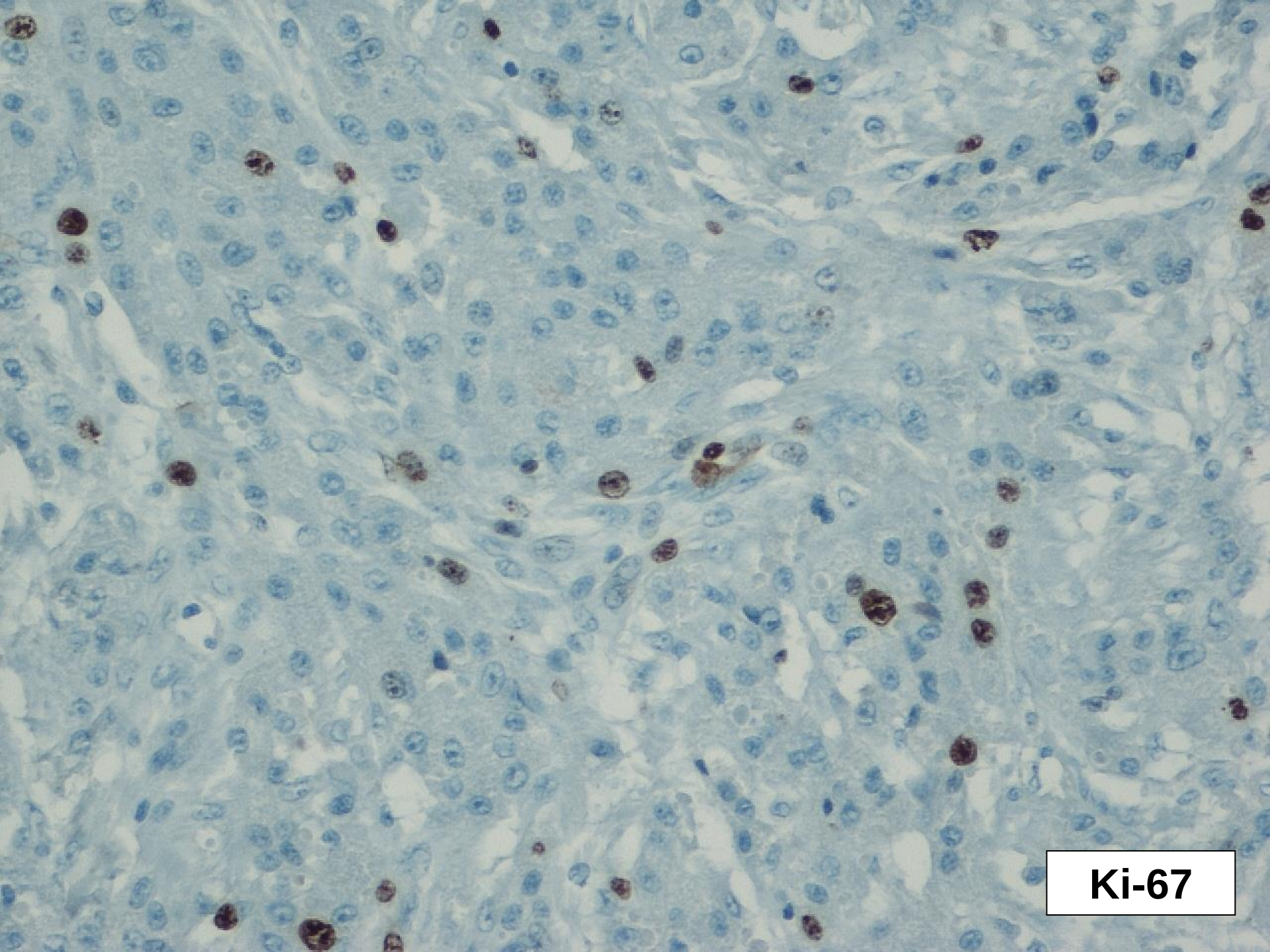

Ki-67

TUMOR DE CÉLULAS GRANULARES MALIGNO

- **Raro** (0,5-2% de todos TCG).
- OMS: Raro **sarcoma** de alto grado con fenotipo schwanniano, compuesto de células granulares malignas con inclusiones lisosomales citoplasmáticas.
- Alta tasa de **recurrencia y metástasis**.
- Frecuente en tejidos blandos de las **ee. y tronco**. Menos frecuente en cabeza y cuello.
- ♀ > ♂ . 3-70 años (40 a).
- Indice de mortalidad **40%**.

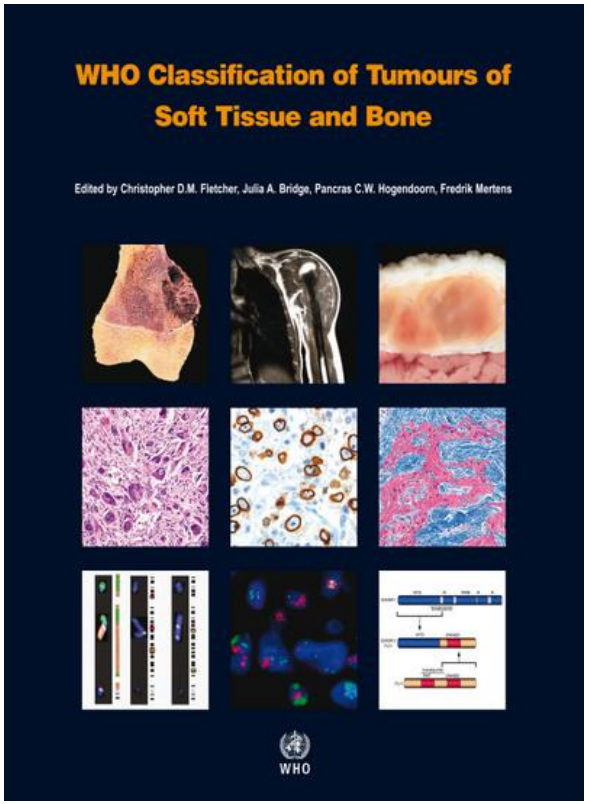
TUMOR DE CÉLULAS GRANULARES MALIGNO

- Hallazgos macroscópicos:
  - **Mayores** (> 5 cm).
  - Masas sólidas, firmes, blanquecina-grisáceas.
  - Más **profundo** (subcutáneas/intramusculares).
- Hallazgos microscópicos:
  - Morfología celular poligonal o **fusiforme** de alto grado con citoplasma granular eosinofílico.
  - A veces características **citológicas similares** a las de su contraparte benigna.

CRITERIOS DE MALIGNIDAD

- Morfología sarcomatoide.
- Núcleo vesicular con nucleolo prominente.
- > 2 mitosis/10 CGA.
- Necrosis geográfica.
- Marcado pleomorfismo.
- Alto índice núcleo/citoplasma.

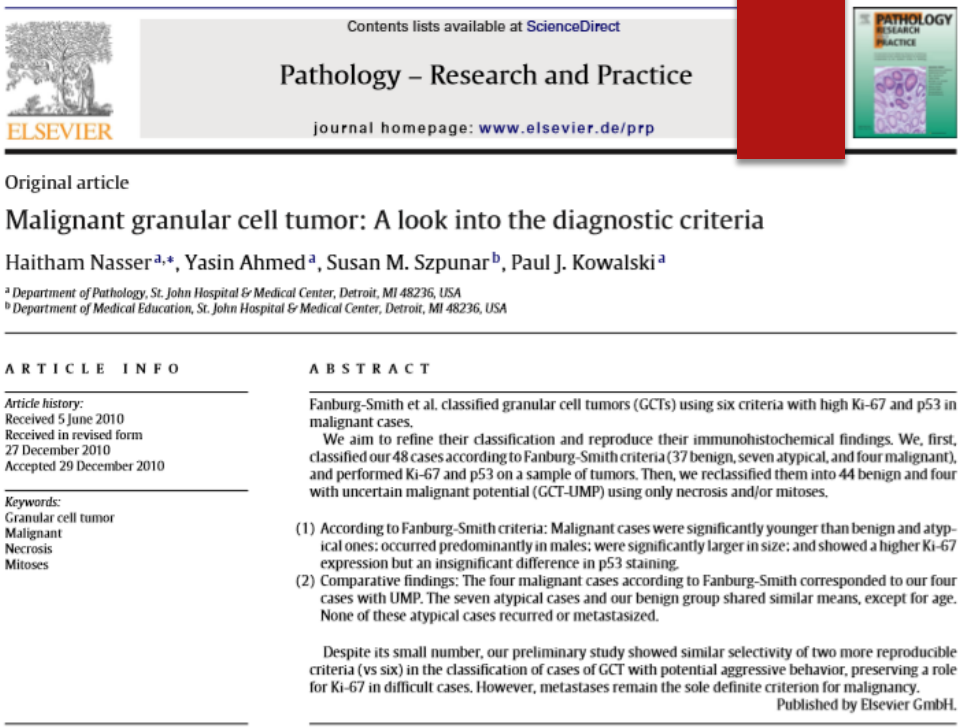
- Estos parámetros **por sí solos no pueden identificar todos los tumores que metastatizan.**



CRITERIOS DE MALIGNIDAD.

- **> 2 mitosis/10 CGA.**
- **Necrosis**.

- TCG BENIGNOS: 0 criterios.
- TCG- UMP: al menos 1 criterio.
- TCG MALIGNO: existencia de **metastasis**.



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Original article

Malignant granular cell tumor: A look into the diagnostic criteria

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ABSTRACT

Fanburg-Smith et al. classified granular cell tumors (GCTs) using six criteria with high Ki-67 and p53 in malignant cases.

We aim to refine their classification and reproduce their immunohistochemical findings. We, first, classified our 48 cases according to Fanburg-Smith criteria (37 benign, seven atypical, and four malignant), and performed Ki-67 and p53 on a sample of tumors. Then, we reclassified them into 44 benign and four with uncertain malignant potential (GCT-UMP) using only necrosis and/or mitoses.

(1) According to Fanburg-Smith criteria: Malignant cases were significantly younger than benign and atypical ones; occurred predominantly in males; were significantly larger in size; and showed a higher Ki-67 expression but an insignificant difference in p53 staining.
(2) Comparative findings: The four malignant cases according to Fanburg-Smith corresponded to our four cases with UMP. The seven atypical cases and our benign group shared similar means, except for age. None of these atypical cases recurred or metastasized.

Despite its small number, our preliminary study showed similar selectivity of two more reproducible criteria (vs six) in the classification of cases of GCT with potential aggressive behavior, preserving a role for Ki-67 in difficult cases. However, metastases remain the sole definite criterion for malignancy.

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TUMOR DE CÉLULAS GRANULARES MALIGNO

- Tratamiento:
  - Exéresis amplia + linfadenectomía regional.
  - Papel de la **QT y RT**: mayoría de artículos publicados describen **poca respuesta**.
  - Ausencia de una pauta de seguimiento específico.

CONCLUSIONES

- **Mitosis** y **necrosis** como criterios histológicos más relevantes.
- La existencia de mitosis y/o necrosis en un TCG no garantiza la malignidad del mismo, siendo precisa la **presencia de metástasis** para el diagnóstico de TCGM.
- El diagnóstico de sospecha malignidad no es posible en **biopsias pequeñas**.

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