Club de partes blandas

Mujer de 21 años con tumor retroperitoneal y pélvico

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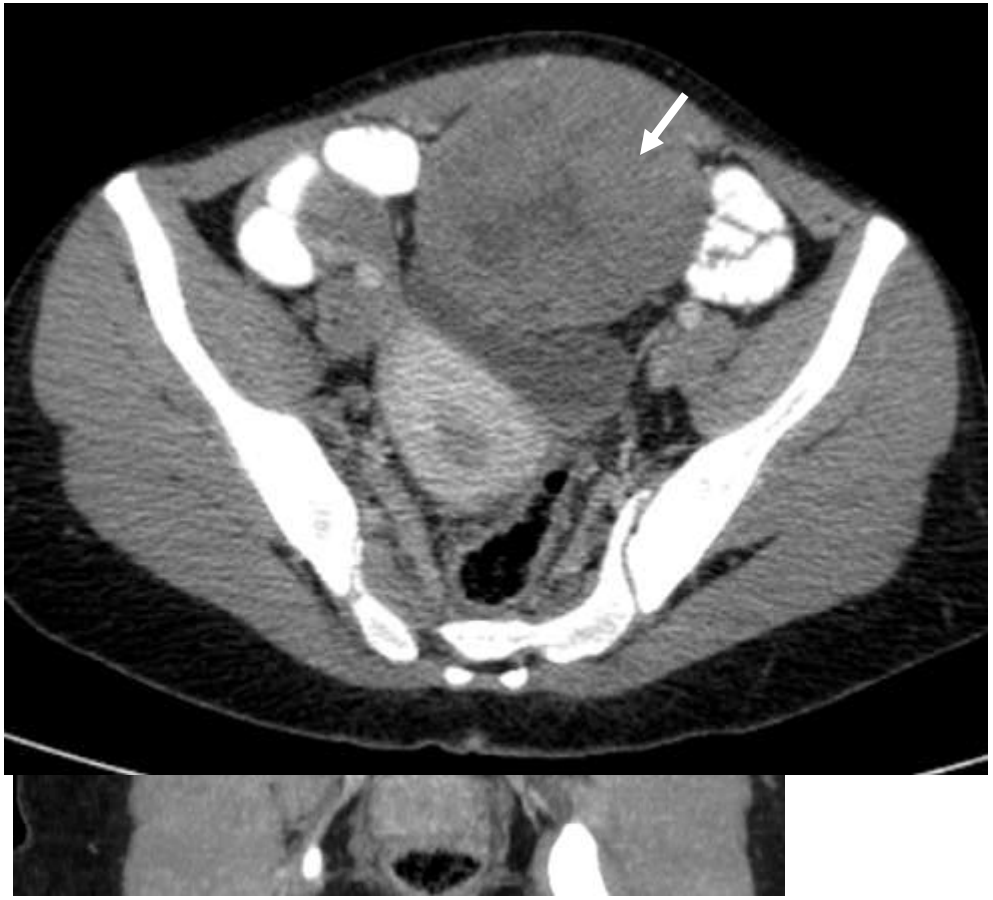
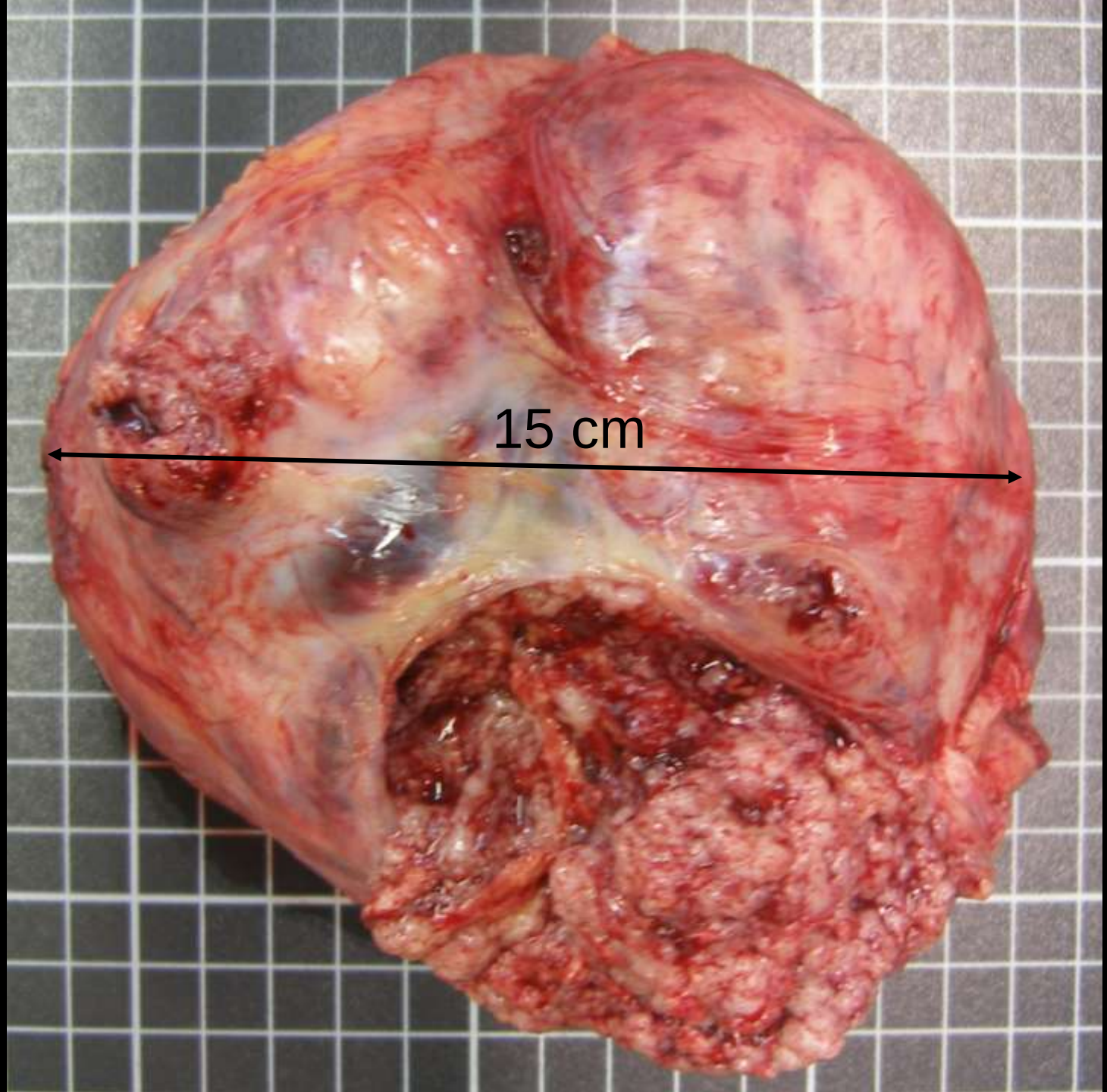
Historia clínica

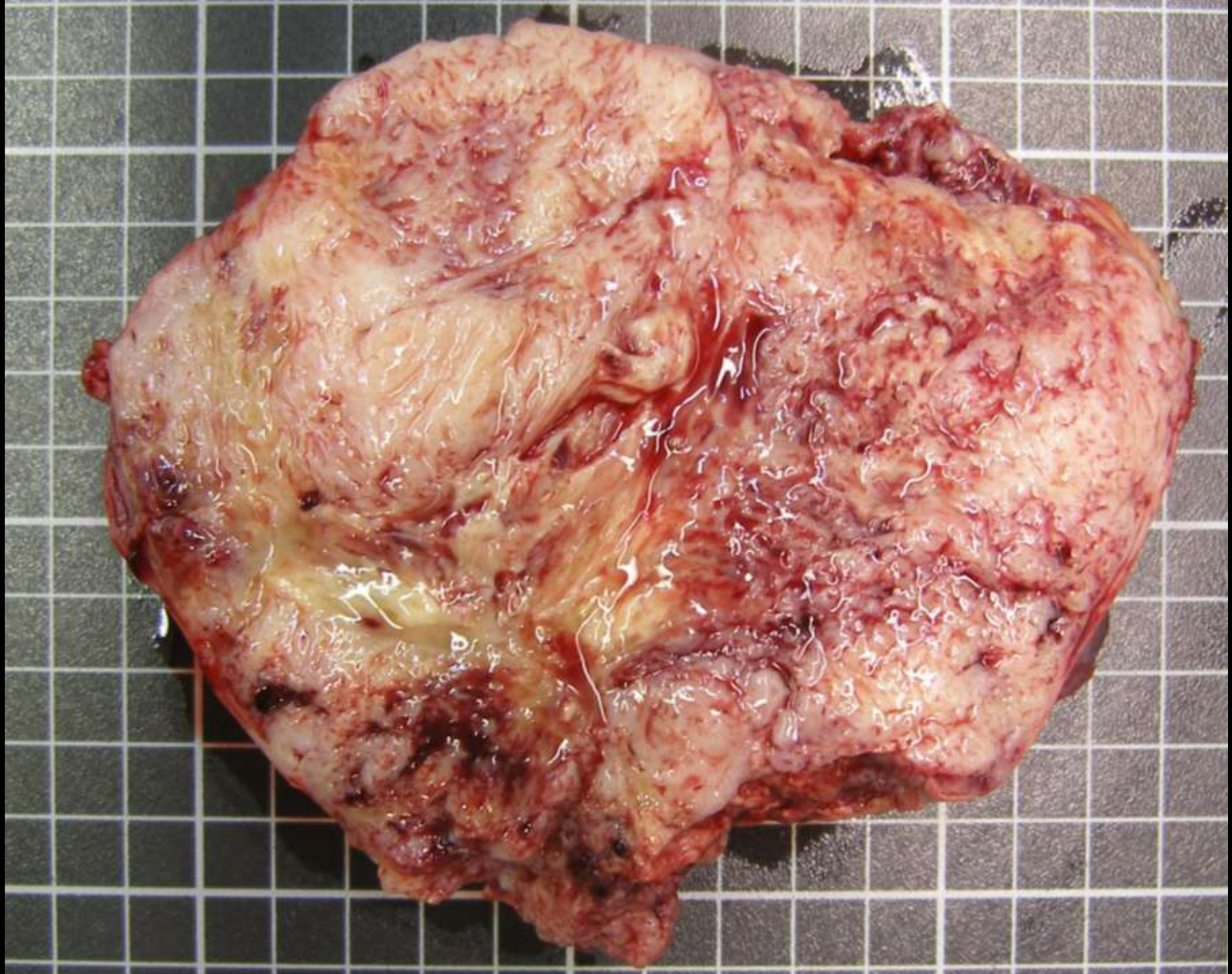


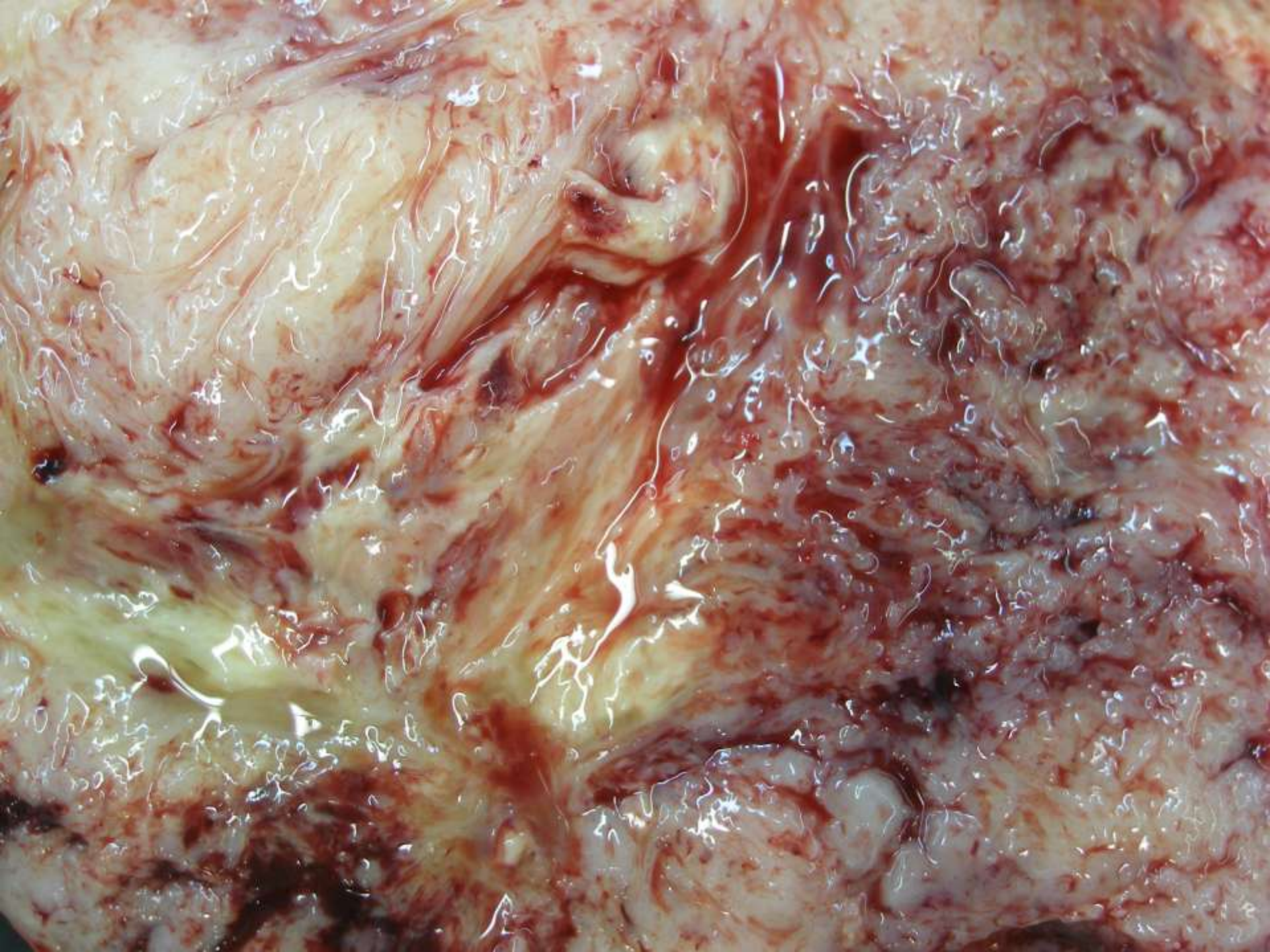
Mujer de **21 años** con antecedentes de **Neurofibromatosis tipo 1**

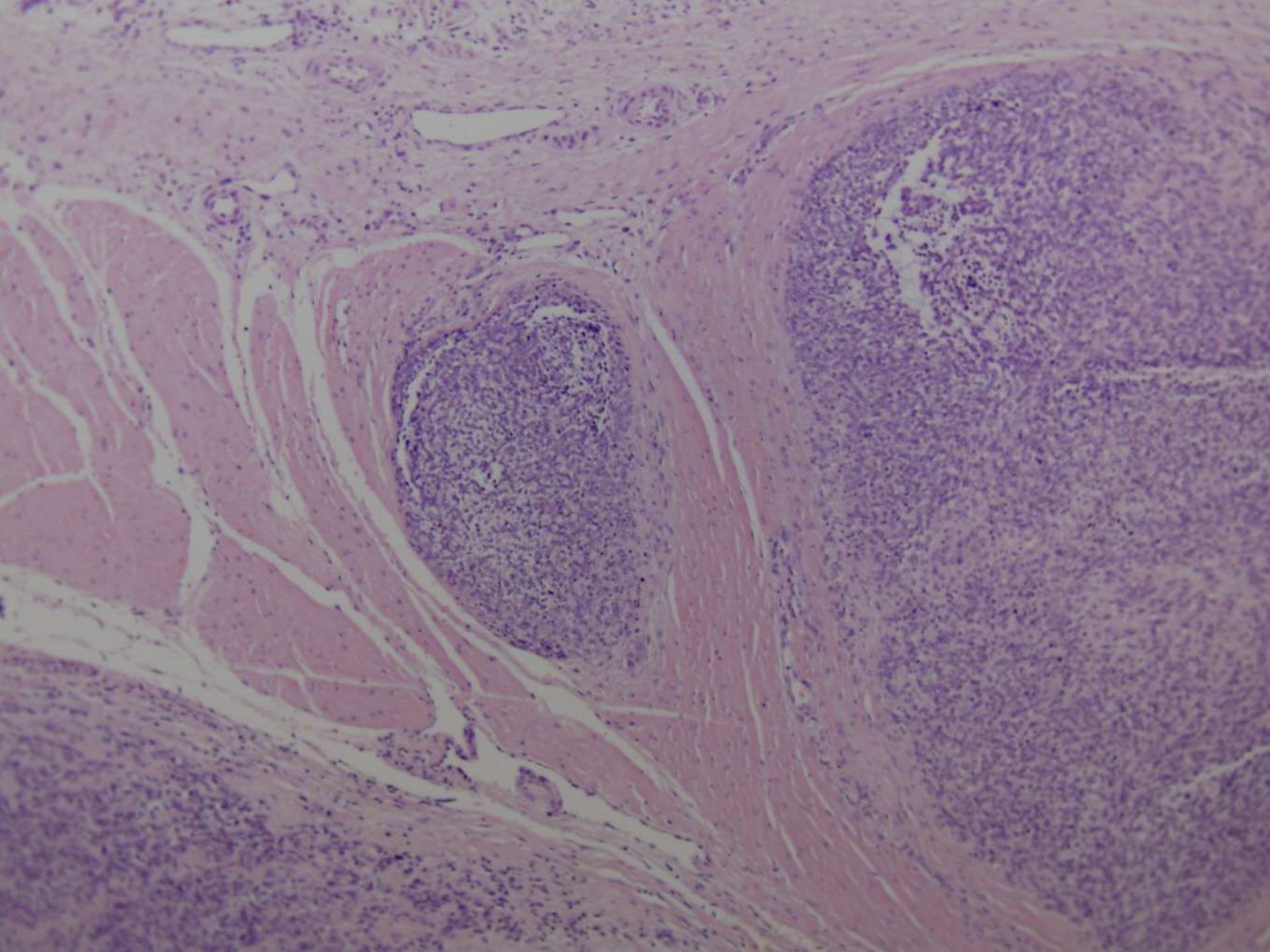
Masa en D11 con afectación del diafragma y que contacta con bazo y riñón izquierdo sin infiltrarlos

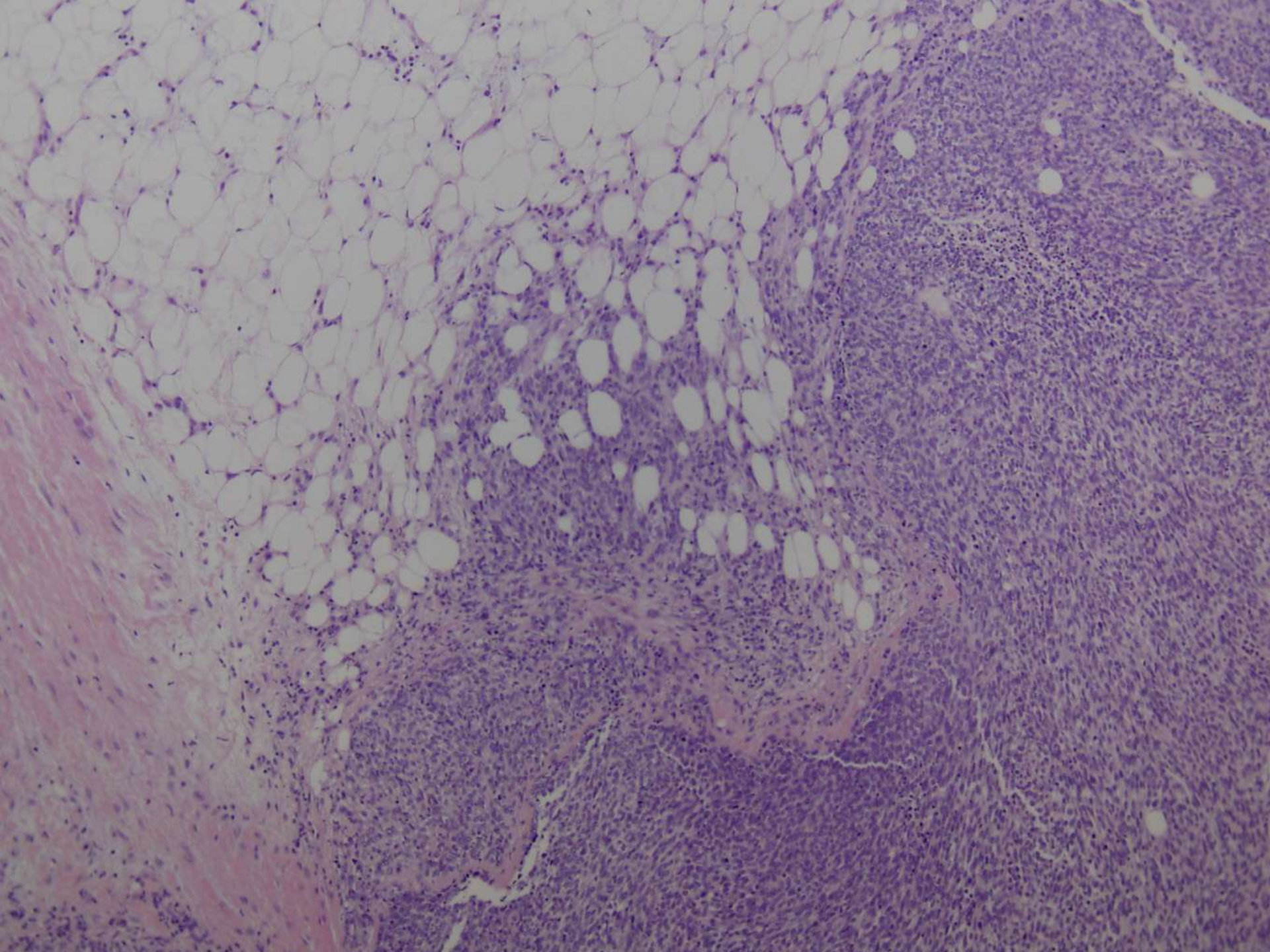
Masa pelviana dependiente de pared abdominal anterior que desplaza a la vejiga y al útero

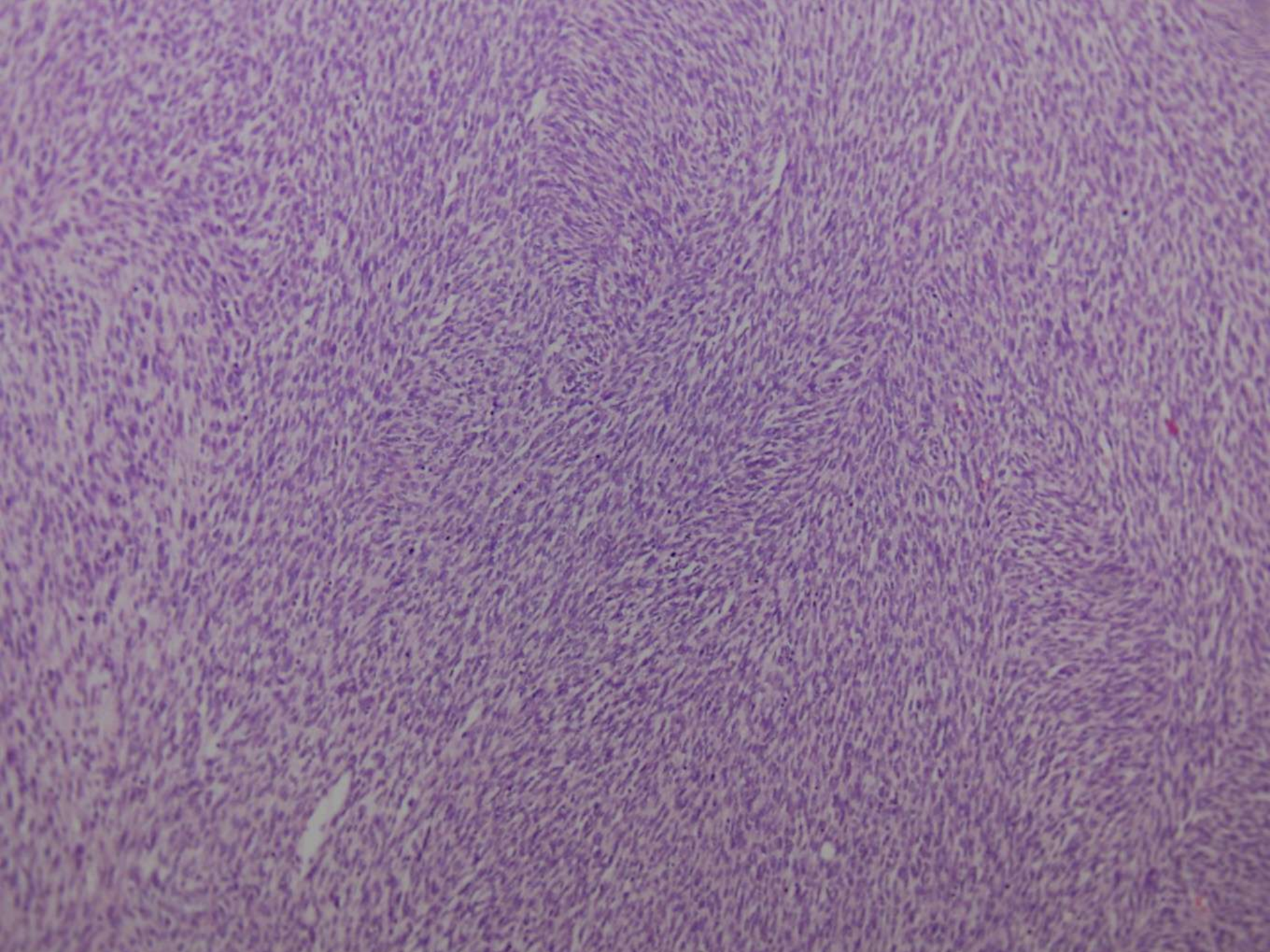

15 cm

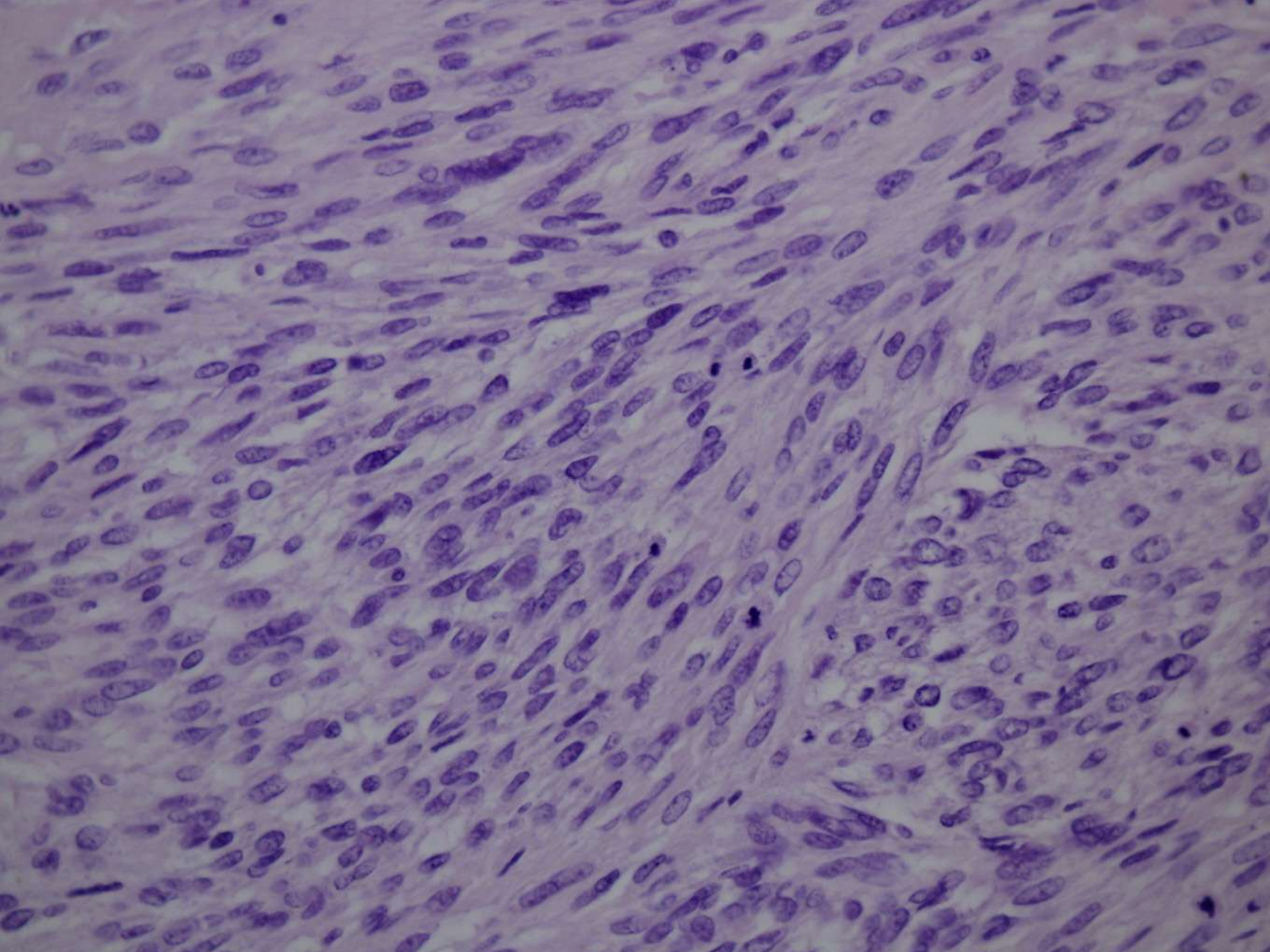


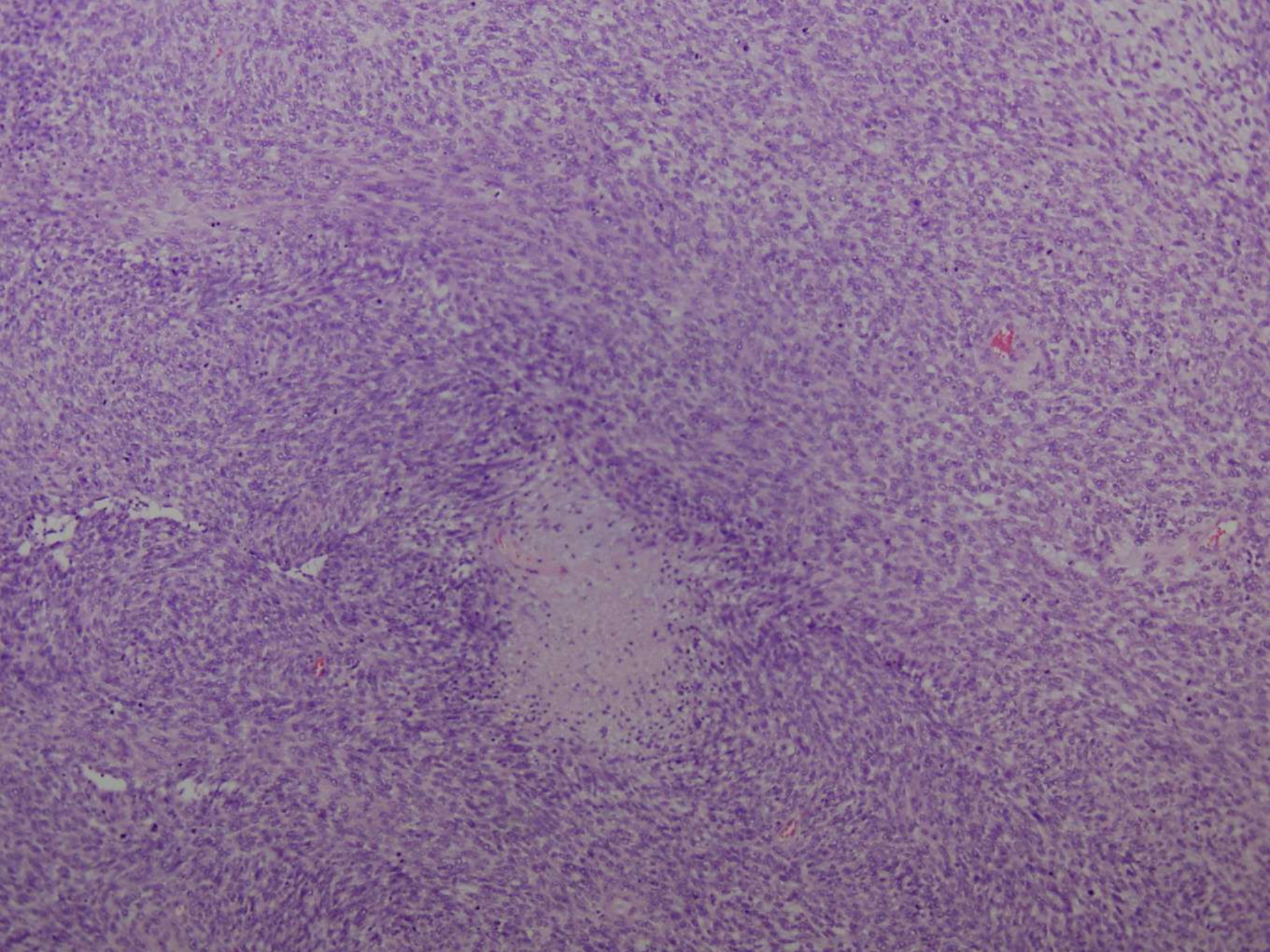


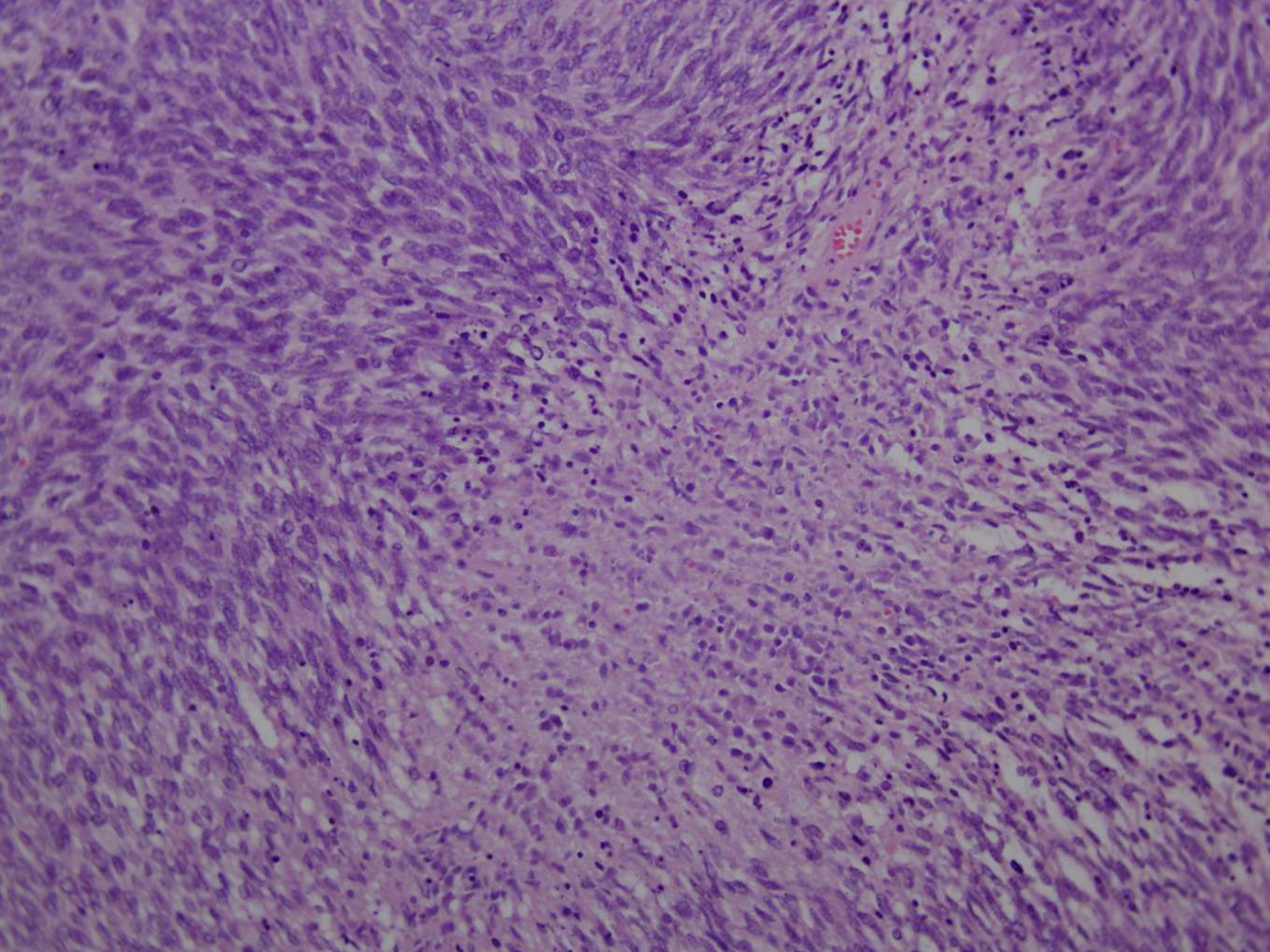


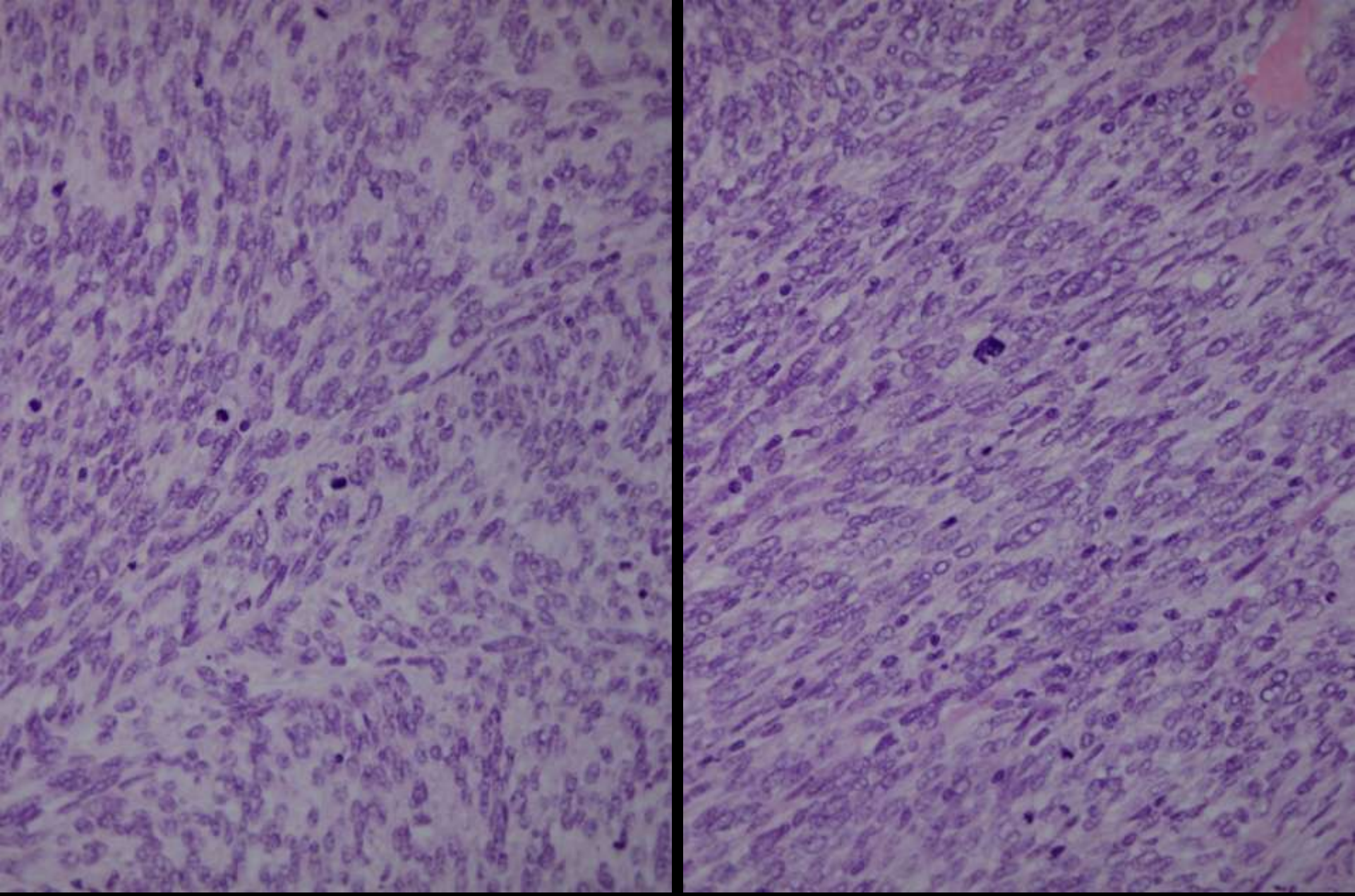


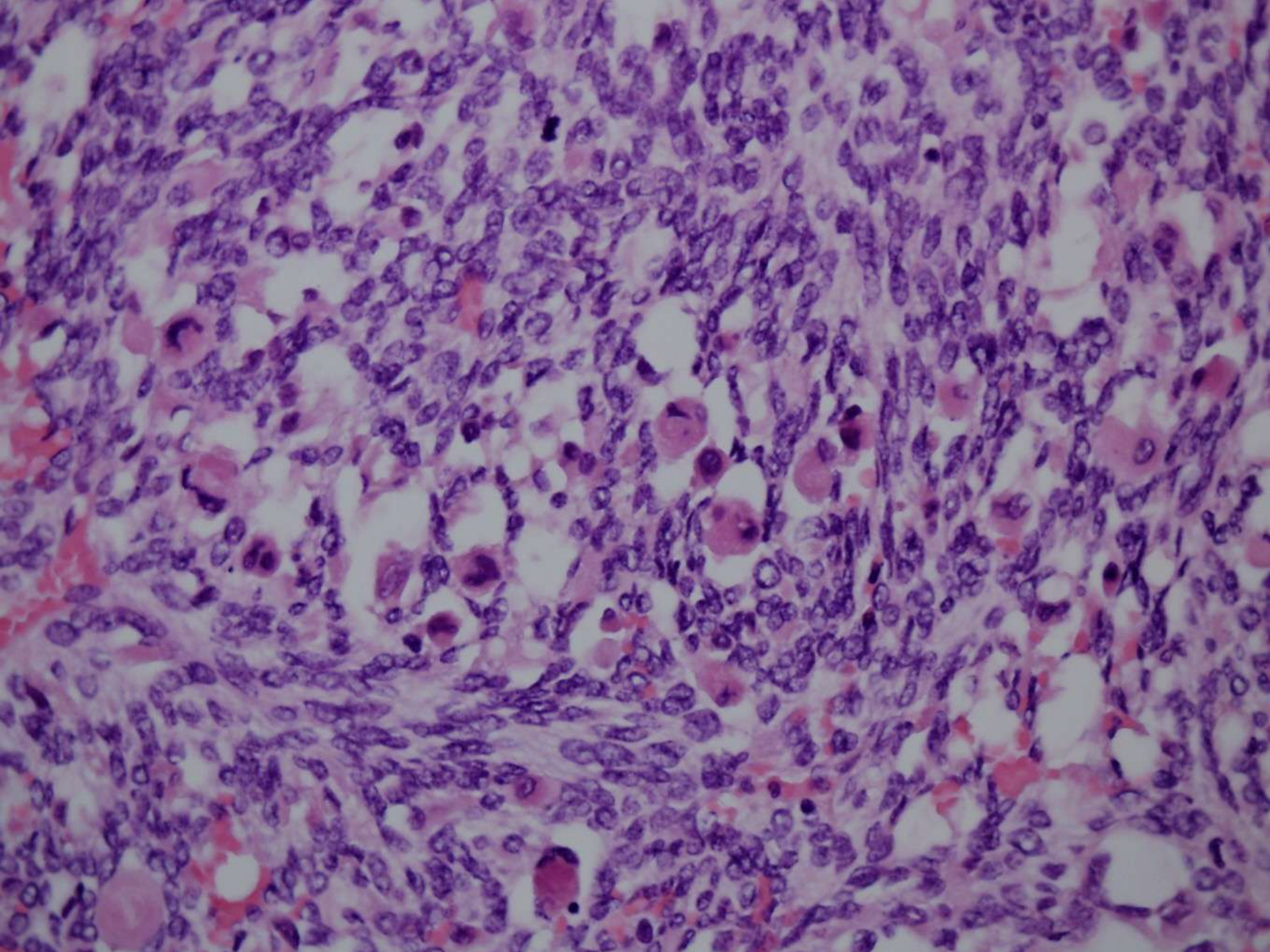


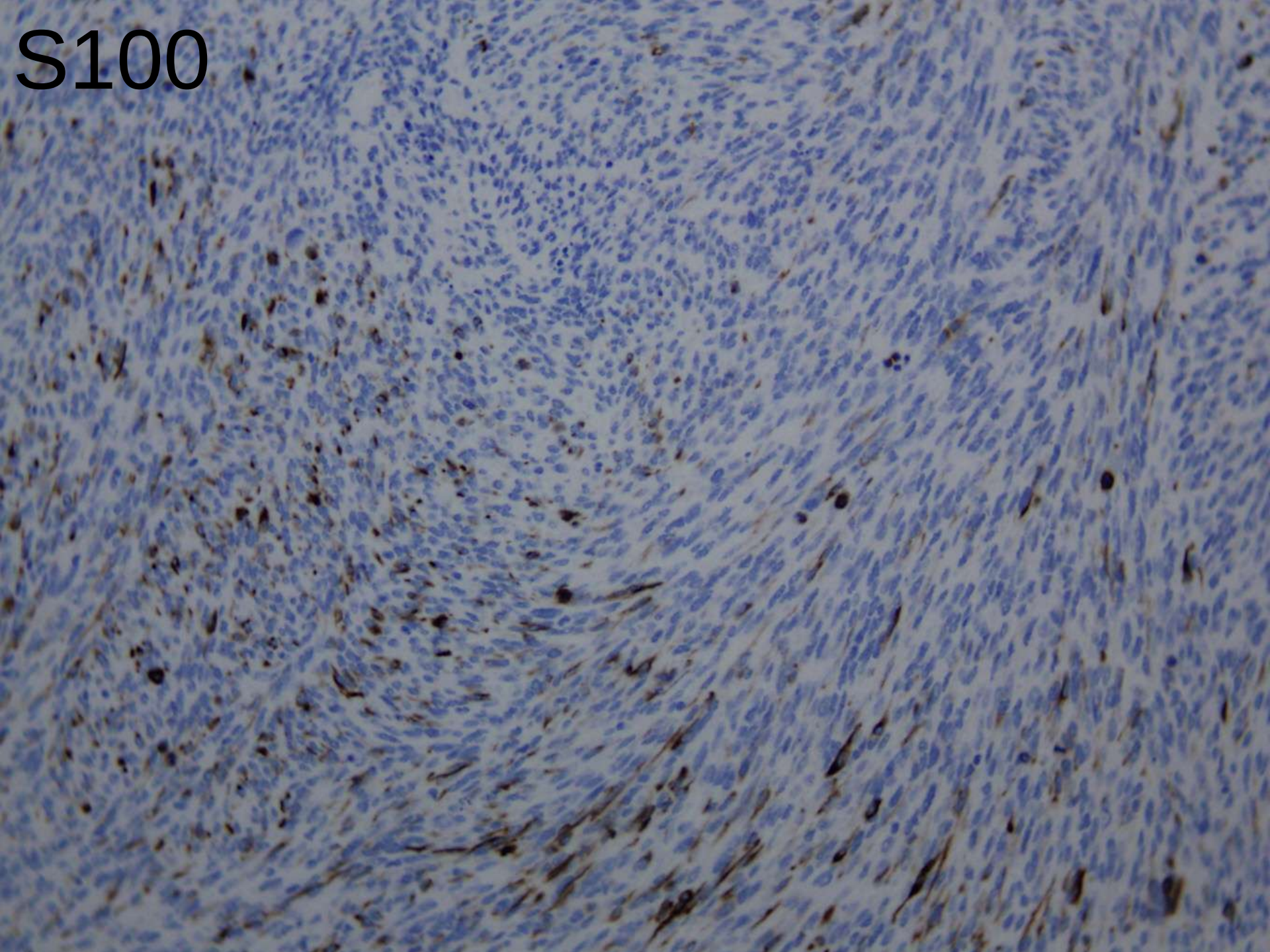


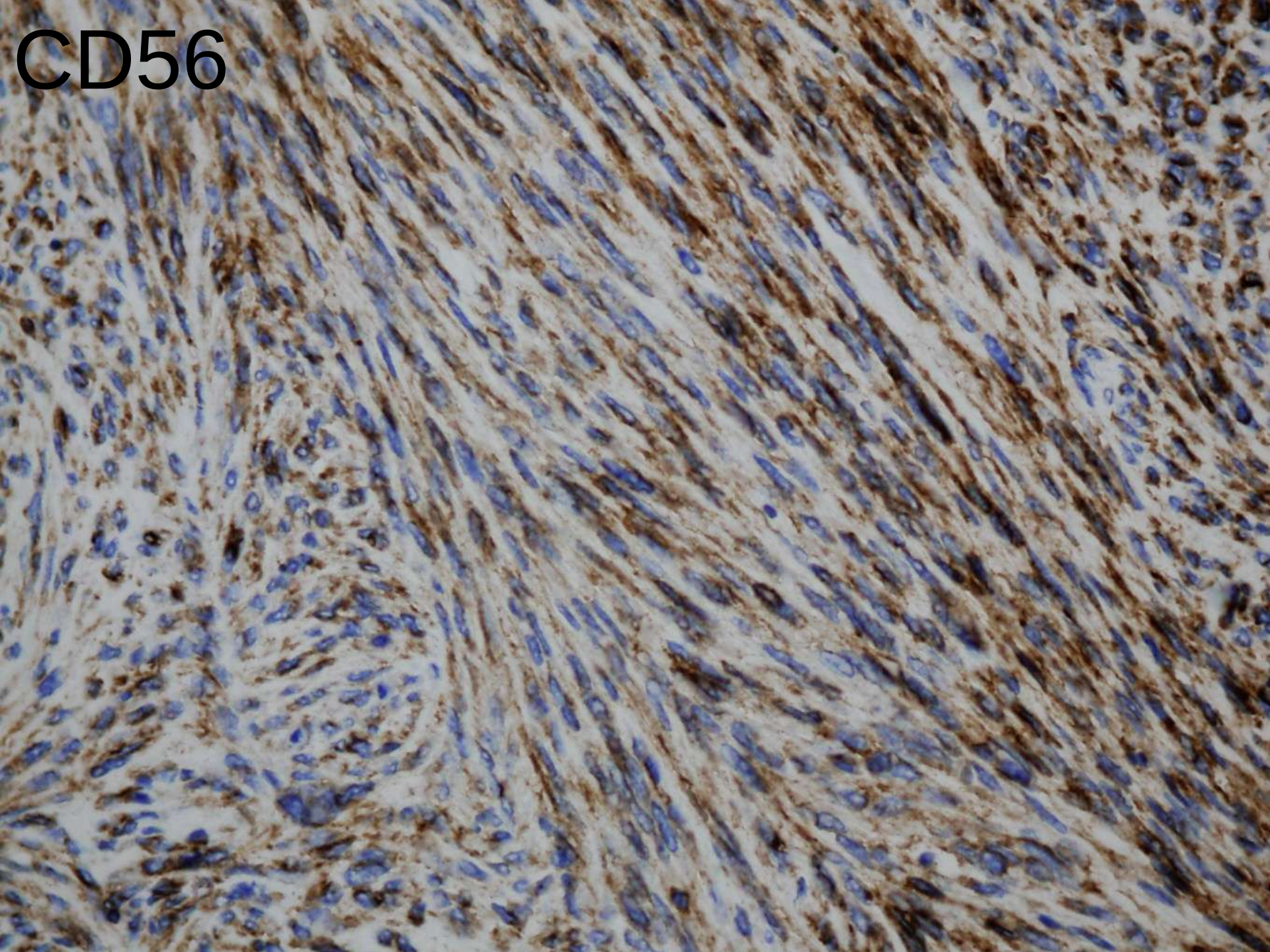


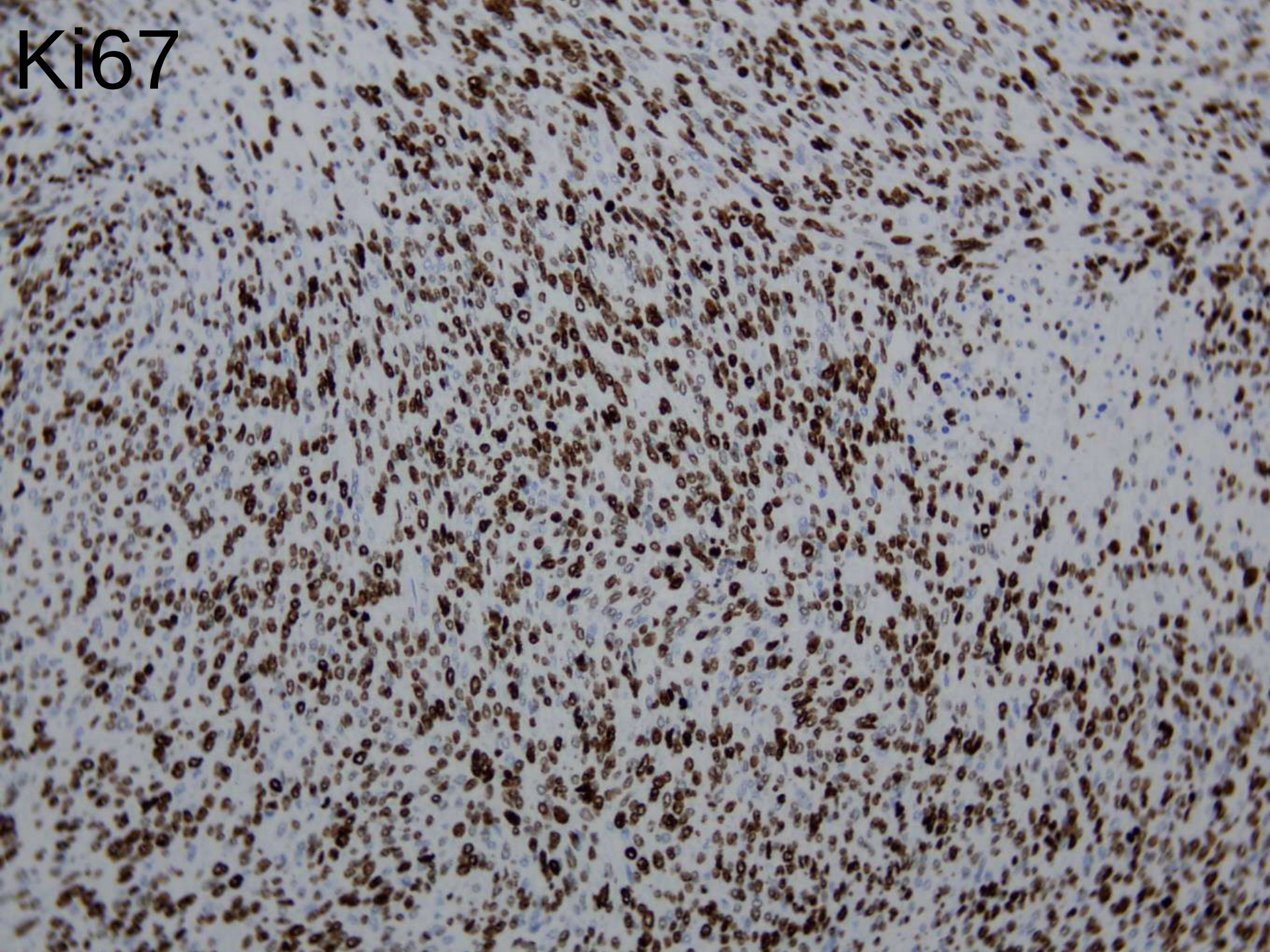




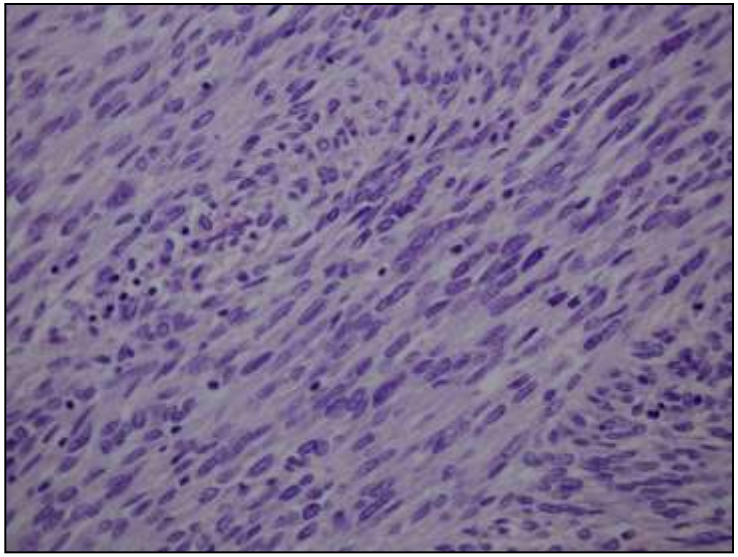


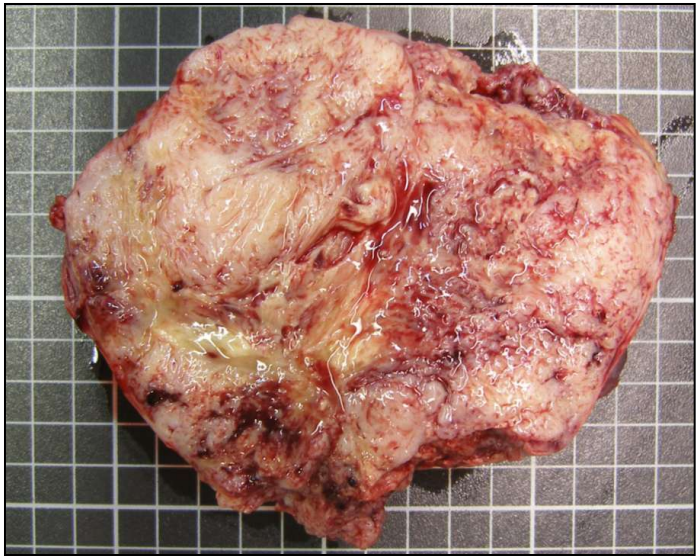

S100

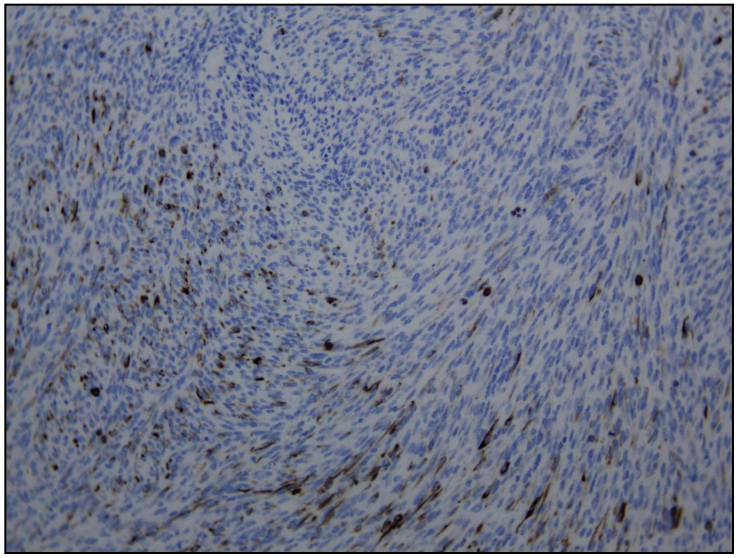

CD56


Ki67

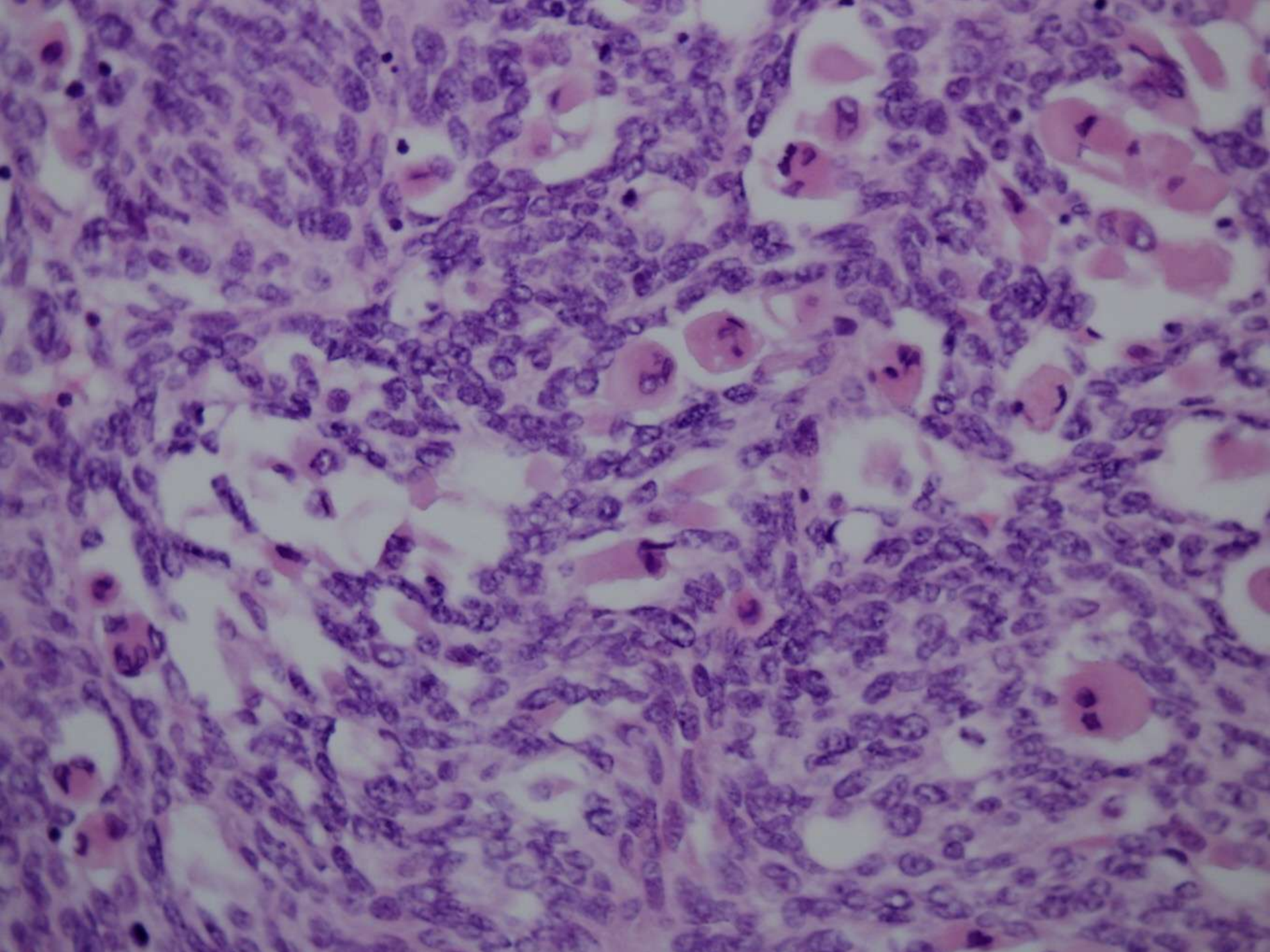
Diagnóstico



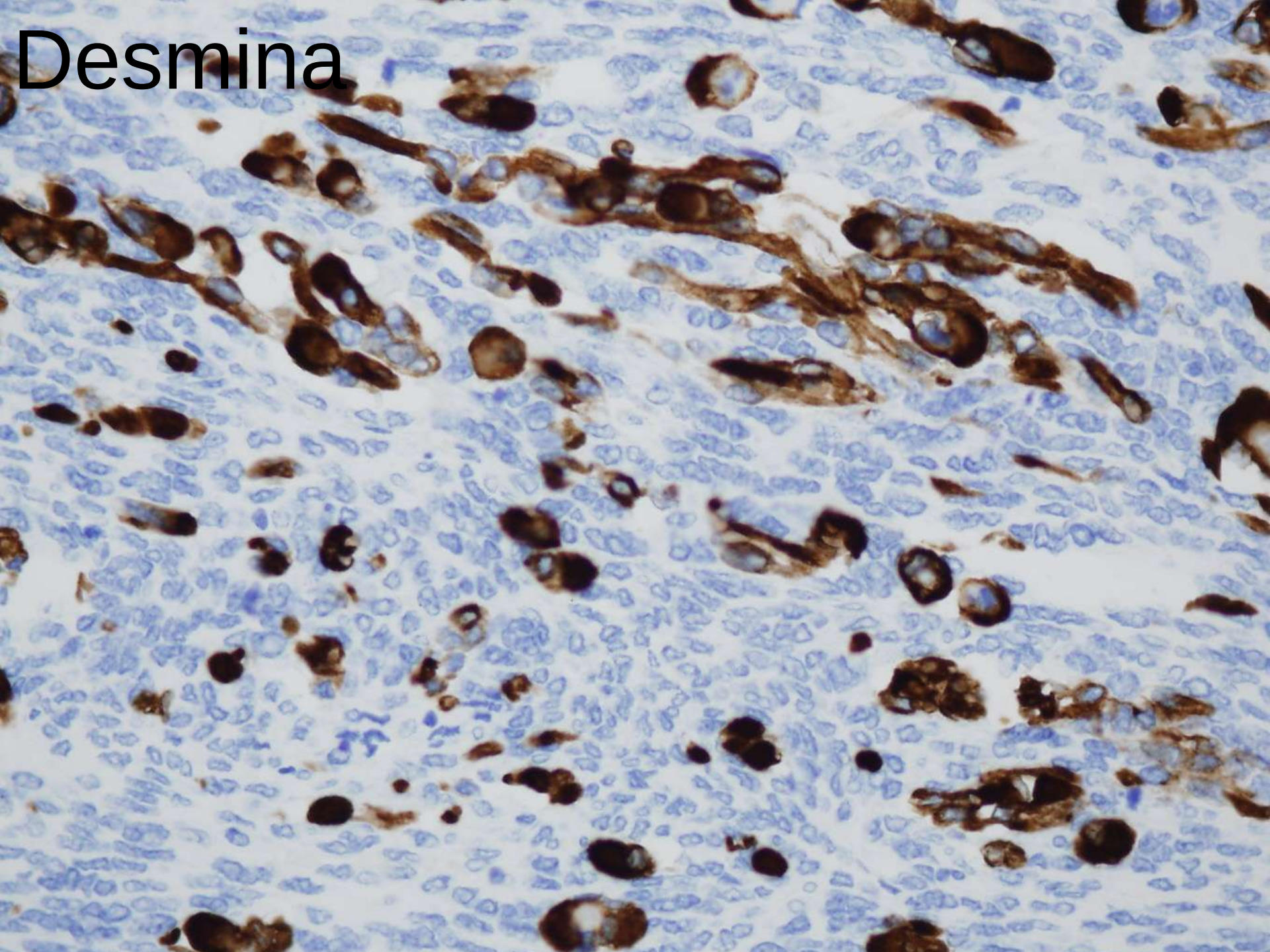


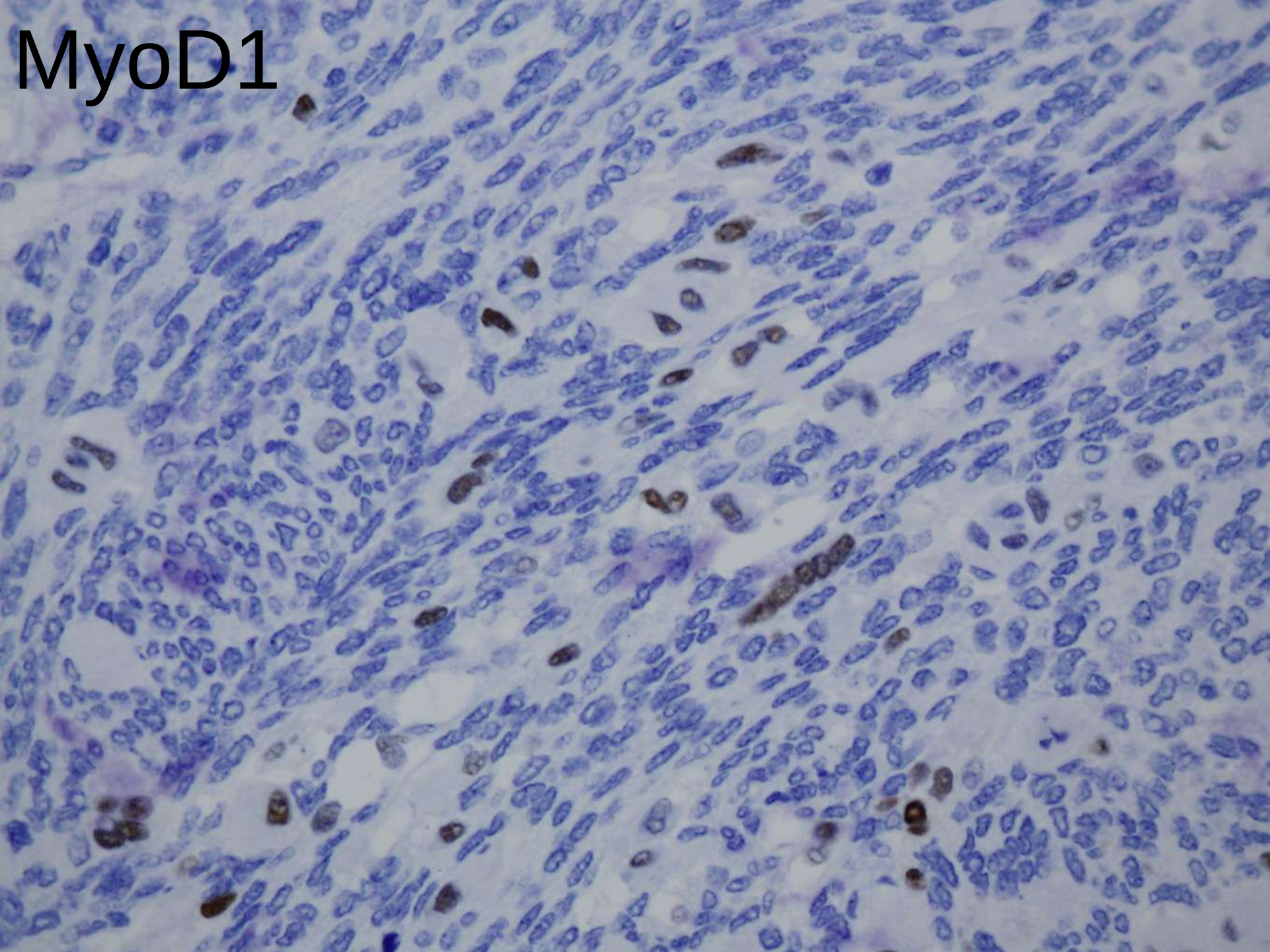


Tumor maligno de vaina nerviosa periférica



Desmina




MyoD1

No expresión inmunohistoquímica

CK AE1-AE3

EMA

CD99

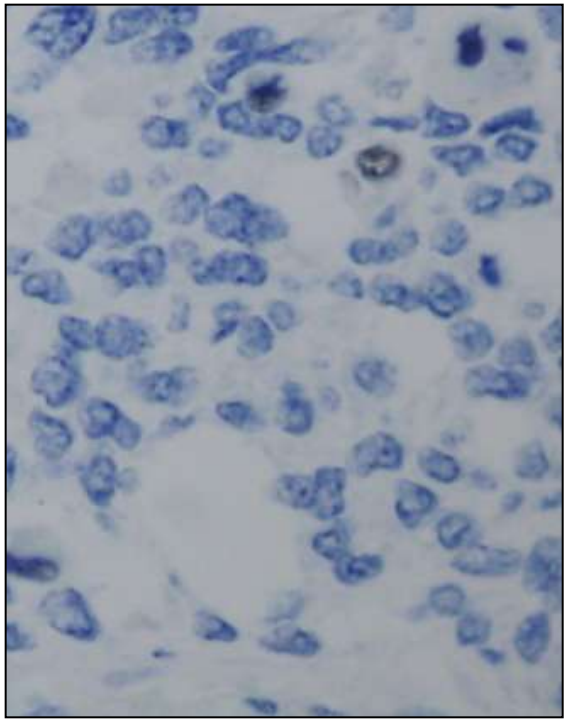
CD34

CD31

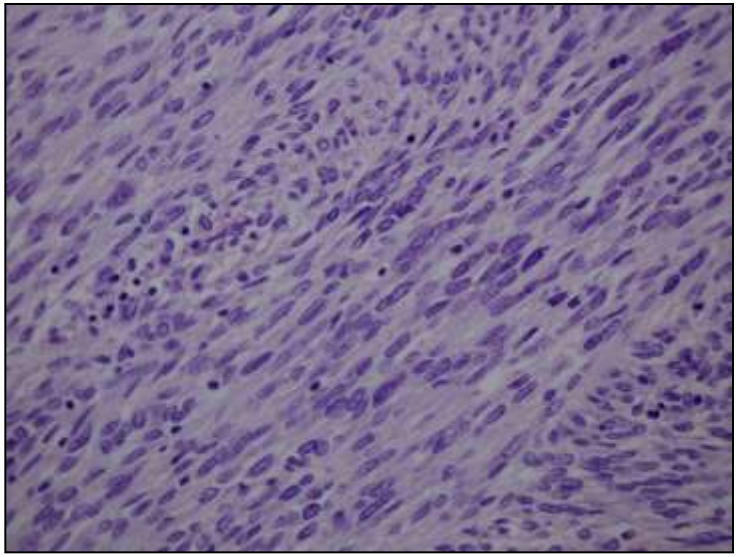
CD57

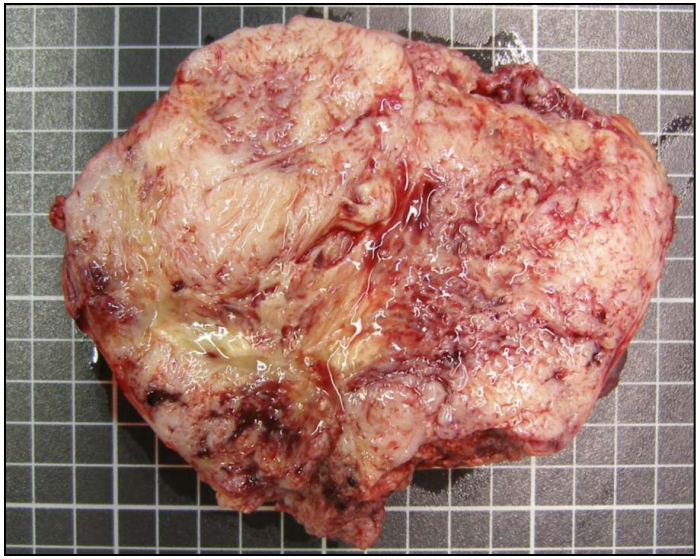
CD117

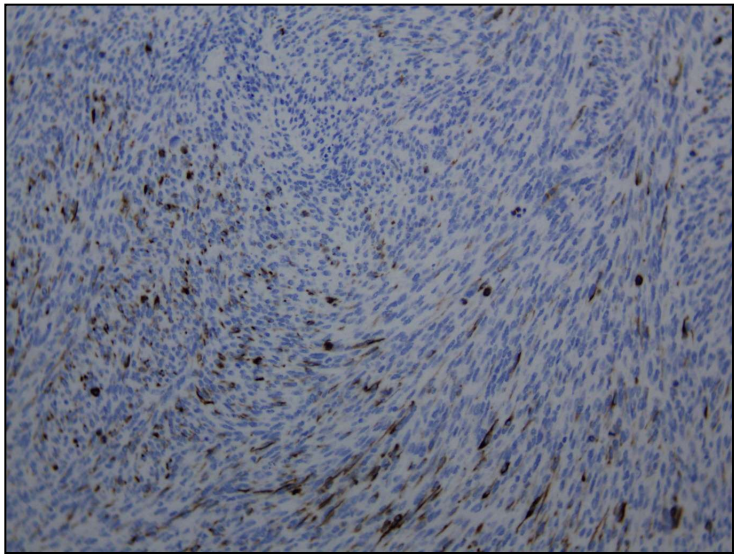
p53



Diagnóstico







Tumor maligno de vaina nerviosa periférica con difrenciación rabdomioblástica

Tumor maligno de vaina nerviosa periférica con difrenciación rabdomioblástica

Tumor Tritón

Masson P, Martin JF. Rhabdomyomes des nerfs. Bull Assoc Fr Etud Cancer **1938**; 27:751.

8% a 13% de neurofibromatosis tipo 1 (0.001% en población general)

Edad al dignóstico 26 años (NF1) y 62 años (esporádico)

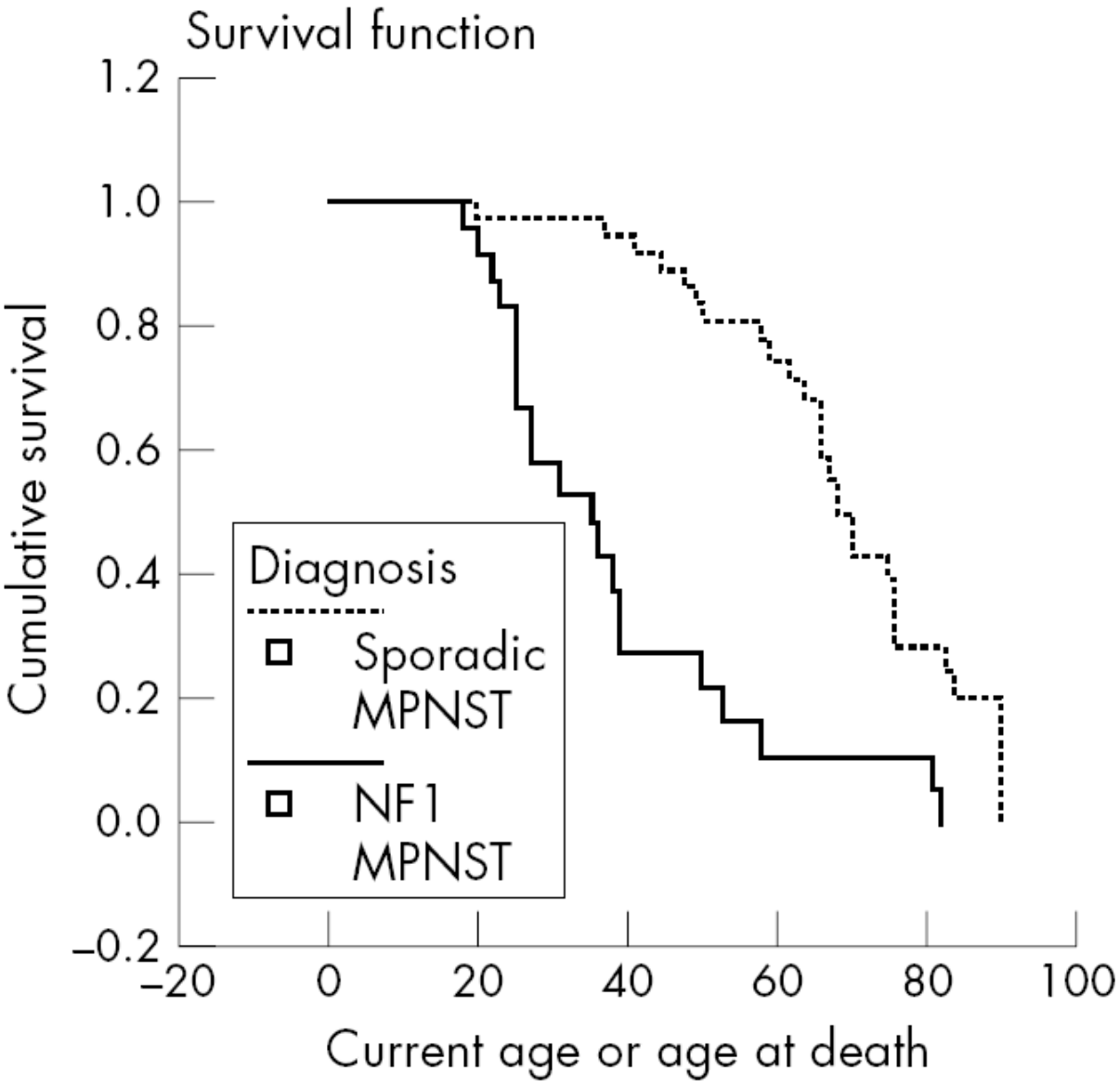
Supervivencia a los 5 años: 21% (NF1) y 42% (esporádico)

Malignant peripheral nerve sheath tumours in neurofibromatosis 1

D G R Evans, M E Baser, J McGaughran, S Sharif, E Howard, A Moran

See end of article for

J Med Genet 2002;**39**:311–314



Malignant peripheral nerve sheath tumours in neurofibromatosis 1

D G R Evans, M E Baser, J McGaughran, S Sharif, E Howard, A Moran

J Med Genet 2002;**39**:311–314

Soft-Tissue Sarcomas in Children and Adolescents With Neurofibromatosis Type 1

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Gianni Bisogno, MD[2]
Alessandra Macaluso, MD[3]
Rita Alaggio, MD[5]
Giovanni Cecchetto, MD[6]
Modesto Carli, MD[2]

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[2] Division of Pediatric Hematology and Oncology, Padova University, Padova, Italy.

[3] Division of Pediatric Hematology and Oncology, Ospedale dei Bambini G. Di Cristina, Palermo, Italy.

[4] Division of Pediatric Hematology and Oncology, Ospedale G. Salesi, Ancona, Italy.

[5] Pathology Department, Padova University, Padova, Italy.

BACKGROUND. Patients affected by neurofibromatosis type 1 (NF1) are at higher risk of developing soft-tissue sarcomas (STS) than the general population. The clinical findings and outcome in 43 children and adolescents with NF1 treated

METHODS. The study included 37 patients with neurogenic sarcomas (36 malignant peripheral nerve sheath tumors [MPNST], 1 triton tumor) and 6 cases of

riod was 43% in the MPNST population and 1% in the RMS group.

RESULTS. Most patients with neurogenic sarcomas had large, invasive tumors. Five-year event-free and overall survival rates were 19% and 28%, respectively. Two of 16 patients with evaluable disease responded to chemotherapy. All 6 RMS patients were $\leq$3 years old and had embryonal subtype, 5 of 6 arising in the genitourinary tract or pelvis (paravesical); 4 were alive in first remission at the time of the analysis, 1 was alive in second remission after a local recurrence, and 1 died of disease.

CONCLUSIONS. The occurrence of STS in pediatric patients with NF1 syndrome in Italy is discussed, confirming that NF1 patients have a high risk of developing STS, and particularly MPNST, often with an aggressive clinical presentation and poor outcome. Cases of RMS tended to have particular features (early age, embryonal histotype, genitourinary site) and their outcome seemed to resemble that of the general RMS population. ***Cancer*** **2007;109:1406–12.**

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