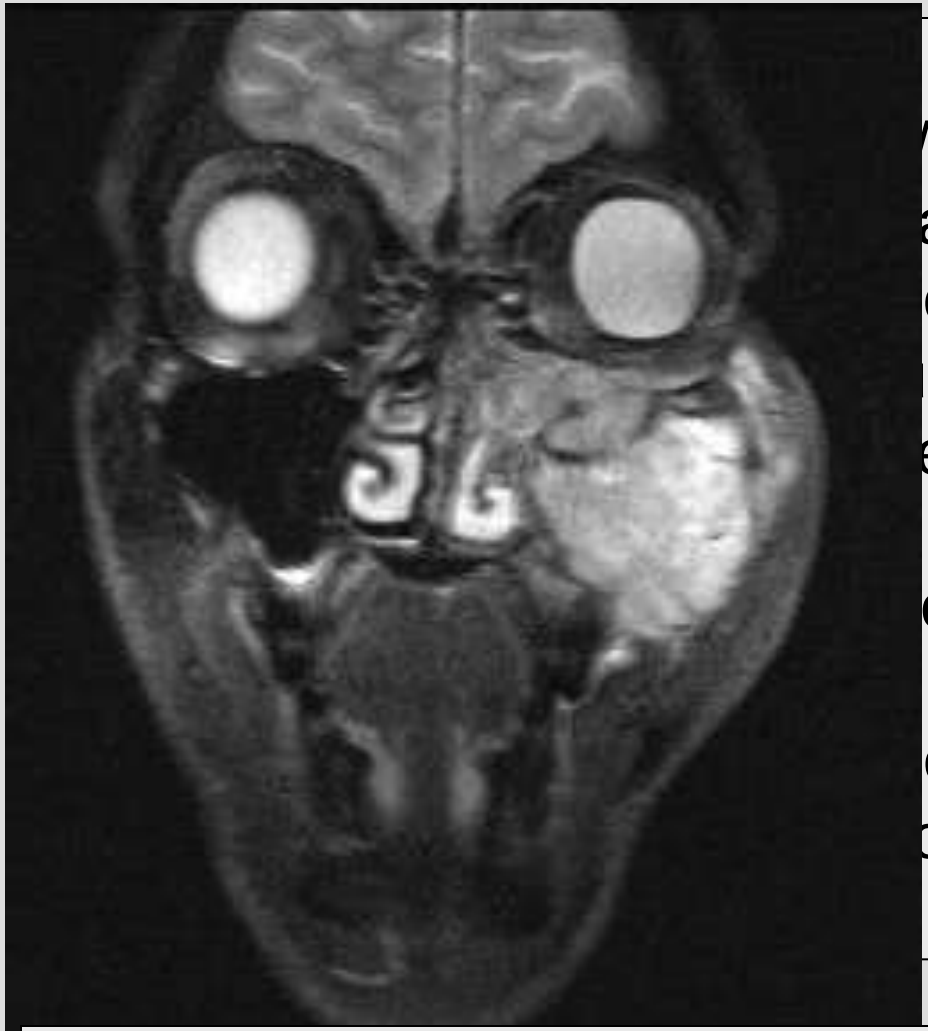
XXV CONGRESO DE LA SOCIEDAD ESPAÑOLA DE ANATOMÍA PATOLÓGICA Y DIVISIÓN ESPAÑOLA DE LA ACADEMIA INTERNACIONAL DE PATOLOGÍA (SEAP-IAP)
XX CONGRESO DE LA SOCIEDAD ESPAÑOLA DE CITOLOGÍA (SEC)
I CONGRESO DE LA SOCIEDAD ESPAÑOLA DE PATOLOGÍA FORENSE (SEPAF)
"Consolidando puentes" *ZARAGOZA 18-21 MAYO, 2011*

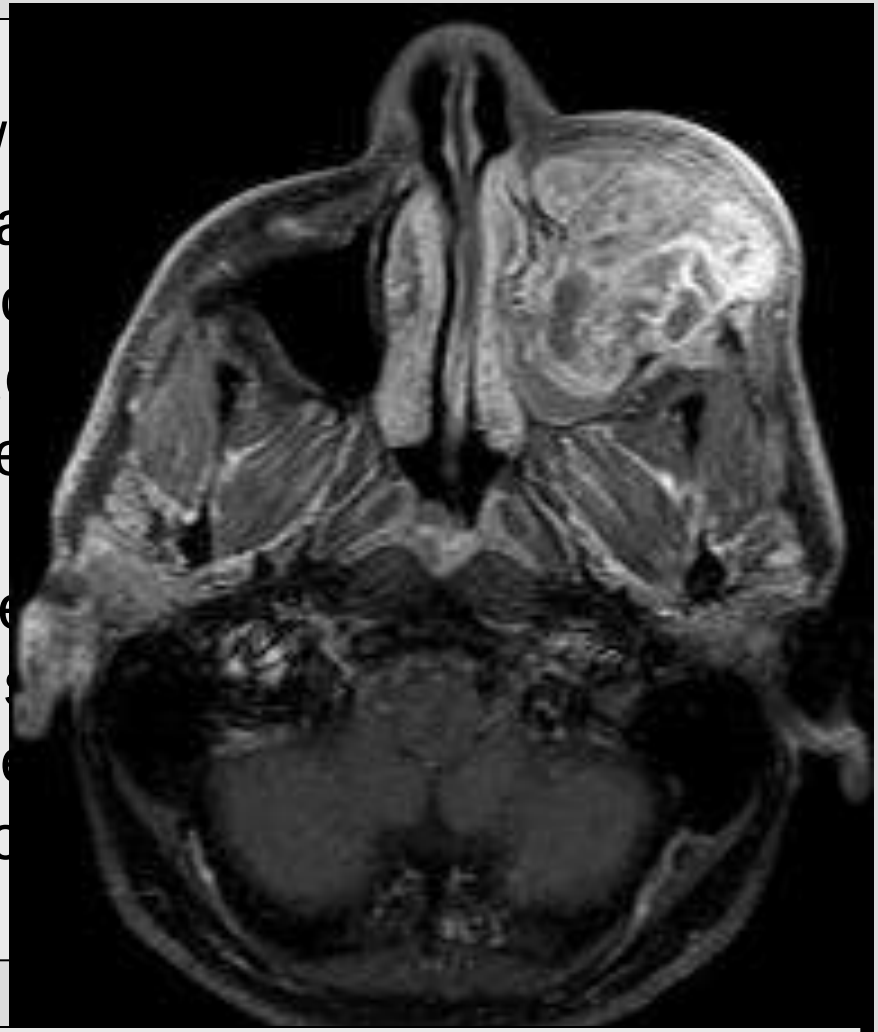
CLUB DE PATOLOGÍA DE CABEZA Y CUELLO

SEMINARIO DE CASOS

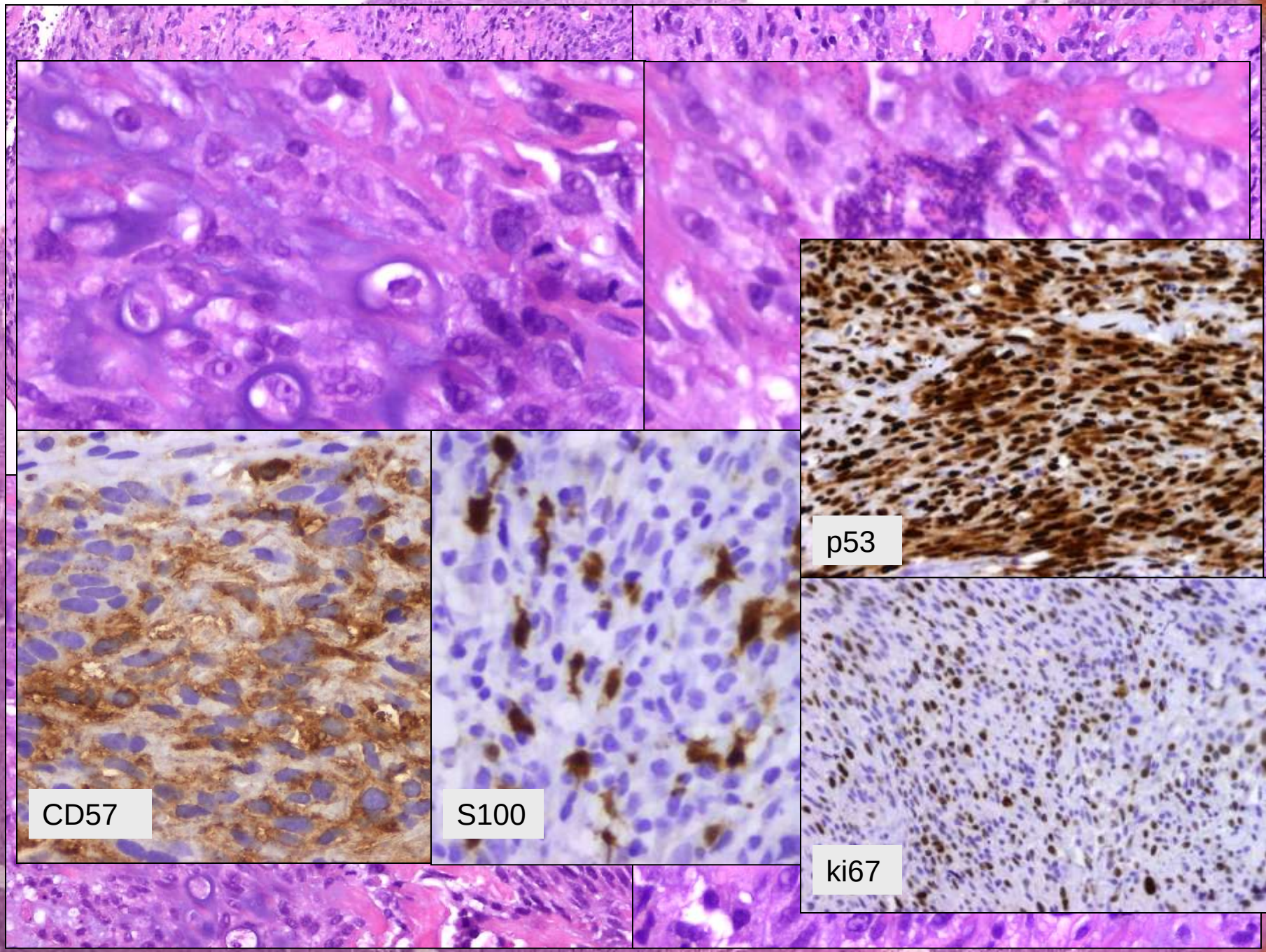
CASO Nº 5

Dr PERE HUGUET
Servei Anatomia Patològica
HOSPITAL VALL D'HEBRON
BARCELONA





- PAAF: tumor de células mesenquimales que precisa de biopsia para su diagnóstico definitivo
- Biopsia-trucut: diagnóstico (se remite imagen de la misma)
- Ingresa para cirugía electiva de exéresis y reconstrucción.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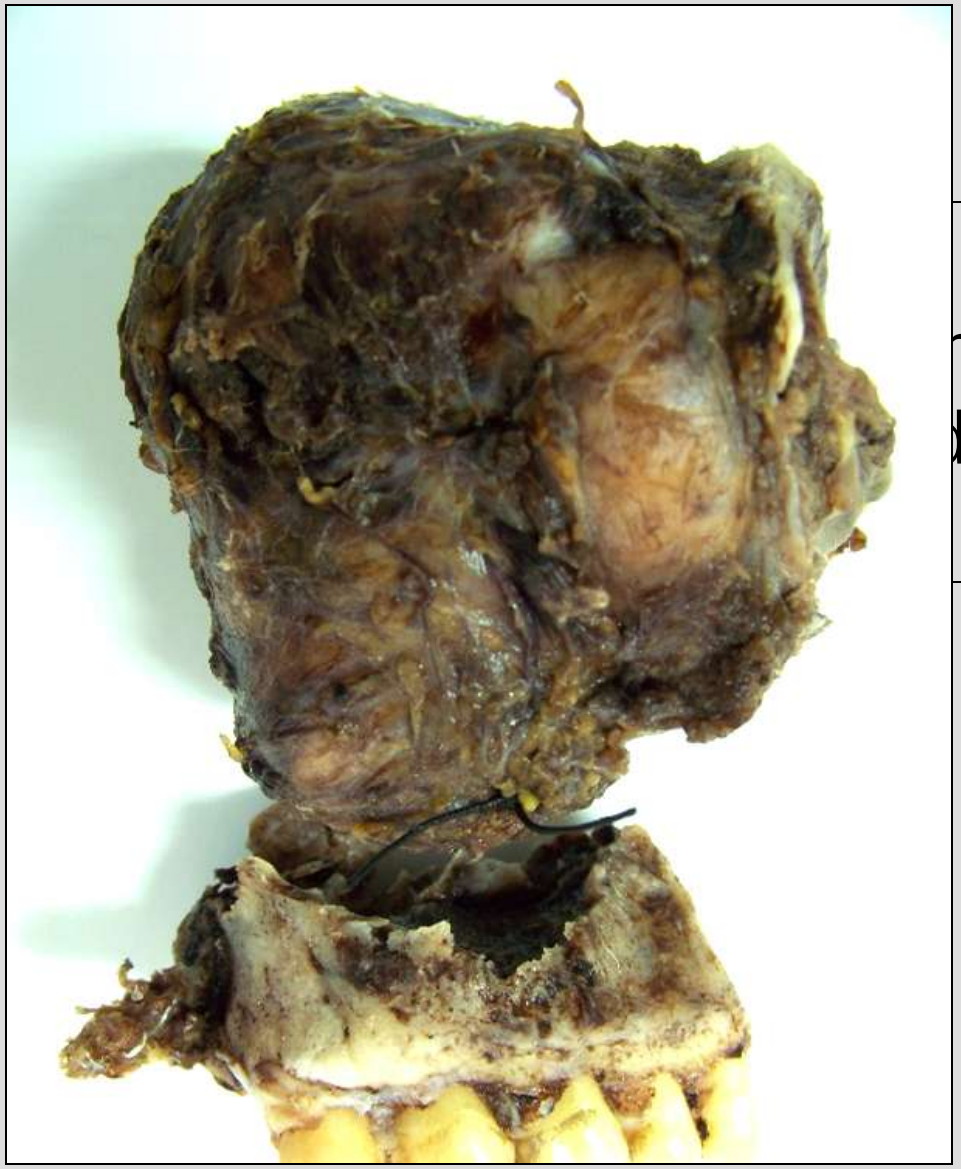

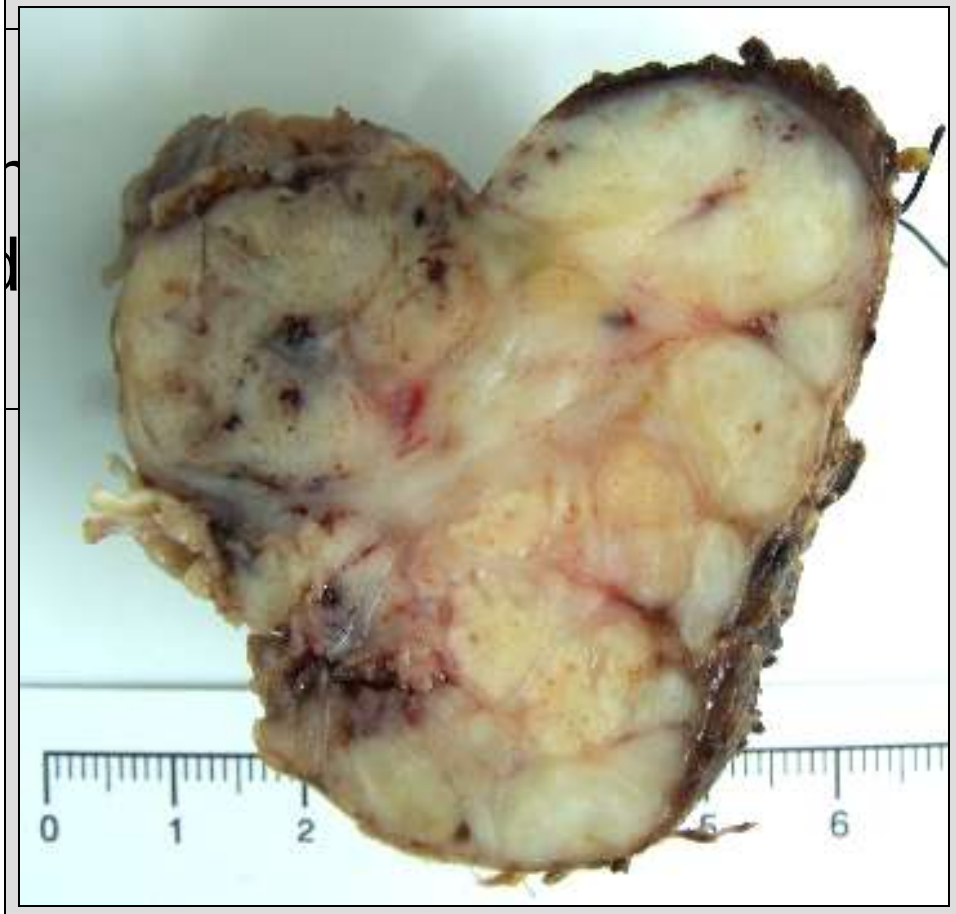
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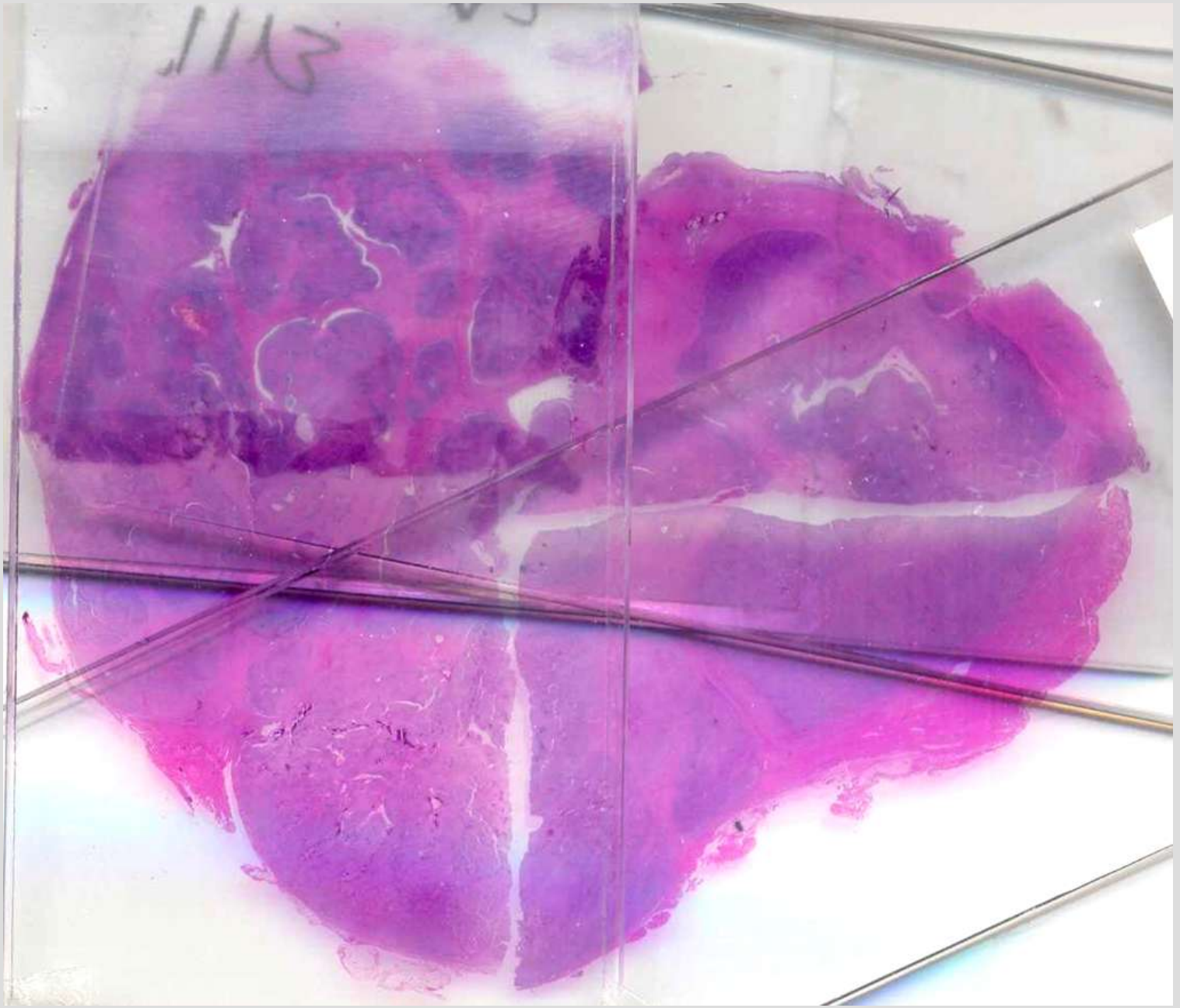

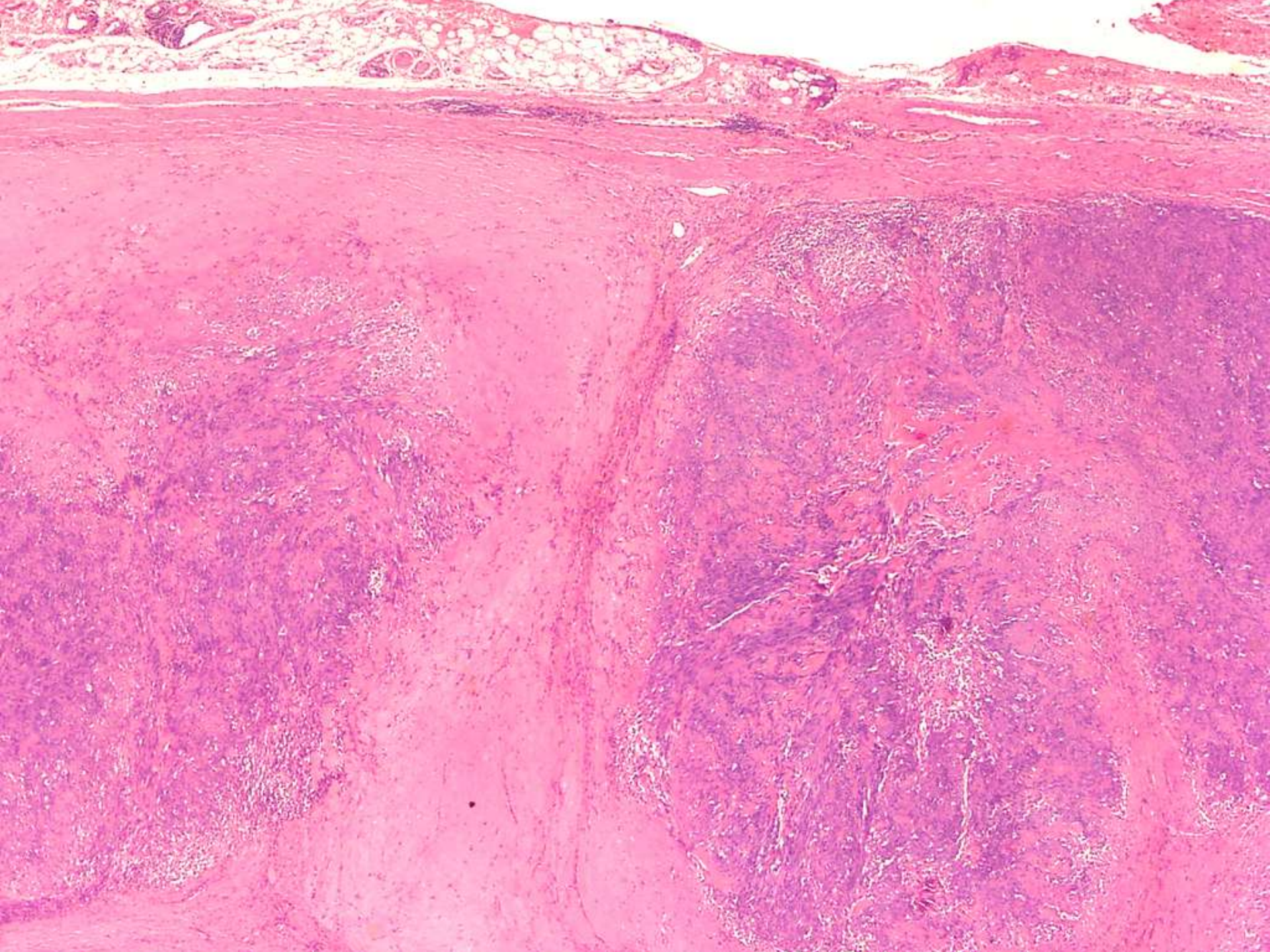

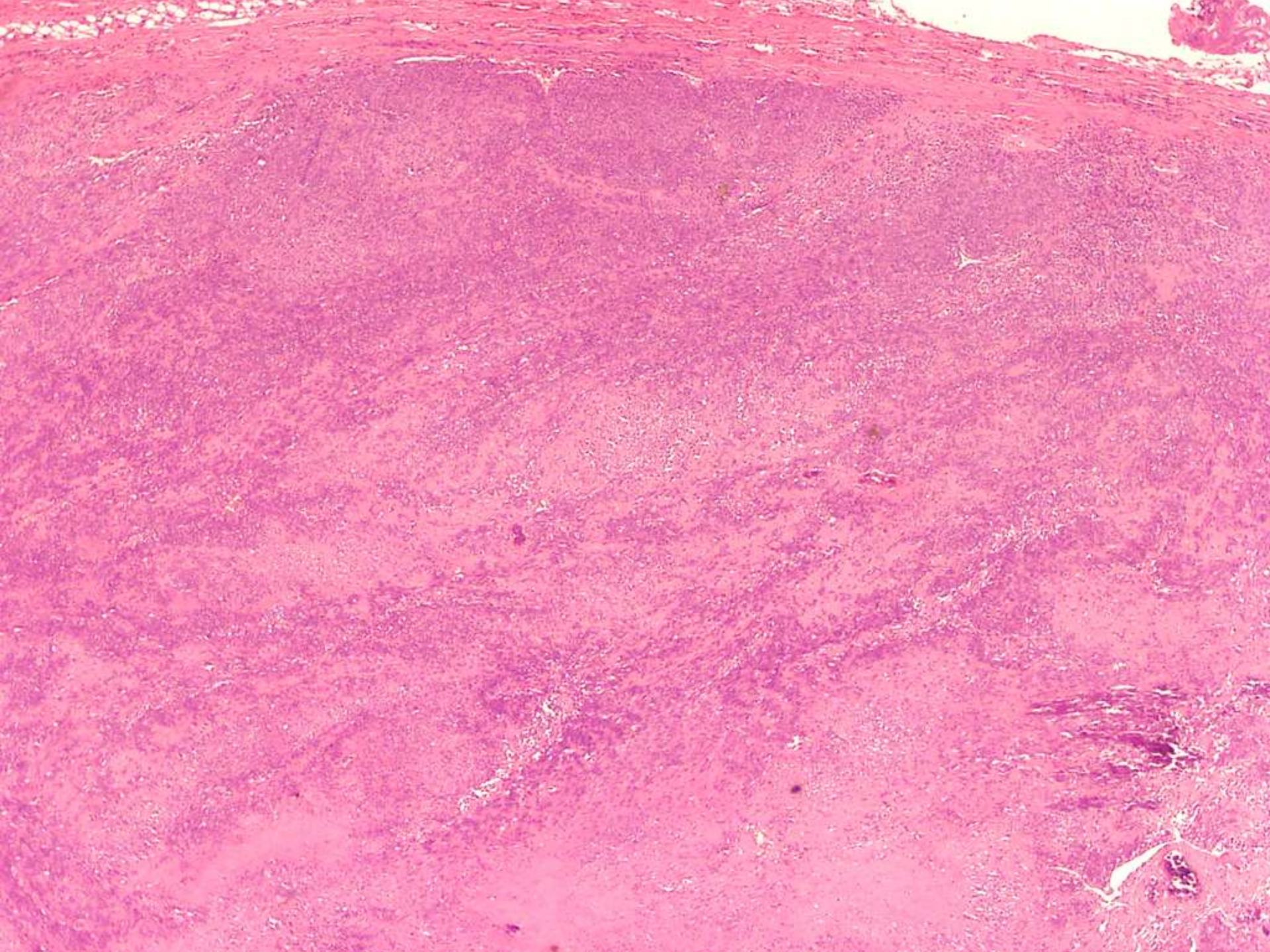

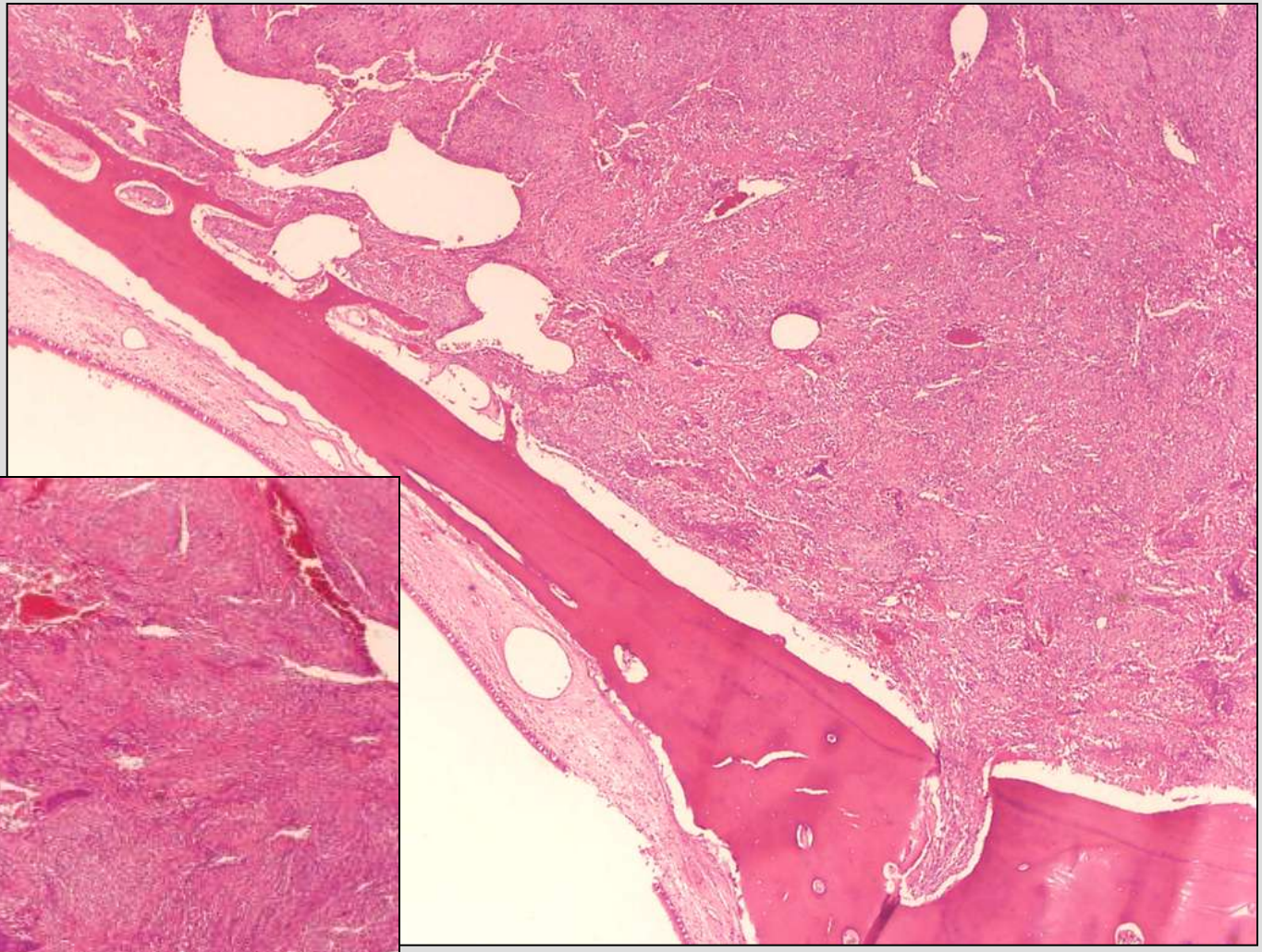

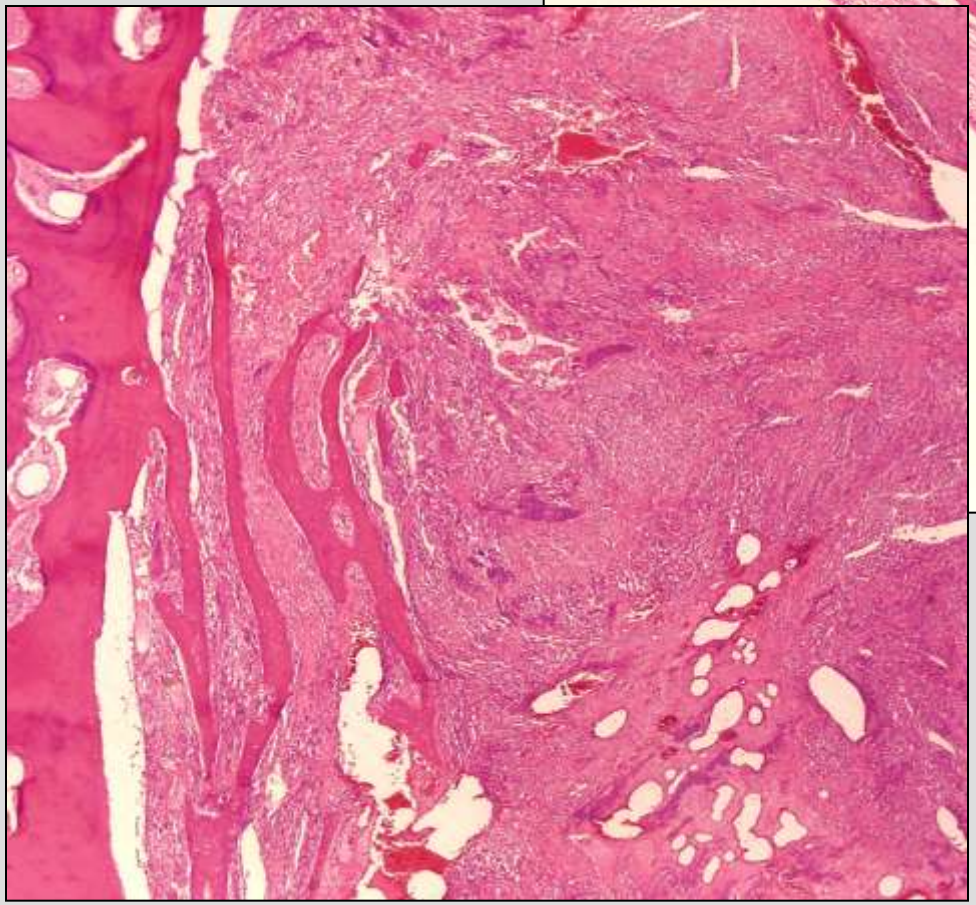

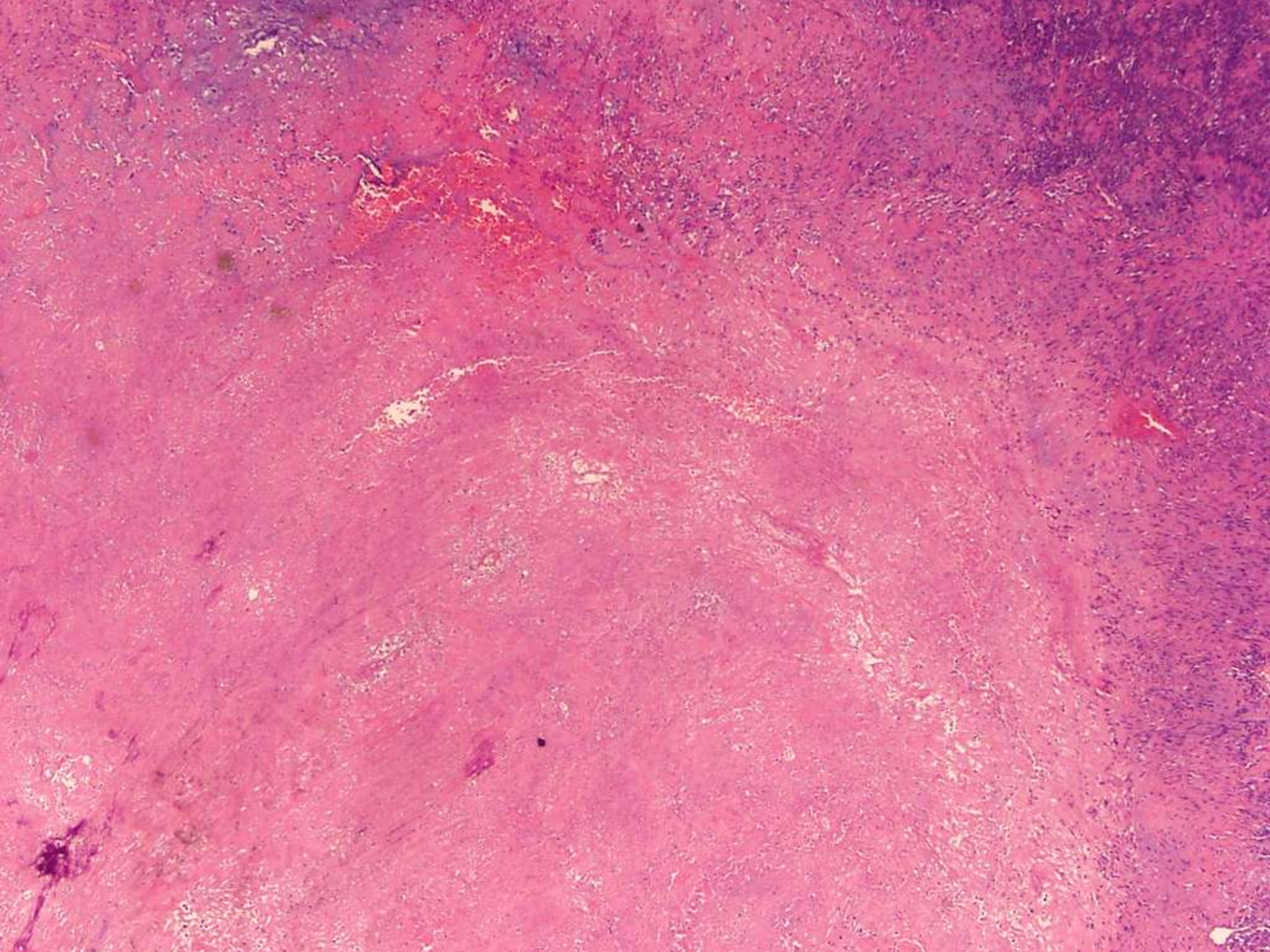

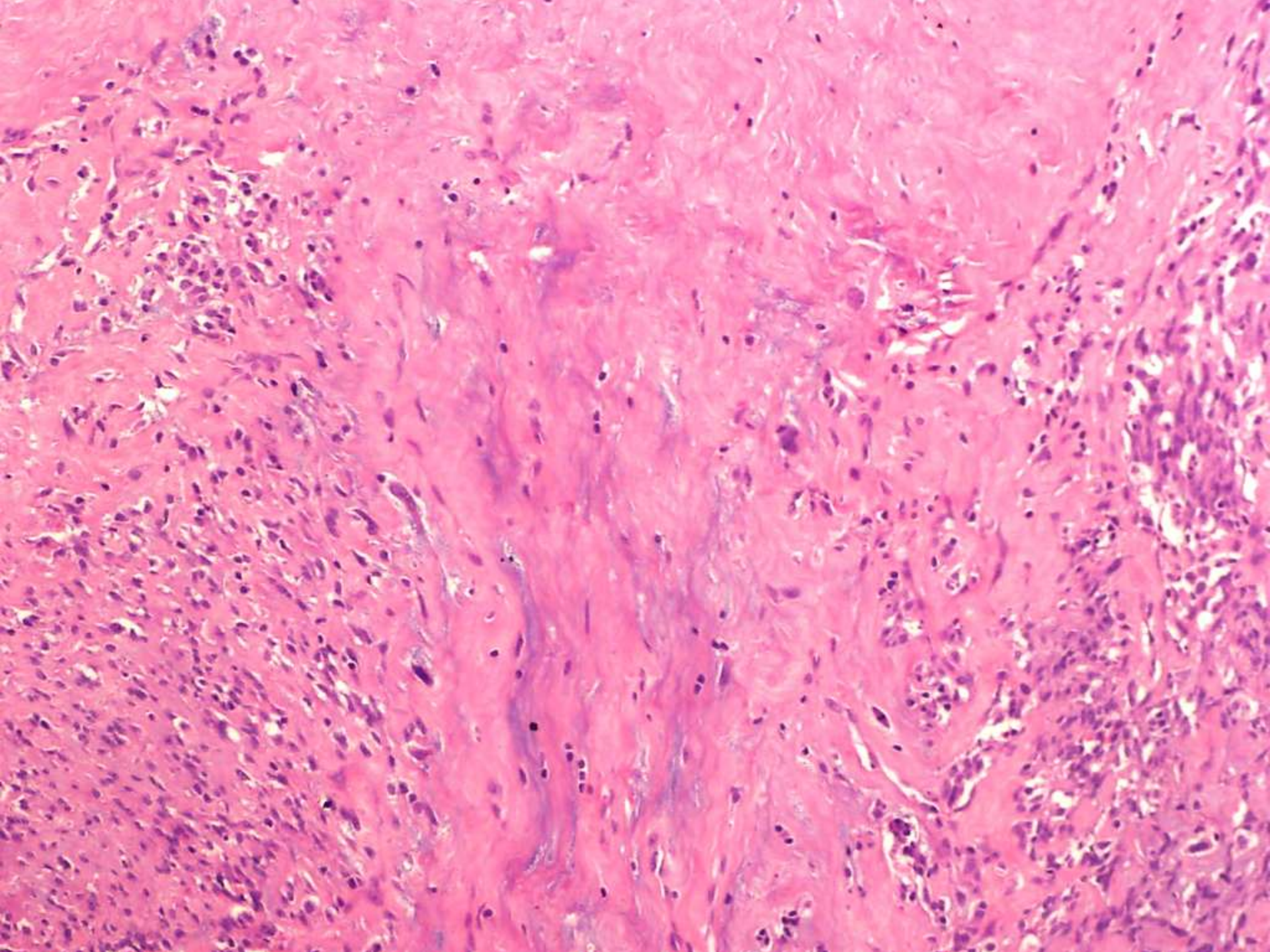

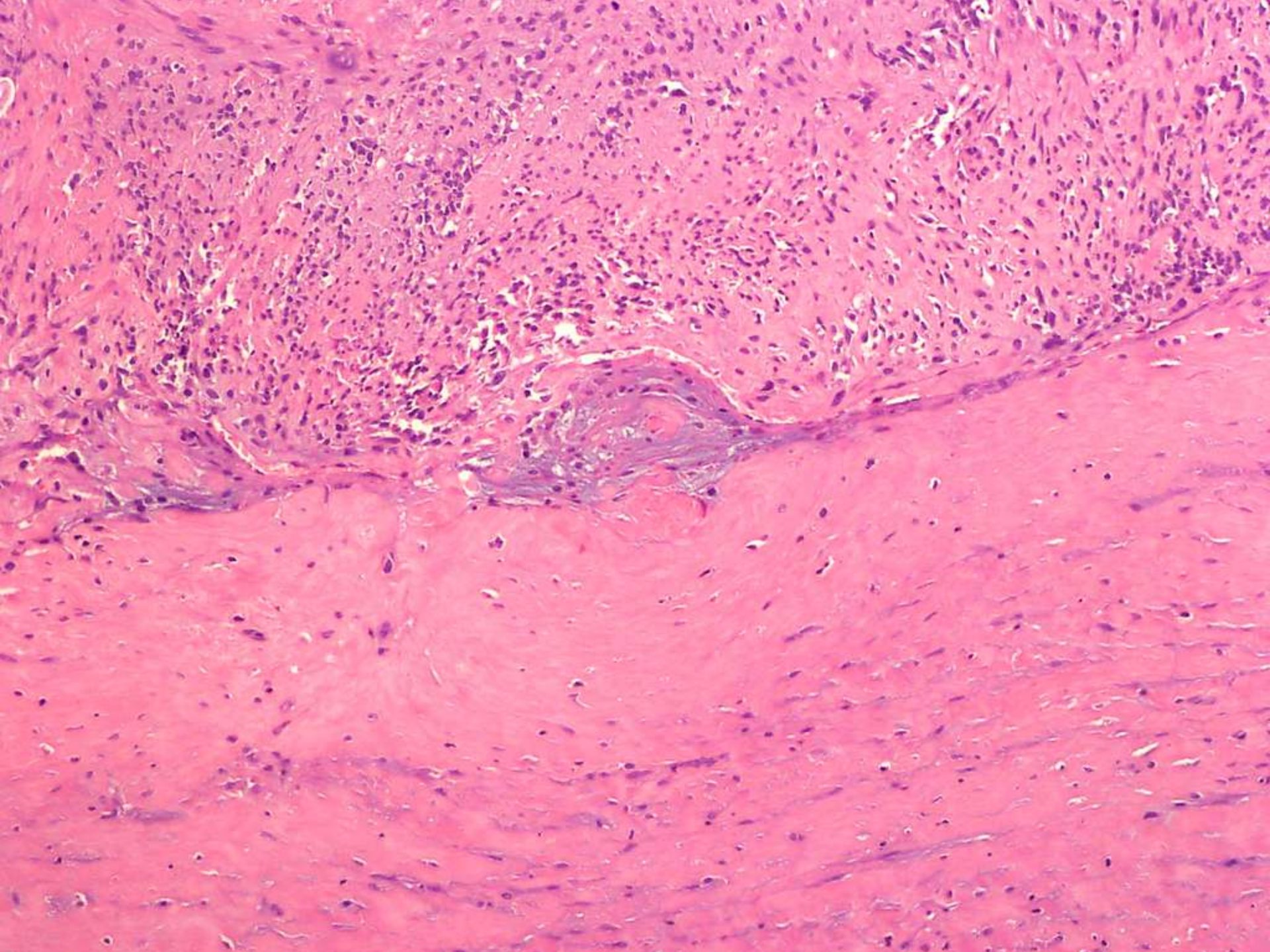

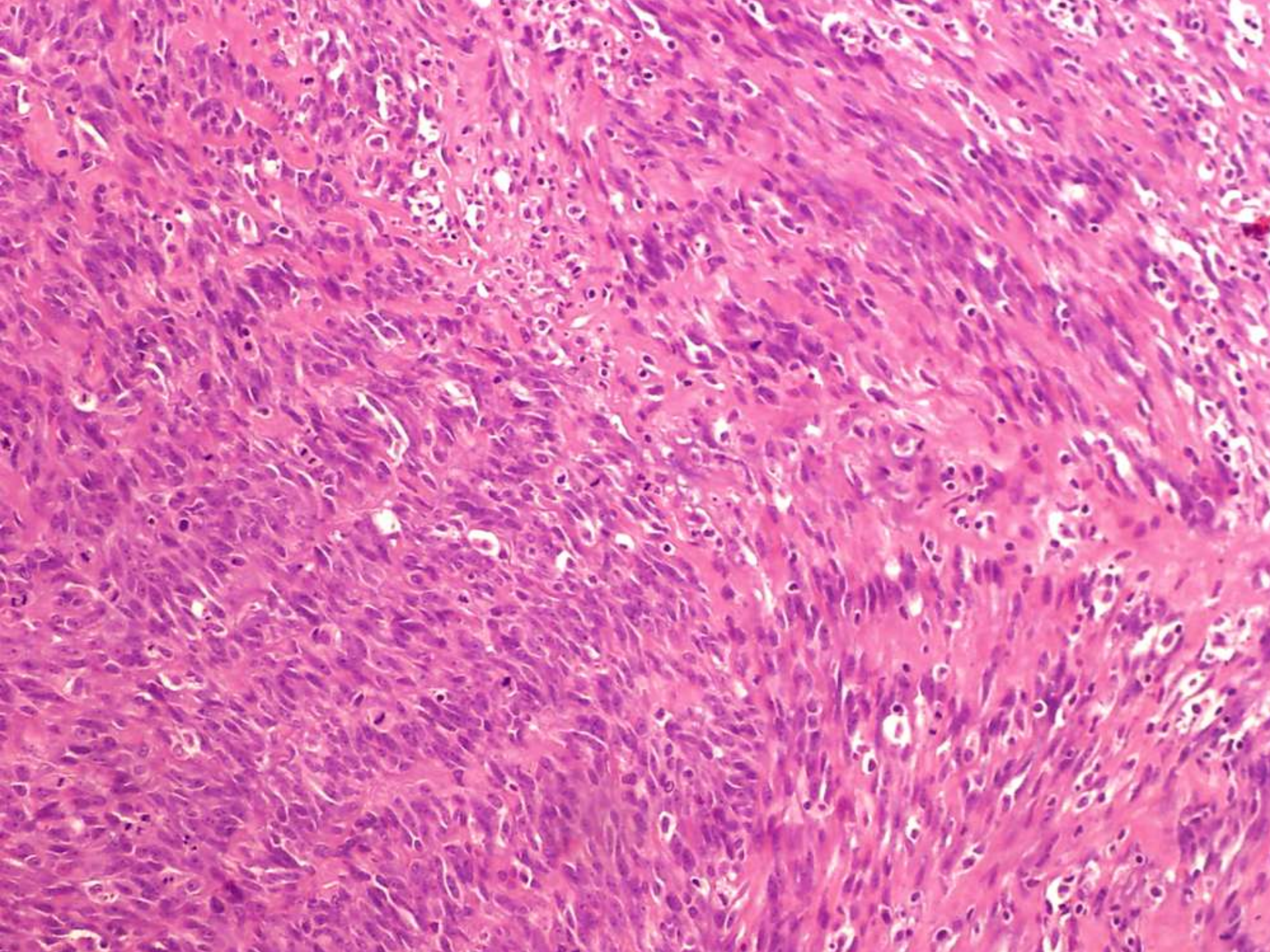

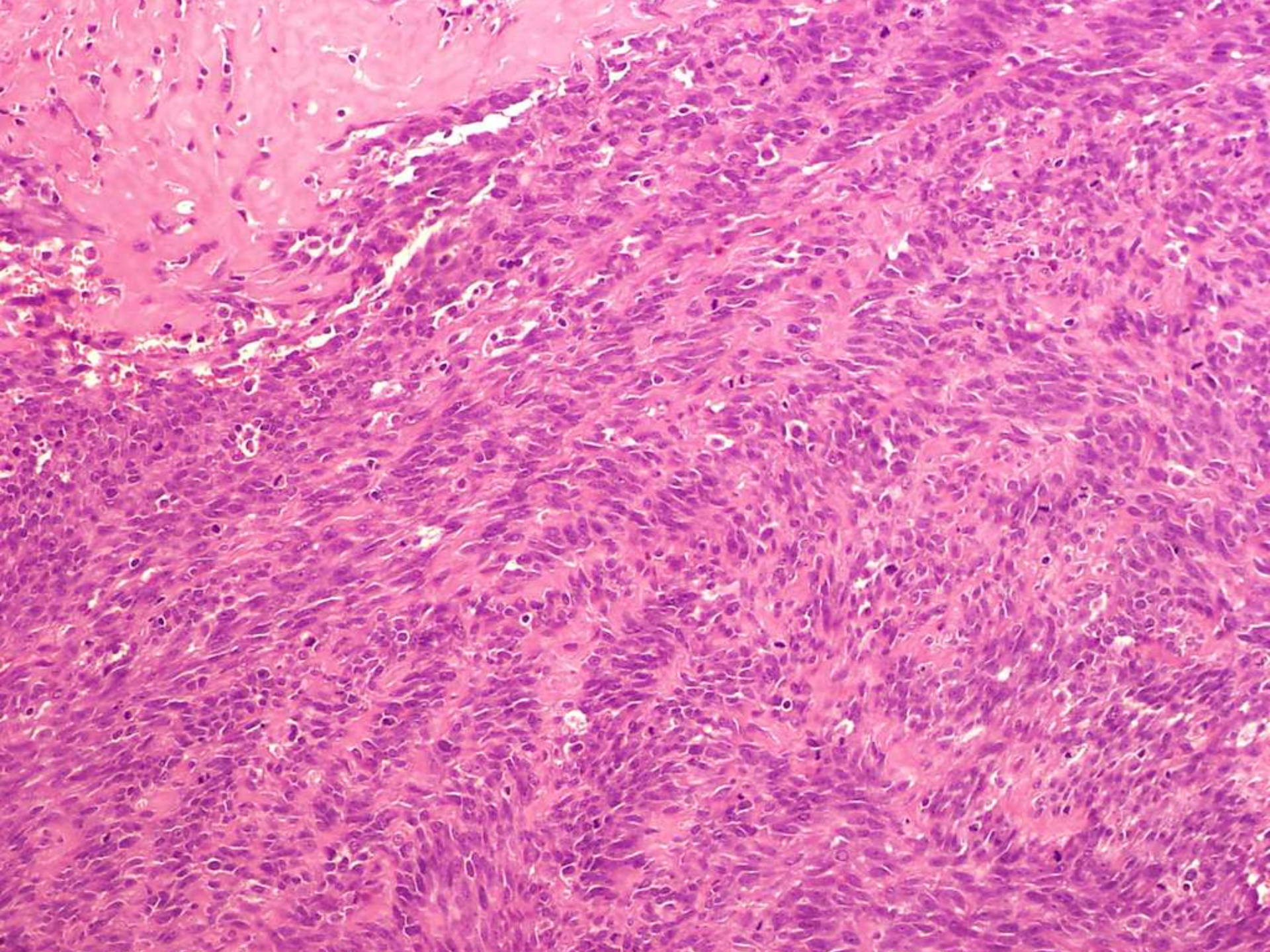

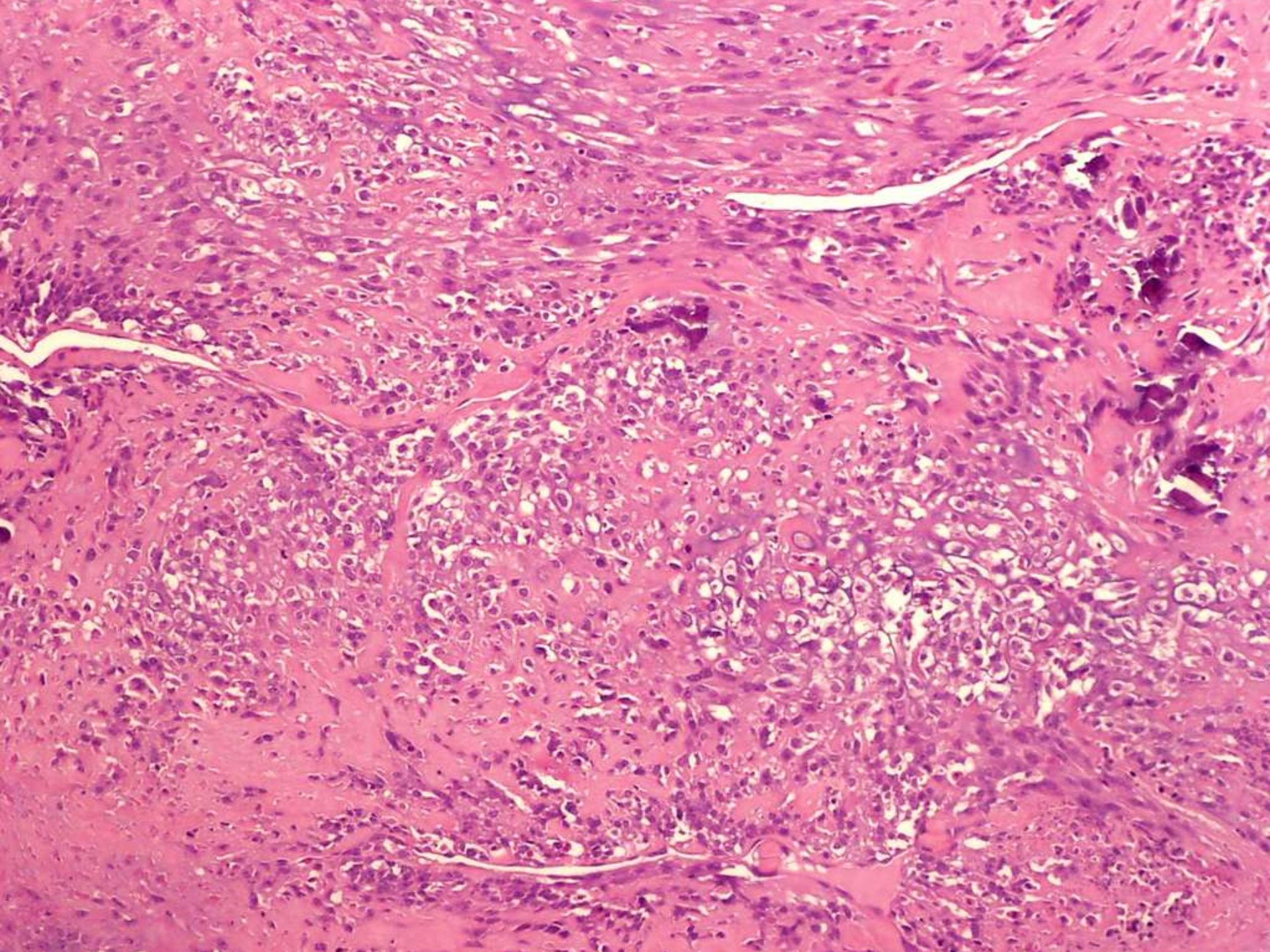

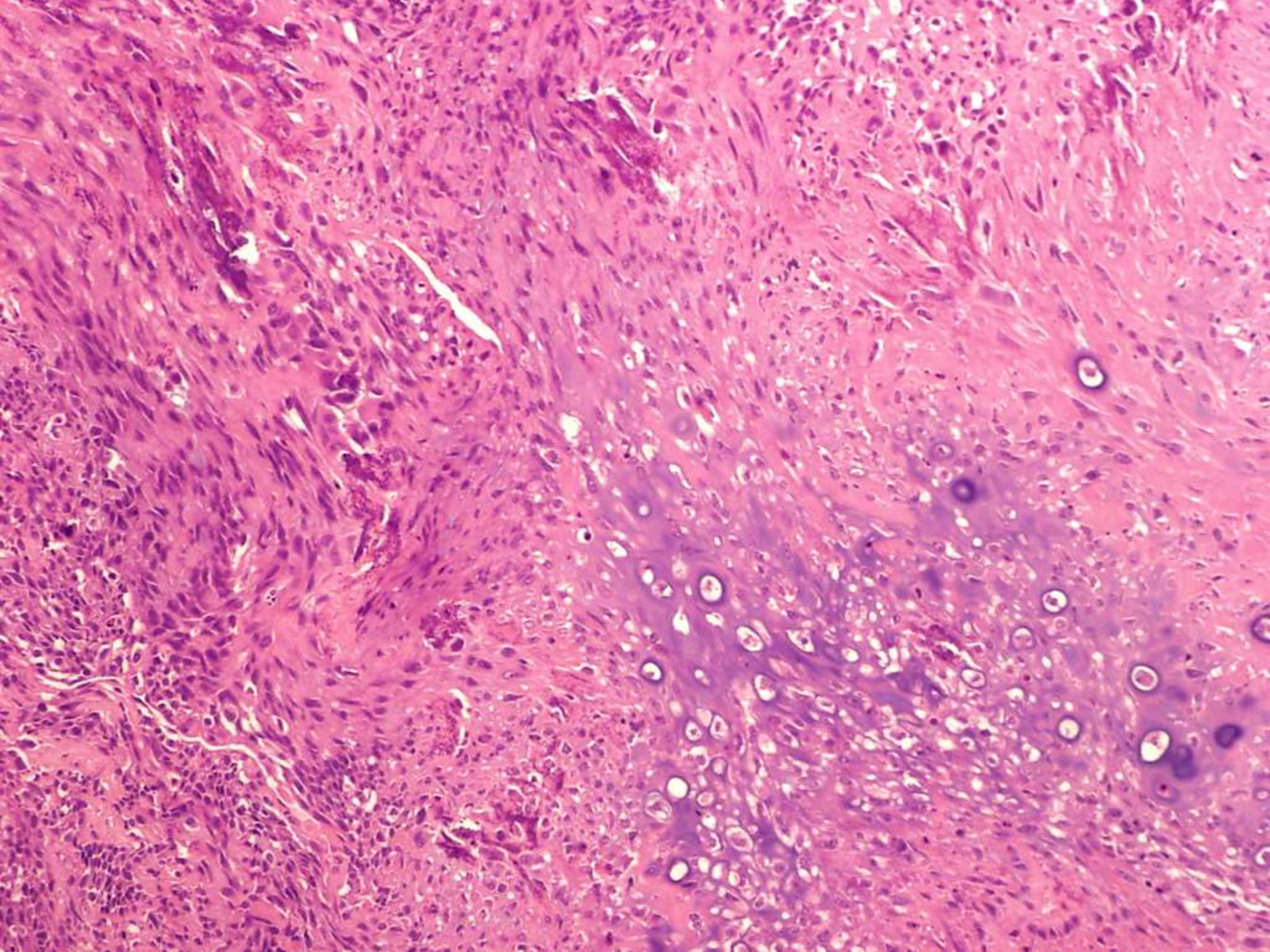

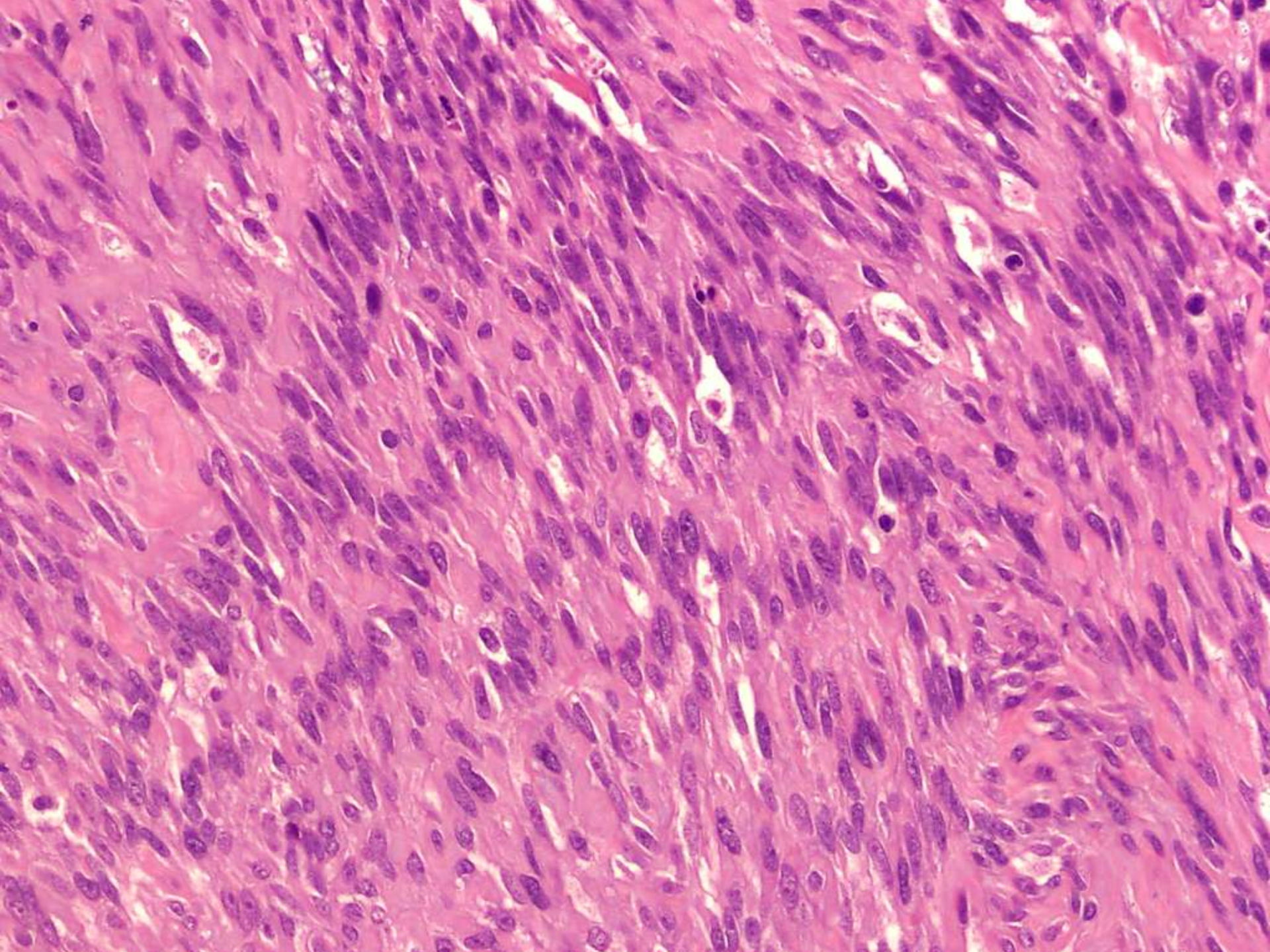

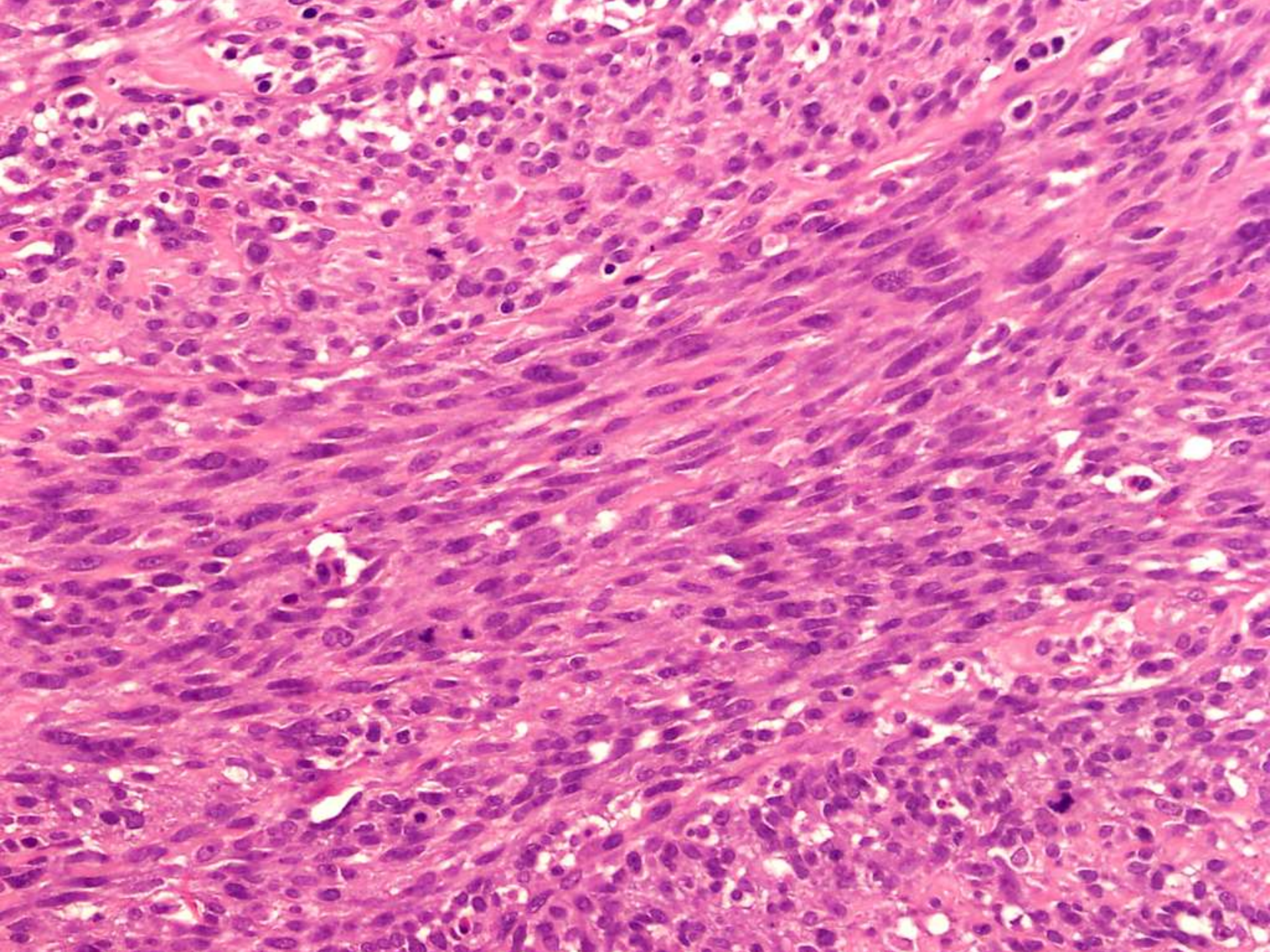

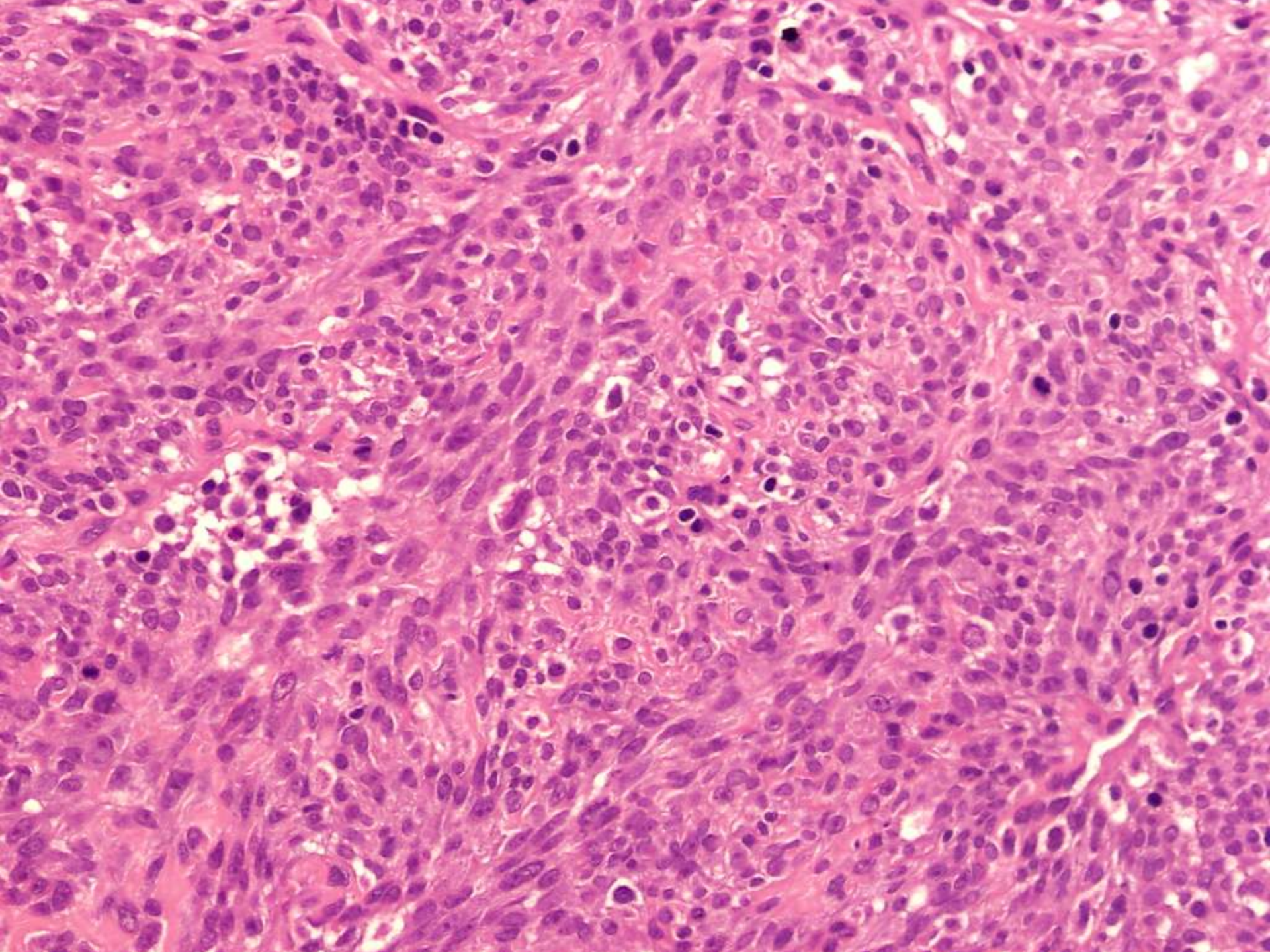

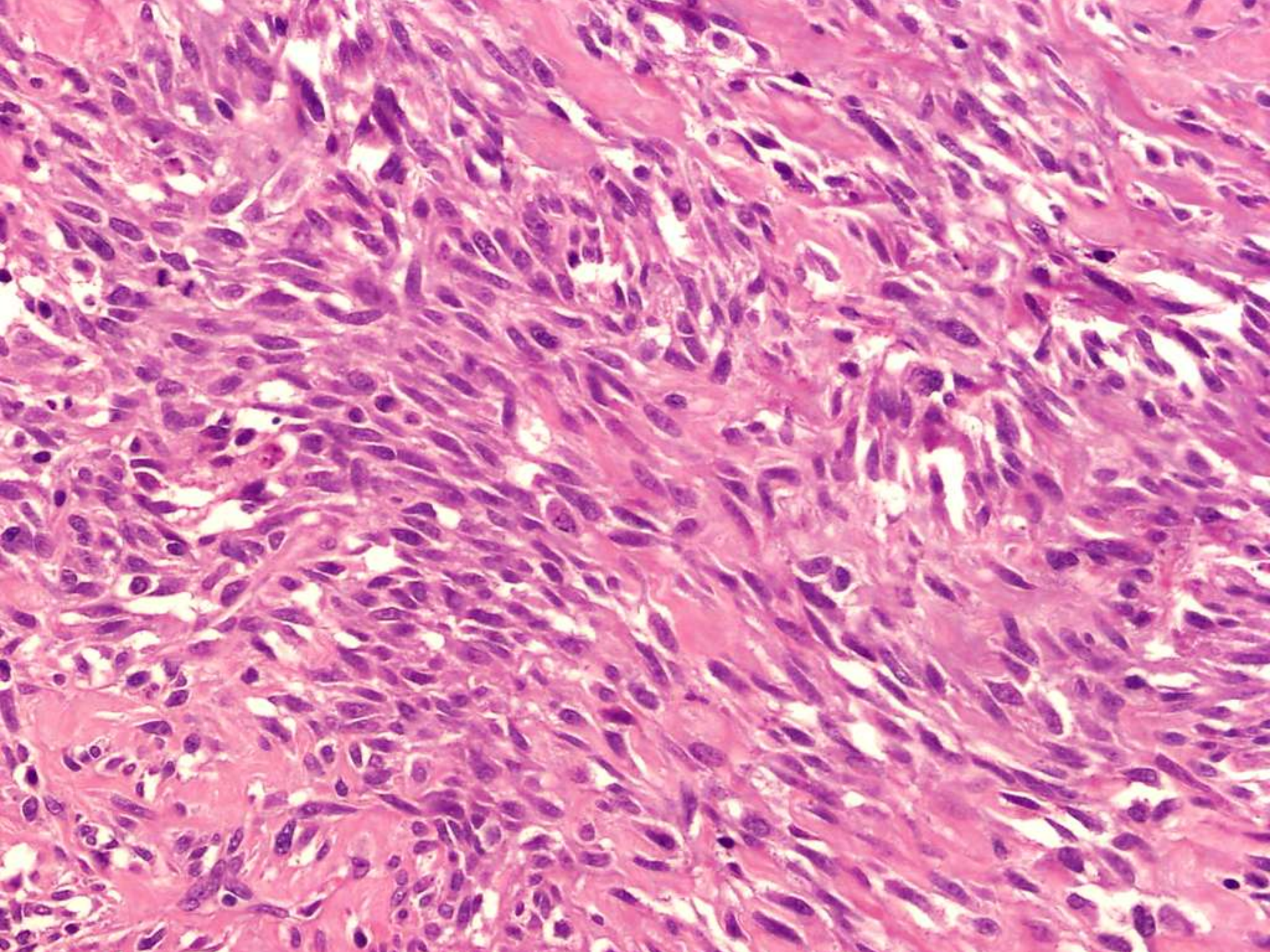

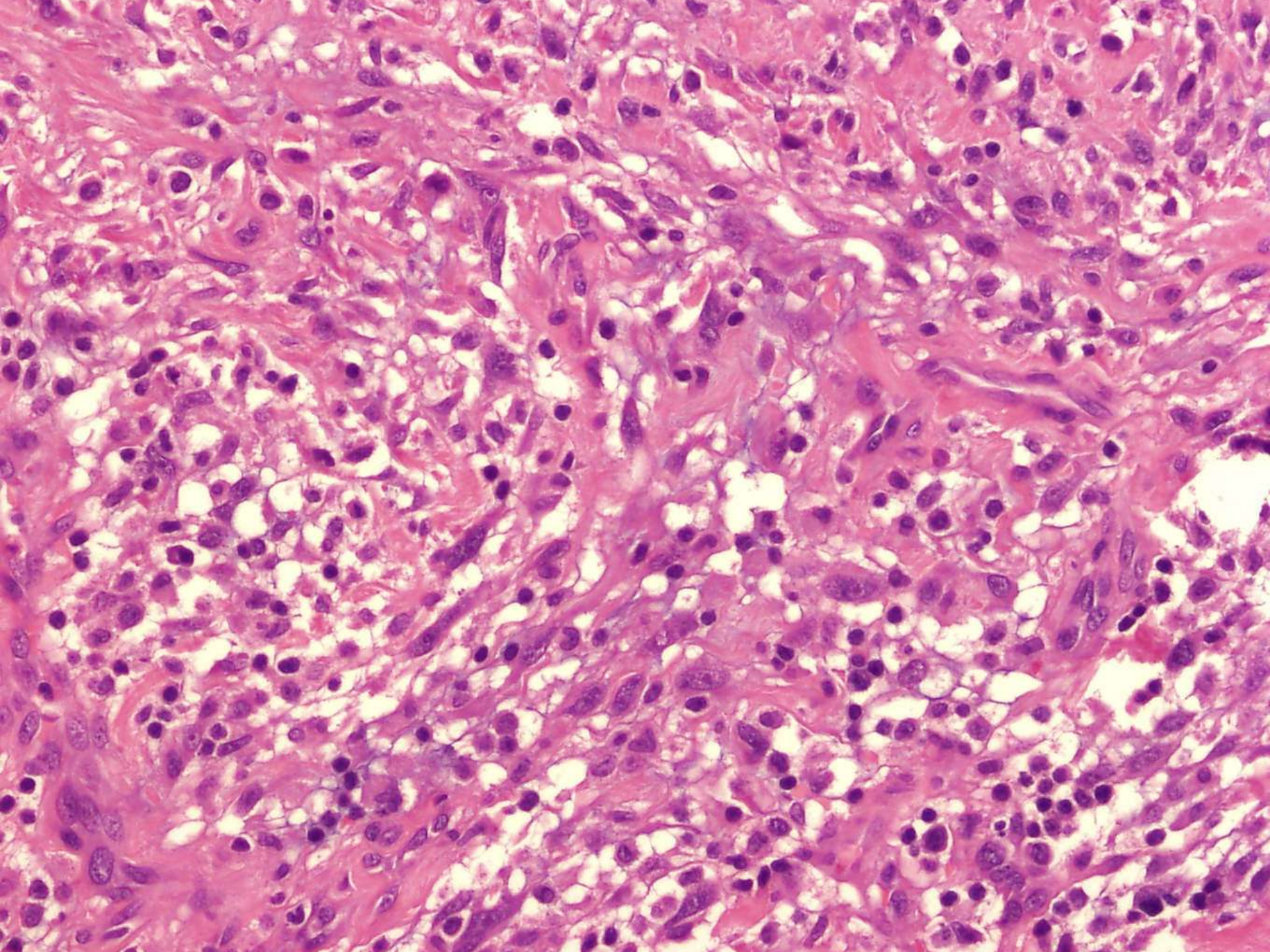

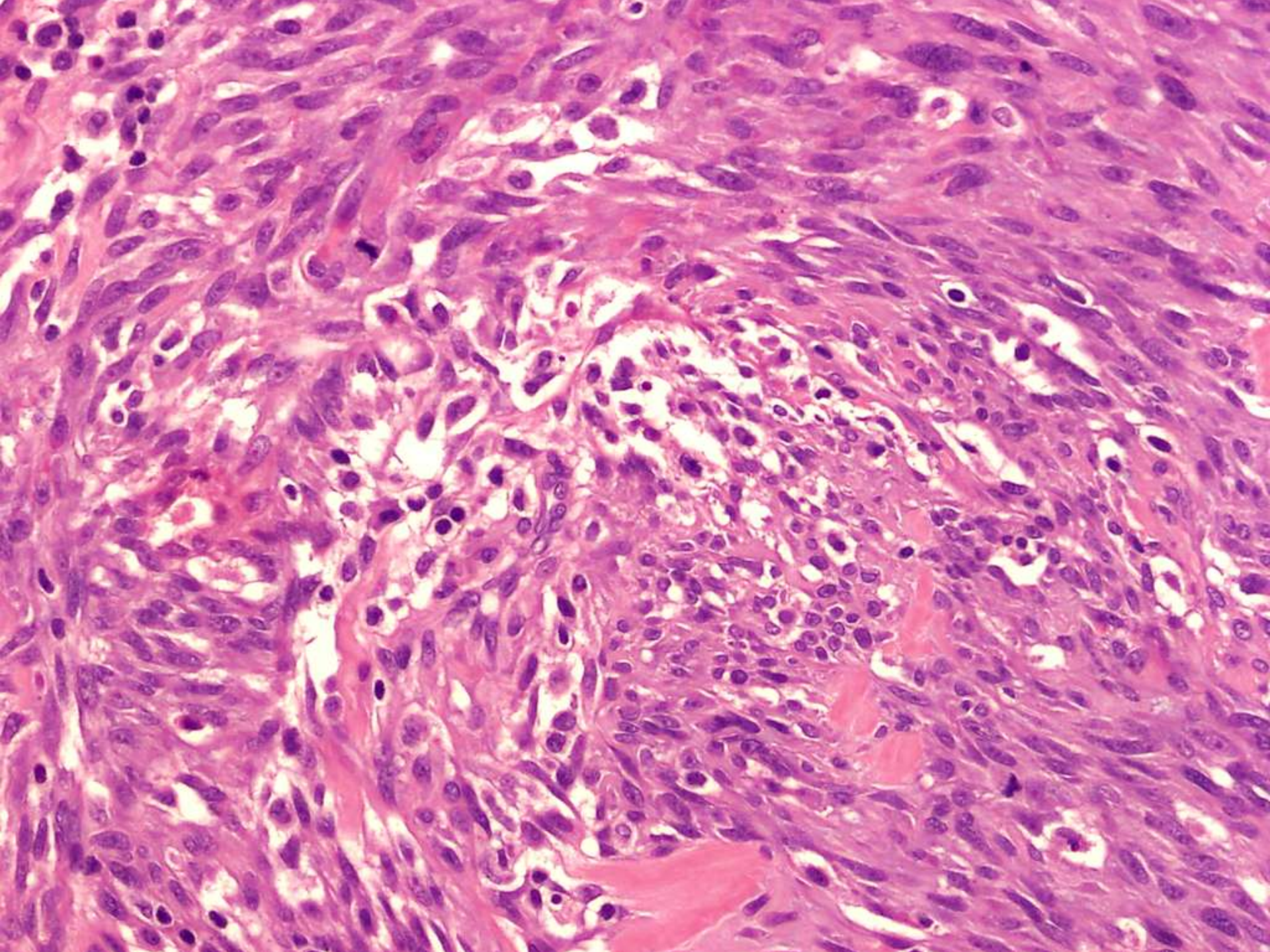

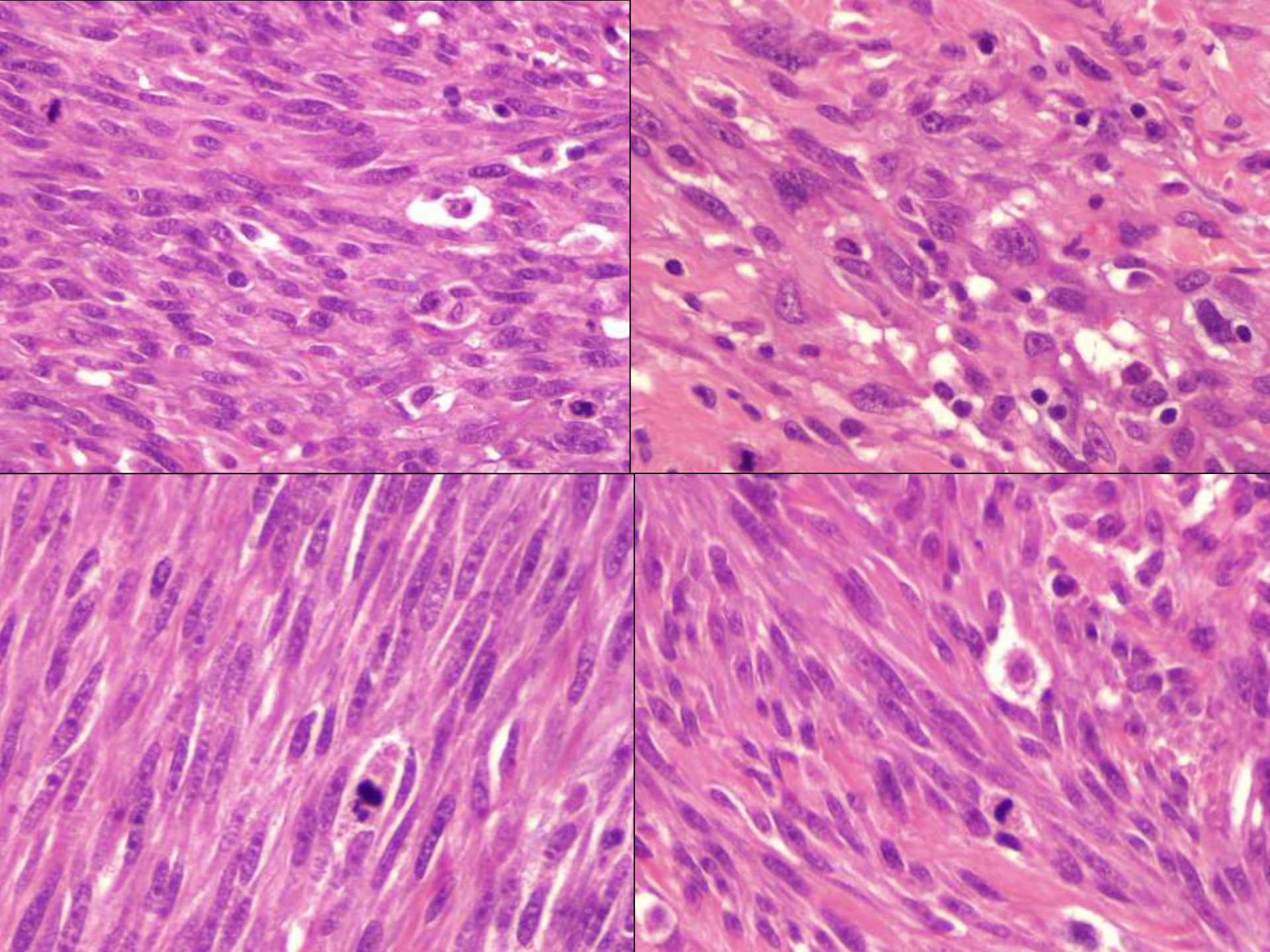

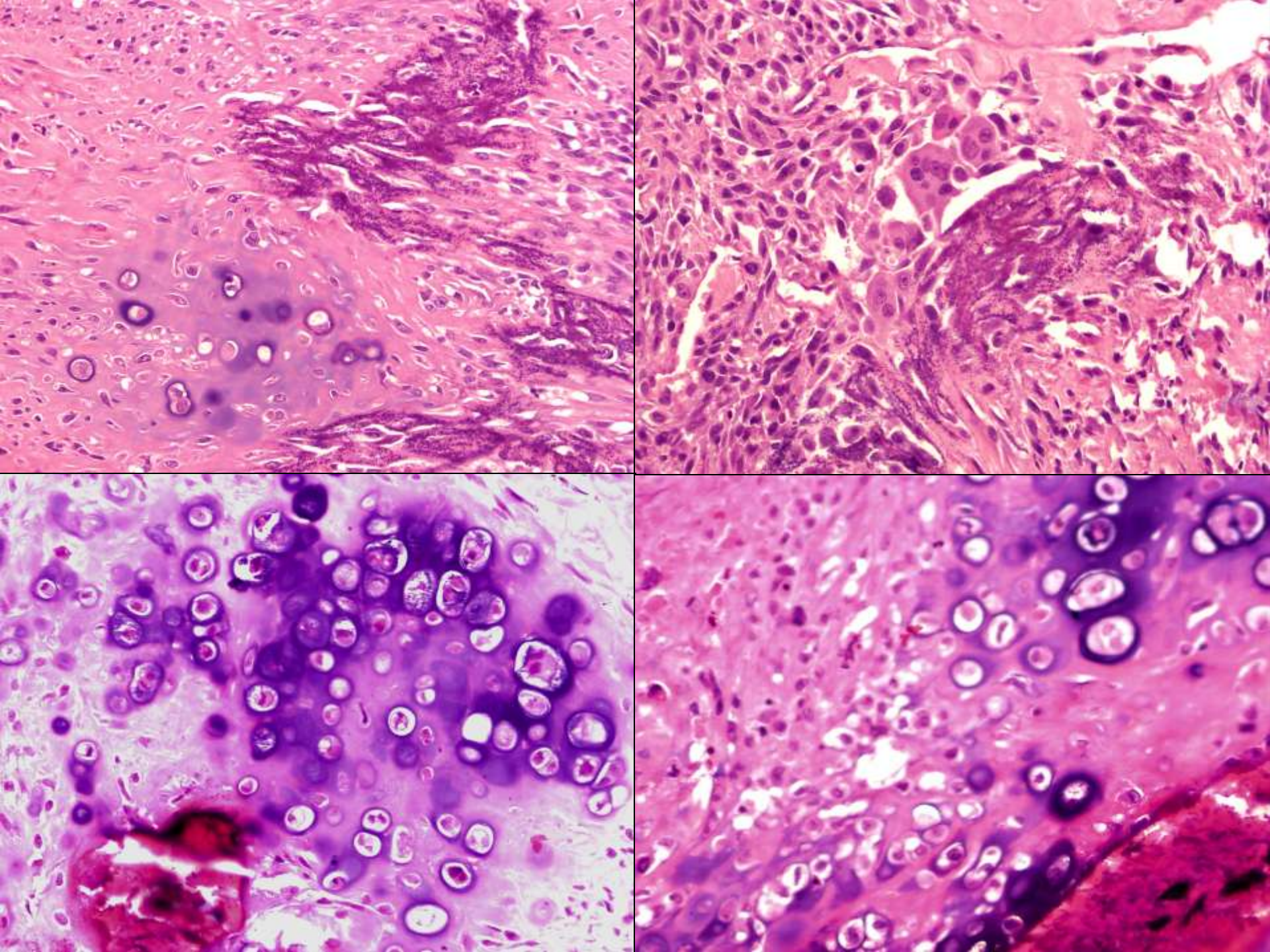

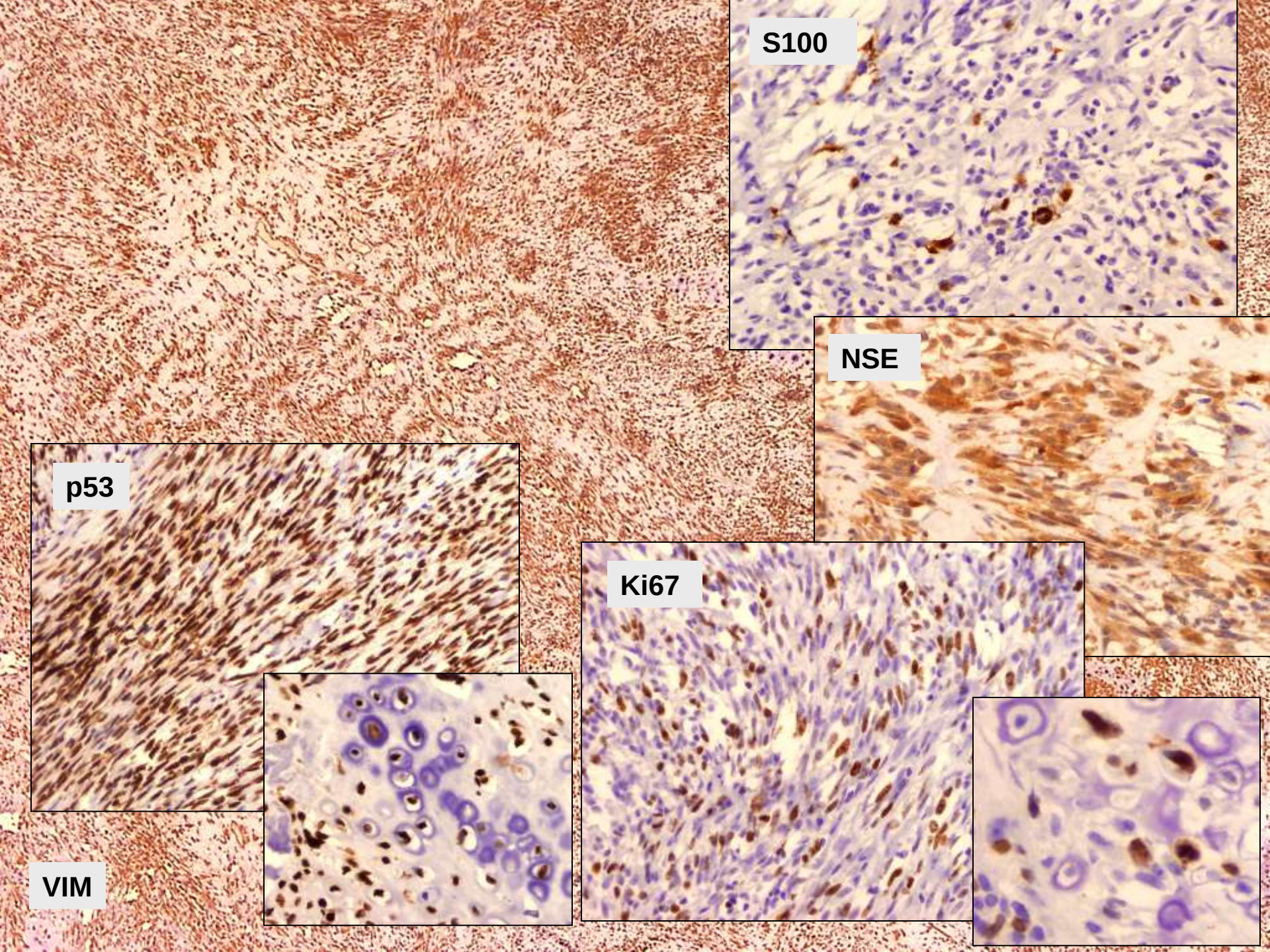
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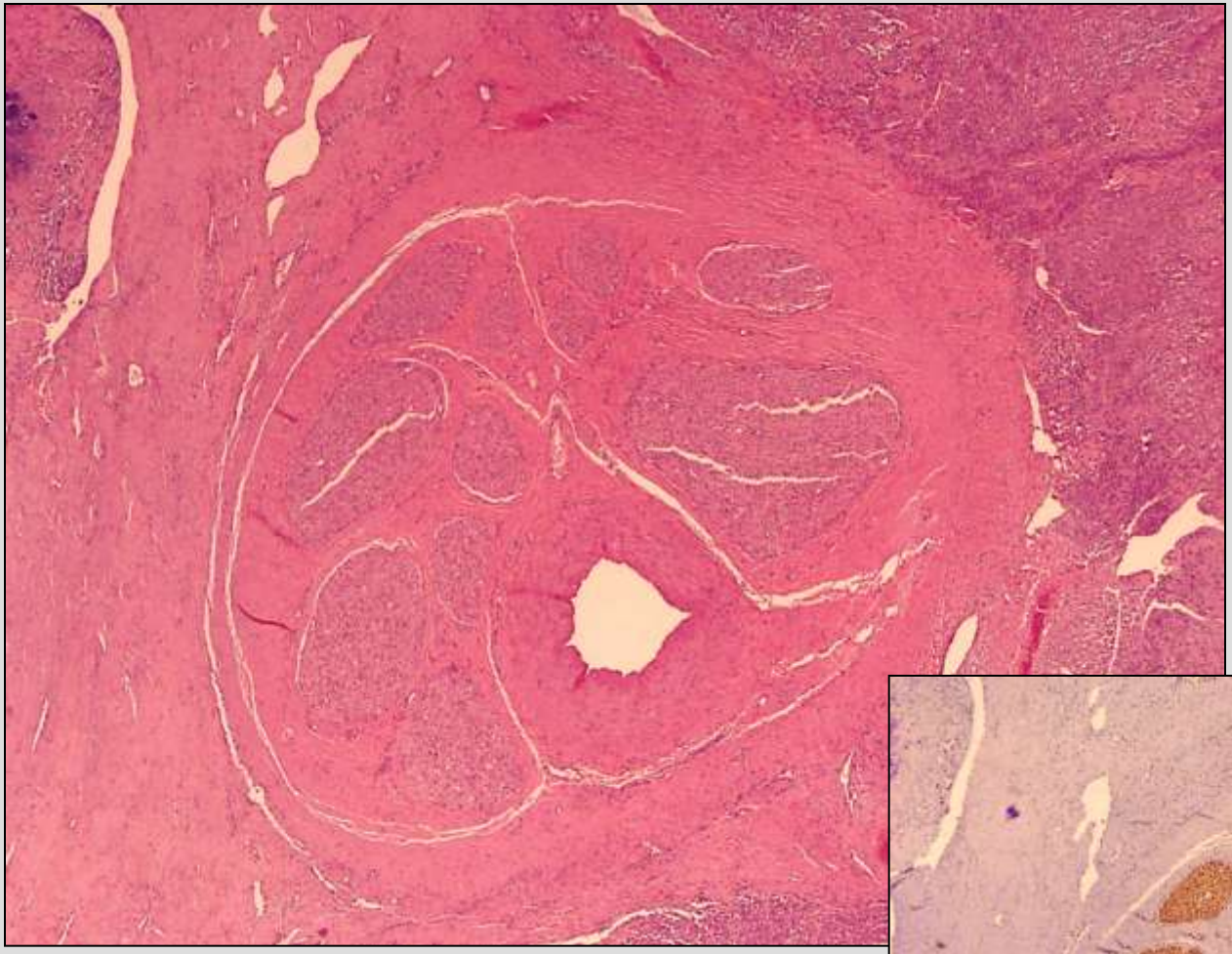


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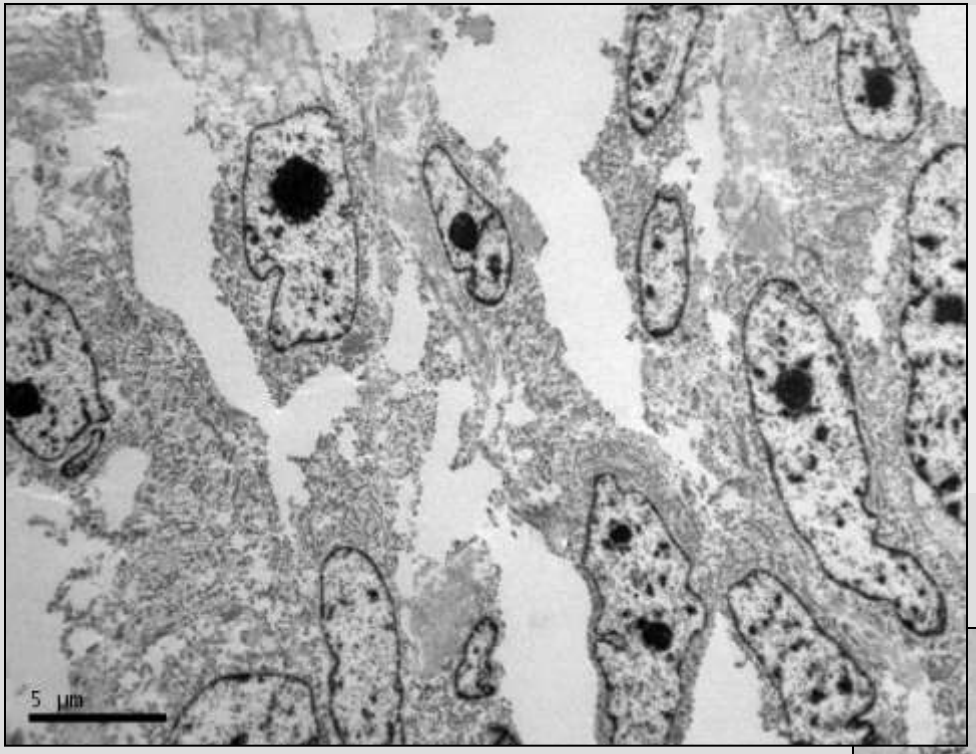
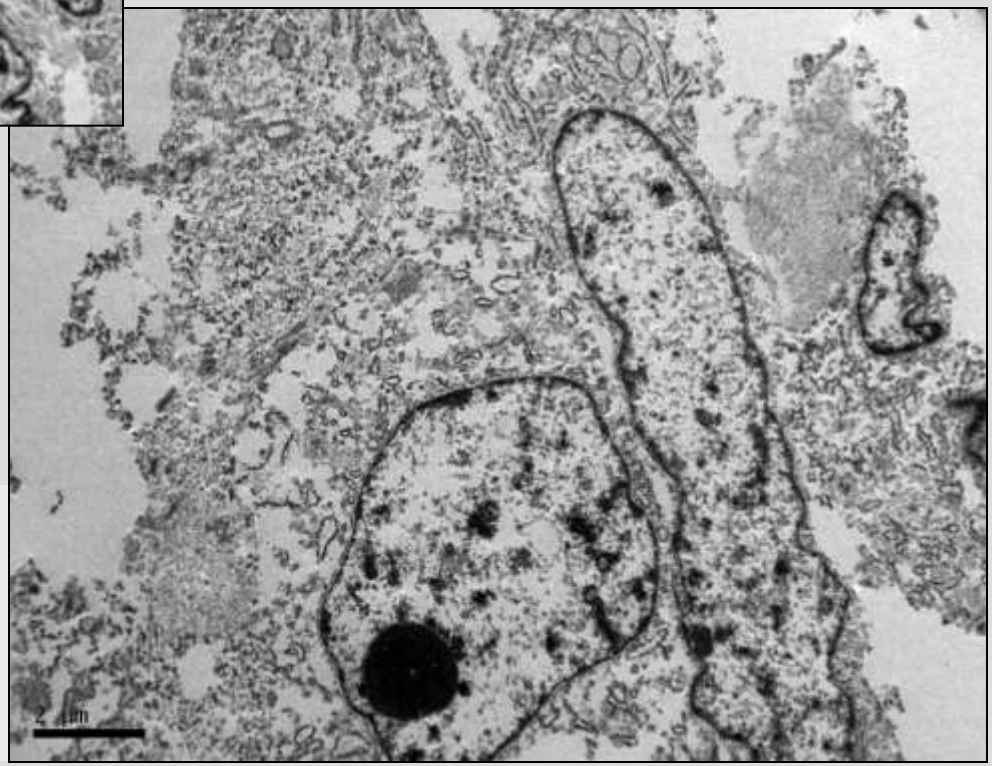


Estudio ultraestructural

- Células fusiformes + RE prominente
- Núcleos irregulares + nucleolos

- No procesos citoplasmáticos
- No uniones intercelulares

- Intenso artefacto de fijación (parafina)



TUMOR MESENQUIMAL FUSOCELULAR SIN DIFERENCIACIÓN EVIDENTE

DIAGNÓSTICO

TUMOR MALIGNO DE LA VAINA DEL NERVIO PERIFÉRICO (TMVNP)*
(sarcoma de alto grado neurogénico, schwannoma maligno) con focos de diferenciación divergente: condro y osteosarcoma.

Probable origen en el nervio infraorbitario o alguna rama del mismo, sin descartar la posibilidad de malignización de un schwannoma benigno previo.

*Se prefiere el término TMVNP por cuanto se identifica con cualquier célula de la vaina neural, no solo la de Schwann sino también fibroblastos y células perineurales. La malignización de schwannomas es excepcional y el término schwannoma maligno la podría sugerir

TMVNP
(schwannoma maligno, neurofibrosarcoma)

•Sarcoma agresivo de origen neural asociado a un nervio periférico, a un neurofibroma (NF) o con características de diferenciación neural.

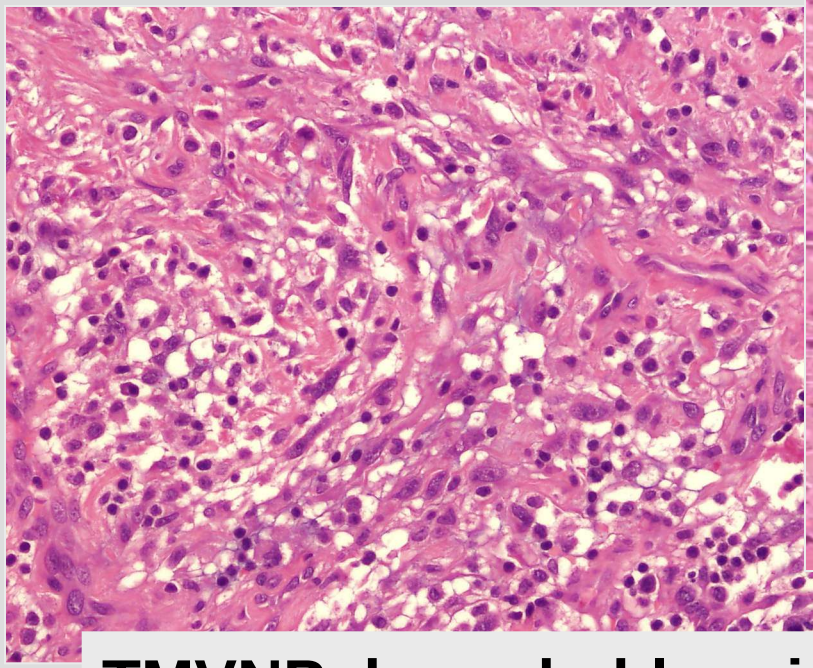
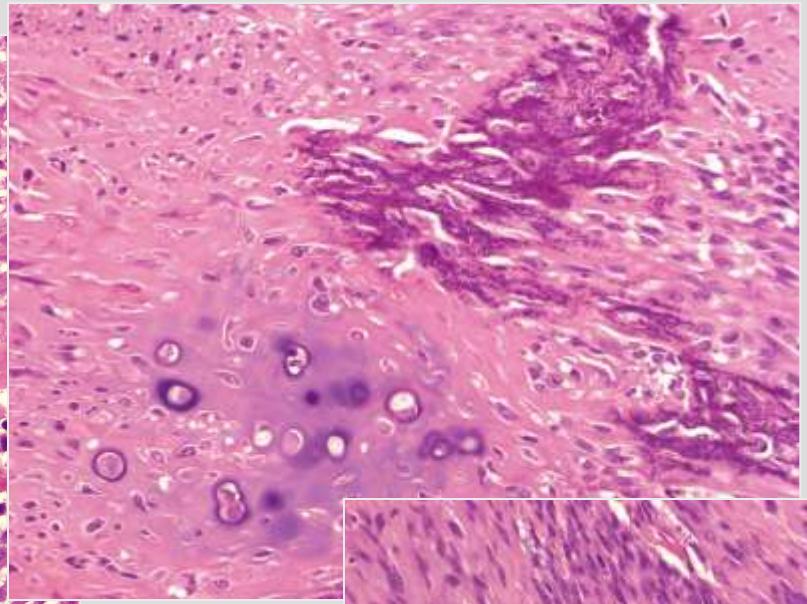
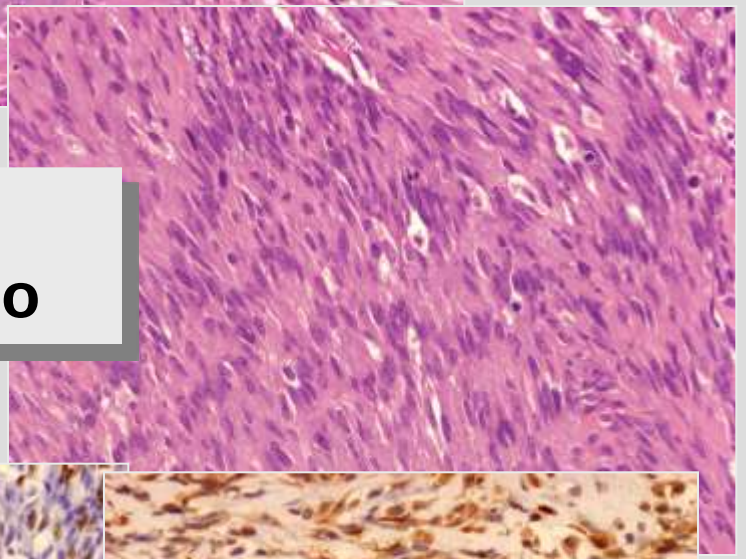
•Morfológicamente se exige la presencia de células fusiformes atípicas de aspecto neural y la demostración de la relación con un tronco nervioso o un NF previo.

•IHQ:
- 50% de TMNP son negativos para proteína S-100
- CD57, mielina básica y prot. glial acídica, variables
- citoqueratina ocasionalmente positiva.

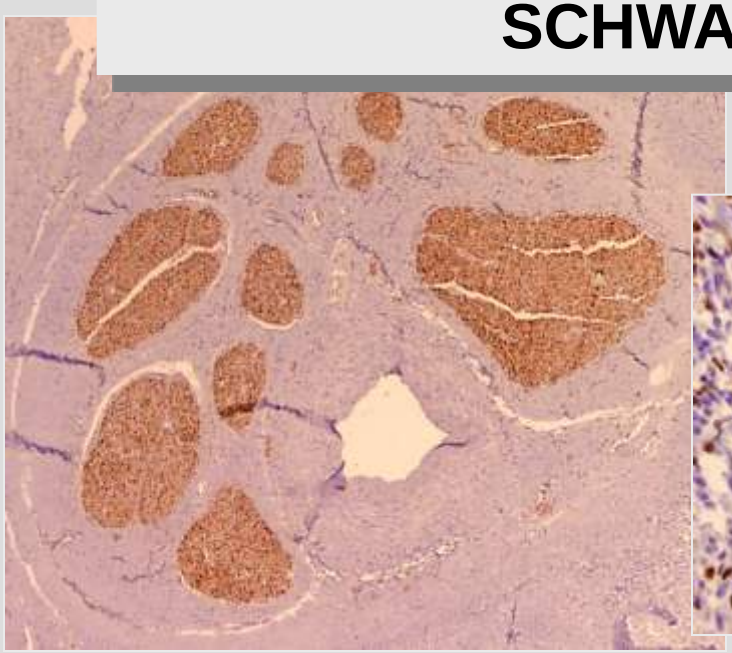
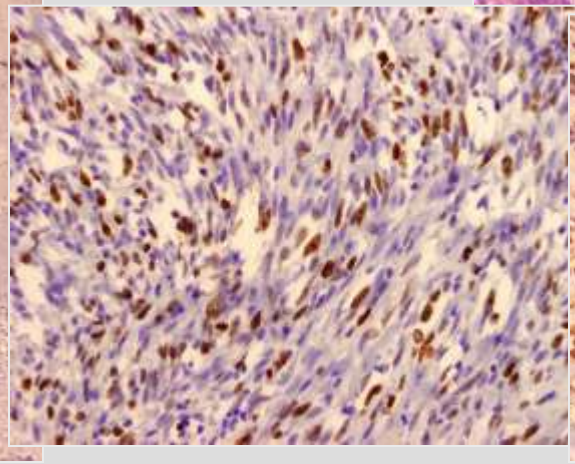
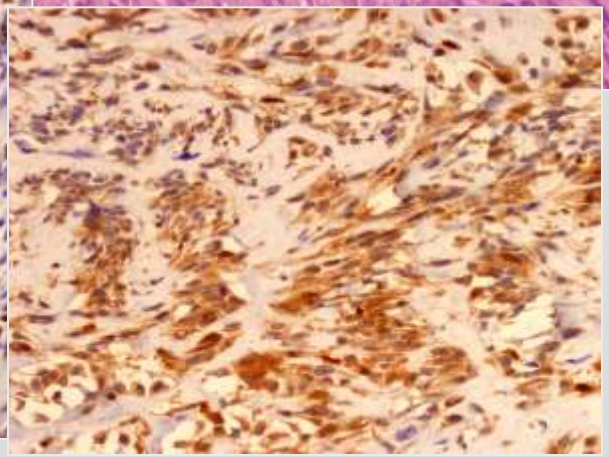
TMVNP Características morfológicas

- Patrón con áreas de distinta densidad celular (“marmoleado”).
- Zonas de estroma mixoide o colagenizado poco celular.
- Haces irregulares de células más o menos onduladas, de núcleos alargados, a veces formando empalizadas.
- Focos de elementos heterólogos, generalmente cartílago y hueso malignos, rabdomioblastos (tumor “triton” maligno) o elementos glandulares o escamosos.
- Atipia variable y abundantes mitosis atípicas.
- Focos de hemorragia y necrosis habituales.

En los raros casos de malignización de schwannomas, las zonas de células epitelioides, las empalizadas nucleares y los focos de condrosarcoma y osteosarcoma son más frecuentes.

TMVNP de probable origen en un SCHWANNOMA previo

SARCOMAS DE CABEZA Y CUELLO

10% SPB en C&C

FRECUENCIA
Histiocitoma fibroso maligno
Fibrosarcoma (20%)
Rabdomiosarcoma (15%)
TMVNP (13%)
Angiosarcoma (13%)

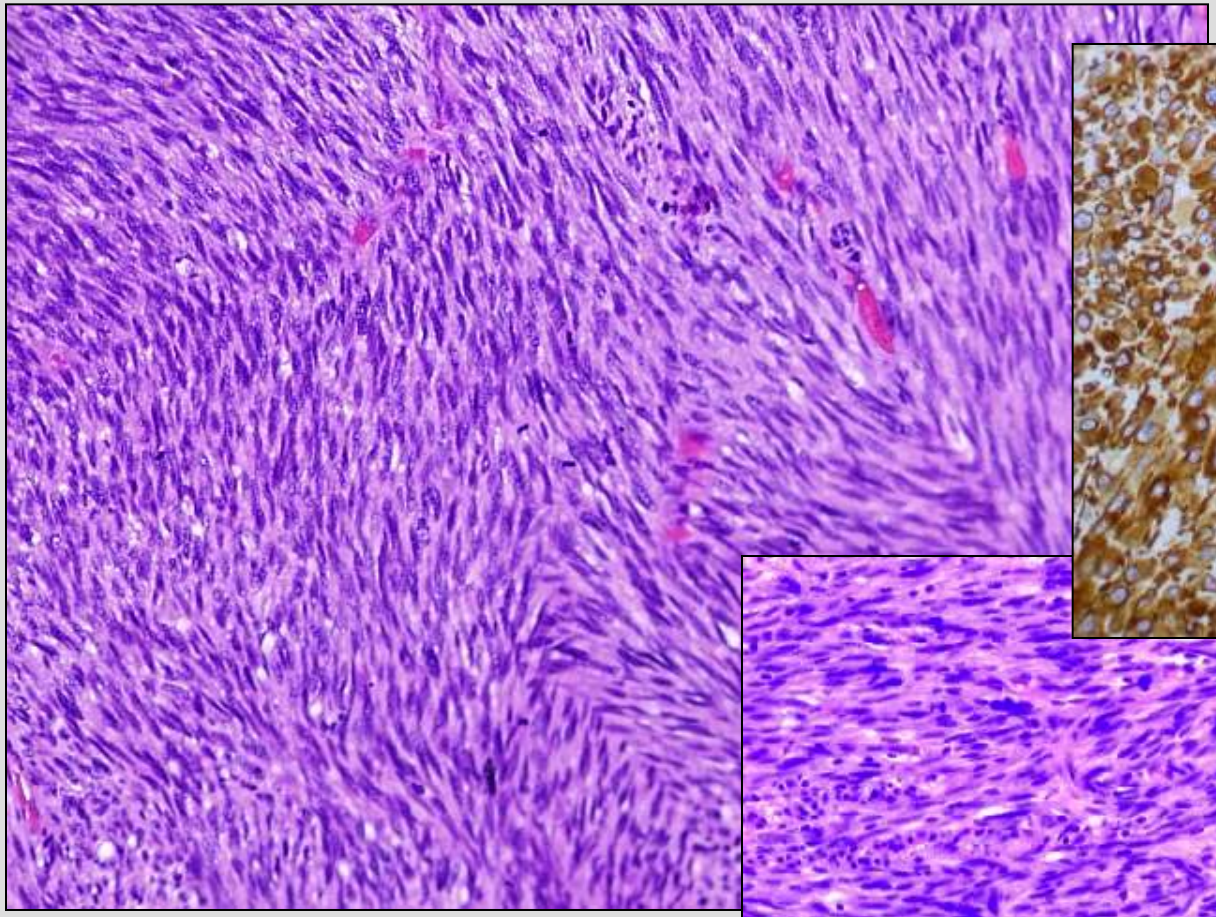
LOCALIZACIÓN
Cuello y parótida (20%)
Cara y frente (18%)
Maxilar y paladar (13%)
Cuero cabelludo (12%)
Mandíbula (11%)
Senos paranasales (7%)
Cavidad oral (5%)
Laringe (2%)

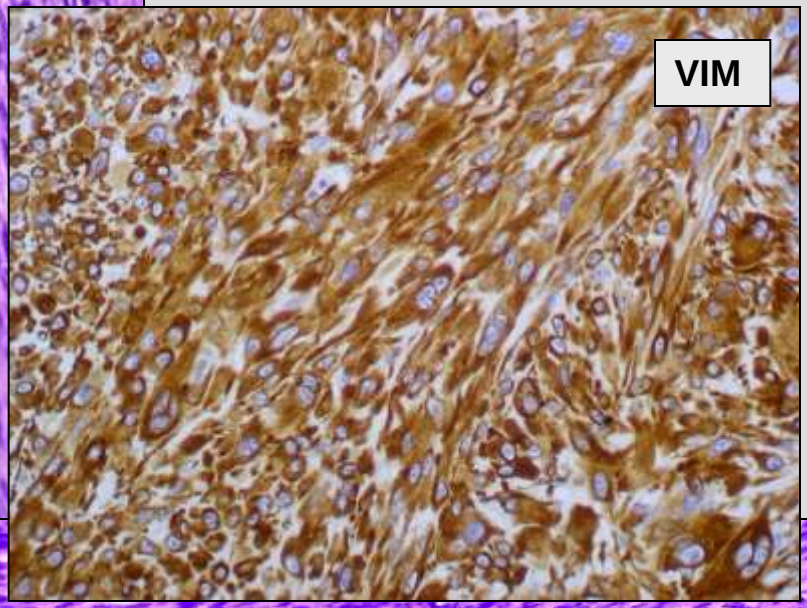
DIAGNÓSTICO DIFERENCIAL
Otros sarcomas
Carcinoma sarcomatoide
Melanoma fusocelular

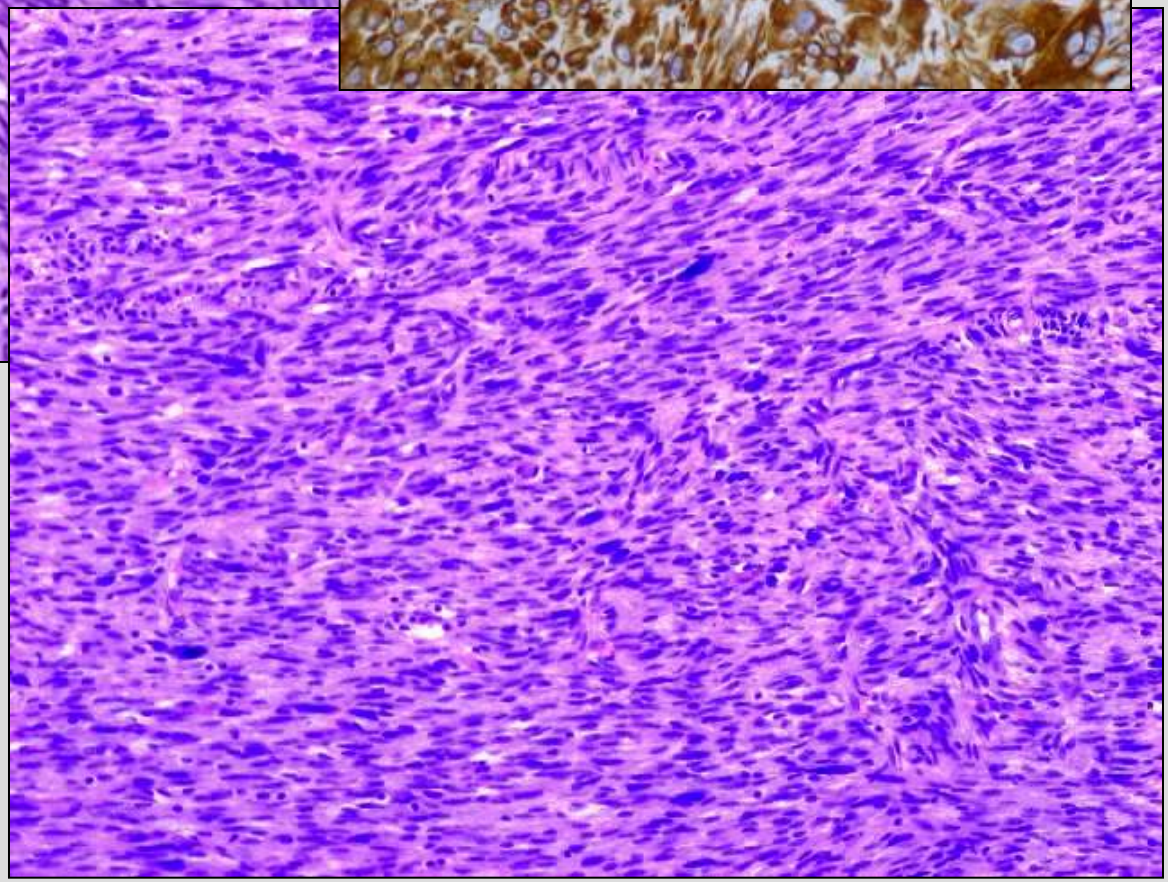
PRONÓSTICO
85% resecables ... ¿anatomía?
Tipo tumoral
Tamaño del tumor
Supervivencia 70 % en 5 años
QRT adyuvantes

SPB/hueso, melanomas y carcinomas sarcomatoides diagnosticados en el Departament d'AP del HVH (Barcelona). Incidencia en C&C y zona sinonasal (2000-2010).

Tumores	***total***	***C&C***	***NS/NF***
Leiomiosarcoma	98	2	0
Fibrohistiocitoma maligno	80	3(2+N1)	1
Osteosarcoma	69	6	2
Condrosarcoma	54	3	1
Rabdomiosarcoma	51	3	1
Carcinoma sarcomatoide	32	5	0
Sarcoma sinovial	31	1	0
Cordoma	28	11	2
Angiosarcoma	28	2	0
Fibrosarcoma	23	2	1
HP / TFS	17	6	1 (HPN)
TMVNP	13	4	1
Condrosarcoma mesenquimal	7	2	1
Melanoma (fusocelular)	5	0	0
Total	***536***	***50*** *(9.5%)*	***11*** *(15.7%)*



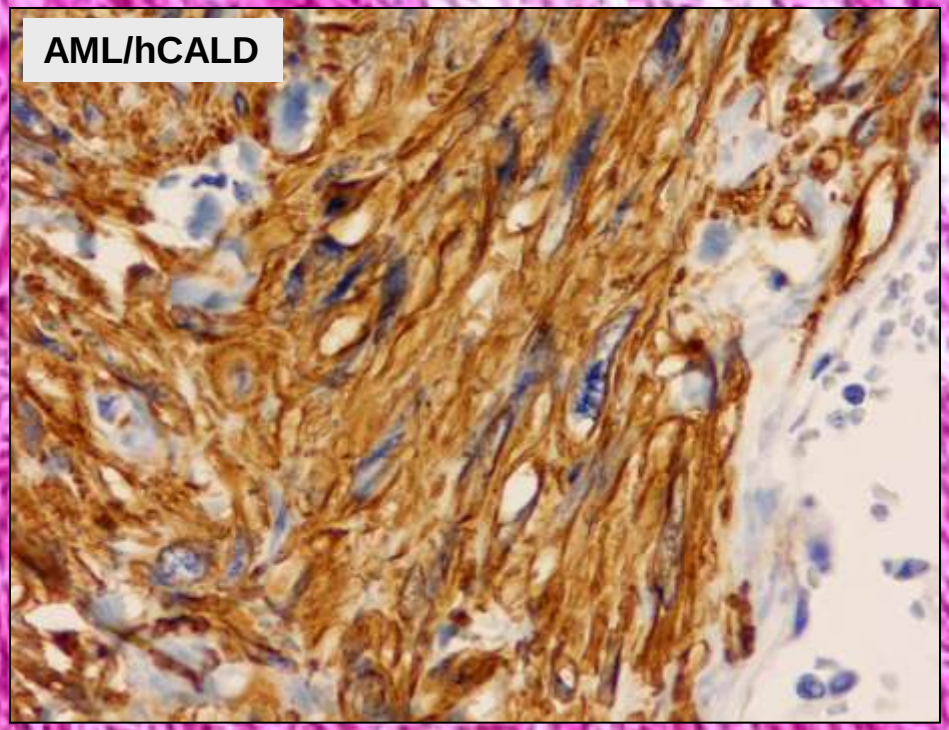




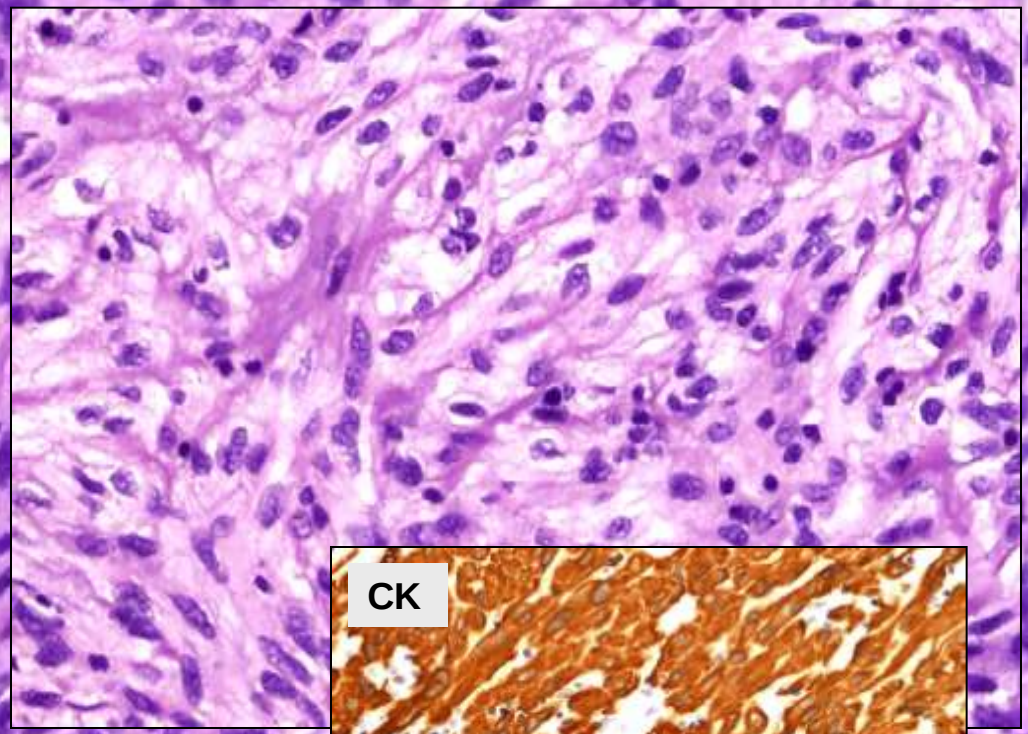
FIBROSARCOMA

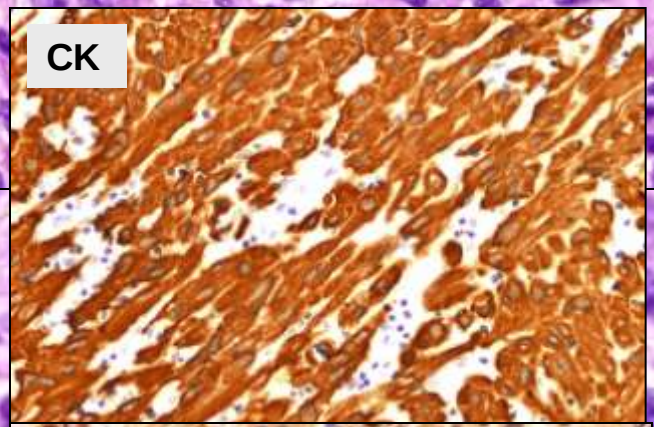
FHM (S. PLEOMÓRFICO INDIFº de AG)

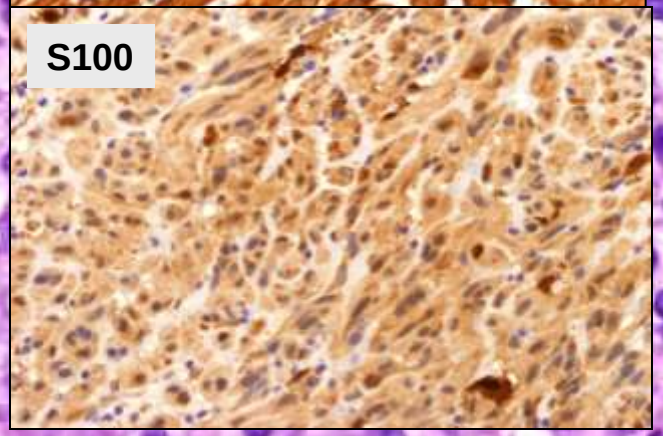
LEIOMIOSARCOMA



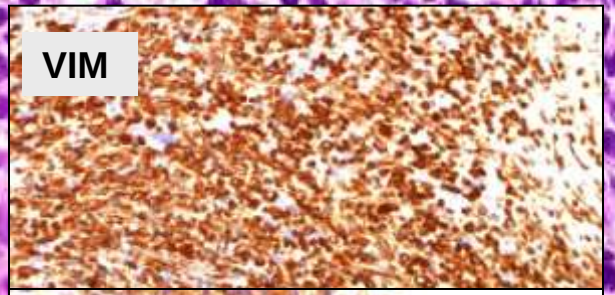
CORDOMA

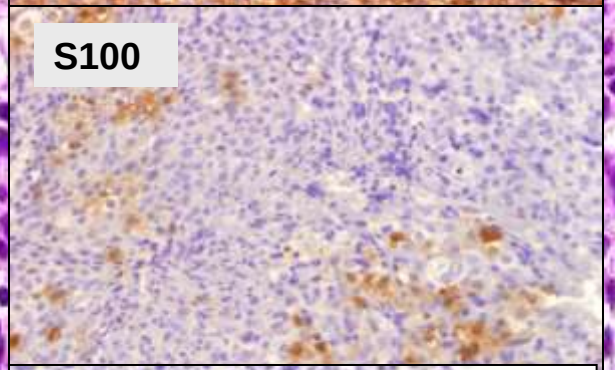


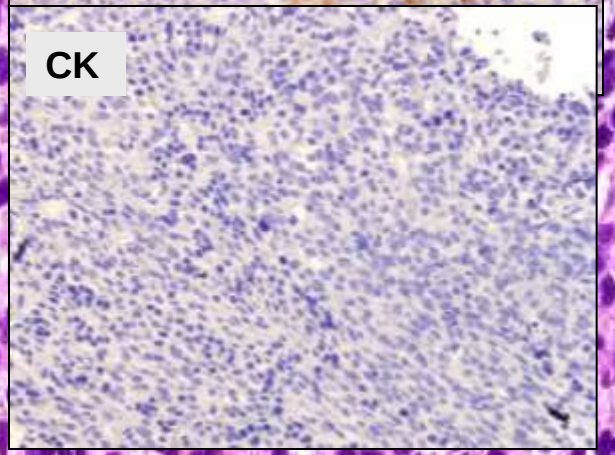




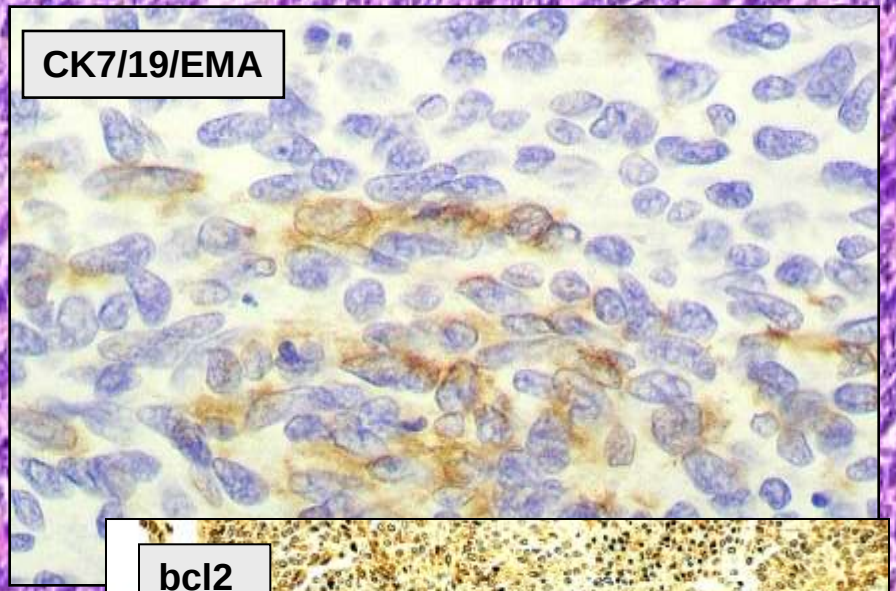
CONDROSARCOMA MESENQUIMAL

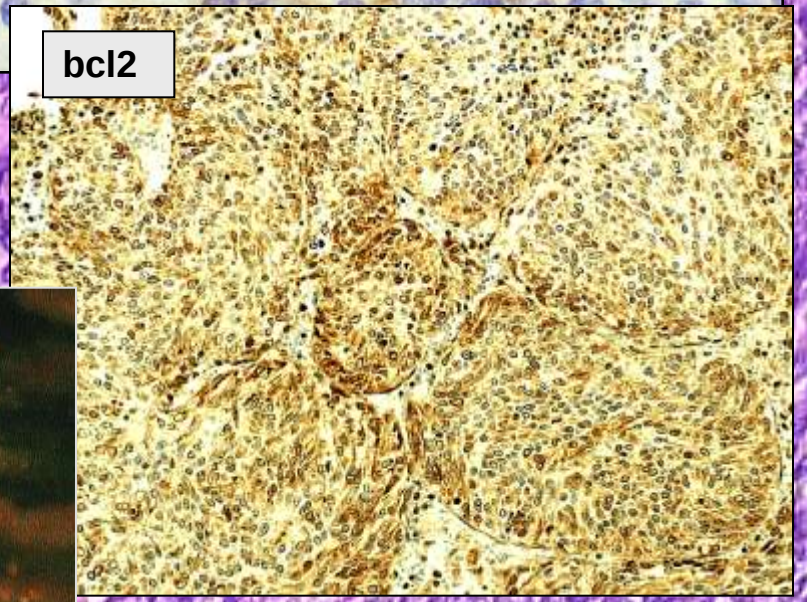


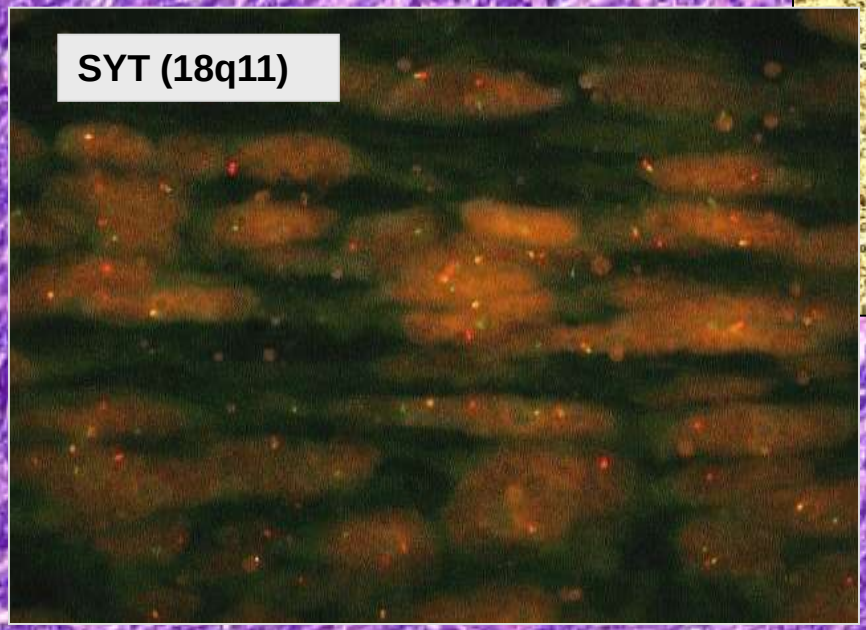




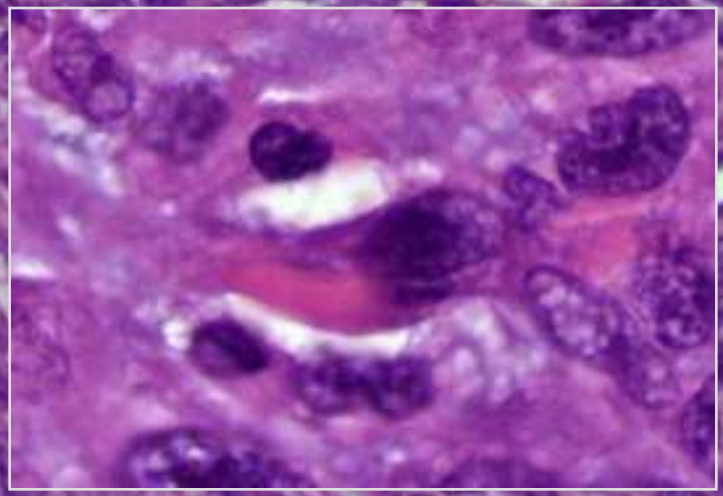
SARCOMA SINOVIAL

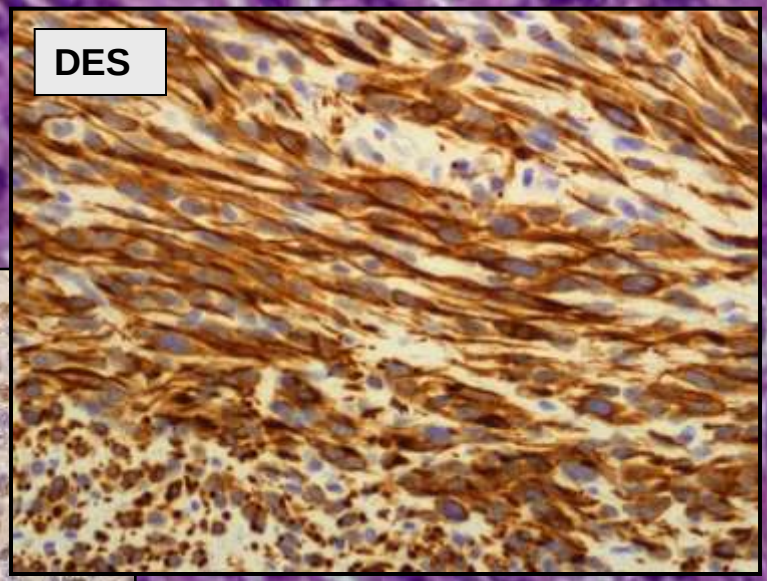


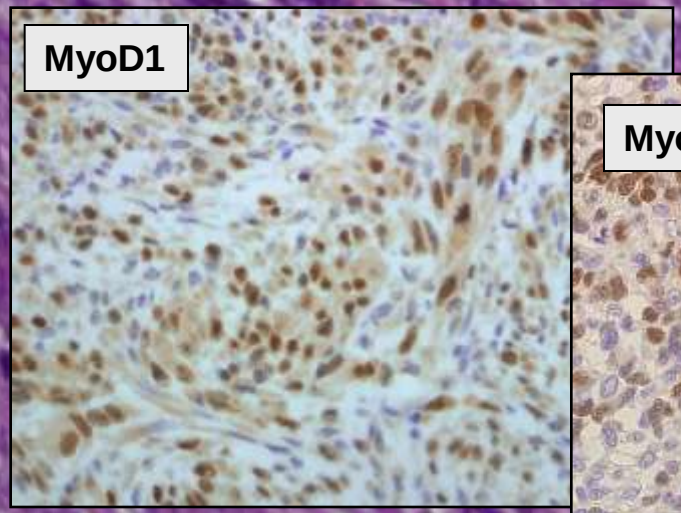


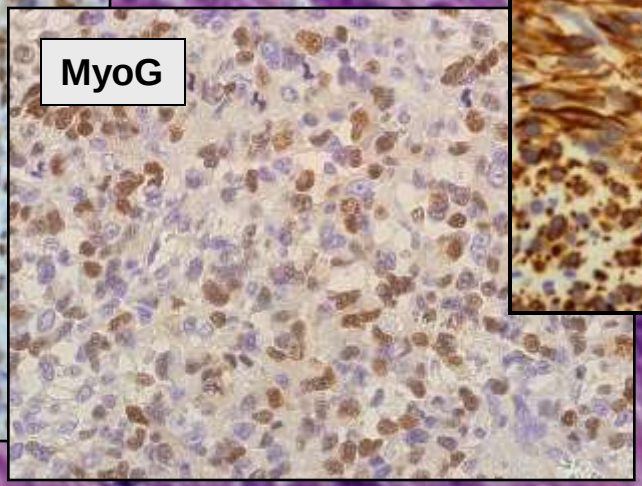


RMS EMBRIONARIO (FUSOCELULAR)

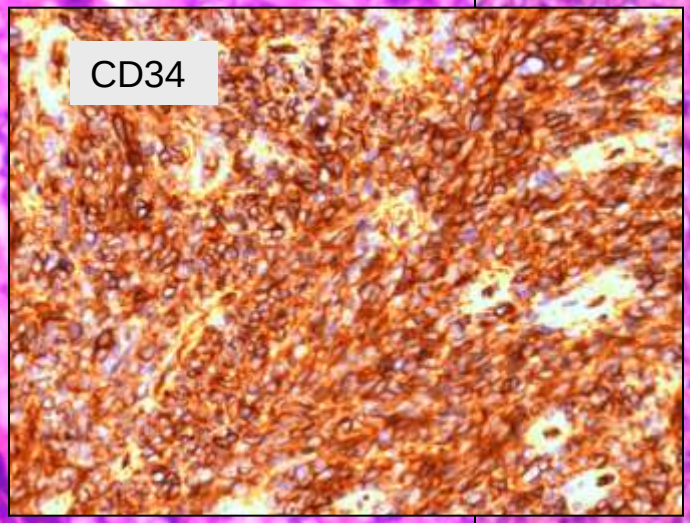


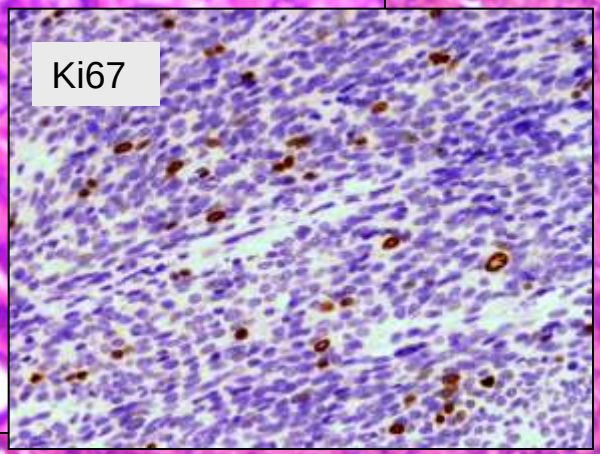




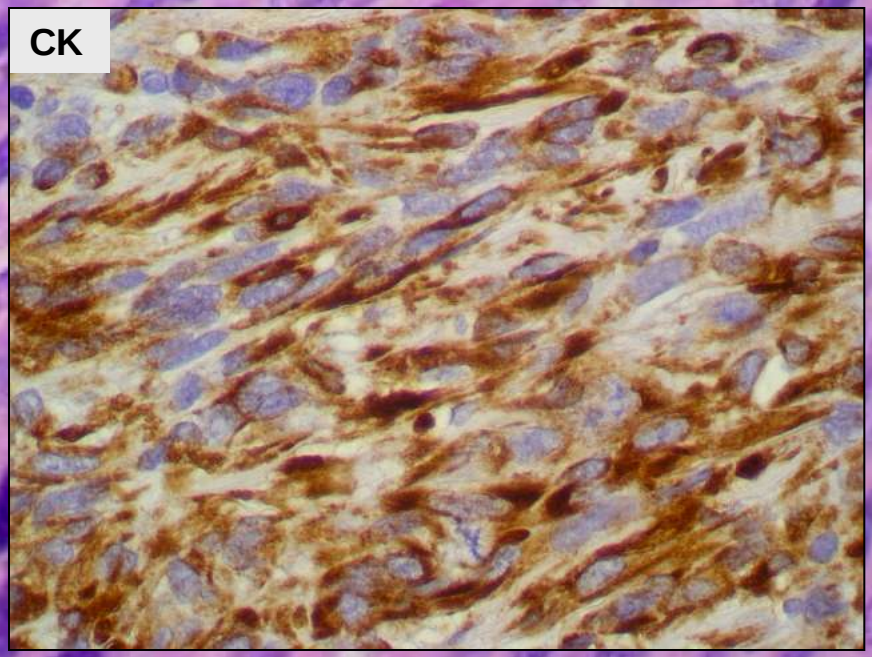


TUMOR FIBROSO SOLITARIO (MALIGNO)

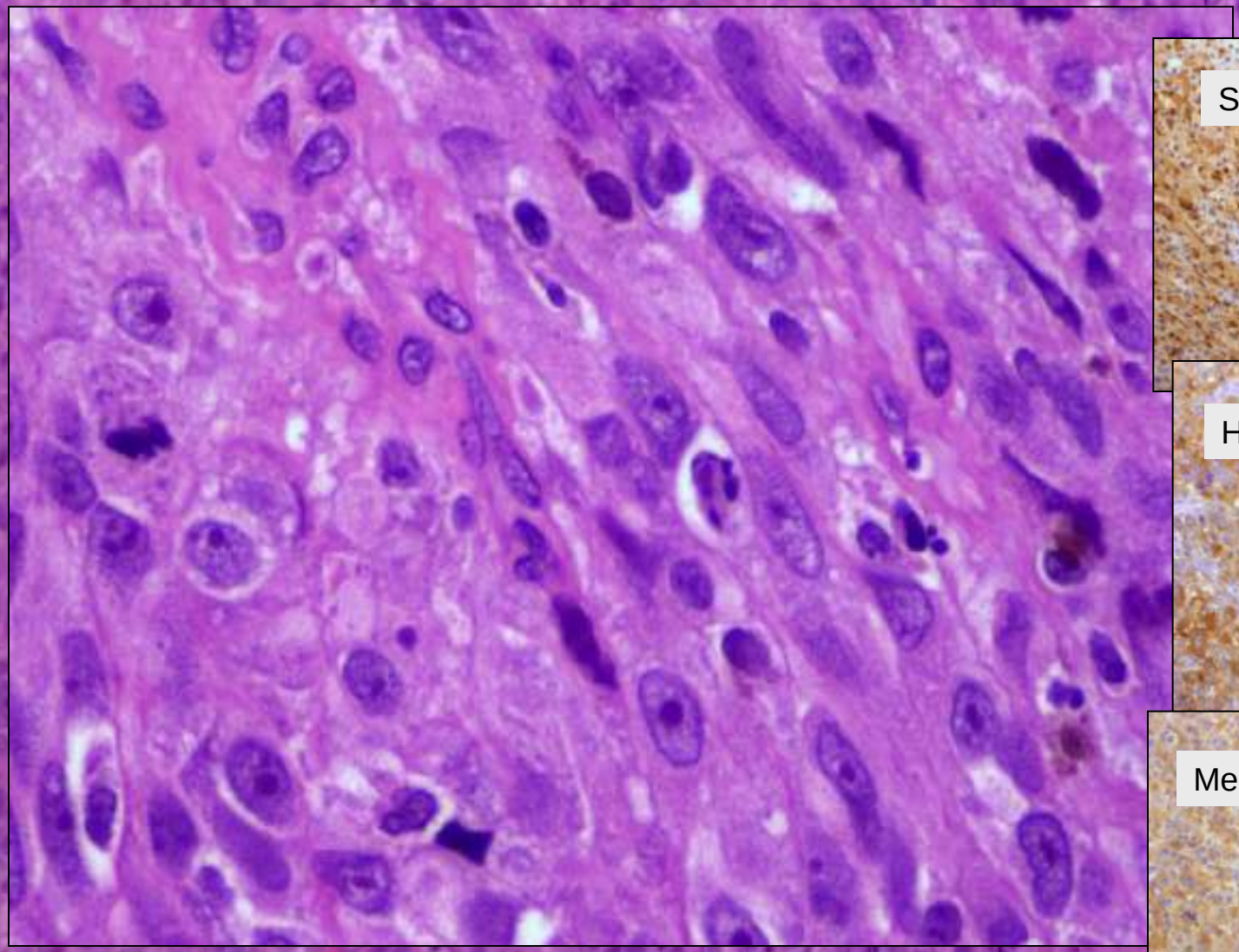


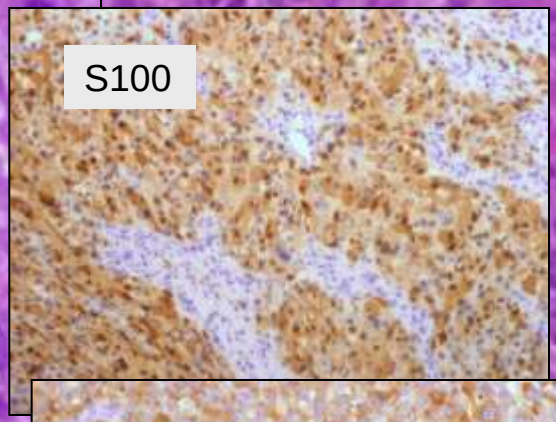


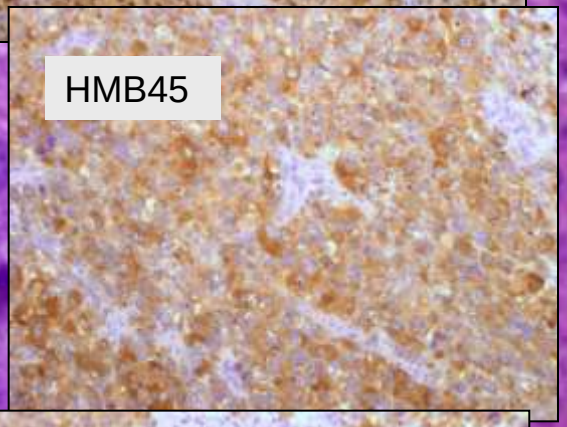
CARCINOMA SARCOMATOIDE

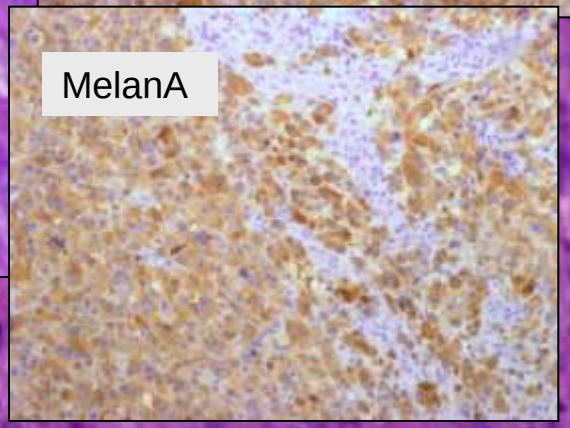


MELANOMA MALIGNO (FUSOCELULAR)









ADÉU