IAP:
Case Presentation 2

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C.S. MOTT CHILDREN'S HOSPITAL
MICHIGAN MEDICINE

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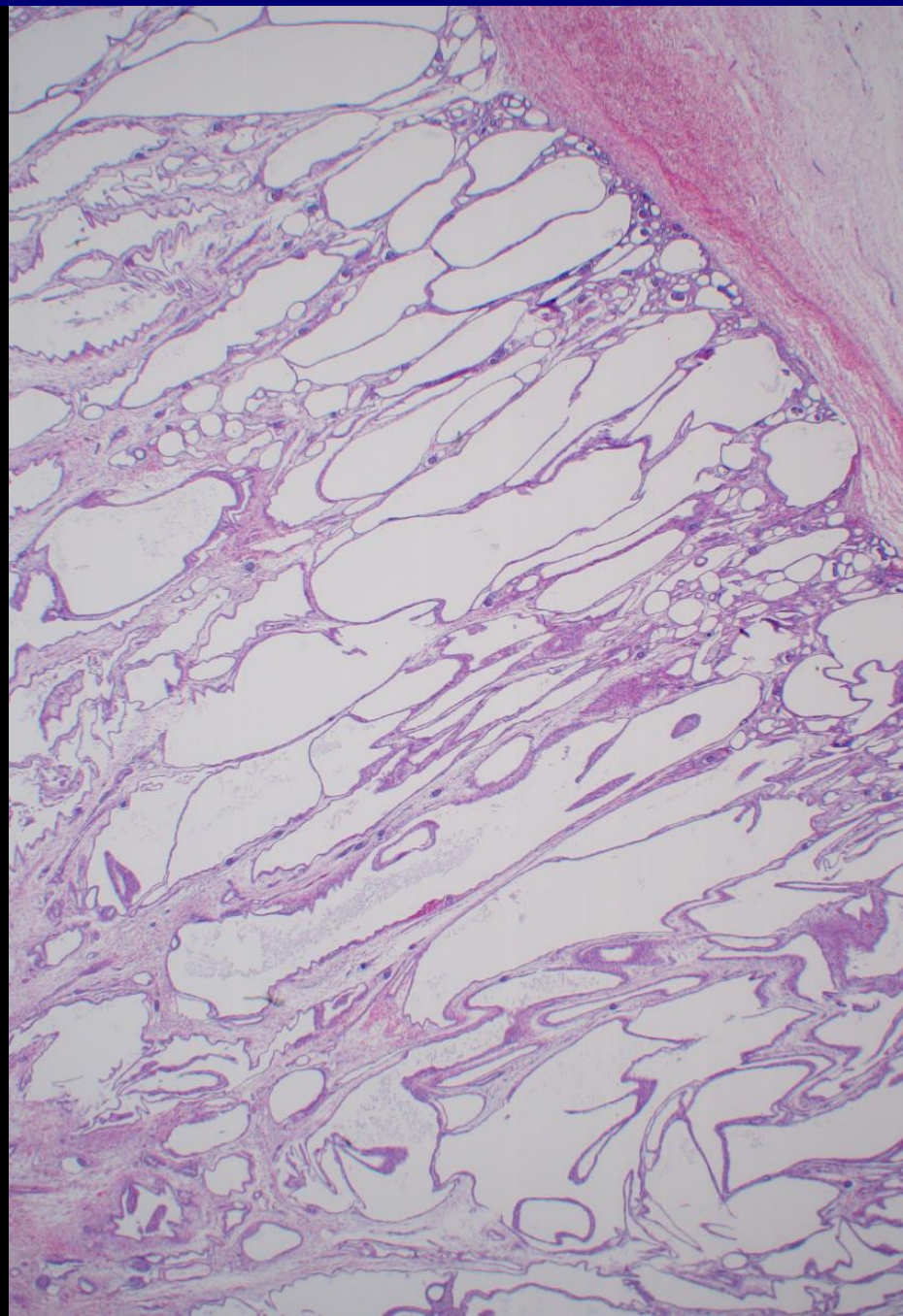
UNIVERSITY OF MICHIGAN

2 year-old boy with history of this disease, presented with incidental vague liver masses



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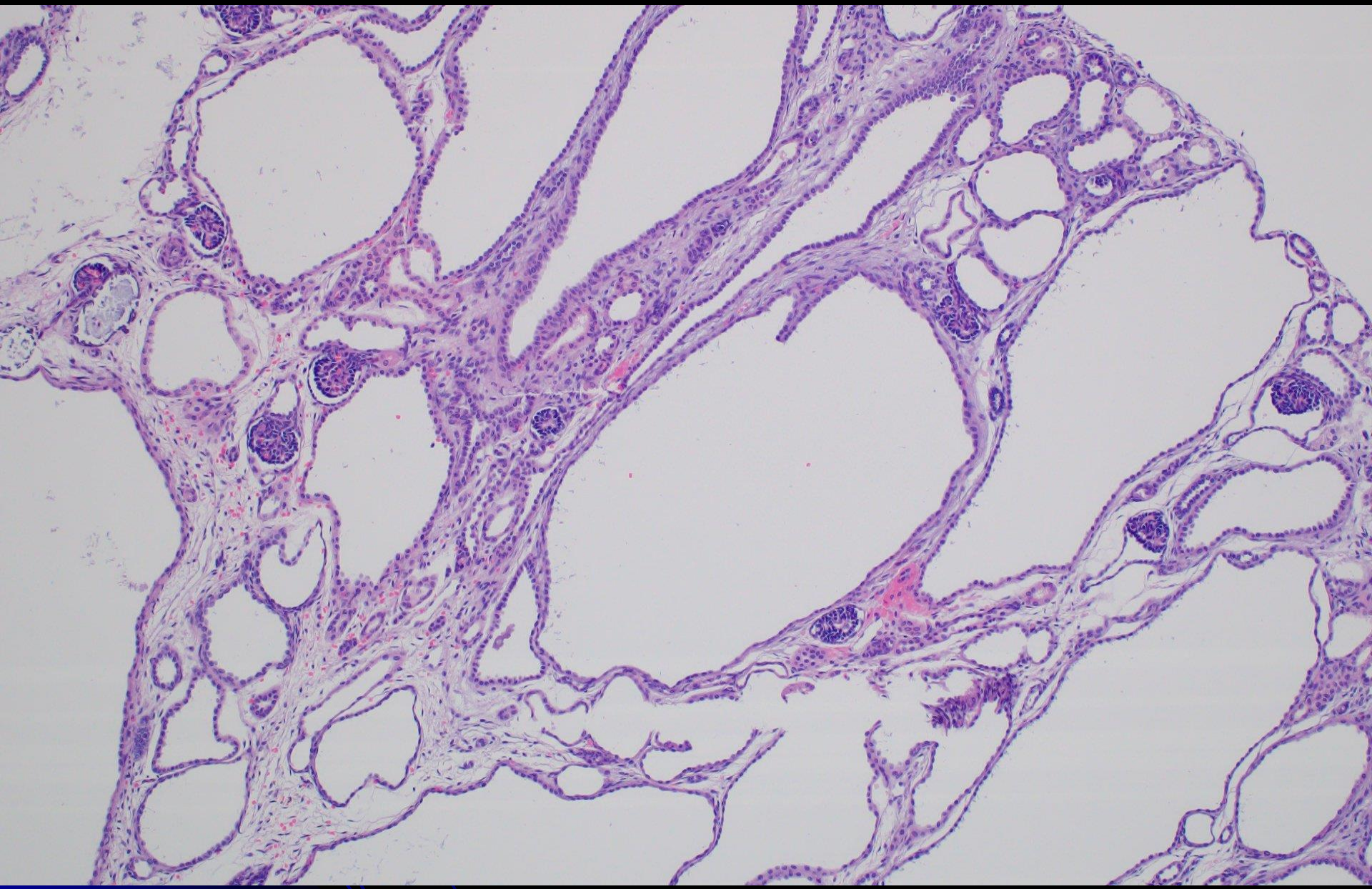

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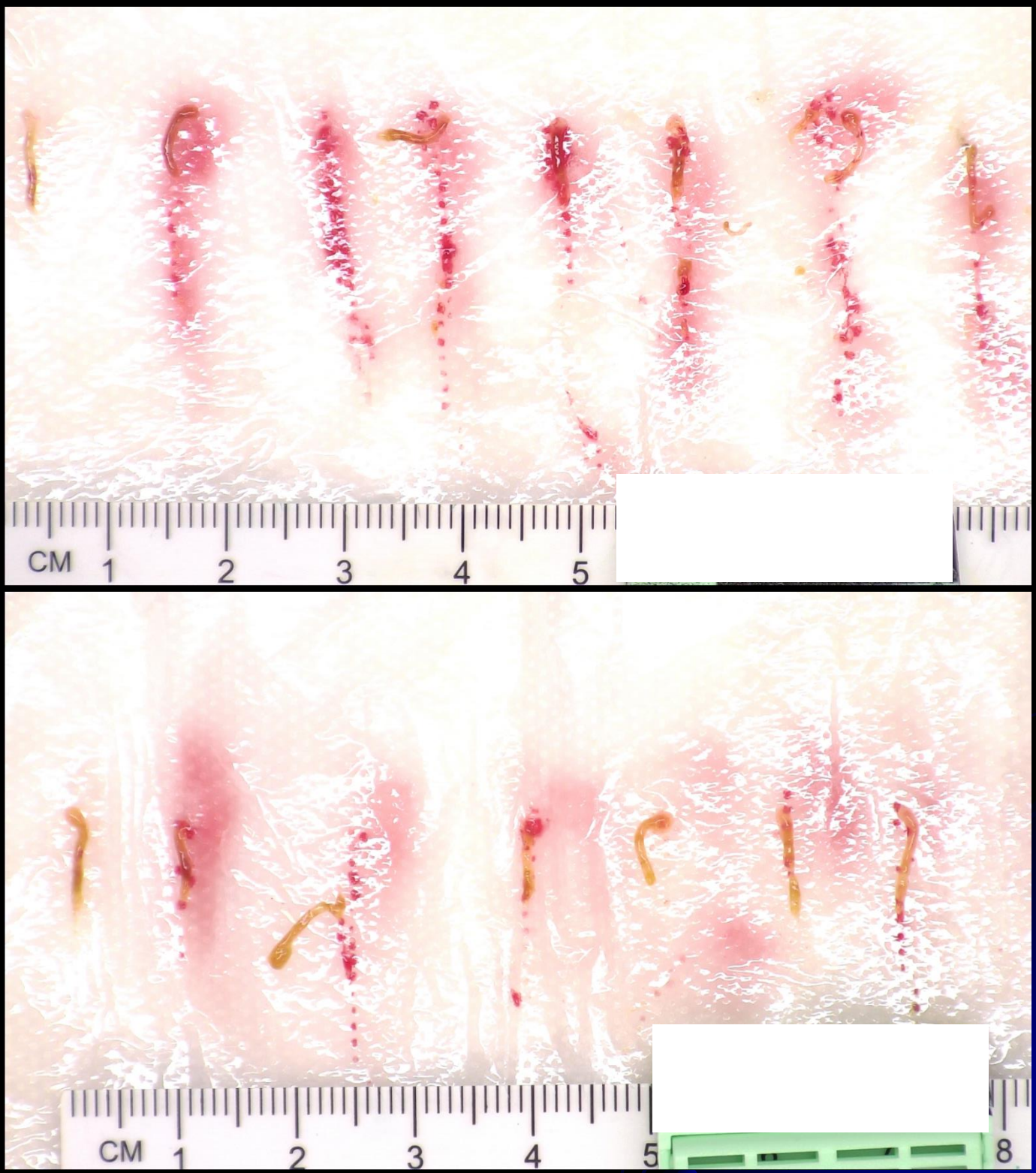
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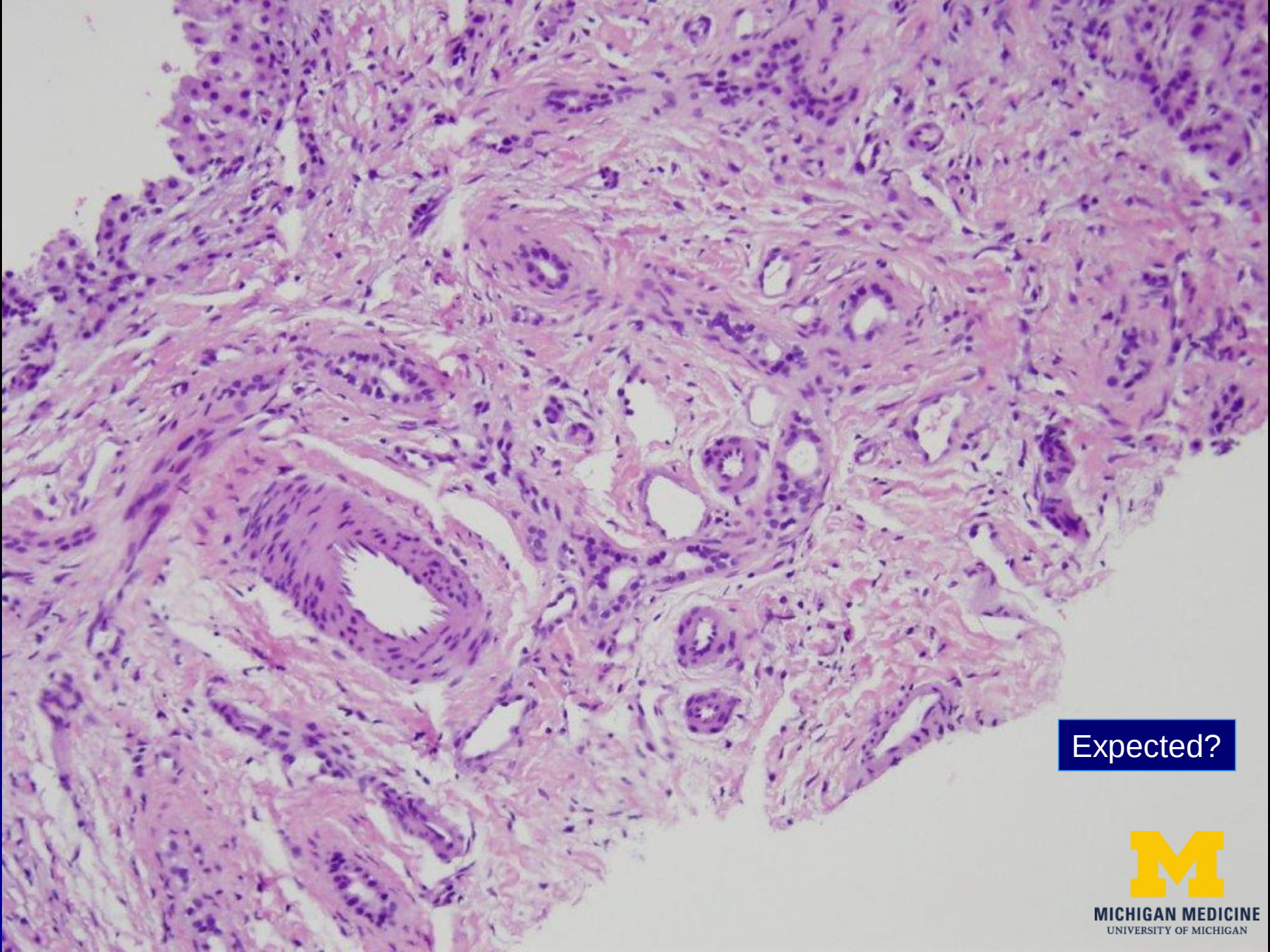

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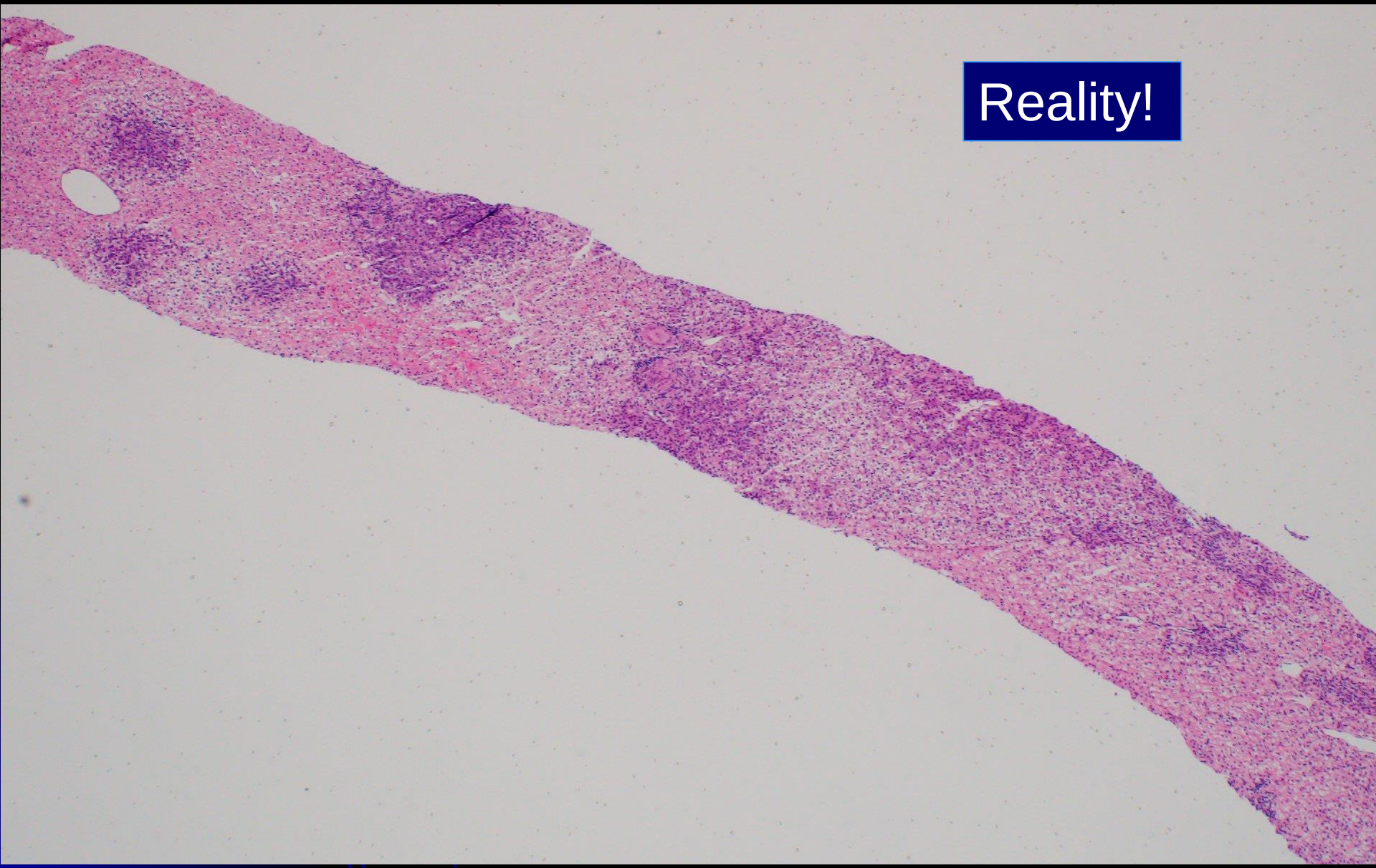

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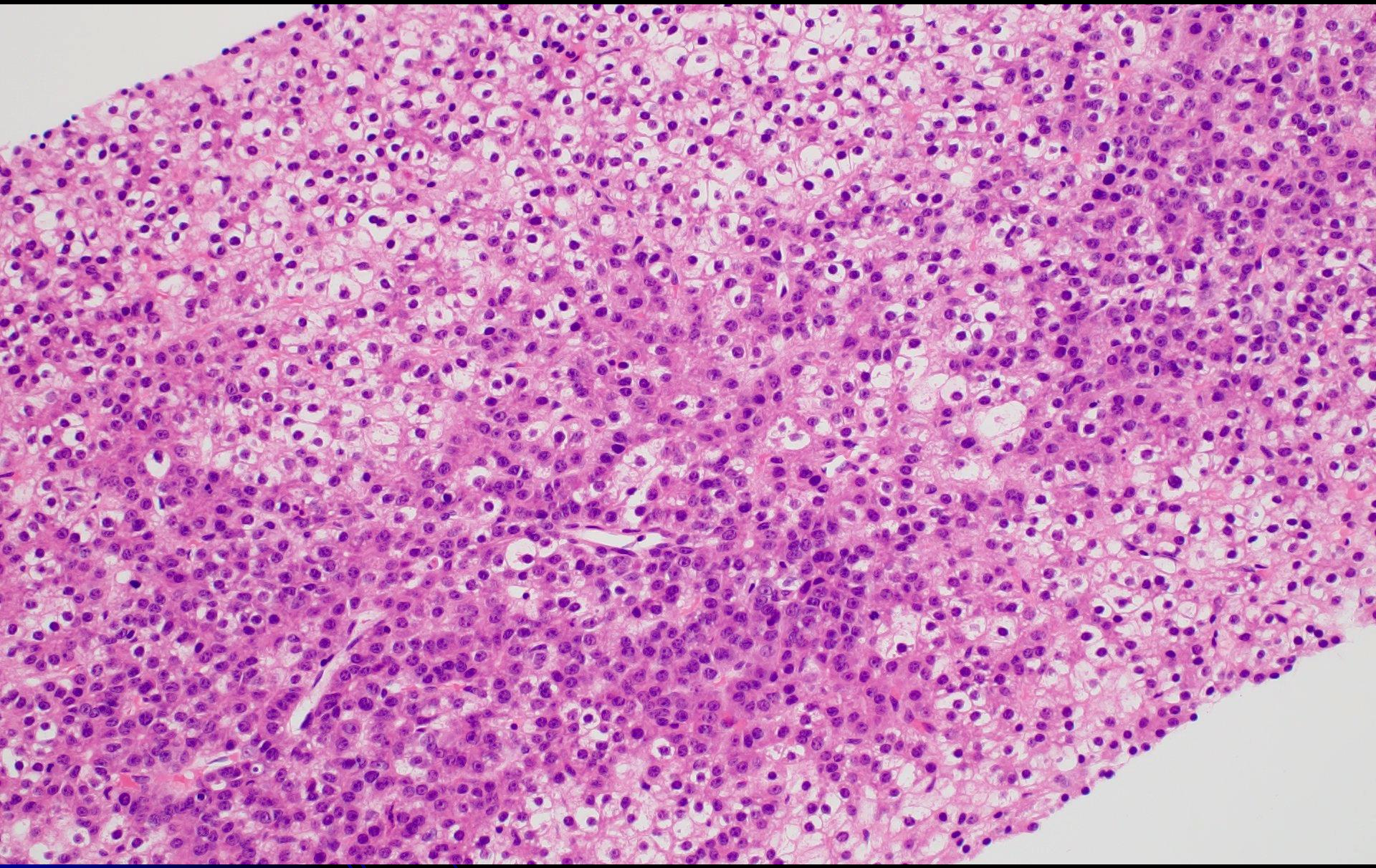

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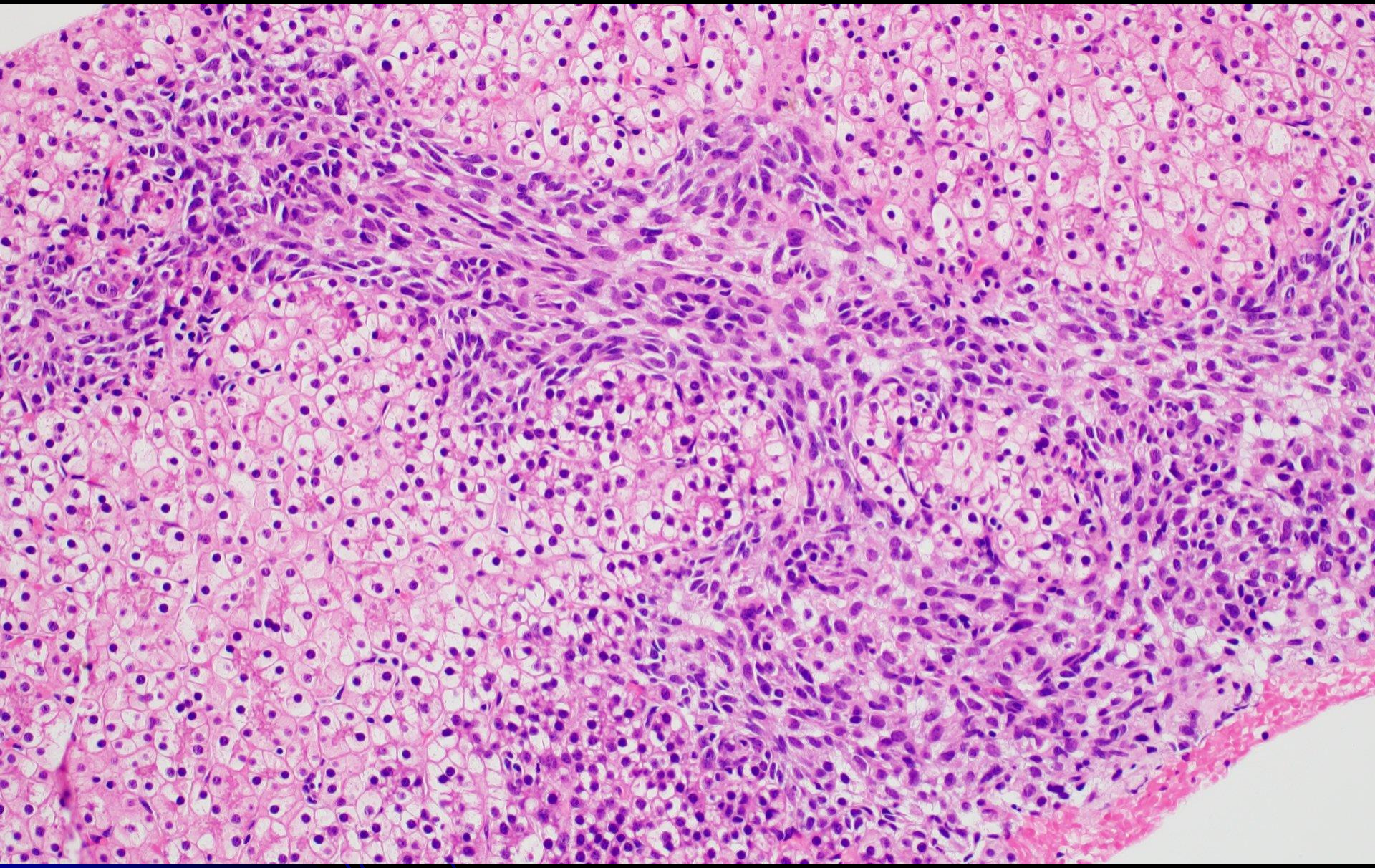

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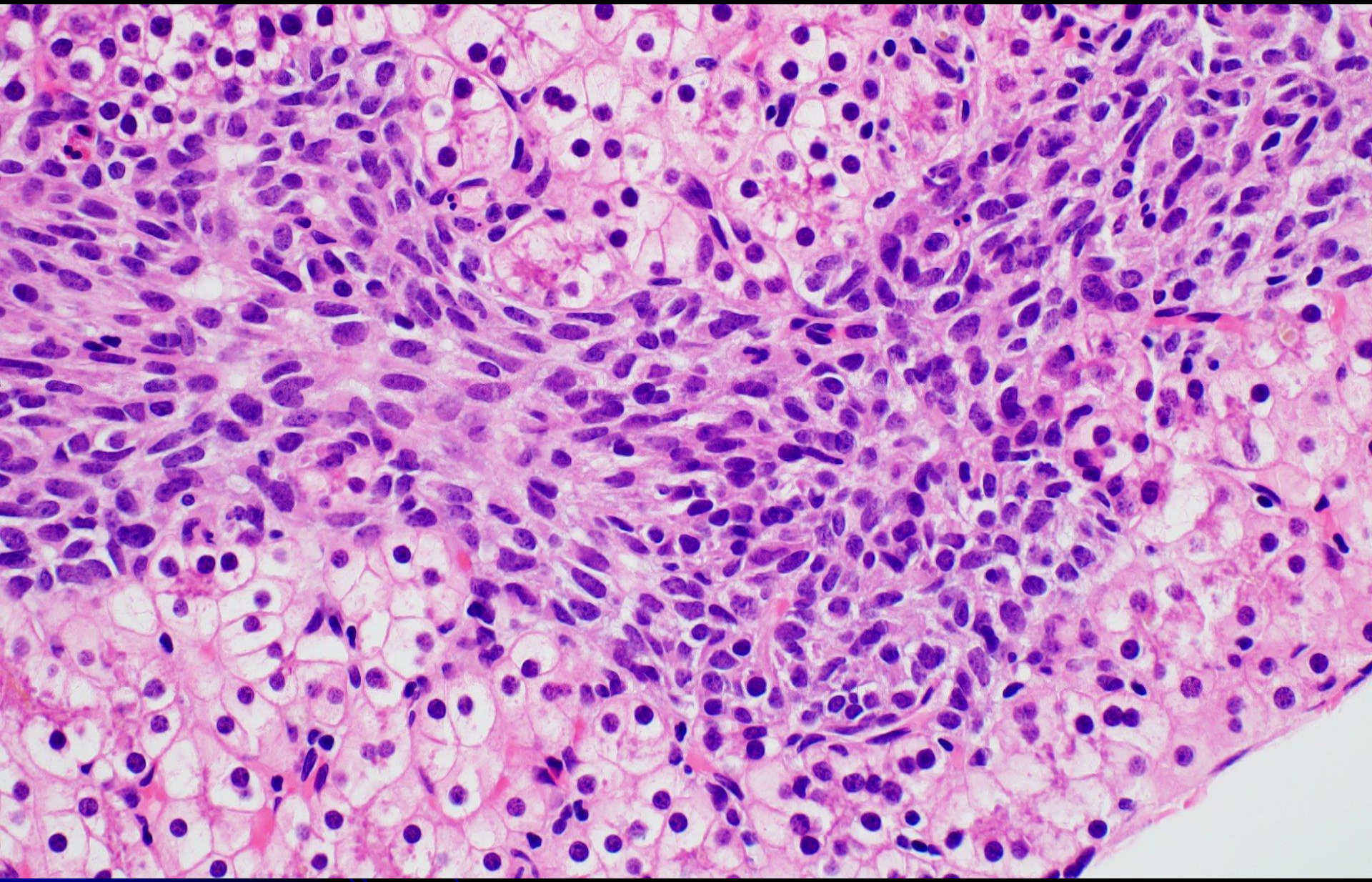

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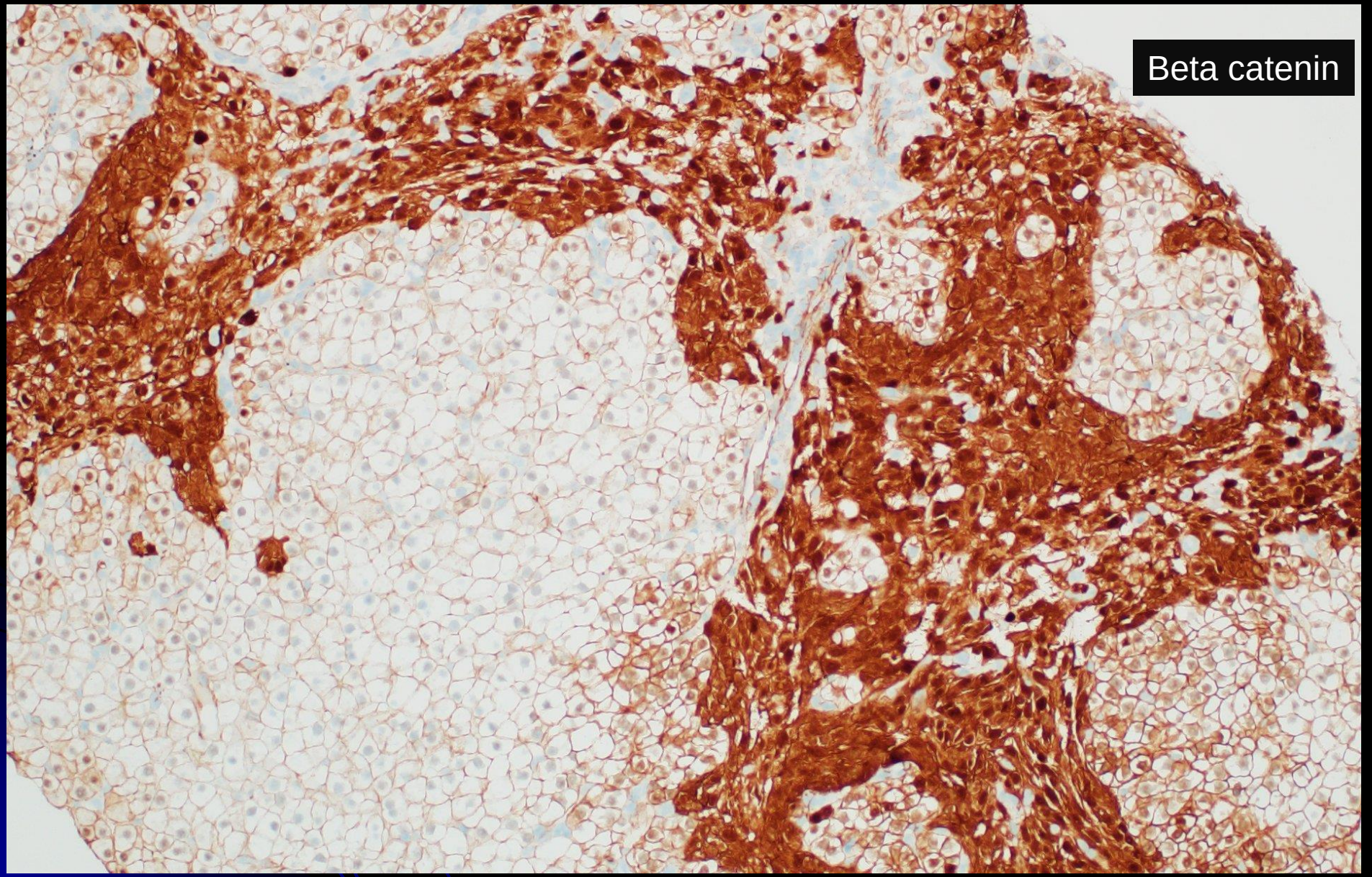
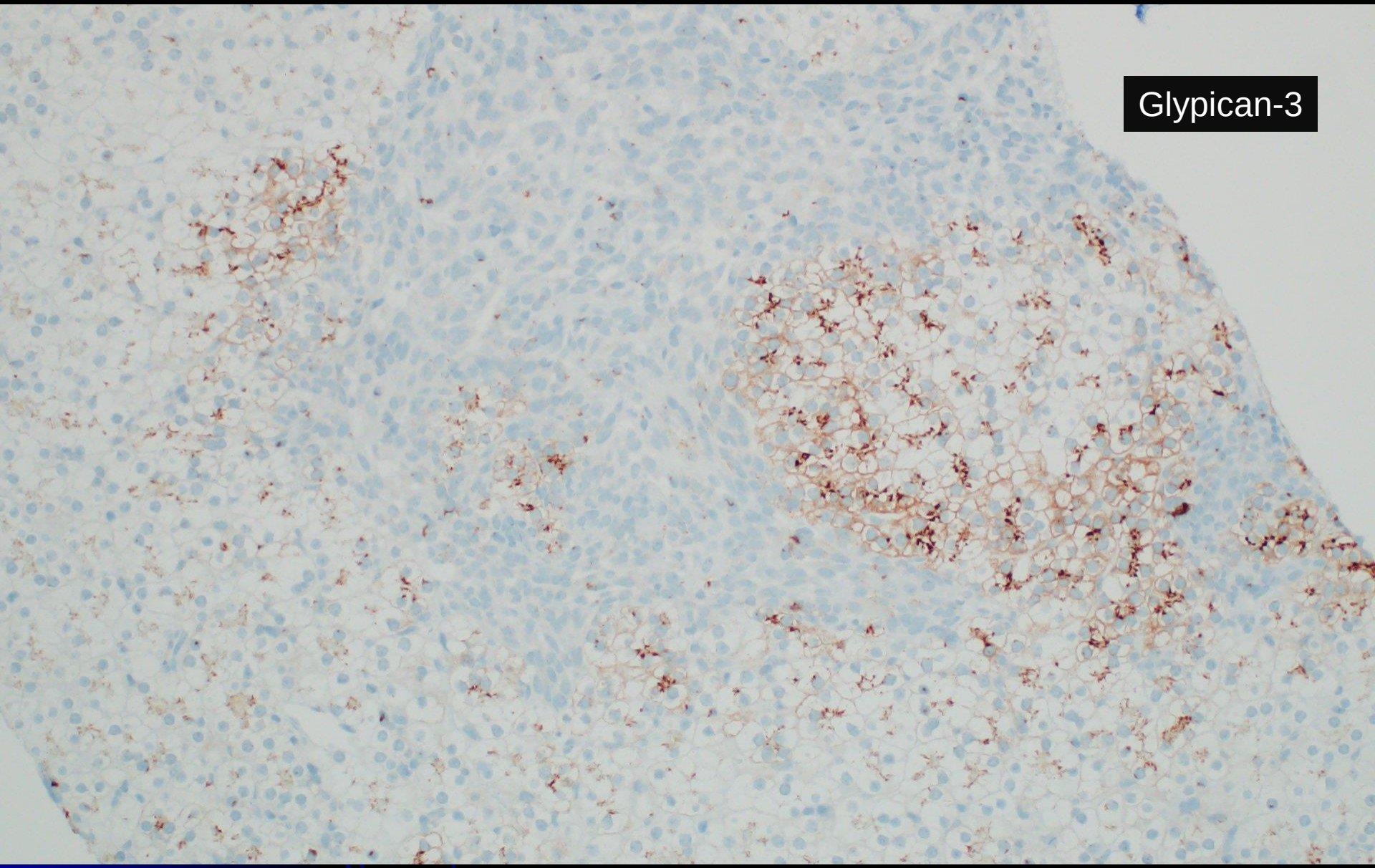
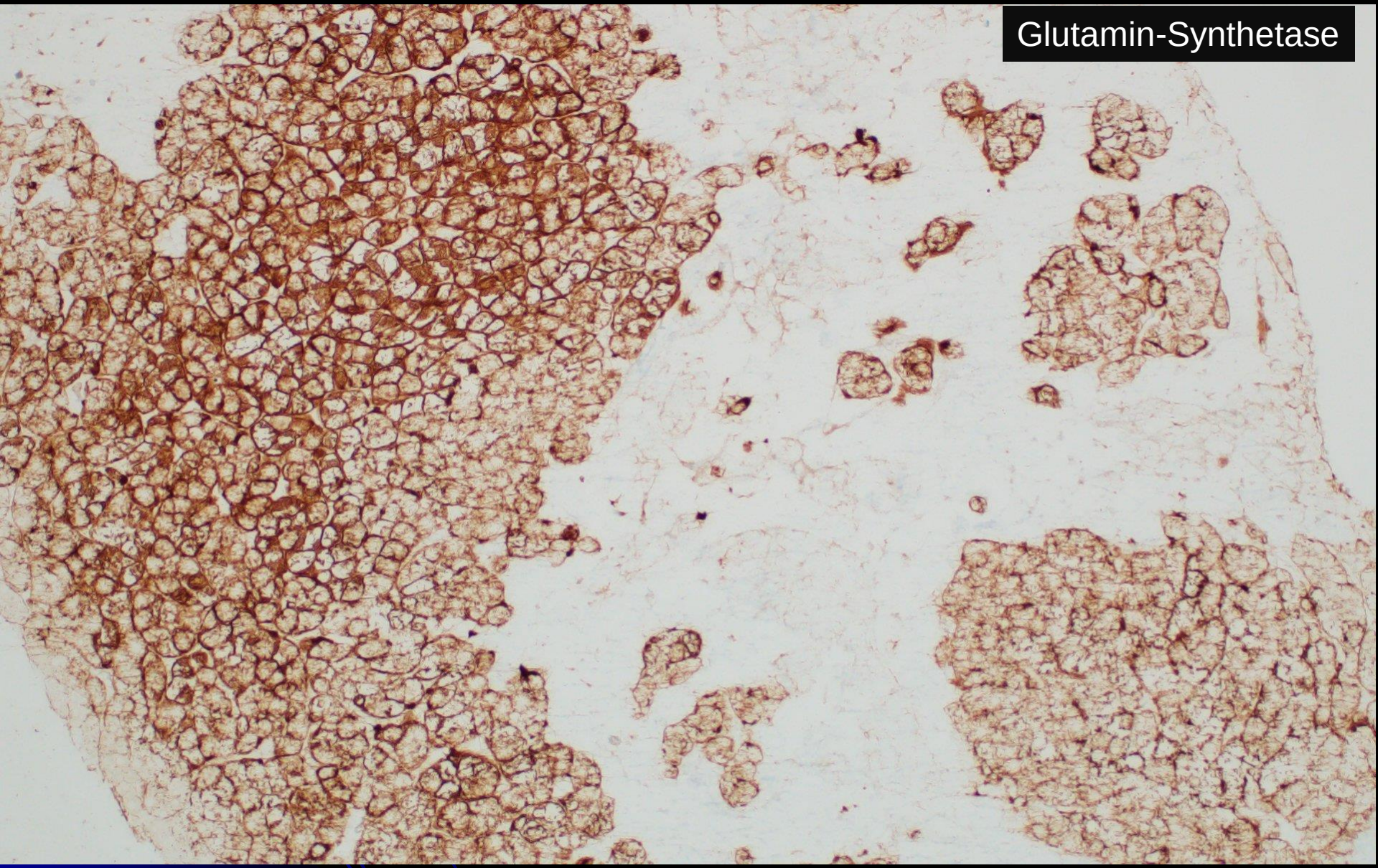
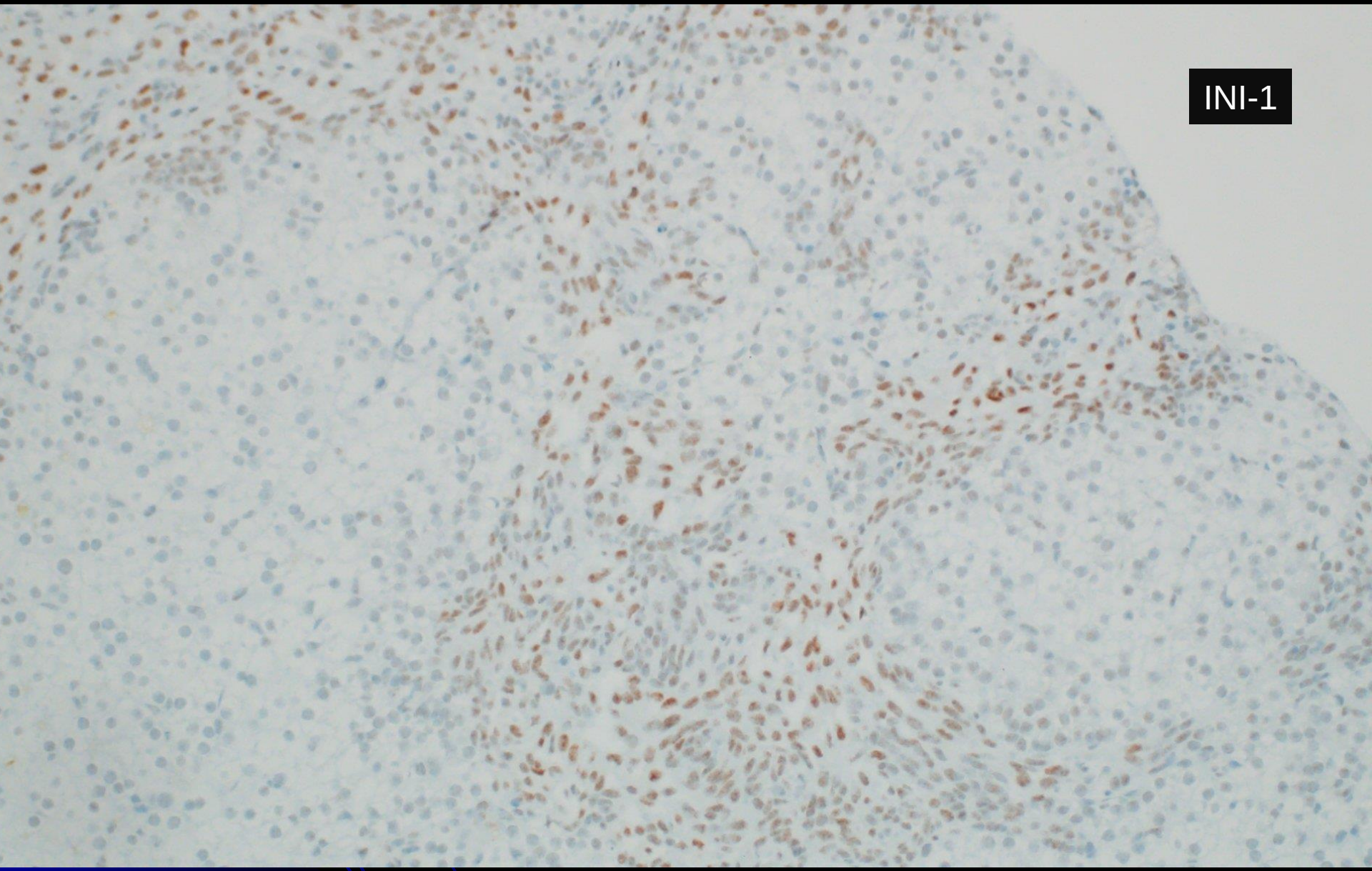
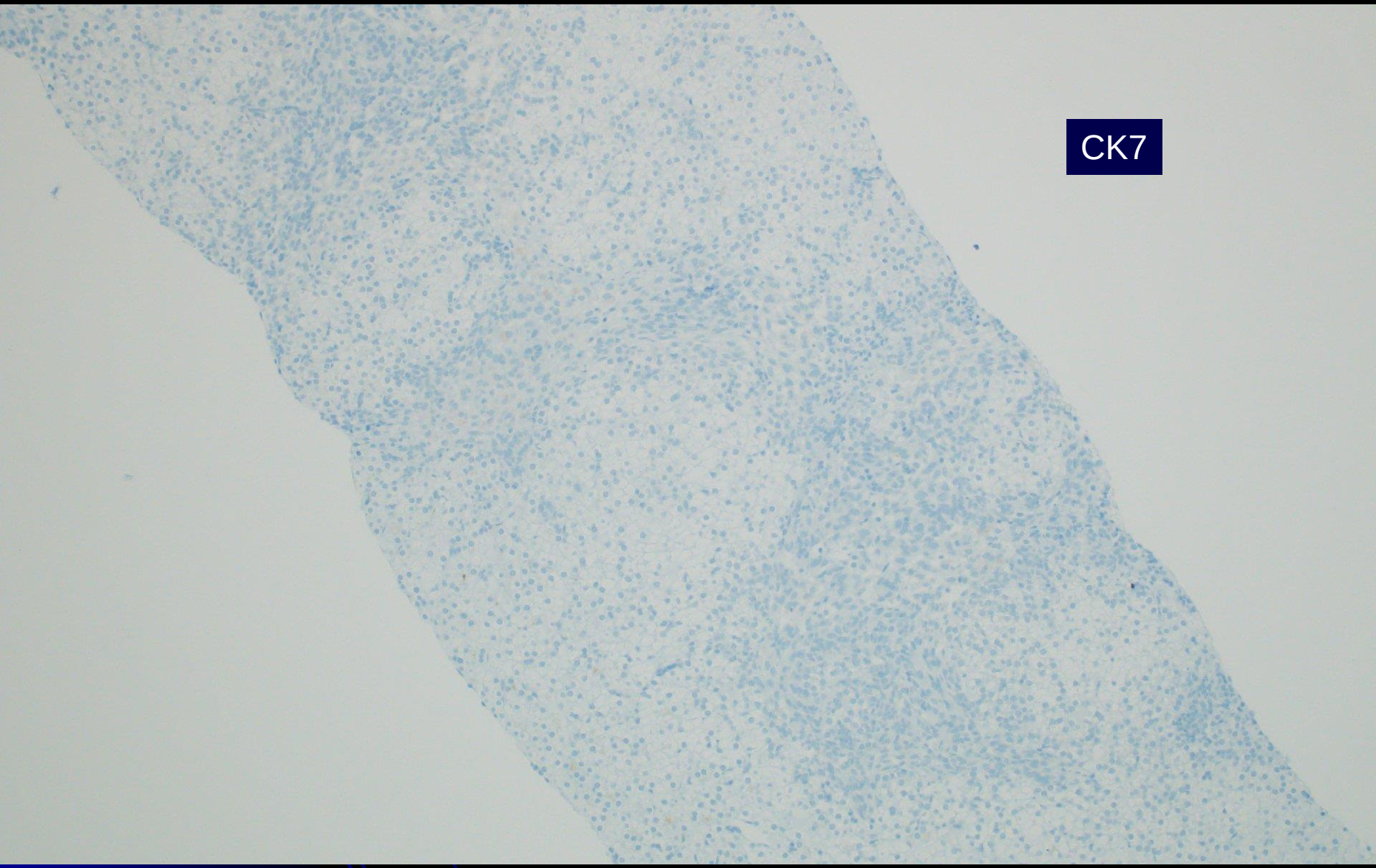
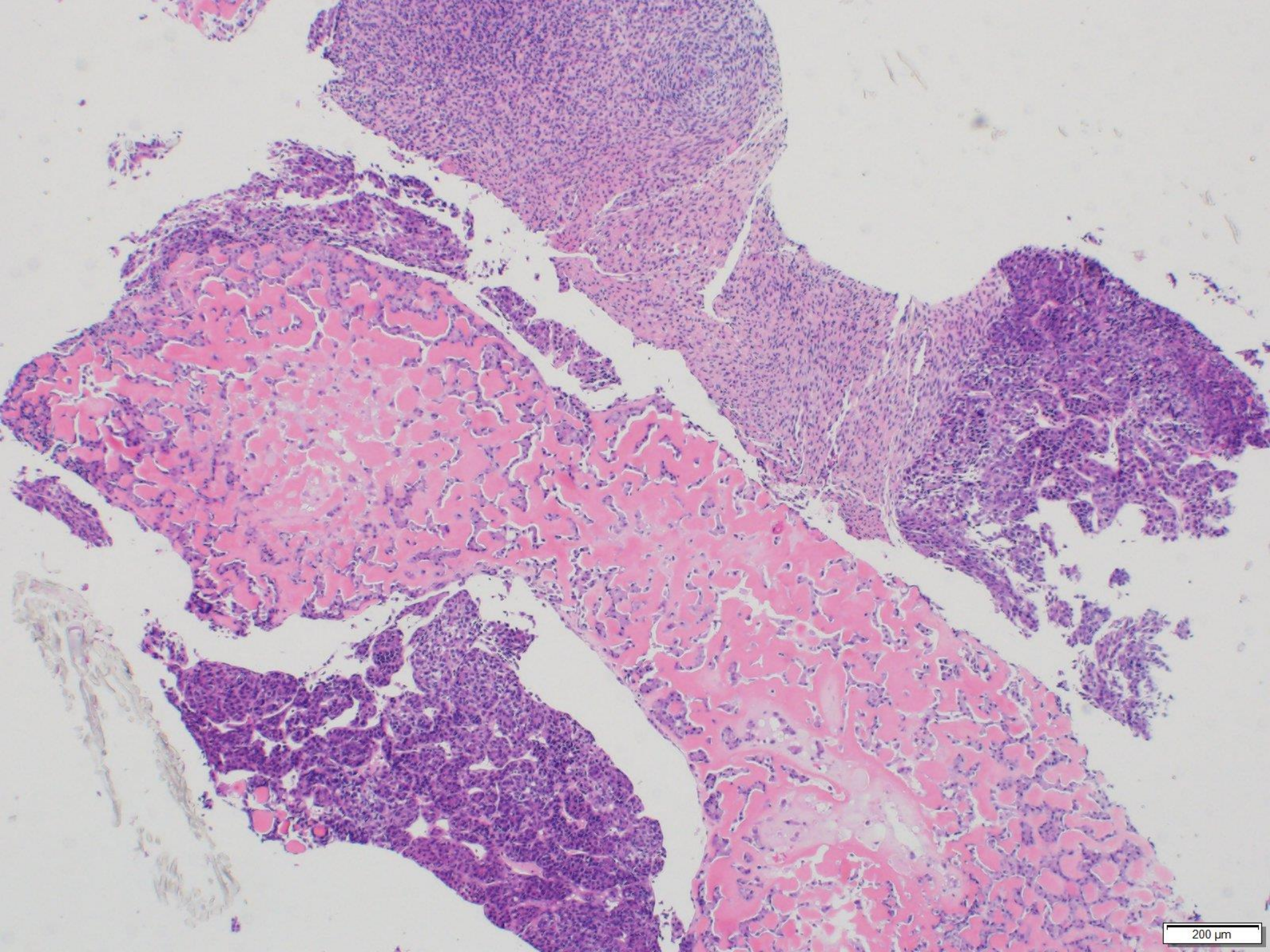


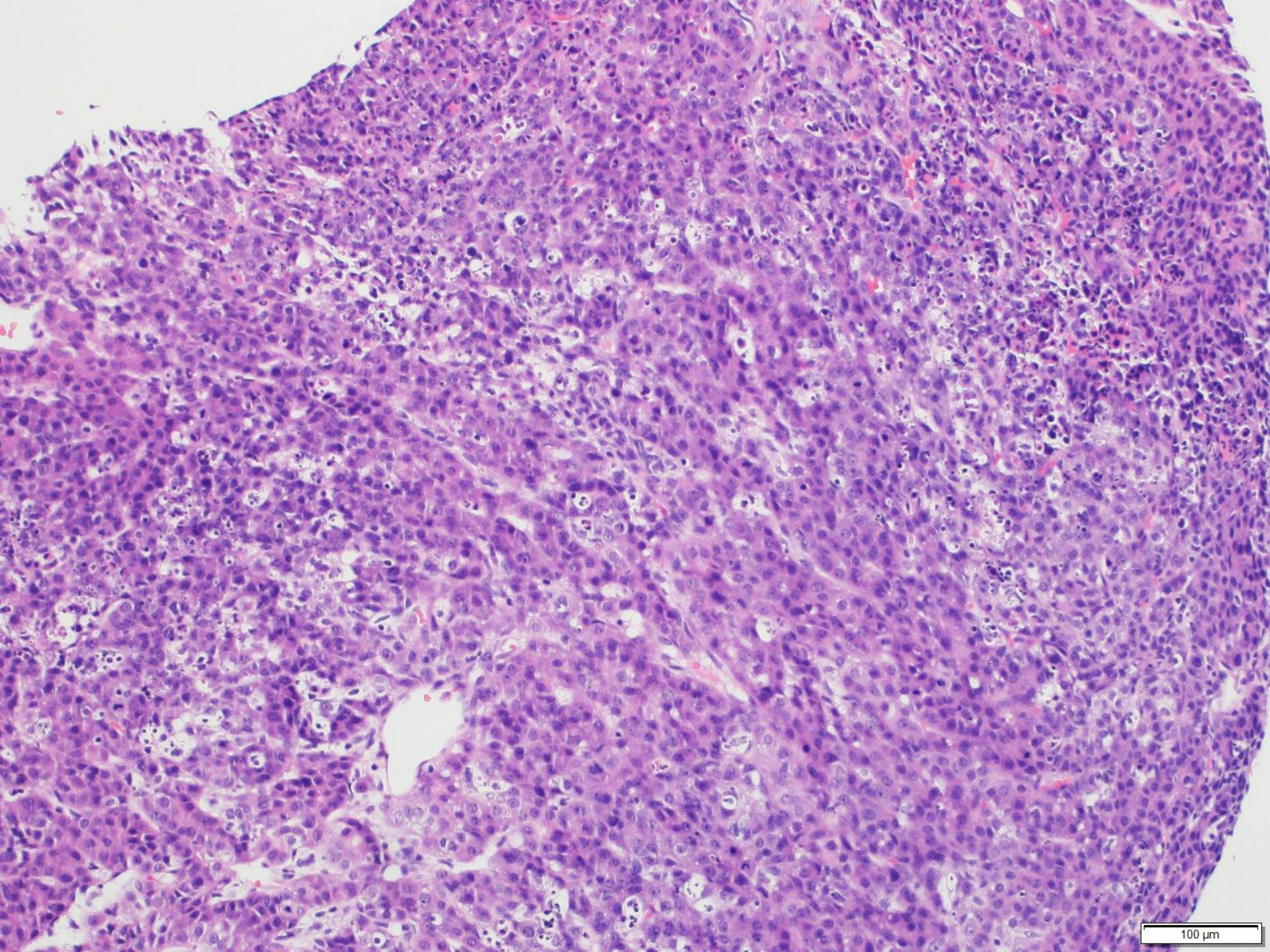


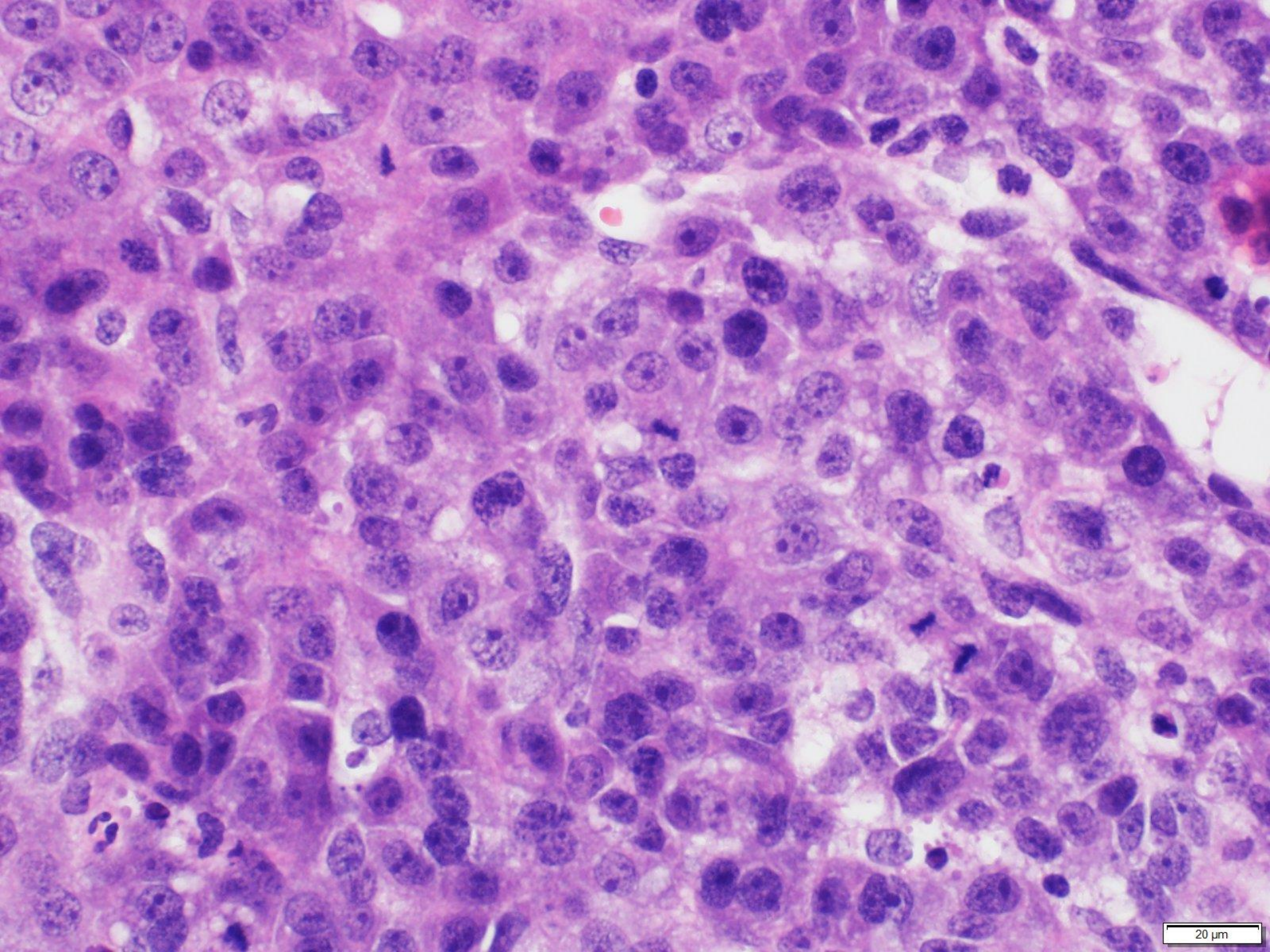


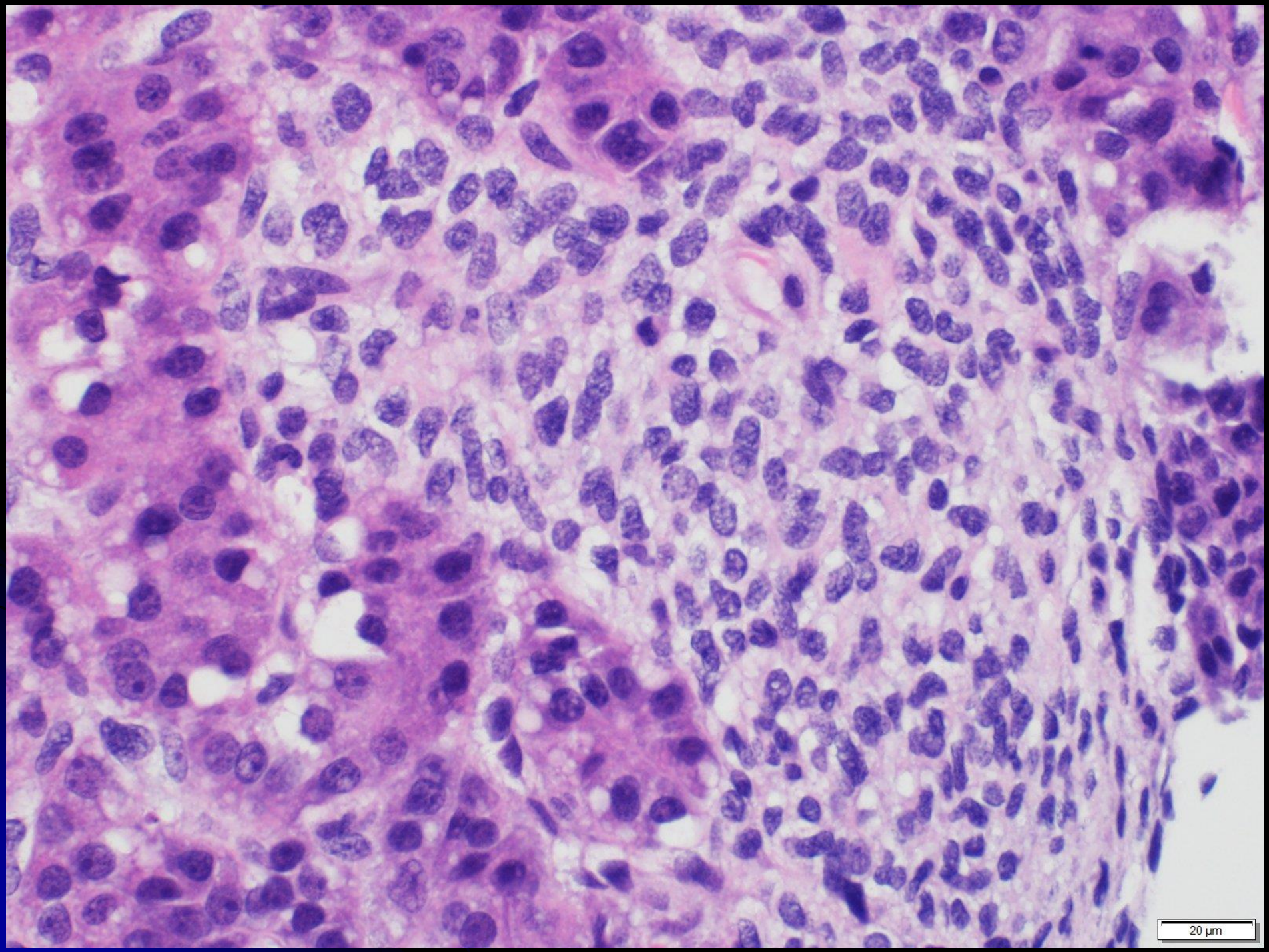


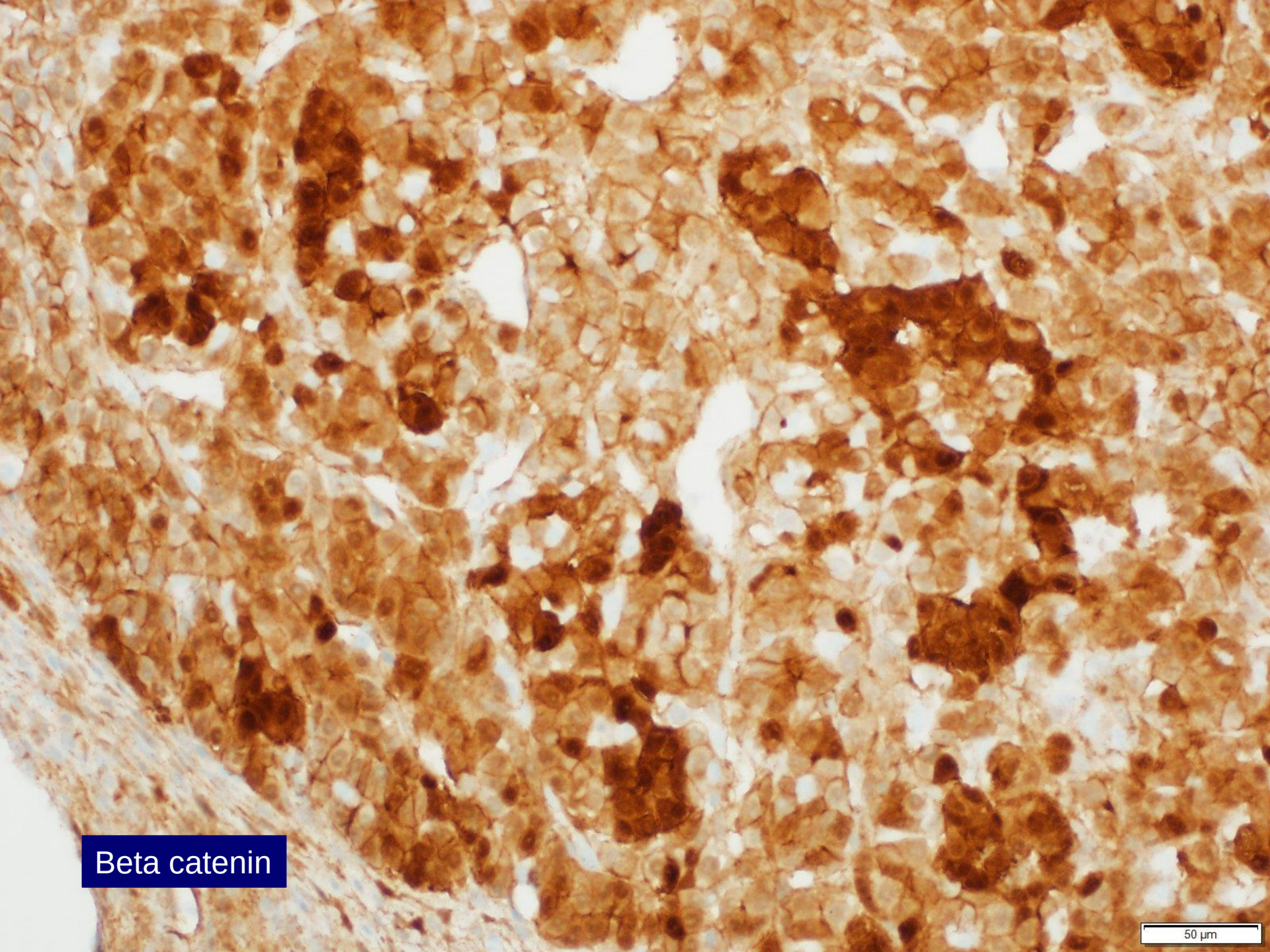


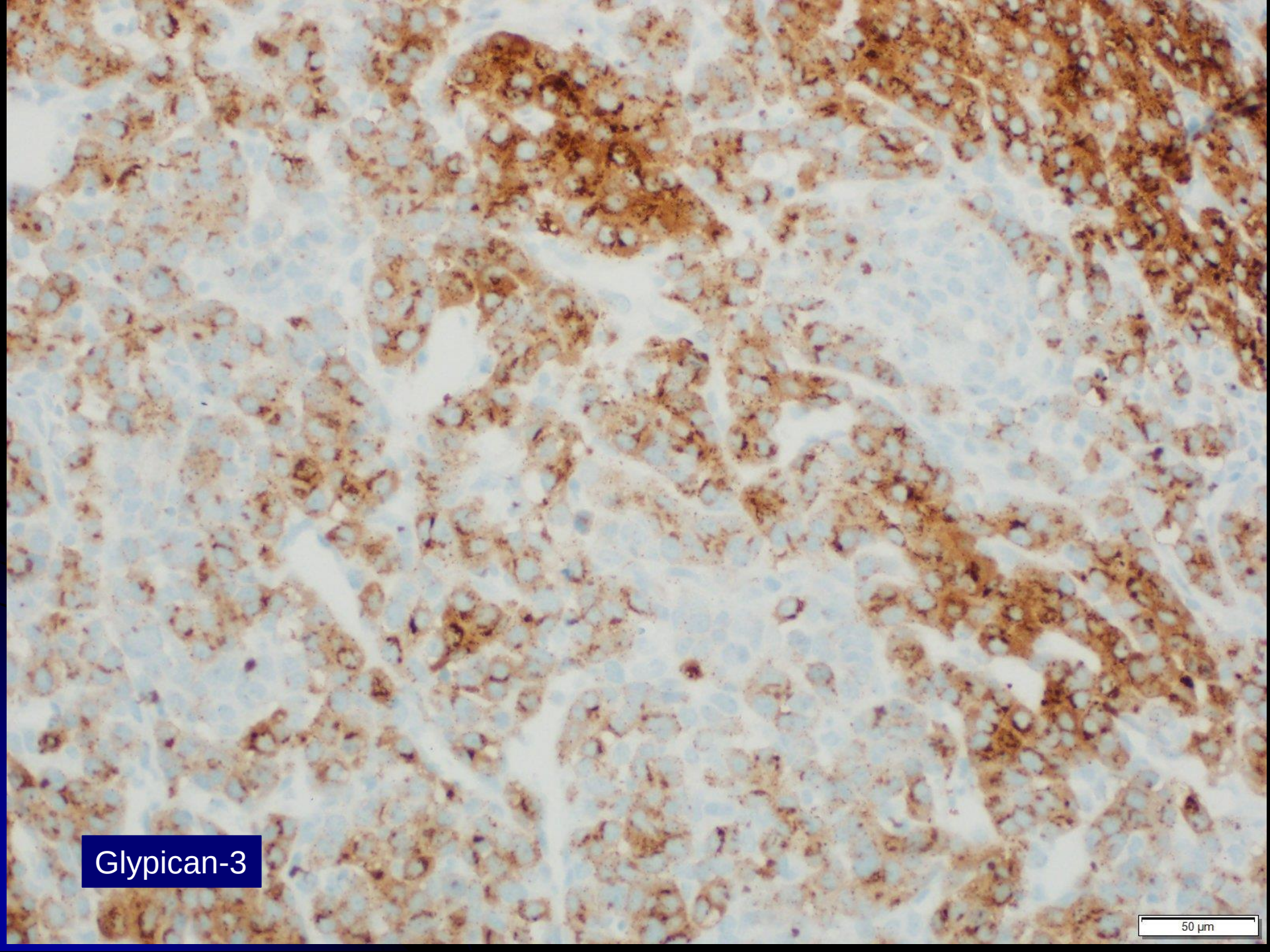

200 µm

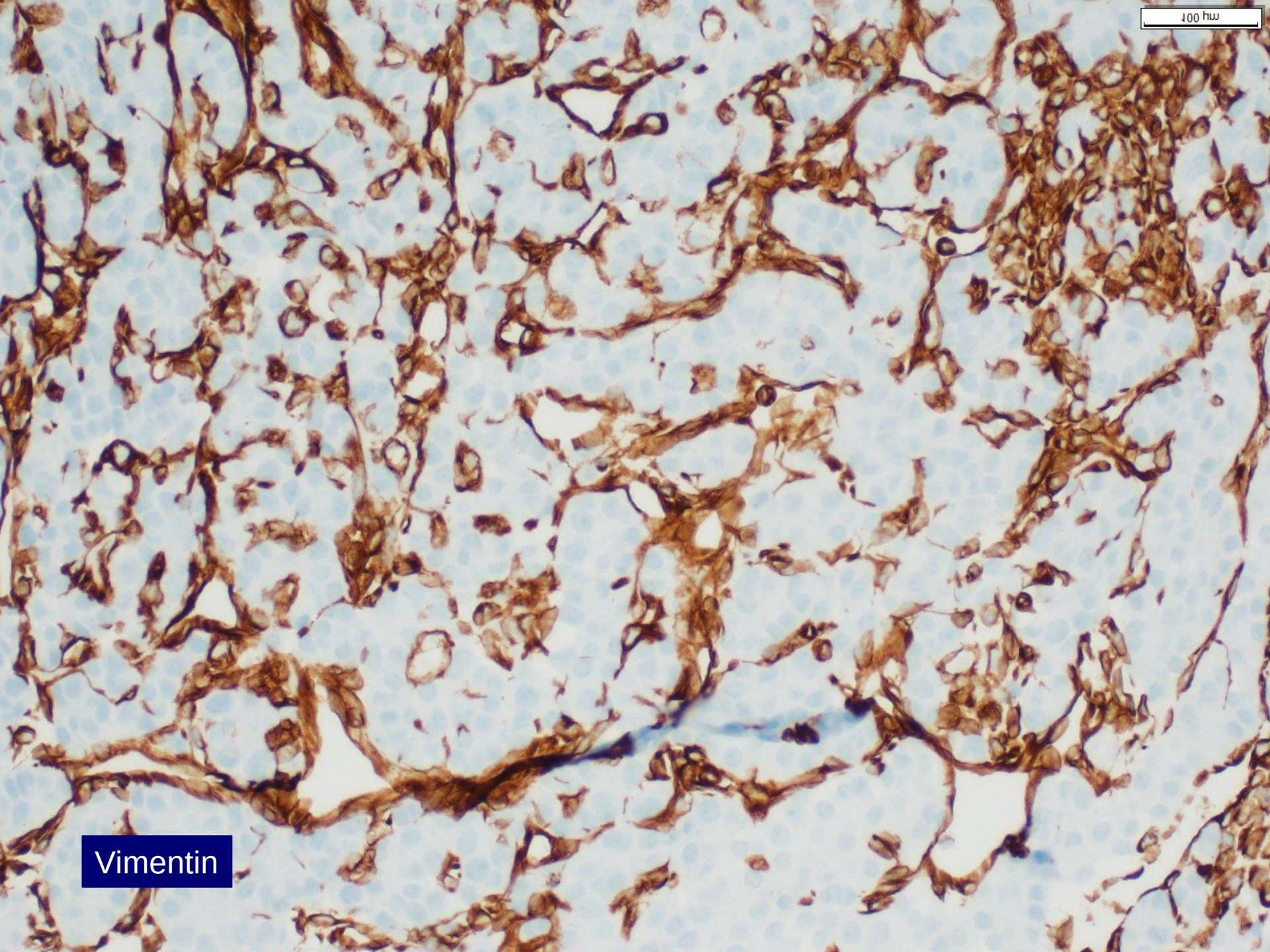

100 µm


20 µm


20 µm


Beta catenin
50 µm


Glypican-3
50 µm


Vimentin

Hepatoblastoma

- Most common primary liver tumor in children
- M>F; most cases by age 5; can be congenital
- AFP, thrombocytosis
- Association: Familial polyposis (APC), Beckwith-Wiedemann, Li-Fruameni, trisomy 18, Glycogen storage disease
- Mets: adrenals, BM, lung, brain, regional LNs

Hepatoblastoma

- Epithelial type >65%; different histologic patterns
  - Fetal; well-differentiated (mitotically inactive, ≤2 mitoses per 10 hpf) treated with surgery alone; mitotically active
  - Embryonal
  - Mixed fetal and embryonal (most common)
  - Macrotrabecular; poor prognosis
- Small cell undifferentiated ; poor prognosis; Is it anymore?
- Rhabdoid, loss of INI-1
- Mixed epithelial and stromal type: osteoid, adipose tissue, other mesenchymal elements
  - Teratoid: Squamous, GI, immature neuroectoderm, melanin, poor prognosis

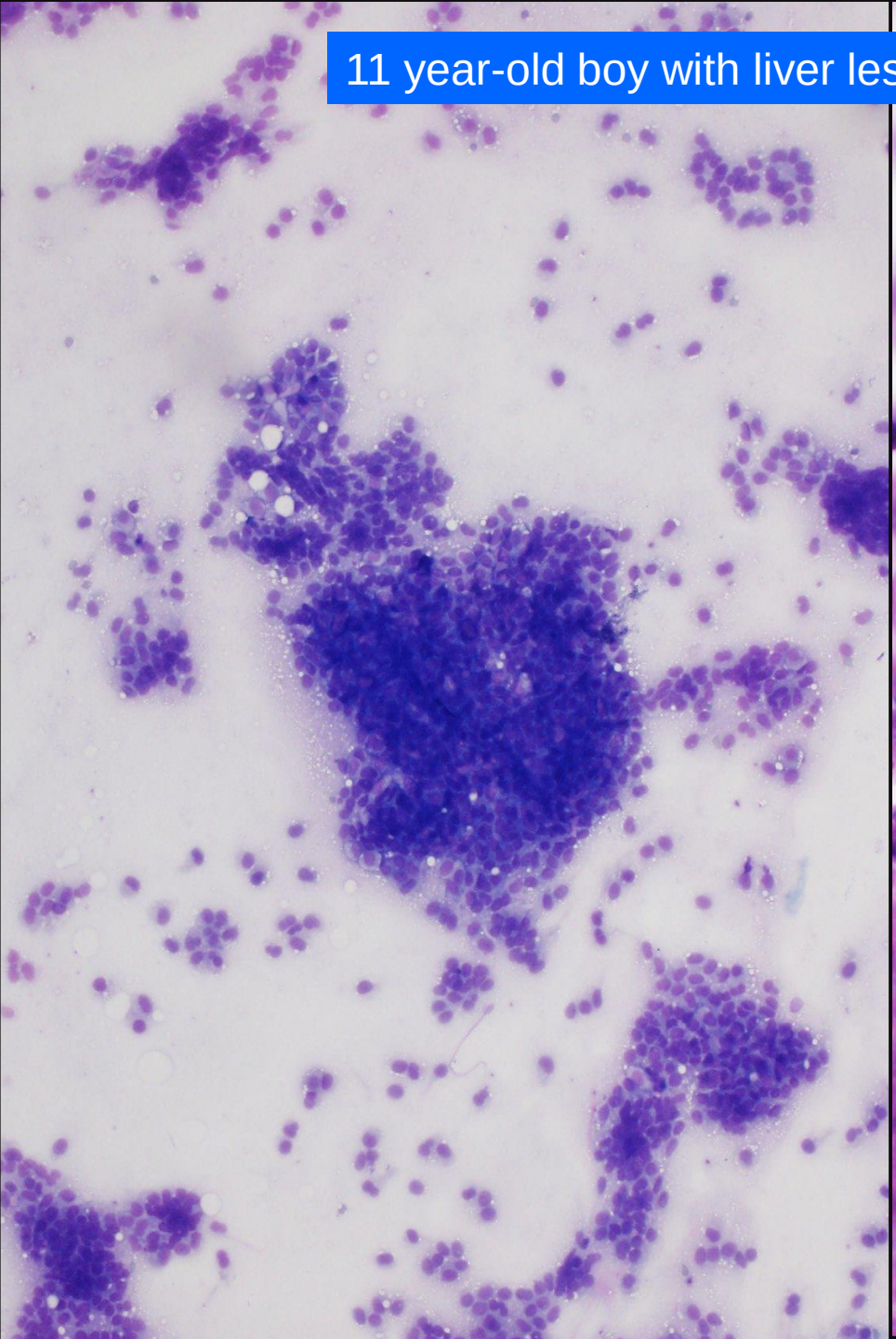
Take Home Points

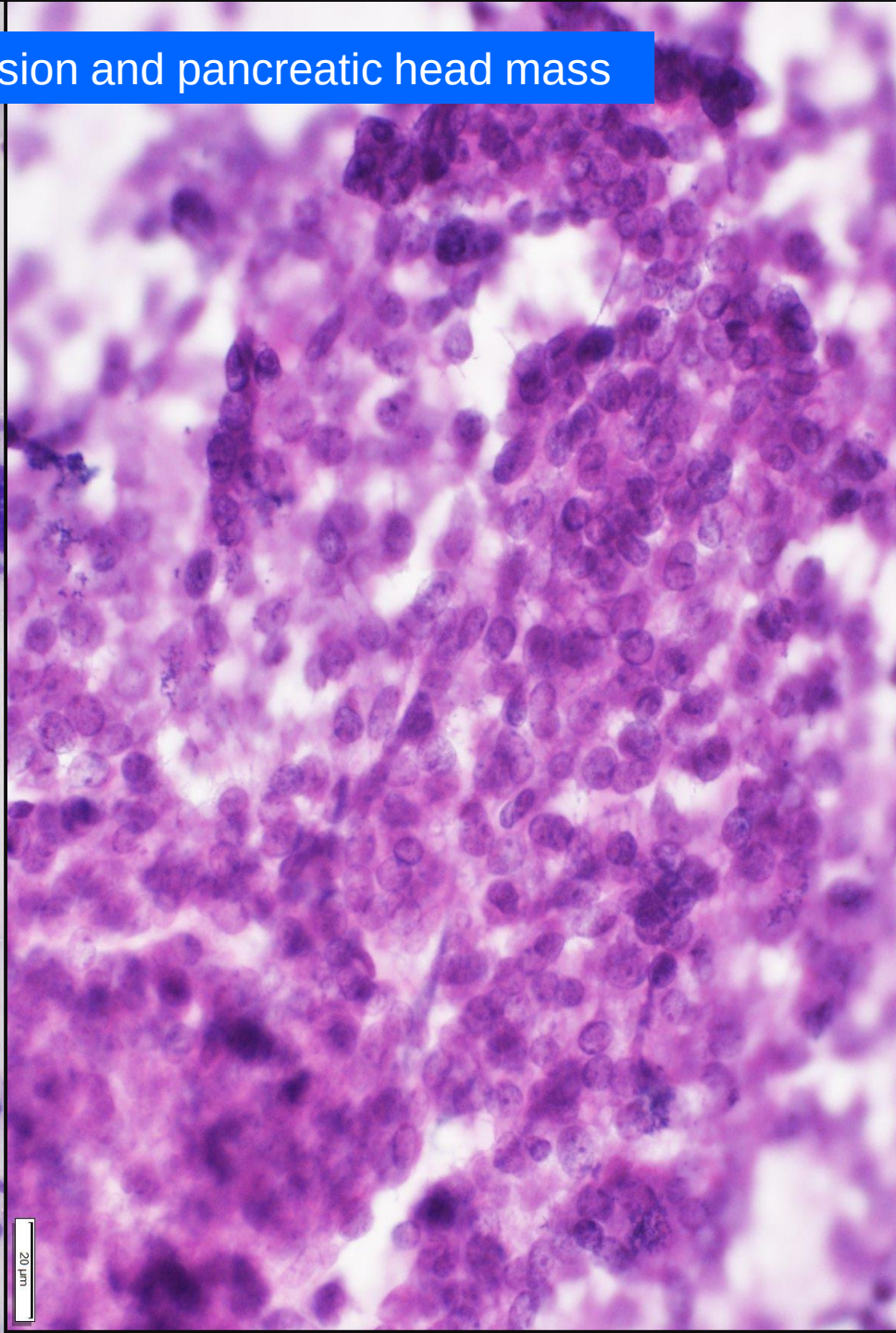
- Hepatoblastoma most common primary liver malignancy in children, AFP
- Hepatoblastoma subtyes: small cell undifferentiated and rhabdoid worse prognosis?
- Small cell undifferentiated component immunoprofile
- Molecular: Trisomies/gains, 2, 20, 8, 6?

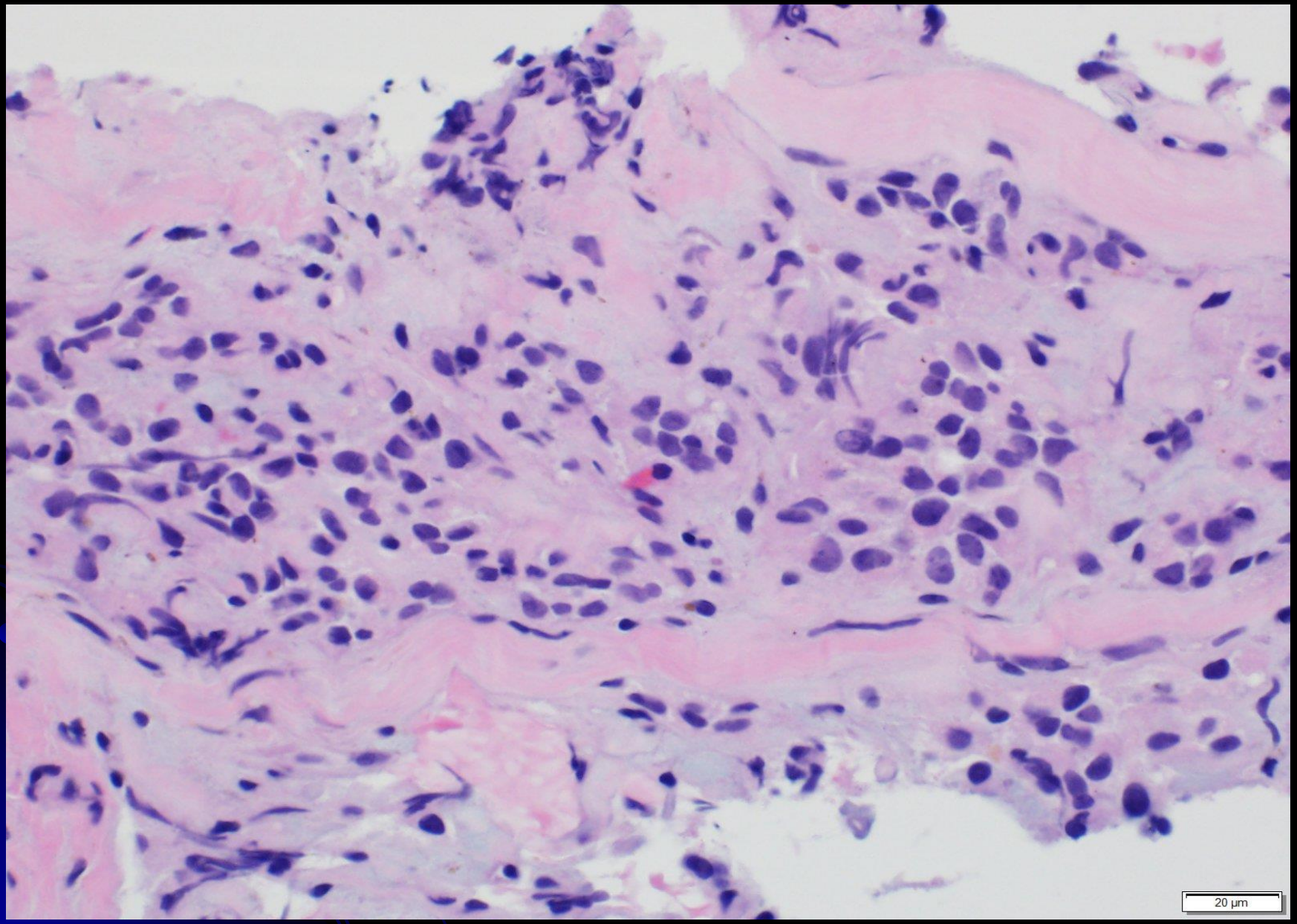

Just take it easy ☺
500 µm

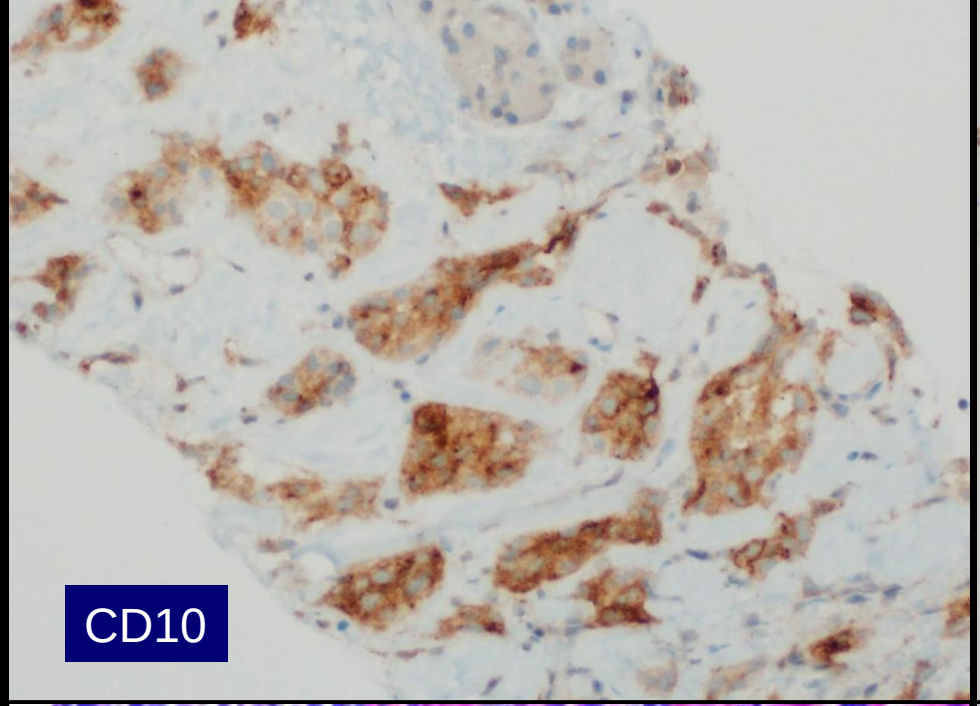
Another Story?

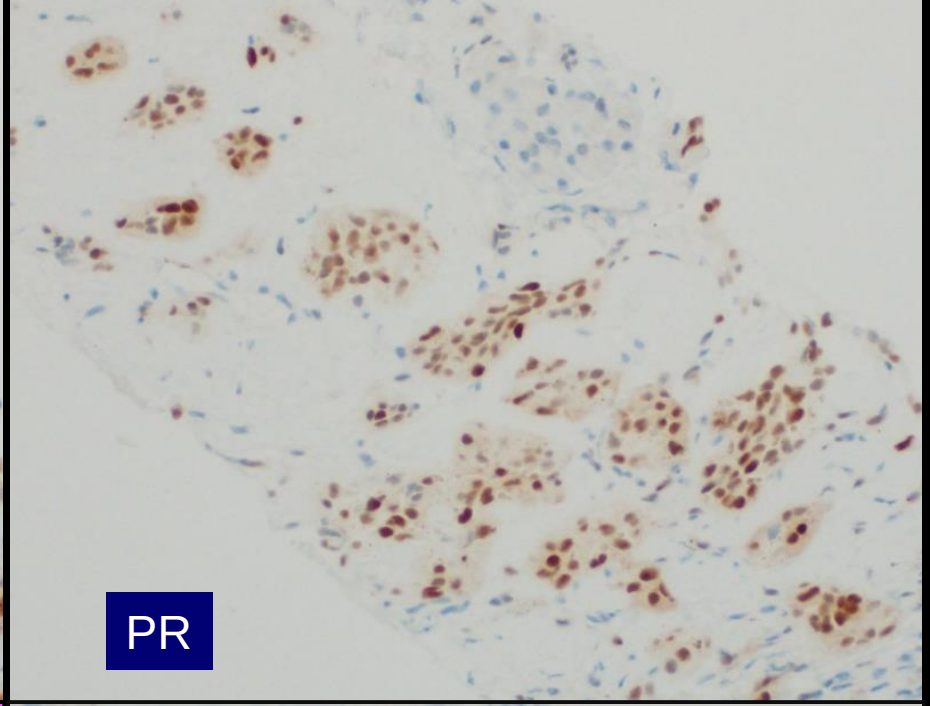
11 year-old boy with liver lesion and pancreatic head mass

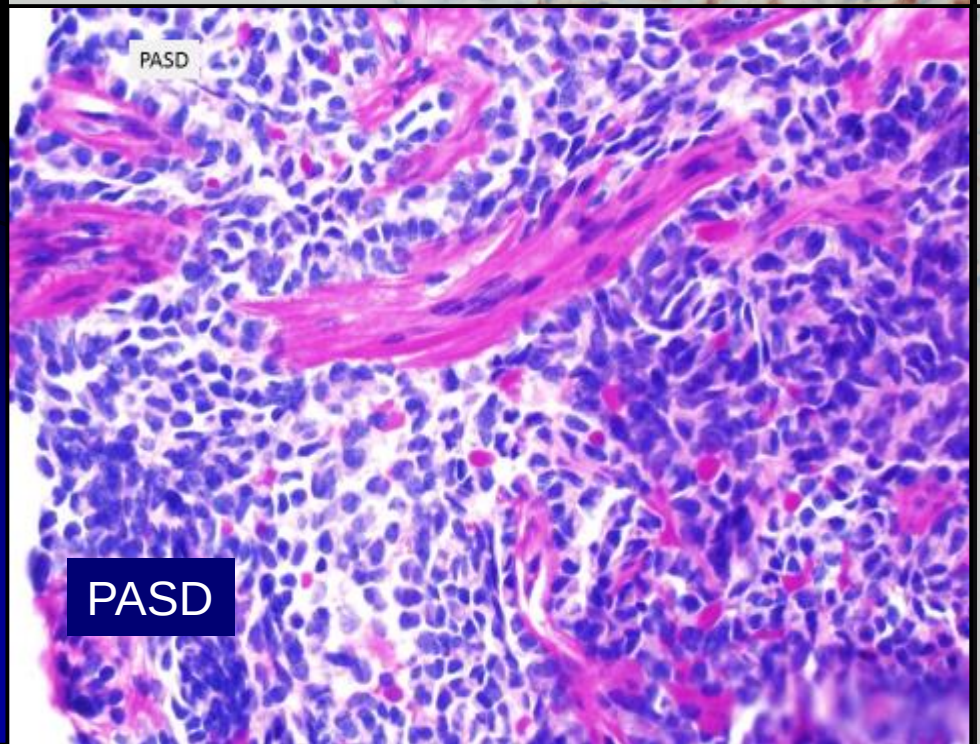


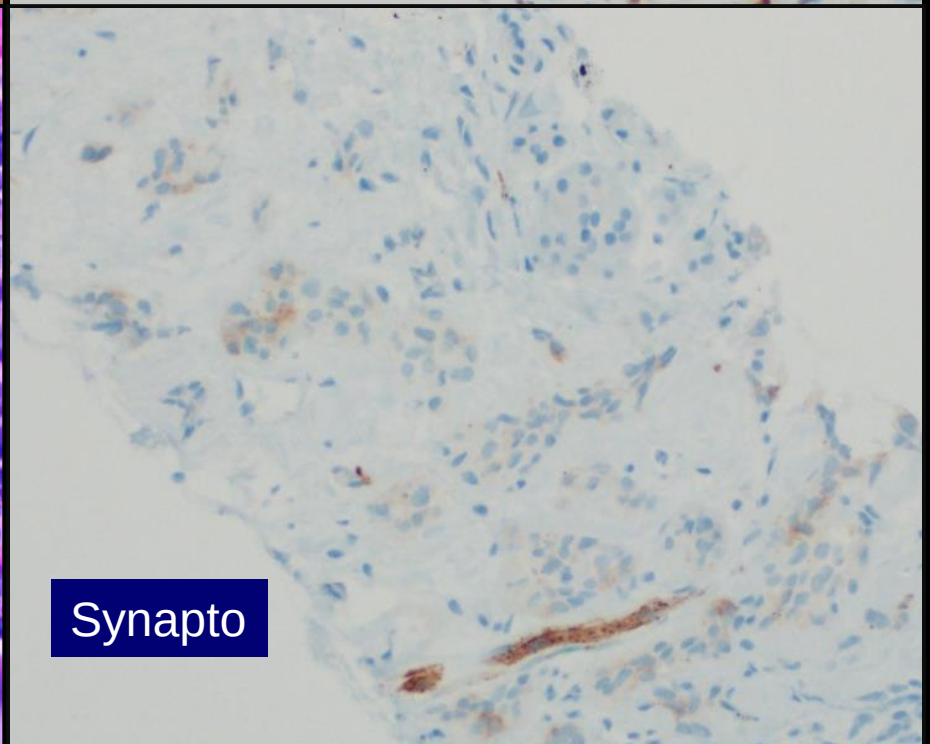


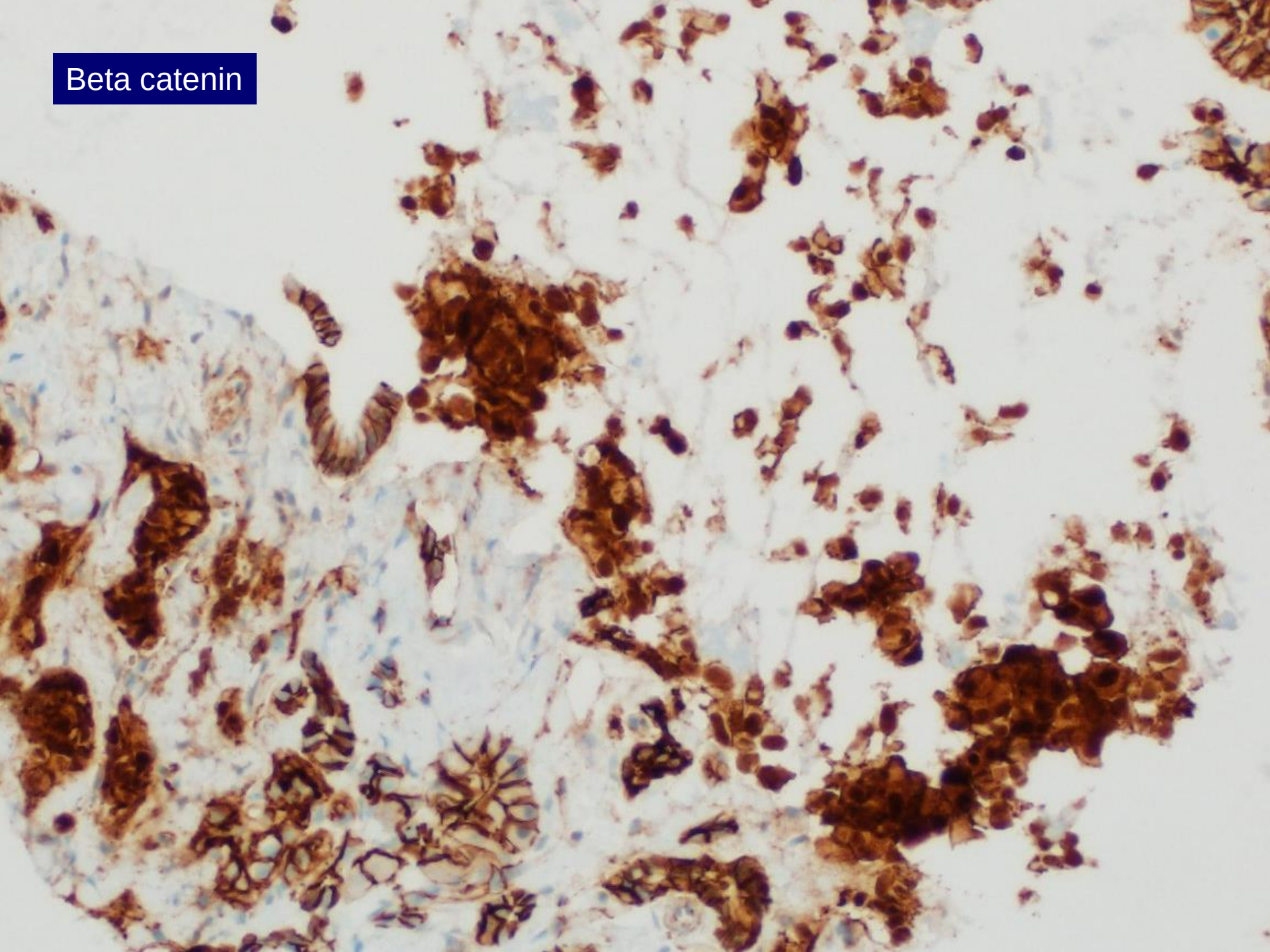

20 µm

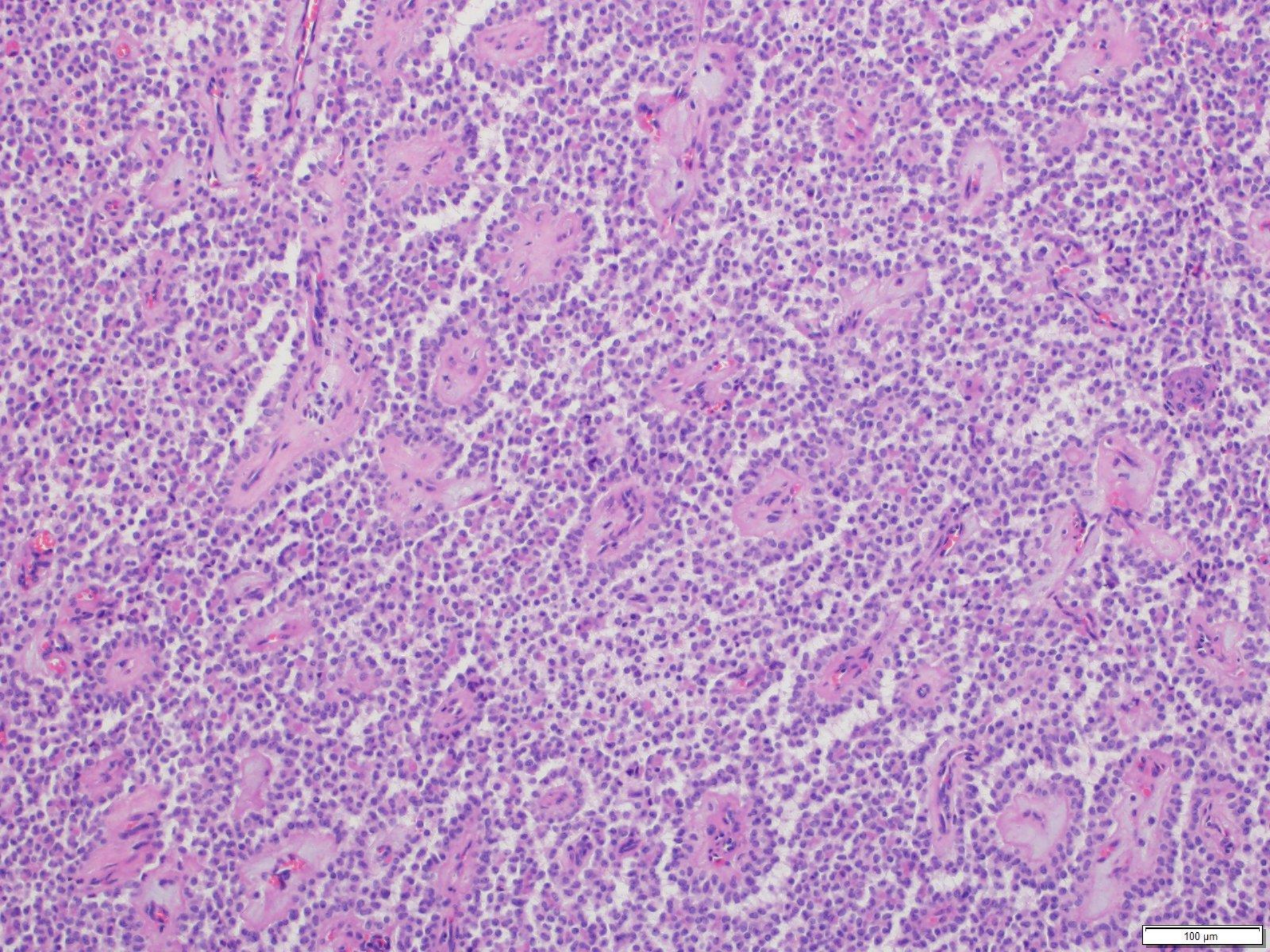

CD10


PR


PASD
PASD

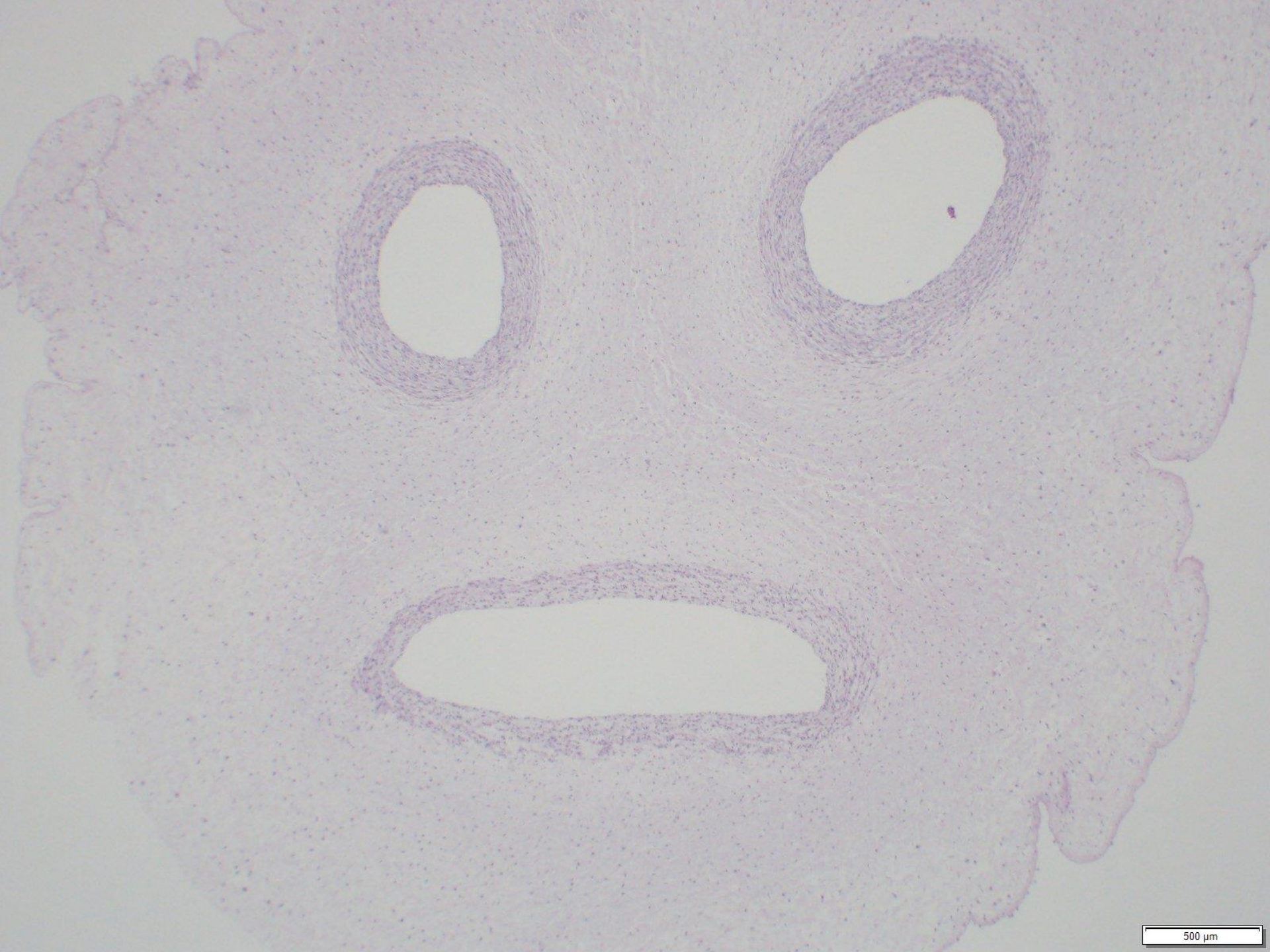

Synapto


Beta catenin

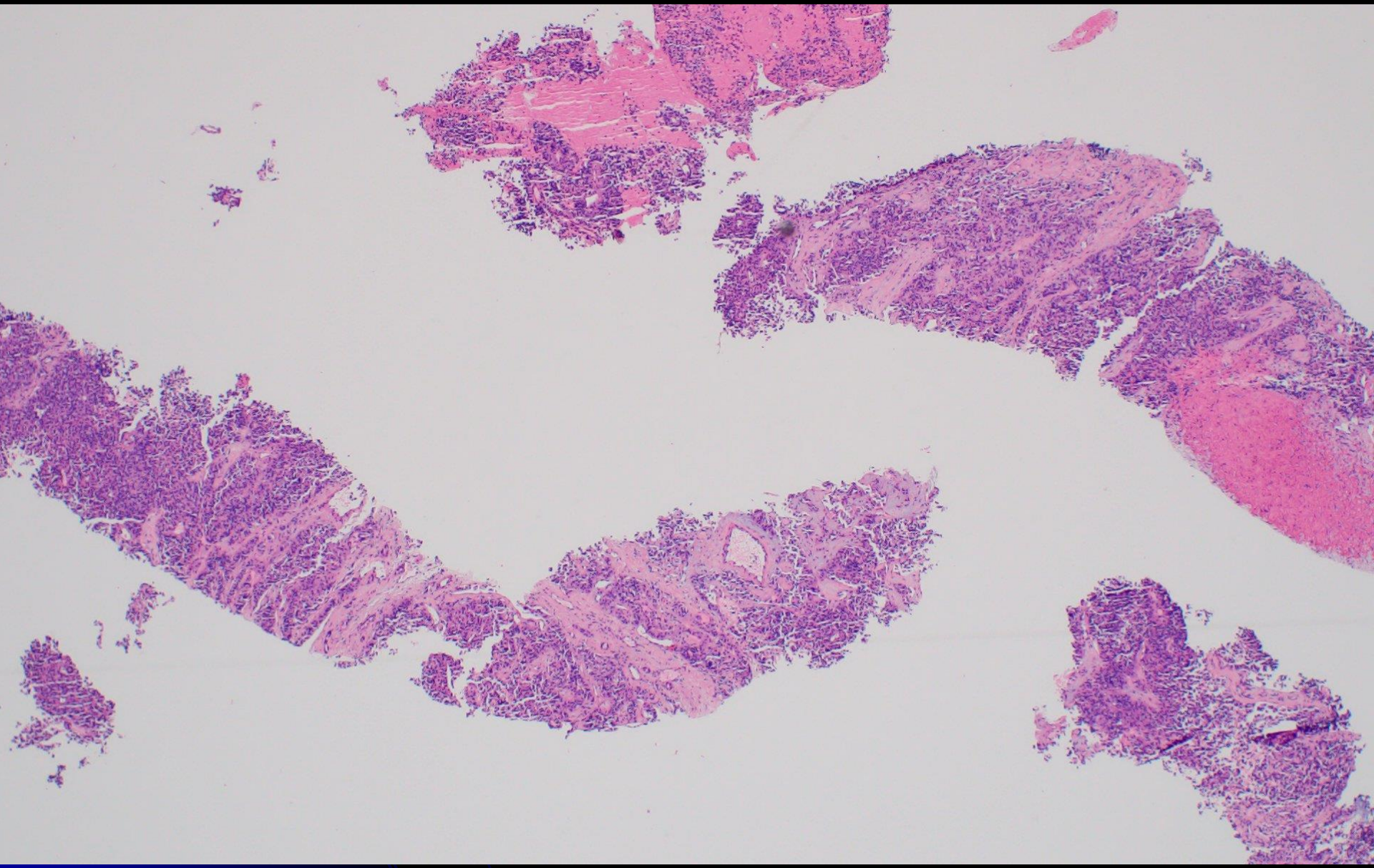

100 µm

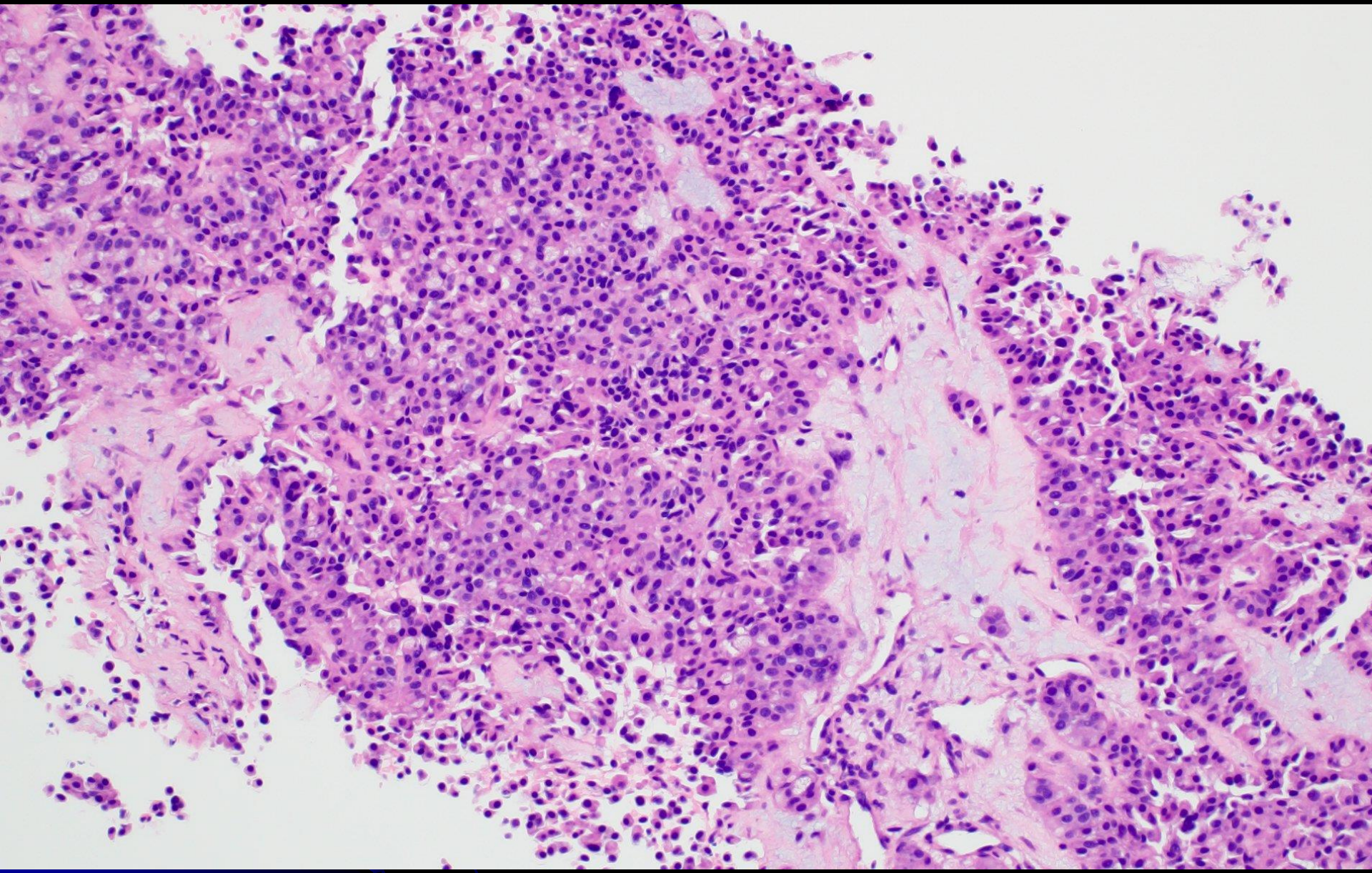
Solid Pseudopapillary Tumor

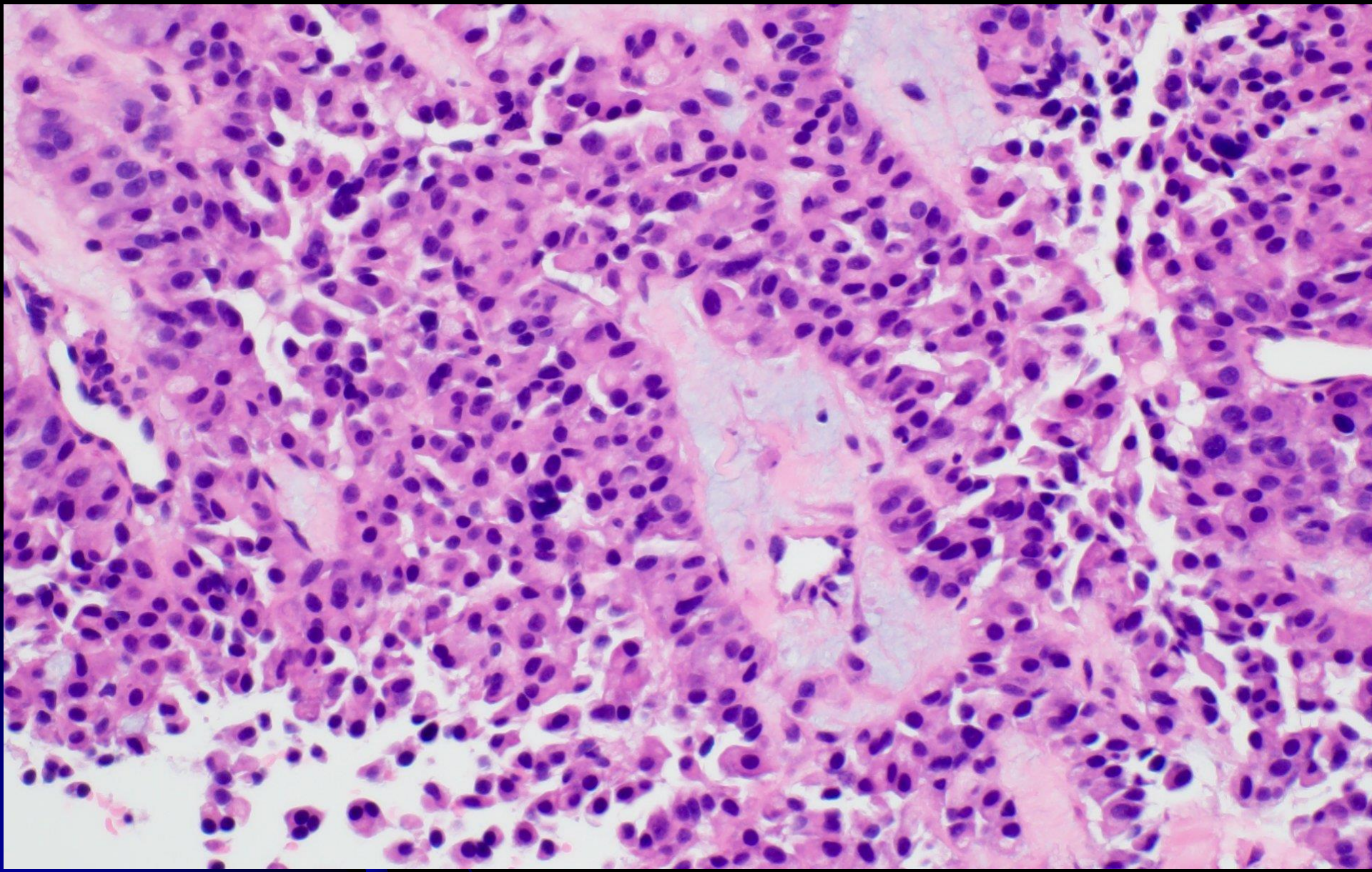
- Common primary pancreatic neoplasm in children
- Low grade malignancy, locally invasive, rare metastasis
- Beta-catenin gene mutation
- Treatment surgical

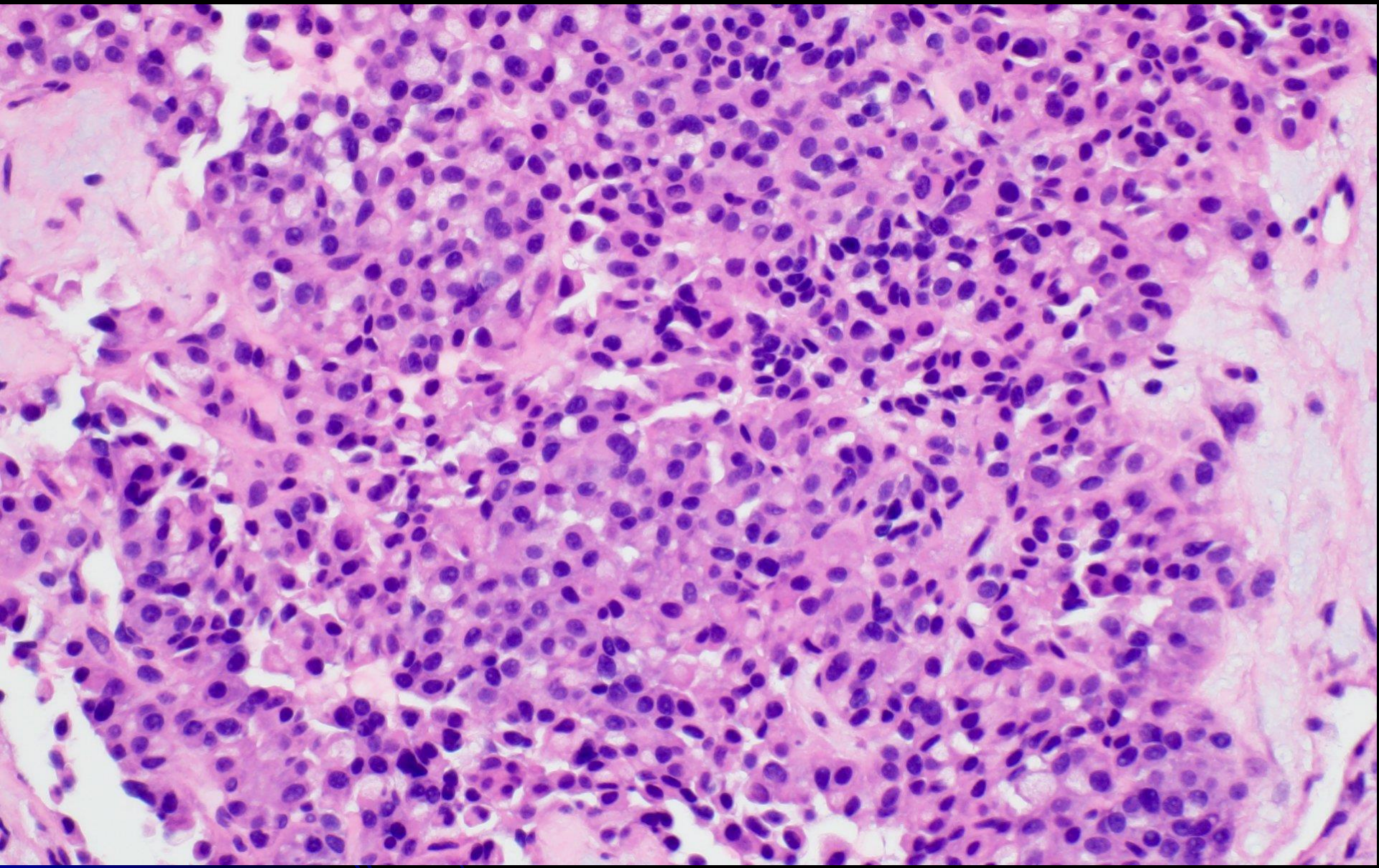

500 µm

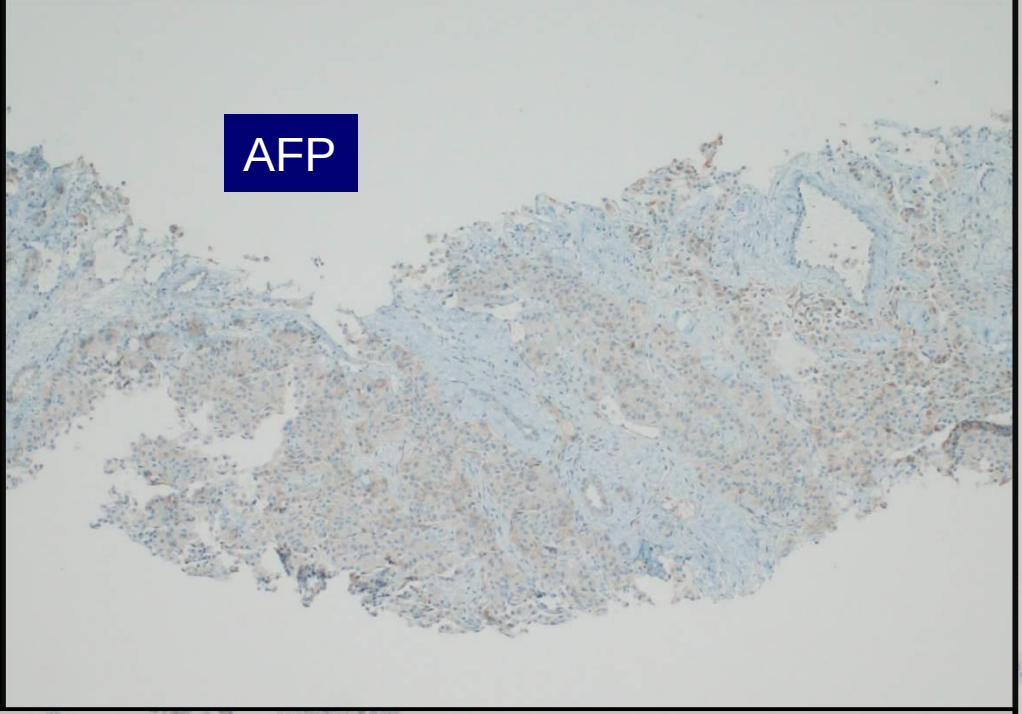
17 year-old girl with liver lesion

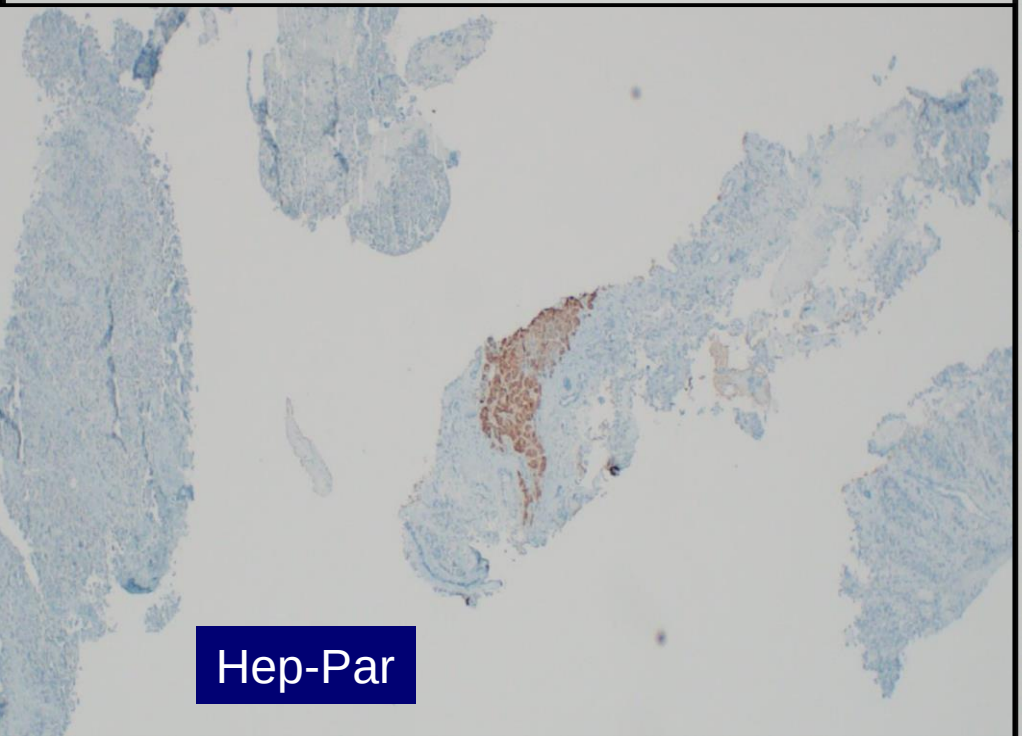


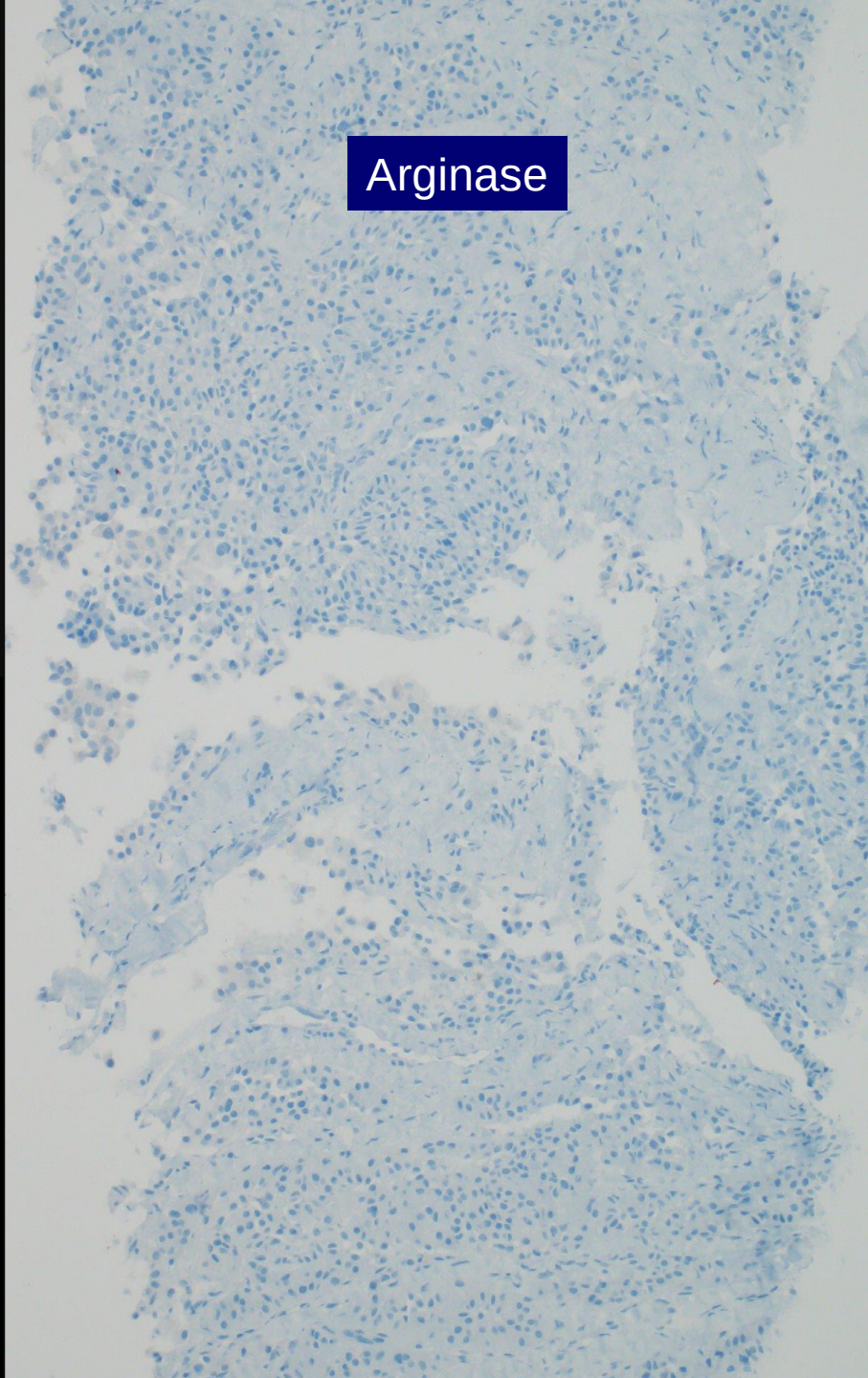


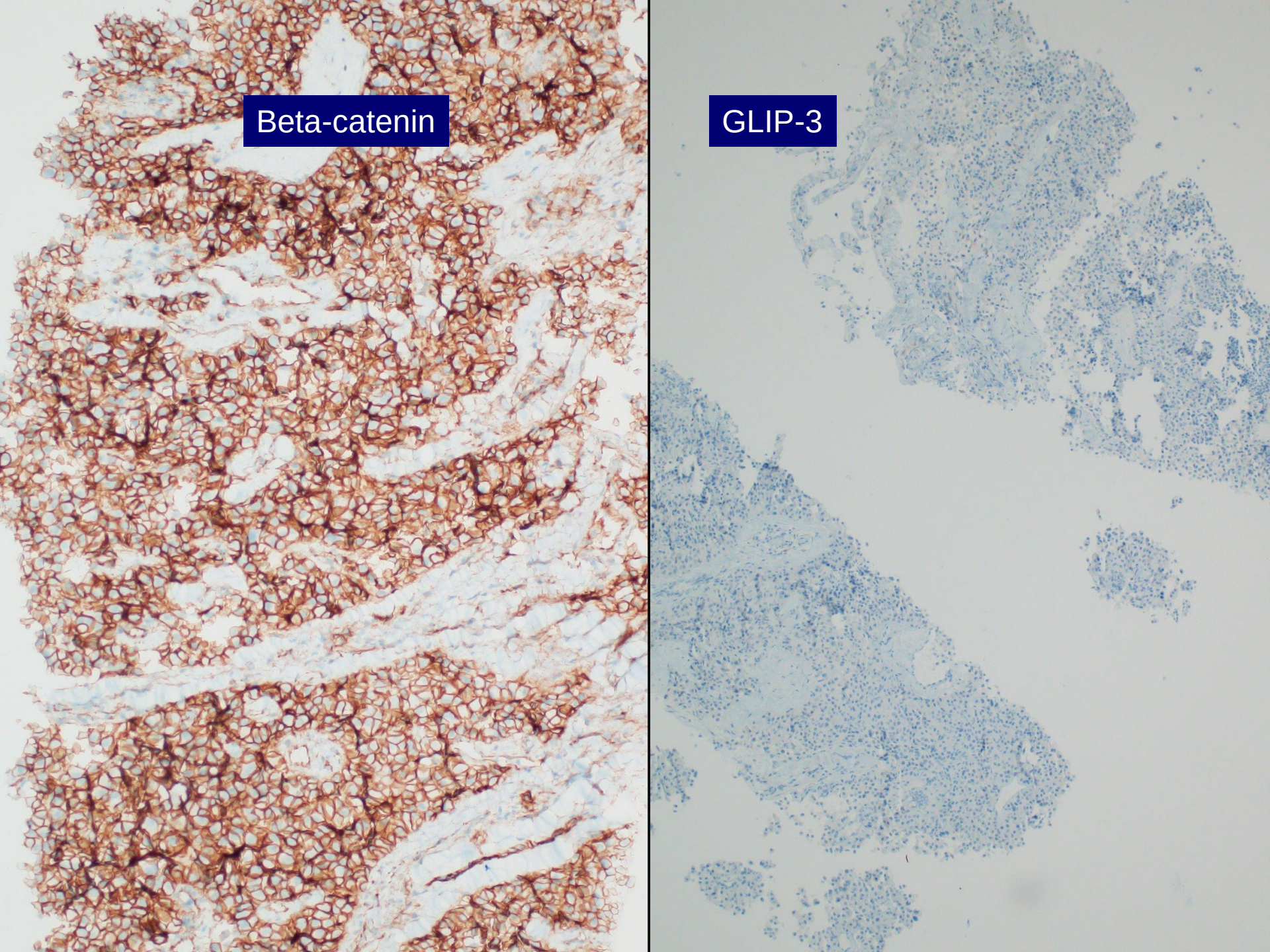


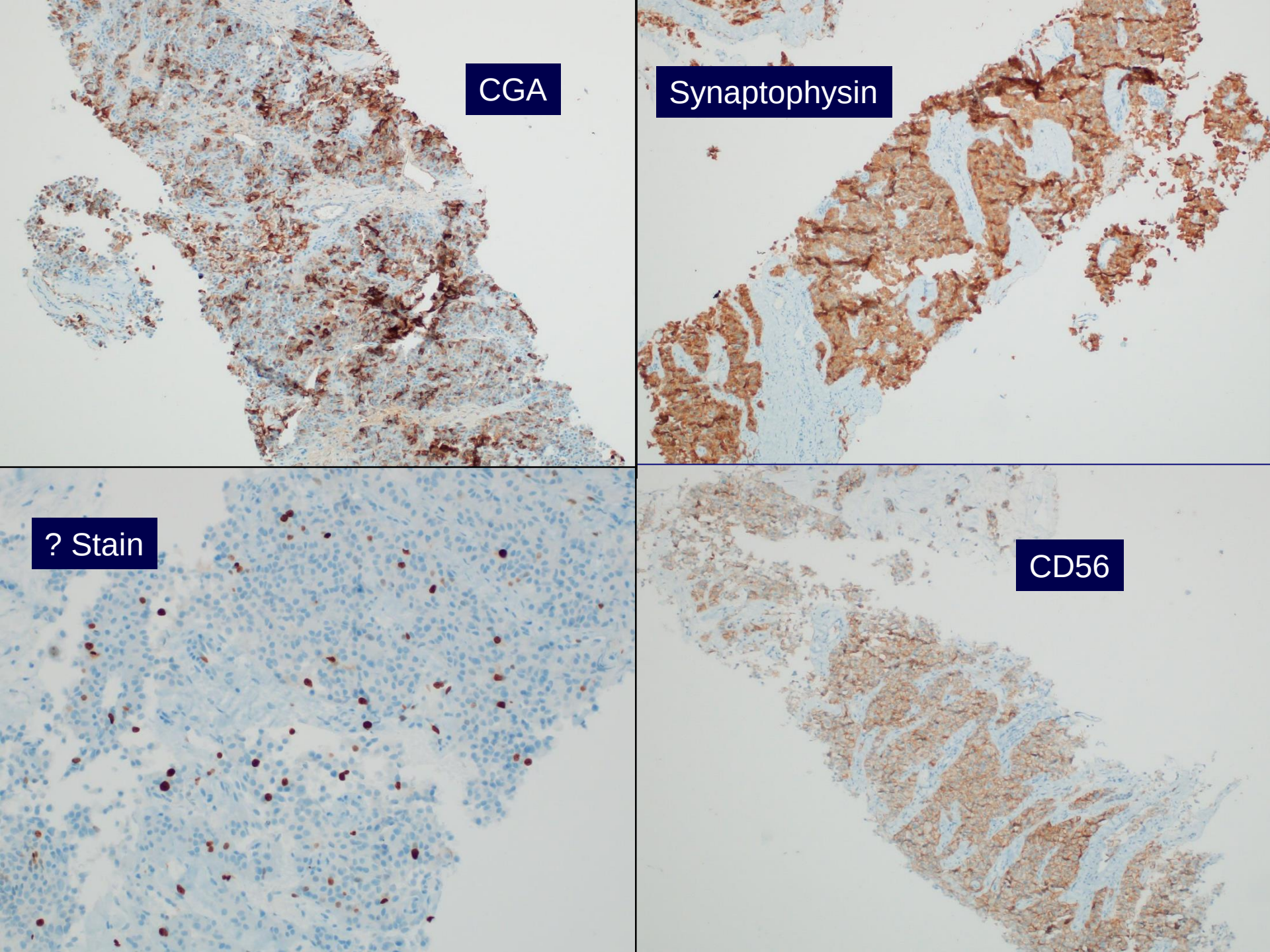



AFP


Hep-Par


Arginase


Beta-catenin
GLIP-3


CGA
Synaptophysin
? Stain
CD56


Thank you!

References

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- Abraham SC, Klimstra DS, Wilentz RE et al. Solid-Pseudopapillary Tumors of the Pancreas Are Genetically Distinct from Pancreatic Ductal Adenocarcinomas and Almost Always Harbor β-catenin Mutations. Am J Pathol. 2002 Apr; 160(4): 1361–1369.