CASE PRESENTATION

Supervised by:

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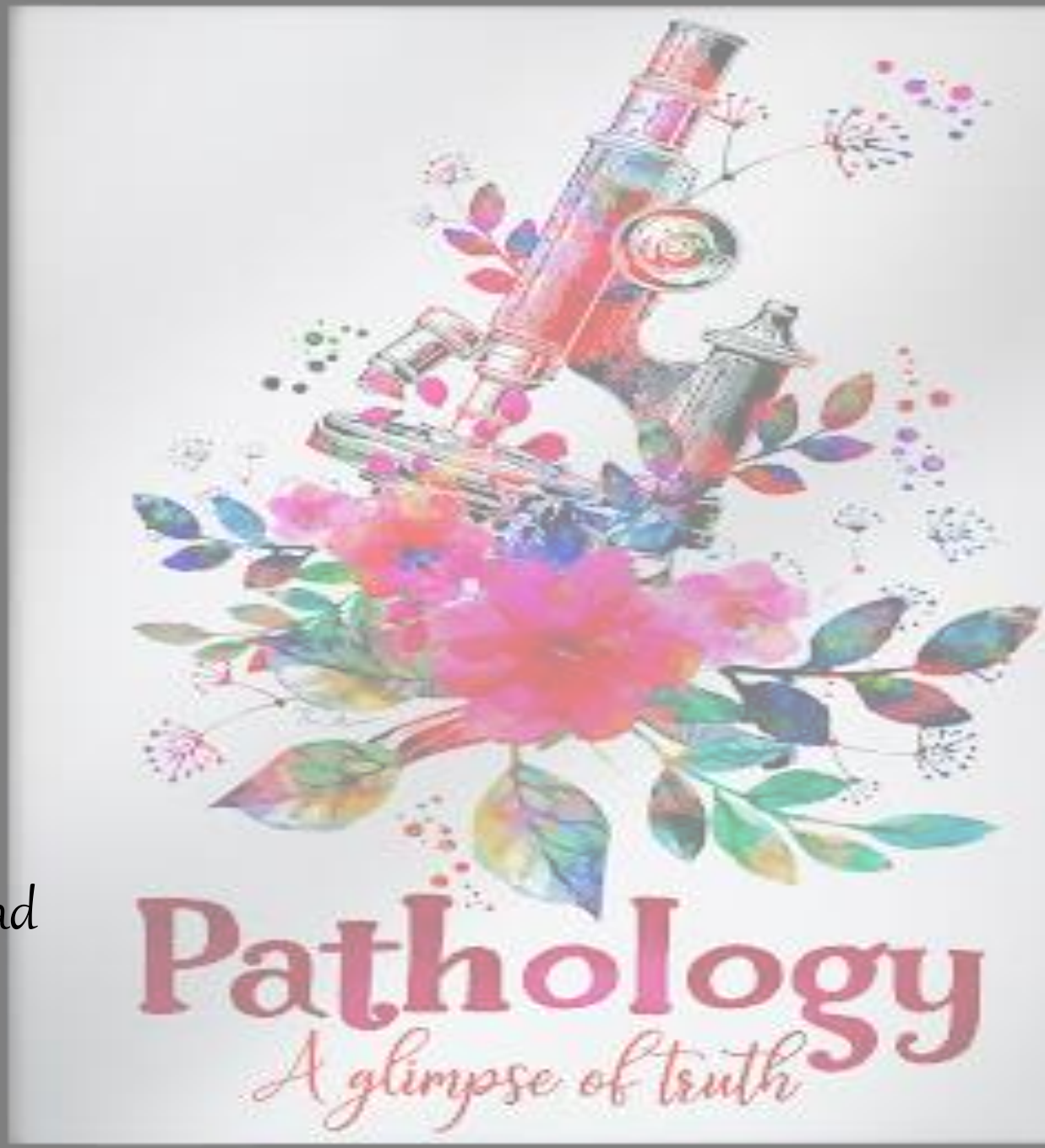
Presented by:Farrin Mahdad



HISTORY

- A 59 year- old female, G3L3, presenting with suprapubic pain, PCB, PCP for 3 months
- 6 year ago menopause
- PMH: HTN-Hypothyroidism-DM
- PSH: 3 NVD
- In physical examination a palpable large abdominopelvic mass and ascites wer noted

سونوگرافی ترانس واژینال رحم و تخمدان ها:

رحم دارای ابعاد 76*40 م م و تغییرات آدنومیوز رویت شد

مخاط اندومتر منظم و ضخامت آن 4 م م است

تخمدان چپ رویت نشد(آتروفیک)

تصویر یک توده سالید به ابعاد 101*87 م م با بوردر نامنظم و دارای فلوی عروقی با PSV= 20cm/s در تخمدان راست رویت شد

مایع آزاد جزیی در لگن مشاهده گردید

مایع آزاد در حفره شکم رویت نشد

سونوگرافی شکم و لگن (TAS/TVS)

کبدحجم واندازه نرمال داشت وفاقدتوده فضاگیربود

اکوی پارانشیم کبد بطور یکنواخت افزایش یافته است که مطرح کننده انفیلتراسیون چربی کبد میباشد .

Mild to moderate fatty liver(grade1to 2)

کیسه صفراجدارصاف ومنتظم باضخامت نرمال داشت وفاقدسنگ بود

مجاری صفراوی داخل و خارج کبدی اتساع غیرطبیعی نداشتند

طحال وپانکراس حجم واندازه واکوی نرمال دارندوفاقدتوده فضاگیربودند

اسپن طحال نرمال و برابربا 80mm میباشد

لنف ادنوپاتی پاراآئورتیک دیده نشد

کلیه هاحجم واندازه نرمال دارندا کوی کورتکسهاوضخامت پارانشیم کلیه ها نرمال بود

طول کلیه راست 104mm و ضخامت پارانشیم کلیه راست و

طول کلیه چپ 109mm و ضخامت پارانشیم کلیه چپ 16mm میباشد

سنگریزه به قطر 2.7mm در کالیس تحتانی و 2.5mm کالیس میانی و 2mm در کالیس فوقانی کلیه راست دیده شد.

هیدرونفروزیا استازادراری درکلیه هادیده نشد

مثانه جدارصاف ومنتظم باضخامت نرمال داشت وفاقدسنگ بود

رحم حجم واندازه واکوی نرمال داشت وفاقد توده فضاگیربود

ابعاد رحم نرمال و برابربا 86x46mm میباشد

اندومتر به ضخامت 4mm میباشد

تصویر یک توده هتروژن تا ایزواکو به ابعاد تقریبی 100x60mm واجد نواحی کیستیک داخلی و فلوی تروقی prominent در محل اددنکس راست دیده شد در مجاورت ان ضایعه مشابه به ابعاد 37x30mm واجد واسکولاریتی مشهود است لذا در درجه اول malignant ovarian lesions مطرح است تطبیق با تومورمارکرهای تخدمان توصیه میشود.(O=5) .

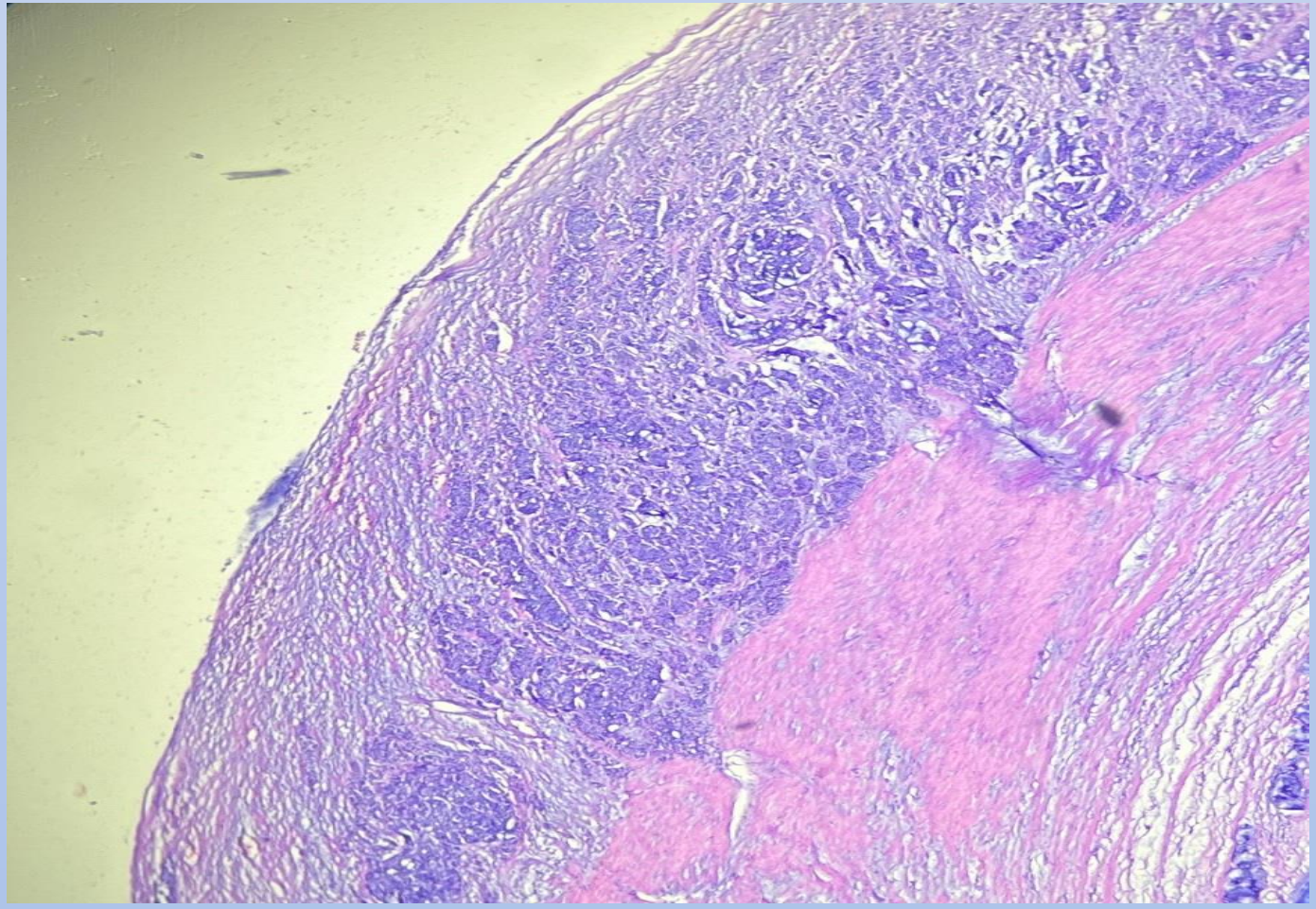
مایع ازاد خفیف در حفره لگن در اطراف توده مذکور دیده شد .

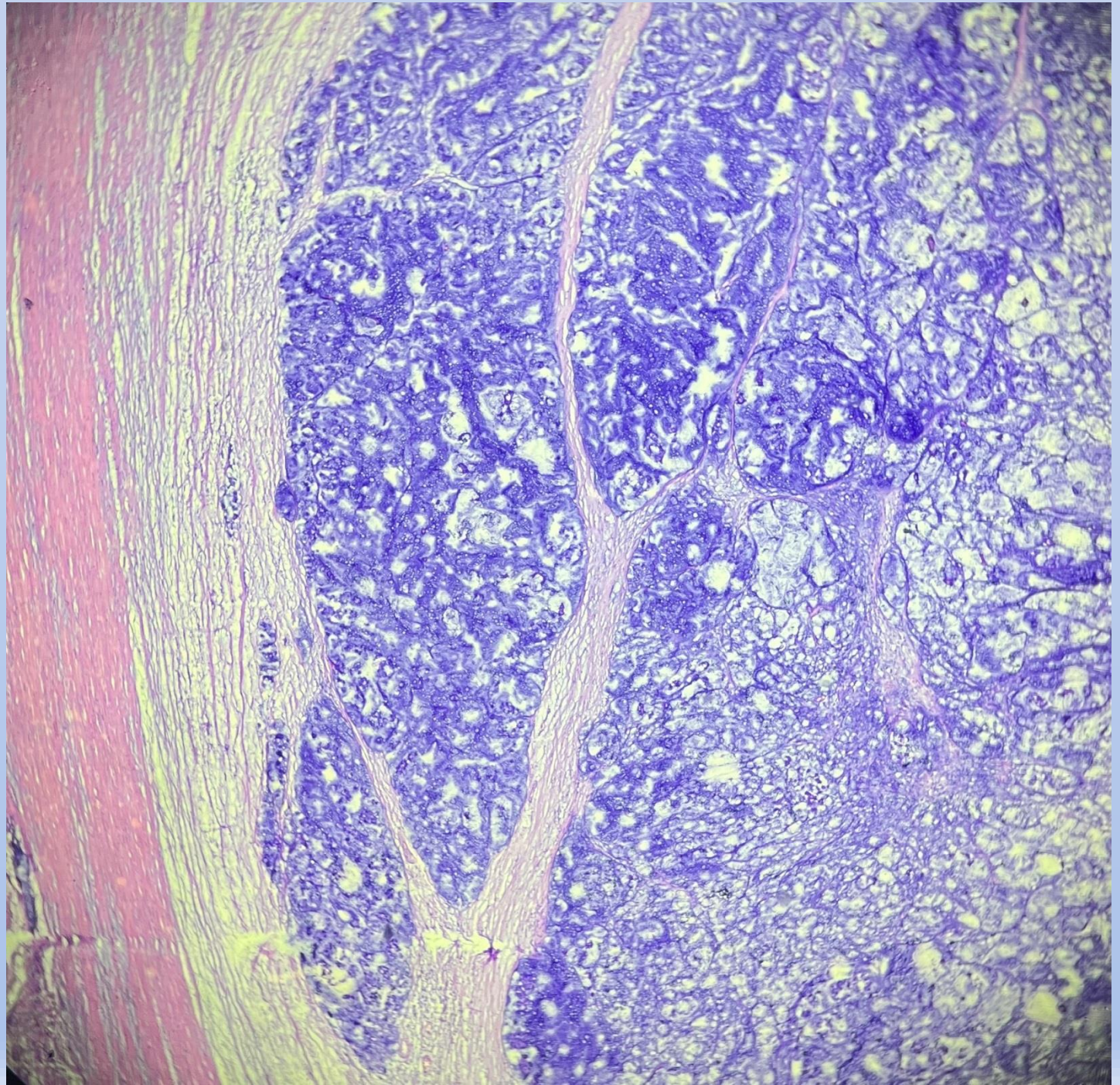
دربررسی حفره شکم مایع ازاد دیده نشد

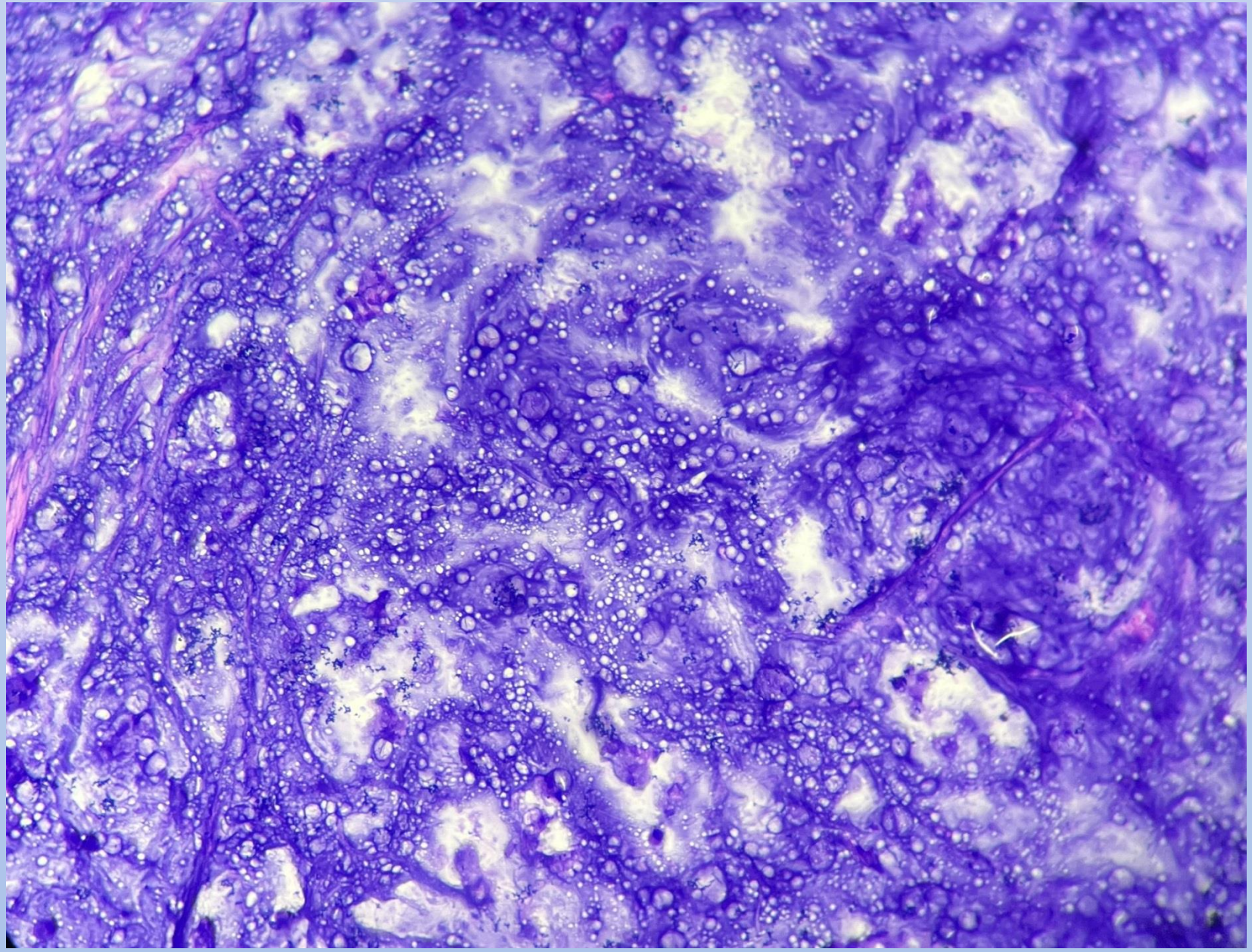
Ovary

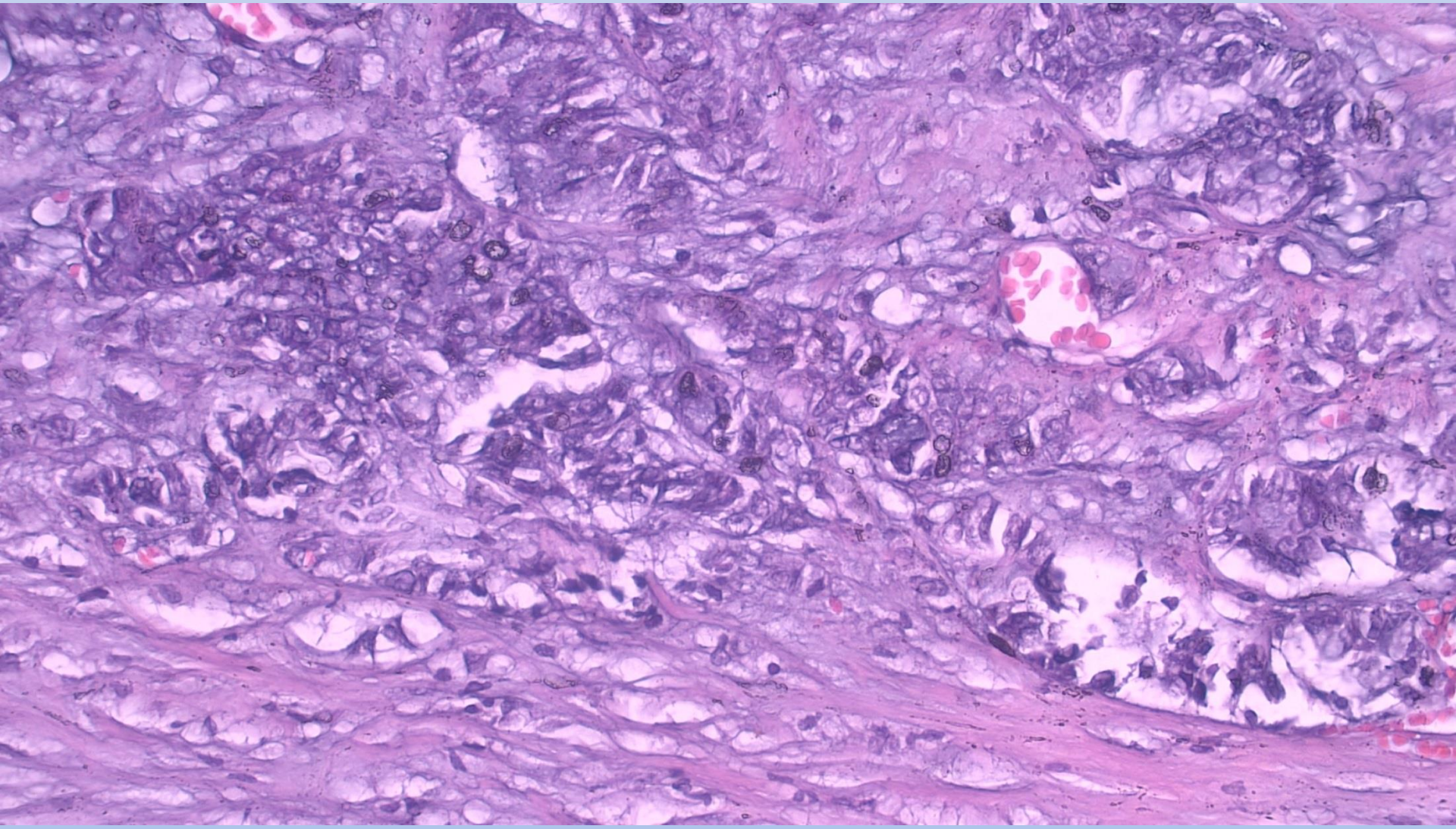
Ovarian cut surface

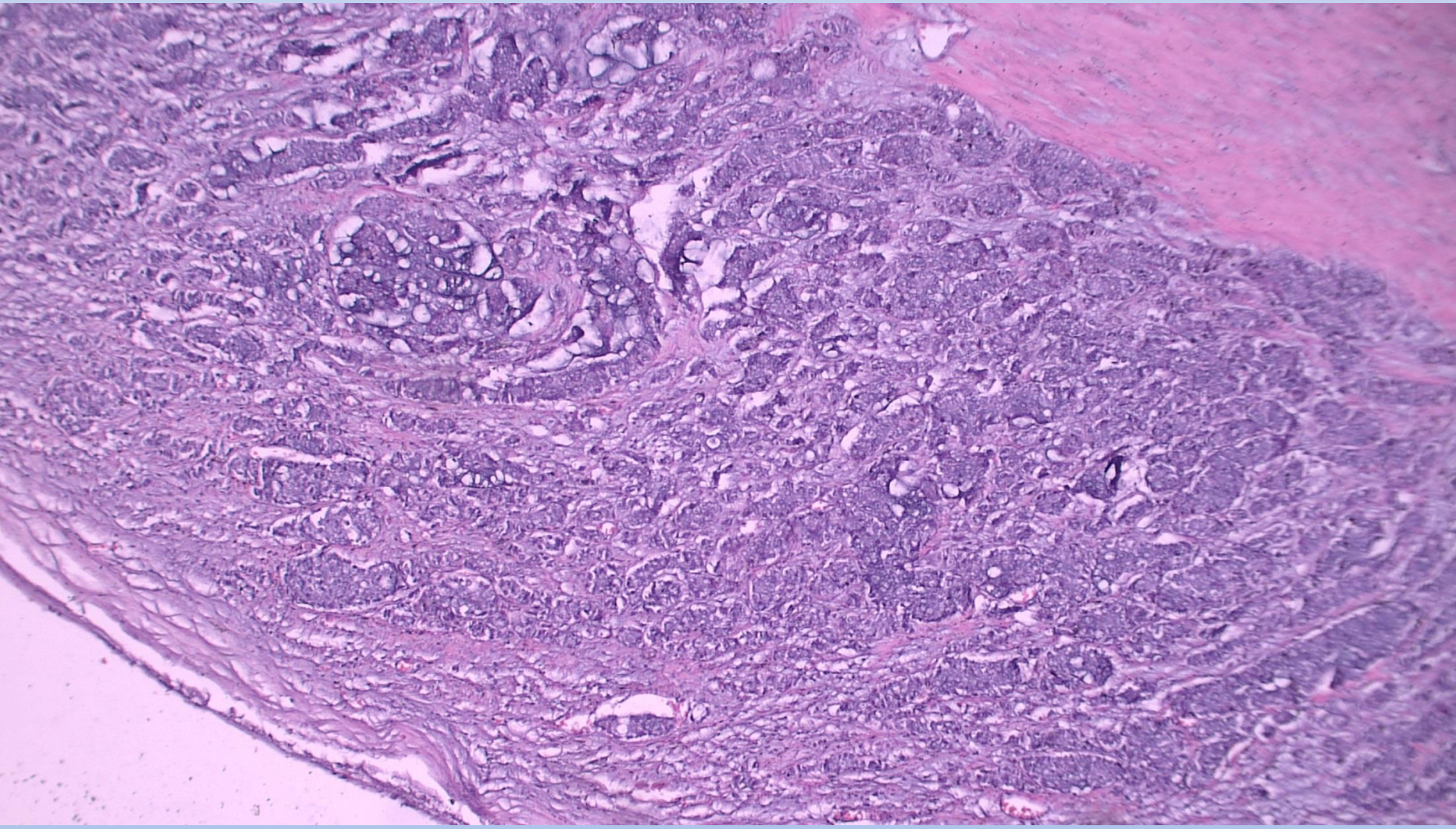
Appendix

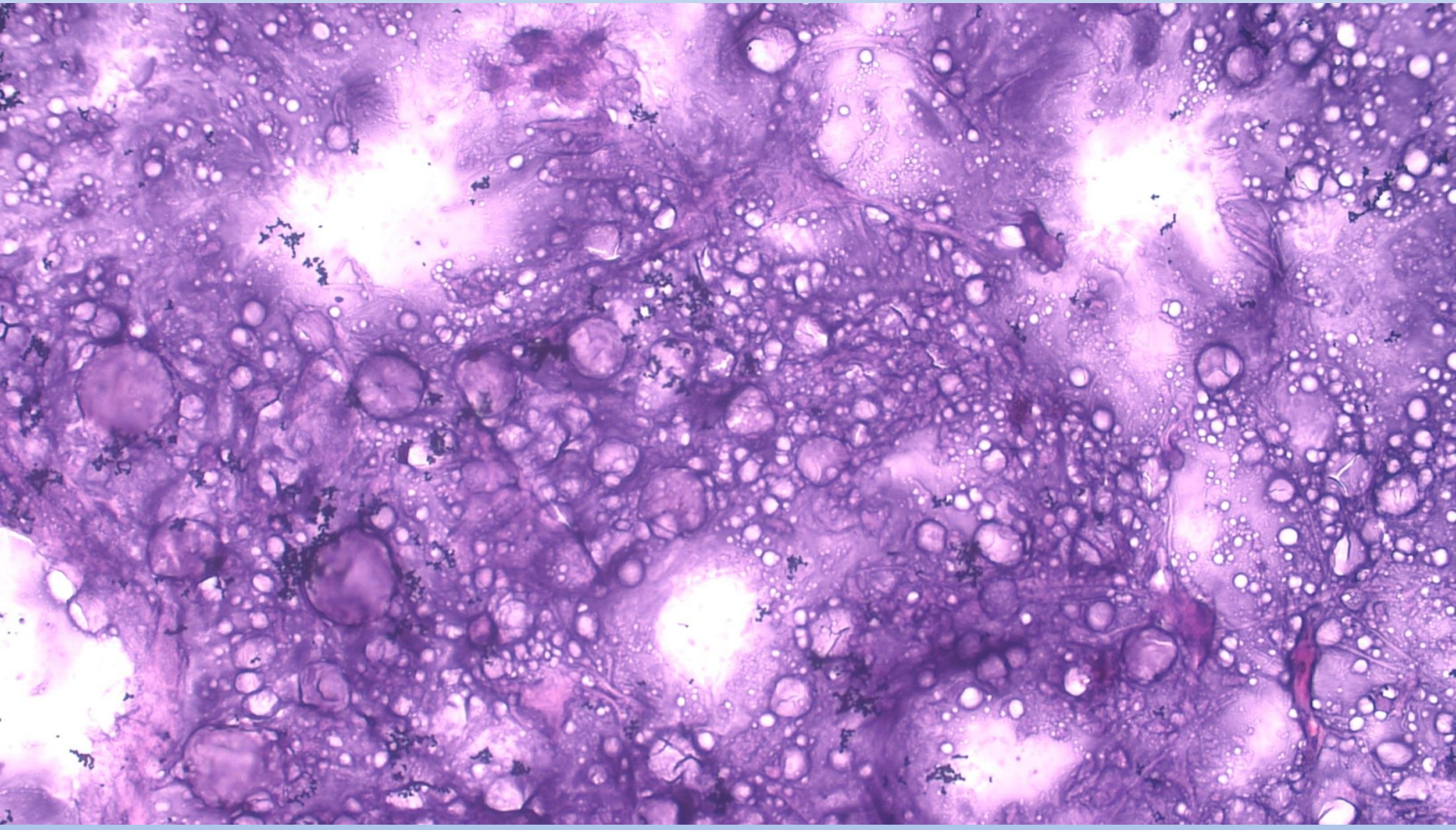


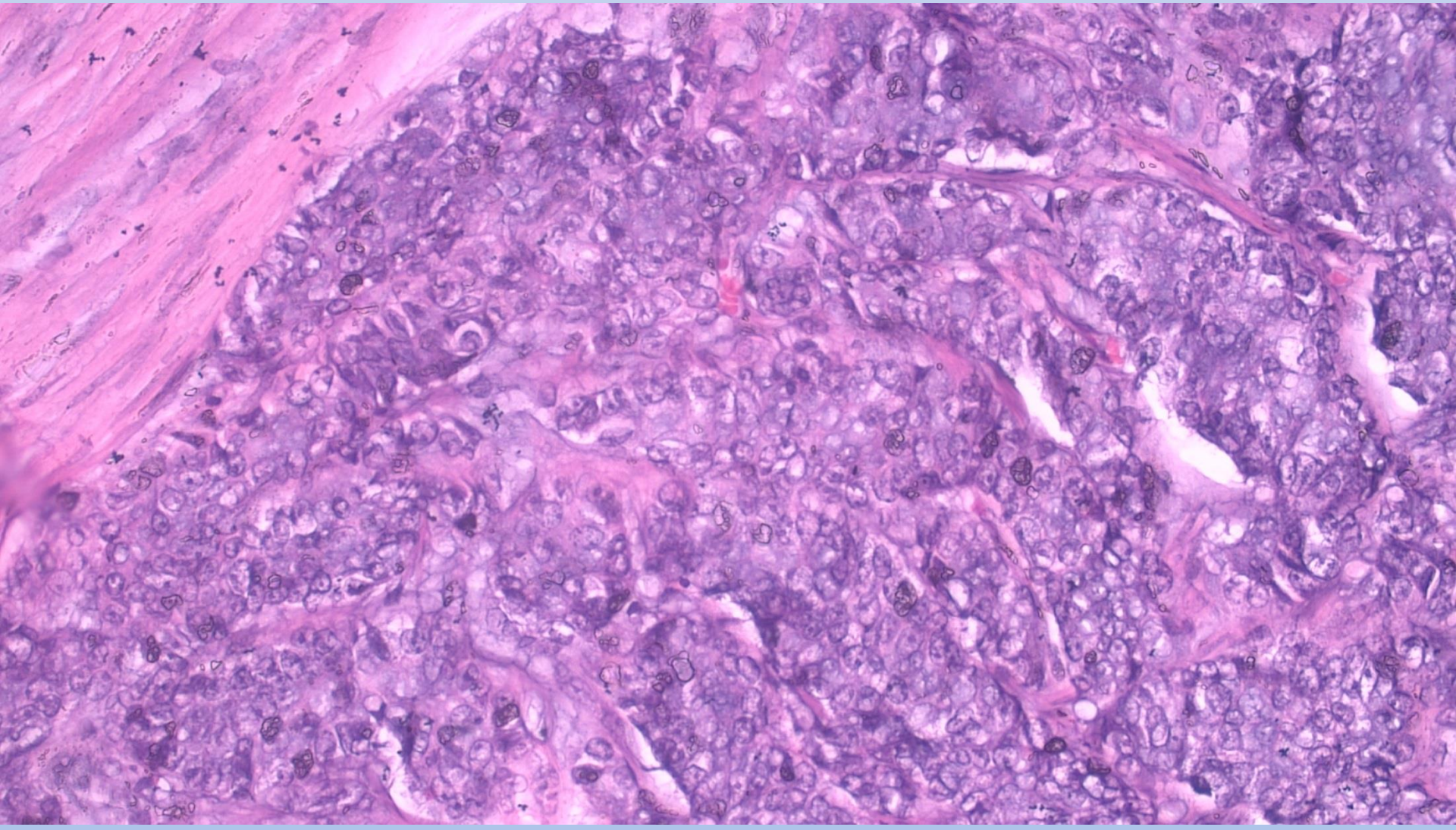


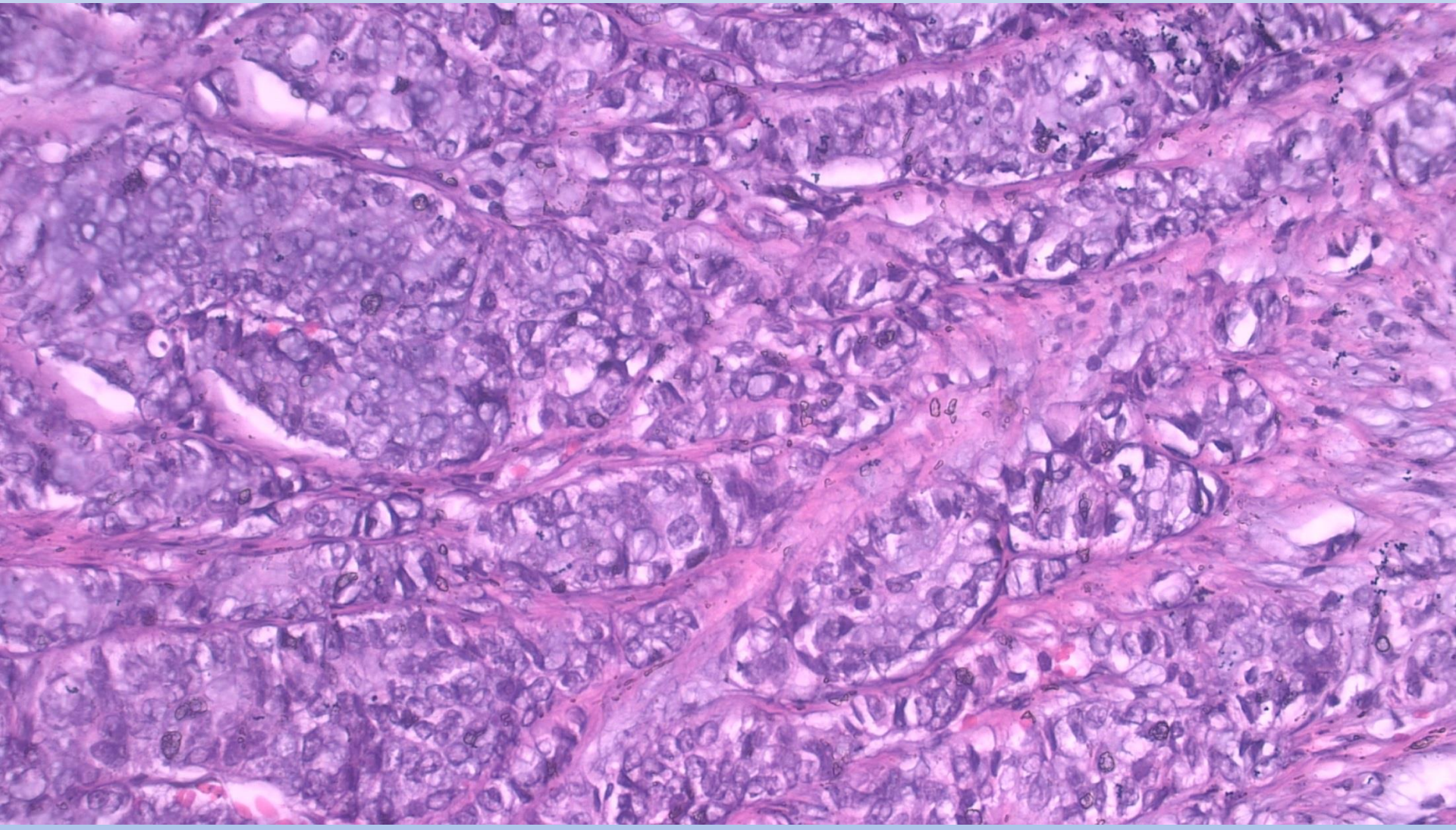


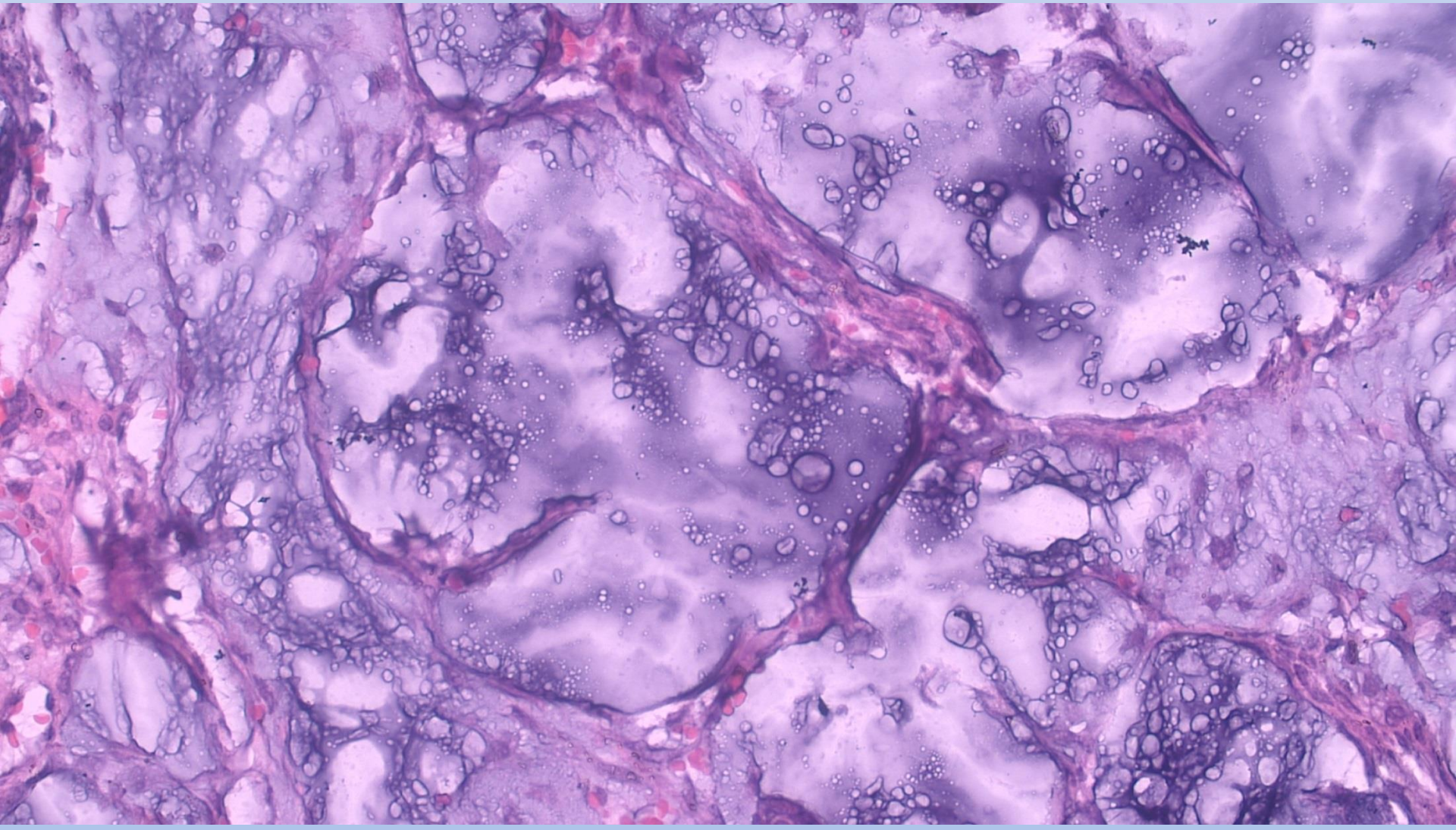


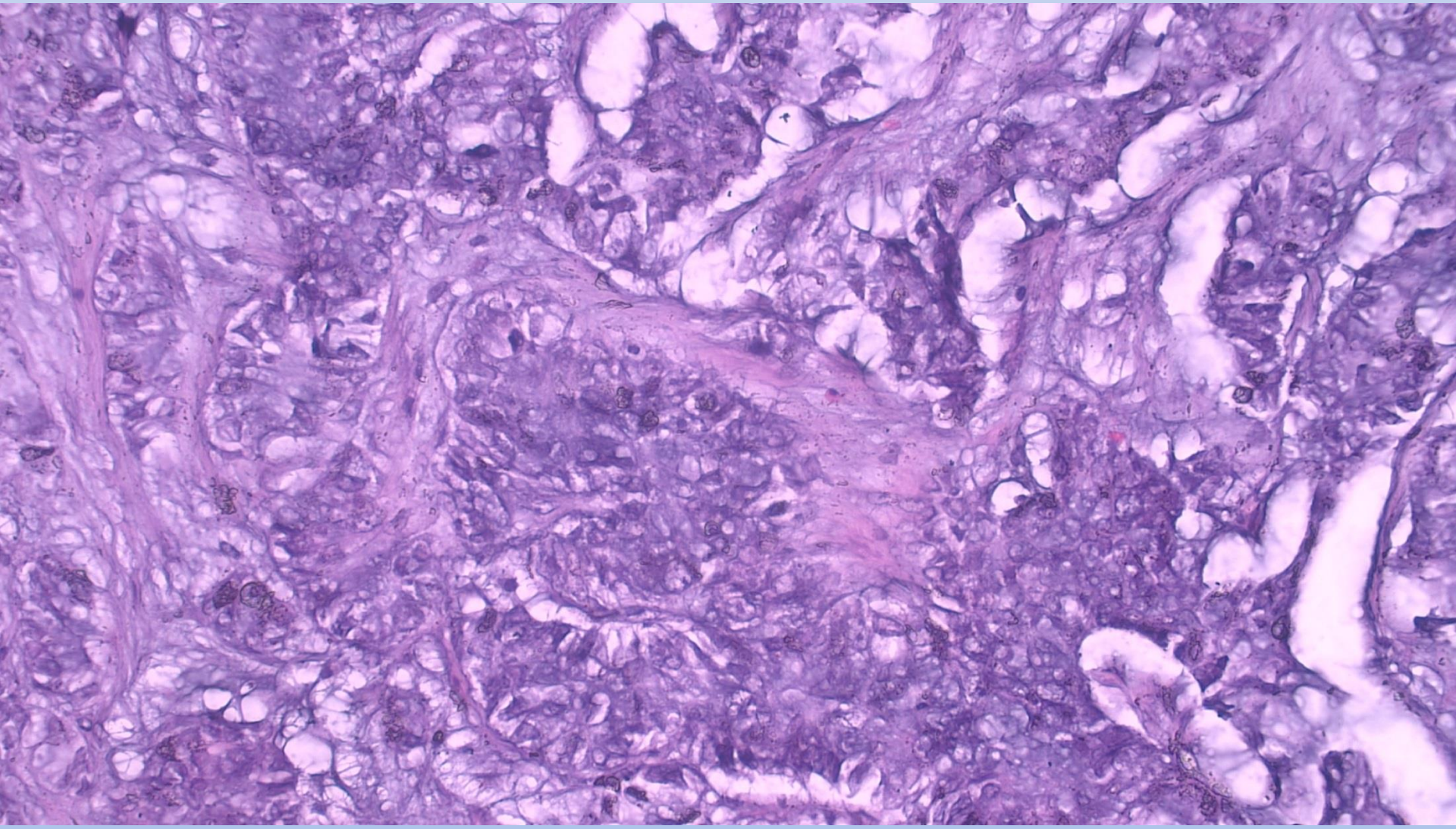


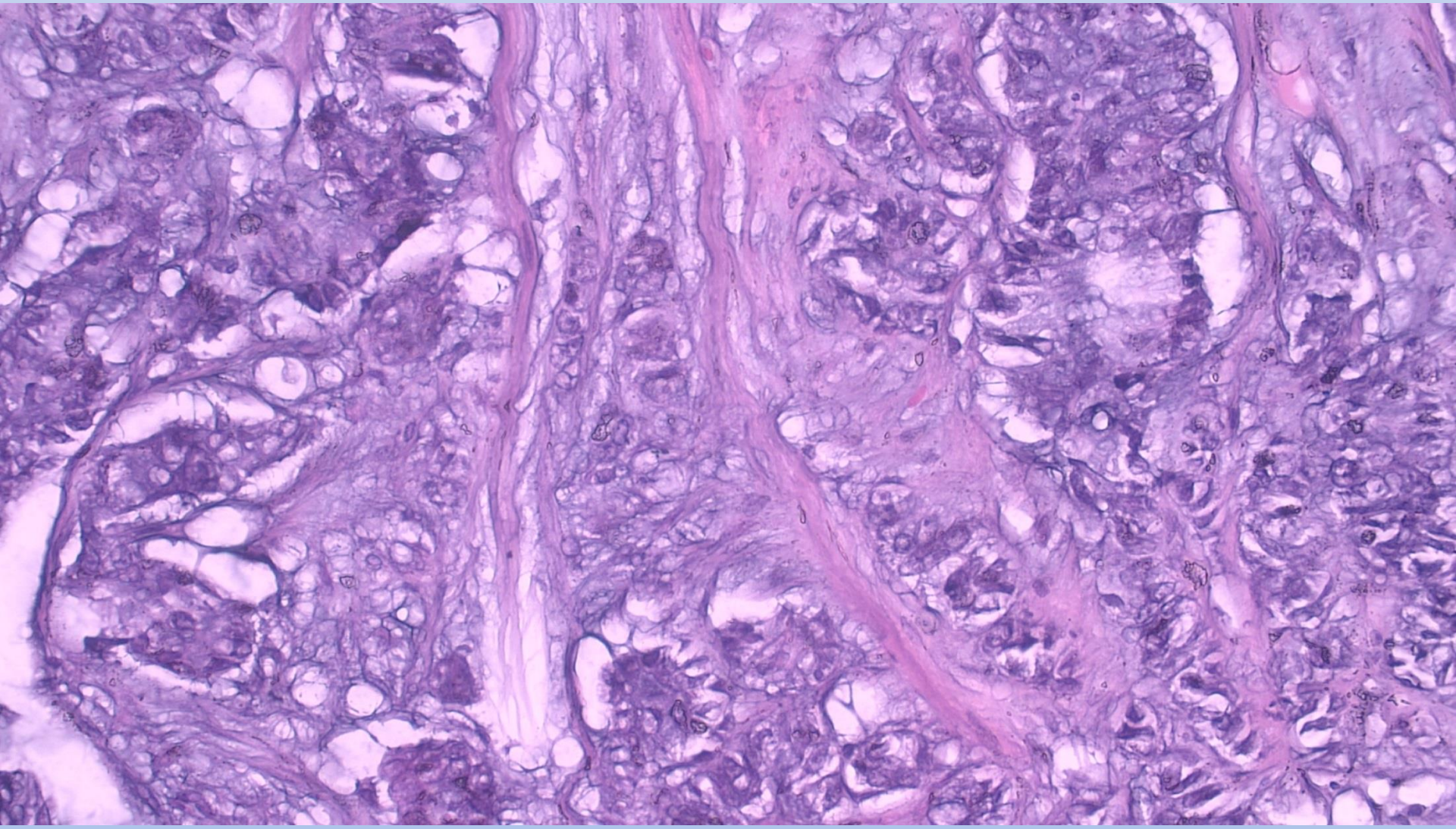


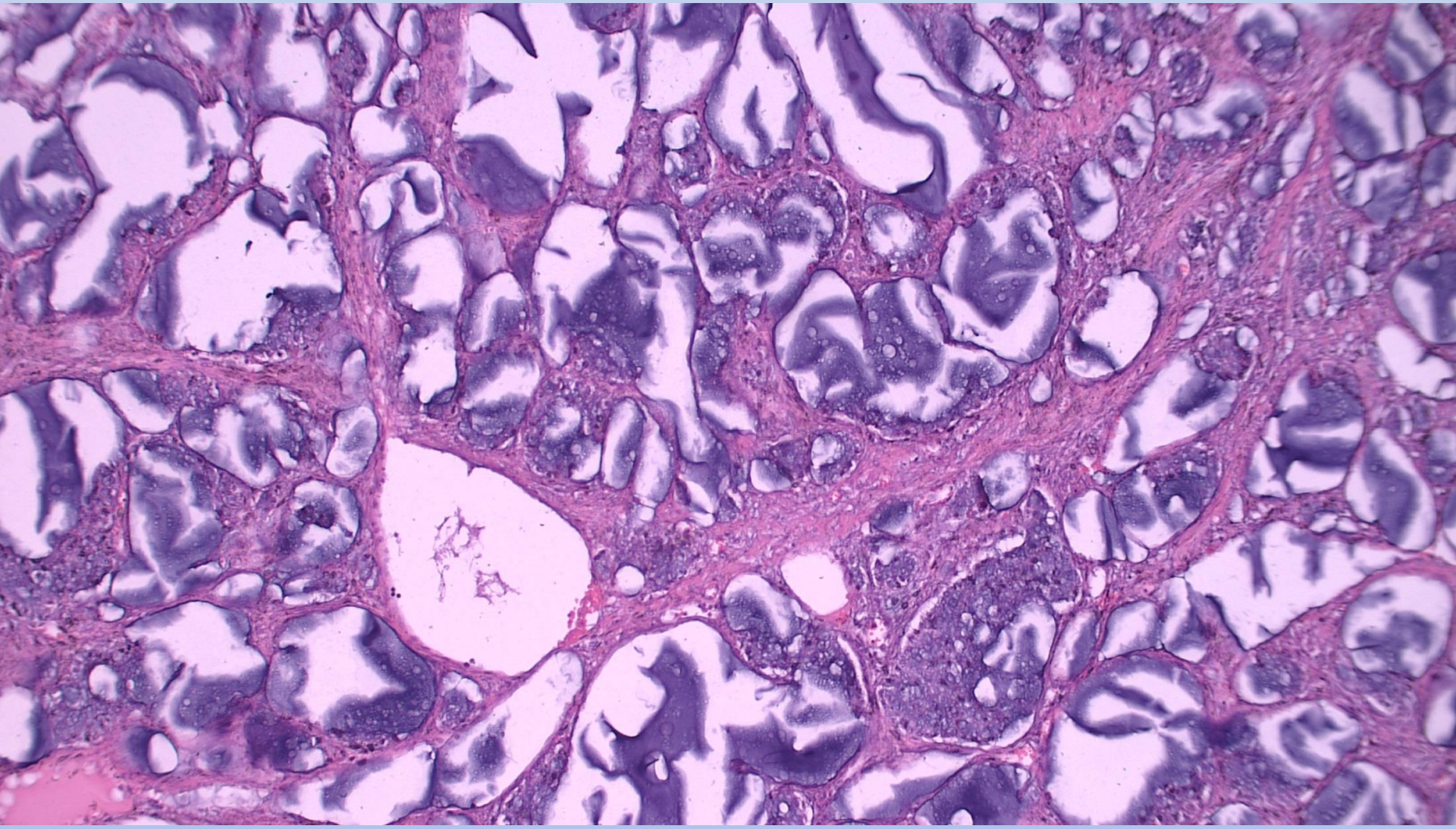


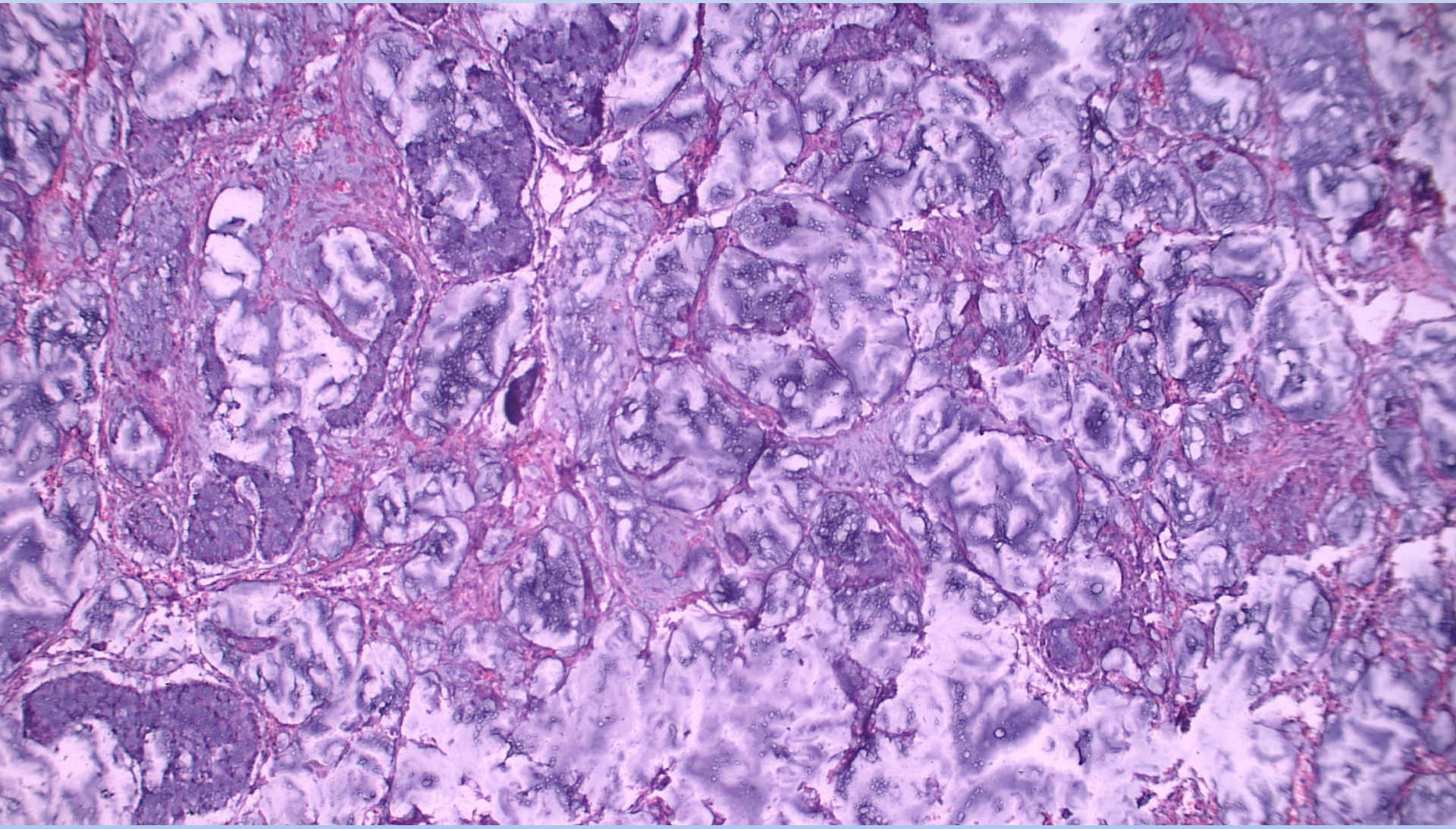


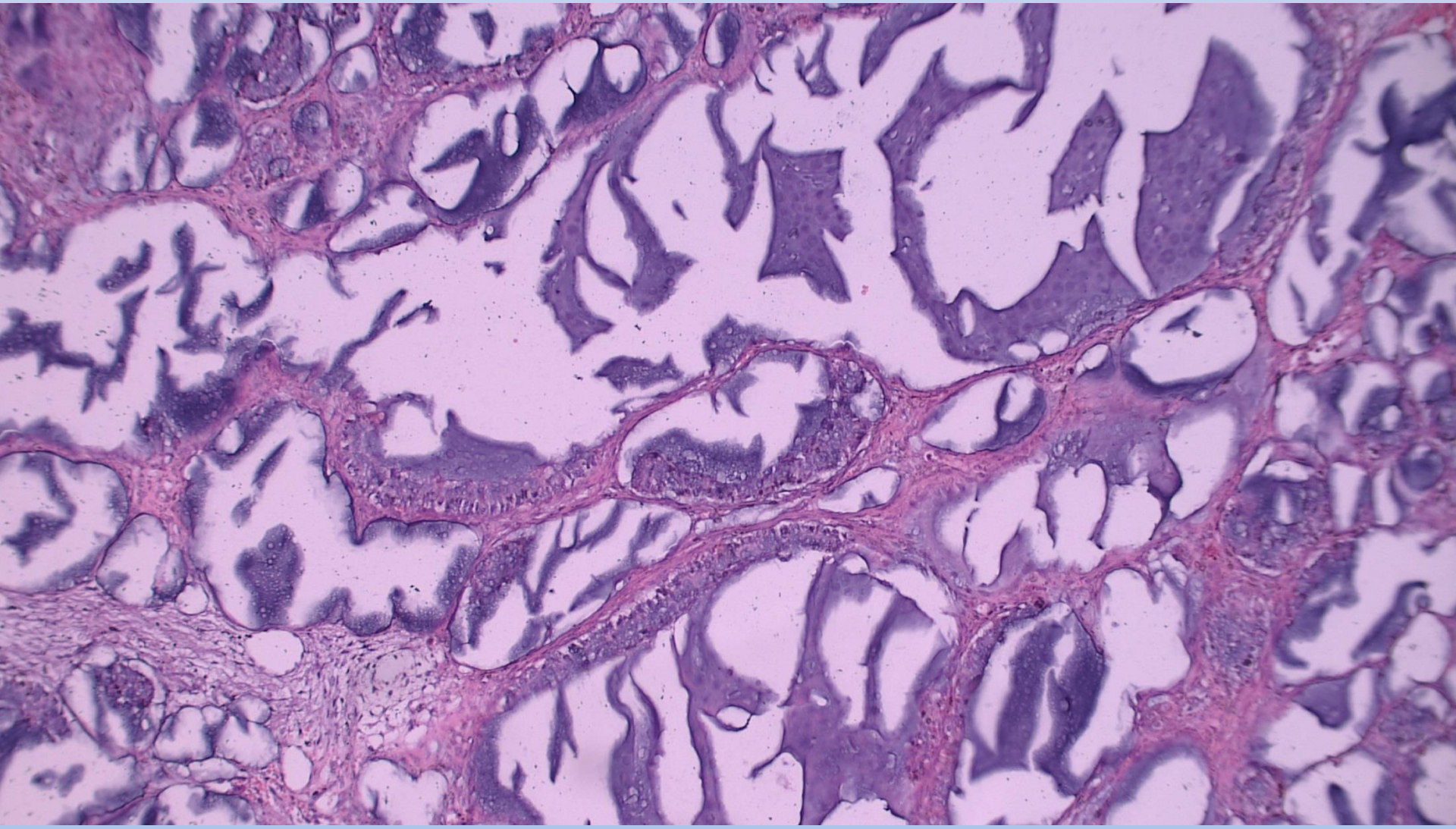


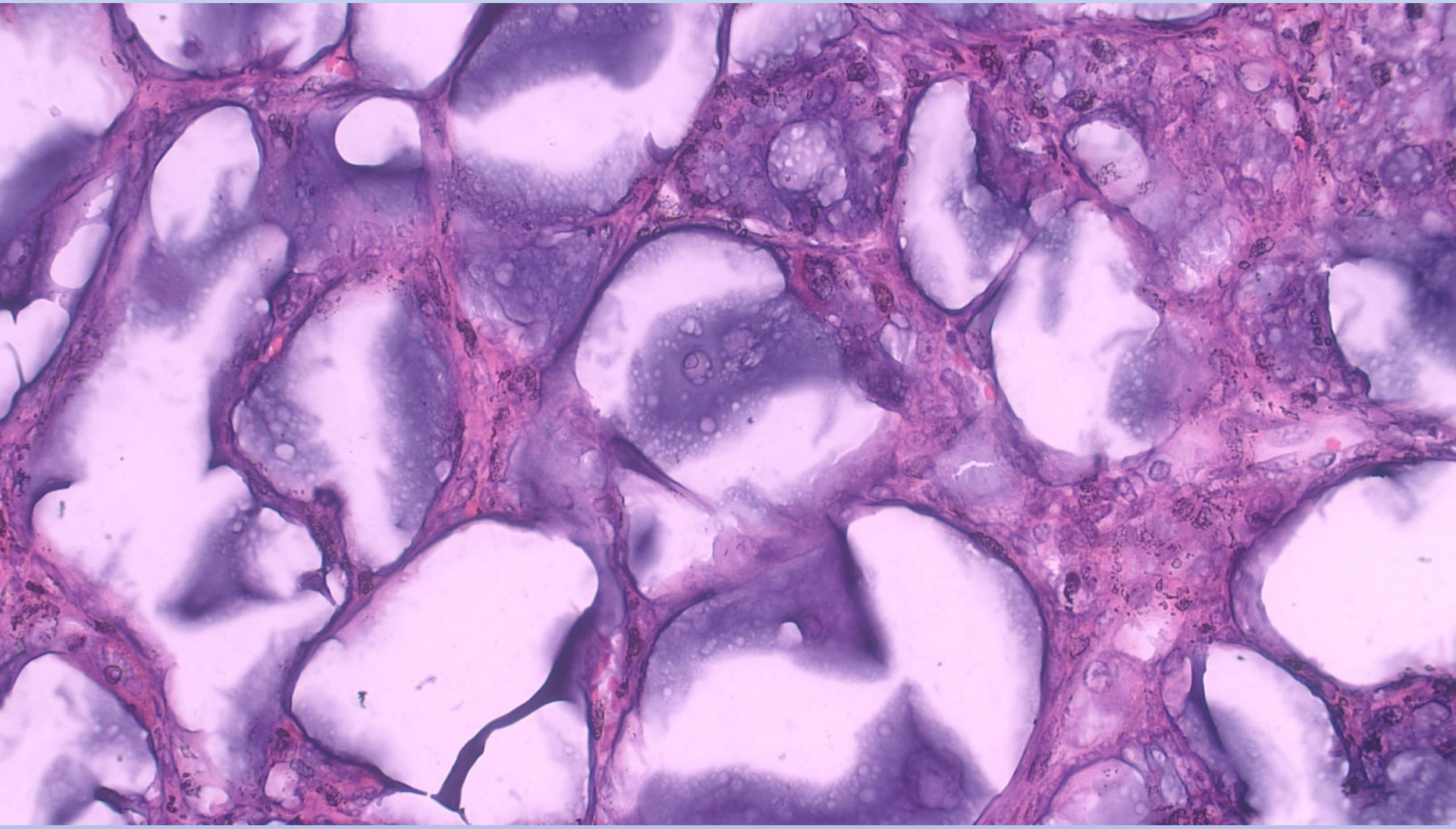


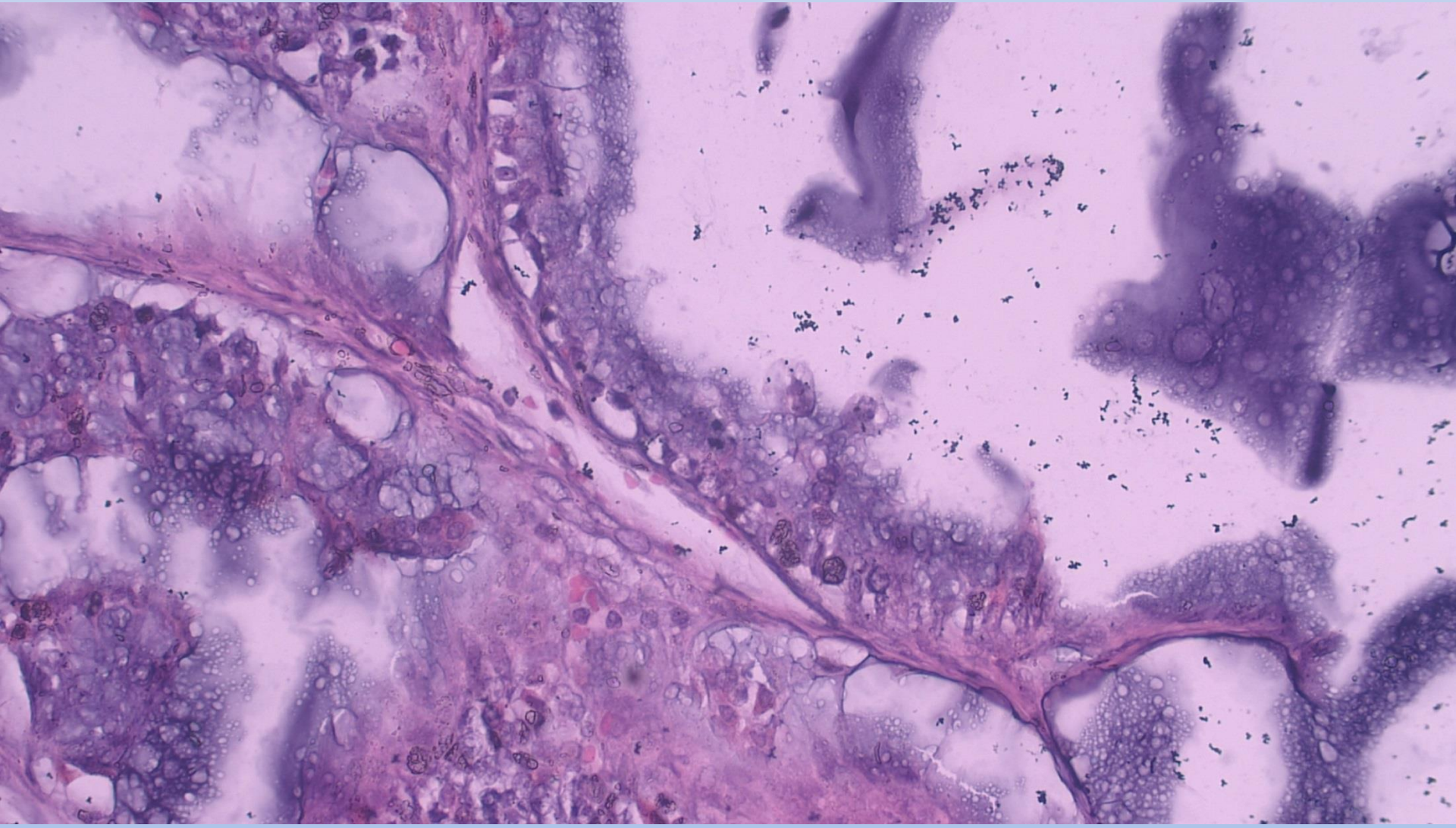


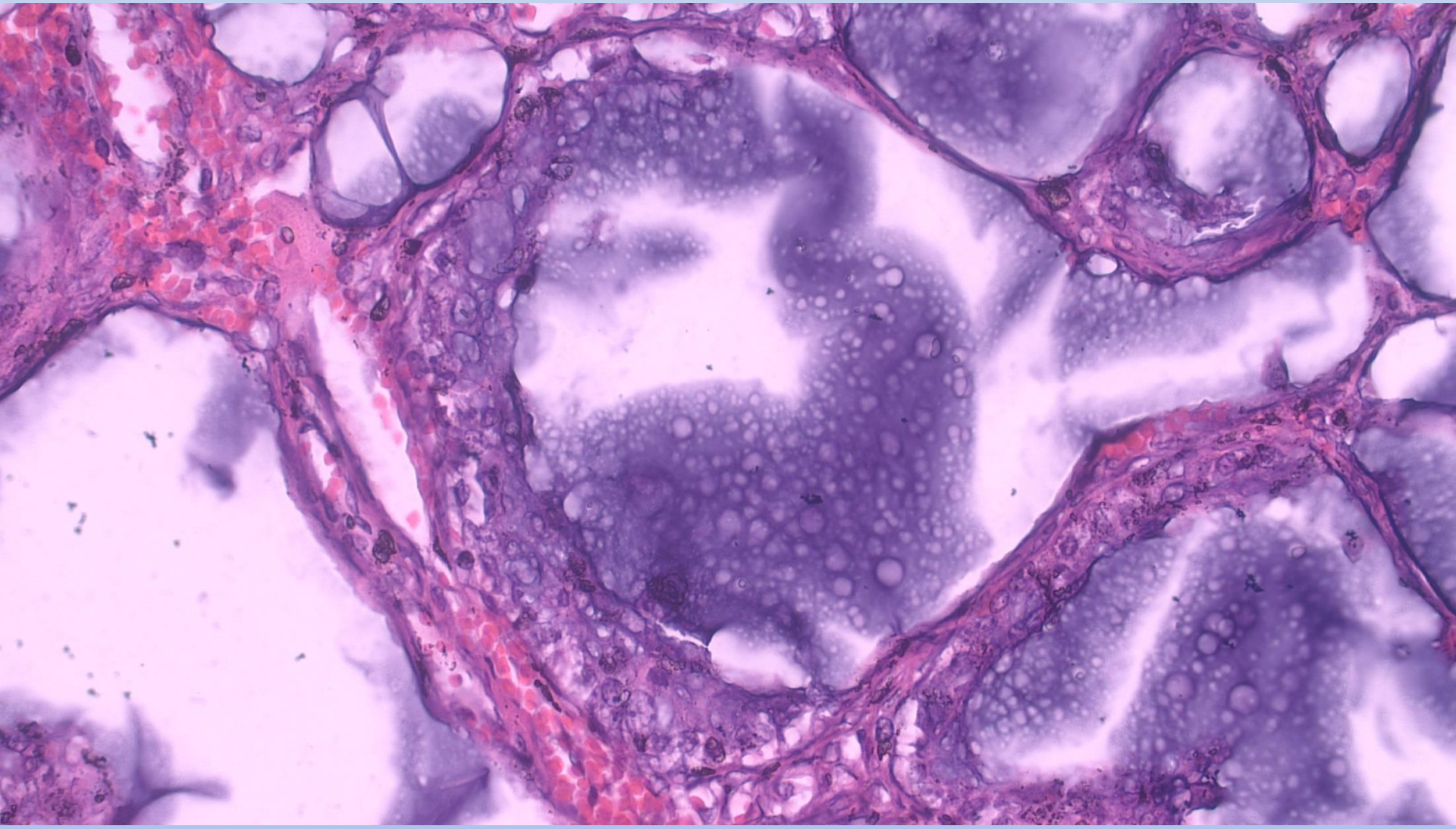


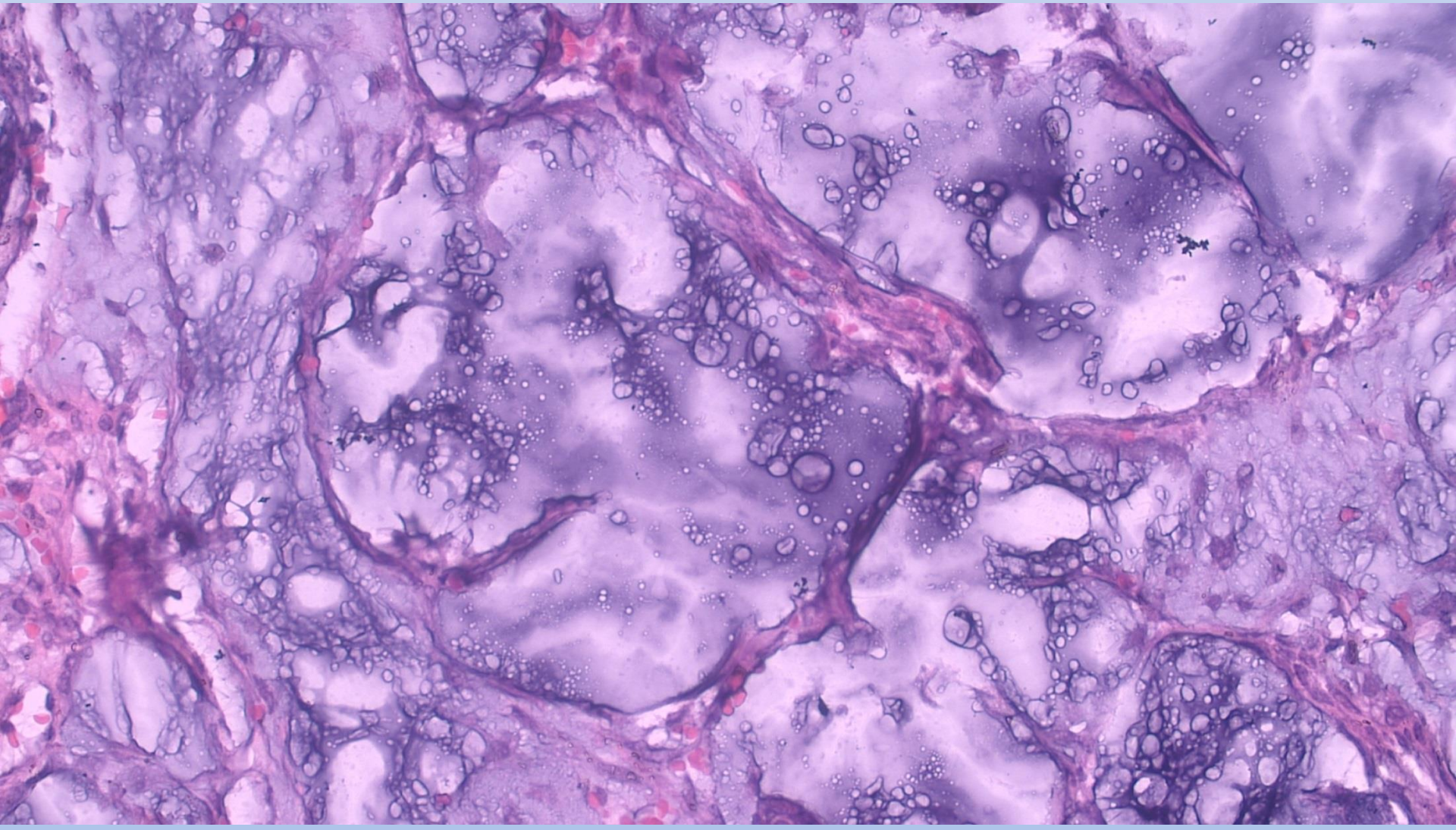




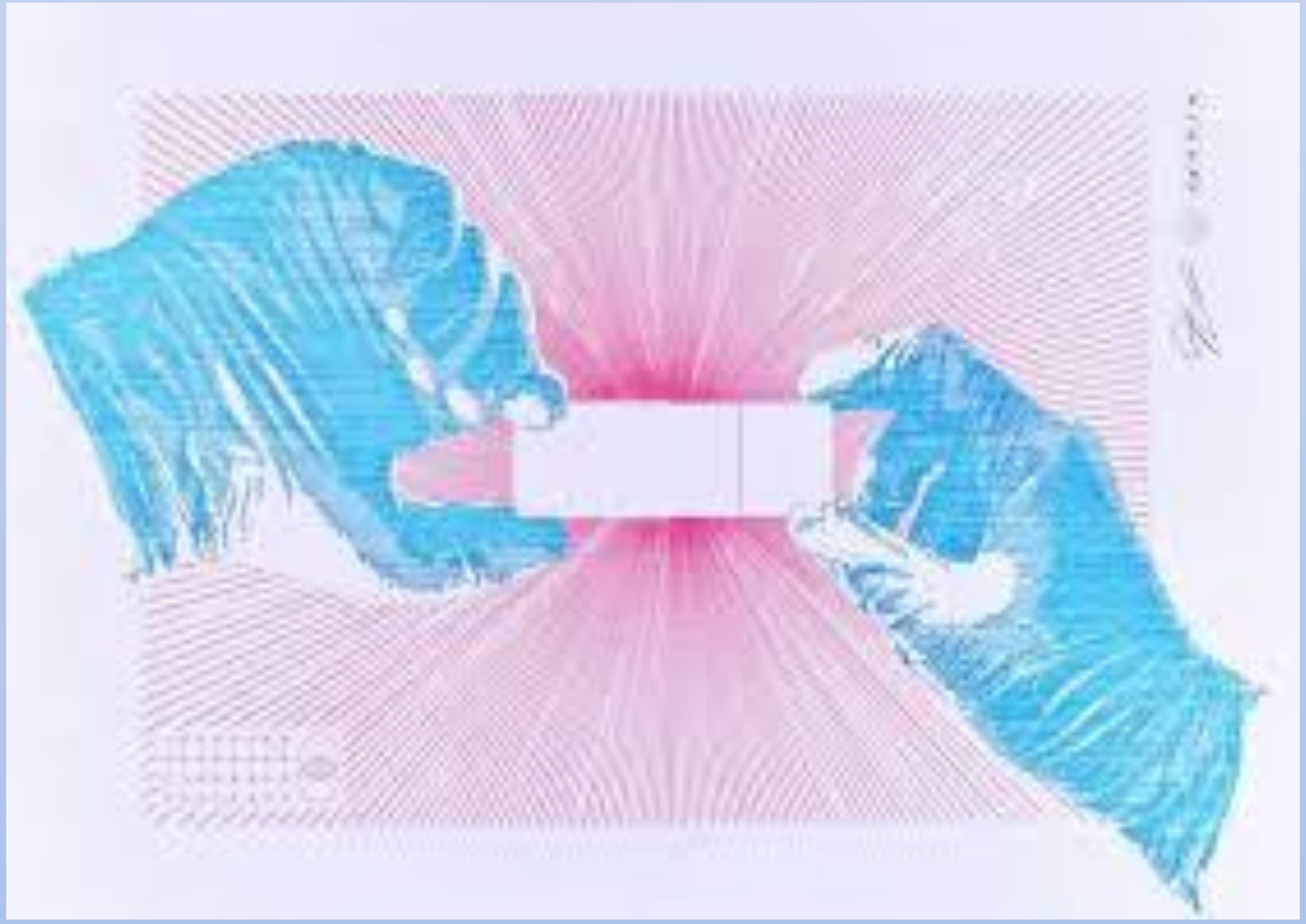








Diagnosis?



Mucinous Adenocarcinoma

- Appendical neoplasms :
    1)Non mucinous
    2)Mucinous
        a) Adenoma/Cystadenoma
        b)LAMN
        c)Mucinous Adenocarcinoma

LAMN

Similar to adenomas, these lesions may be grossly unremarkable or produce a cystically dilated appendix filled with mucin. Grossly visible mucin on the serosal surface or in the mesoappendix is an important finding and, if present, should be carefully documented and submitted entirely in order to establish the presence or absence of extra appendiceal neoplastic epithelium.

The low-grade epithelium of LAMNs is similar to cystadenomas and is characterized by tall mucinous epithelial cells in a villous or flat configuration. The villi are usually slender with straight edges. The epithelium is almost completely denuded in many cases, and multiple sections may be required to find neoplastic epithelium. LAMN have a distinct manner of invasion of the appendiceal wall, which features a broad pushing front (rather than infiltrative neoplastic glands with desmoplasia) with effacement of the muscularis mucosae and mural atrophy, fibrosis, and/or calcification.

If high- grade cytology is observed, the diagnosis is not LAMN, and the classification is based on the depth of invasion. If it is limited to the mucosa, the diagnosis is adenoma, otherwise the diagnosia is mucinous adenocarcinoma

Mucinous Adenocarcinoma

Peritoneal mucinous tumors with high-grade features, such as solid cell clusters that display nuclear enlargement and hyperchromasia, single cell necrosis, and mitotic activity, are more often associated with a mucinous adenocarcinoma.

The AJCC cancer staging manual classified the appendical mucinous tumors as a three- tier grading system: **G1**(well differentiated), **G2**(moderately differentiated), **G3**(poorly differentiated), based on cytologic features, tumor cellularity and signet ring components).

Appendiceal G1 (well-differentiated) tumors are low-grade cytology, usually lacking infiltrative invasion, and essentially refers to LAMN.

Given that mucinous adenocarcinoma is characterized by infiltrative invasion and almost always exhibits at least focal areas of high-grade cytologic features, typical mucinous adenocarcinomas should be classified as either G2 (moderately differentiated) or G3 (poorly differentiated) tumors.

Mucinous adenocarcinomas often show complex architecture, such as cribriform and complex papillary structures.

The presence of signetring cells is an indication of infiltrative mucinous adenocarcinoma.

Poorly differentiated (G3) mucinous adenocarcinoma demonstrates infiltrative invasion, with most having signet-ring cells. Signet-ring cells, characterized by prominent intracytoplasmic mucin displacing the nucleus, may infiltrate single cells or as aggregates, classified as grade G3 (poorly differentiated). Most G3 tumors are almost entirely composed of signet-ring cells, while a minority of cases are composed of mixed signet-ring cells and glandular structures.

If cancer comprises ≤ 50% signet-ring cells, the WHO terminology is mucinous adenocarcinoma with signet-ring cells or mucinous adenocarcinoma, poorly differentiated.

If the tumor contains > 50% signet-ring cells, the WHO terminology is signet-ring cell carcinoma.

Most mucinous adenocarcinomas have clinically aggressive behavior, infiltrating through the appendiceal wall (pT4), and frequently metastasize to the abdominal or pelvic peritoneum at diagnosis. After an appendectomy specimen, diagnosis of mucinous adenocarcinoma should result in subsequent right hemicolectomy to evaluate lymph node metastases.

Grading appendiceal mucinous neoplasms

Tumor grade	In the appendiceal primary tumor	In the peritoneal metastasis
1	Low grade cytology with a pushing margin (low grade appendiceal mucinous neoplasm)	Hypocellular mucinous deposits Neoplastic epithelial elements have low grade cytology No infiltrative type invasion
2	High grade cytology with a pushing margin (high grade appendiceal mucinous neoplasm) Invasive mucinous adenocarcinoma without a signet ring cell component	Hypercellular mucin deposits as judged at 20x magnification High grade cytological features Infiltrative type invasion characterized by jagged or angulated glands in a desmoplastic stroma or a small mucin pool pattern with numerous mucin pools containing clusters of tumor cells
3	Signet ring cell adenocarcinoma with numerous signet ring cells in mucin pools or infiltrating tissue	Mucinous tumor deposits with signet ring cells

IHC staining

Appendiceal mucinous neoplasms and pseudomyxoma peritonei typically express cytokeratin 20, CDX-2, and MUC2; cytokeratin 7 is variably positive.

Molecular

A limited number of studies have reported that the vast majority of *KRAS* mutations occurred in G1 and G2 tumors, and most *KRAS* mutations occur in 50% to 60% of Tis(LAMN).

These data suggest that *KRAS* mutations are important in tumor initiation but may be less critical for aggressive highgrade tumor progression. *GNAS* mutations, known as essential in abundant mucin production, are also presented in G1 and G2 tumors but less commonly in G3 tumors. The co-mutation of *GNAS* and *KRAS* is identified in between 65% to 85% of cases .


Thank
You!