

Topics for today

- Von Meyenburg Complex/ Biliary Hamartoma
- Bile Duct Adenoma
- Cholangiocarcinoma
 - Large duct type (type 1)
 - Small duct type (type 2)
- Biliary Intraepithelial Neoplasia (BillIN)
- Differential Diagnosis
 - Metastatic Adenocarcinoma



Biliary Hamartoma

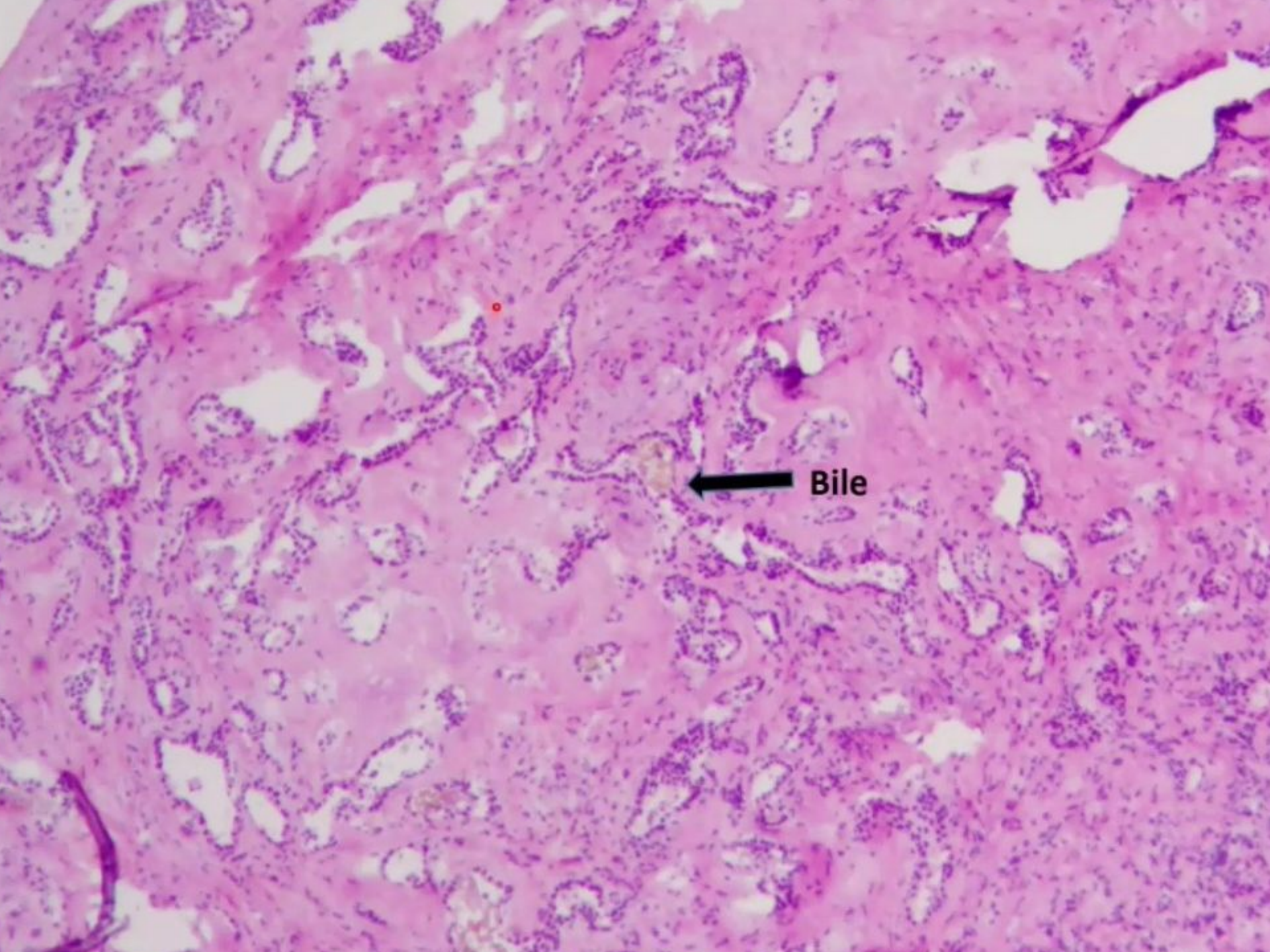
- Benign lesion, developmental
- Remnant of ductal plate, due to incomplete modeling
 - Focal, in one or few portal tracts
 - Numerous in fibro-polycystic diseases (autosomal dominant/ recessive polycystic disease, congenital hepatic fibrosis)
- Incidental finding in biopsies or resections
- Frequently biopsied during abdominal surgery for cancer for frozen section/ Intraoperative diagnosis

Case 1

75 year old female undergoing surgery for pancreatic cancer

Surgeon noticed small white lesions on the liver capsule

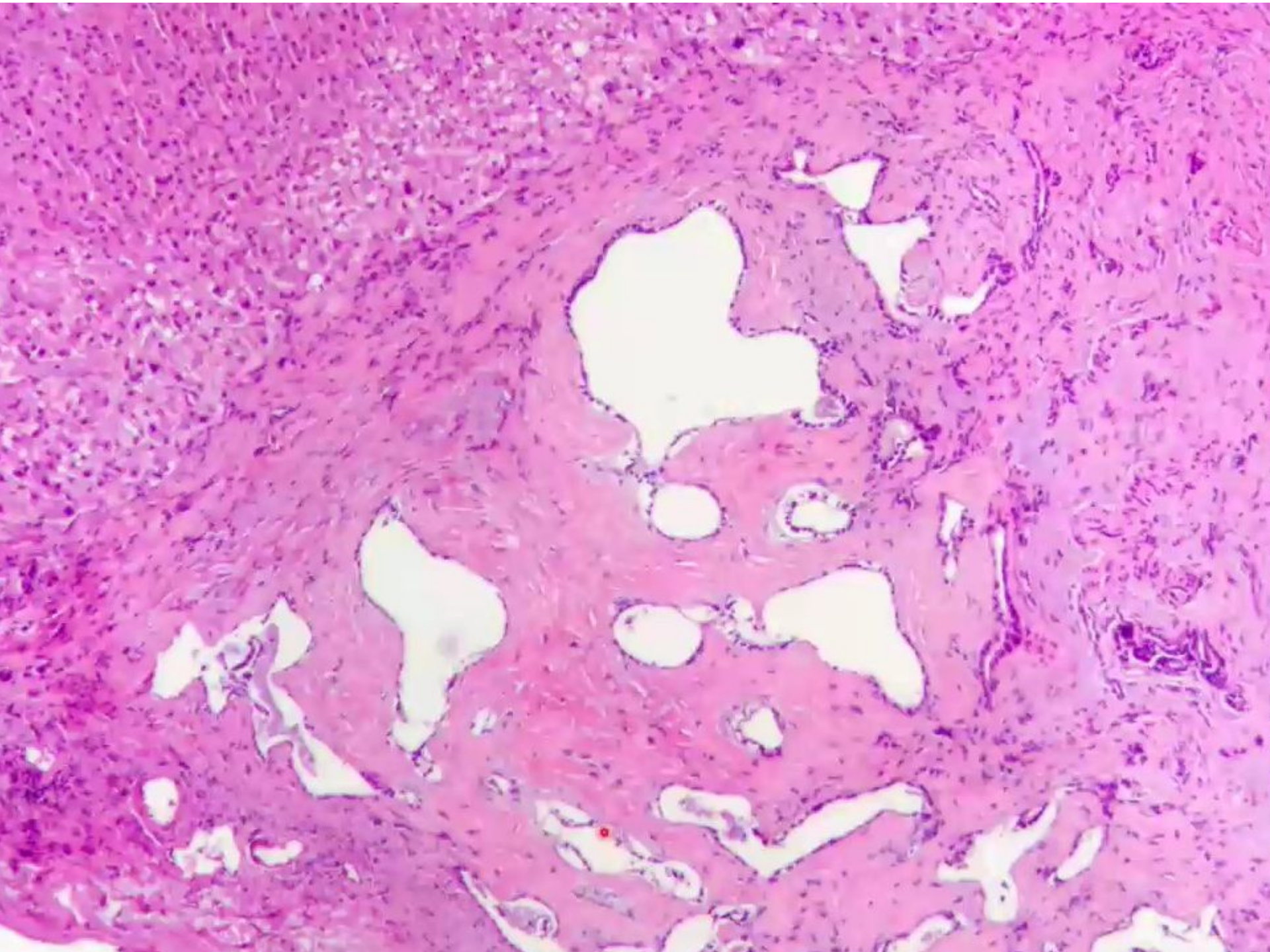
He sent one for frozen section/ intraoperative diagnosis

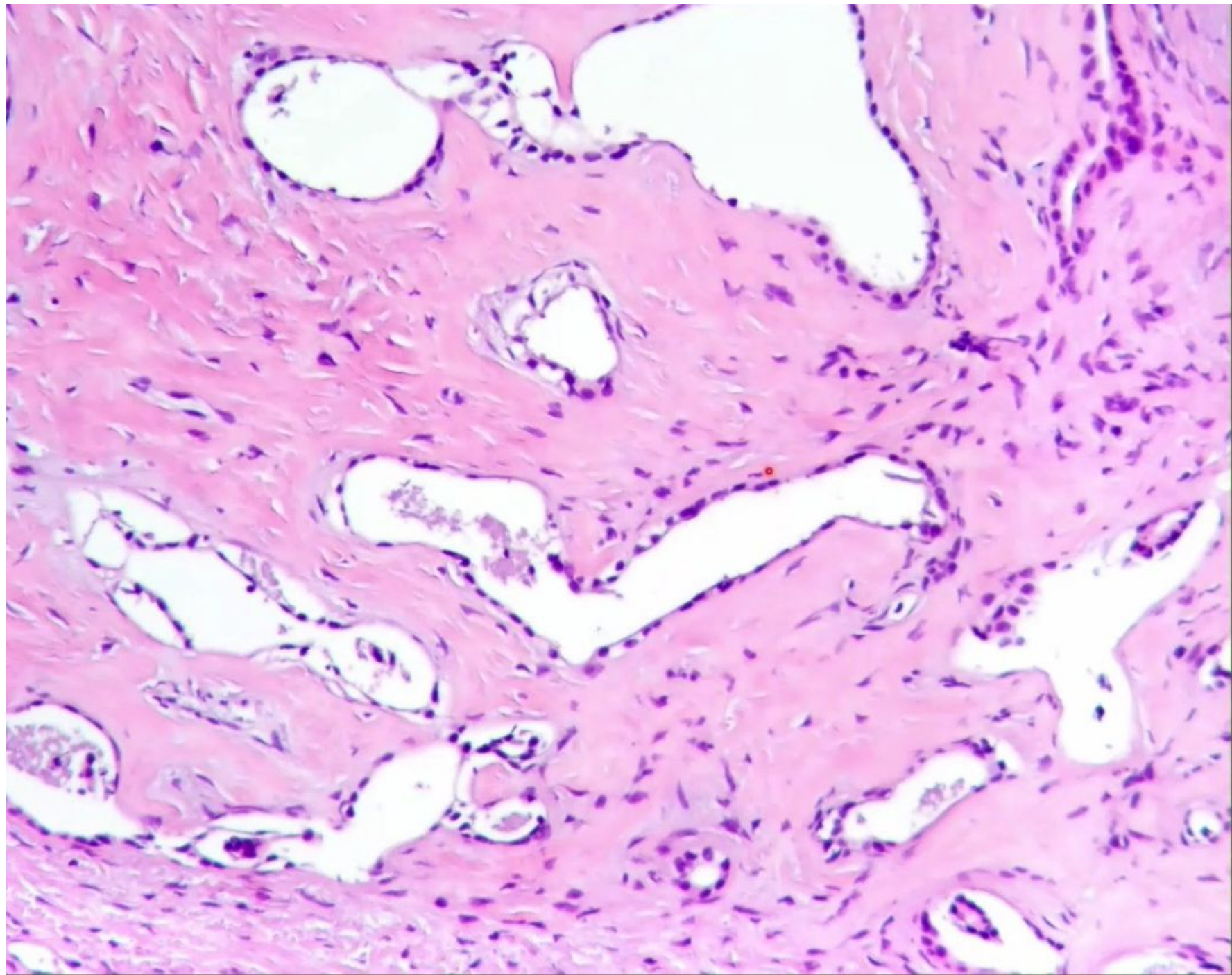


Bile

Von Meyenburg Complex – Frozen section

- **Benign ductal structures with irregular shapes**
- **Single or double layer of flat or low cuboidal epithelium**
- **Open, cystically dilated lumina**
- **Often contain bile**

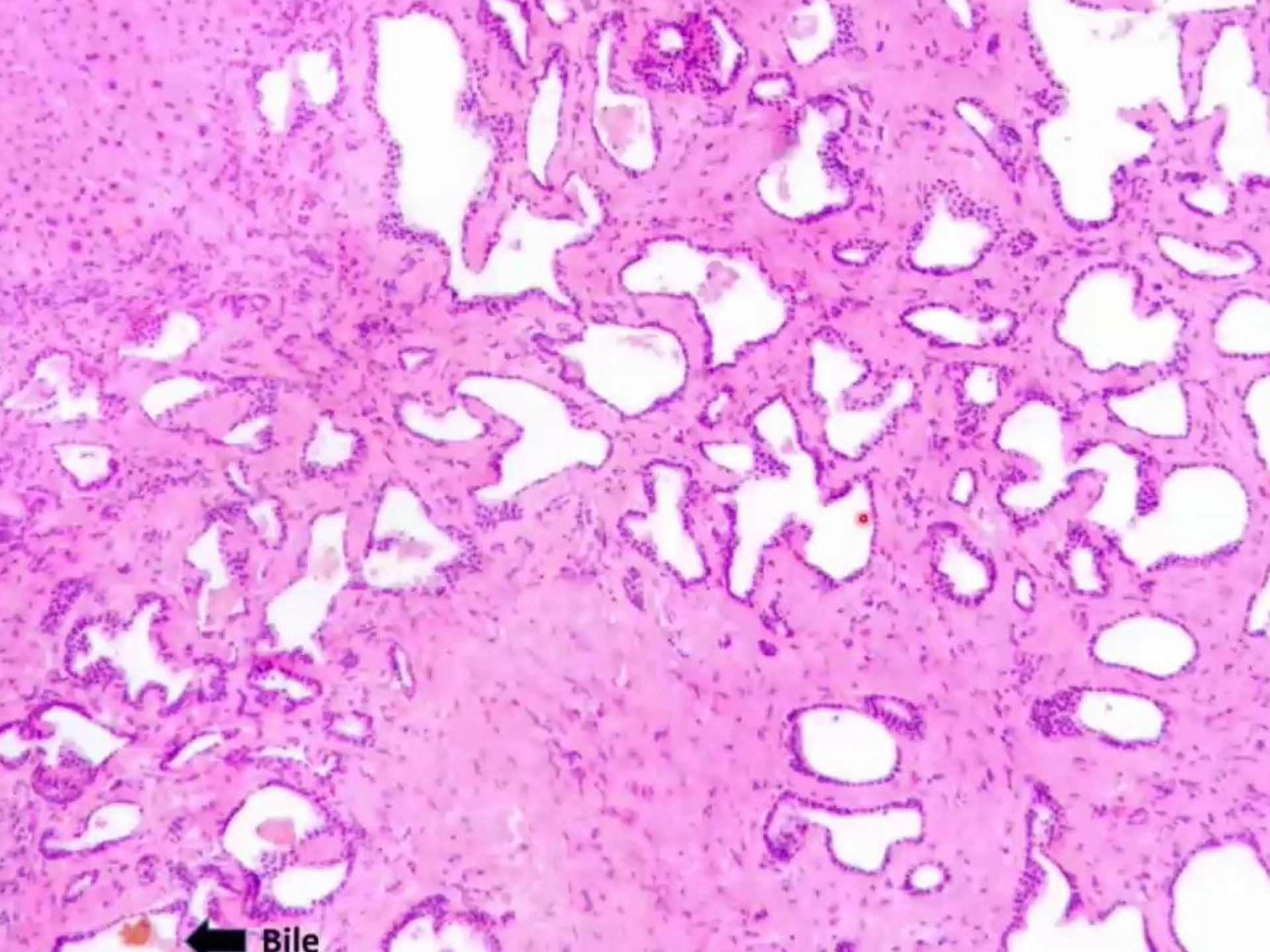




**Von Meyenburg Complex –
Permanent (formalin fixed,
paraffin embedded)
section**

**Von Meyenburg Complex –
Permanent (formalin fixed,
paraffin embedded) section**

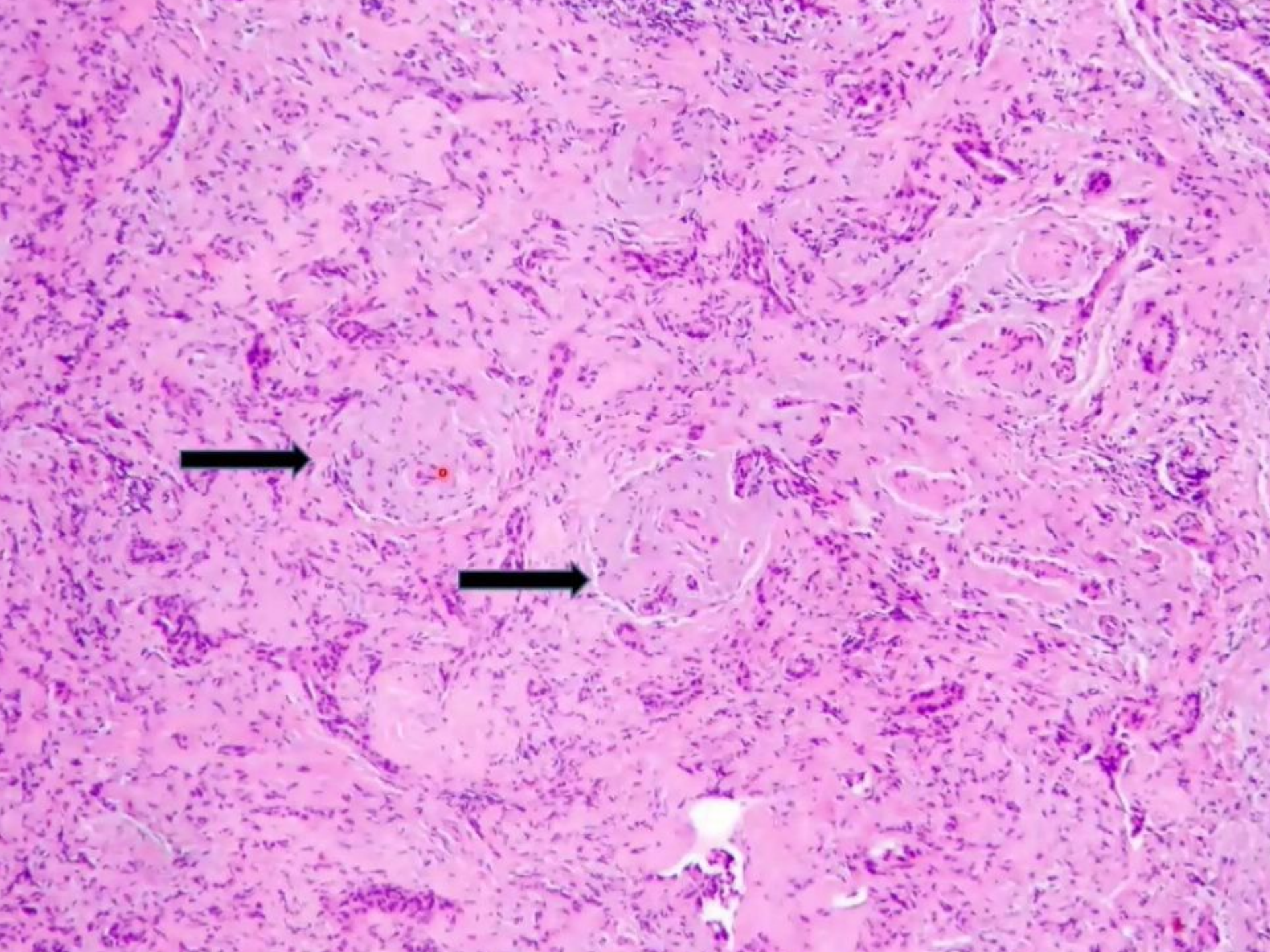
- Irregular ductal structures with cystically dilated lumina
- Single layer of flat epithelial cells

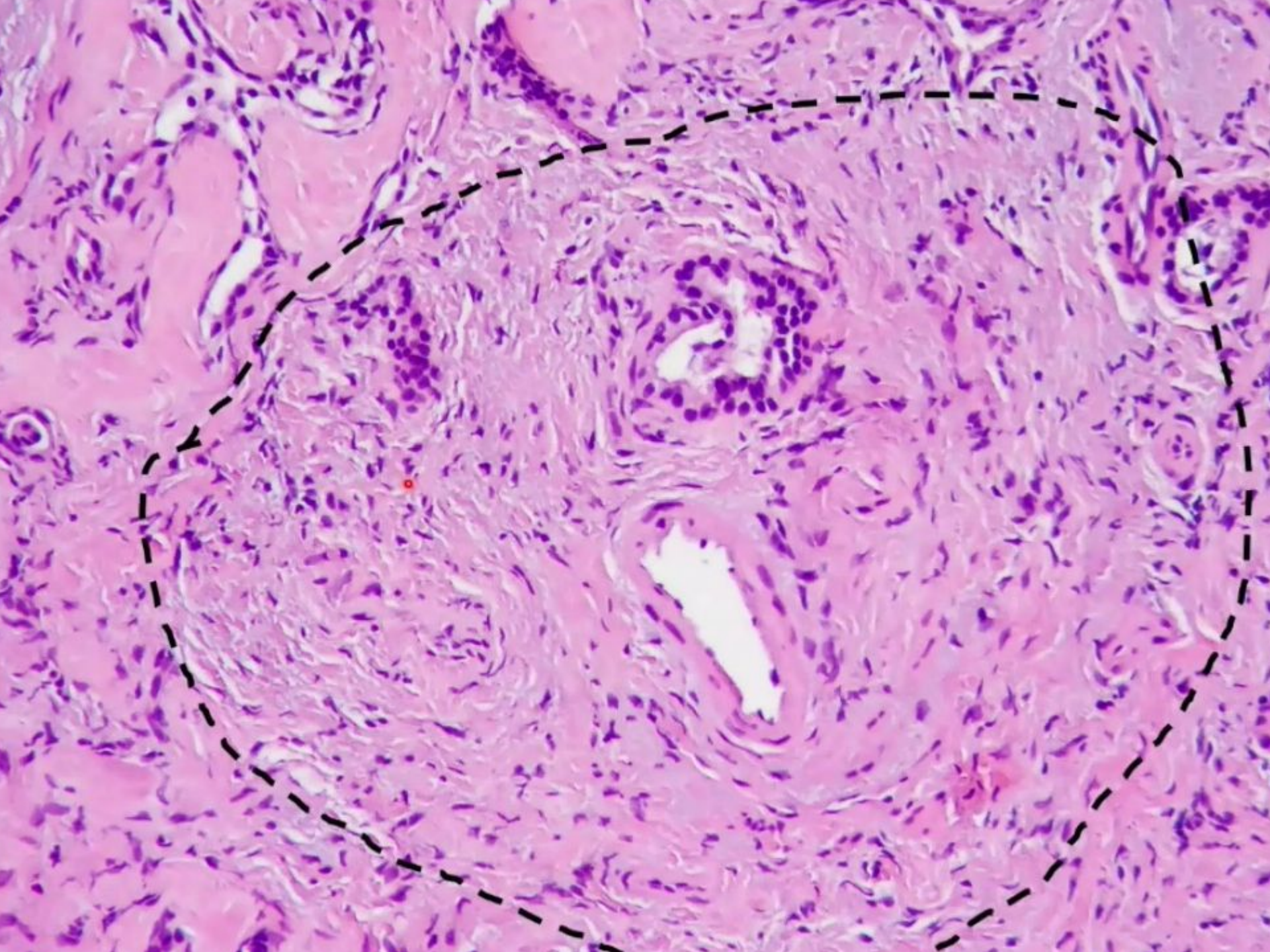


← Bile

Case 2

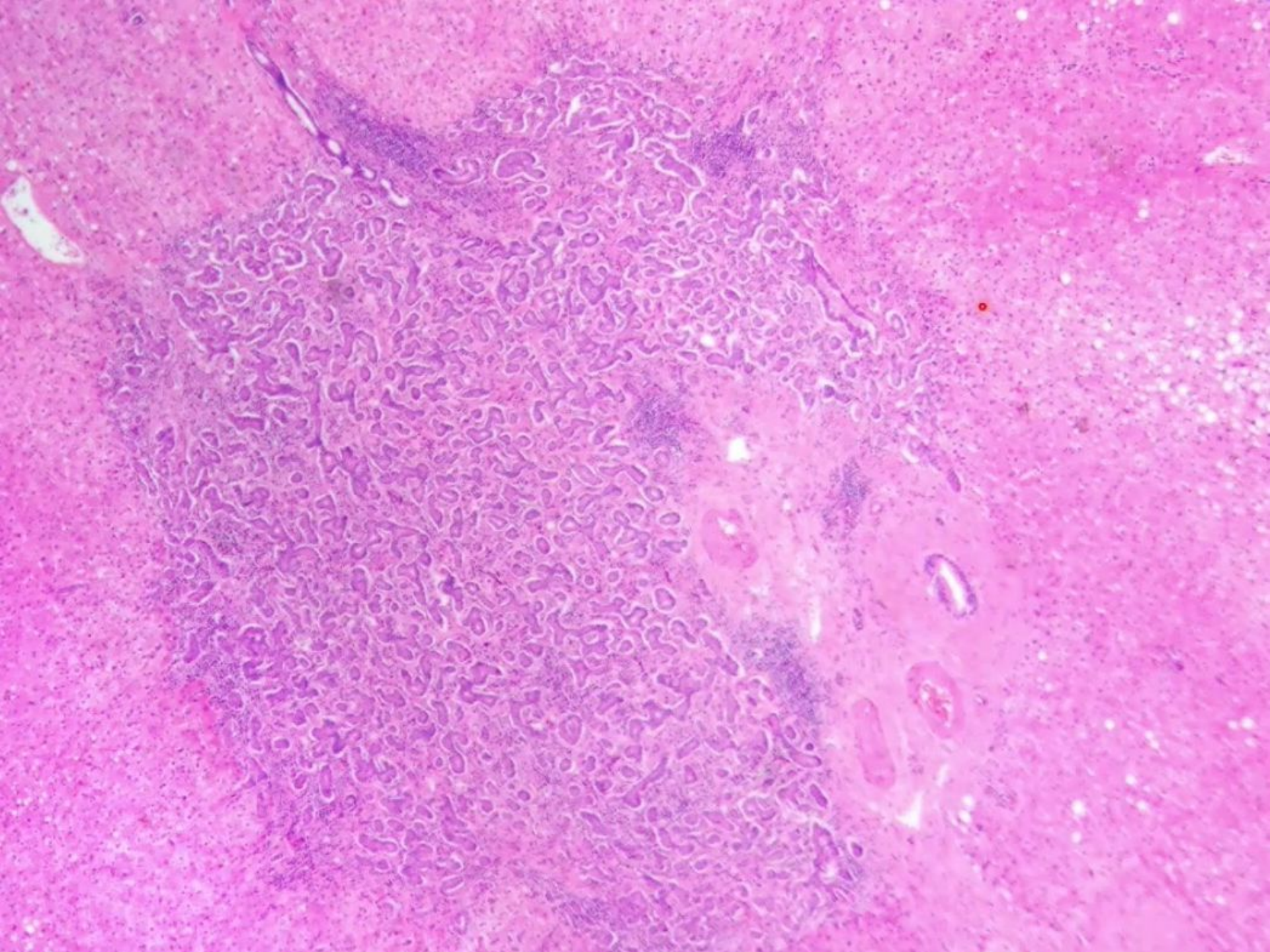
- 70 year old patient undergoing surgery for gastric cancer
- Surgeon noticed small whitish lesions on surface of liver
- He sent one for frozen section diagnosis

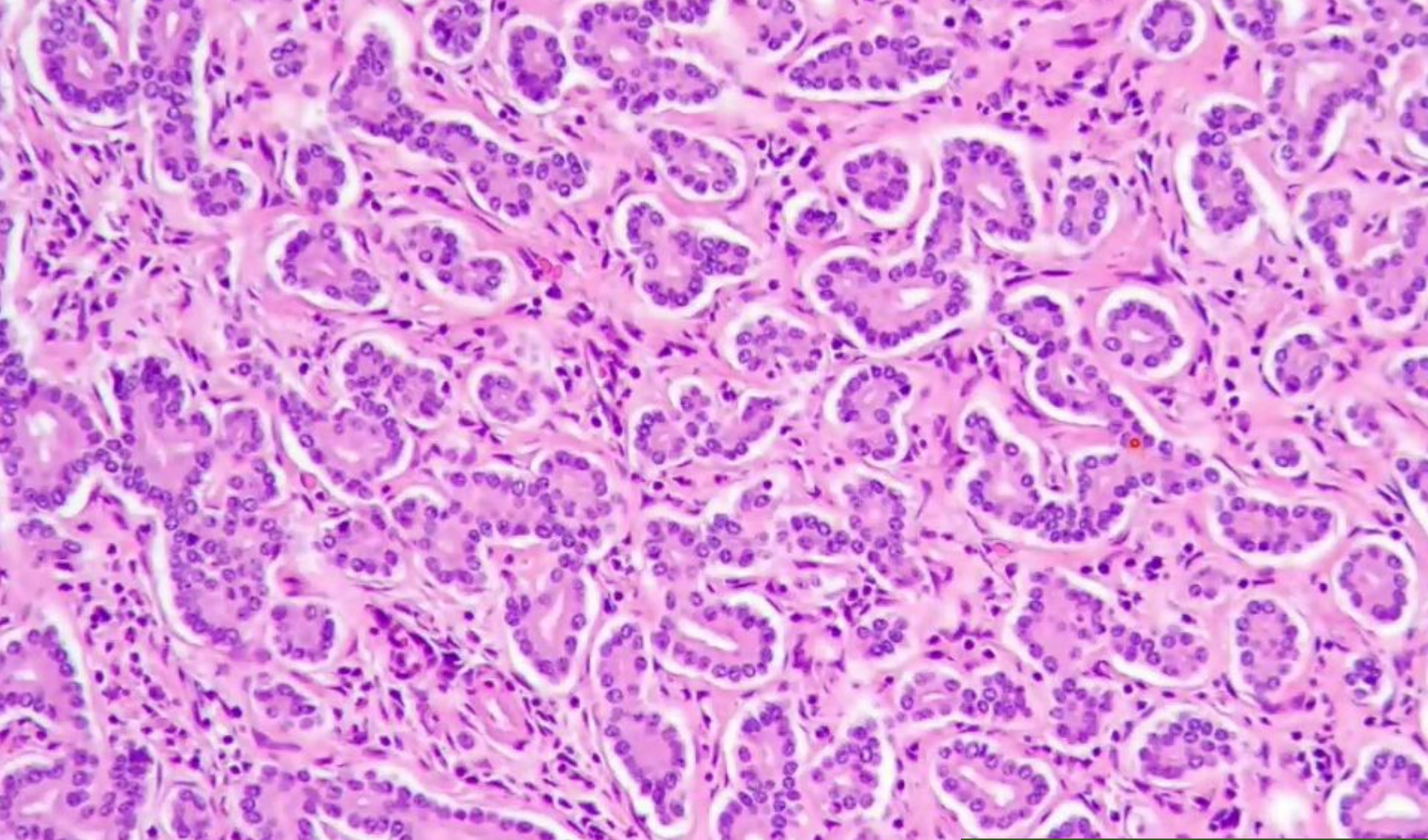




Bile Duct Adenoma
Permanent (formalin
fixed, paraffin embedded)
section

Small ducts with open lumina in
fibrous stroma
Entrapped portal tract





**Incidental Bile Duct
Adenoma in an explant
during transplantation**

Bile Duct Adenoma

Biliary Hamartoma

Benign tumor

Developmental lesion, not a tumor

Proliferation of bile ducts

Remnants of embryonic ductal plate

Small ducts in dense fibrous stroma

Irregular shaped, cystically dilated ductal structures

Entrapped portal tracts common

Occurs within portal tract

May contain bile

May contain bile

Distinguishing between these 2 lesions is not clinically important; both are benign and no further action is required. Do not cause symptoms

Cholangiocarcinoma

A tumor arising from bile ducts and derived from cholangiocytes

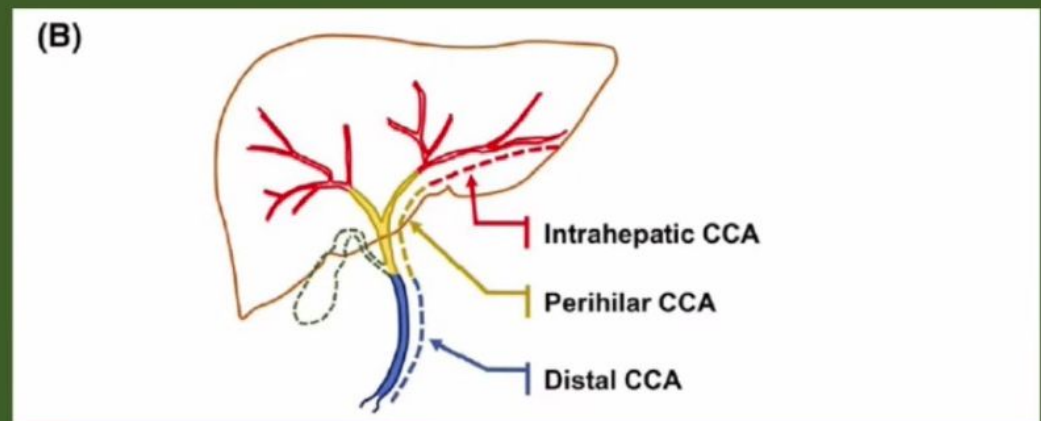
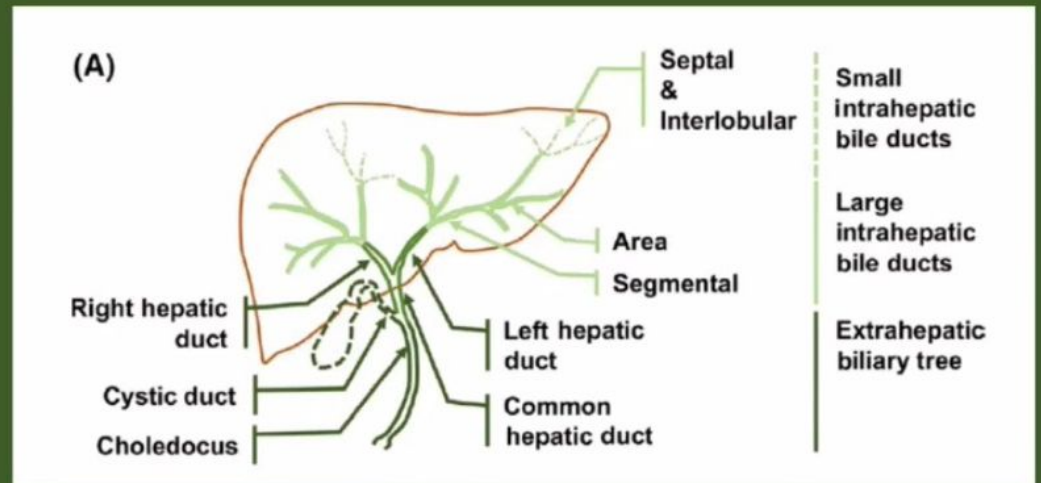


Classification

- Anatomic Location
- Macroscopic Growth Pattern
- Microscopic

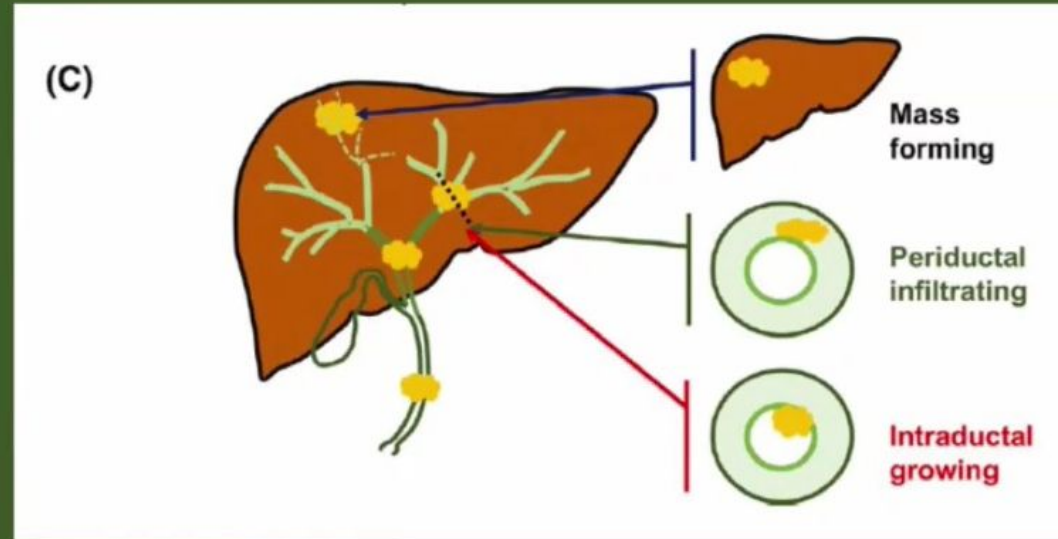
Anatomic Classification

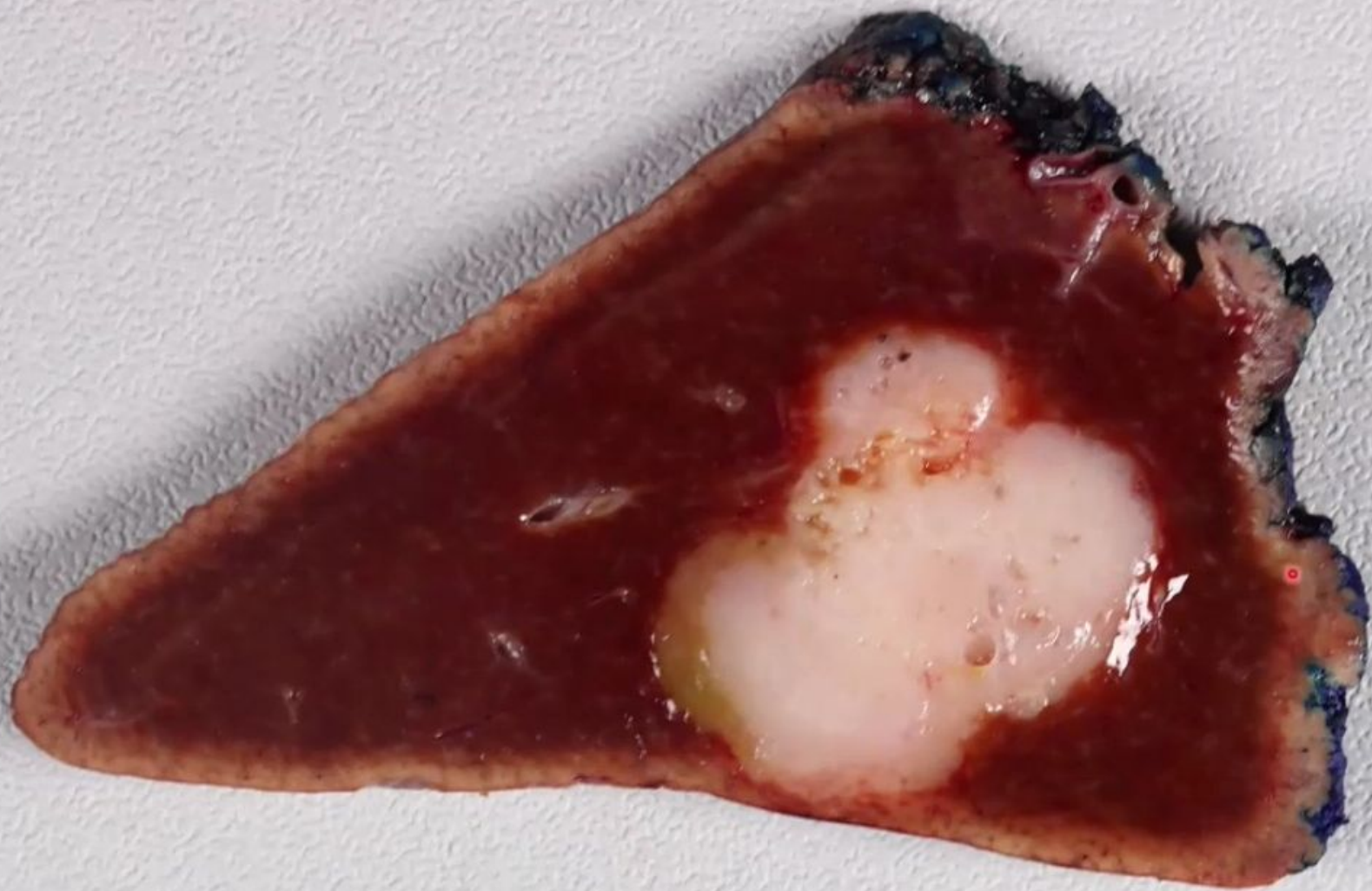
- Intrahepatic
- Perihilar
- Distal



Macroscopic Growth Pattern

- Mass Forming
- Periductal
- Intracystic
- Mixed

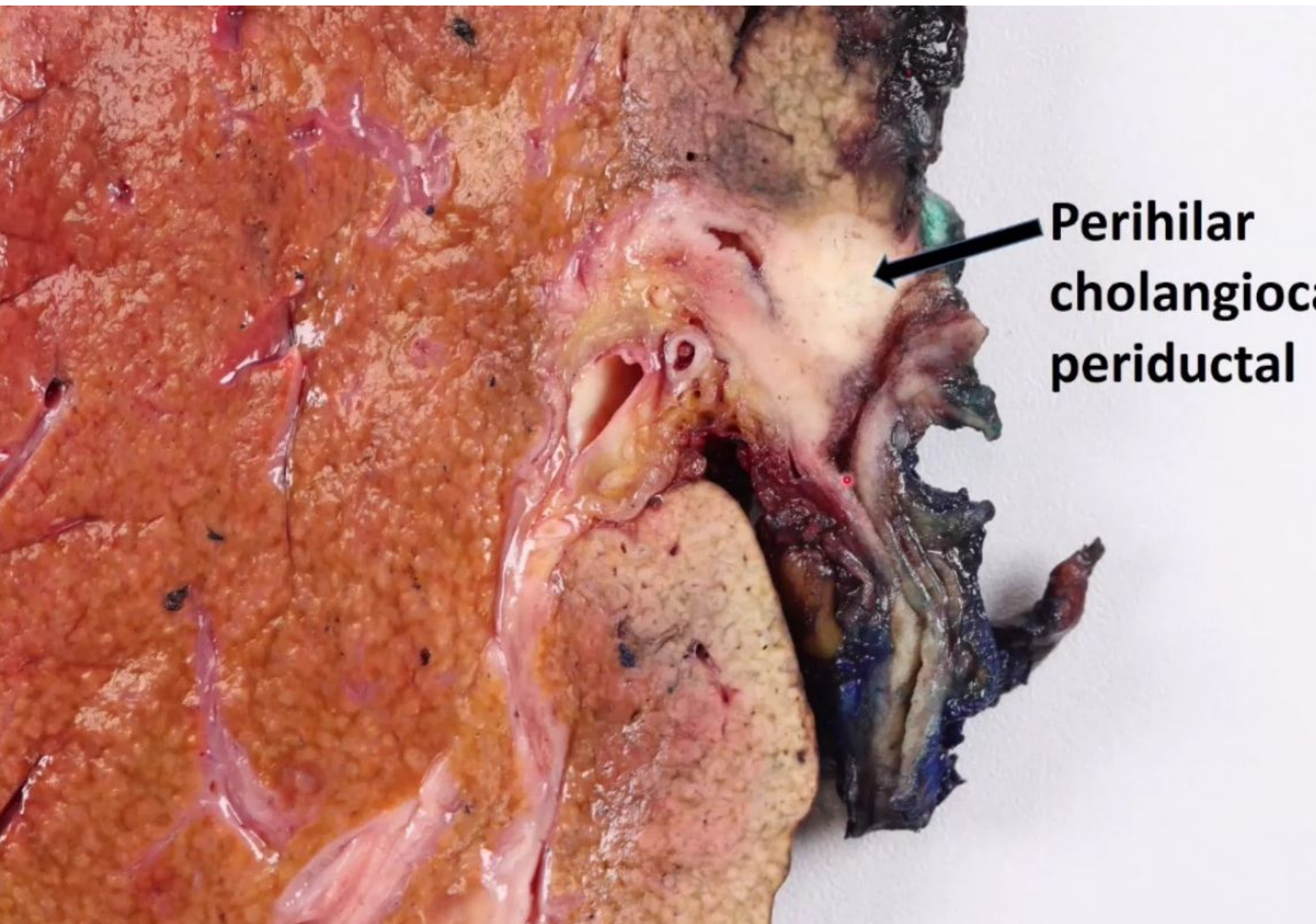




Intrahepatic cholangiocarcinoma, Mass forming

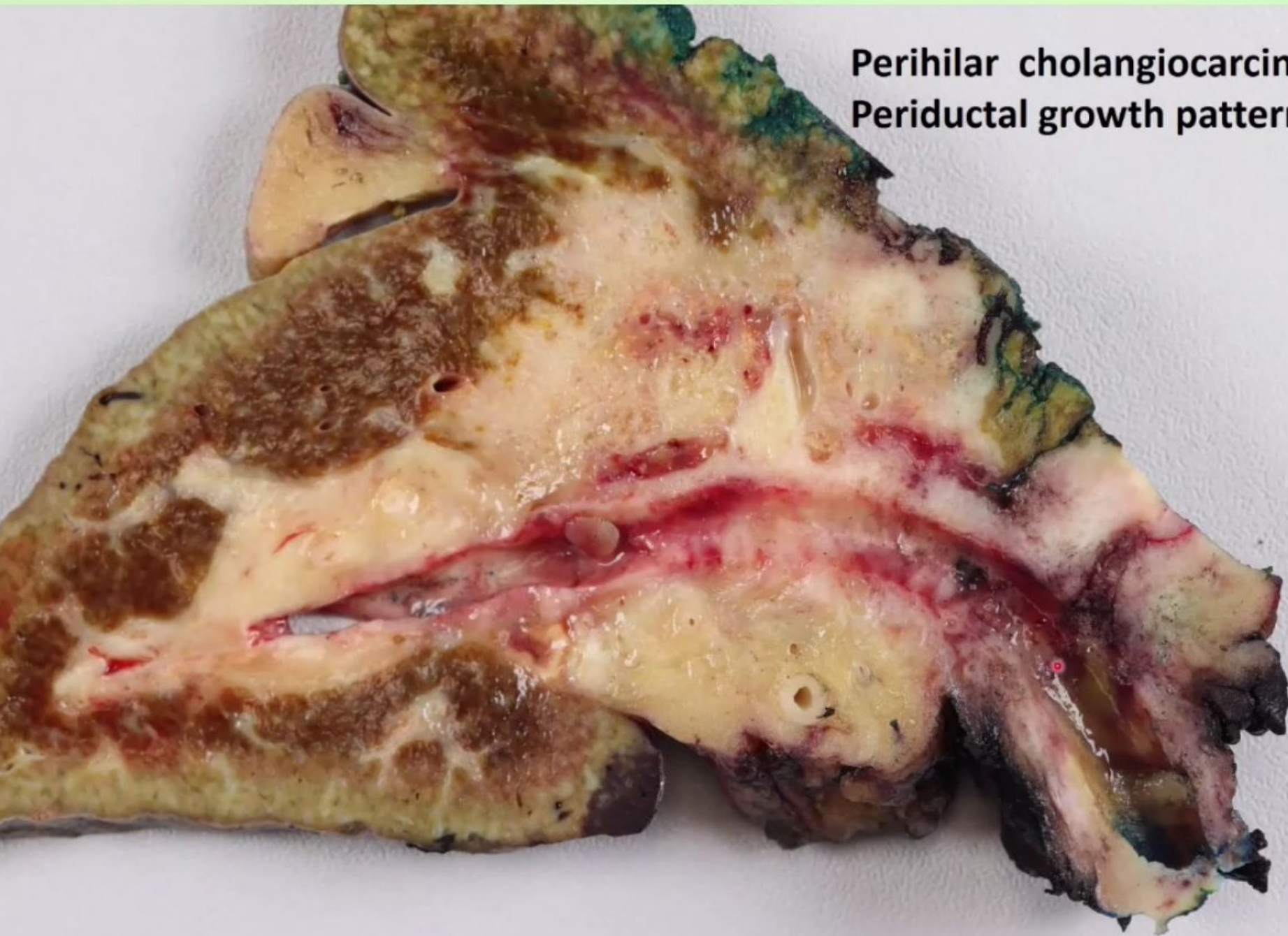


Perihilar,



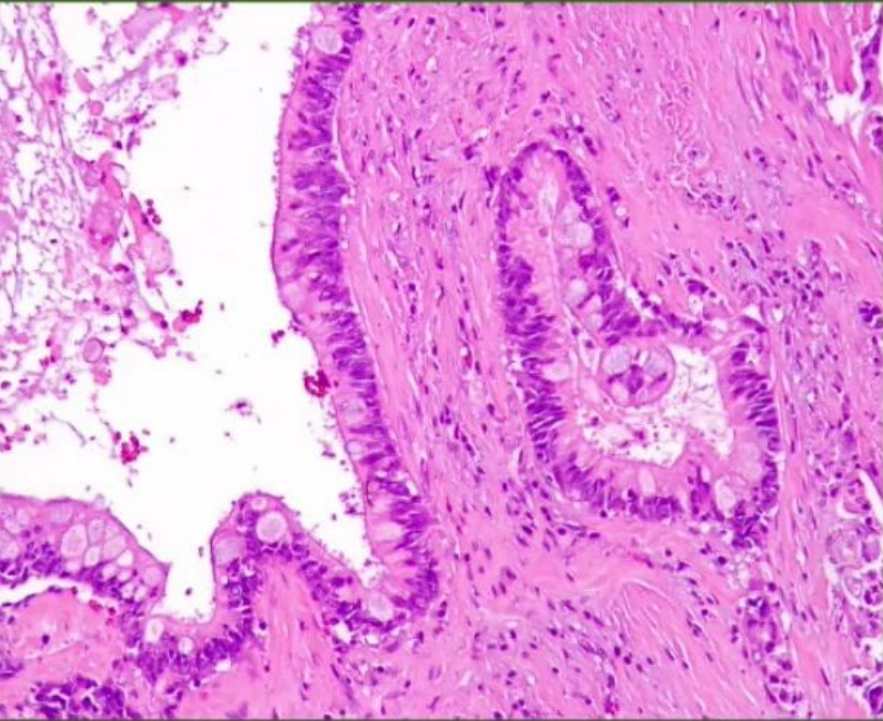
**Perihilar
cholangiocarcinoma
periductal**

Perihilar cholangiocarcinoma
Periductal growth pattern

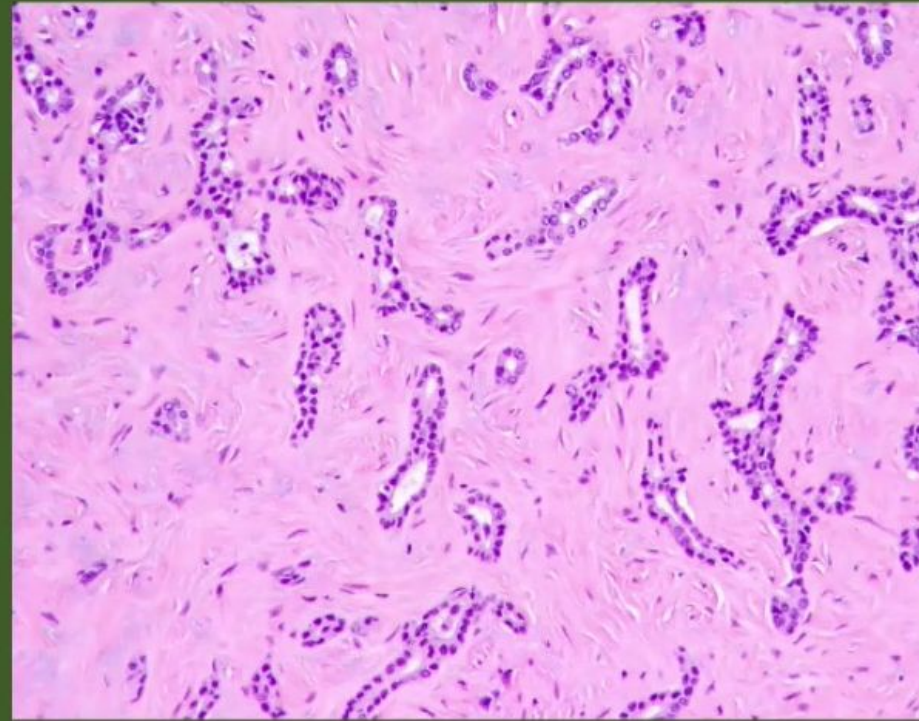


Microscopic Classification

Large Duct (type 1)



Small Duct (type 2)



Large Duct Type (type 1)

Adenocarcinoma

- Pancreatobiliary
- Intestinal
- Gastric Foveolar

Biliary Intraepithelial Neoplasia (BillN) may be seen

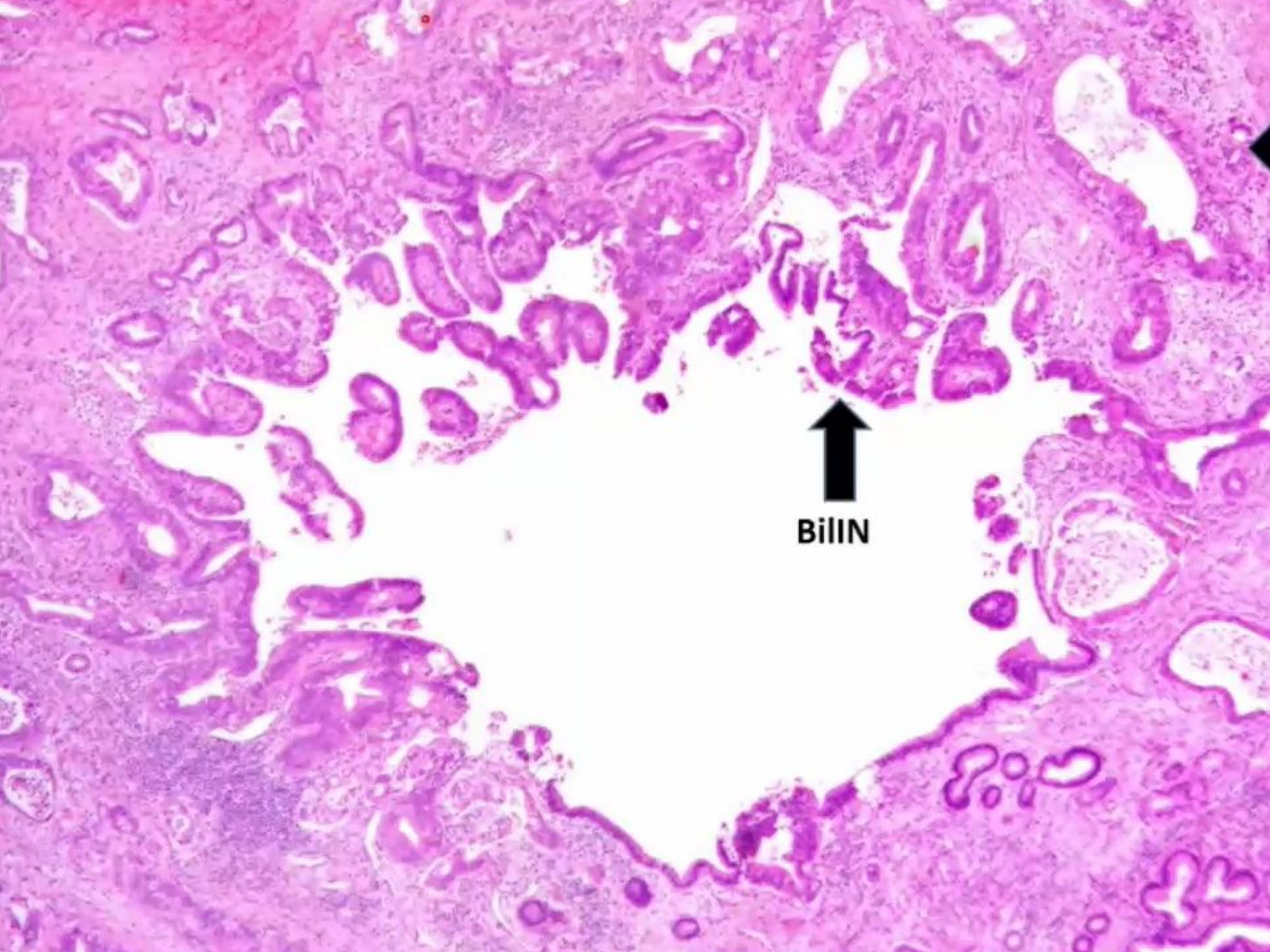
Perineural, lymphovascular, lymph node metastasis common

Biliary Intraepithelial Neoplasia (BiIN)

- Flat, premalignant intraepithelial neoplasia of bile ducts
- Field change, multiple foci
- Precursor lesion of cholangiocarcinoma
- Risk factors: hepatolithiasis, PSC (primary sclerosing cholangitis), chronic liver fluke infection

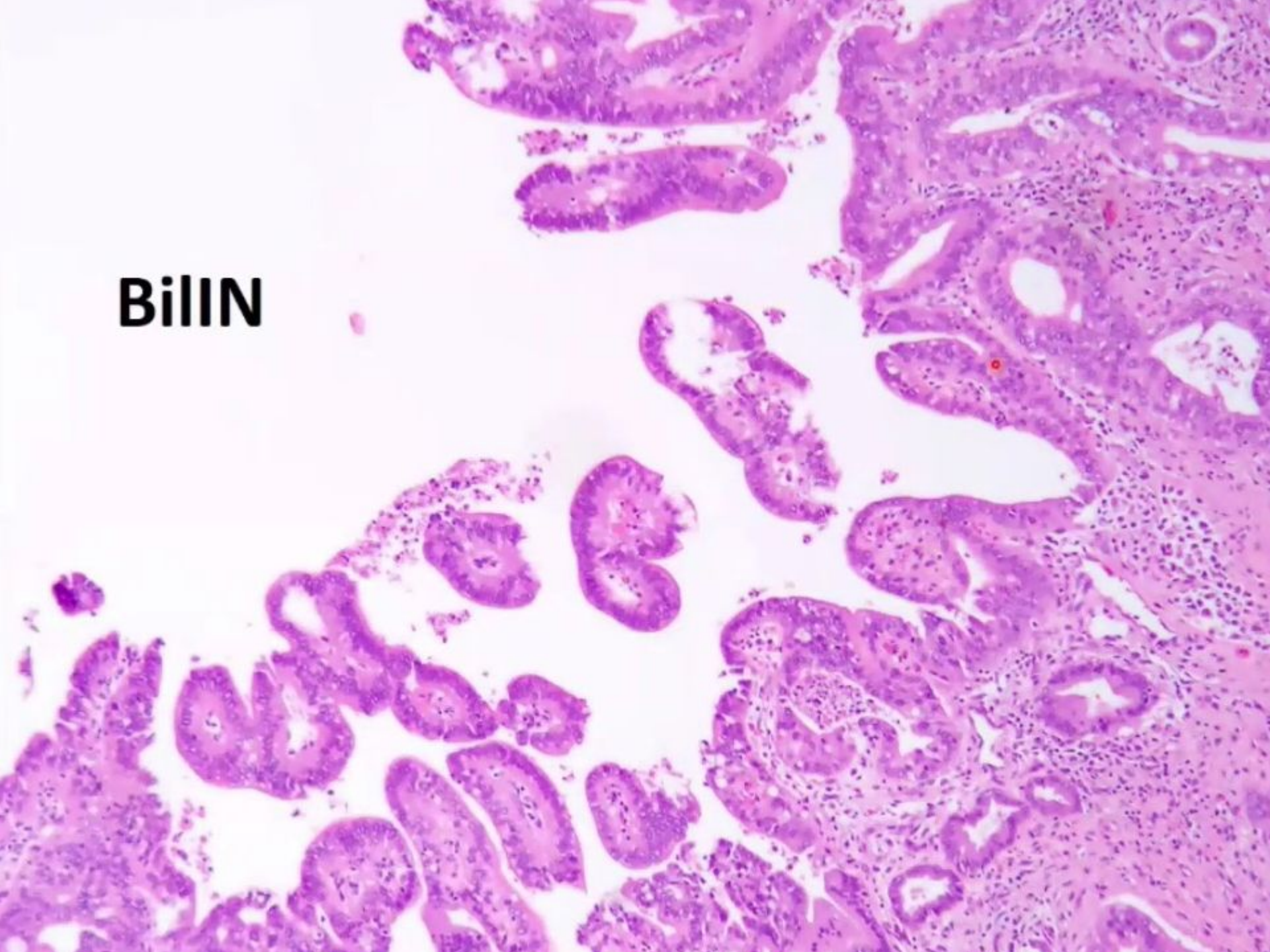
Microscopic Features (BillN)

- Nuclear multilayering, pleomorphism, hyperchromasia, loss of polarity
- Flat or micropapillary projections into duct lumen
- Graded as 3 tier (low, intermediate or high grade, BillN 1,2,3) or 2 tier (low or high grade)

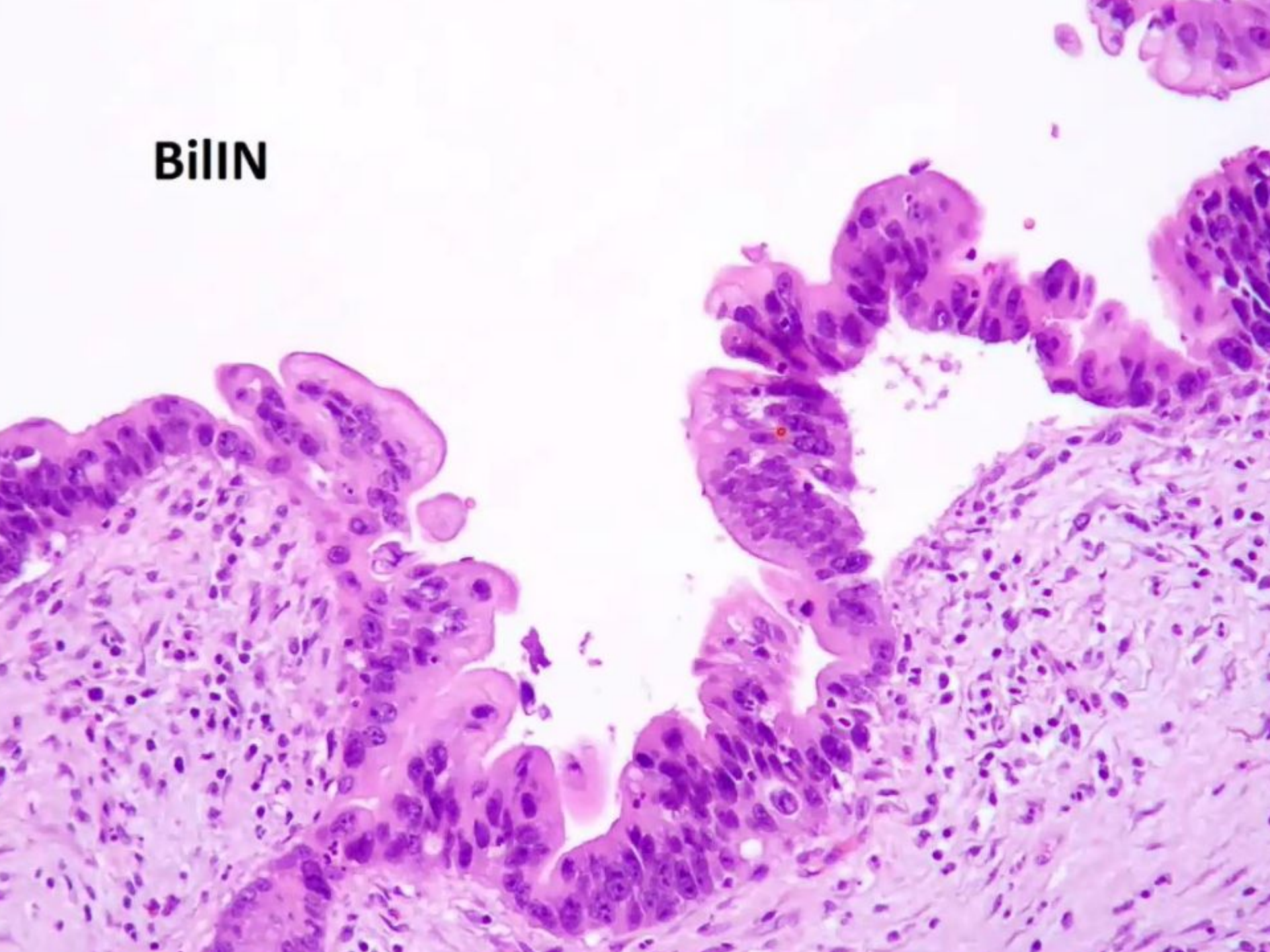


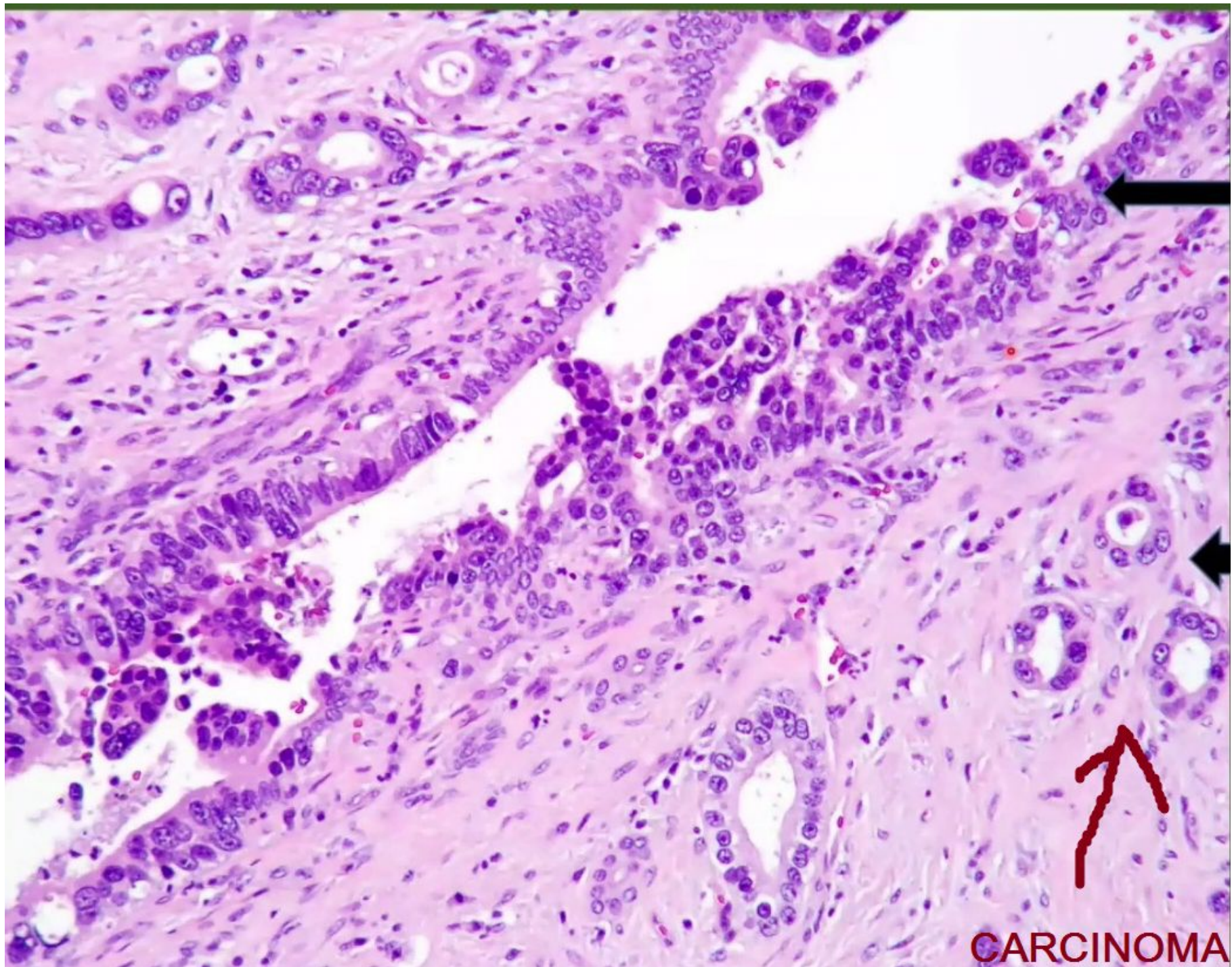
Bilin

BilIN

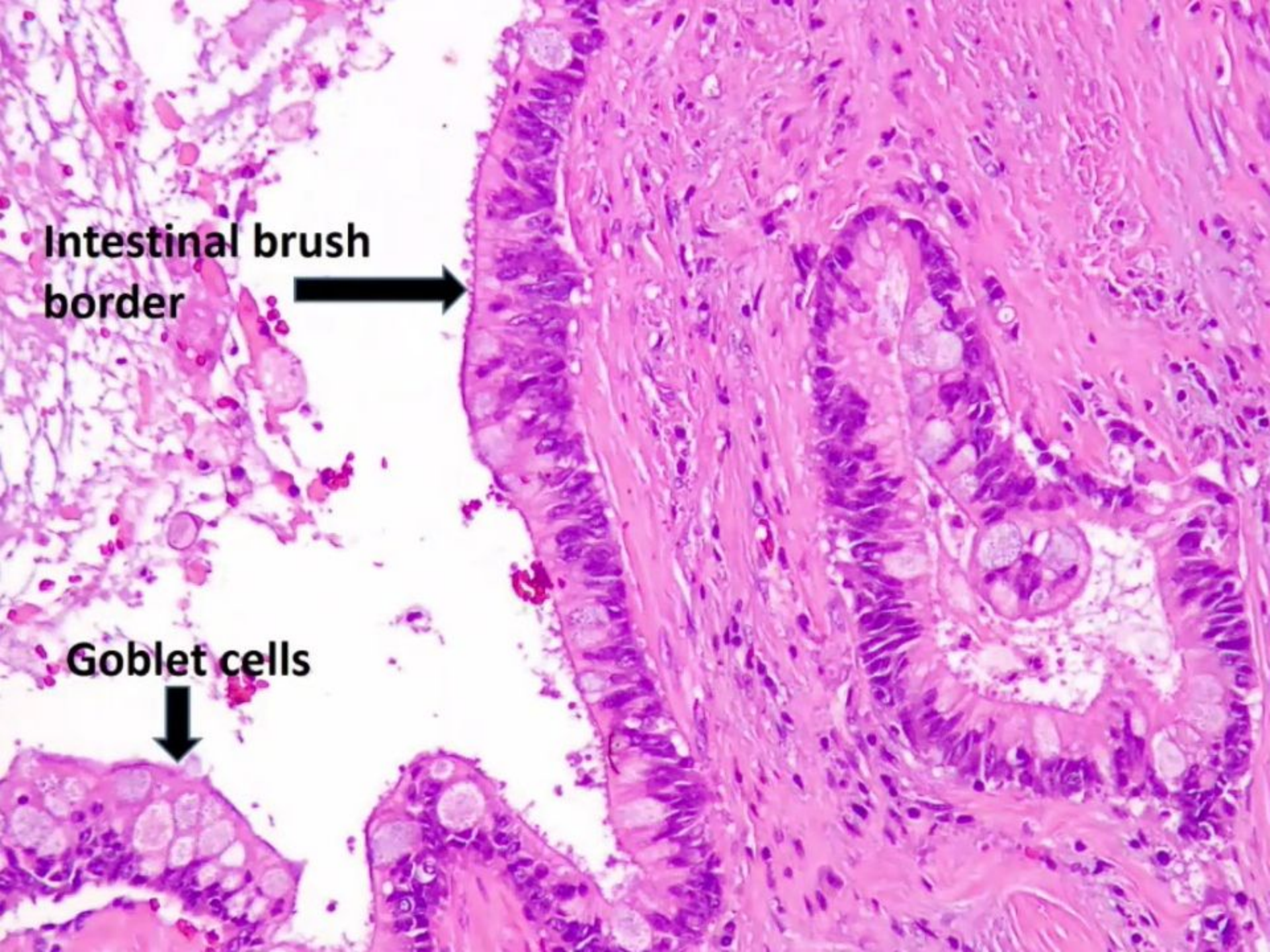


BilIN





CARCINOMA

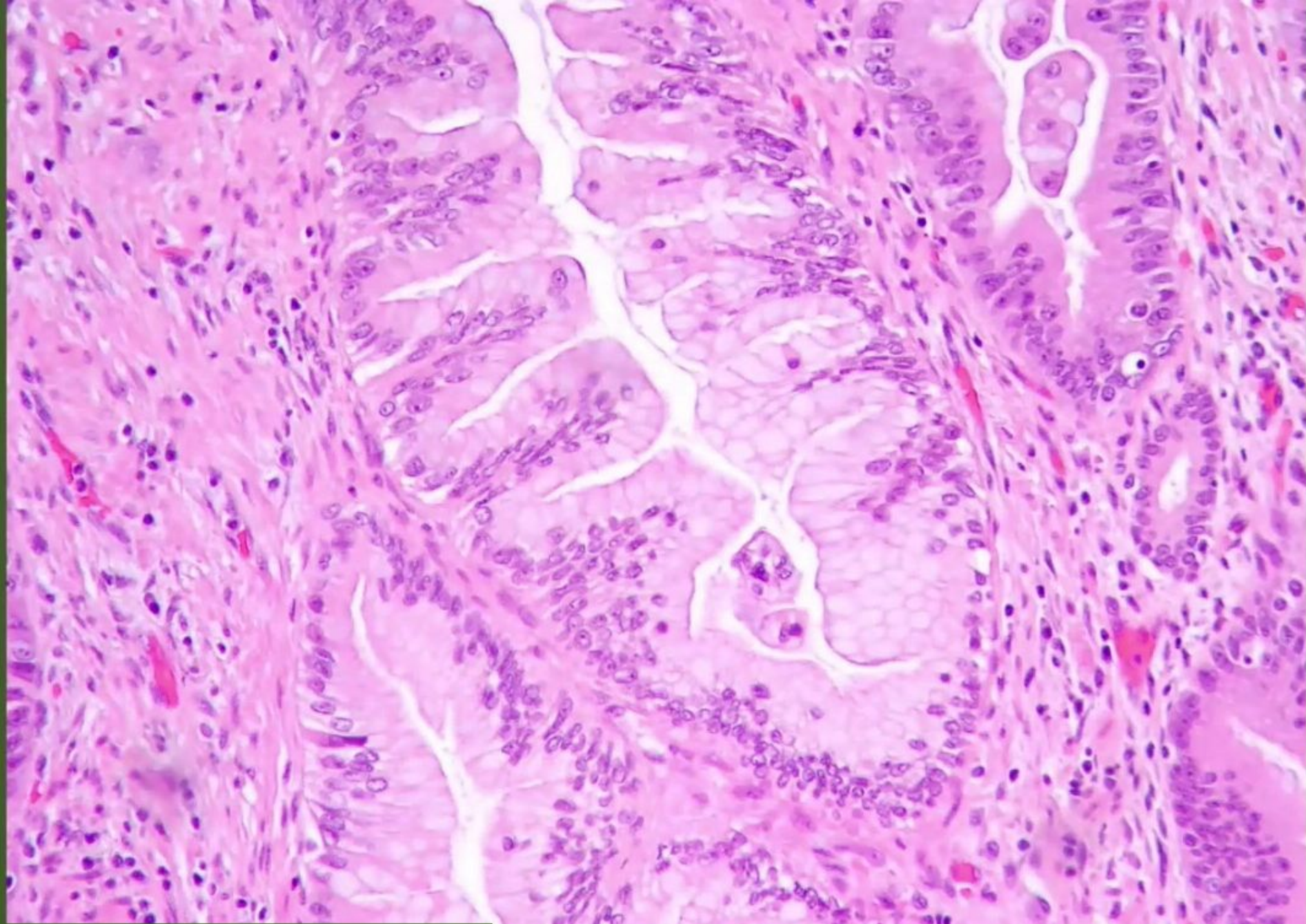


**Intestinal brush
border**

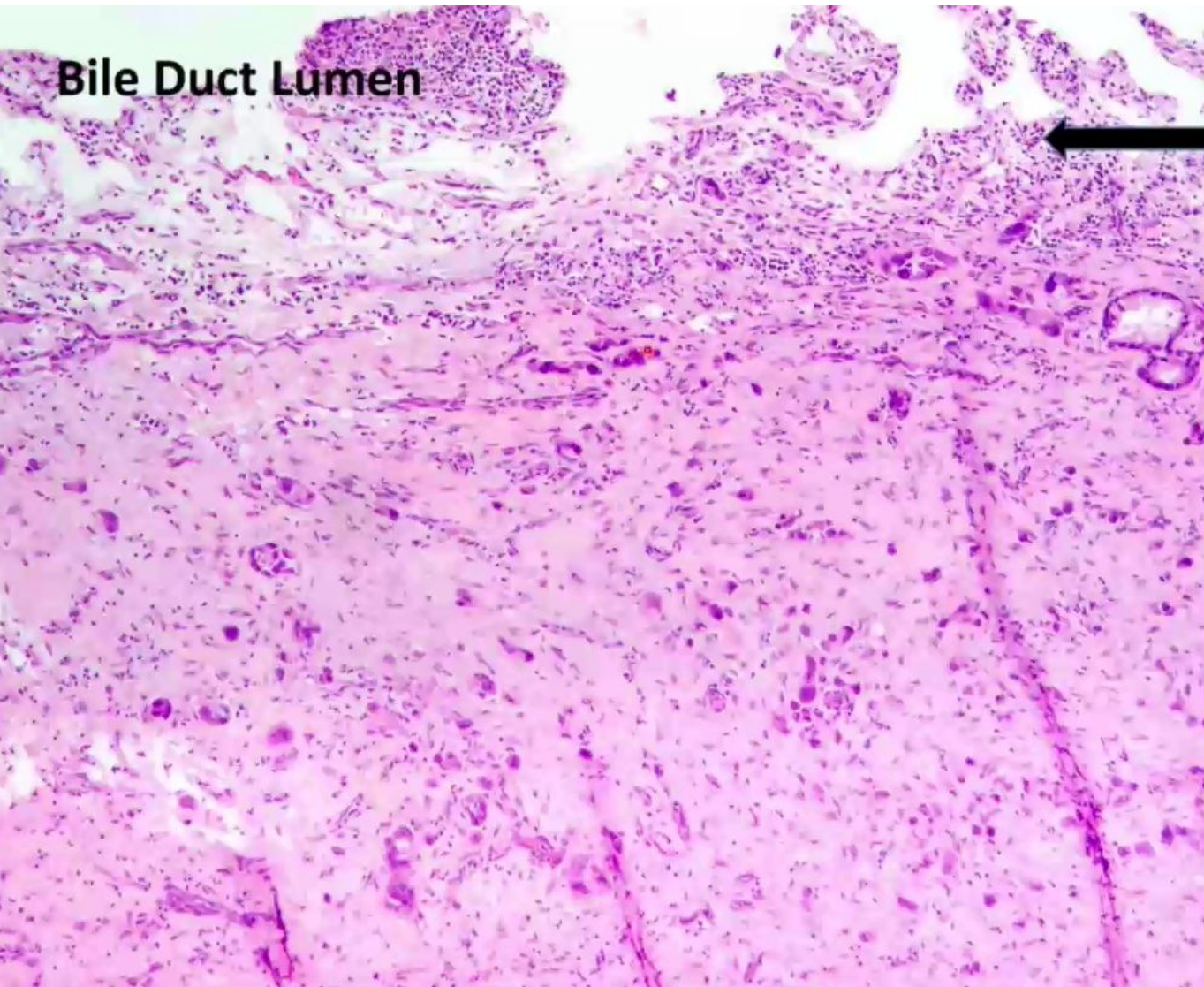


Goblet cells





Foveolar epithelium

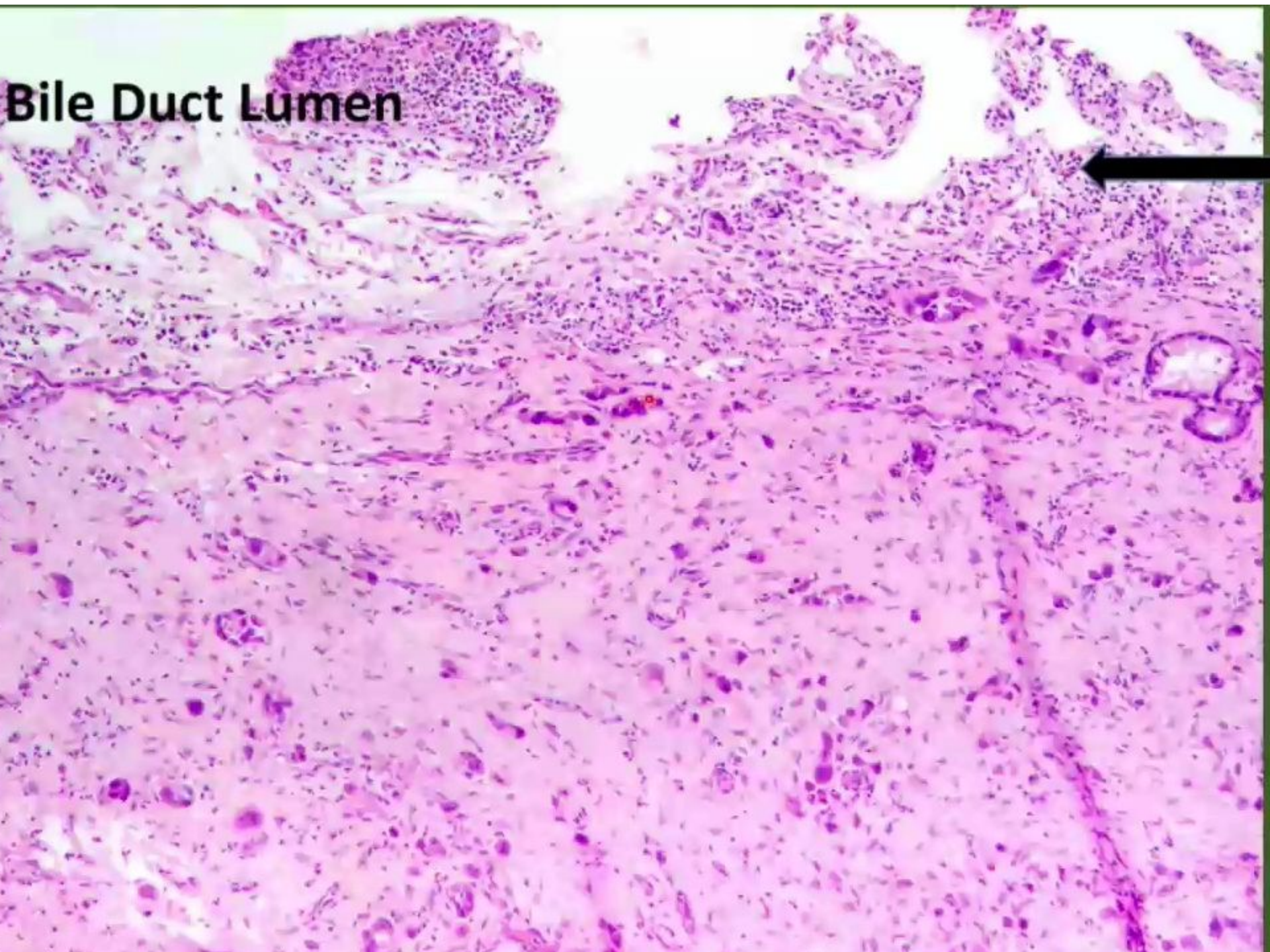


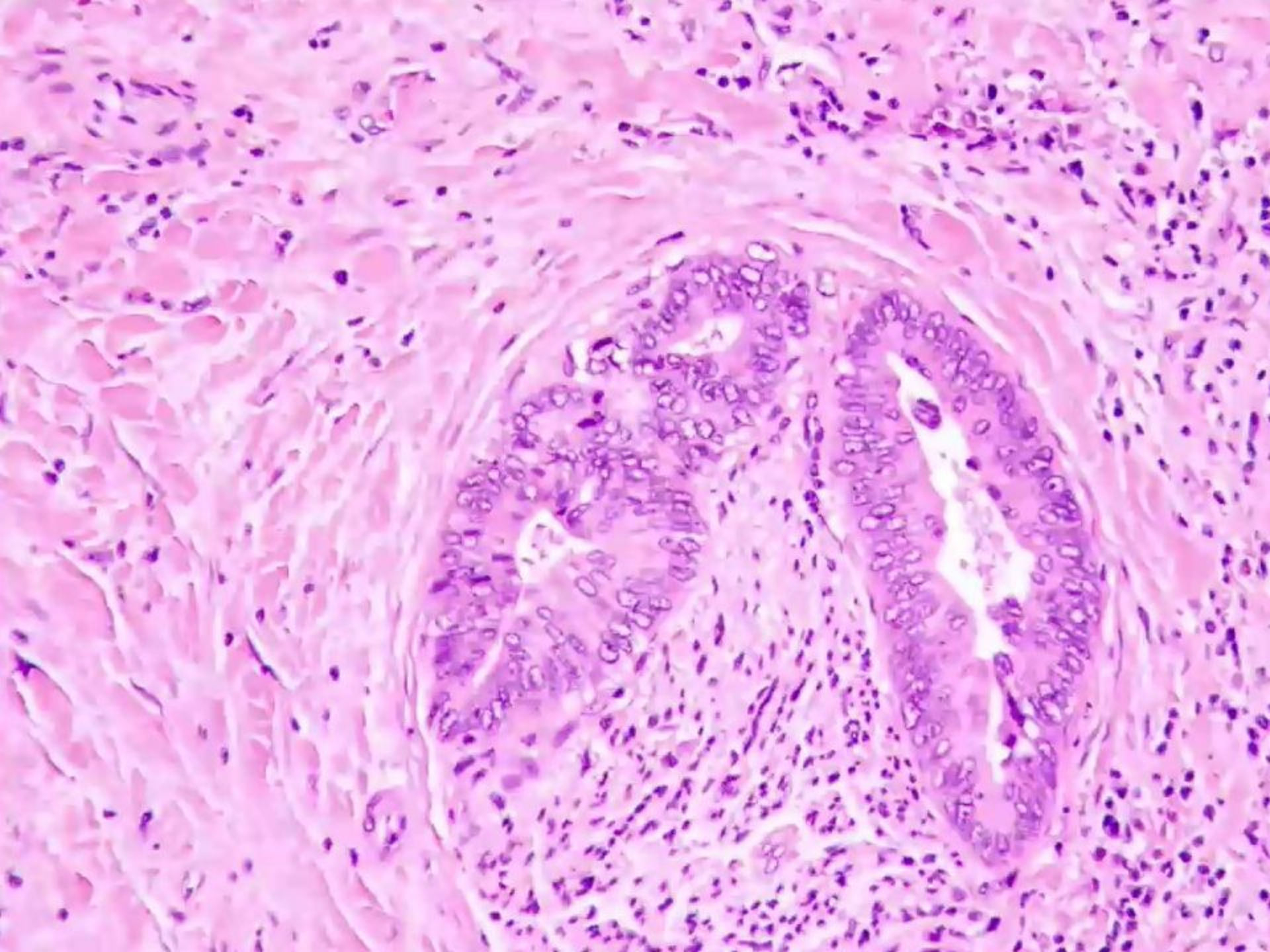
Bile Duct Lumen

Denuded biliary epithelium

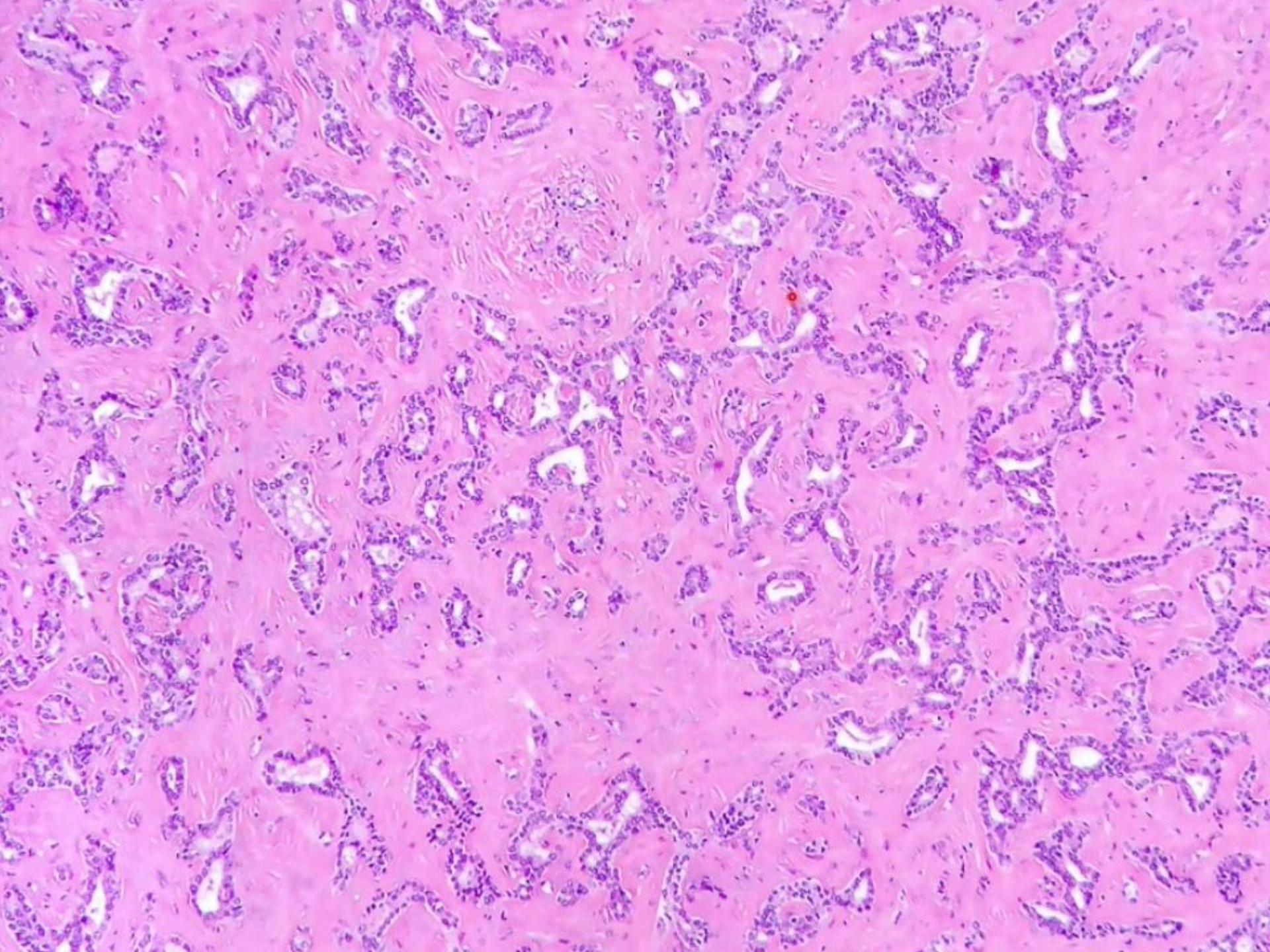
Poorly differentiated adenocarcinoma

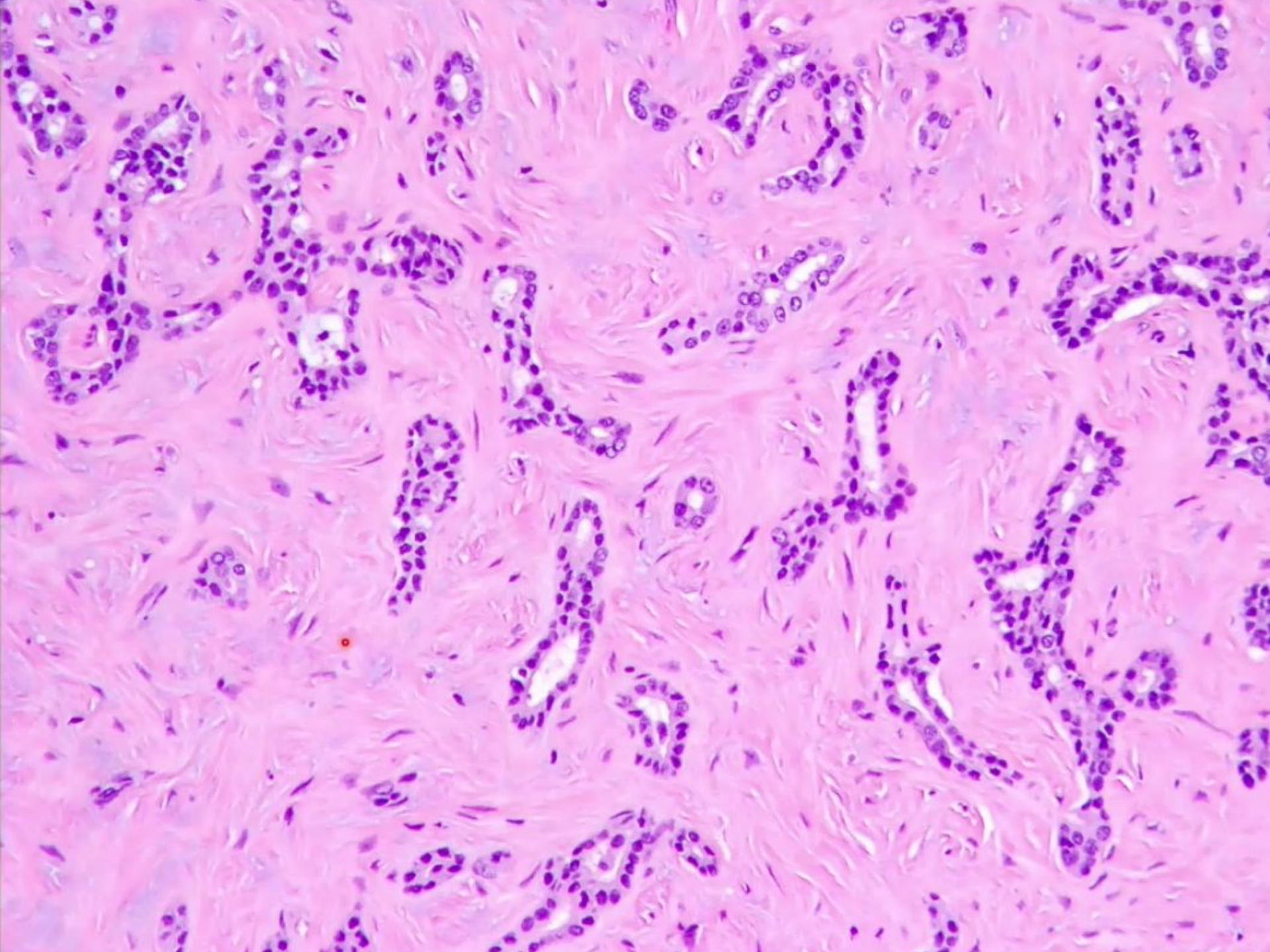
Bile Duct Lumen





Small duct type (type 2)	Usual type (large duct, type 1)
Intrahepatic (peripheral)	Perihilar, distal, intrahepatic
Chronic liver disease with advanced fibrosis (stage 3, 4) – chronic viral hepatitis, steatohepatitis	Chronic biliary disease (PSC, hepatolithiasis, fluke infections)
No biliary intraepithelial neoplasia (BillIN)	BillIN usually present
Mass forming	Any/ all pattern(s)
Well differentiated tubules, anastomosing glands	Large glands with foveolar, pancreatobiliary, intestinal epithelium ± mucin production
No perineural invasion	Perineural invasion common
Lymphovascular, lymph node invasion very rare	Lymphovascular, lymph node invasion common
Pushing tumor borders	Infiltrative tumor borders
CD 56 (NCAM) +	S100, TFF3 + (TreFoil Factor)
KRAS mutations, FGFR2 translocations	IDH1/IDH2 mutations







БЛАГОДАРЮ
СЕРДЕЧНО



САКСЕНА
РОМИЛ