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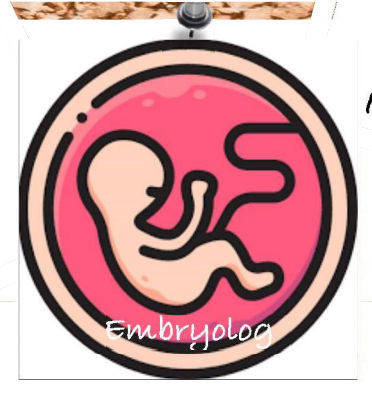
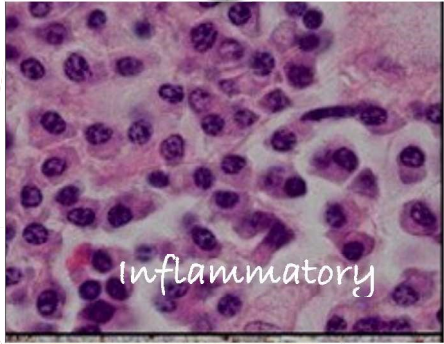


# Uncommon tumours of the liver

Course on liver tumours

Dr. Laurine Verset MD, PhD the 3th December 2022, Antwerp

# Uncommon liver tumours



Infectious

# Liver tumours

## Diagnostic approach

- **Importance of context:**

- ✓ Cirrhosis or not

- ✓ Gender

- **Lesion characteristics:**

- ✓ Solid or cystic

- ✓ Unique or multifocal

- ✓ Containing fat tissue

- ✓ Vascularisation



# Ciliated hepatic foregut cysts

## Clinical features

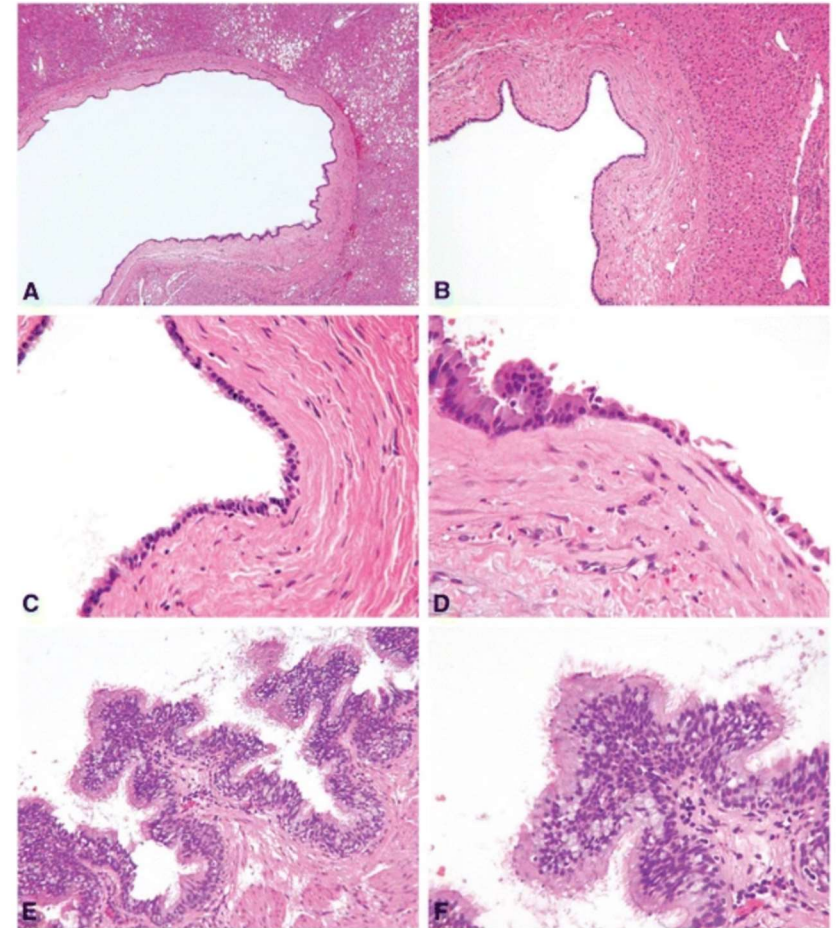
- Rare **cystic** lesions of **embryological** origin
- Small, benign
- Unilocular
- Subcapsular
- Located in segment IV (left lobe)



# Ciliated hepatic foregut cysts

## Microscopic aspects

- **Consist of four layers:**
  - ✓ An inner layer of ciliated pseudostratified column epithelium
  - ✓ Loose lamina propria
  - ✓ Smooth muscle band (one to three layers in thickness)
  - ✓ Outer fibrous capsule



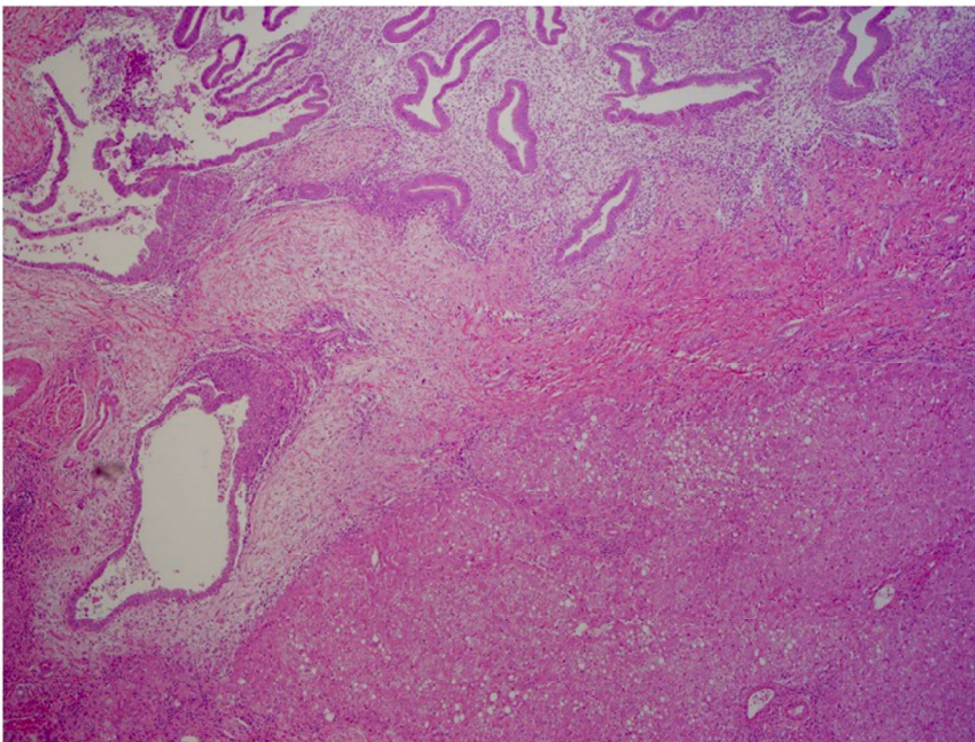
# Endometriosis

## Clinical features

- Common gynecologic disease
  - ✓ presence of endometrial glands and stroma outside of the uterus
  - ✓ 5-15% of women of reproductive age
- Extrapelvic endometriosis
  - ✓ gastrointestinal tract, urinary system, thoracic cavity, kidneys and pancreas
  - ✓ Exact prevalence is unknown
  - ✓ older population with median age of 34-40 years

# Endometriosis

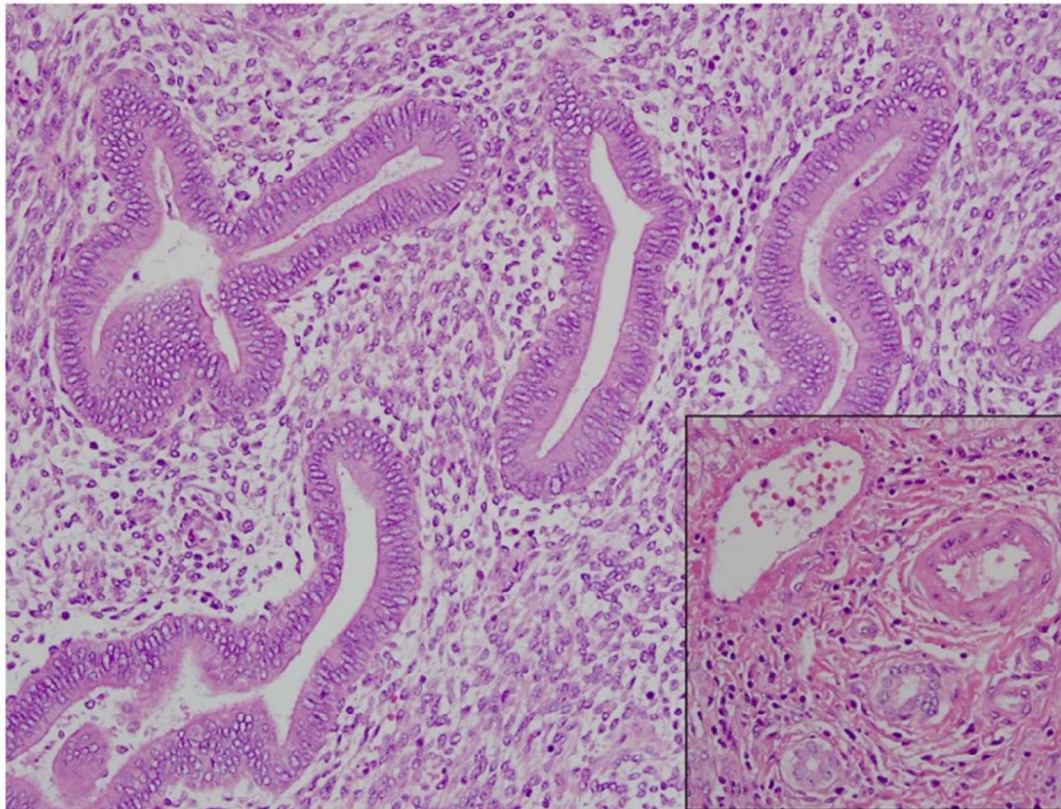
## Microscopic aspects





# Endometriosis

## Microscopic aspects





# Mucinous cystic neoplasm

## Essential features

- Exclusively in **woman**
- Cyst-forming epithelial neoplasm
  - ✓ **Multilocular** cystic lesion
  - ✓ **No communication with the bile duct**
  - ✓ Cuboidal and columnar neoplastic epithelia with variable atypia; **ovarian-like stroma** (at least focally positive for ER and/or PR)

# Mucinous cystic neoplasm

## Clinical aspects and pathophysiology

- **F >>>>** (M)
- Mean age: 51 y
- **Liver (left lobe)** but occasionally extrahepatic biliary system
- **Solitary**
- **KRAS mutations** in 20% of MCNs (rare in case of MCN with low grade dysplasia)
- Etiology unknown

# Mucinous cystic neoplasm

## Macroscopic aspects

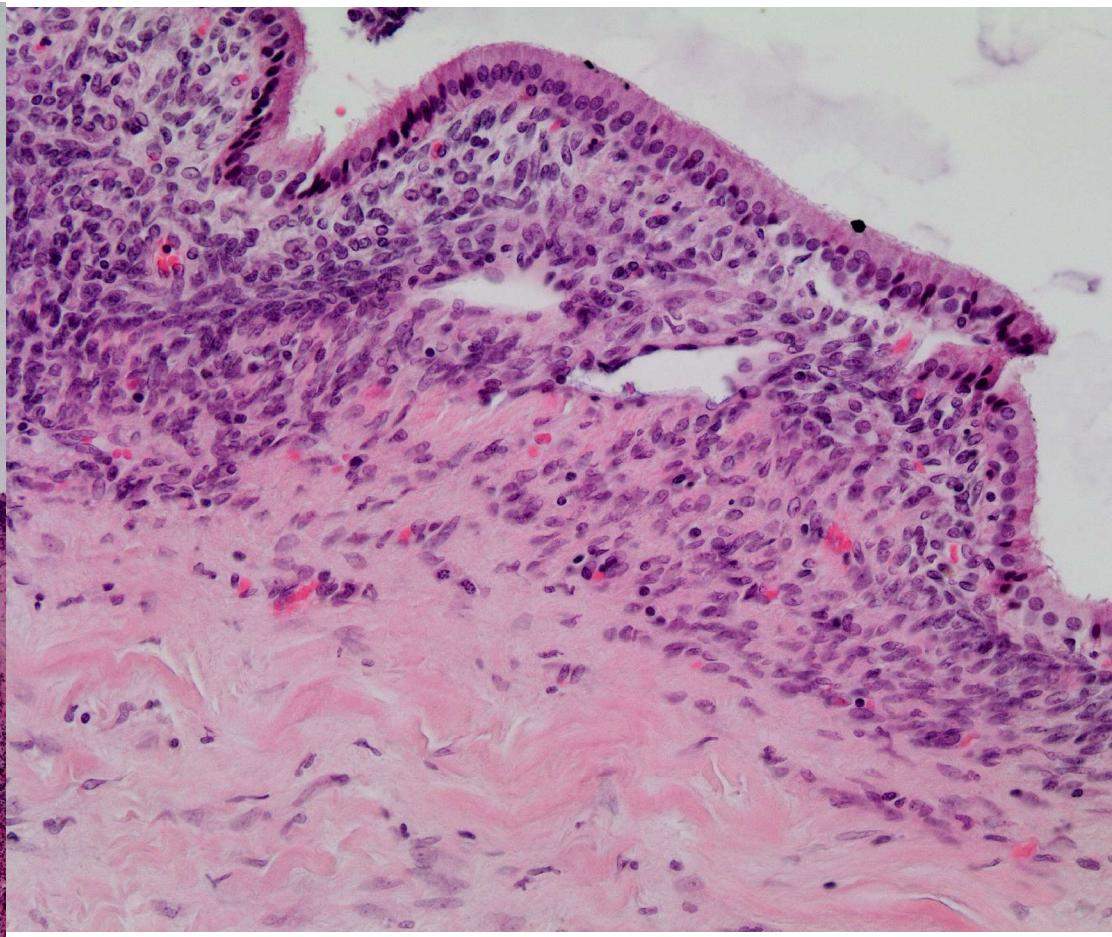
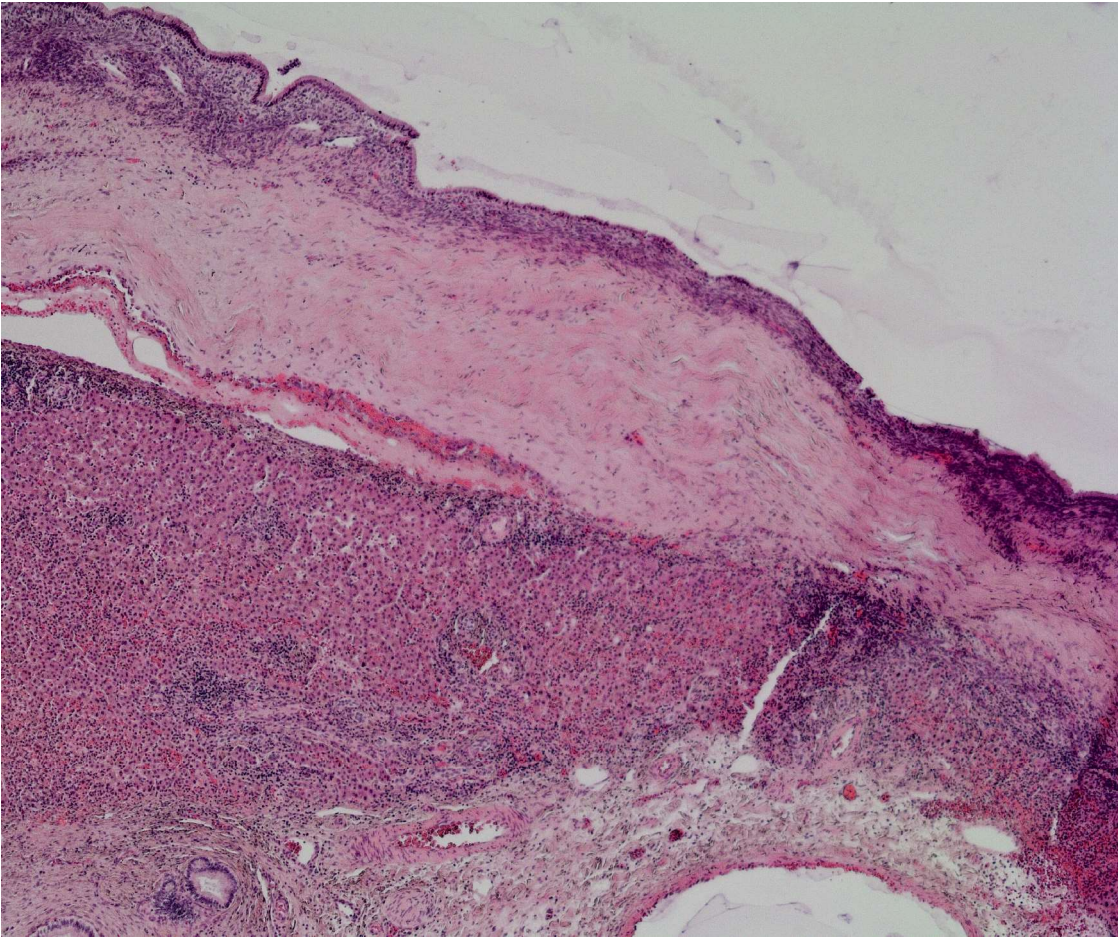
- Well demarcated
- Multilocular
- Size: 5 à 29 cm (mean: 11 cm)
- Mucinous, clear or hemorrhagic fluid
- Solid area (in case of associated invasive carcinoma)





# Mucinous cystic neoplasm

## Microscopic aspects





# Mucinous cystic neoplasm

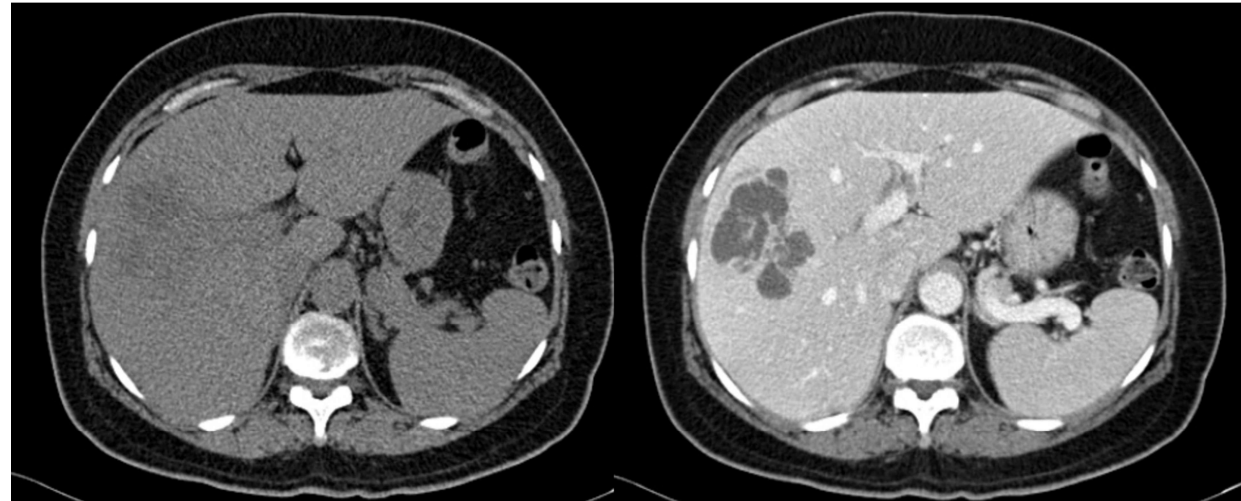
## Staging and prognosis

- Excellent if complete excision is possible
- MCN with associated adenocarcinoma follows the TNM classification for intrahepatic cholangiocarcinoma
  - ➡ !!!! sampling of solid area because invasive component should be focal

# Liver abscess

## Clinical features

- Pyogenic liver abscess
- Most common cause of bacterial liver abscess:
  - ✓ **Biliary disease**
  - ✓ Septicaemia
  - ✓ Traumatic liver injury
  - ✓ IBD
  - ✓ Diverticulitis
  - ✓ Ruptured appendix

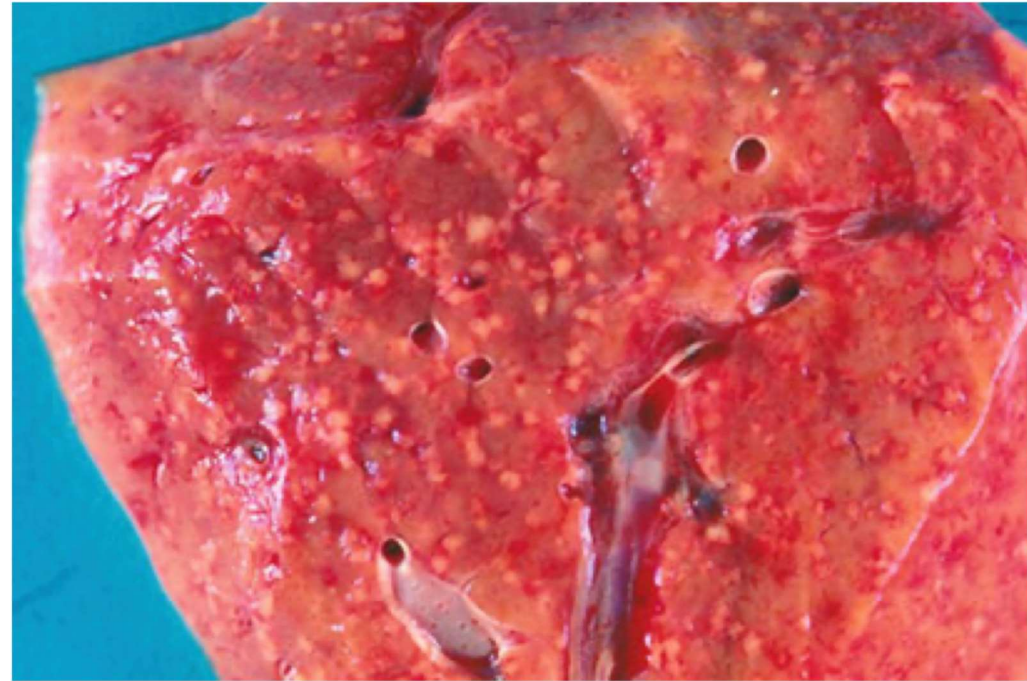
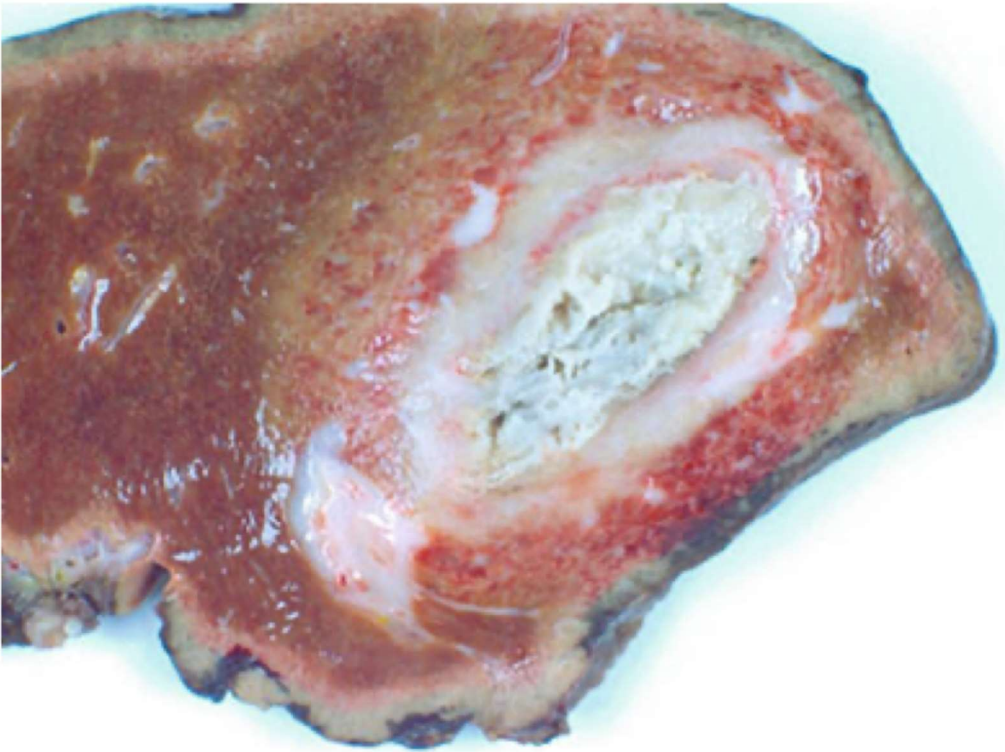


### Radiological features:

1. Cystic lesion, multiloculated with blurred limits
2. CT+C: enhancement of pericystic wall
3. Peripheric oedema

# Liver abscess

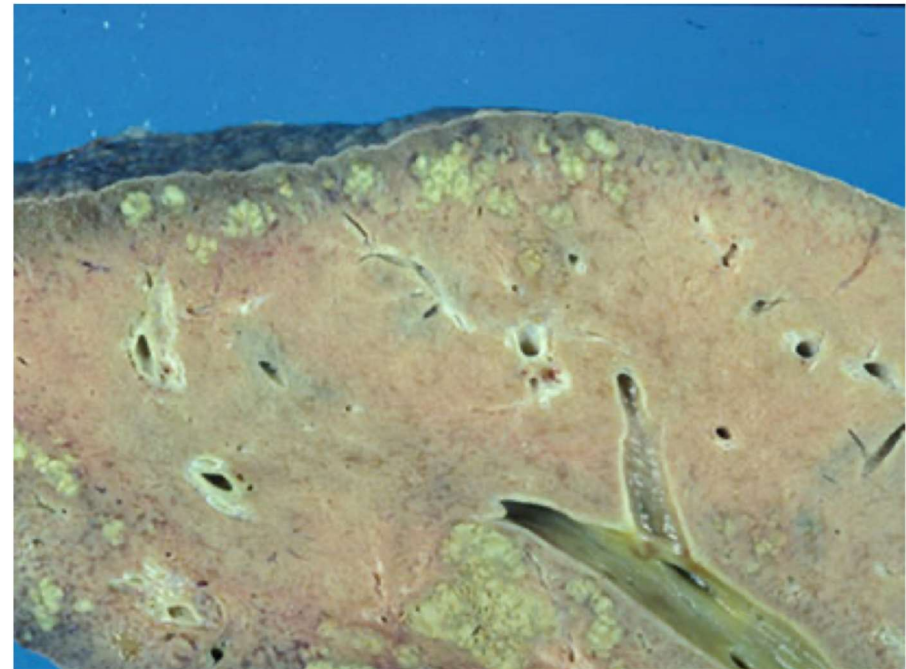
## Macroscopic aspects



*Zimmermann, A. (2017). Liver Abscesses as Pseudotumoral Lesions. In: Tumors and Tumor-Like Lesions of the Hepatobiliary Tract. Springer, Cham.*

# Liver abscess

## Macroscopic aspects

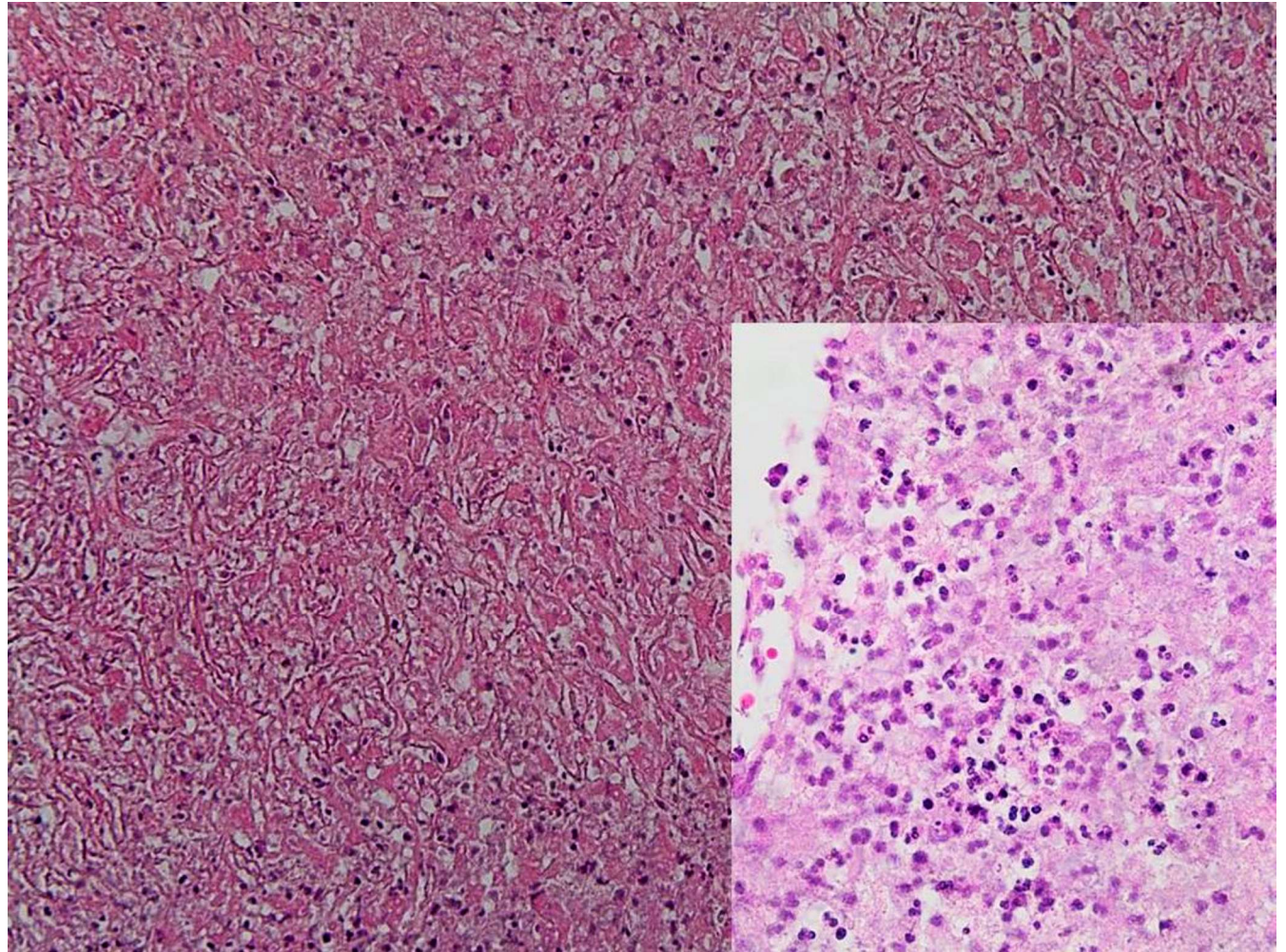


*Zimmermann, A. (2017). Liver Abscesses as Pseudotumoral Lesions. In: Tumors and Tumor-Like Lesions of the Hepatobiliary Tract. Springer, Cham.*



# Liver abscess

## Microscopic aspects

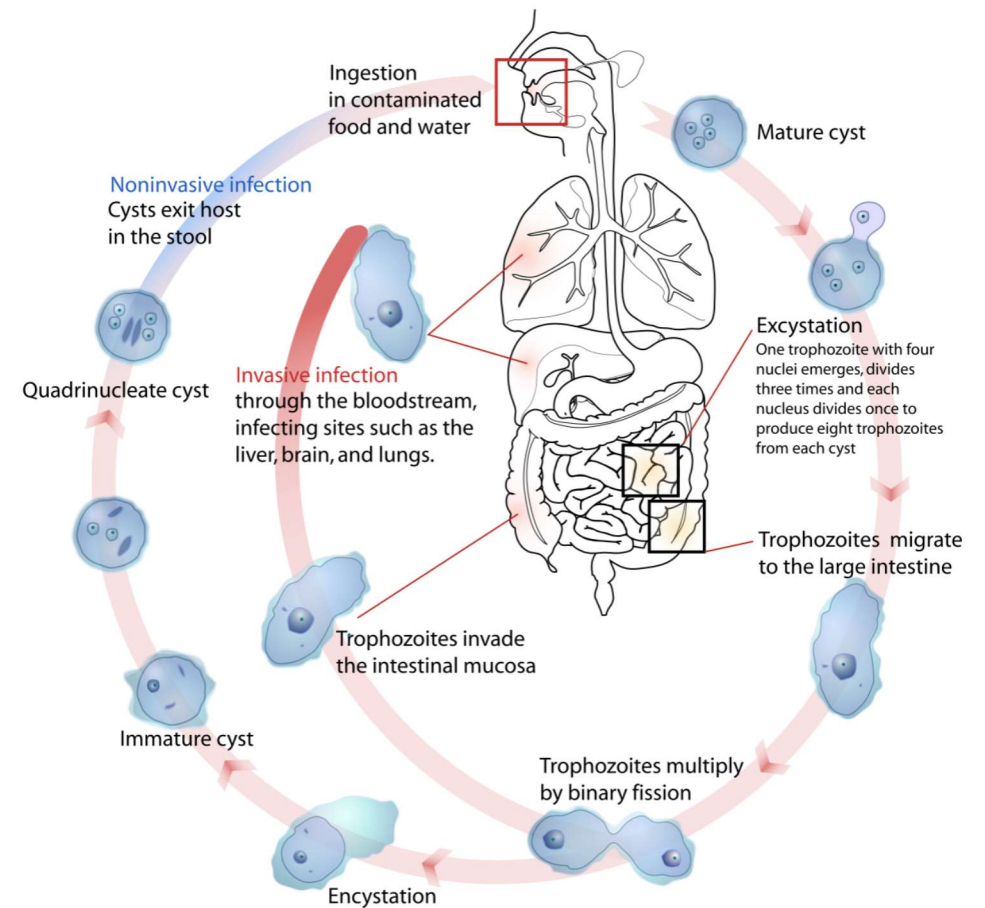
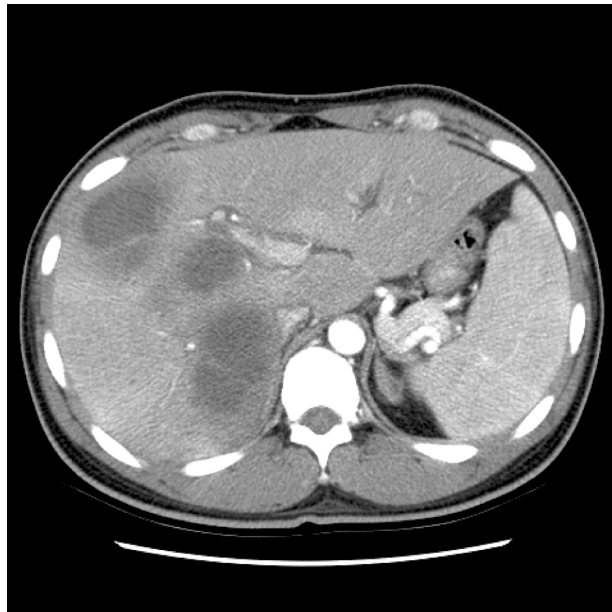


# Liver abscess

## Clinical features

- Entamoeba Histolytica (Amebic) abscess

✓ Usually fecal oral spread

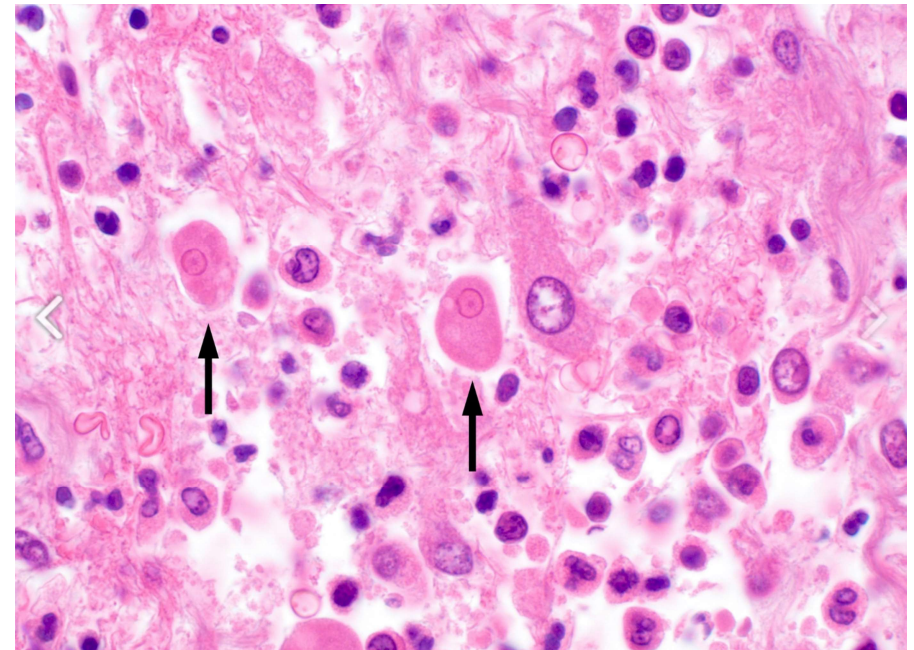




# Liver abscess

## Microscopic aspects

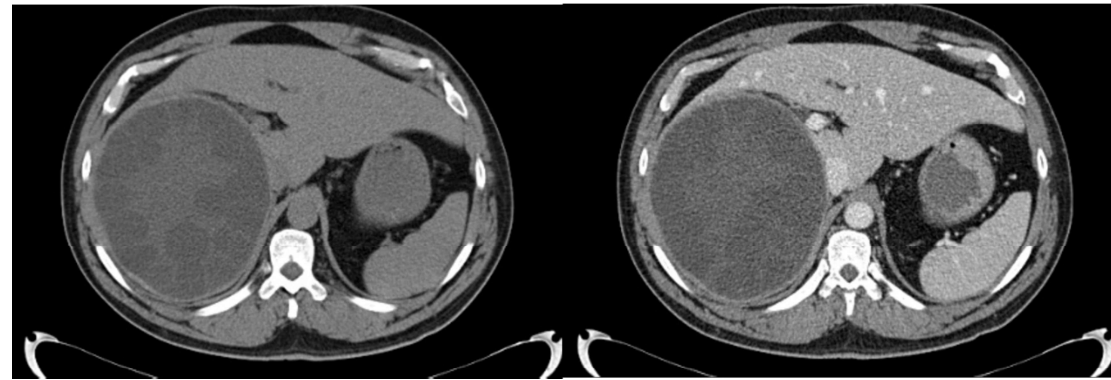
- Entamoeba Histolytica (Amebic) abscess
- ✓ Amebic trophozoites
  - Round nucleus with peripherally condensed ring of chromatin and central dot-like karyosome



# Parasitic cyst

## Clinical features

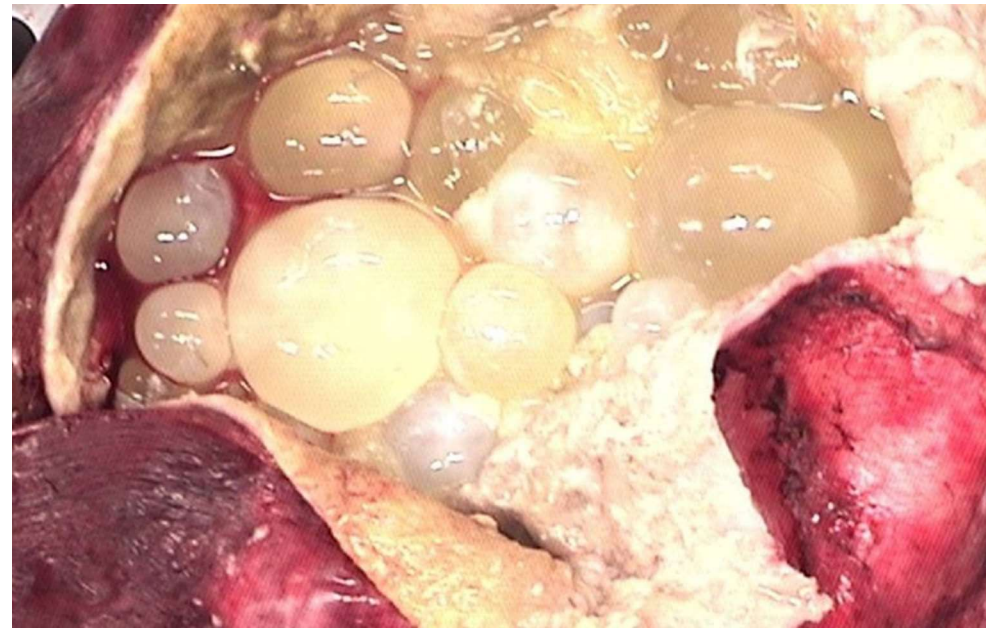
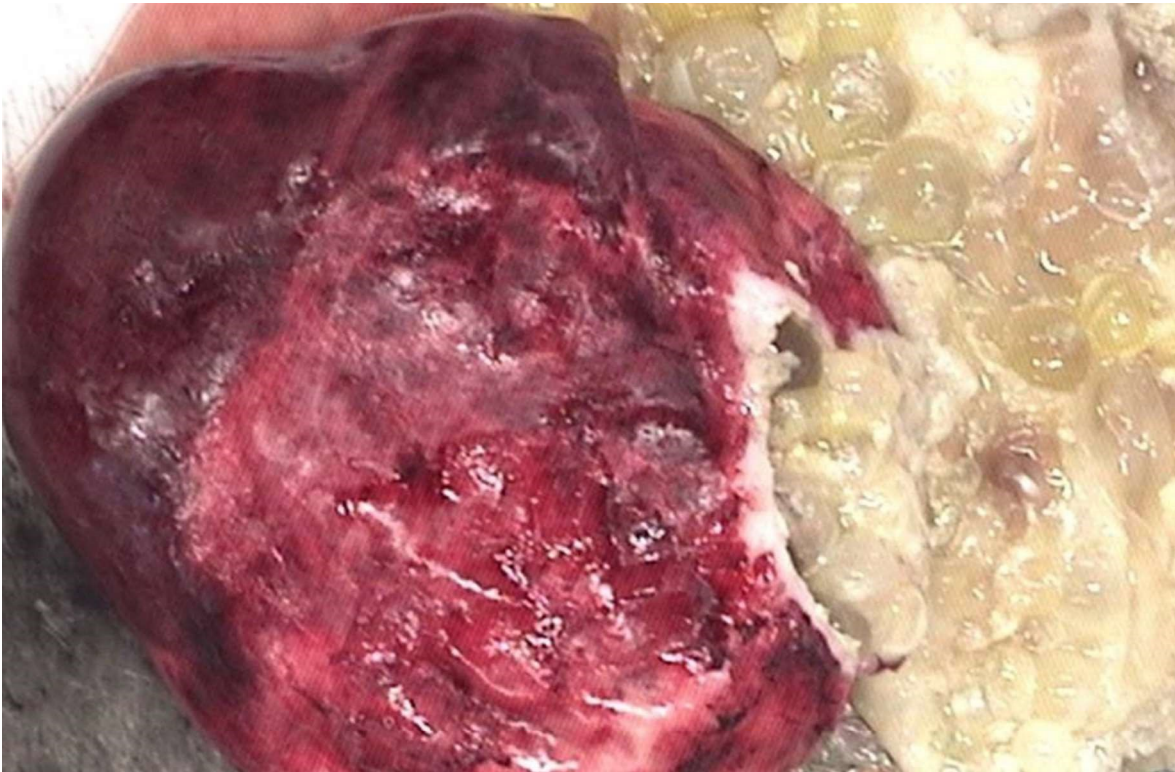
- **Hydatid cyst (echinococcosis)**
  - Zoonotic disease
- **4 forms:**
  - **Cystic echinococcosis** (hydatidosis) caused by *Echinococcus granulosus*
  - **Alveolar echinococcosis** caused by *E. Multilocularis*
  - Polycystic (*E. Vogeli*) and unicystic (*E. Oligarthrus*)





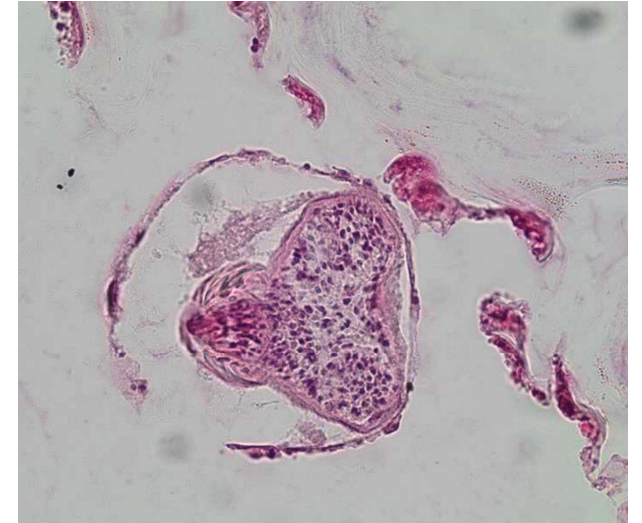
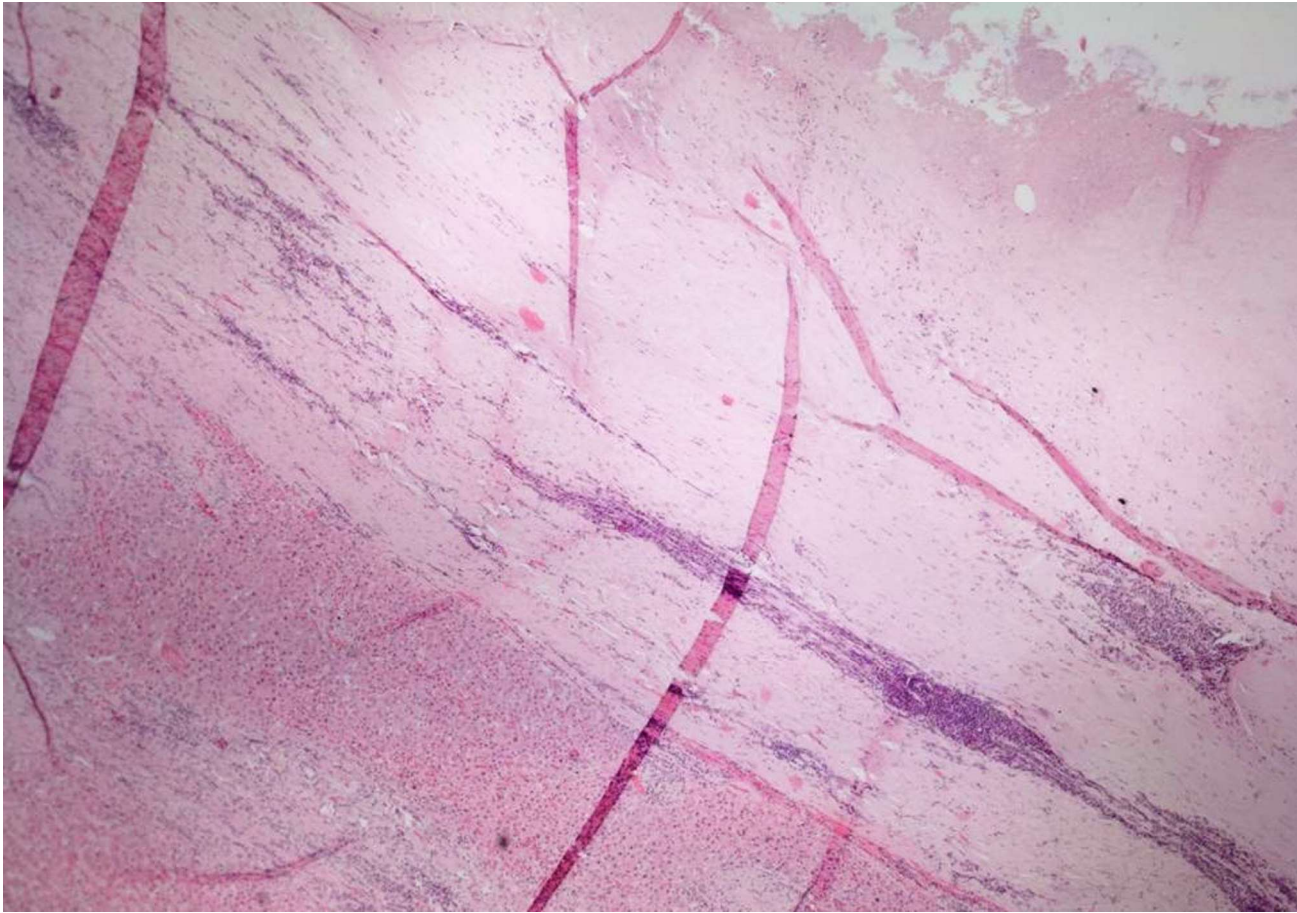
# Parasitic cyst

## Macroscopic aspects



# Parasitic cyst

## Microscopic aspects



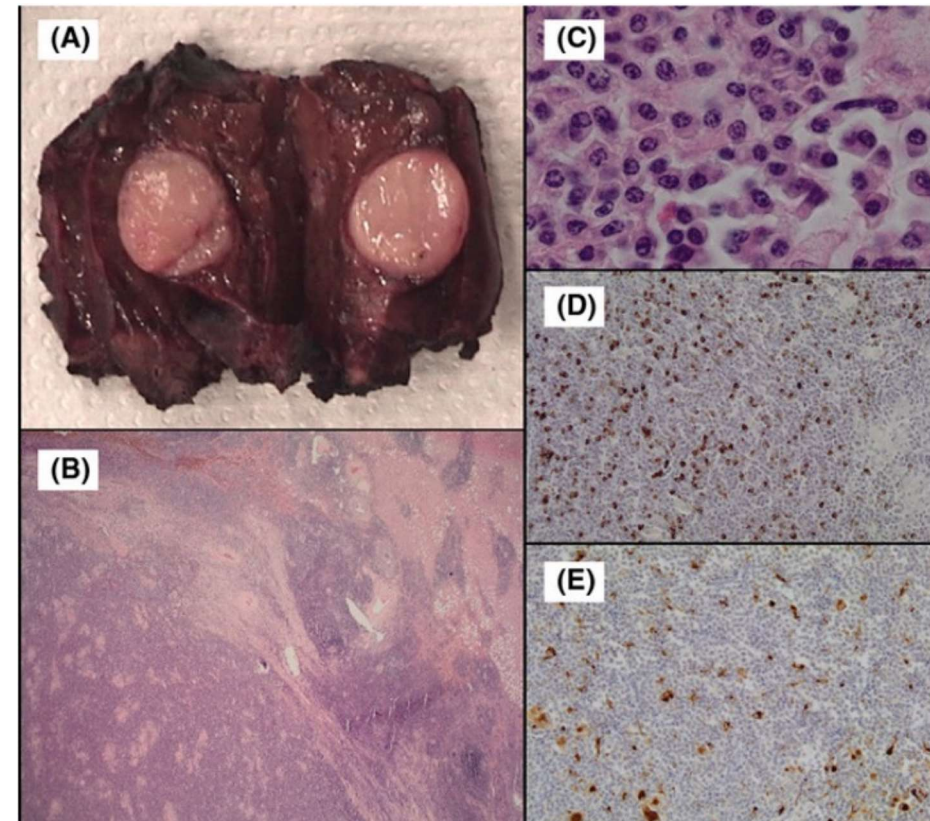
Protoscolex



# IgG4 related inflammatory pseudotumor

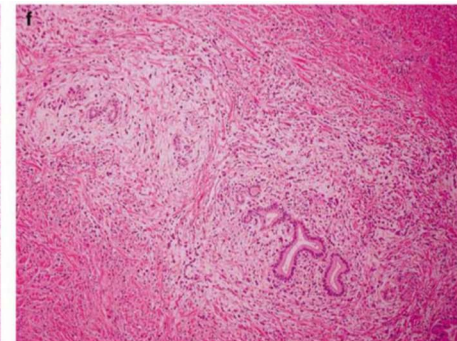
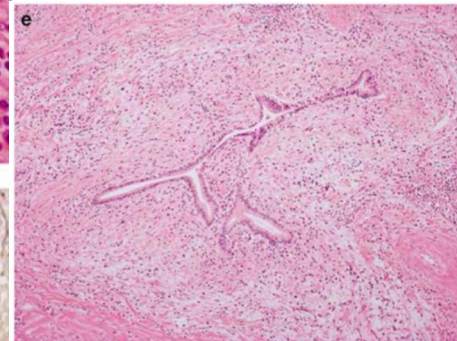
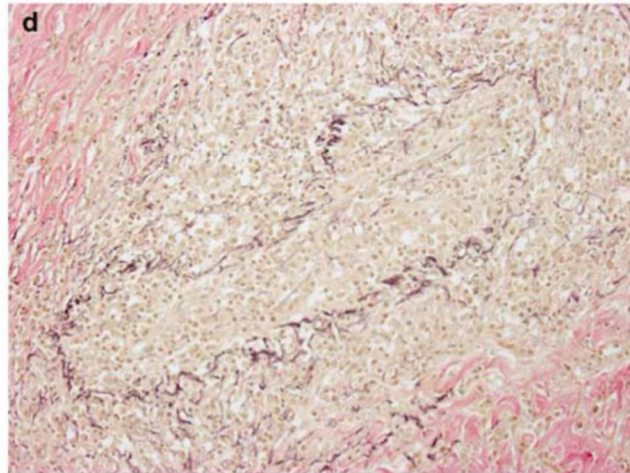
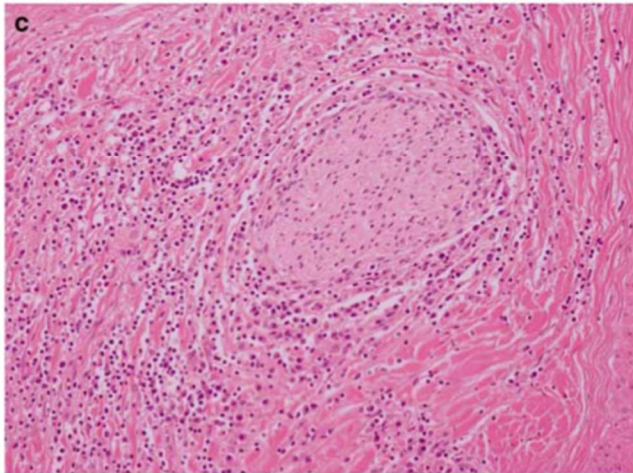
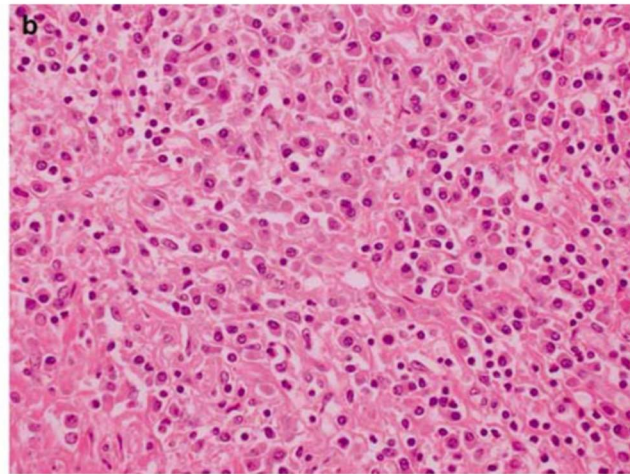
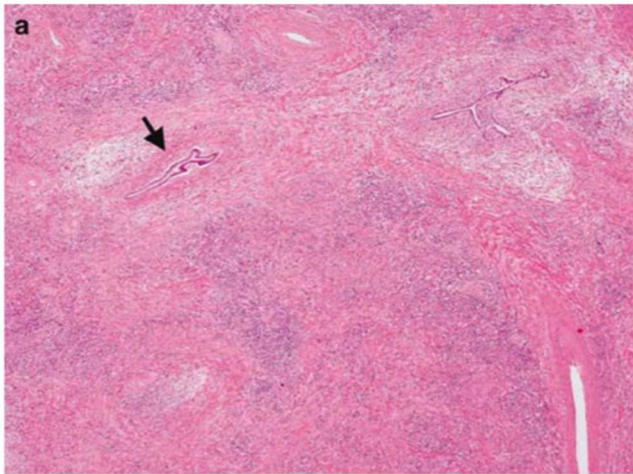
## Clinical aspects

- IgG4 is an **immune-mediated fibroinflammatory** condition that is capable of affecting multiple organs
- Common forms of presentation:
  - ✓ Type 1 (IgG4-related) autoimmune pancreatitis
  - ✓ Sclerosing cholangitis
  - ✓ Salivary gland disease
  - ✓ Orbital disease
  - ✓ Retroperitoneal fibrosis
  - ✓ Mass lesion: pancreas, biliary tree, orbits, lungs, kidneys, major salivary gland or lacrimal gland



# IgG4 related inflammatory pseudotumor

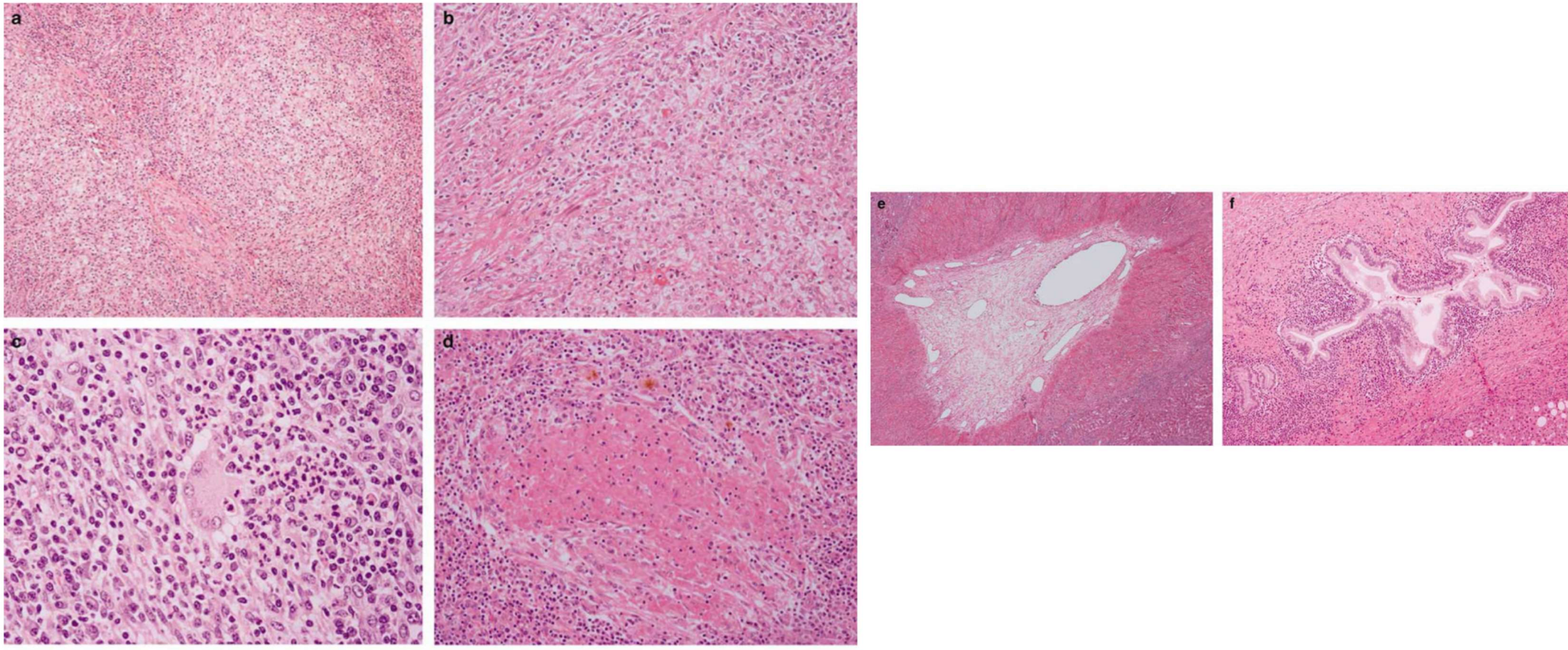
## Microscopic aspects - Lymphoplasmacytic type





# IgG4 related inflammatory pseudotumor

## Microscopic aspects - Fibrohistiocytic type



# IgG4 related inflammatory pseudotumor

## Comparison of microscopic aspects

**Table 2** Comparison of clinical characteristics between hepatic inflammatory pseudotumors of fibrohistiocytic and lymphoplasmacytic types

	<i>Fibrohistiocytic type (n = 10)</i>	<i>Lymphoplasmacytic type (n = 6)</i>	<i>P-value</i>
Average age (range)	68 (59–82)	67 (56–79)	0.586
Male/female ratio	5/5	6/0	0.431
Nodular size (cm)	2.8 (1.5–4.5)	3.4 (2.0–4.8)	0.231
<i>Hepatic lobes</i>			0.197
Left lobe	5 (50%)	5 (83%)	
Right lobe	5 (50%)	1 (17%)	
<i>Location</i>			0.003
Peripheral liver	8 (80%)	0	
Hilar bile ducts	2 (20%)	6 (100%)	
<i>Shapes</i>			0.003
Mass-forming type	8 (80%)	0	
Periductal infiltrating type	2 (20%)	6 (100%)	
<i>Clinical presentation</i>			0.017
Subjective symptoms	8 (80%)	0	
Liver dysfunction by laboratory test	2 (20%)	6 (100%)	
Extrahepatic lesion	Carcinoma of the Vater's ampulla (1)	Autoimmune pancreatitis (1) Chronic viral hepatitis (HCV) (1)	

n, number of cases; (1), one case having this extrahepatic lesion.

# Angiomyolipoma

## Essential features

- Benign **mesenchymal tumour** composed of (in variable amounts and growth pattern):
  - ✓ Smooth muscle cells
  - ✓ Adipocytes
  - ✓ Blood vessels
- Usually **sporadic** (5-10% tuberous sclerosis, multiple with renal angiomyolipoma)
- **Rare malignant behavior** (recurrence, metastasis) reported (difficult to determine)
- Smooth muscle component (usually predominant) express HMB45, SMA and MelanA

# Angiomyolipoma

## Clinical aspects and pathophysiology

- F > M
- Mean age: 56 y
- Right lobe > Left lobe > caudate lobe
- Mutation *TSC2* gene (sporadic)
- Tumour included in Perivascular Epithelioid Cell Tumor (PEComa)

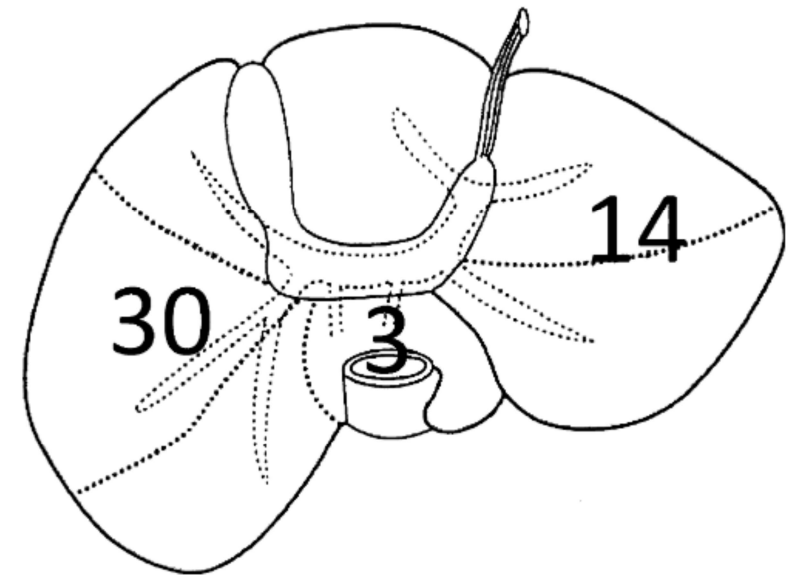


Figure 1. Localization of 47 hepatic angiomyolipomas.

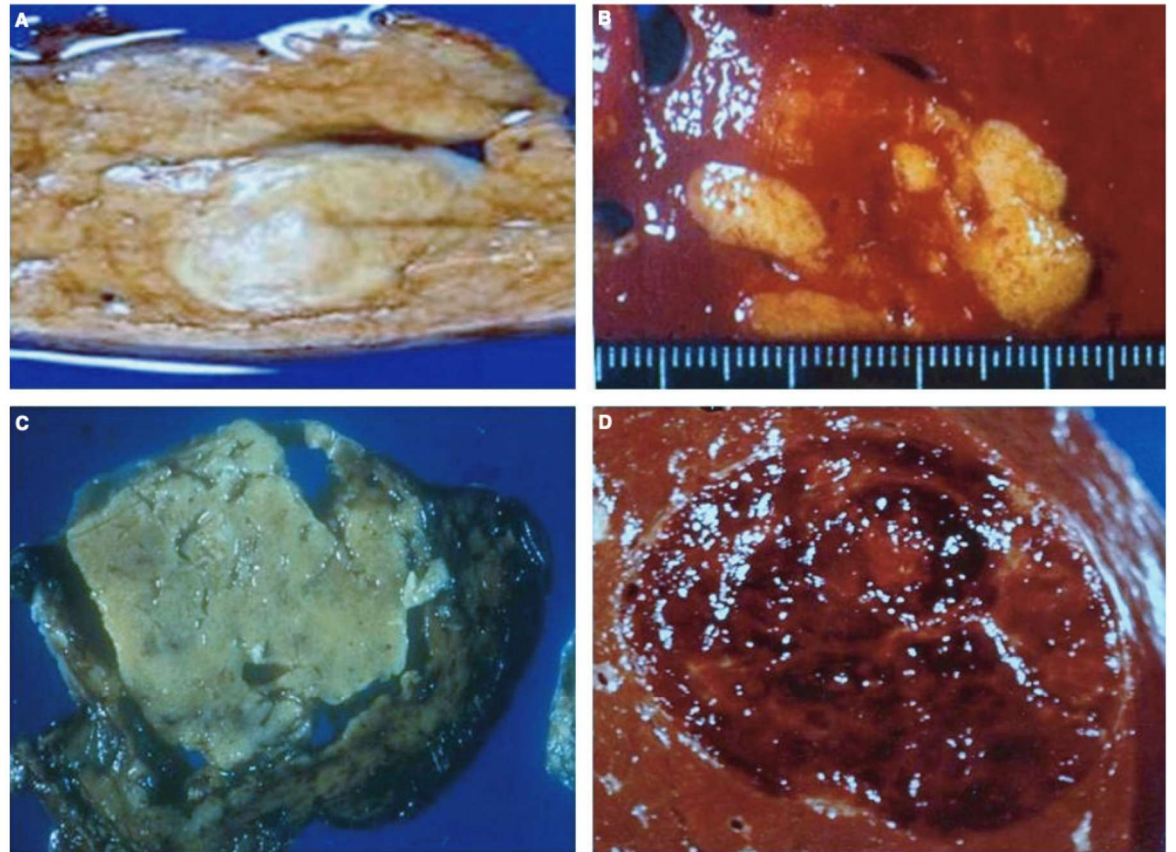
*Nomomura A et al. Histopathology 2012*  
*Yan Z et al Histopathology 2018*



# Angiomyolipoma

## Macroscopic aspects

- Well circumscribed
- Unencapsulated or partially encapsulated
- Myomatous
- Mixed (conventional)
- Lipomatous
- Angiomyomatous
- Non cirrhotic liver



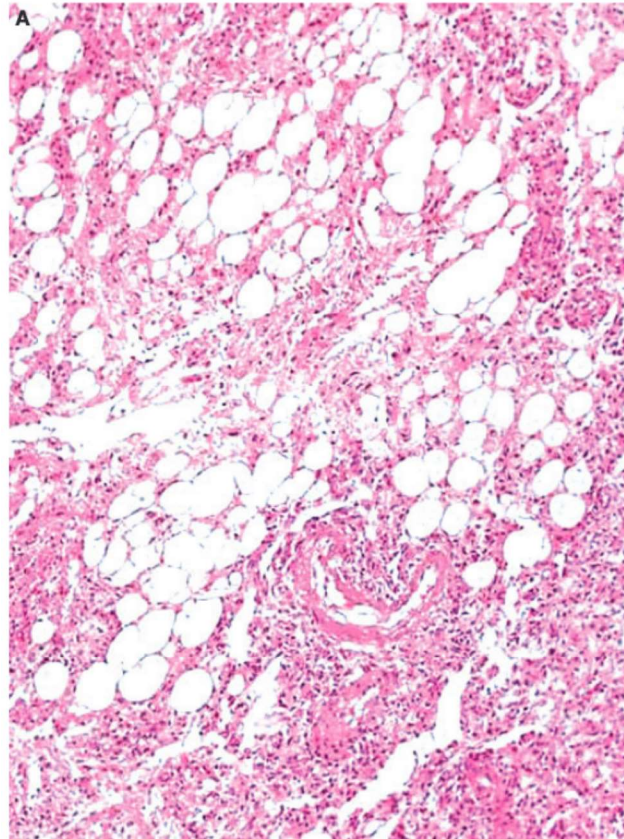
**Figure 2.** Macroscopic appearance of hepatic angiomyolipoma, showing myomatous (A), mixed (conventional) (B), lipomatous (C) and angiomyomatous (D) types.

# Angiomyolipoma

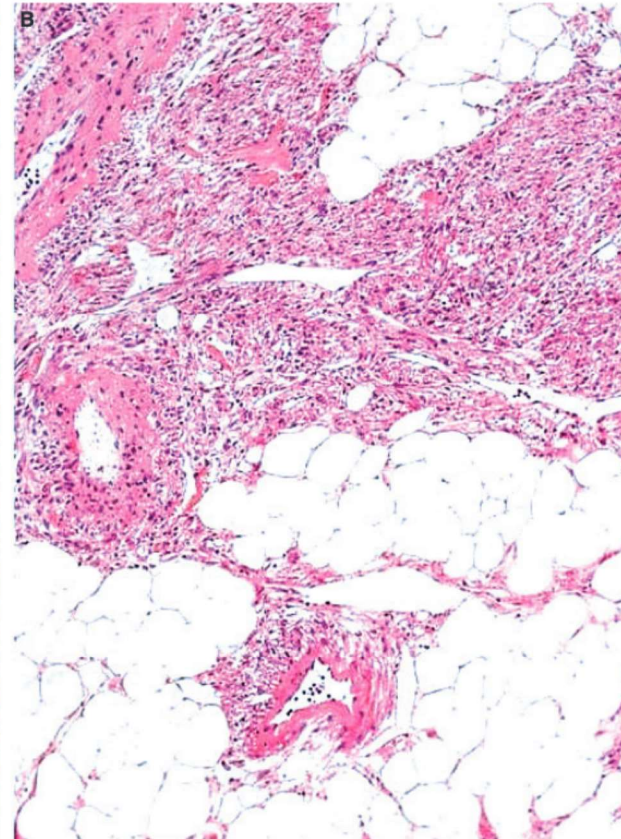
## Microscopic aspects

Mixed  
(conventional) type

Intermingled type



Mosaic  
(geographical) type

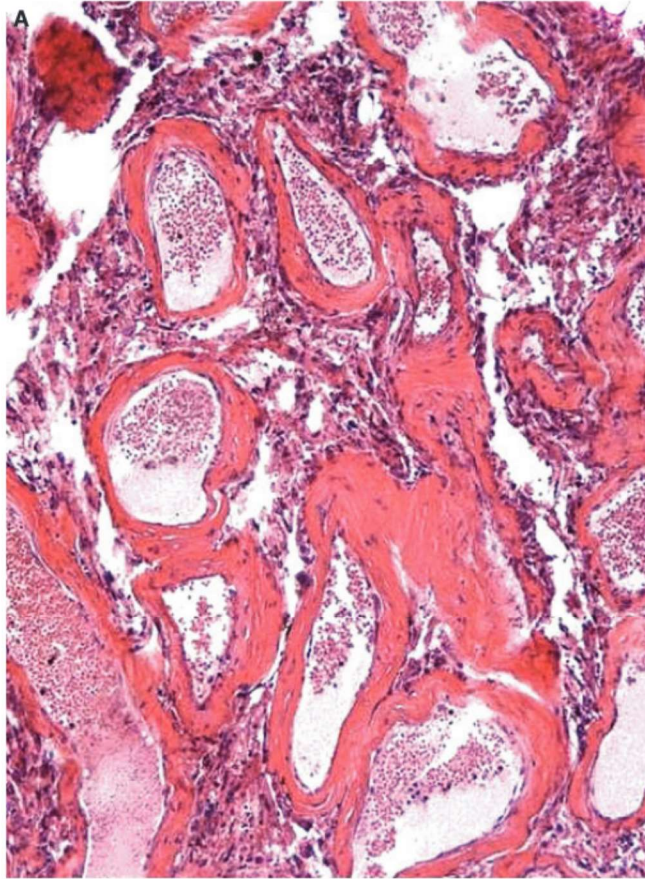




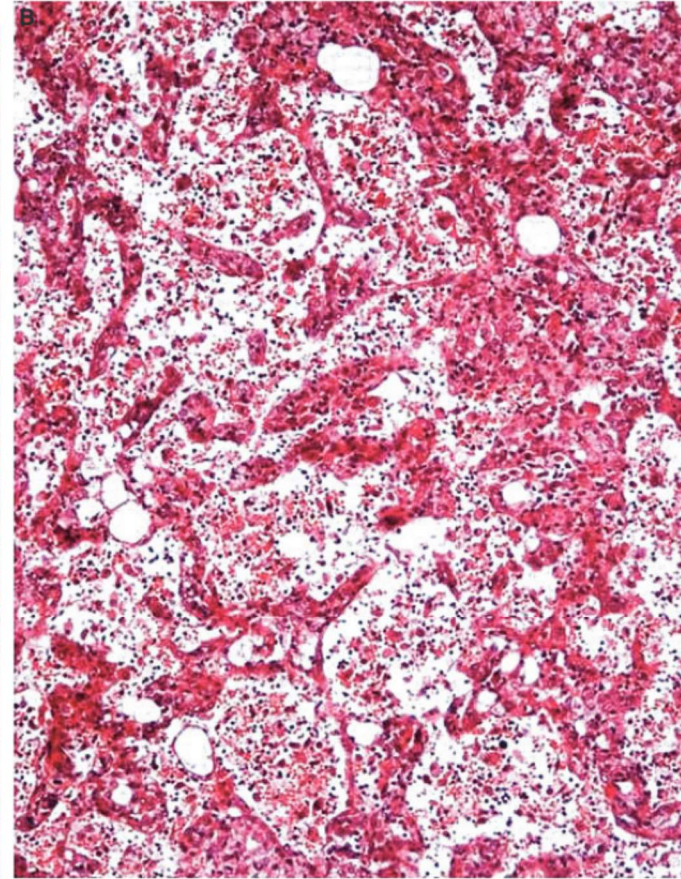
# Angiomyolipoma

## Microscopic aspects

Angiomatous blood vessels



Dilated sinusoidal blood vessels

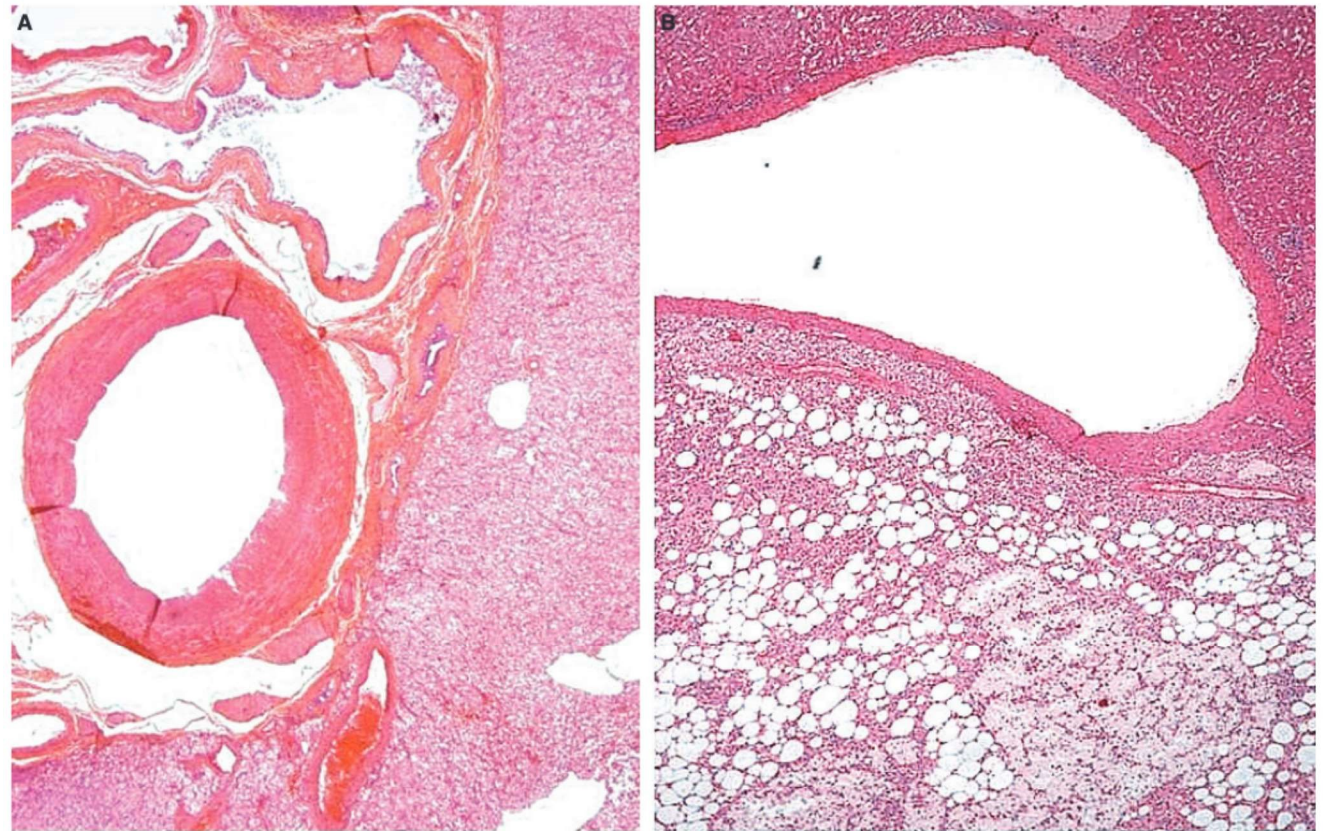




# Angiomyolipoma

## Microscopic aspects

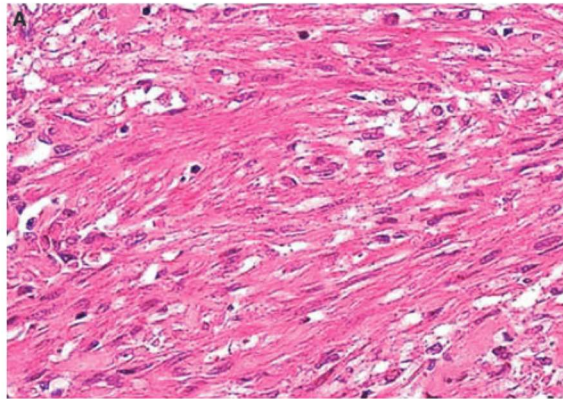
Large blood vessels could be observed in the adjacent liver parenchyma



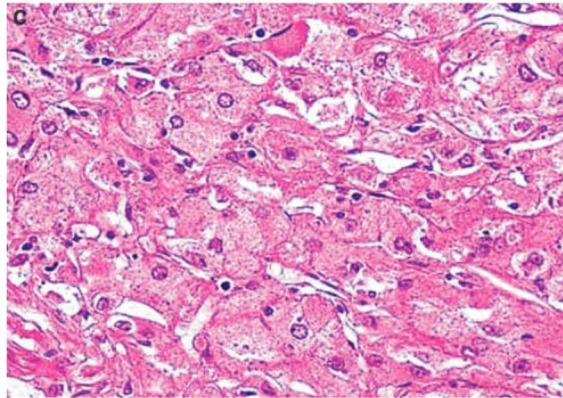
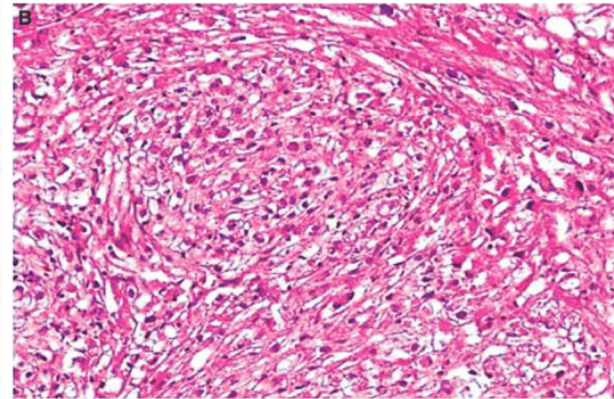
# Angiomyolipoma

## Microscopic aspects

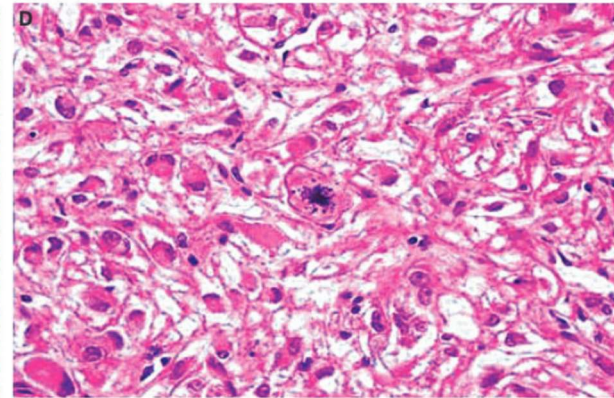
Spindle cells



Nodular or whorled pattern



Epithelioid



Pleiomorphic

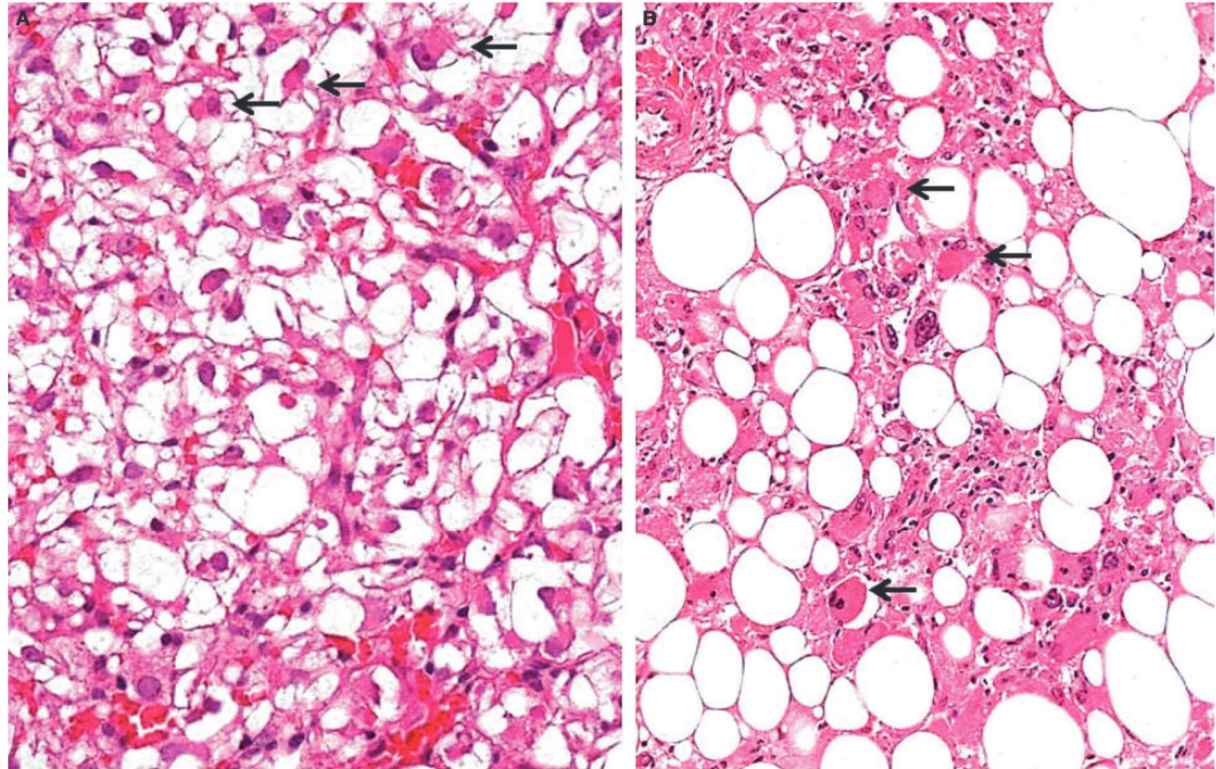


# Angiomyolipoma

## Microscopic aspects

Epithelioid smooth muscle could display:

- Clear cell feature
- Rhabdoid feature
- Severe nuclear pleomorphism



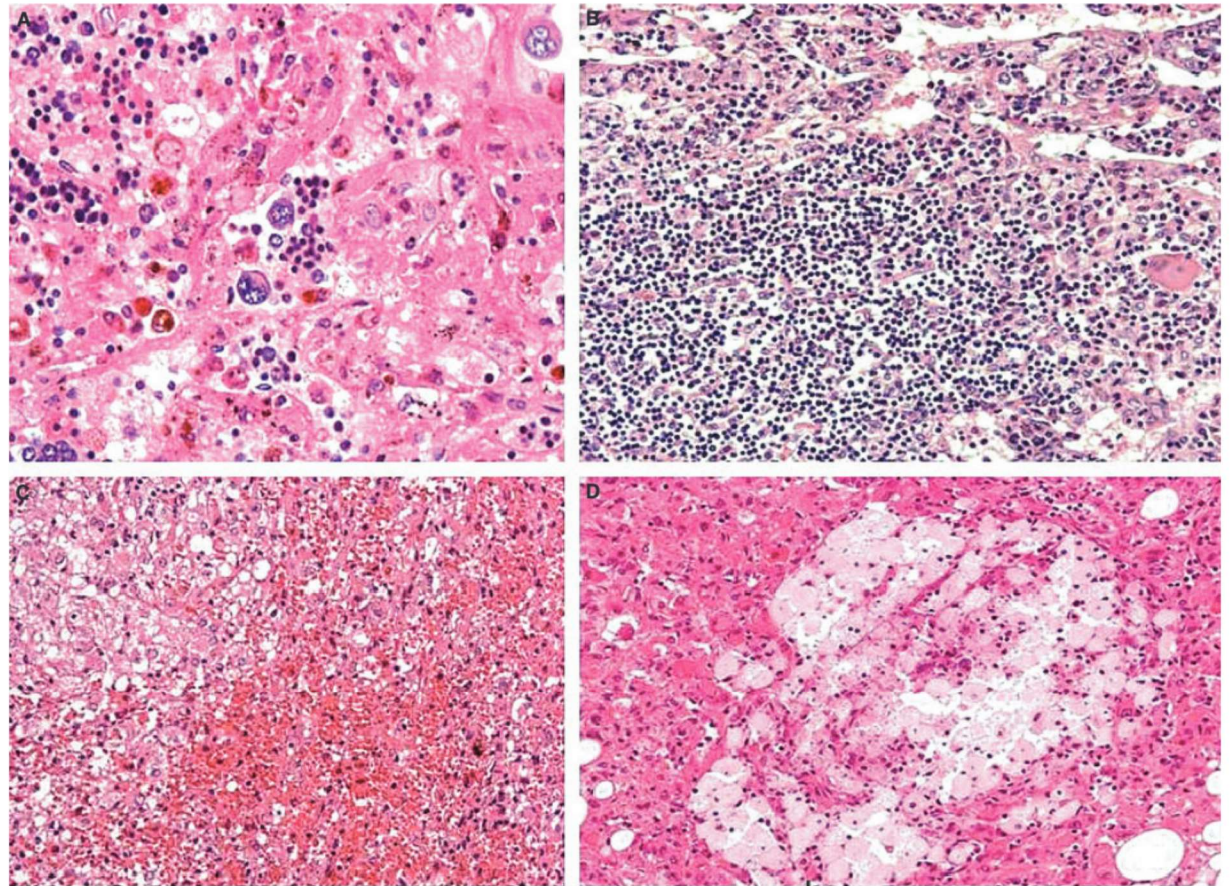


# Angiomyolipoma

## Microscopic aspects

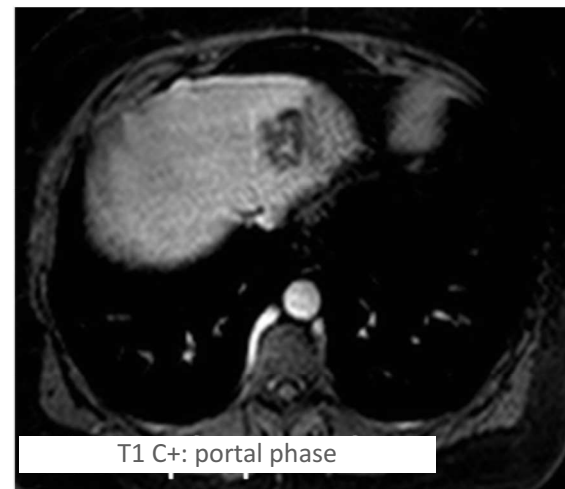
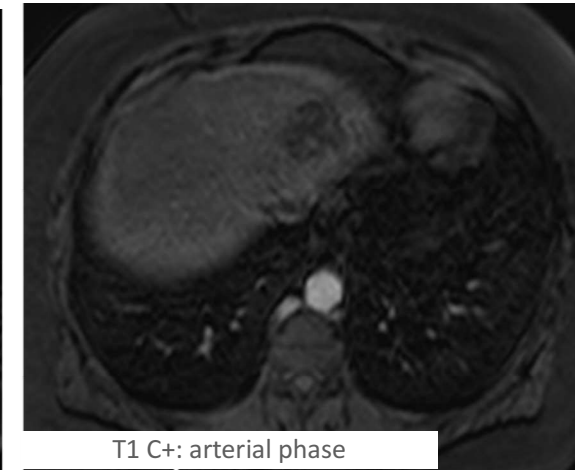
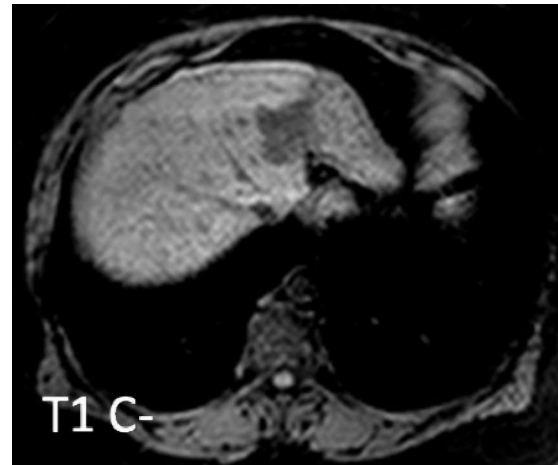
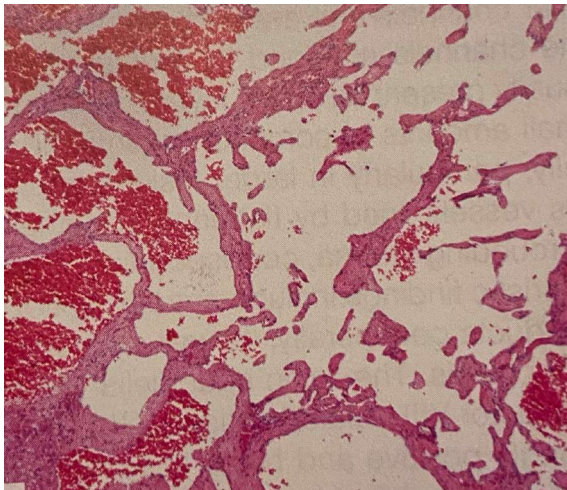
### Additional findings

- Extramedullary hematopoiesis
- Aggregates of lymphocytes
- Necrosis and/or hemorrhage
- Foamy macrophages



# Haemangioma

## Essential features



# Epitheloid haemangioendothelioma

## Essential features

- Rare **malignant vascular neoplasm** composed of epitheloid endothelial cells
- Often presents as multifocal disease (may be confused for metastatic disease)
- **WWTR1-CAMTA1 gene fusion** (most cases) = characteristic feature
- YAP1-TFE3 gene fusion (rare)



# Epitheloid haemangioendothelioma

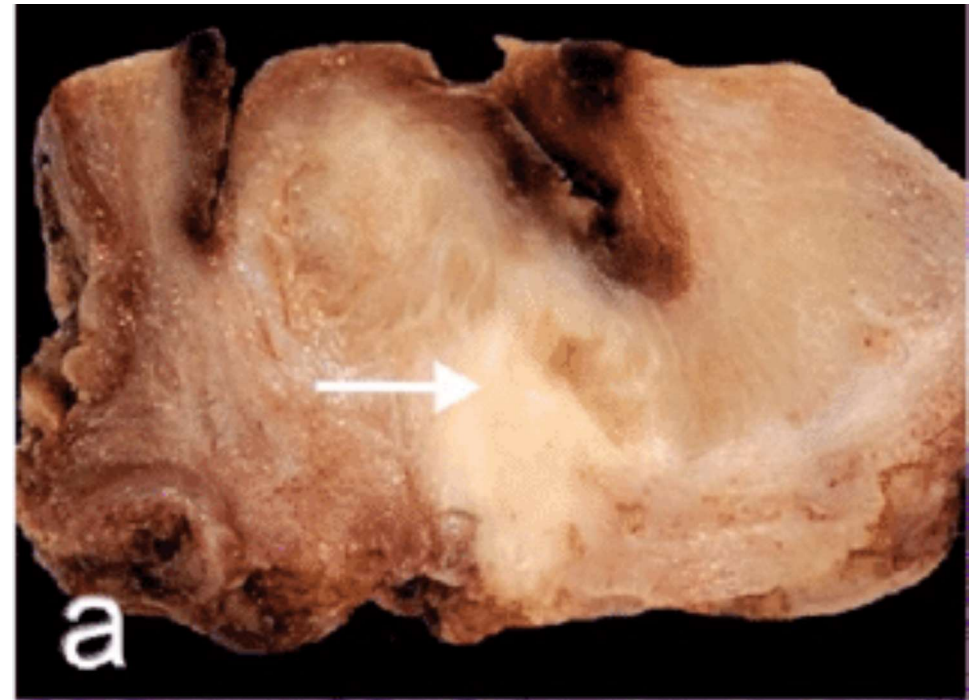
## Clinical aspects and pathophysiology

- May arise in the liver, lungs, bone or soft tissue
- **Rare** tumour
- Slight **female** predominance
- **Middle-aged** adults (children are rarely affected)
- Sporadic tumour

# Epithelioid haemangioendothelioma

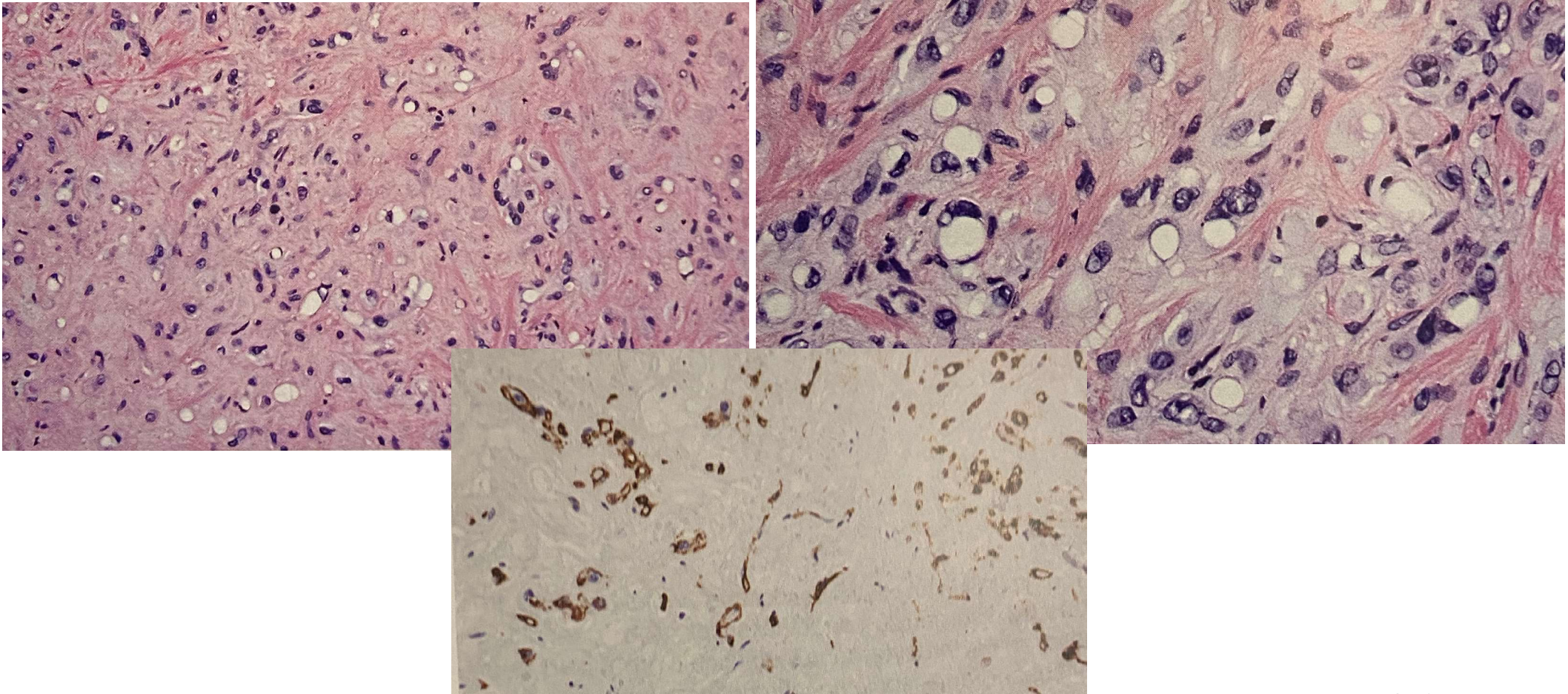
## Macroscopic aspects

- Size vary from small nodules to large coalescing masses > 10 cm
- The cut surface is typically white and firm



# Epithelioid haemangioendothelioma

## Microscopic aspects





# Conclusion

## Uncommon liver tumours

- Wide variety of lesions developing within the liver but with no hepatocytic or biliary origin
- Importance of clinical data:
  - Cirrhosis
  - Gender
  - Fever
- Importance of lesion characteristics: radiologist role!

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