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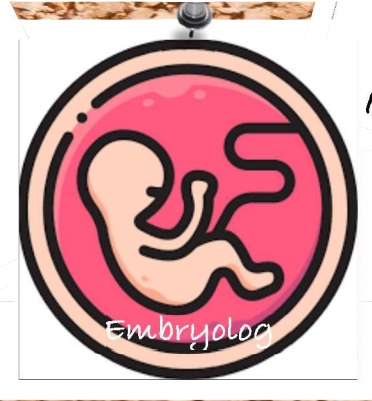
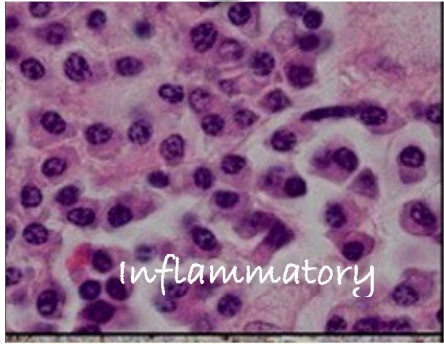
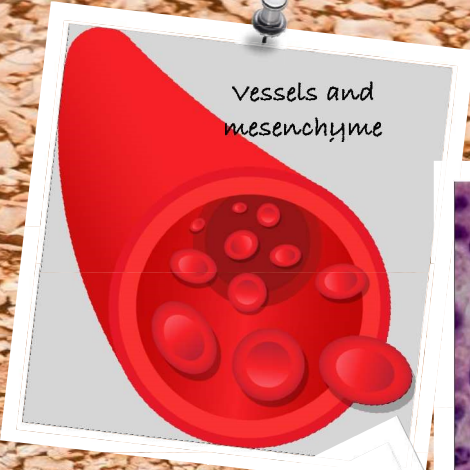


Uncommon tumours of the liver

Course on liver tumours

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

Uncommon liver tumours



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

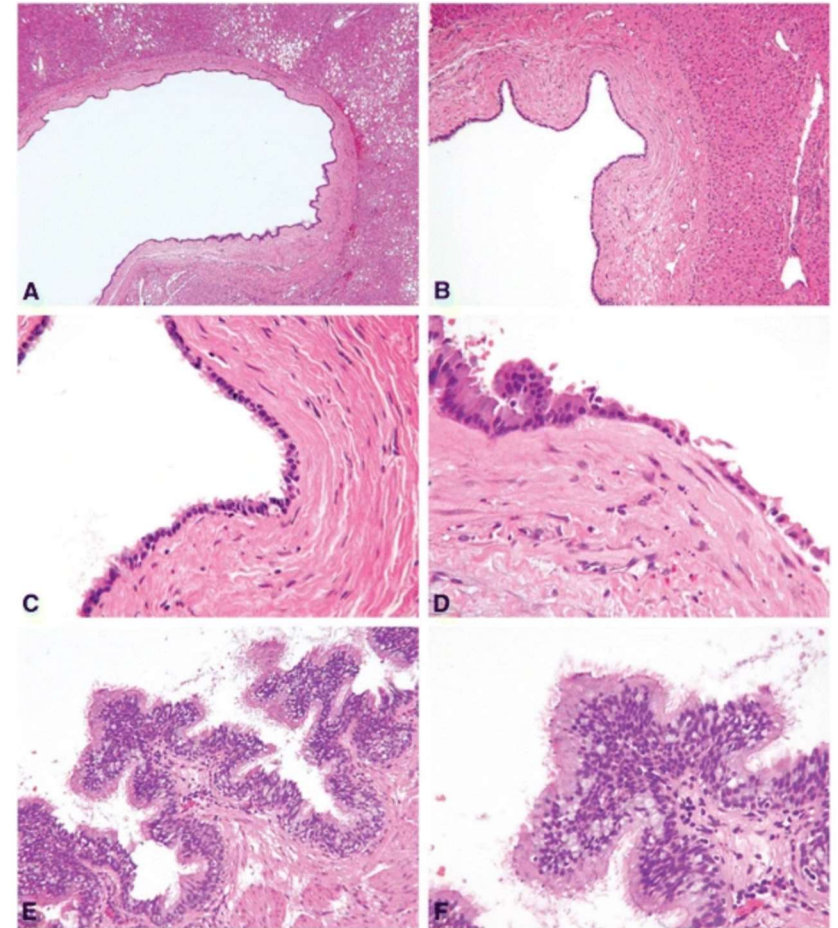
Clinical features

- 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



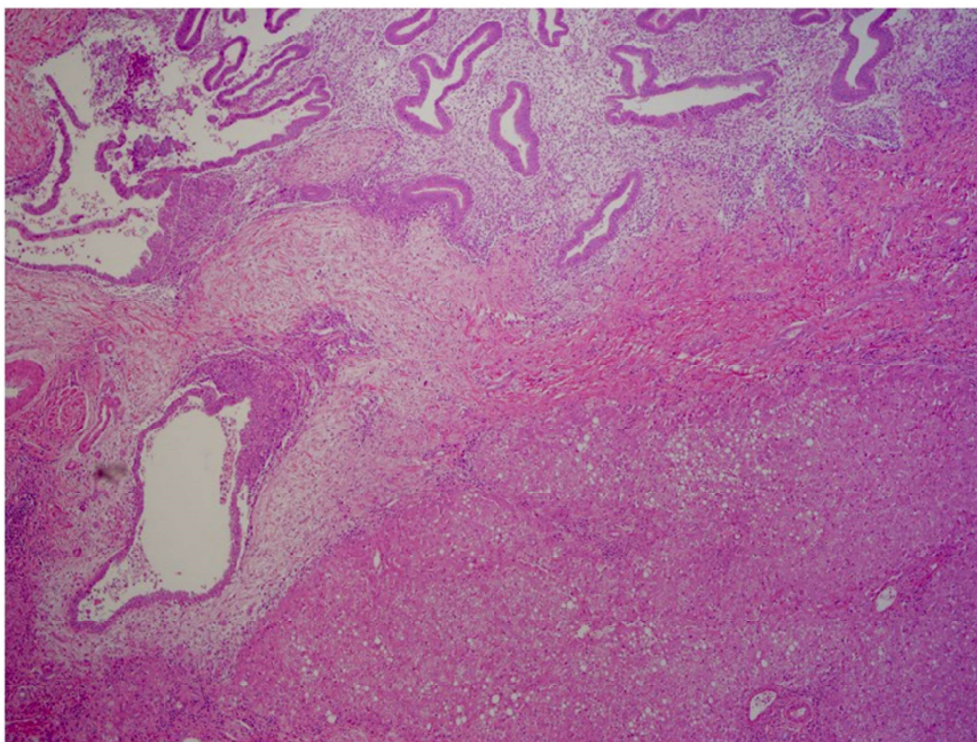
Endometriosis

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

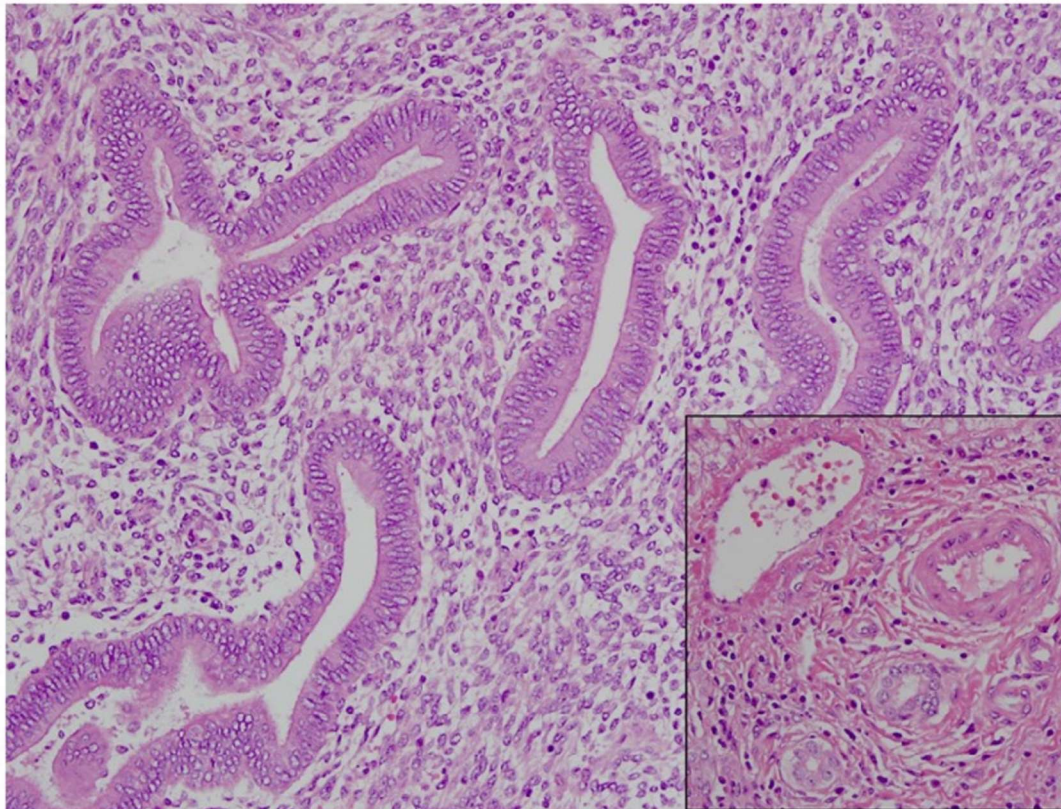
Endometriosis

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

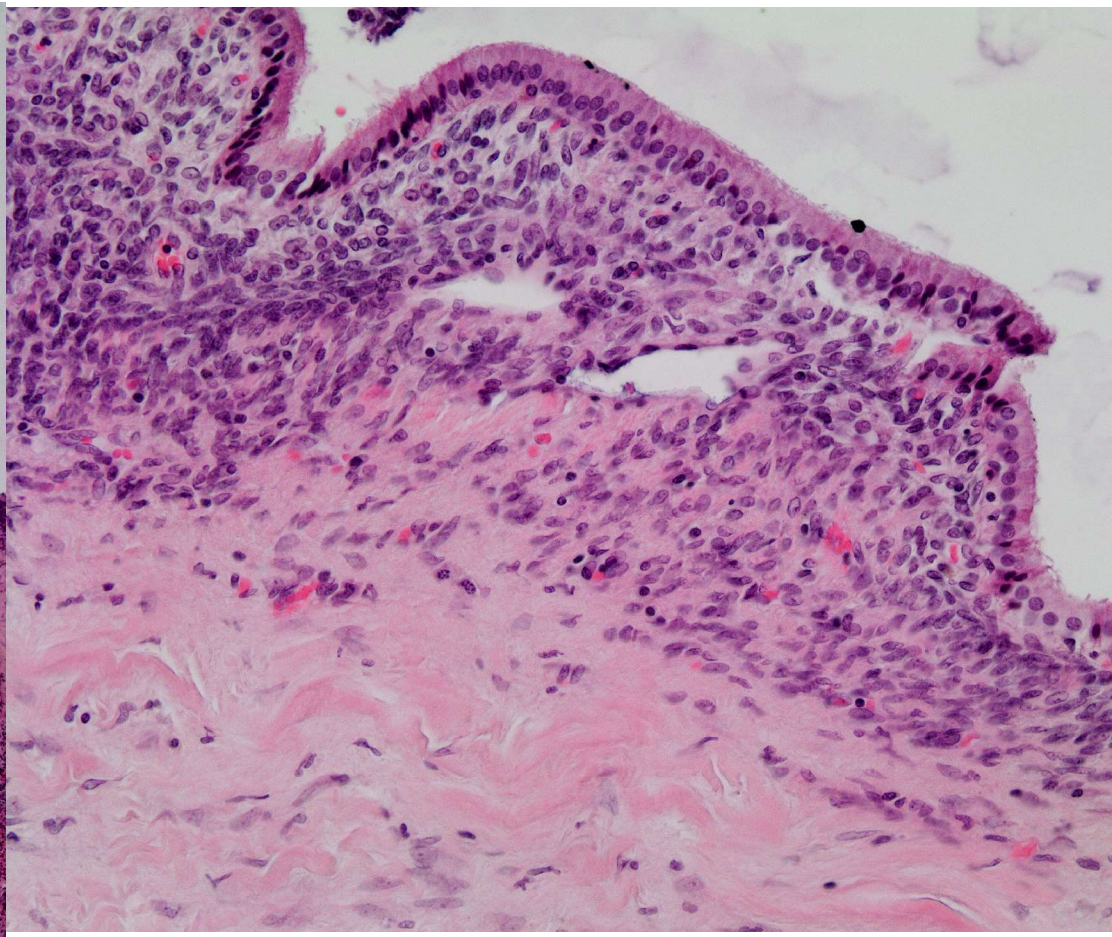
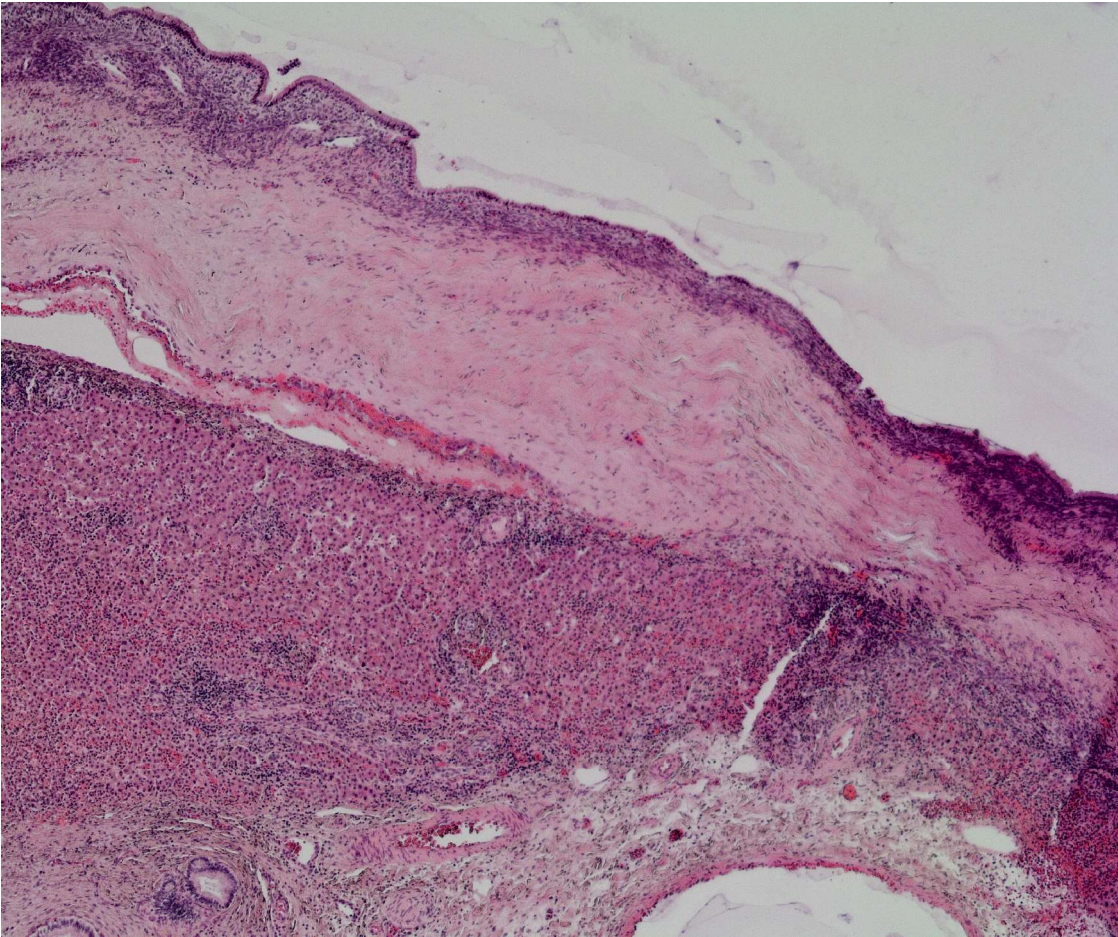
Macroscopic aspects

- 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

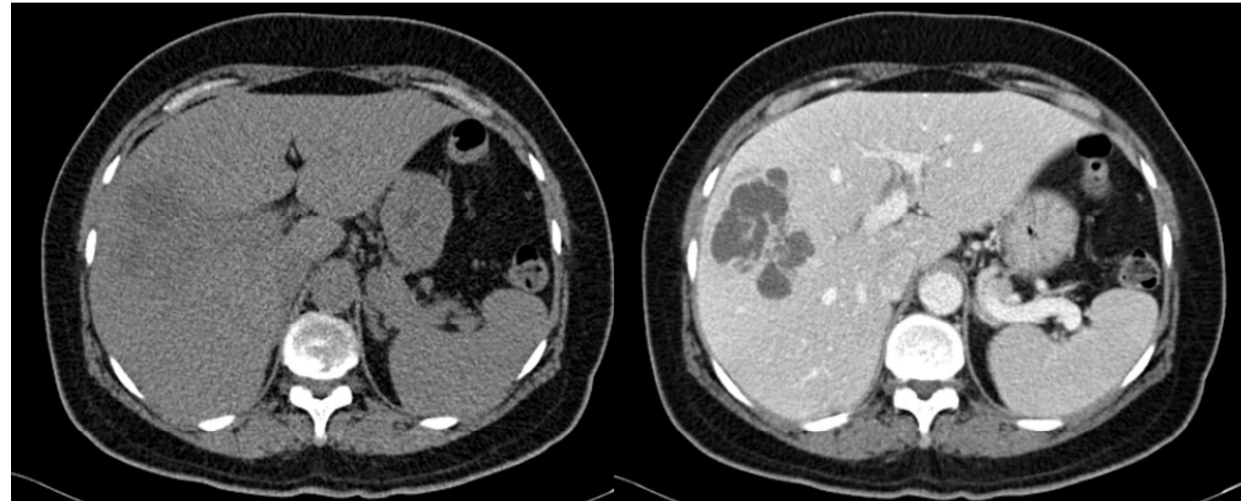
Staging and prognosis

- 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

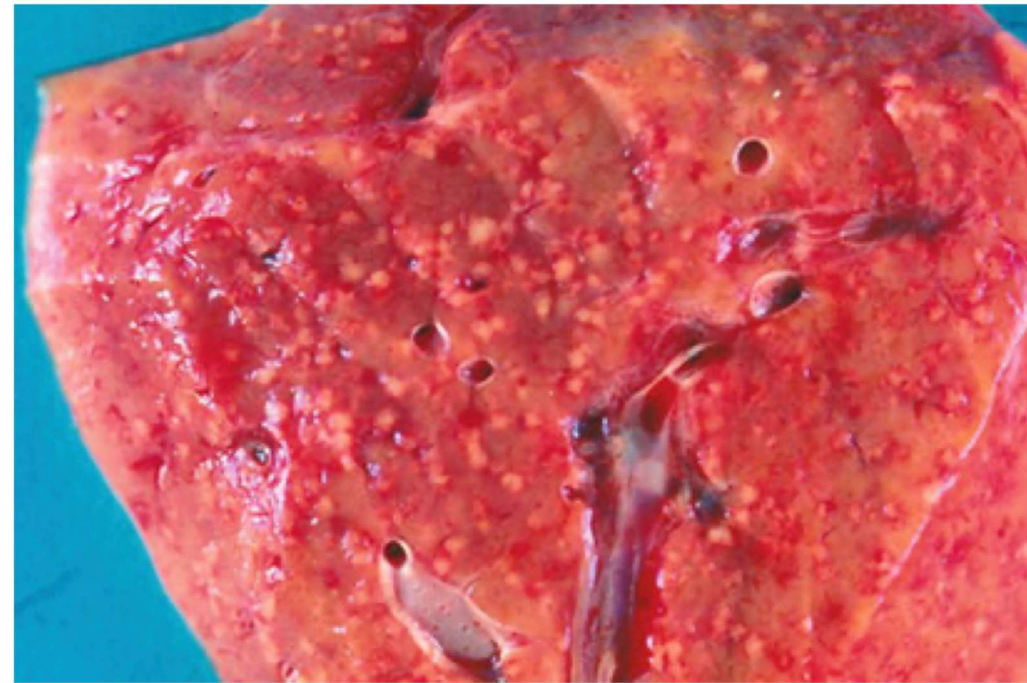
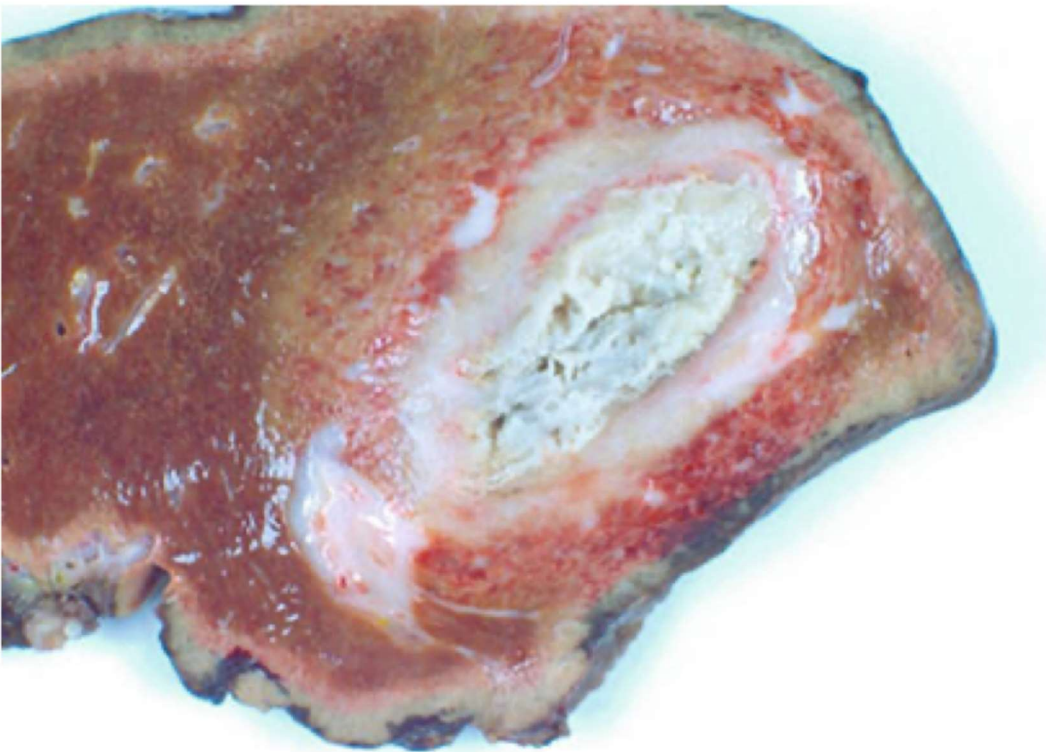


Radiological features:

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

Liver abscess

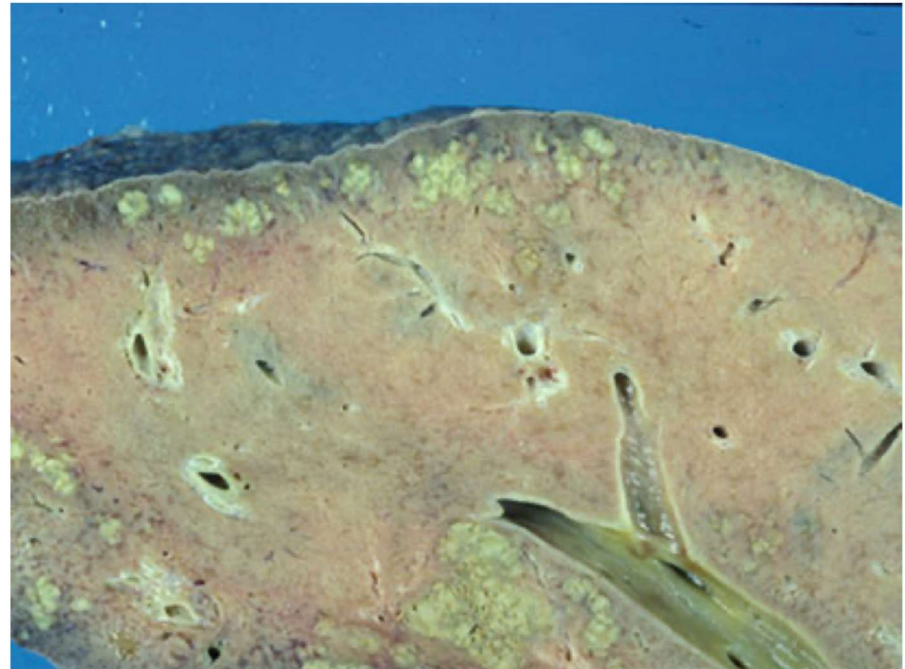
Macroscopic aspects



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

Liver abscess

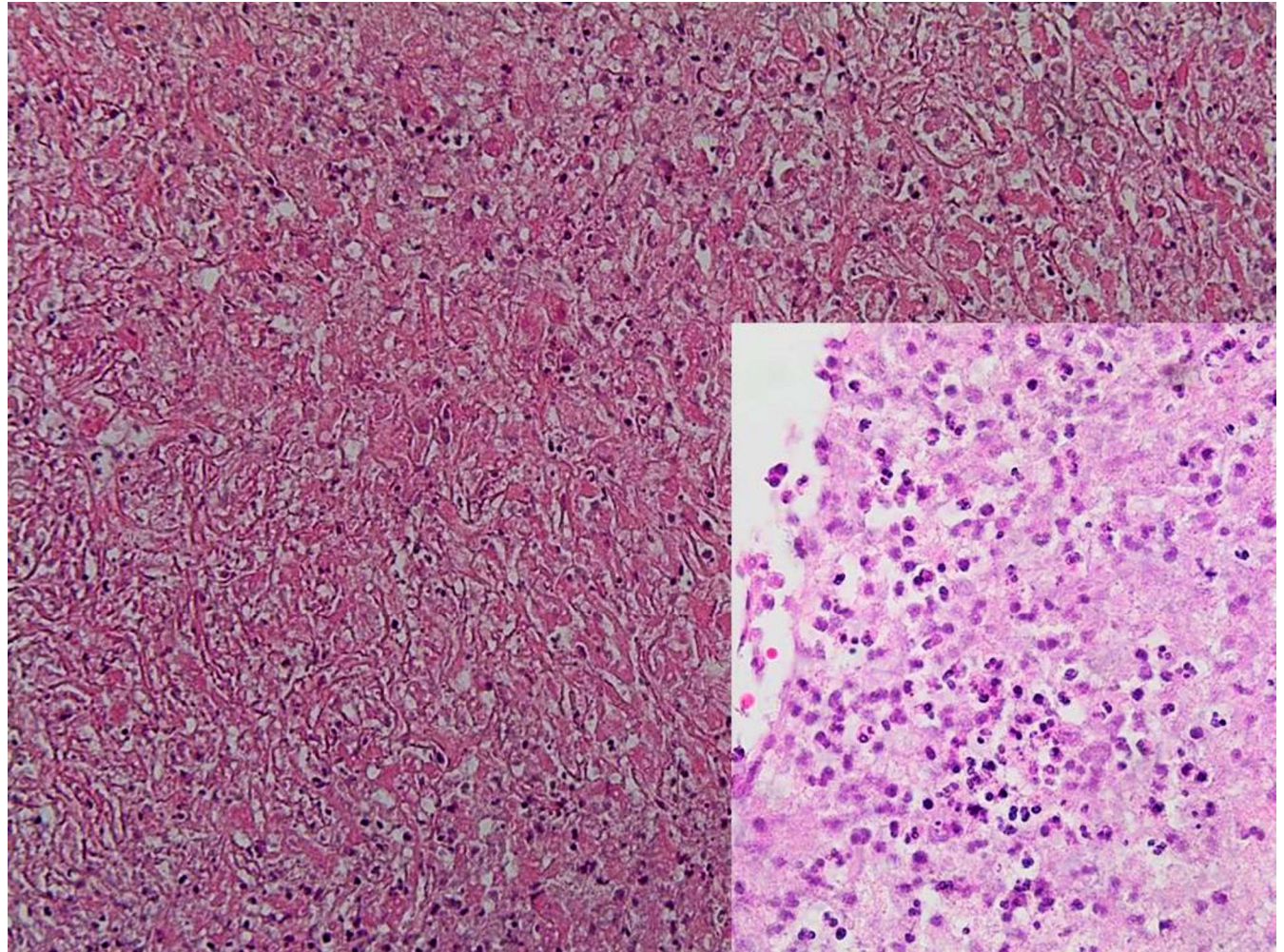
Macroscopic aspects



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Liver abscess

Microscopic aspects

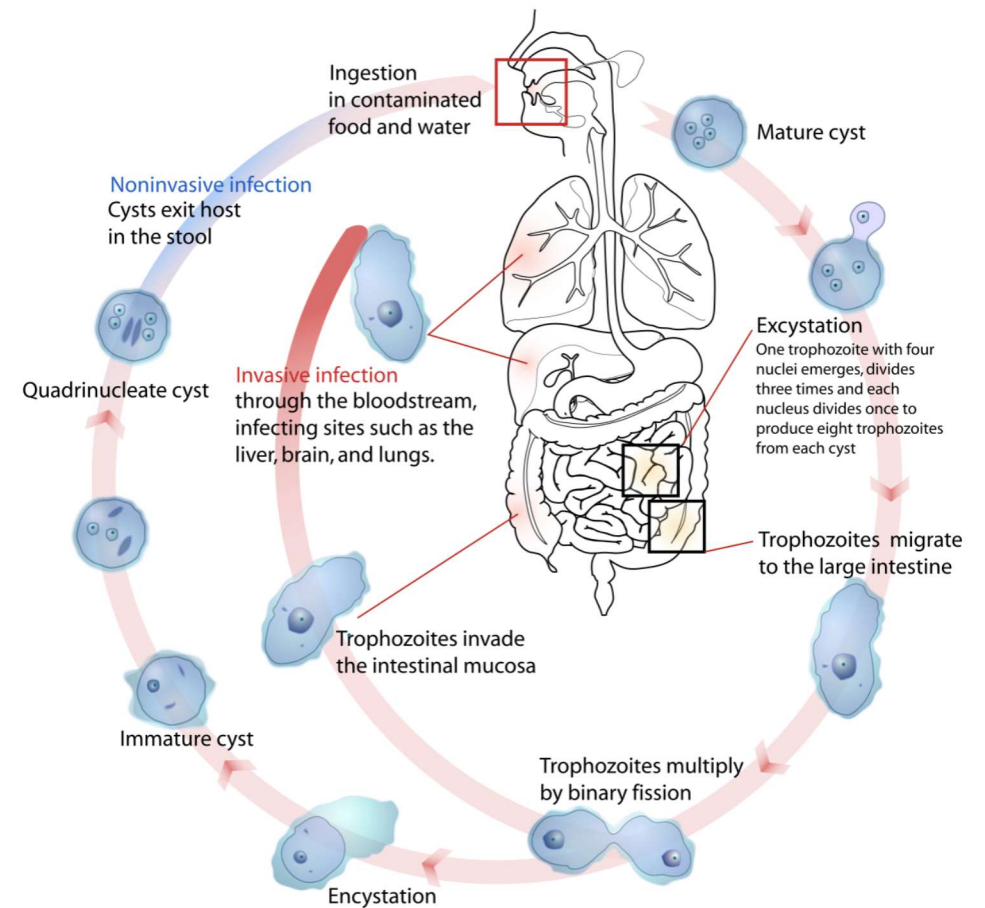
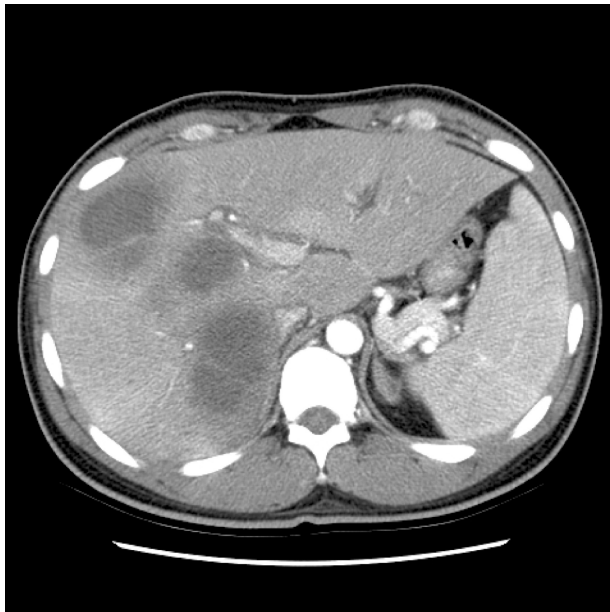


Liver abscess

Clinical features

- Entamoeba Histolytica (Amebic) abscess

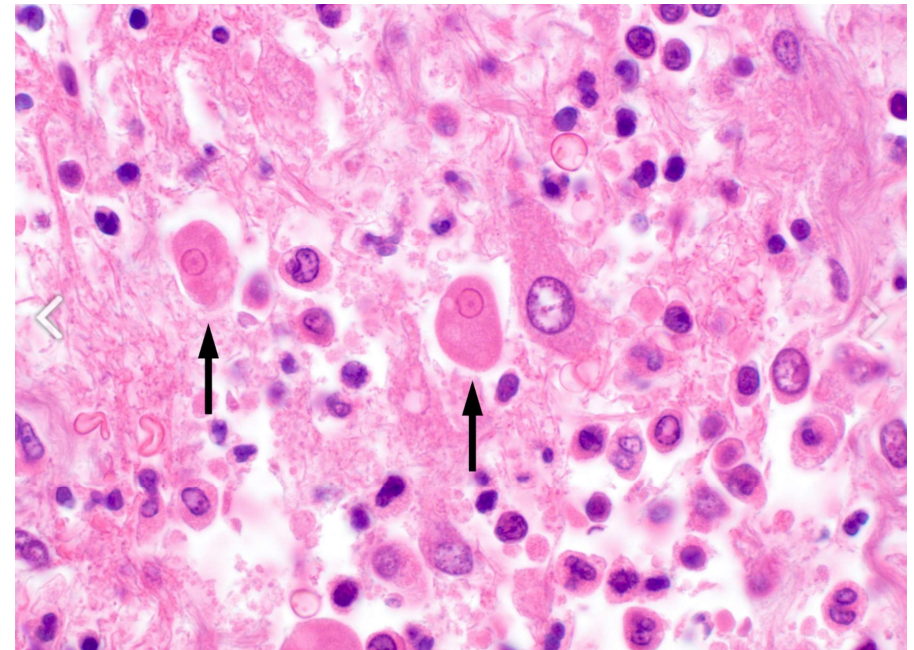
✓ Usually fecal oral spread



Liver abscess

Microscopic aspects

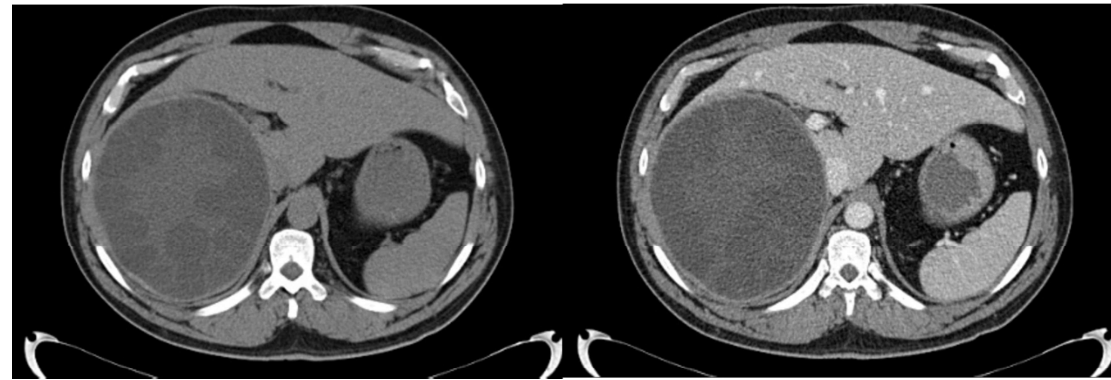
- Entamoeba Histolytica (Amebic) abscess
- ✓ Amebic trophozoites
 - Round nucleus with peripherally condensed ring of chromatin and central dot-like karyosome



Parasitic cyst

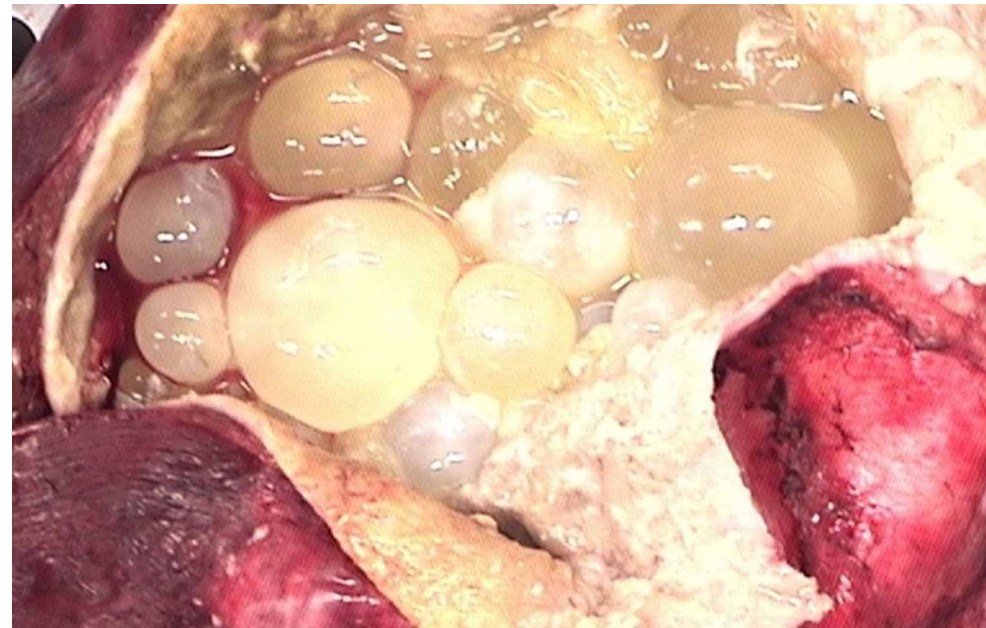
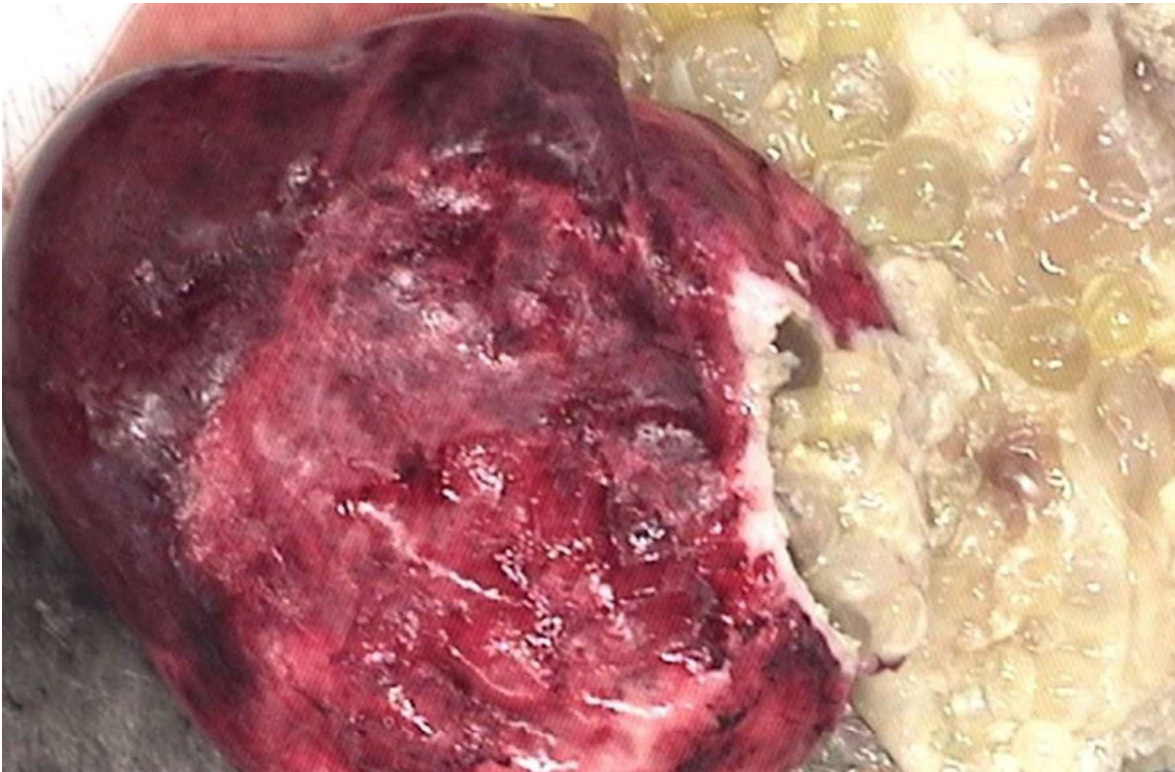
Clinical features

- **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*)



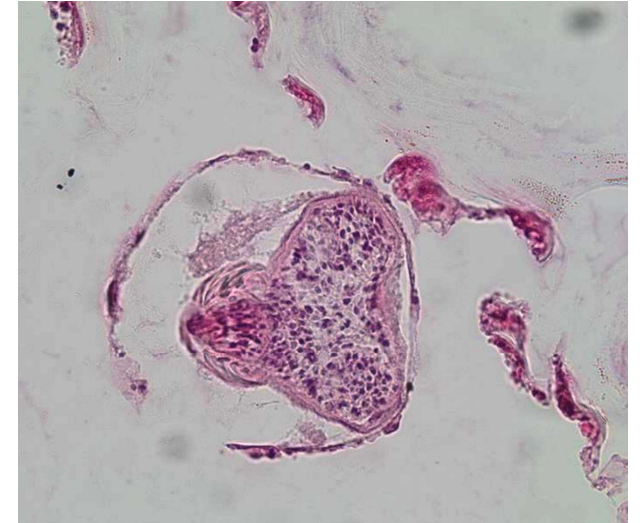
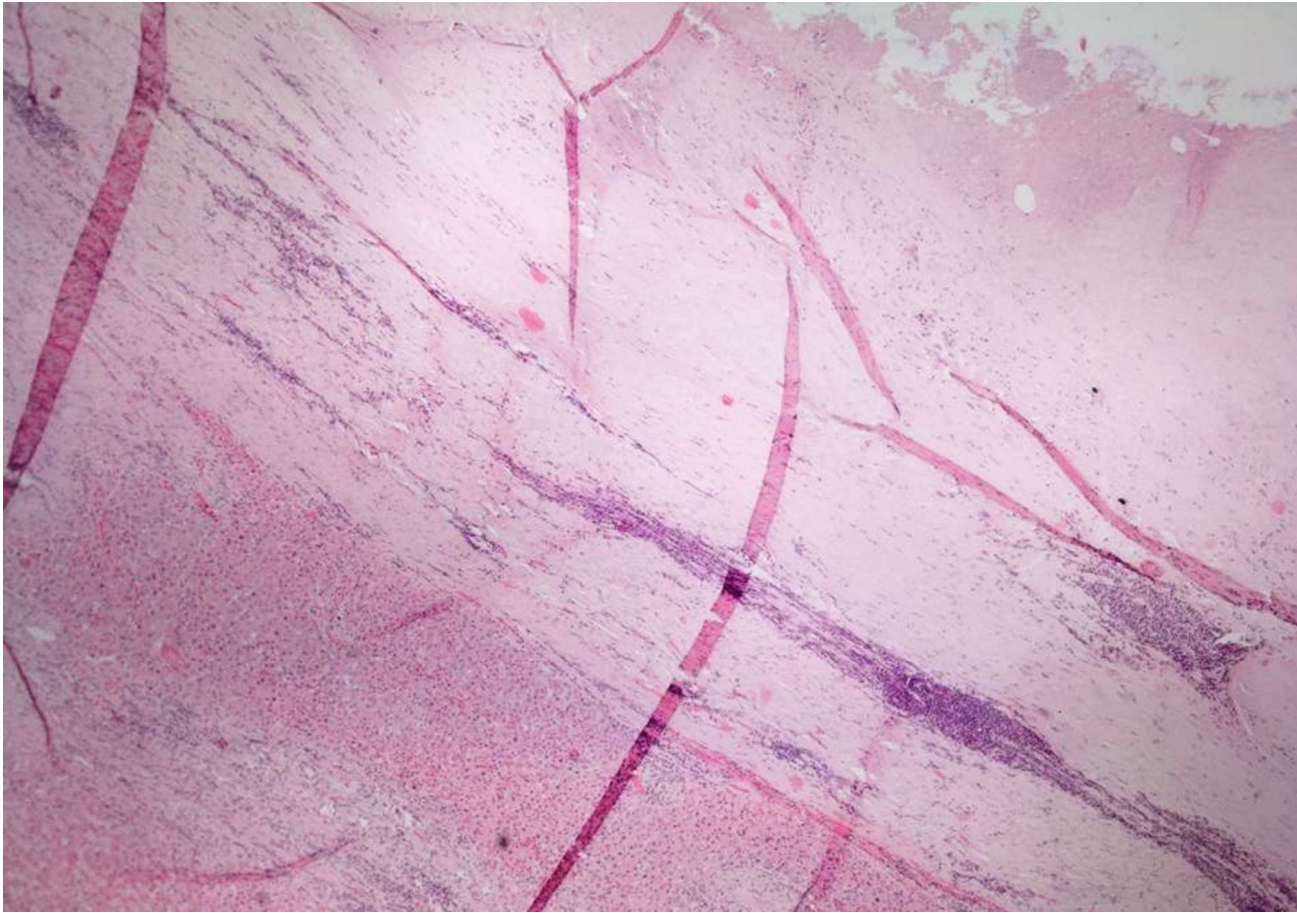
Parasitic cyst

Macroscopic aspects



Parasitic cyst

Microscopic aspects

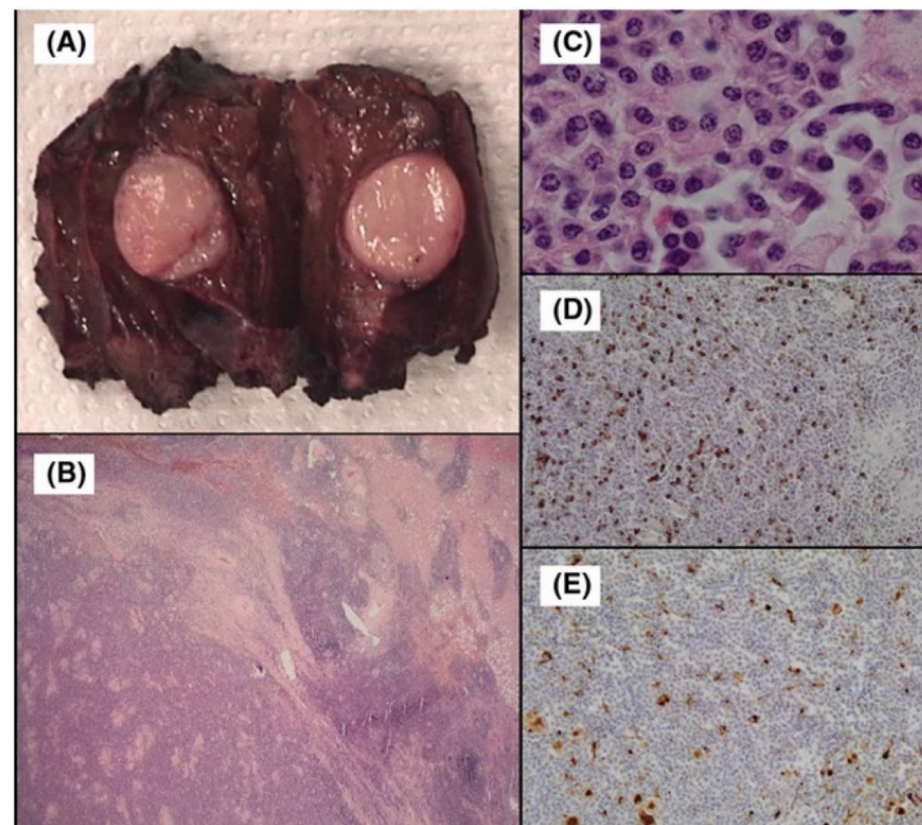


Protoscolex

IgG4 related inflammatory pseudotumor

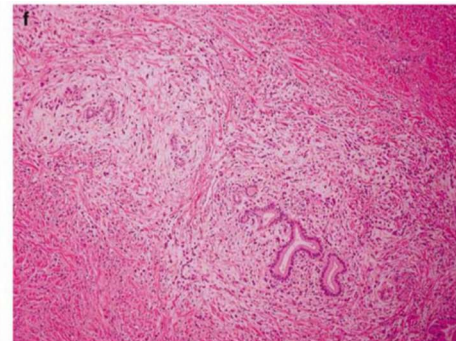
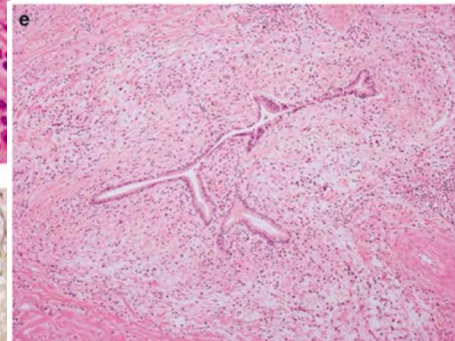
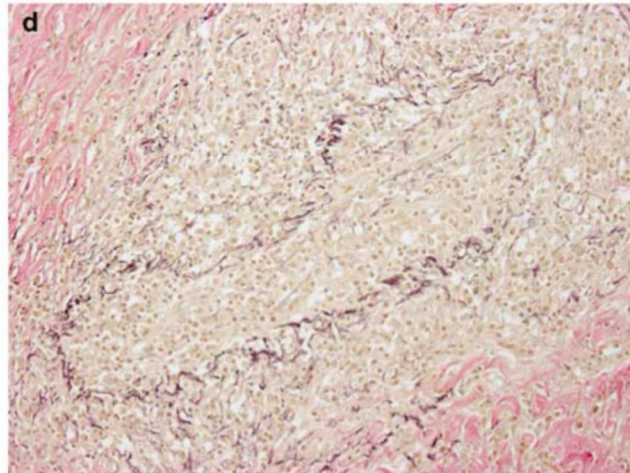
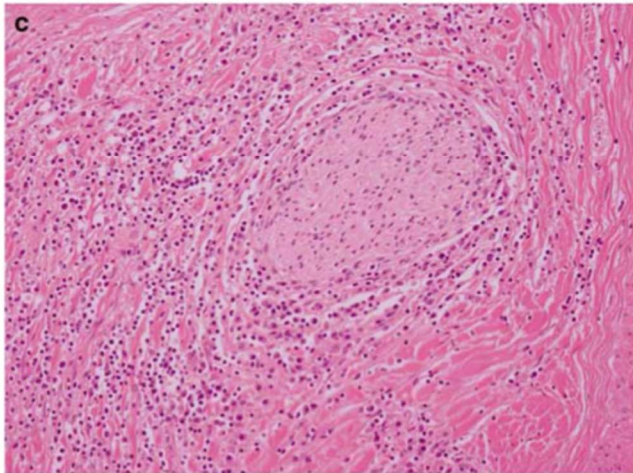
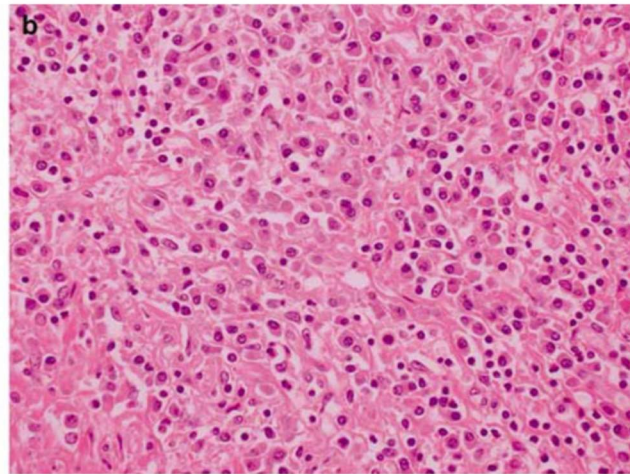
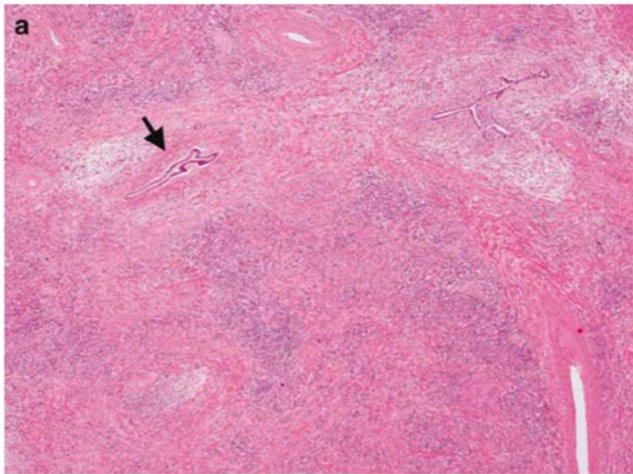
Clinical aspects

- 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



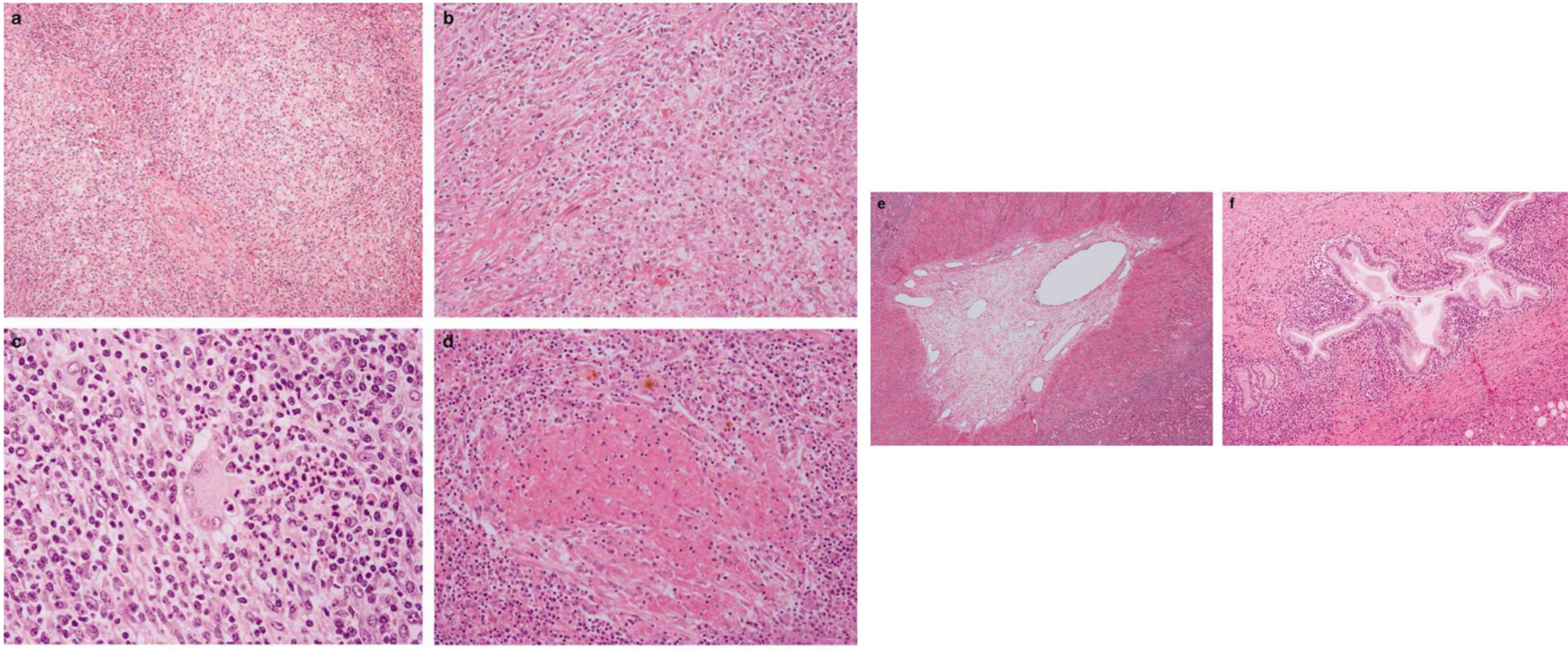
IgG4 related inflammatory pseudotumor

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)

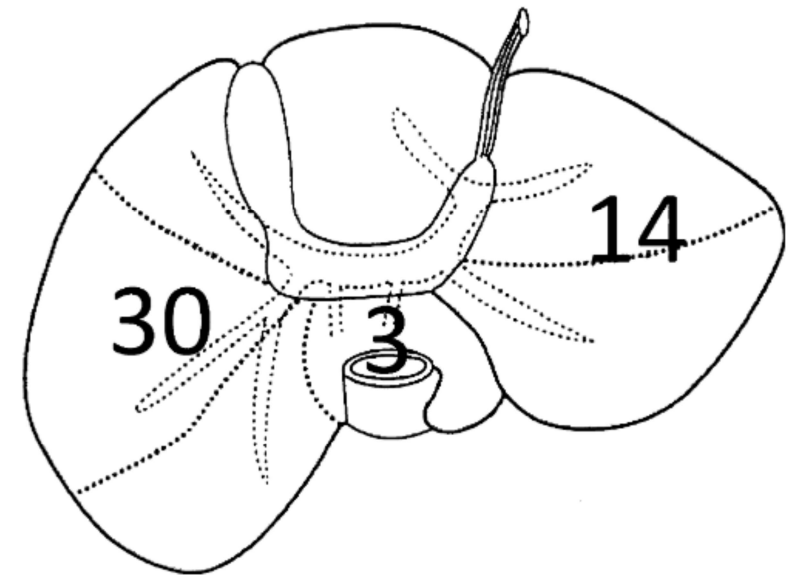


Figure 1. Localization of 47 hepatic angiomyolipomas.

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

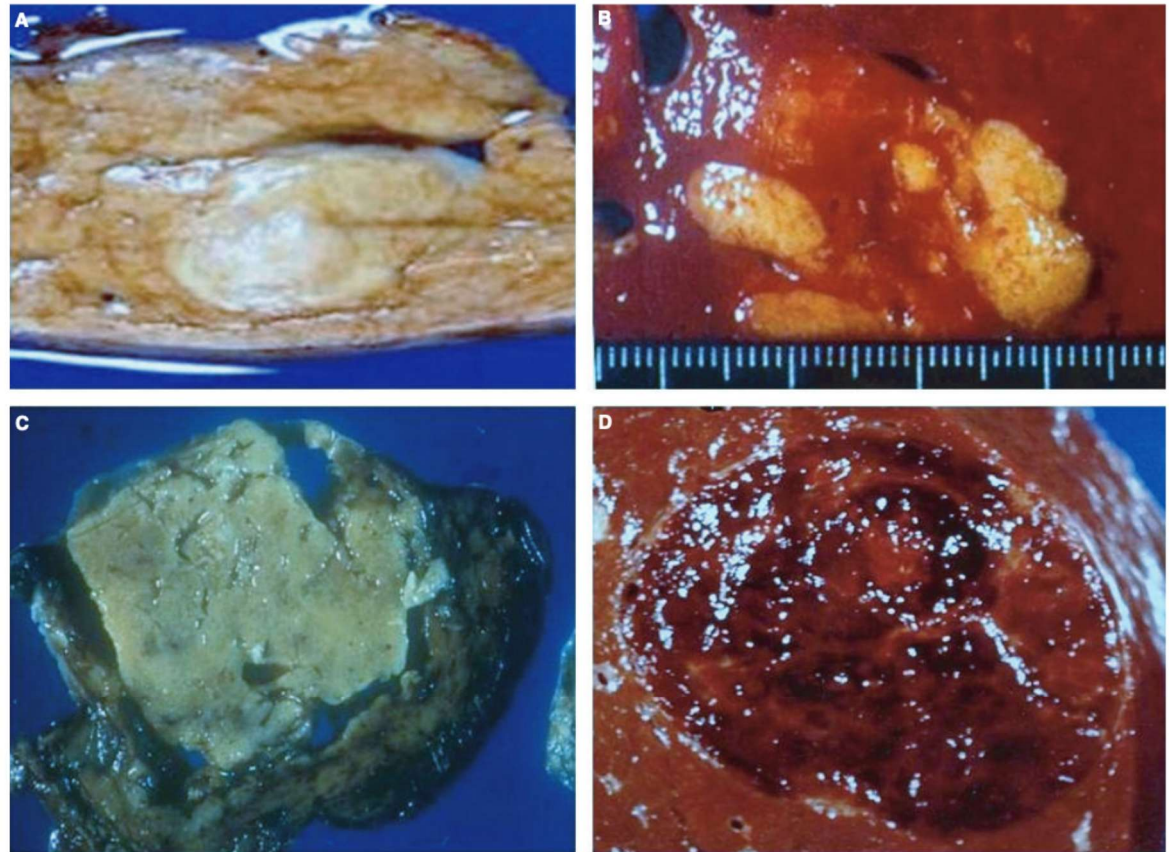


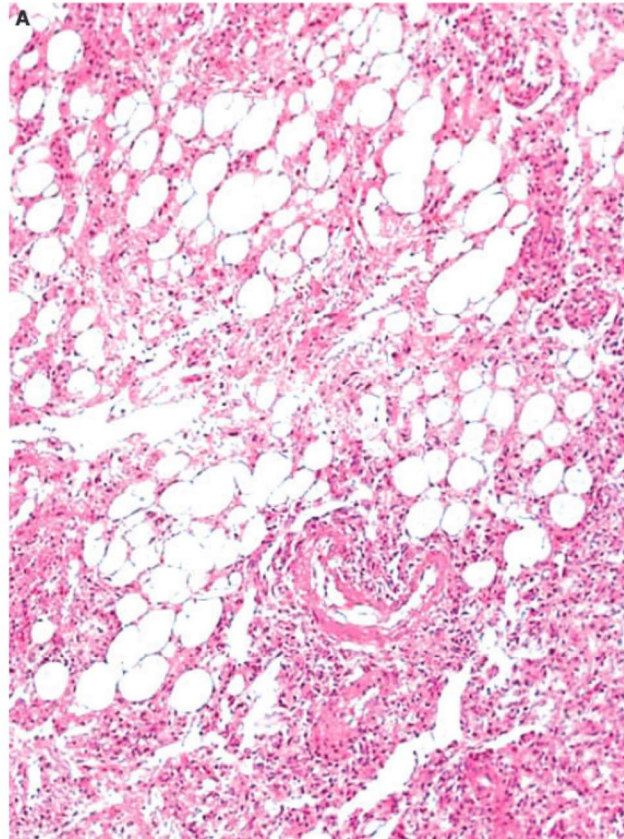
Figure 2. Macroscopic appearance of hepatic angiomyolipoma, showing myomatous (A), mixed (conventional) (B), lipomatous (C) and angiomyomatous (D) types.

Angiomyolipoma

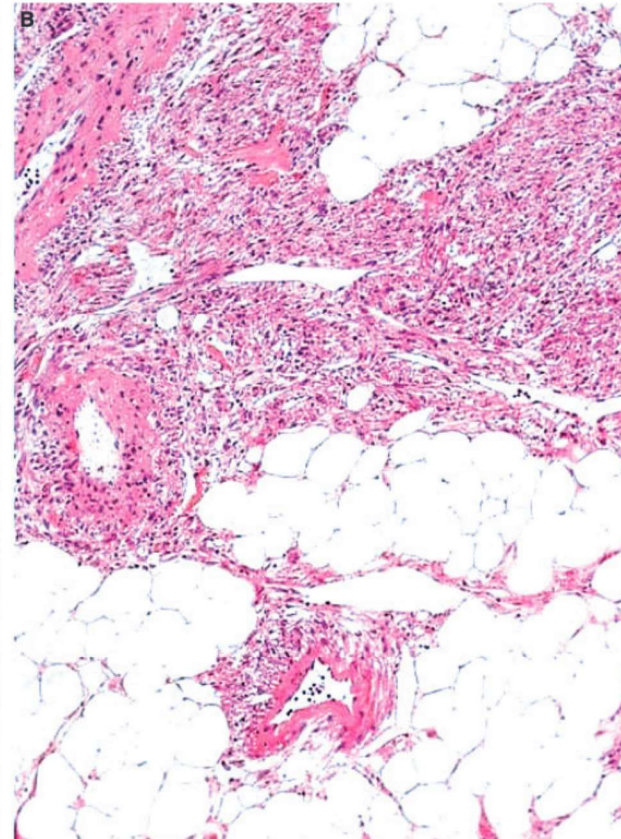
Microscopic aspects

Mixed
(conventional) type

Intermingled type



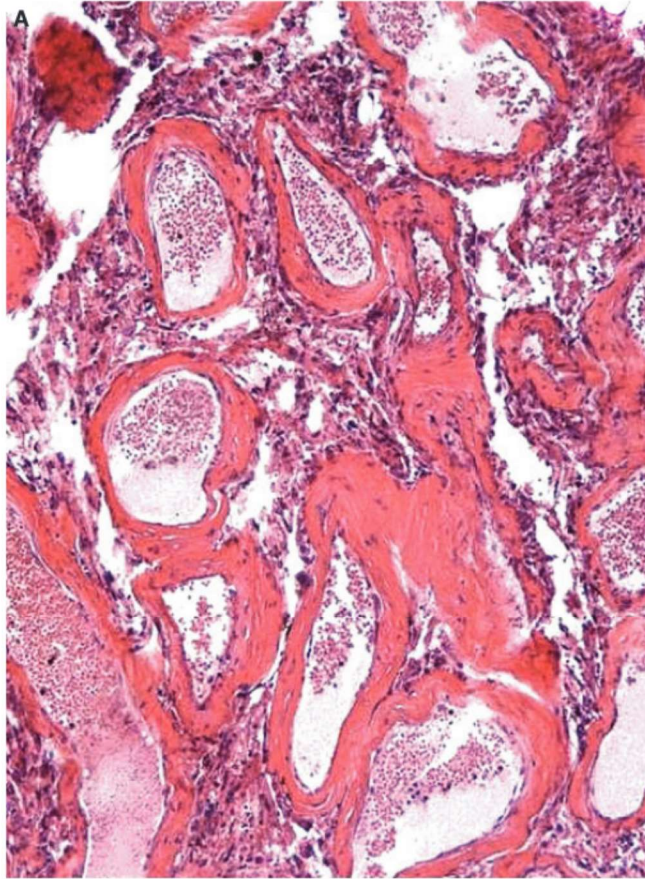
Mosaic
(geographical) type



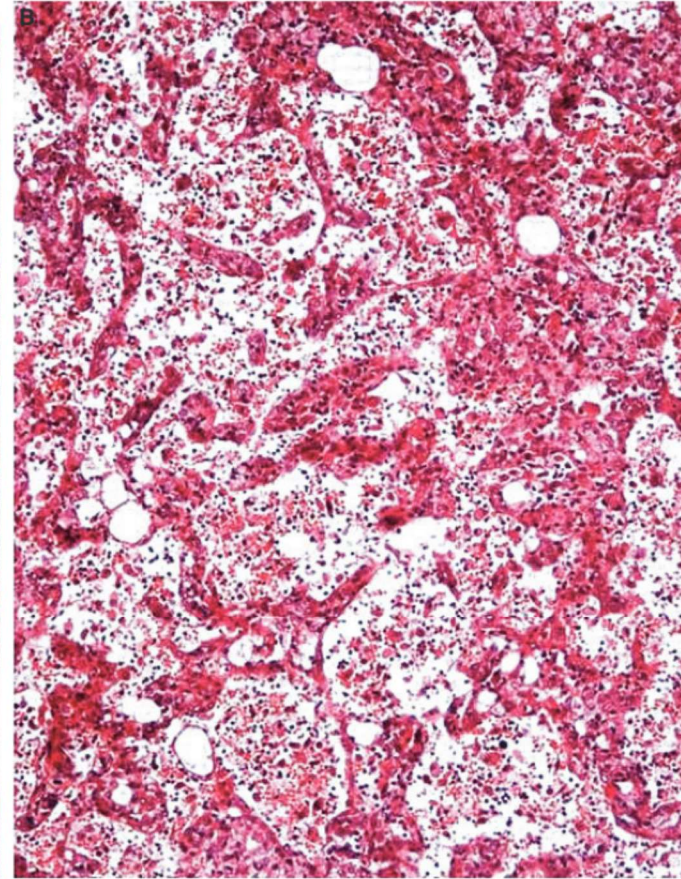
Angiomyolipoma

Microscopic aspects

Angiomatous blood vessels



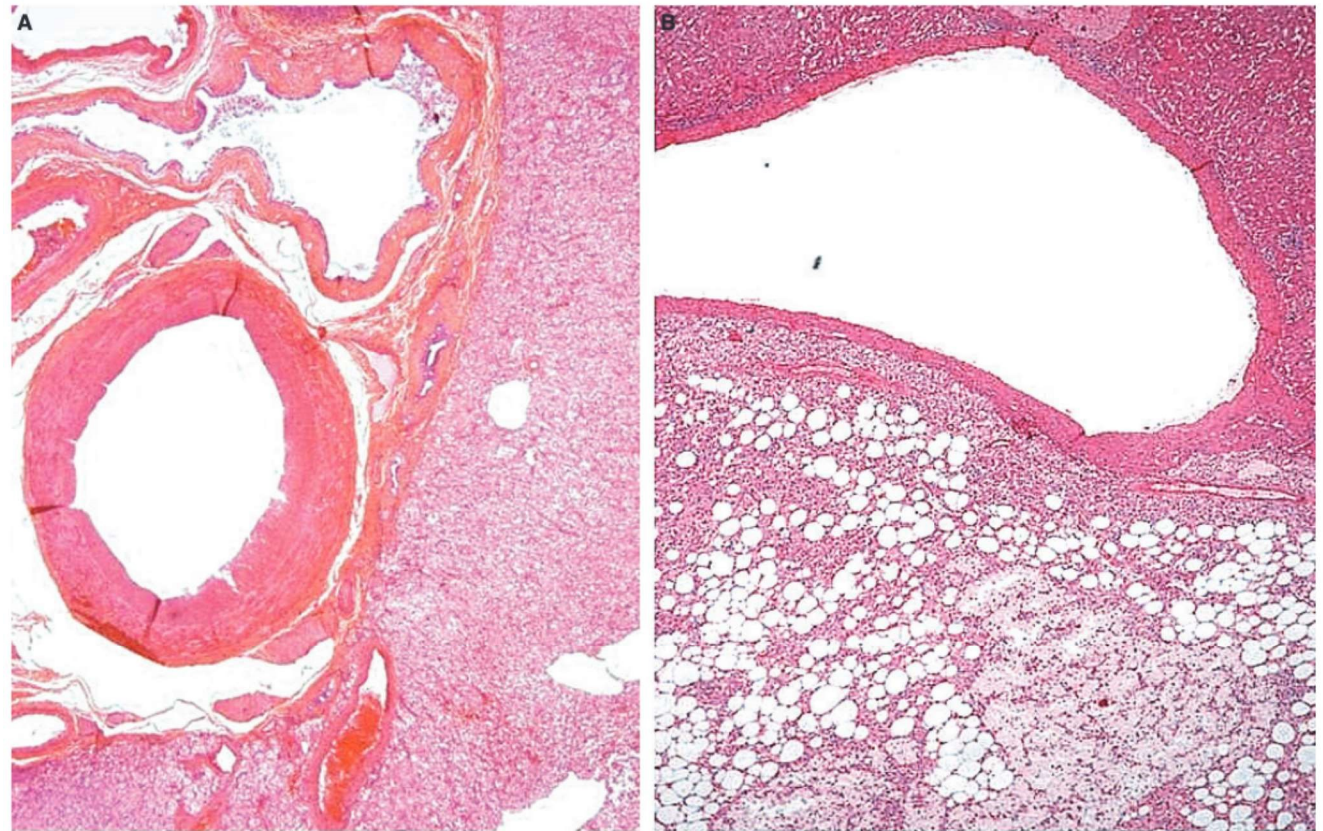
Dilated sinusoidal blood vessels



Angiomyolipoma

Microscopic aspects

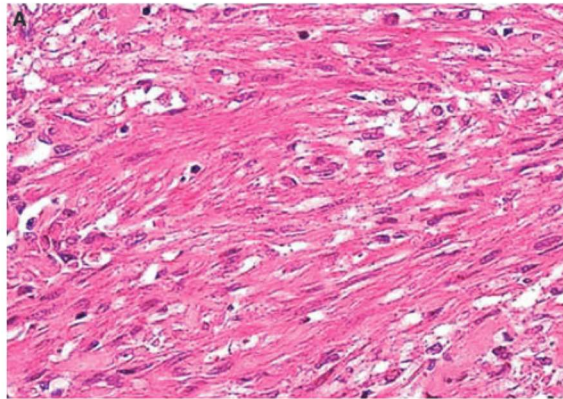
Large blood vessels could be observed in the adjacent liver parenchyma



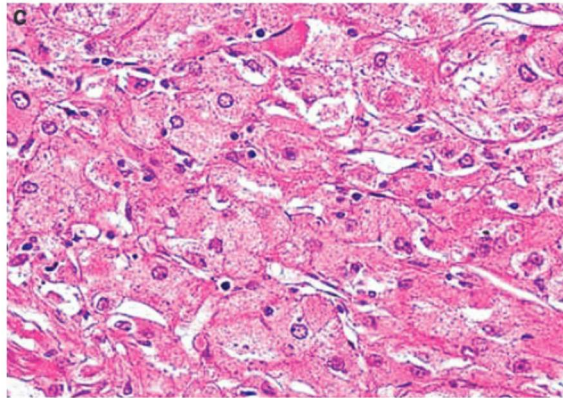
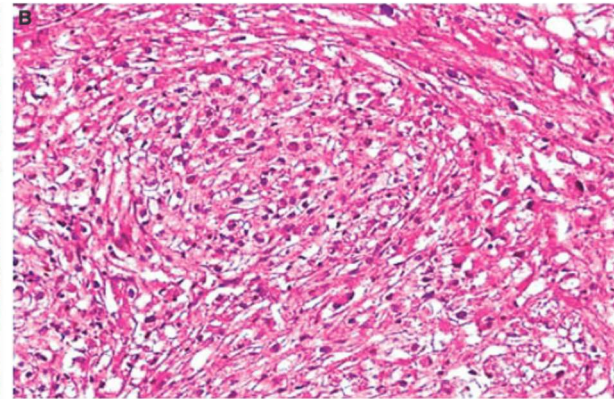
Angiomyolipoma

Microscopic aspects

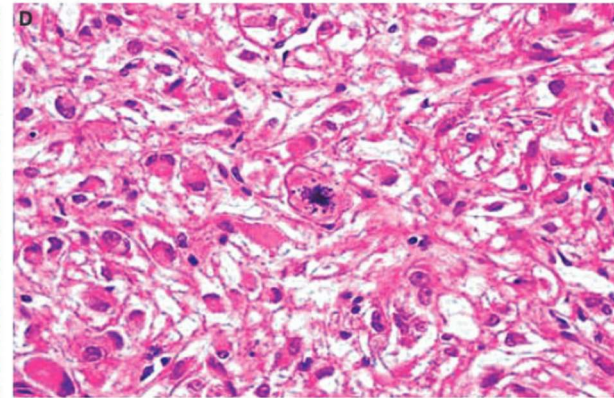
Spindle cells



Nodular or whorled pattern



Epithelioid



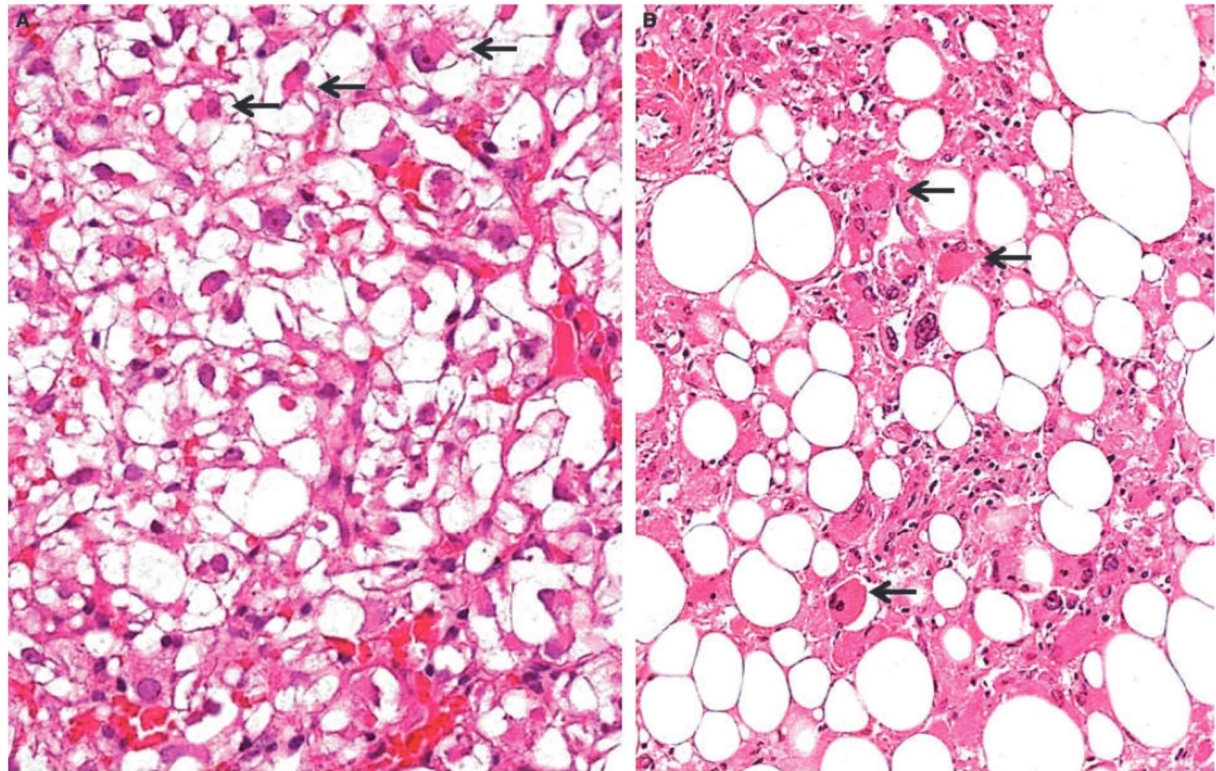
Pleiomorphic

Angiomyolipoma

Microscopic aspects

Epithelioid smooth muscle could display:

- Clear cell feature
- Rhabdoid feature
- Severe nuclear pleomorphism

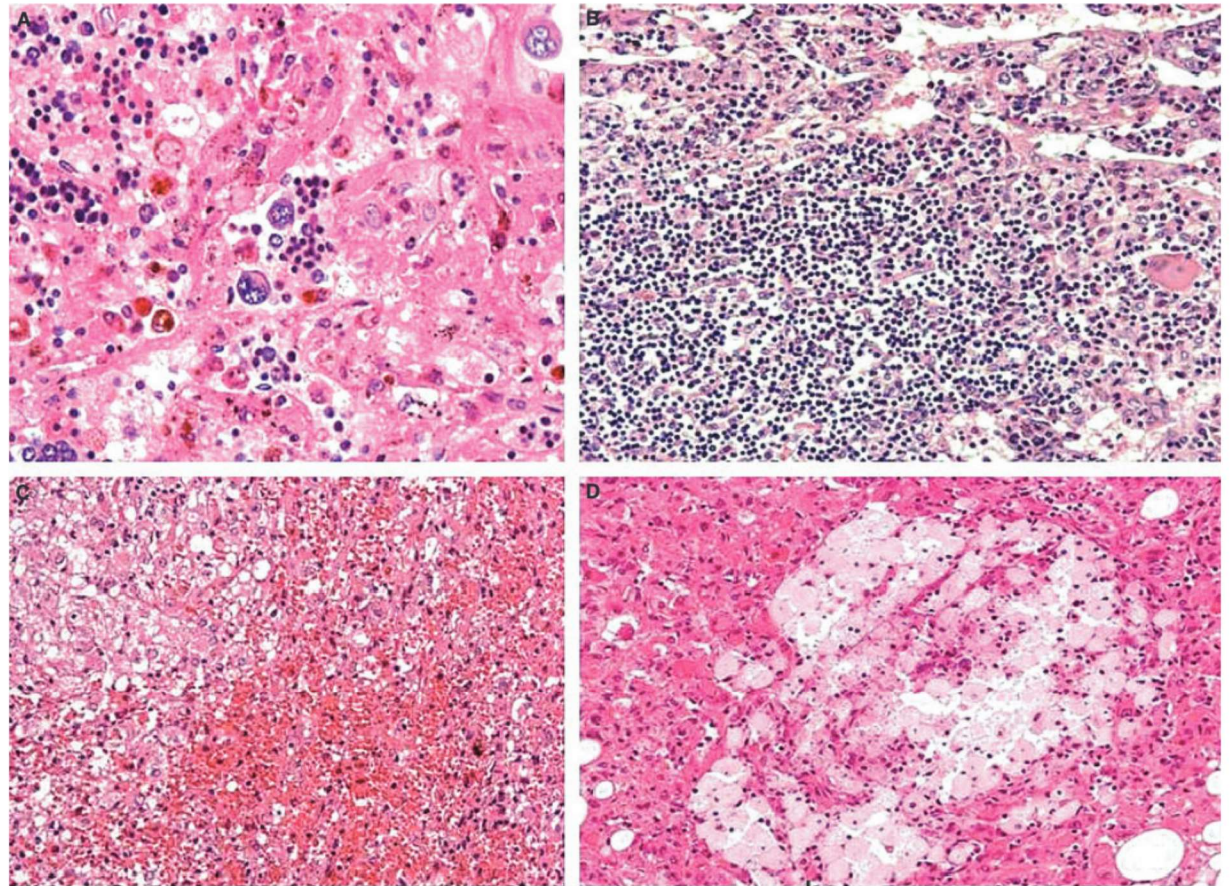


Angiomyolipoma

Microscopic aspects

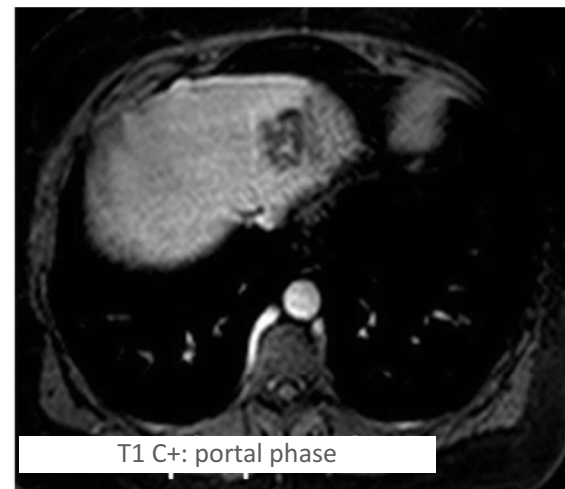
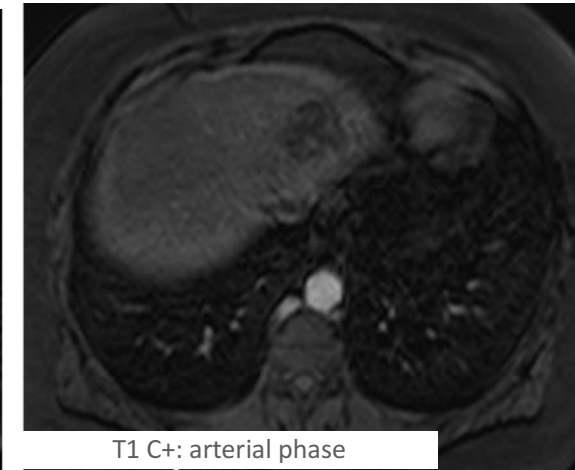
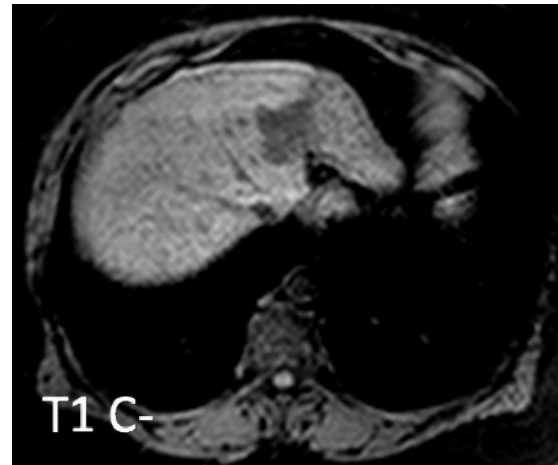
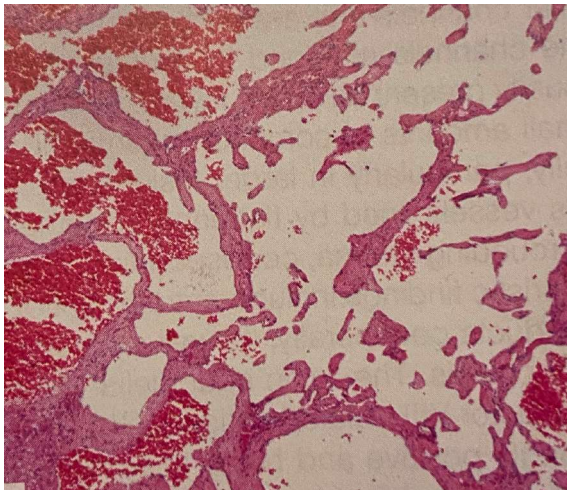
Additional findings

- 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

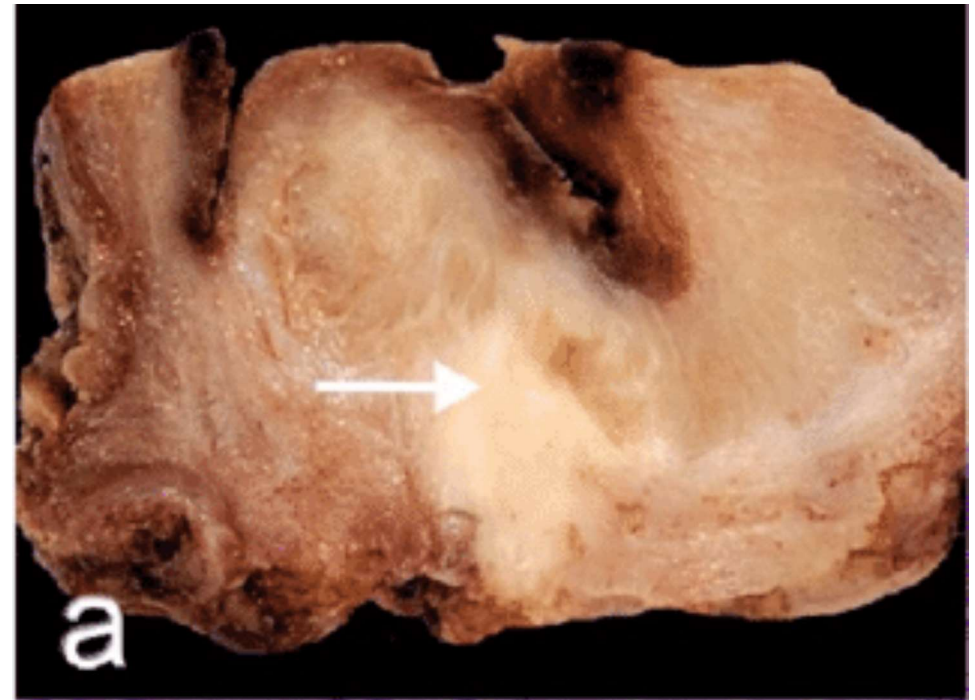
Clinical aspects and pathophysiology

- 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

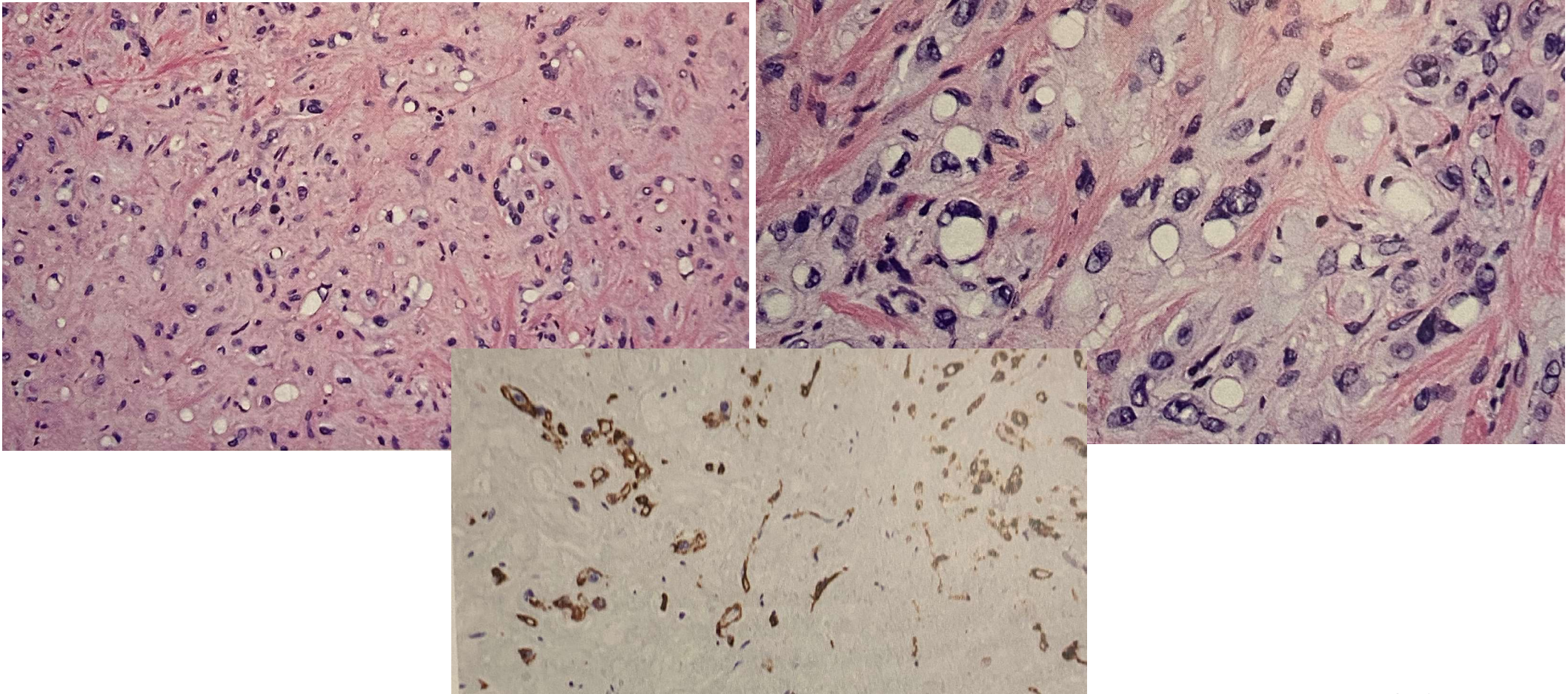
Macroscopic aspects

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



Epithelioid haemangiioendothelioma

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!

References

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