LUDWIG INSTITUTE FOR CANCER RESEARCH

Imperial College London

Case 2

Digestive Diseases Course
Bucharest 2016

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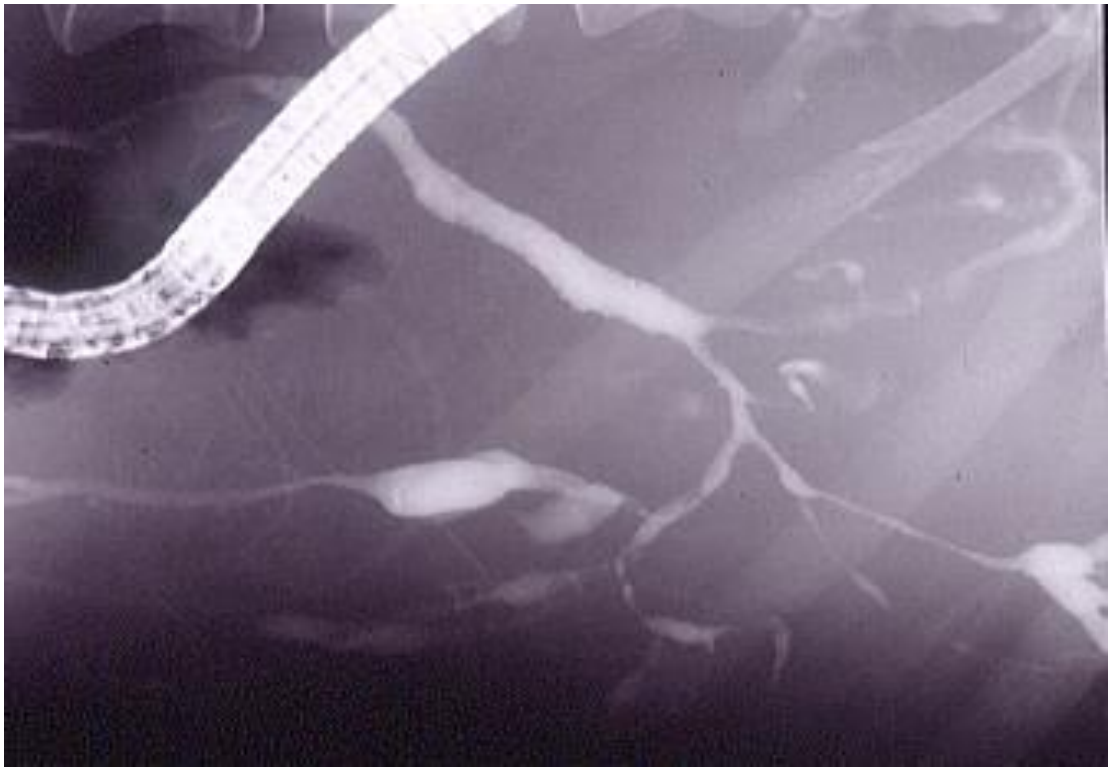
- 39 year old gay male
- Malaise, anorexia, weight loss and RUQ pain
- Pale stools and dark urine
- HIV positive for 10 months
- CD4 $0.042 \times 10^9/l$

- Jaundiced
- Hepato-splenomegaly
- Generalised lymphadenopathy

- Bilirubin 74umol/l
- ALT 239 IU/l
- Alk Phos 651 umol/l
- Hb 11.7g/dl
- WCC 4.2 x 10^9/l
- Platelets 169 x10^9/l

- US: no intrahepatic bile duct dilatation, extrahepatic bile duct 1cm in diameter
- Lymph node biopsy: reactive changes only
- CT: dilated bile ducts, enlarged abdominal lymph nodes, splenomegaly
- ERCP: “irregular beaded biliary outline and multiple, intrahepatic and extrahepatic strictures”



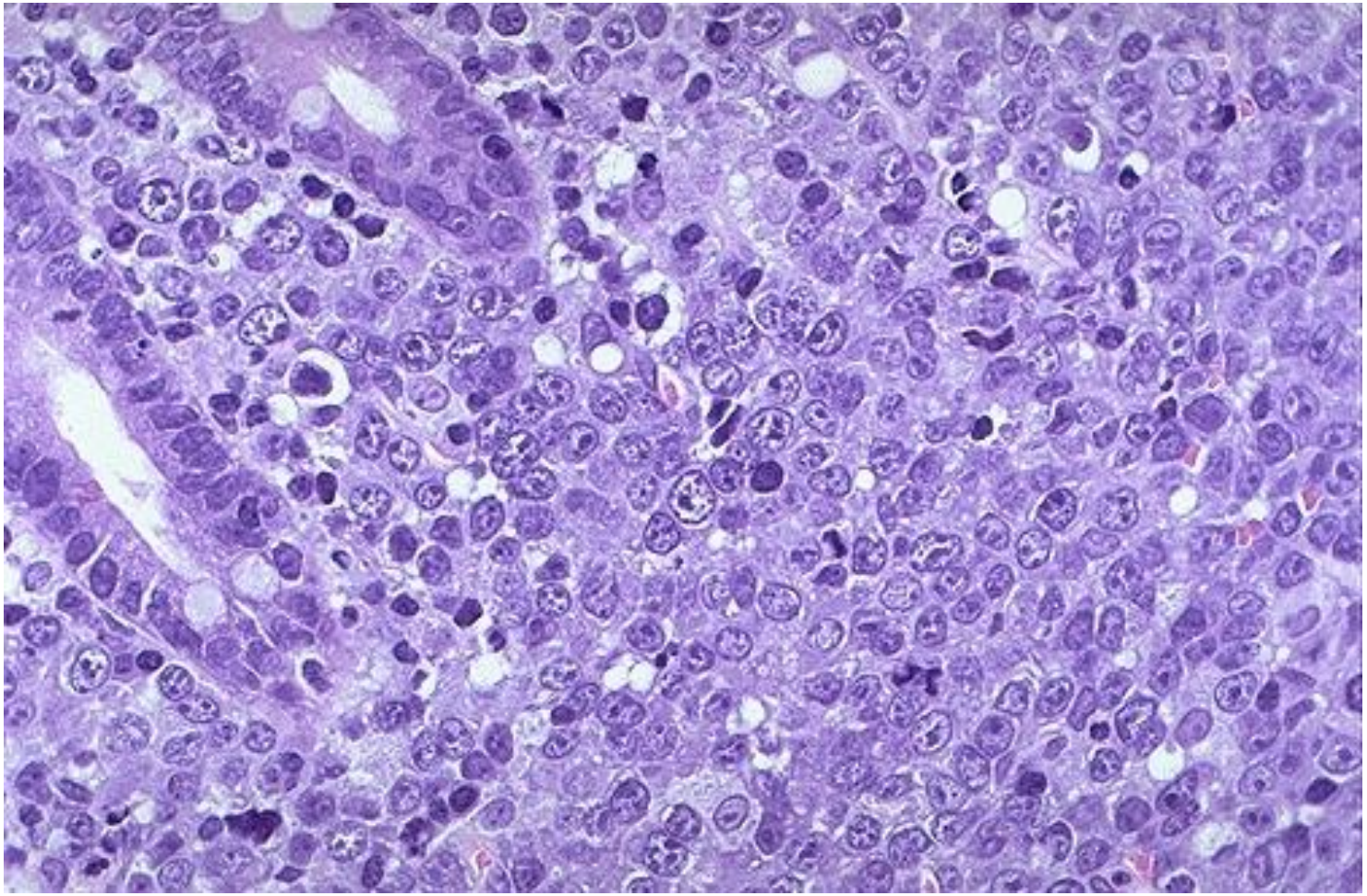
- A diagnostic procedure was performed …..

- Ampullary biopsy:

High grade B cell lymphoma

- Bile no organisms on microscopy and culture sterile
- Bone marrow:

Reactive changes only

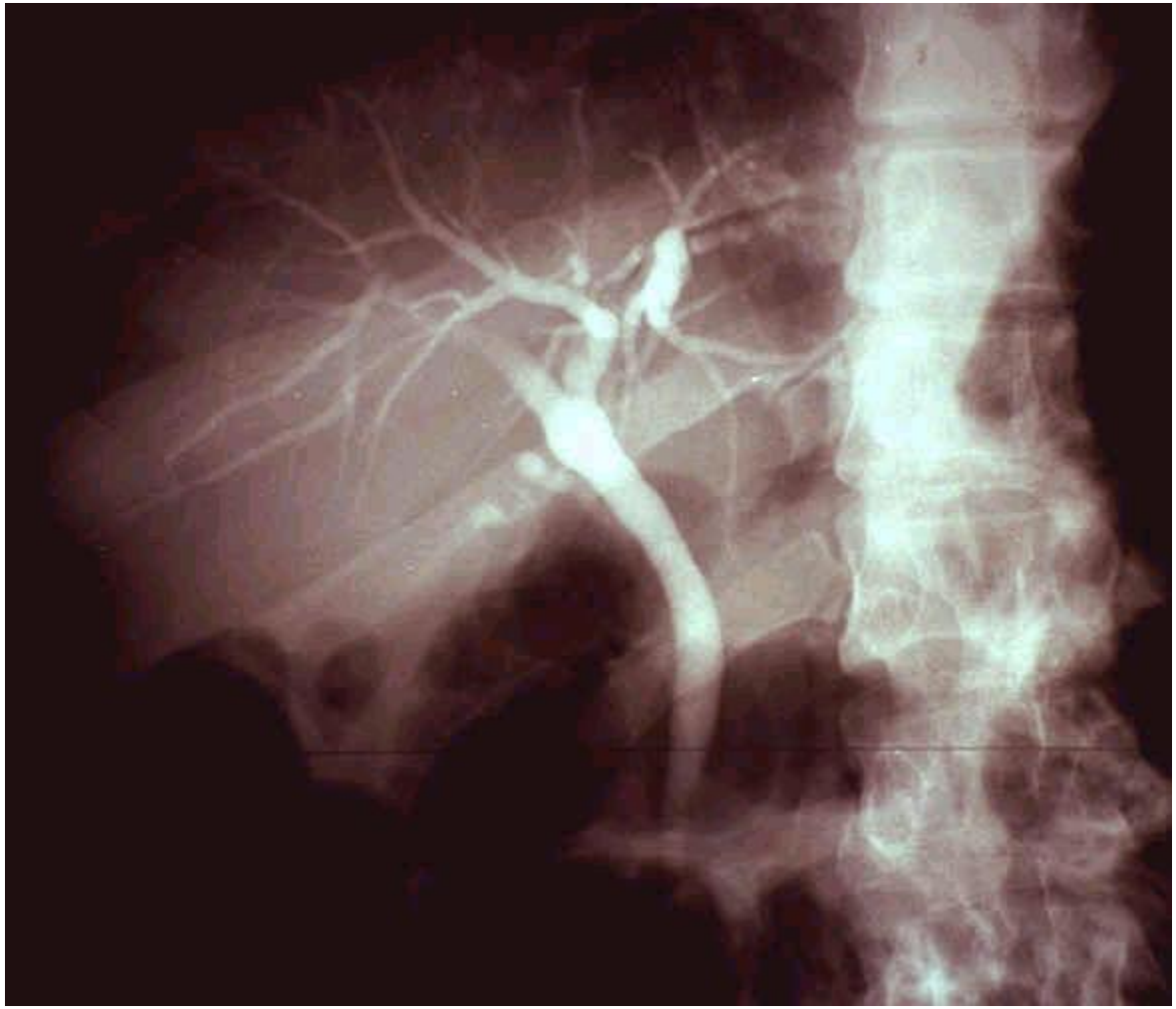


Diagnosis

- AIDS cholangiopathy secondary to high grade B cell lymphoma

Follow-up

- 3 cycles of combination chemotherapy (CHOP)
- Dramatic clinical, biochemical and radiological



Biliary tract disease in HIV

1. Non-HIV associated diseases
2. Acalculous cholecystitis
3. AIDS cholangiopathy:

   cryptosporidiosis, microsporidiosis, CMV, giardia
5. Malignancy
6. Mycobacteria

AIDS cholangiopathy

Resembles PSC

Associated with advanced very low CD4 counts

Fever, abdominal pain and jaundice

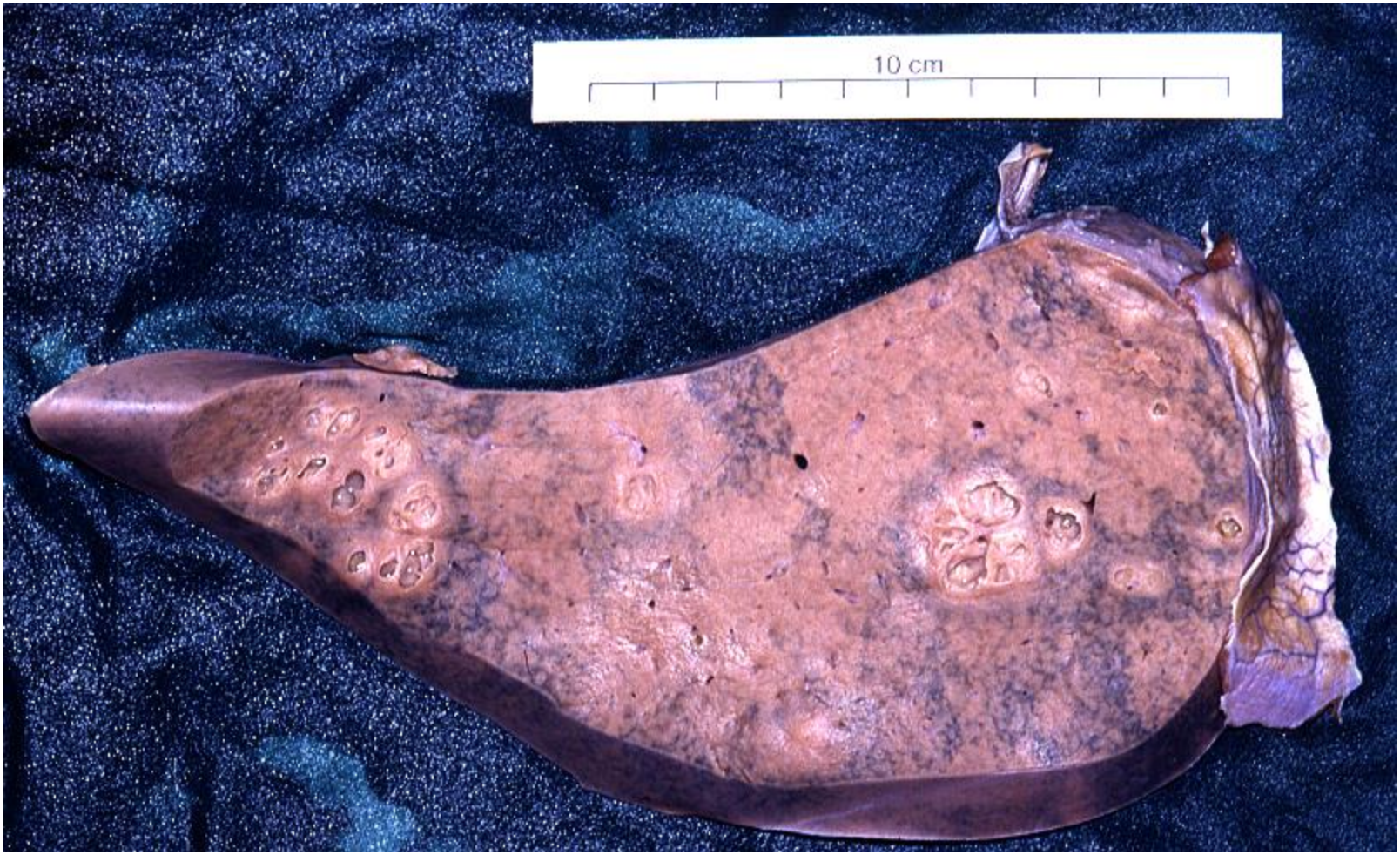
ERCP demonstrates intra- and extra-hepatic strictures

Liver biopsy shows ductopaenia and fibrosis

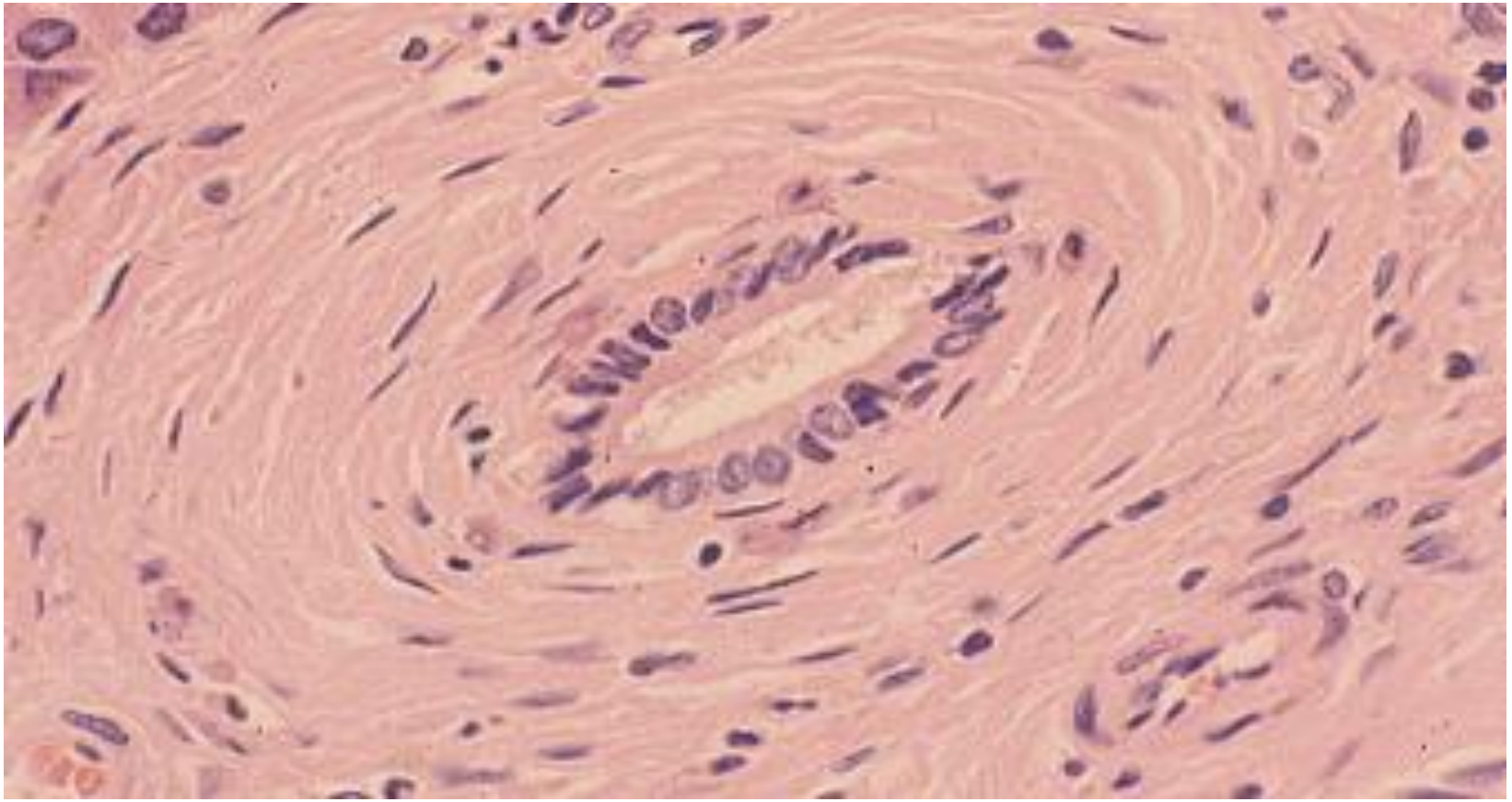
Underlying causes:

Cryptosporidium, Microsporidium and CMV

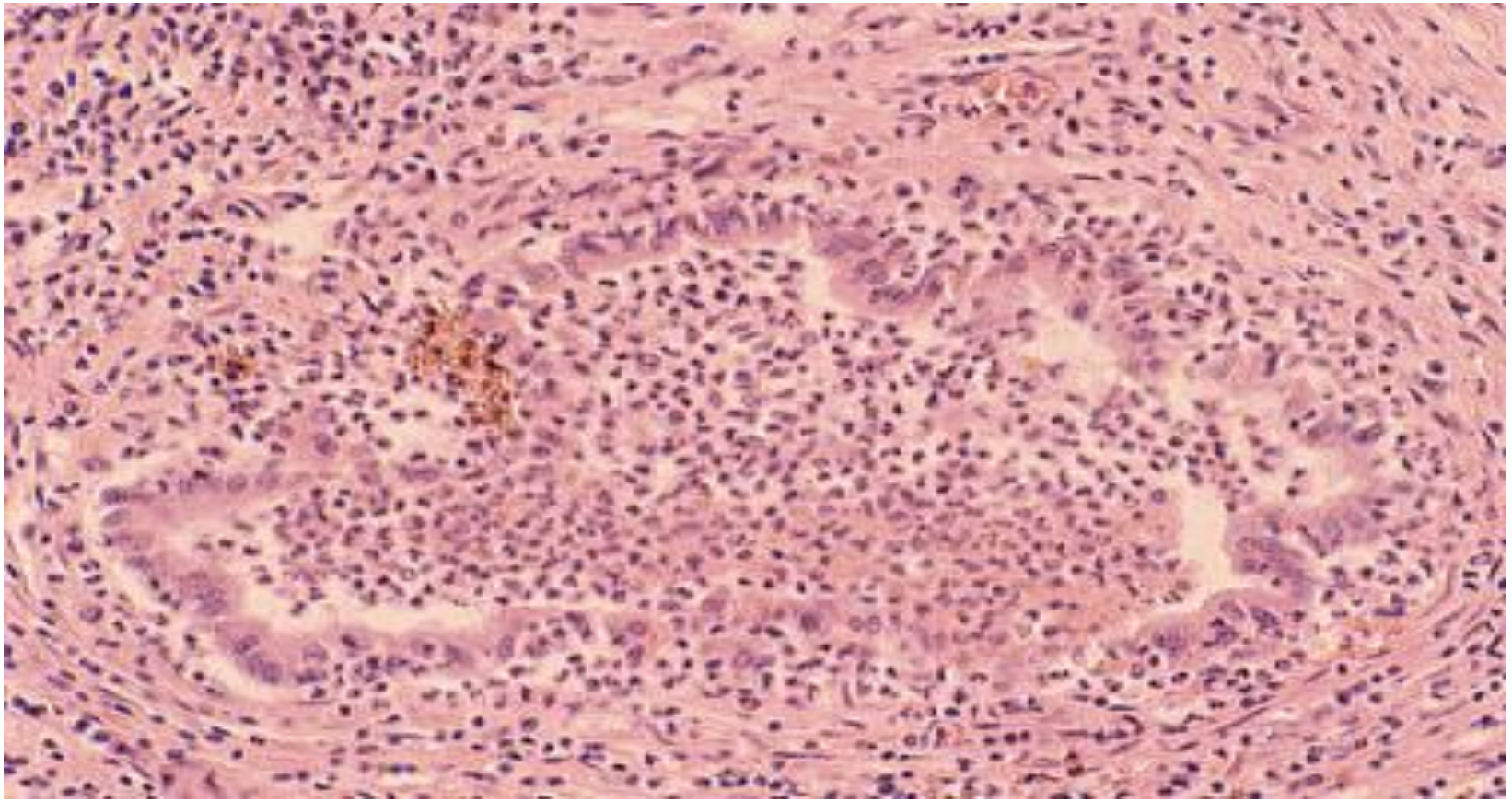
AIDS Cholangiopathy



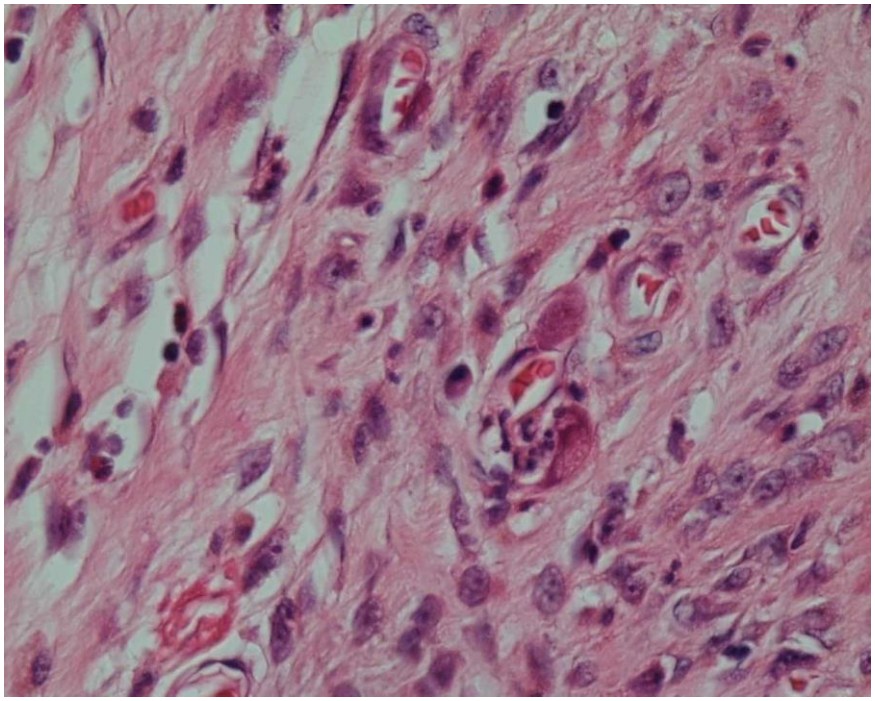
Aids Cholangiopathy



Aids Cholangiopathy



CMV



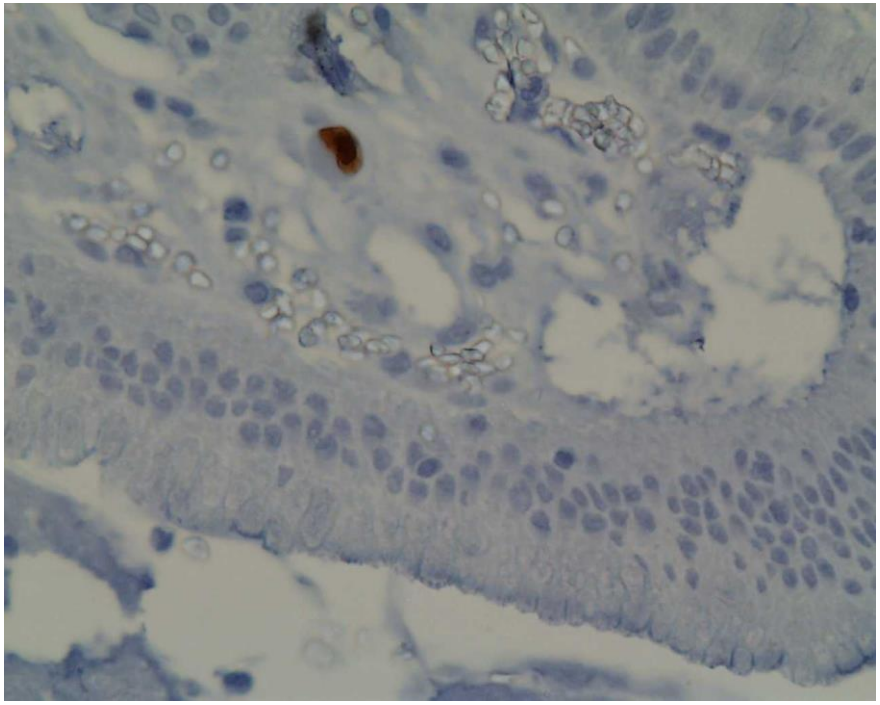


TABLE. Classification of the Cholangiopathies[a]

Genetic
Alagille syndrome
Caroli syndrome
Cystic fibrosis
Polycystic liver disease
ADPLD
ADPKD
ARPKD
Idiopathic
Autoimmune cholangitis
Biliary atresia[b]
Idiopathic childhood/adulthood ductopenia
IgG4-associated cholangitis
Primary biliary cirrhosis[b]
Primary sclerosing cholangitis[b]
Malignant
Cholangiocarcinoma
Secondary sclerosing cholangitis
ABCB4 deficiency
Abdominal trauma (surgical or blunt)
AIDS cholangiopathy
Amyloidosis
Chemical/drugs (ie, 5-fluorouracil)
Choledocholithiasis
Eosinophilic or mast cell cholangitis
Graft-vs-host disease involving the liver
Iatrogenic biliary strictures
Portal hypertensive biliopathy
Recurrent pyogenic cholangitis
Sarcoidosis
Sickle cell disease
Vascular/ischemic (ie, hepatic artery stenosis after liver transplant)

[a]ADPLD = autosomal dominant polycystic liver disease; ADPKD = autosomal dominant polycystic kidney disease; AIDS = acquired immunodeficiency syndrome; ARPKD = autosomal recessive polycystic kidney disease.

[b]For the genetic component of these diseases, see Supplemental Table 2.

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