
Slide seminar:
Liver and Pancreatobiliary pathology


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# Clinical presentation

- 37-year old man with acute cholestatic syndrome complaining of jaundice and pruritus the last five days before admission
- Medical and family history non significant
- No use of medications or exposure to toxic chemicals
- Alcohol consumption: ~ 320g twice/week, last 8 years
- Occasional cannabis user, cocaine inhaler for a 5-month period until 2 months before the icteric episode

# Laboratory findings

- **Cholestatic biochemical profile**
  (tbil:9.9 mg/dL, ALP 315 IU/L, GGT 1082 IU/L)
- **Elevated aminotransferase levels** (AST 122 IU/L, ALT 245 IU/L)
- Prothrombin, serum albumin: normal
- White and red blood cell count: normal
- Viral serological markers: undetectable
- Autoantibodies: negative
- IgG, IgA, IgM values, protein electrophoresis: normal

- **Ultrasound and MRCP**

  No parenchymal abnormalities, no dilation or stenosis of extra-or intrahepatic bile ducts

- **Gastroduodenoscopy,colonoscopy**

  No signs of inflammatory bowel disease

# Predominant histological findings

- Majority of interlobular bile ducts:
  **pleomorphic cholangitis** rarely with epithelial destruction
  *(infiltration of the epithelium by neutrophils and eosinophils, degenerative epithelial changes with nuclear pleomorphism, cell crowding and flattening*)

- Three bile ducts:

  **sclerosing cholangitis** *(pericholangitis, concentric periductal fibrosis, focally severely damaged epithelium)*

- No bile duct loss

# Secondary findings

- Portal oedema and inflammatory infiltrates of mild to moderate density with predominance of neutrophils or lymphocytes, a variable number of eosinophils and only few plasma cells (CD 138 +).
  IgG4 immunohistochemically undetectable

- Mild steatohepatitis and mild cholestasis

# Summary

Biliary disease combining features of diffuse pleomorhic cholangitis and focal sclerosing cholangitis affecting small interlobular bile ducts

# Clinical outcome

The patient responded to corticosteroid treatment and remains asymptomatic without any signs of cholestasis 6 months after completion of therapy.

# Differential diagnosis of small-duct biliary diseases in immunocompetent patients

- Drug induced bile duct damage
- Hepatitis C virus-associated bile duct loss
- Primary biliary cirrhosis/Autoimmune cholangitis
- Steroid responsive IgG4 -associated cholangitis
- Secondary sclerosing cholangitis complicated by recurrent or longstanding episodes of suppurative cholangitis
- Primary small- duct sclerosing cholangitis

→ **Excluded by clinical, laboratory findings and histology**

- **Idiopathic adulthood ductopenia (IAD)**

**Exclusion criteria of primary small-duct PSC**:

- acute clinical onset
- no IBD
- response to steroids
- coexistence of diffuse pleomorphic cholangitis

**Exclusion criteria of secondary cholangitis complicated by acute ascending cholangitis:**

- absence of clinical and laboratory findings of bacterial infection
- absence of suppurative cholangitis histologically

**Diagnosis proposed** :

Idiopathic adulthood ductopenia (IAD)

# IDIOPATHIC ADULTHOOD DUCTOPENIA

- Heterogeneous entity
- Diagnosis one of exclusion
- Young or middle aged adults (median 30 years)
- Distinct male predominance
- Chronic cholestatic disease
- Episodic jaundice and pruritus in 1/3 of the patients

# Diagnostic criteria

- Negative AMA
- No large bile duct involvement
- No history of drug use
- No evidence of IBD
- *Occasional response to steroids*

# Possible aetiologies

- Viral cholangitis
- Autoimmune cholangitis with absence of autoantibobies
- Small- duct PSC  (variant?)
- Late onset of non-syndromic paucity of intrahepatic bile ducts

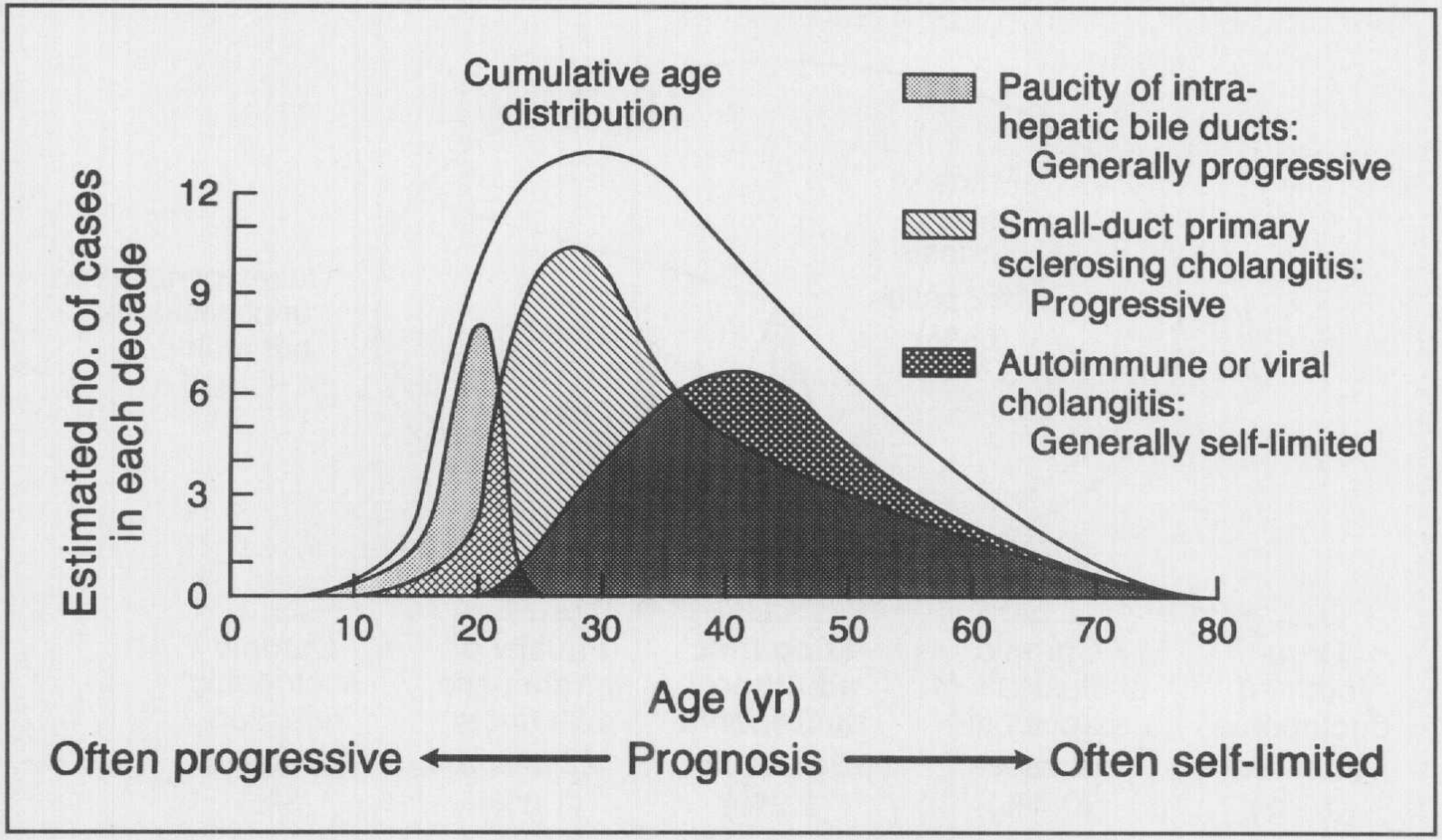


Fig. 4. Putative etiologic groups and their likely age distribution and prognosis in idiopathic adulthood ductopenia.

Ludwig J. *Mayo Clinic Proceedings 1998; 73 (3):* 285

# Diversity of histology

- Ductopenia (diagnostic criterion)
- Pleomorphic destructive cholangitis
- Lymphocytic cholangitis (rare)
- Small duct PSC

# Conclusion

- The case meets the exclusion and inclusion criteria of **IAD** regarding age, sex, clinical history, clinical presentation and laboratory findings

- The combination of two different bile duct lesions probably reflects different stages of a rapid evolving disease starting as pleomorphic cholangitis and progressing to small-duct PSC

- The effectiveness of corticosteroids points towards an immune mediated process

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