Primary Biliary Cirrhosis

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Primary Biliary Cirrhosis

- Autoimmune cholestatic liver disease.
- Destruction of intrahepatic bile ducts.
- Incidence: 1-50/1000000/year
- Predominantly in middle aged women.

PBC- Etiology

- **Unknown but various factors have been implicated**

- **Autoimmunity –**
- **Autoantibodies (AMA) against pyruvate dehydrogenase complex is found in 95% of patients**
- **Anti nuclear Ab's (ANA) in approximately 50%**
- **Elevated serum levels of IgM**
- **Association with a variety of autoimmune- diseases: thyroiditis; scleroderma; CREST - calcinosis cutis, Raynaud phenomenon, esophageal motility disorder, sclerodactyly, and telangiectasia)**

PBC- Etiology

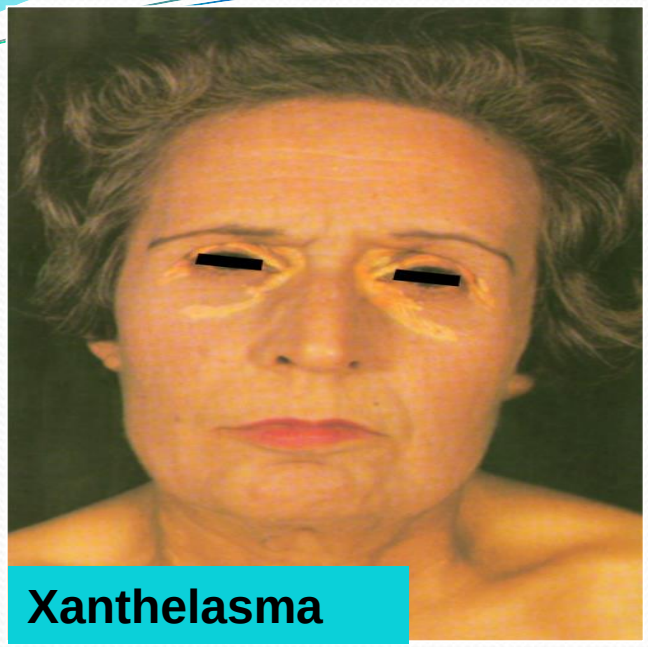
- **Genetic - First-degree relatives have a 570-1000-fold increased chance of developing PBC.**
- **Enviromental - Microorganism infection with organisms of the family Enterobacteriaceae: Cross-reactivity between antigens on the bacterial wall and the mitochondria. Patients with PBC present with an increased incidence of gram-negative UTIs.**
- **Xenobiotics**
- **Others – chemicals, cigarete smoke, exogenous estrogens etc.**

PBC - Pathophysiology

- A continuous destruction of intrahepatic bile ducts, mediated by activated lymphocytes. As a result - chronic cholestasis .
- The retention of toxic substances, such as bile acids and copper, can cause a further secondary destruction of the bile ducts and the hepatocytes.
- Cirrhosis develops late in the course of the disease.

PBC-signs and symptoms

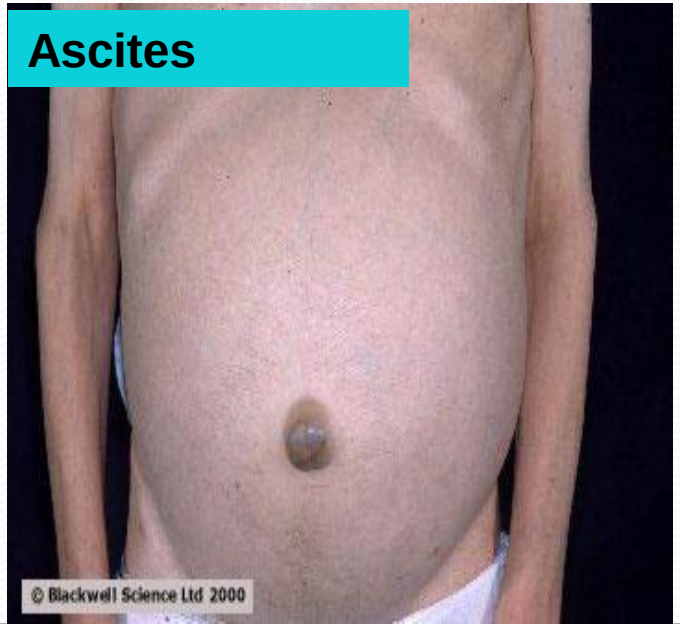
- Asymptomatic 50%
- Fatigue- most common symptom
- Pruritus
- Sicca syndrome
- Jaundice
- Xanthoma /Xanthelasma
- Hepato/splenomegaly
- Signs of cirrhosis: spider nevi, palmar erythema, ascites, temporal and proximal muscle wasting, and peripheral edema.



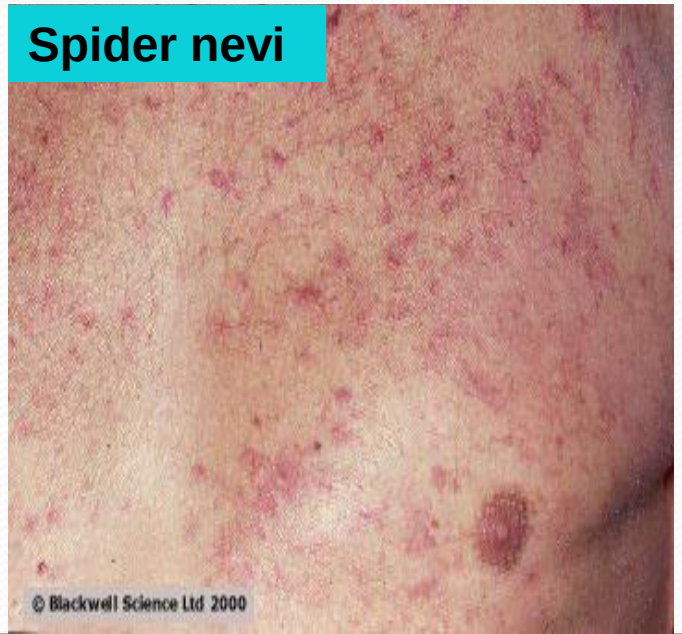
Xanthelasma

Palmar erythema

Ascites



Spider nevi



PBC - Lab

- Anti- Mitochondrial Ab's
- ANA
- IgM elevated
- Alkaline phosphatase elevated
- Cholesterol elevated

PBC-treatment

The goals of treatment are to slow the progression rate of the disease and to alleviate the symptoms.

- **Ursodeoxycholic acid (UDCA)- suppresses hepatic synthesis & secretion of cholesterol , inhibits intestinal absorption of cholesterol.**
- **Suggested but not proved**
  - **Corticosteroids?**
  - **Colchicine?**
  - **MTX?**
  - **Antipruritic treatment**
- **Liver transplantation – high recurrence rate.**

PBC - prognosis

- **Biopsy - to confirm diagnosis and determine stage.**
- **Mayo Clinic Score – survival probability based on:**

  - Age
  - Bilirubin level
  - Albumin level
  - INR
  - Edema
  - Use of diuretics
- **Once symptoms develop, life expectancy is about 10 years.**
- **Cirrhosis develops late in the course of the disease requiring liver transplant.**


MUCHAS
GRACIAS!