Primary Biliary Cholangitis (Primary Biliary Cirrhosis)

Primary Biliary Cirrhosis (PBC) is an inflammatory autoimmune reaction leading to destruction of intrahepatic bile ducts and liver cirrhosis. This disease typically happens to middle-aged women who present with symptoms such as fatigue, pruritus and skin hyperpigmentation.



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Mechanism

Autoimmune Destruction of Bile Ducts
Auto-in-moon Destroys Liver Bile-duck
The pathophysiology of primary biliary cholangitis involves autoimmune destruction, with the specific mechanism still unknown. It is proposed that autoreactive T cells and anti-mitochondrial antibodies target bile ducts and bile duct epithelium, leading to destruction.

Intrahepatic bile ducts
Enter-sign-liver
It is important to note that the autoimmune destruction in primary biliary cholangitis occurs via intrahepatic bile ducts damage, with a proposed mechanism that autoreactive T cells and anti-mitochondrial antibodies target bile ducts and bile duct epithelium, leading to destruction.

Cholestasis
Coal-stop-sign
With destruction of intralobular bile ducts, bile cannot be properly transported, resulting in cholestasis.

Occurs in Middle-Aged Females
Middle-aged Woman in the Middle
Primary biliary cholangitis occurs mostly in middle-aged women and peak incidence is between the ages of 40-50.

Signs and Symptoms

Fatigue
Sleepy-guy
This disease is insidious in onset, but often the initial presenting symptom is fatigue, occurring in approximately 65% of patients with this disease.

Pruritus
Prairie-dog
Due to cholestasis and bile absorption, around 55% of patients become pruritic, which is often another initial presenting symptom.

Skin Hyperpigmentation
Hiker-pig with Hyperpigmentation
In addition to jaundice from bile absorption, hyperpigmentation occurs in 25-40% of patients due to increased melanin deposition. Patients can also complain of dark urine and light stools.

Hepatomegaly
Liver-balloon
Primary biliary cholangitis can lead to significant liver enlargement in approximately 25% of patients, though likelihood increases and enlargement worsens as the disease advances. Hepatomegaly can also be found in previously asymptomatic patients.

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