Primary Sclerosing Cholangitis Mechanisms

This disease results in inflammation and fibrosis of intrahepatic and extrahepatic bile ducts. It is visible as “beading” on radiographs of the biliary tree and in symptomatic patients they present with pruritus and fatigue that can lead to jaundice and hepatosplenomegaly.



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Mechanism

Unknown Mechanism
Question-mark Mechanism
It is believed that primary sclerosing cholangitis may be caused by autoimmune destruction of bile ducts. However, there are competing theories and the mechanism is not fully understood.

"Onion Skin" Fibrosis
Onion
Histologically, fibrosis of bile ducts resembles the layers of an onion. This is known as concentric periductal fibrosis.

"Beading" of Bile Ducts
Beading at Bile-duck with Beads
This is visible on radiographs of the intrahepatic and extrahepatic biliary tree. "Beading” is a manifestation of the strictures and dilations of the bile ducts that occur due to inflammation and fibrosis.

Symptoms

Pruritus
Prairie-dog
Itchiness, along with fatigue, are often presenting symptoms of primary sclerosing cholangitis. However, similar to other cholestatic causes of pruritus, this mechanism is poorly understood.

Hepatosplenomegaly
Liver-spleen-balloon
Chronic and extensive liver inflammation and fibrosis that is progressive and pervasive in primary sclerosing cholangitis lends itself to the development of hepatosplenomegaly.

Jaundice
Jaundice-janitor
As the disease process and liver dysfunction progress, bilirubin metabolism and excretion malfunctions and accumulates, resulting in the characteristic yellow discoloration of the skin called jaundice. Similarly, dark urine may also be present in these patients, manifesting kidney excretion of excess bilirubin.

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