Primary sclerosing cholangitis

Primary sclerosing cholangitis

PSC is a chronic cholestatic liver disease of unknown aetiology , although a genetic predisposition is likely owing to its asso ciation with ulcerative colitis. It produces di ff use, progressive inflammation and fibrosis with structuring of the intra- and extrahepatic biliary tree and mainly a ff ects young men in their thirties. The exact worldwide prevalence is unclear, but it appears to a ff ect 1.5/100 /uni00A0 000 men and 0.5/100 /uni00A0 000 women. - In patients with PSC and ulcerative colitis, the condition usually progresses even following colectomy . The diagnosis is principally based on the finding of irregular, narrow ed bile ducts at cholangiography involving both the intra- and extra - hepatic biliary tree (Figure 69.17), but if the radiological appearances are equivocal a liver biopsy is required. There is no specific treatment and patients usually progress inexora - bly with progressive cholestasis and fatal liver failure. Isolated areas of intrahepatic sclerosing cholangitis can occasionally be resected but di ff use disease usually requires liver transplanta - tion. There is a strong predisposition to cholangiocar cinoma and gallbladder cancer, which should be considered when a new or dominant stricture is demonstrated on cholangiogra - phy or when gallbladder ‘polyps’ are identified. T he di ffi culty in the clinical setting is distinguishing scleros - ing cholangitis from a malignant process, particularly multi-). focal cholangiocarcinoma. Imaging cannot reliably di ff erentiate between inflammatory and malignant strictures and rarely demonstrates a mass lesion even in patients with advanced c holangiocarcinoma. Diagnosis often requires biliary brush TM cytology or direct endoscopic inspection (SpyGlass). Serum cancer antigen (CA) 19-9 levels may be increased but the sensitivity of CA 19-9 in detecting cholangiocarcinoma in PSC is only 60%. Temporary relief of obstructive jaundice owing to a dominant bile duct stricture can be achieved by biliary stenting, although there is considerable risk of cholangitis. Patients with good liver function, no dominant strictures and negative biliary cytology are monitored for disease progression. Liver transplantation produces excellent results if performed before the development of malignancy .

Figure 69.17 Typical appearance of primary sclerosing cholangitis with a ‘beaded’ appearance of the intrahepatic ducts and diffuse widespread strictures. The intrahepatic ducts usually do not dilate owing to the pathological process involving the whole of the biliary tract.

Revision #1
Created 2025-12-31 15:25:45 UTC by Omar Ayman
Updated 2025-12-31 15:25:45 UTC by Omar Ayman