PRIMARY SCLEROSING CHOLANGITIS

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PSC is a rare idiopathic and progressive biliary tract disease characterised by inflammation and destruction of the intrahe patic and extrahepatic bile ducts that can lead to liver fibrosis and cirrhosis. Association with hypergammaglobulinaemia and markers such as anti-smooth muscle antibodies and anti-nuclear factor suggest an immunological basis; cystic fibrosis transmembrane conductance regulator (CFTR) gene muta tions have been associated with the development of PSC. The majority of patients are between 30 and 60 years of age. There appears to be a male predominance and a strong association with inflammatory bowel disease (IBD), especially ulcera colitis (IBD in PSC, 80%; PSC in IBD, 5%). Patients may be asymptomatic, but common symptoms include pruritus, fever, fatigue, right upper quadrant discom fort, jaundice and weight loss. Liver function tests reveal a Summary box 71.5 Causes of benign biliary stricture /uni25CF /uni25CF - /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF Summary box 71.6 Radiological investigation of biliary strictures /uni25CF /uni25CF /uni25CF - /uni25CF /uni25CF - cholestatic pattern, elevated serum ALP and gamma-glutamyl transpeptidase (GGTP) and smaller rises in the aminotransfer - ases; bilirubin values can be variable. MRCP (or ERCP) may demonstrate stricturing and beading of the bile ducts (Figure tive 71.37). Liver biopsy is helpful to confirm the diagnosis (con - centric periductal ‘onion skinning’) and may help guide ther - apy by excluding cirrhosis. Important di ff erential diagnoses are - secondary scler osing cholangitis, immunoglobulin G4 (IgG4) cholangitis, autoimmune hepatitis, human immunodeficiency

(a) A T-tube in situ with a stone in the bile duct. (c) The stone is extracted from the duct along the Congenital In /f_l ammatory Biliary atresia Stones Cholangitis Bile duct injury at Parasitic surgery Pancreatitis Cholecystectomy Sclerosing cholangitis Choledochotomy Radiotherapy Gastrectomy Hepatic resection Trauma Transplantation Idiopathic USG MRCP ERCP (with brush cytology in cases of a dominant stricture) PTC CT

virus (HIV) cholangiopathy and cholangiocarcinoma. The last may arise in patients with PSC and is di ffi cult to diagnose; a high index of suspicion is required, especially in the setting of unexplained clinical deterioration. Patients with PSC are at increased risk for cholangio carcinoma and gallbladder cancer, as well as colon cancer in those with concurrent IBD. Medical management with antibiotics, vitamin K, cholestyramine, steroids and immunosuppressant drugs may not relieve symptoms. Endoscopic stenting of dominant strictures and, in selected patients with pr edominantly extrahepatic disease, operative resection may be worthwhile. For patients with cirrhosis, liver transplantation is the best option; 5-year survival following transplantation in high-volume centres is in excess of 80%. Screening for malignancies involving the gallbladder (polyp)

and bile ducts and colonoscopy for IBD or malignancy are therefore critical, and bone densitometry for bone density is mandatory .

Figure 71.37 Sclerosing cholangitis in a patient with ulcerative colitis, visualised by endoscopic retrograde cholangiopancreatography.

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