



FIGURE 44-6 Cholangiogram obtained on endoscopic retrograde cholangiopancreatography demonstrates the characteristic beading of the intrahepatic and extrahepatic bile ducts in a patient with primary sclerosing cholangitis.

men (two thirds of patients are younger than 45 years of age), often in association with ulcerative colitis. Approximately 70% of patients with PSC have ulcerative colitis. The clinical spectrum of PSC is broad, ranging from asymptomatic patients with abnormal liver enzyme levels (typically an elevated alkaline phosphatase concentration) to patients with recurring episodes of fever, chills, abdominal pain, and jaundice. The diagnosis of PSC is made by MRCP or ERCP, which show characteristic changes (beading) of the intrahepatic and/or extrahepatic bile duct (Fig. 44-6).

No proven therapy exists for PSC, although ursodeoxycholic acid and methotrexate are being used in some centers. Other forms of therapy include prophylactic antibiotics for prevention of recurrent bacterial cholangitis, treatment of pruritus, and repletion of fat-soluble vitamins. Endoscopic dilatation of a dominant biliary stricture during ERCP is an effective treatment of

cholestasis in selected patients. Most patients with advanced PSC eventually progress to end-stage liver disease, and evaluation for liver transplantation is appropriate in advanced disease. One third of patients with PSC will develop cholangiocarcinoma; therefore, thorough clinical and laboratory studies (liver function tests and cancer markers such as CA 19-9) and radiologic follow-up are warranted.

Sphincter of Oddi Dysfunction

Sphincter of Oddi dysfunction is a benign motility disorder that leads to noncalculous obstruction of the flow of bile or pancreatic juice at the level of the pancreaticobiliary junction. Patients typically have unexplained biliary-type abdominal pain, with or without elevated results on liver function tests and with or without bile duct dilation. In a selected group of patients, endoscopic or surgical sphincterotomy is of value.

For a deeper discussion of this topic, please see Chapter 155, "Diseases of the Gallbladder and Bile Ducts," in Goldman-Cecil Medicine, 25th Edition.

SUGGESTED READINGS

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