Macroscopically-localized hepatic lesions in sclerosing cholangitis[1]

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A first report is presented of lesions on the liver surface mimicking liver metastases, seen at laparotomy in a patient with mild ulcerative colitis. Biopsy revealed focal sclerosing cholangitis.

Sclerosing cholangitis is an uncommon disease of unknown aetiology affecting the biliary apparatus. It has a variable course and is usually slowly progressive (Chapman *et al.* 1980, Wiesner & La Russo 1980). The disease may however be asymptomatic (Chapman *et al.* 1981, Thompson *et al.* 1982).

The diagnosis of sclerosing cholangitis must be considered in patients with ulcerative colitis who develop upper abdominal pain and altered liver function tests. The disease usually affects both the intrahepatic and extrahepatic ducts. Endoscopic retrograde cholangiopancreatography is essential.

Case report

Mrs KF, aged 33, had suffered from intermittent diarrhoea for nine months when a diagnosis of ulcerative colitis was made in 1979. In May 1980 she lost her appetite and developed epigastric pain. On examination there was epigastric tenderness and guarding. Liver function tests showed a raised serum alkaline phosphatase of 1365 IU/l (upper limit of normal 170). The antimitochondrial antibody titre was negative. An intravenous cholangiogram appeared to show gallstones in a small contracted gallbladder.

She was admitted nine weeks later for elective cholecystectomy, at which time the serum bilirubin was normal, although the serum alkaline phosphatase was still raised at 652 IU/l.

At laparotomy the gallbladder and extrahepatic ducts appeared normal. On the upper surface of the left lobe of the liver there was a 4 cm diameter raised variegated lesion, grey/purple/brown in colour. There were two similar circumscribed lesions on the surface of the right lobe of an otherwise normal liver. No other abnormality was found. Biopsies were taken of the hepatic lesions which were thought to be metastases. No further procedure was carried out.

Histology showed almost total destruction of the normal architecture (Figure 1). Most of the tissue was fibrous, surrounding the remaining islands of liver cell trabeculae. There was a variable chronic inflammatory cell infiltrate, which in places surrounded the remnants of bile ducts and ductules. There was no evidence of malignancy. The appearances were of a focal form of sclerosing cholangitis.

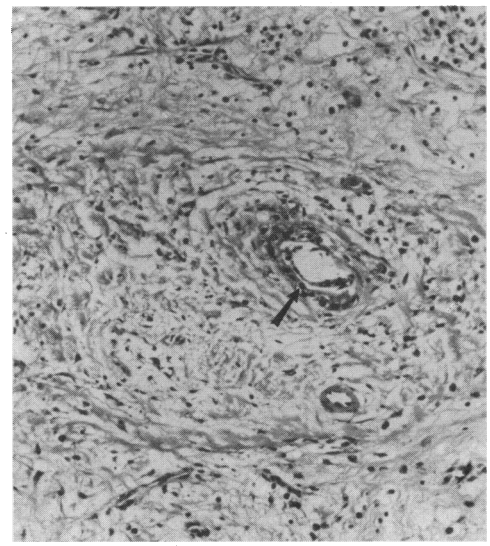

Figure 1. Liver biopsy showing fibrosis and inflammation around atrophic bile ducts. (H&E × 220)

Endoscopic retrograde cholangiopancreatography (ERCP) in January 1981 showed a moderately-dilated common bile duct with a diameter of 18 mm, with no evidence of obstruction distally. The intrahepatic ducts were small and showed definite beading (Figure 2), establishing a diagnosis of sclerosing cholangitis (Williams & Schoetz 1981, Bonnel *et al.* 1982, Lefkowitch 1982). Since this time she has remained clinically well, but her liver function tests continue to be abnormally raised.

Discussion

The macroscopic findings at laparotomy of discrete lesions mimicking secondary carcinomatous deposits (especially in a patient with ulcerative colitis), with the rest of the liver appearing normal macroscopically, do not appear to have been reported previously.

The association between ulcerative colitis and sclerosing cholangitis is now well recognized, but not explained. Ulcerative colitis has been reported to be present in 20–30% of cases of sclerosing cholangitis; however, only 1% of patients with ulcerative colitis develop sclerosing cholangitis (Williams & Schoetz 1981). Patients with ulcerative colitis who develop sclerosing cholangitis often present with weight loss, pruritis, jaundice,

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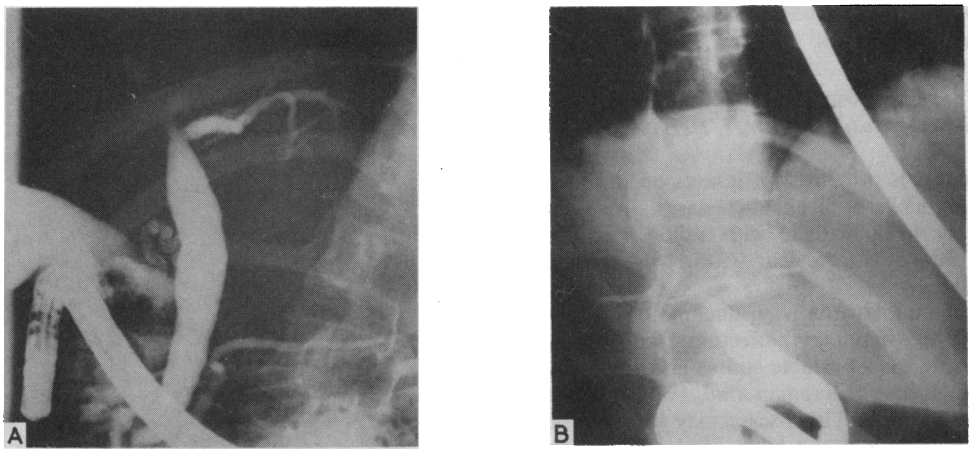

Figure 2A & B. Endoscopic retrograde cholangiopancreatography demonstrating small intrahepatic ducts with beading

fever and pain (Thompson *et al.* 1982). The alkaline phosphatase is usually raised disproportionately to the other liver function tests, being more than three times the upper limit of normal in 75–85% of cases (Chapman *et al.* 1980, Wiesner & La Russo 1980).

Intravenous cholangiography is usually unrewarding in patients with sclerosing cholangitis (Wiesner & La Russo 1980, Kotel *et al.* 1979, *British Medical Journal* 1976). Percutaneous transhepatic cholangiography using a skinny Chiba needle may be of use diagnostically (Chapman *et al.* 1980) but it is difficult technically, because the bile ducts are usually narrowed in sclerosing cholangitis (Kotel *et al.* 1979, *British Medical Journal* 1976).

The diagnostic criteria have widened since 1976, when it was suggested that sclerosing cholangitis required the features of progressive obstructive jaundice with generalized thickening and stenosis of the biliary system (*British Medical Journal* 1976, Williams & Schoetz 1981). Patients previously labelled as having pericholangitis have the bile duct abnormalities of sclerosing cholangitis (Blackstone & Nemchauskey 1978, Chapman *et al.* 1980), namely, multiple stricturing of the biliary tree with beading of the intrahepatic bile ducts.

The grossly visible hepatic masses represented localized areas of fibrosis. The characteristic microscopic changes in liver biopsy specimens of primary sclerosing cholangitis are bile duct proliferation, periductal fibrosis and inflammation, ductal obstruction and actual loss of bile ducts. However the hepatic histology may be non-diagnostic (Ludwig *et al.* 1981).

The wider availability of ERCP has increased the diagnosis of sclerosing cholangitis two-fold (Wiesner & La Russo 1980). It is the least invasive investigation which can reliably provide the diagnosis. Laparotomy should be used selectively to relieve bile duct obstruction if it becomes complete, or to manage bile duct carcinoma, or to remove biliary sludge and stones.

Although laparotomy provided a tissue diagnosis in the present case and indicated that there was no other abnormality in the abdomen, the prior investigation by ERCP would have provided the diagnosis.

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References

Blackstone M O & Nemchauskey B A
(1978) *American Journal of Digestive Diseases* **23,** 579–585
Bonnel D, Grenier P, Cornud F & Nahum H
(1982) *Gastroenterologie Clinique et Biologique* **6,** 73–79
British Medical Journal (1976) ii, 1090–1091
Chapman R W G, Arborgh B A M, Rhodes J M, Summerfield J A, Dick R, Scheuer P J & Sherlock S
(1980) *Gut* **21,** 870–877
Chapman R W G, Burroughs A K, Bass N M & Sherlock S
(1981) *Digestive Diseases and Sciences* **26,** 778–782
Kotel C M, Baddeley H & Salmon P R
(1979) *Clinical Radiology* **30,** 445–450
Lefkowitch J H
(1982) *Archives of Internal Medicine* **142,** 1157–1160
Ludwig J, Barham S S, La Russo N F, Elveback L R, Wiesner R H & McCall J T
(1981) *Hepatology* **1,** 632–640
Thompson H H, Pitt H A, Tompkins R K & Longmire W P
(1982) *Annals of Surgery* **196,** 127–136
Wiesner R H & La Russo N F
(1980) *Gastroenterology* **79,** 200–206
Williams L F & Schoetz D J
(1981) *Surgical Clinics of North America* **61,** 951–962