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Sclerosing Cholangitis: Surgical Significance

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The authors' experience with the rare entity of sclerosing cholangitis suggests that several anatomical patterns of involvement of the extrahepatic biliary tract may occur. The involvement may include the intraluminal ducts segmentally or diffusely, or the process may primarily be external to the ducts as pericholedochitis. It may or may not represent one component of a systemic disease, particularly autoimmune disease.

Although involvement may be diffuse, progression may be slow and compatible with many years of life.

Long time followup is essential before any value can be assigned to a given modality of treatment. Carcinoma of the ductal system may be found after a more exhaustive examination or at autopsy.

All efforts should be provided operatively, however, to relieve biliary tract obstruction. If this is not possible or provides only incomplete relief, steroid therapy should be administered.

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A COMMON duct stone or carcinoma of the head of the pancreas is usually suspected in a patient with jaundice. When the surgeon operates, a thickened gallbladder is found but no stones are palpable. The wall of the common duct is very thick, and the thickness encroaches on the lumen so that it is difficult to introduce instruments or a T-tube. Operative cholangiography reveals narrowing of the extrahepatic biliary system and the intrahepatic ducts may also be involved. There is no evidence of calculous disease, and malignancy of the pancreas or bile ducts seems unlikely.

In such a patient, the tentative diagnosis of "primary sclerosing cholangitis" is justified. The entity was well described by Schwartz and Dale in 1958[1] although the honor of having described the first case is frequently given to the French surgeon Delbet.[2] Obviously, it is not important whether there have been 100 or only 50 cases reported to the present time. The important consideration is whether or not the surgeon of the 1970s is apt to encounter a "bona fide" case of sclerosing cholangitis and whether or not he will render optimal treatment.

Most authors consider the following criteria necessary for inclusion of a case:[1-4]

1. Absence of previous biliary tract operations
2. Absence of gallstones
3. Diffuse, generalized involvement of the extrahepatic biliary ducts

4. Exclusion of cholangiocarcinoma

Warren and his group[5] admitted that their criteria for classification were much less rigid than others.

Although this entity is rare, we encountered among 369 patients six for whom the diagnosis of sclerosing cholangitis was initially given. All had been treated for extrahepatic obstructive lesions. In two of the six the intraoperative findings were similar but the patients eventually died. Absence of postmortem examinations made confirmation of the intraoperative diagnosis impossible. Our experience suggests that sclerosing cholangitis may be characterized by at least three patterns of anatomic involvement of the bile ducts.

1. Segmental extrahepatic, intraluminal obstructive sclerosing
2. Diffuse sclerosing, commonly associated with chronic ulcerative colitis
3. Diffuse, primarily extrahepatic sclerosing pericholedochitis

Segmental Sclerosing Cholangitis

This variety of sclerosing cholangitis results in recurrent bouts of cholangitis which are relieved if operative procedures to by-pass the obstruction are feasible. At operation, stenosis is limited to a segment, usually located distally, at the extrahepatic bile ducts. No other disease process has been evident and the biliary tract sclerosis has not appeared to be progressive in the patient whose case illustrates this clinical pattern. This permits a good outlook for his future.

Case 1: A man, aged 64 years, had had five episodes of chills, fever, and jaundice during the 21 months preceding his examination. Each of these bouts lasted several weeks. He had been taking only thyroid and vitamin medications and consumed no alcohol. He was initially treated for suspected infectious hepatitis. A liver biopsy done six months previously showed microscopic evidence of a marked cellular infiltration in the portal spaces but the hepatic parenchyma appeared normal except for an increase of pigment in the hepatic cells. When the patient was seen a few weeks after his last episode of cholangitis, his physical examination showed a liver enlarged to two fingers below the right costal margin. He recalled a brief period of painless jaundice approximately 40 years before.

At the time of his examination, the patient's total serum bilirubin varied from 2.08 to 5.45 mg % and the serum alkaline phosphatase varied from 9.9 to 17.2 Bodansky units % (normal 1.5 - 4.0). The serum antinuclear factor test was positive (weak to medium homogeneous). The prothrombin time, SGOT, and serum immunoglobulin determinations were all within normal limits. The intravenous cholangiogram showed the faint outline of a slightly dilated proximal common bile duct.

At operation, the liver appeared normal but the gallbladder appeared distended. No stones were found in the biliary tract. The pancreas appeared normal. The segment of the common bile duct within the pancreas was stenotic with dilation present proximal to this (Figure 1). A silver wire probe could not

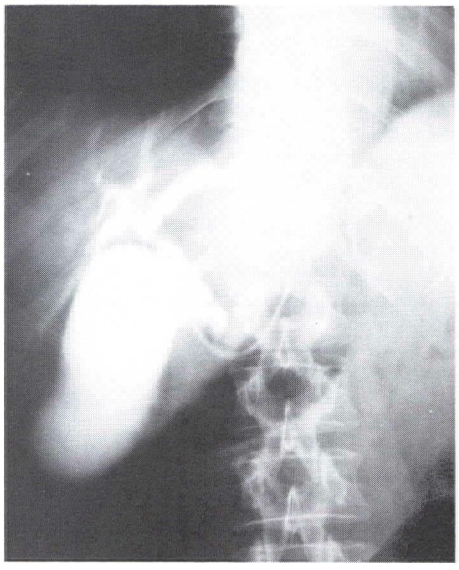

Figure 1
Operative cholangiogram showing obstruction in distal common bile duct due to segmental sclerosing cholangitis.

be passed through the stenotic segment. Biopsies from the stenotic portion showed evidence only of acute and chronic inflammation with fibrosis (Figure 2). A side-to-side choledochoduodenostomy and a cholecystoduodenostomy were performed.

Postoperatively, the patient's serum alkaline phosphatase and serum bilirubin returned to normal levels. The patient has remained asymptomatic six years later.

Diffuse Sclerosing Cholangitis Associated with Chronic Ulcerative Colitis

Sclerosing cholangitis involving the entire biliary tract, both intrahepatic and extrahepatic, is associated with other diseases, particularly chronic ulcerative colitis.[2] This clinical variety is also only slowly progressive in many patients who remain asymptomatic for many years.[3]

Case 2: Two cholesystograms done in 1968 revealed a nonfunctioning bladder in a 40-year-old female who had recurrent episodes of right upper abdominal pain radiating to the back. A total colectomy and ileorectostomy had been performed 11 years previously (in 1957) because of disabling manifestations of chronic ulcerative colitis for the preceding eight years. Following this operation the patient had been relatively asymptomatic except for an anal fistula requiring a fistulectomy in 1963.

At operation for the biliary tract difficulty the gallbladder was found to be thick-walled, the cystic duct thick and scarred, and the extrahepatic bile ducts also thickened and nodular to palpation. An operative cholangiogram showed an irregular small lumen throughout the entire extrahepatic biliary tract (Figure 3). One fleck of a pigment debris was found in the wall of the excised gallbladder but no calculi otherwise. Microscopically, the gallbladder wall was involved by a heavy infiltrate of inflammatory cells. Fibrosis around bile ducts, but normal hepatic cells, were seen in a liver biopsy.

Postoperatively, the patient had two episodes of mild pain in the right upper abdomen during the first year but no further biliary tract symptoms during the subsequent five years. However, she has required excision of the rectal stump with an ileostomy because of a rectovaginal fistula.

Figure 2
Photomicrograph of biopsy from bile duct involved by segmental sclerosing cholangitis. The endothelial lining is covered by a thick band of tissue. H and E x115.

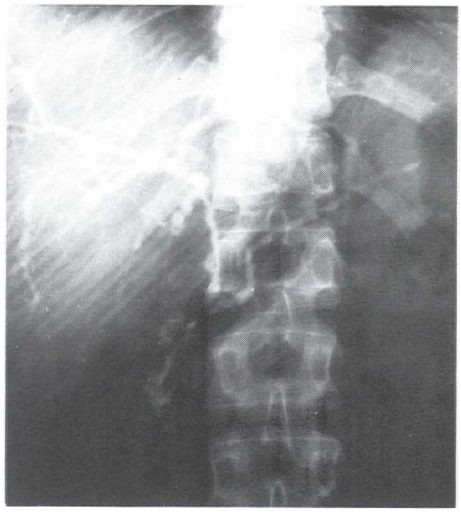

Figure 3
Operative cholangiogram from patient having diffuse sclerosing cholangitis associated with chronic ulcerative colitis. The duct is narrowed by intraluminal irregularities and beading.

Primary Fibrosing Pericholedochitis

This variety of chronic inflammation and fibrosis, involving the extrahepatic biliary tract, is difficult to distinguish from manifestations of carcinoma or the consequences of biliary lithiasis or other lesions predisposing to chronic inflammation in this region, such as duodenal ulcer disease and pancreatitis. Biopsies which show only scarring and chronic inflammation are not reliable for excluding an underlying carcinoma. The subsequent clinical course after operative findings must be reviewed to establish the diagnosis.

Case 3: A woman, aged 47 years, was seen in 1968 because she had had pruritis for four months and jaundice for one month. The only medication she had taken was Donnatal for abdominal gaseousness during the preceding year. She did not drink alcohol and had no pain. There was no history of abdominal operations. Her liver was slightly enlarged but not tender.

Laboratory studies showed the total serum bilirubin ranged from 12.2 to 19.8 mg % and the alkaline phosphatase 7.6 to 46 Bodansky units. The SGOT varied from 135 to 700 units. The IgM component of the serum immunoglobulins was elevated to 161 units (normal 30-120) and the IgG was elevated to 1556 units (normal 600-1400). The ANF test was positive, showing medium speckle-like threads. The direct and Coombs' test, serum amylase and lipase as well as other usual laboratory studies were negative or within normal limits. A liver scan using 131-I Rose Bengal failed to show excretion from the liver into the intestinal tract.

At operation, the region of the porta hepatis and gallbladder, as well as adjacent structures including the duodenum, hepatic flexure and associated mesocolon were involved in an inflammatory process with much fibrosis. The liver was dark but had a smooth surface and showed none of the features of primary biliary cirrhosis. No fluid could be obtained by aspirating the gallbladder and no stones were palpable. A segment of the common bile duct was isolated but found to be entirely incorporated in a fibrotic process. Biopsies from the gallbladder and portal hepatis and a lymph node adjacent to the cystic duct showed only chronic inflammation and fibrosis. A definitive operative procedure did not appear feasible.

Postoperatively, the patient's jaundice cleared spontaneously, and although the serum bilirubin returned to normal levels, the serum alkaline phosphatase remained elevated. However, painless jaundice recurred five months later. The ANF test remained positive and the serum immunoglobulins IgG and IgM remained elevated. After instituting steroid therapy the jaundice again rapidly disappeared. She has remained asymptomatic for the subsequent four years as steroid therapy was continued. The alkaline phosphatase was 8.9 units after receiving steroids for 18 months.

Discussion

By applying indiscriminately the first three diagnostic criteria listed previously, many cases of primary sclerosing cholangitis, such as the segmental variety, will be excluded. Of course sclerosis of the extrahepatic ducts may be secondary to trauma, including blunt, penetrating and operative. It may also result from inflammatory processes arising from adjacent structures.

In certain situations, it may be difficult to be certain whether sclerosis of the extrahepatic ducts is primary or secondary to other disease processes or of iatrogenic origin. On occasion it may be difficult even at operation to identify sclerosing cholangitis as primary or secondary.

Case 4: A 25-year-old female was first seen in 1971 because of jaundice. Two years previously she had been operated on in another hospital for evidence of biliary tract symptoms. No stones were found in the biliary tract, a cholecystectomy was performed, and evidence of sclerosis of the extrahepatic ducts was noted. Two months postoperatively the patient developed jaundice and exploration of the common bile duct was performed. Evidently diffuse sclerosis of the bile ducts prevented the T-tube, placed in the common bile duct, from providing relief of jaundice. No evidence of primary liver disease was observed.

At the time of our examination in 1971, the patient demonstrated features of obstructive jaundice with a serum bilirubin of 11 mg %. At operation, a lumen for the extrahepatic biliary tract could not be demonstrated. The

entire tract was involved with dense scarring and chronic inflammation. Also, chronic inflammation surrounding cholangioles was evident during a liver biopsy. Drains were placed to the divided scar at the hilum of the liver.

The patient developed a biliary cutaneous fistula postoperatively. Injection of radio-opaque media showed the presence of a narrow duct entering the liver with narrow intrahepatic radicals (Figure 4). Because of recurrent bouts of cholangitis, the patient again underwent operation. A duct within the hilum of the liver was identified and a Roux-en-y hepaticojejunostomy accomplished (Figure 5). The patient subsequently improved, and except for several episodes of fever, remained asymptomatic for 18 months. She then became jaundiced and febrile. Suspected obstruction at the anastomosis led to a revision of the hepaticojejunostomy. She recovered following this operation and has been asymptomatic for the last six months.

It is never possible to identify sclerosing cholangitis preoperatively. The manifestations do not readily distinguish this entity from more common varieties of chronic disease of the extrahepatic biliary tract. Percutaneous hepatic cholangiography may be helpful, but not specifically diagnostic. Present use of transduodenal endoscopy has become useful in suggesting lesions of the pancreatic duct or in carcinomas of the bile ducts (Figure 6). In time, its continuous application may provide certain features suggesting the possibility of sclerosing cholangitis. At operation the thickening of the extrahepatic tract is usually diffuse, but in some patients it is more pronounced in segments. The external appearance of the ducts is not increased. Operative cholangiography is of real value in the assessment of the anatomic situation. In a few patients stones can be expected since obstructive factors predispose to stone formation.

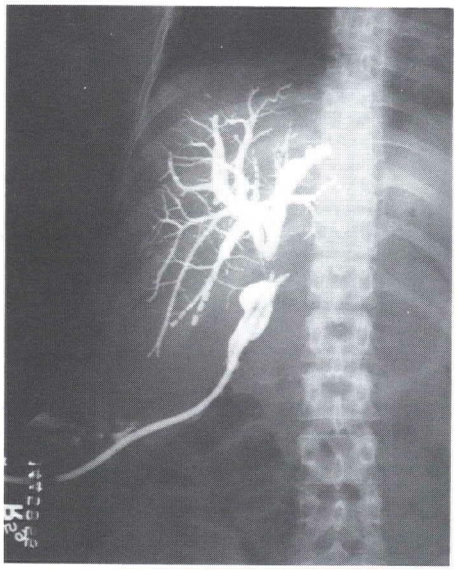

Figure 4
Radio-opaque media in biliary cutaneous fistula showing narrow intra- and extrahepatic duct system in patient with probably diffuse sclerosing cholangitis.

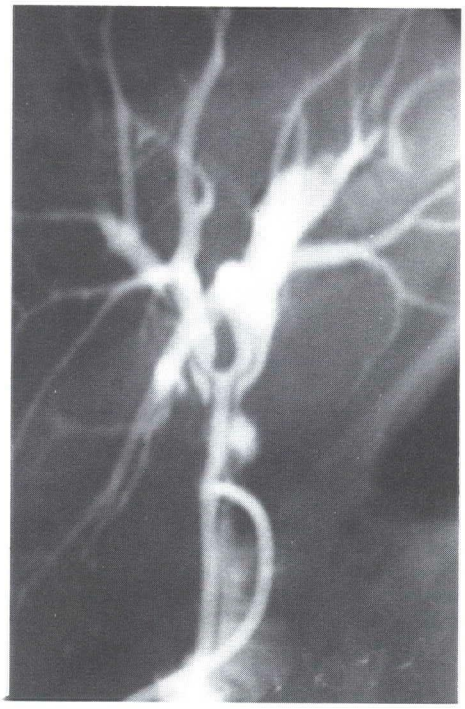

Figure 5
Cholangiogram performed through a Catell tube shows adequate opening of hepaticojejunostomy needed to relieve biliary obstruction.

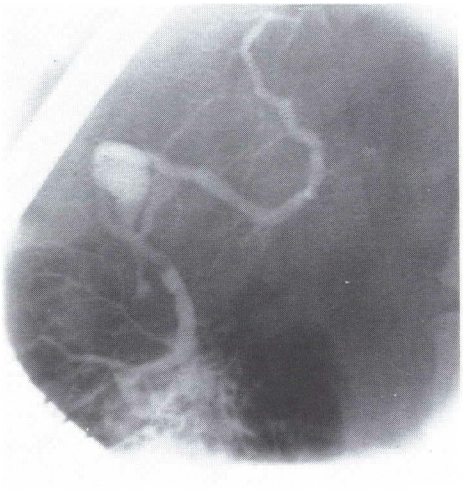

Figure 6
Transduodenal pancreatic duct cannulation showing also marked stenosis of lower end of common bile duct. At surgery, cholangiocarcinoma.

But, strict criteria for the diagnosis require absence of stones.

The etiology of the lesion is not known. It is evident that many cases represent one manifestation of a systemic disease process.[4,6,7] As demonstrated by two of our patients, serum immunoglobulin and anti-nuclear factor determinations indicate the presence of an auto-immune response by the patient. Perhaps the rare condition of stenosis of the sphincter of Oddi should be indicated as a component of sclerosing cholangitis.[9]

The diffuse involvement of the extrahepatic biliary tract, as well as intrahepatic involvement, make the ultimate outlook poor for the sclerosing cholangitis patient. However, our experience, as well as that reported by others, indicates that in many instances the disease is compatible with many years of life.[5] Therefore, all possible avenues of mechanical relief of obstruction by operative procedures should be provided. In addition, steroids should be administered when operative aid is not possible or is only incompletely effective.

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