CASE REPORTS

Congenital Absence of the Gallbladder and Cystic Duct Associated with Primary Carcinoma of the Common Bile Duct

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CONGENITAL absence of the gallbladder and cystic duct is a rare anomaly. Its association with primary carcinoma of the common bile duct has been described in a case reported by Robertson, Robertson and Bower.[1] A similar case is presented in the following report.

A 73-year-old white woman was admitted to the Toronto East General Hospital on December 25, 1964, with a one-week history of abdominal pain and jaundice. She had been well until a few months before, when there was a gradual onset of anorexia, weight loss and generalized weakness. A few days before admission she experienced a fairly sudden onset of mid-abdominal pain which was more severe on the left side and radiated through to her back. It was accompanied by vomiting and the development of jaundice. Examination on admission revealed a chronically ill elderly woman with jaundiced sclerae. Her abdomen was soft, with slight right upper quadrant tenderness. There were no abdominal scars.

Her past history revealed that 30 years previously she had been subject to repeated attacks of abdominal pain which was aggravated by certain foods. A cholecystectomy was planned but was cancelled because of a close friend's death following the same procedure. These attacks recurred infrequently for 20 years and then spontaneously subsided, and for the 10 years prior to this terminal episode she had no specific gastrointestinal complaints. She was a mild diabetic, controlled on tolbutamide for three years. She was hypertensive and had suffered a right hemiplegia in 1961 from which she had recovered well. She had never had an abdominal operation.

Laboratory examinations showed evidence of obstructive jaundice; the urine contained bile and no urobilin. The serum bilirubin level varied from 12 mg. % to 30 mg. %. The prothrombin time was 43 seconds (corrected with vitamin K given intramuscularly). The serum glutamic oxaloacetic transaminase (SGOT) was 600 units, the alkaline phosphatase was 80 King-Armstrong units, serum protein and cephalin-cholesterol flocculation (CCF) were normal, and her hemoglobin was 80%. An abdominal flat plate was normal. An upper gastrointestinal series showed an indentation in the base of the duodenal cap which was felt to be due to a prolapse of gastric mucosa. It was otherwise normal.

Her jaundice continued to deepen, and a laparotomy was performed on January 10, 1965. At laparotomy the anterior wall of the stomach and the omentum were adherent to the liver along the area of the gallbladder bed, but no gallbladder was palpable. The stomach and omentum were freed and the porta hepatis was visualized. The common bile duct was opened, releasing a small amount of white bile, and a probe was passed distally into the duodenum without any obstruction. Proximally, however, the lumen of the common bile duct was markedly narrowed by intraluminal tumour. This was particularly extensive toward the right hepatic duct. Graduated sounds were passed proximally and entered the left hepatic duct, and one arm of a T-tube was inserted. The other arm was placed in the lower portion of the common bile duct. A cholangiogram through the T-tube showed a normal common bile duct below the level of the straight arm of the T-tube, the dye readily passing distally into the duodenum. The proximal arm of the T-tube contained dye which entered the left hepatic duct and its branches. The right hepatic duct was not seen. No gallbladder or cystic duct were visualized. The pancreas, stomach and duodenum were normal to palpation.

Postoperatively her T-tube drained well and her serum bilirubin fell from 30 mg. % to 17 mg. %. She was discharged to a nursing home on February 25. She deteriorated in the nursing home and was readmitted on April 15 with severe jaundice, confusion and weakness. She died on April 21.

At autopsy, the body was that of a deeply jaundiced white woman. There was a recent healed abdominal scar, but no old scars were observed. A large amount of bile-stained ascitic fluid was present. The liver was slightly enlarged and on the right side there was a large subphrenic abscess which contained about 500 c.c. of purulent bile. Smaller abscesses were noted throughout the substance of the liver, particularly on the right side. Some of these abscesses were in relationship to the markedly distended bile radicles. The distal portion of the common bile duct and its opening into the duodenum were normal. Proximally, however, a firm white tumour was obstructing the common duct. This originated just proximal to the bifurcation of the common bile duct and extended well up into the substance of the liver, particularly on the right side where it had almost completely destroyed the right hepatic duct. No gallbladder, cystic duct or gallbladder fossa were seen. The location of the carcinoma indicated that it had not destroyed a pre-existing gallbladder or cystic duct. The T-tube had been removed immediately after death but its tract was present in the expected position. The pancreas and duodenum were normal. A few small nodules of metastatic carcinoma were present on the serosal surface of the stomach. The intestine contained clay-coloured stool. Microscopic sections (Fig. 1) of the tumour from the common bile duct showed an adenocarcinoma with closely packed glands that varied in size and shape. In some areas

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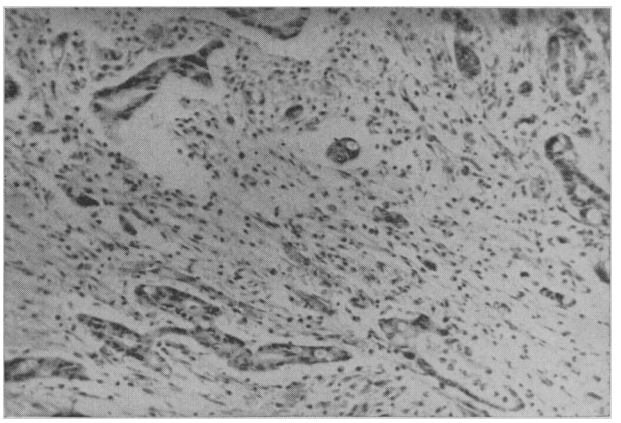

Fig. 1.—Photomicrograph showing adenocarcinoma of the common bile duct. (× 400.)

the tumour took on a scirrhous appearance with dense fibrosis. In others, the growth was more anaplastic without clear glandular differentiation. Sections of the liver showed numerous abscesses and adenocarcinoma similar to that in the common bile duct.

DISCUSSION

Congenital anomalies of the biliary tree are very common and are encountered in approximately 10% of necropsies, but congenital absence of the gallbladder and cystic duct in the presence of a normal hepatic and common duct system is one of the rare biliary tract anomalies. The first pathological report of this type of abnormality was published by Lémery *fils* in 1701.[2]

The incidence of congenital absence of the gallbladder and cystic duct has been reported in several series. Kirshbaum[3] found one case (0.03%) in a series of 3669 autopsies. McIlrath, Re Mine and Baggentoss[4] found 10 cases (0.04%) in a series of 26,531 autopsies. Malmström[5] found one case (0.042%) in a series of 2365 cholecystograms. Tallmadge[6] reported an incidence of 0.065% in a review of 18,500 autopsies. Seifert[7] reviewed the literature in 1962 and was able to find 143 published cases.

The congenital absence of a gallbladder may be explained in two ways: The hepatic diverticulum arises from the foregut to form the liver, the extrahepatic ducts and the gallbladder. The gallbladder and cystic duct normally arise from the diverticulum at the 3-mm. fetus stage as an outpouching, but the failure of the latter development would result in complete absence of the gallbladder and cystic duct. A second mechanism by which absence of the gallbladder might be explained comes into effect at the 15-mm. fetus stage, when the caudal portion of the liver diverticulum fails to develop a lumen and only a vestigial gallbladder represented by a white fibrous cord is seen.

The diagnosis of congenital absence of the gallbladder may only be made at laparotomy with cholangiography, or at postmortem examination. The incidence and mechanism of production of symptoms are not accurately known. When they occur, symptoms are typical of biliary tract disease with attacks of epigastric pain, nausea, vomiting, food intolerance and jaundice. The symptoms are usually due to complicating factors such as stones or hypoplasia of the common bile duct. However, some patients probably do have symptoms in the absence of complicating factors, and these may be due to the altered biliary drainage with pressure changes in the bile ducts and disturbed kinetics of the sphincter of Oddi. Latimer, Mendez and Hage[8] found a history of biliary tract symptoms in 34 of their 71 cases. In McIlrath, Re Mine and Baggentoss's[4] series of 10 patients eight were free of biliary tract symptoms, whereas Dixon and Lichtman[9] found that all of their 10 patients had symptoms suggestive of biliary tract disease. In the latter authors' review of 60 cases they found that 27% had common bile duct stones and that 48% were jaundiced. The average age of their patients was 46 years, and 73% of those over this age had symptoms similar to those of gallbladder disease.

It is believed that this is the second reported case of the association of congenital absence of the gallbladder with primary carcinoma of the common bile duct. The first case was reported by Robertson, Robertson and Bower[1] in a 45-year-old white man with a history of heavy alcohol intake. He had not had a previous history of biliary tract symptoms. At postmortem examination a primary carcinoma of the common bile duct with extension into the liver was found. No stones were present.

In view of the lack of reports linking these two conditions, it would appear that their association is fortuitous rather than of etiologic significance.

SUMMARY

A case of congenital absence of the gallbladder and cystic duct is described, in association with a primary carcinoma of the common bile duct. A brief review of the literature is presented. It is concluded that the association of these two conditions is fortuitous rather than of etiologic significance.

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