Caroli's Disease Associated with a Gastric Diverticulum

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ABSTRACT

Caroli's disease or communicating ectasia of the intrahepatic biliary tree is a rare disease with unknown aetiology. The coexistence of this along with the uncommon condition of a gastric diverticulum has never been reported before. A deficiency in the fibromuscular matrix of both the bile ducts and the gastric wall may explain why these two pathologies may coexist in a single patient.

INTRODUCTION

Communicating ectasia of the intrahepatic biliary tree, originally described by Caroli, is a rare clinical entity of obscure aetiology (1). Family studies have suggested an autosomal recessive mode of inheritance (2). It is often associated with autosomal recessive polycystic kidney disease and congenital hepatic fibrosis (3). We report the first case of Caroli's disease associated with the uncommon entity of a gastric diverticulum. This case raises certain interesting points on the aetiology of the disease.

CASE REPORT

In April 1977, a 34-year-old female of African descent presented with symptoms suggestive of reflux oesophagitis. She had no history of jaundice, fever, rigours, diarrhoea or constipation. Retrosternal pain and regurgitation of bitter fluid had occurred frequently over the preceding 2 years.

On physical examination, this well-looking, afebrile, anicteric patient had an enlarged liver, extending 7 cm below the right costal margin. It was firm, smooth and non-tender. No other masses were palpable and ultrasound imaging was not available at the time.

Laboratory studies revealed a haemoglobin of 13.2 gm/dl and a white cell count of 5.2×10^9. The liver function tests were normal. Barium meal showed a diverticulum on the posterior aspect of the stomach near the gastro-oesophageal junction (Fig 1).

At laparotomy, the diverticulum was excised but it was noticed that the enlarged liver was studded with numerous small compressible cystic protrusions. The extra hepatic biliary tree, pancreas and kidneys were normal. There was no gross evidence of cirrhosis or portal hypertension and a liver biopsy was done which confirmed Caroli's disease histologically (Fig 2).

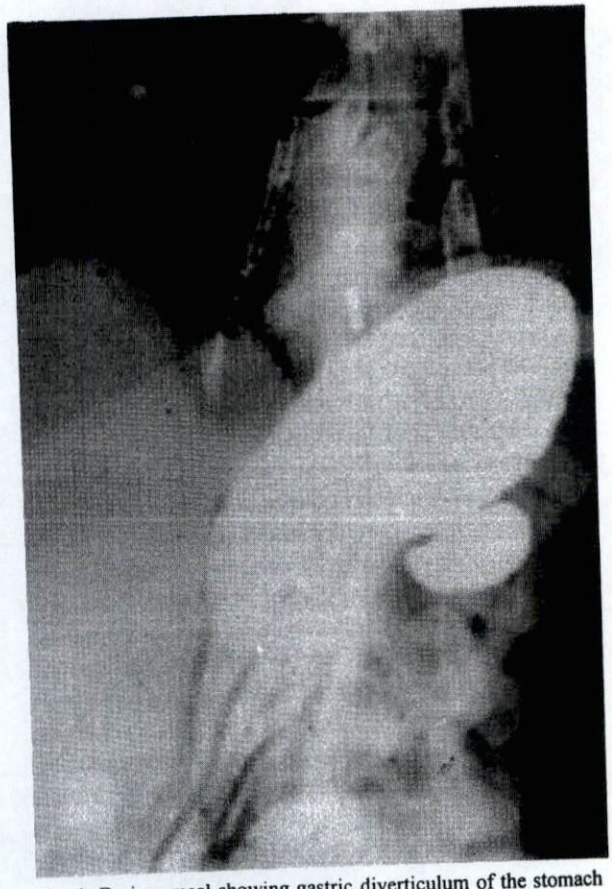

Fig. 1: Barium meal showing gastric diverticulum of the stomach

The patient recovered well post-operatively, had no further symptoms of reflux, and no clinical evidence of biliary tract disease subsequently. Intravenous pyelogram was normal. She has been followed in the out-patient clinic for the last 20 years and has developed no complications of liver disease.

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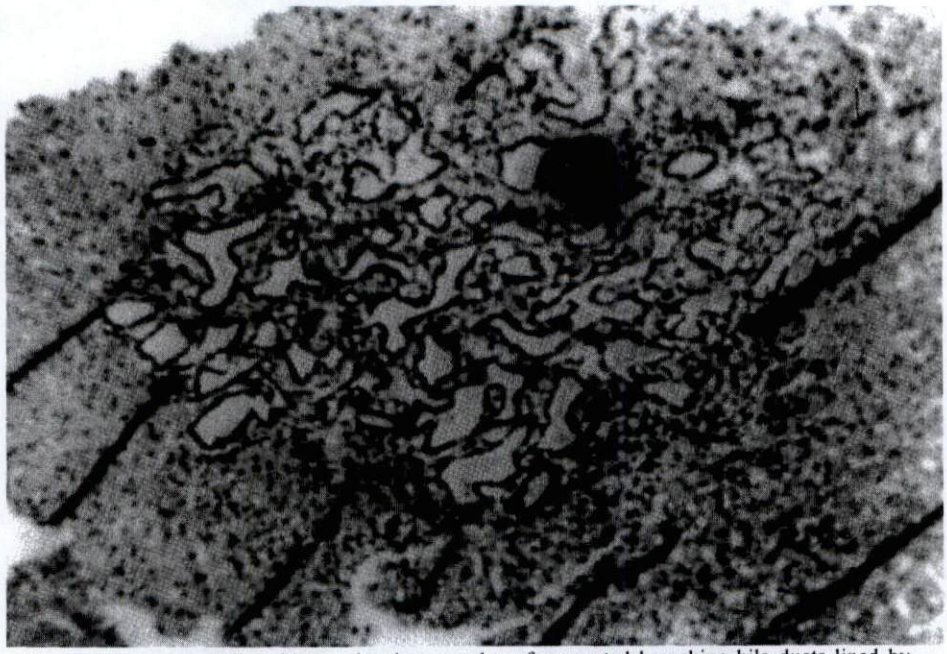

Fig. 2: Micrograph of liver biopsy showing complex of connected branching bile ducts lined by normal bile duct epithelium. The peripheral hepatocellular parenchyma shows no abnormalities including signs of cholestasis or inflammation. (Magnification x 40: H&E stain)

DISCUSSION

Congenital cystic dilation of the intrahepatic bile ducts (Caroli's Disease) is a rare clinical entity of dubious aetiology. Glenn and McSherry suggested that defective fibromuscular matrix of the duct wall was responsible for segmental or diffuse dilation of the biliary tree (4). The possible congenital origin of this anomaly was mentioned by Caroli in 1958 (5). An increased incidence of medullary sponge kidney and cysts of the pancreas and kidney are associated (5). It has also been associated with a cystic dilation of the extrahepatic bile duct (6).

This is the first reported association of Caroli's disease with a gastric diverticulum which is in itself a rare finding (7). It is possible that this is yet another manifestation of a fundamental weakness in the supporting matrix of smooth muscle in these patients. This is also one of the very few cases in which the diagnosis was made incidentally and before the patient developed symptoms related to the dilated intrahepatic ducts.

It would be advisable, therefore, to investigate thoroughly any patient with Caroli's disease for defects in the muscular layers throughout the gastrointestinal tract, as well as for renal and pancreatic disorders. The occurrence of a gastric diverticulum should also be an indication for investigating the patient for Caroli's disease, as one may then be able to make an early diagnosis and treat its complications.

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