distal pancreas, Roux-en-Y pancreaticojejunostomy had been recommended[8], and was performed.

The other surgical problem was the traumatic pseudocyst. Pseudocysts per se may have major complications, including haemorrhage, infection, rupture and compression of surrounding viscera[6]. Several techniques are available for treatment of pancreatic pseudocysts including percutaneous drainage, endoscopic drainage, surgical drainage and pancreatic resection[6]. Percutaneous drainage is very effective for treatment of traumatic pseudocysts in children, but is less so in adults[7]. In this case, with a known duct transection and a laparotomy already decided on, surgical drainage by posterior cystgastrostomy was performed.

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(Accepted 15 October 1991)

Budd-Chiari syndrome in association with polycystic disease of the liver and kidneys

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Keywords: Budd-Chiari syndrome; polycystic disease

Case presented to Clinical Section, 10 May 1991

The first reported case of polycystic disease of the liver and kidneys associated with Budd-Chiari syndrome is described.

Case report

A 47-year-old woman with a 7 year history of asymptomatic polycystic disease of the liver and kidneys was referred to the department of gynaecology because of ascites and a uterine mass. Investigations showed haemoglobin 10.1 g/dl (normal range 11.5-16.5), white cell count 5.6×10^9 (normal range $4.0\text{-}11.0\times10^9$), platelet count 289×10^9 (normal range $150\text{-}400\times10^9$). Serum vitamin B12, folate and ferritin were normal. The urea and electrolytes and liver function tests were normal with a plasma albumin of 44 g/l (normal range 35-50 g/l). The ascitic protein content was 58 g/l, and cytology and microbiological investigations were negative. Computerized tomography of the pelvis confirmed a uterine mass with inconclusive fibroid-like features. Laparotomy confirmed the fibroid and ascites but no other pelvic abnormality. Following hysterectomy she was well for one month before ascites recurred.

On examination a cystic liver was easily palpable below the umbilicus. Routine biochemistry remained normal but the EDTA clearance was 65 ml/min (predicted GFR 83 ml/min). The 24-h urinary protein excretion was normal (< 0.1 g/l). The serum alphafetoprotein was normal.

Further investigations were directed to the portal and hepatic vessels. Contrast venography demonstrated occlusion of the left renal vein (Figure 1) and the left hepatic vein (Figure 2).

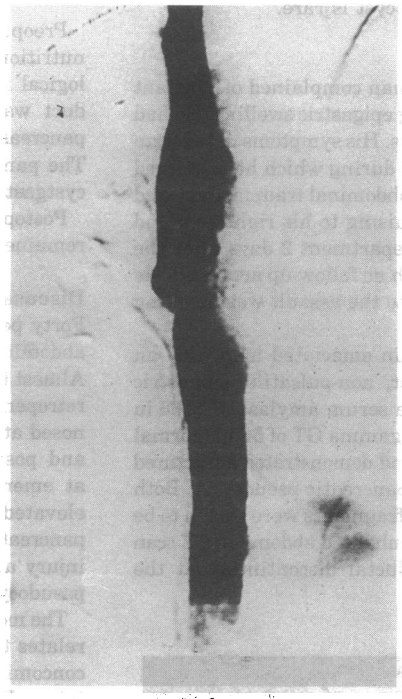

Figure 1. This inferior vena cavagram reveals the absence of the left renal vein suggestive of left renal vein thrombosis

Evaluation for a hypercoagulable state revealed that the antithrombin III, cardiolipin antibody, protein C and S, Ham's test and clot lysis time were all normal and the test for the lupus anticoagulant was negative. The serum fibrinogen was 5.2 g/l (normal range 2-4 g/l), and remained persistently elevated. Her platelet count was now noted to be elevated. Marrow aspirate was normal and cultures showed no excess of megakaryocyte colonies (technical problems precluded erythroid cell line assessment).

Treatment with diuretics, low sodium diet and warfarin failed to prevent reaccumulation of ascites and repeated paracenteses were necessary. Because the platelet count continued to rise (to between 500 and 700×10^9/litre) she was therefore started on aspirin and later hydroxyurea was given. However, hepatic synthetic function gradually deteriorated and she was referred to King's College Hospital for liver transplantation. Unfortunately she developed severe hypotension during the reperfusion phase of the operation from which she could not be resuscitated. Histology on her liver confirmed the diagnosis of the Budd-Chiari syndrome.

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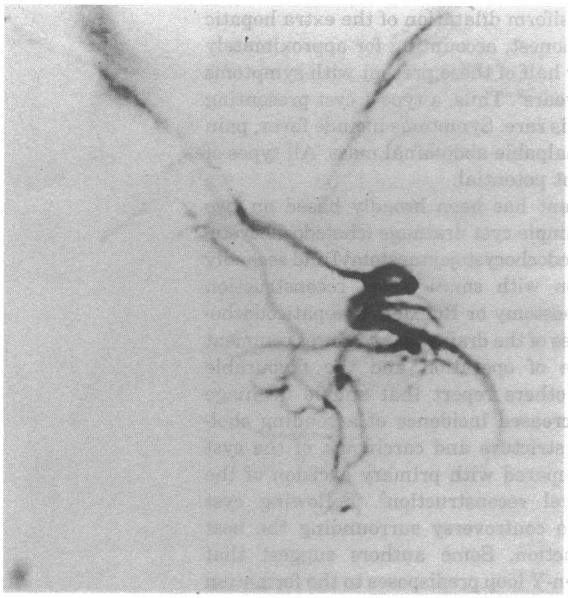

Figure 2. Venography of the left hepatic vein reveals a characteristic lace like spider-web pattern which is suggestive of hepatic vein thrombosis

Discussion

Diagnosis of the Budd-Chiari syndrome should prompt a careful search for a cause. Mitchell *et al.*[1], found a cause in 70% of 253 cases; the commonest being a hypercoagulable state. The responsible myeloproliferative disorders are often occult but the formation of spontaneous/autologous erythroid cultures in erythropoietin-poor media may be diagnostic[2,3]. In our patient the megakaryocyte cultures were negative, however the erythroid cultures were unsuccessful due to technical problems.

Symptoms caused by polycystic disease of the liver occur in only 10-15% of the cases[4], usually abdominal pain or fullness. Rarely obstructive jaundice, portal hypertension, cholangitis, or liver abscess may occur. Ascites is described usually with end-stage liver disease[5]. The Budd-Chiari syndrome has not been previously associated with polycystic disease although a case of inferior vena caval compression by hepatic cysts has been described[6].

We believe that this case of Budd-Chiari syndrome with polycystic disease of the liver and kidneys represents a true association rather than the coincidental occurrence of two rare disorders. It is probable that the numerous hepatic cysts compressed the left hepatic vein and left renal vein predisposing to thrombus formation. A hypercoagulable state secondary to the high serum fibrinogen and thrombocythaemia is likely to have been contributory.

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(Accepted 5 November 1991)

Choledochal cyst in adulthood

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Keywords: choledochal cyst; biliary tract; cystic dilatation

Choledochal cysts are rare congenital cystic dilatations of the biliary tract. They usually present in childhood.

Case report

A 44-year-old West Indian woman presented with a 6 month history of intermittent right upper quadrant pain, sweats and rigors. She had lost a stone in weight and complained of lethargy and malaise. There was no history of jaundice, steatorrhoea or bilirubinuria, and she was previously asymptomatic. Examination was unremarkable. She had a mild normochromic normocytic anaemia. Her serum alkaline phosphatase level was eight times normal and serum aspartate transaminase level was twice normal. Her serum bilirubin, amylase and tumour markers were normal.

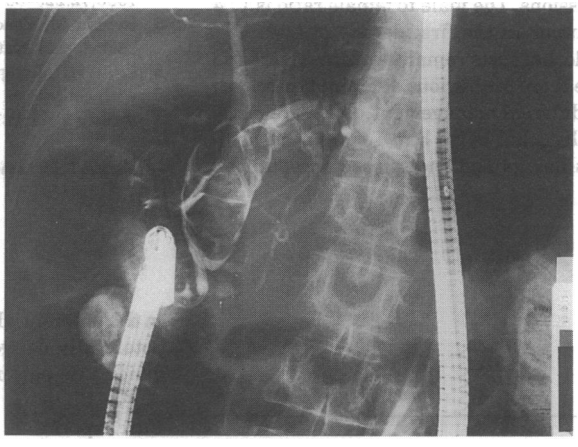

Figure 1. ERCP demonstrating the type 1 cyst with multiple filling defects representing the stones

Endoscopic retrograde cholangio-pancreatogram (Figure 1) and percutaneous transhepatic cholangiogram (Figure 2) demonstrated cystic dilatation of the common bile duct with gallstones in the gallbladder and common bile duct.

At laparotomy there was a fusiform dilatation of the common bile duct. This had a 5 mm thick woody hard wall and contained many large stones. The gallbladder also contained stones. There was no macroscopic evidence

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Case presented to Clinical Section, 13 December 1991

0141-0768/92/050297-02/$02.00/0