Ruptured giant liver cyst: a rare cause of acute abdomen in a haemodialysis patient with autosomal dominant polycystic kidney disease

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Abstract

Autosomal dominant polycystic kidney disease (ADPKD) is a common hereditary disorder. Although liver involvement is the most frequent extra-renal manifestation, serious complications due to liver cysts are very rare. We report the occurrence of an acute abdomen caused by massive haemoperitoneum resulting from rupture of a giant liver cyst in ADPKD. Data suggest that chronic anticoagulation therapy should be avoided where possible in the presence of a giant liver cyst.

Introduction

Autosomal dominant polycystic kidney disease (ADPKD) has multiple systemic manifestations, including the renal, hepatic, cardiovascular, cerebrovascular and gastrointestinal systems. Therefore, these patients may develop various unusual complications.

Liver involvement is the most frequent extra-renal manifestation of ADPKD. However, unlike renal cysts, which may cause infection or intracystic bleeding, serious complications due to liver cysts are very rare.** We report the fourth case in the literature of acute abdomen due to rupture of a giant liver cyst.

Case report

In December 1999, a 76-year-old man was admitted at the emergency department of our hospital with progressive abdominal pain. There was no nausea or vomiting. Micturition and defecation had been impaired for 24 hours. His medical history revealed end-stage renal disease (ESRD) due to ADPKD for which he had been receiving chronic haemodialysis for four hours three times a week since June 1999. His medication included oral anticoagulation (acenocoumarol) because of a polytetrafluoroethylene (PTFE) shunt in the left arm.

Upon physical examination, we saw a severely ill patient. His blood pressure was 75/50 mmHg, pulse rate 100 beats/minute and body temperature 37°C. Abdominal inspection revealed no distension; bowel sounds were sparse and inactive. Severe tenderness was noted in the upper abdomen with diffuse rebounding pain. No masses were palpable. Rectal examination revealed no abnormalities.

Relevant laboratory investigation included the following (normal ranges in brackets): C-reactive protein 210 mg/l (<10); haemoglobin level 6.1 mmol/l (8.5-11.0); platelet count 195 x 10⁹ /l (150-400); white cell count 12.2 x 10⁹ /l (4-10) with normal differential count. Serum urea amounted to 18.7 mmol/l (2.5-7.5); creatinine 848 mmol/l (75-110); sodium 142 mmol/l (135-145) and potassium 5.5 mmol/l (3.5-5.0). Liver biochemistry tests were abnormal: ALAT 198 U/l (<30); ASAT 137 U/l (<30); LDH 419 U/l (<320); γGT 27 U/l (<40) and alkaline phosphatase 56 U/l (<100).

Blood gas analysis revealed: pH 7.34; pCO₂ 39.8 mmHg; base excess -4.1 mmol/l; bicarbonate 20.7 mmol/l; pO₂ 86 mmHg; O₂ saturation 96%. Serum lactate was 1.70 (<2.4). As the International Standardised Ratio (INR) amounted to 7.9, 20 ml of prothrombin complex (coagulation factors II, VII, IX and X) was administered. Two days prior to admission, the INR had been 2.9.
Abdominal radiography showed no subdiaphragmatic air or distension of the bowel. Computed tomography of the abdomen disclosed multiple cysts in both kidneys and a large cyst with a diameter of nine centimetres in the right lobe of the liver (figure 1a). As the debris in the lower CT slices was visible on the dorsal site of the cyst (i.e., following gravity with the patient lying down during CT scanning), bleeding was suspected (figure 1b). In addition, free fluid was noted in the lower abdomen. No aortic aneurysm was present. Exploratory laparotomy revealed massive haemoperitoneum due to a ruptured giant liver cyst. Placing omentum over the ruptured cyst stopped the bleeding. There were no signs of intra-abdominal infection or infection of (other) liver cysts. The postoperative course was eventful with several serious complications (persistent haemodynamic instability, arrhythmias, bacterial pneumonia) eventually leading to the patient’s death four weeks after admission.

Discussion

ADPKD is one of the most common hereditary disorders. It is characterised by cyst formation in ductal organs, particularly in the kidney. Patients with ADPKD represent 3 to 10% of patients treated for end-stage renal disease (ESRD). Cyst formation may occur in the liver, pancreas, ovary, spleen and central nervous system. However, contrary to the present case, these patients usually develop complications associated with renal cysts, such as complaints related to the mass effect or more seriously intracystic infection or bleeding with gross haematuria and abdominal pain. The prevalence of hepatic cysts in ADPKD increases with age: 10% of patients 20 to 29 years of age compared with 75% of patients over the age of 60. Therefore, liver cysts are more likely to be present in the age group on haemodialysis. Women have more extensive hepatic cysts than men and pregnancy is strongly associated with extensive liver cysts. Female steroid hormones appear to influence liver cyst formation. Only 5 to 15% of patients with liver cysts develop symptoms related to the mass effect of cysts such as abdominal fullness, swelling, intermittent or continuous abdominal pain, dyspnoea due to elevation of the diaphragm, heartburn and change in bowel movements. Liver cysts can also become infected, presenting with fever and right upper quadrant tenderness. The incidence and complications of hepatic cyst haemorrhage in ADPKD patients on haemodialysis has not been well assessed but is probably rare. Patients with liver cyst bleeding may present with signs and symptoms similar to those in cystic infection: right abdominal pain and fever. To date, only three cases with haemoperitoneum due to rupture of a liver cyst have been described. In one case, the cyst was 11 cm, in the other two cases the size of the ruptured cyst was not mentioned. As in the present case, one patient was also on chronic anticoagulation therapy. Surgical treatment, i.e. marsupialisation of the cyst, was undertaken in two cases, whereas in the third case no surgery could be performed because of rapid clinical deterioration due to complicating abdominal sepsis and death. Of note, other serious complications

Figure 1a
Abdominal computed tomography revealing a massive subcapsular cyst with a diameter of 9 cm in the right upper lobe of the liver. The medial wall of the cyst contains a dense configuration, which obliterates the border of the cyst.

Figure 1b
In a lower coupe the borders of the cyst are clearly shown. The dense configuration caused by debris was visible on the dorsal site of the cyst (i.e., following gravity with the patient lying down during CT scanning), which is suggestive of intracystic bleeding.
related to liver cysts have also been reported in anecdotal case reports, such as portal hypertension\textsuperscript{11} and obstructive jaundice.\textsuperscript{14}

There is no accurate assessment of the influence of hepatic complications on the prognosis of haemodialysed patients. However, a review of 50 ADPKD patients on haemodialysis suggests limited influence of hepatic manifestations on their outcome,\textsuperscript{11,16} but death related to liver cyst complications has been reported.\textsuperscript{19} Our patient also died after he presented with acute abdomen and hypovolaemic shock due to massive haemoperitoneum.

Although not readily recognised by the authors as such, the size of the liver cyst may have been a risk factor for cyst bleeding.\textsuperscript{1} However, it remains unclear whether regular ultrasound monitoring of the size of asymtomatic liver cysts should be performed. Several surgical options for significantly enlarging liver cysts are available (i.e. cyst aspiration, cyst fenestration or marsupialisation, enucleation). However, the associated risks with these surgical interventions are probably higher than the risk of spontaneous life-threatening bleeding from a large liver cyst.\textsuperscript{12,13,16}

Another risk factor for bleeding in chronic haemodialysis patients is uraemic bleeding tendency, repeated systemic anticoagulation during dialysis procedures and/or the use of chronic oral anticoagulation therapy to prevent thrombosis of (graft) fistulas. On reviewing the patient’s charts, no careful evaluation was noted of the benefits and risks of the use of oral anticoagulation with regard to the presence of large liver cysts. Therefore, in the cases of large or enlarging liver cysts, we suggest that the need for oral anticoagulation should be weighed carefully against the risk of bleeding in these patients. The presence of a giant liver cyst, if not treated surgically as described above, may perhaps be regarded as a relative contraindication for chronic anticoagulation treatment. As such, Cinimino-Brescia fistulas should probably be preferred in these patients because of a significantly lower risk of thrombosis as compared with graft fistulas.\textsuperscript{17} In addition, heparin-free or regional anticoagulation haemodialysis should be considered in patients with giant liver cysts.

In conclusion, we describe a rare but life-threatening complication of ADPKD, i.e. rupture of a giant liver cyst with massive haemoperitoneum. Data suggest that asymptomatic giant liver cysts may be at significant risk of rupture. Therefore, if no surgical treatment of the cyst is performed, we feel that chronic anticoagulation therapy should be avoided if possible in these patients.

REFERENCES