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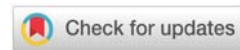
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Case Report

Severe polycystic liver disease: An unusual cause of chronic Budd- Chiari Syndrome

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Abstract

Autosomal dominant polycystic kidney disease is a hereditary disease, characterized by the development of cysts in the renal parenchyma with extra-renal manifestations. Liver damage is rarely a source of complications. A Budd-Chiari syndrome could occur following the compression of the supra-hepatic veins by the cysts. It is an exceptional mechanical complication.

We present a case of a 54 year old woman, with a diagnostic of an asymptomatic autosomal dominant polycystic kidney disease since childhood was admitted to our hospital due to significantly increased abdominal girth. The physical examination showed grade III ascites. A paracentesis for relief at admission disclosed an exudative fluid. A abdominal computed tomographic scan showed multiples cystic lesions in the kidneys and liver, with a large hepatic cyst responsible for compression of the suprahepatic veins and the inferior vena cava resulting in chronic Budd-Chiari syndrome. The treatment was radiological drainage followed by percutaneous sclerosis of cysts to alleviation of the compression. Unfortunately, the patient died a few days after an intraperitoneal cystic rupture.

Introduction

Polycystic liver disease represents the most frequent extra-renal manifestation of autosomal dominant polycystic kidney disease (ADPKD) [1,2]. It rarely causes complications. However, its symptoms are often related to mass effect with compression of the hepatic parenchyma and adjacent structures, including the vena cava, portal portal or bile ducts [3,4]. A Budd-Chiari syndrome could occur following the compression of the supra-hepatic veins by the cysts. This is an exceptional mechanical complication, but possible. In the literature, we found only a few reported historical cases [5-8]. We report in this article the case of a 54-year-old patient with ADPKD complicated by chronic Budd-Chiari syndrome revealed by ascites and we will provide a review of the literature.

Case report

A 54-year-old woman, she had four children, without any

history of abortion, with a diagnostic of an asymptomatic autosomal dominant polycystic kidney disease since childhood was admitted to our hospital due to significantly increased abdominal girth since four months in a context of apyrexia and of asthenia. She had no positive family history of ADPKD, and no other medical history. Her physical examination had objectified grade III ascites with a positive flow sign without an abdominal collateral circulation. At admission, a 15 Litre paracentesis improved abdominal discomfort and revealed an exudative fluid (protein level at 41 g/l). The serum albumin and ascites gradient was low (< 1.1g/dl). There were no obvious signs of spontaneous bacterial peritonitis (leukocytes were 18 elements/mm³), it was also negative for neoplastic cells.

Laboratory tests revealed the following results: polycythemia with a hemoglobin level at 19 g/dl, hematocrit raised to 56% with red blood cells at 6.8 T/L, a platelet level at 241,000/Ul. There was no renal insufficiency with a serum

creatinine level at 6.7 mg/l, serum albumin at 3.57 g/dl, ALT at 21 IU/L, AST at 30 IU/L, γ -GGT at 127 IU/L, alkaline phosphatase at 131 IU/L, serum total protein: 6,9 g/L, serum albumin: 3.9 g/L.

Abdominal computed tomography (Figure 1) was performed, showing multiples cystic lesions of variable sizes in the kidneys and liver, a massive hepatomegaly with a large hepatic cyst measuring 170×200×240 mm, causing compression of the supra-hepatic veins and the inferior vena cava. The diagnosis of chronic Budd-Chiari syndrome was retained. Abdominal ultrasound with Doppler of the supra-hepatic veins showed compression of the veins by the hepatic cysts without signs of endoluminal thrombosis. Echocardiography had objectified a slight diastolic dysfunction with a preserved ventricular ejection fraction, without any pericardial effusion. The upper digestive endoscopy had shown an aspect of extrinsic compression of the stomach, probably linked to the mass effect of the hepatic cysts, without notable macroscopic abnormalities.

The therapeutic options were percutaneous alcoholization or surgical treatment. The evolution was marked a few days after the diagnosis by the sudden onset of diffuse abdominal pain, with on physical examination showed a tachycardia a generalized abdominal defense and. A cystic rupture was suspected, and the patient was scheduled for an emergency laparotomy. Unfortunately, his death occurred a few minutes later.

Discussion

Autosomal Dominant Polycystic Kidney Disease (ADPKD) is the most frequent hereditary nephropathy, with an incidence of 0.25% to 1%, and a prevalence of 1/2500 in the general

population [9]. It is characterized by the development of renal cysts, associated with extra-renal manifestations, the most common being polycystic liver disease, present in nearly 94% of patients [10]. ADPKD is due in 80% of cases to the mutation of the PKD1 gene, located on chromosome 16p13, while in 5 to 10% of cases it is linked to a mutation of the PKD2 gene, which is located on chromosome 4q21- 22 [9]. Currently the GANAB gene mutation is reported to be responsible for ADPKD.

Liver cysts result from abnormal growth of the biliary epithelium (cholangiocytes), due to the persistence of embryonic biliary structures in the liver [11]. The proliferation of bile ductules results in the formation of biliary micro-hamartomas, also called Von Meyenburg complexes. These biliary micro-hamartomas can be subjected to progressive dilation, leading to the formation of hepatic cysts. The cystic epithelium thus retains the characteristics of the biliary epithelium, but with increased secretory and proliferative activities [5].

Unlike kidney damage, which ultimately leads to destruction of the renal parenchyma with the risk of kidney failure, liver damage is often asymptomatic [3]. However, in some cases the expansion of the cysts can lead to the compression of the neighboring structures, at the origin of the clinical manifestations [12]. Female gender and increased exposure to estrogen during the reproductive period are major risk factors for hepatic cyst growth, and consequent compressive complications [13]. Clinical presentation includes transient right hypochondrial pain, abdominal distension, early satiety, dyspnea and back pain related to hepatomegaly [1,14].

Ascites is an unusual clinical manifestation of hepatorenal polycystosis. In this case, ascites is usually observed in advanced stages of the disease. It preferentially develops when there is massive hepatomegaly. In the literature, we found only a few historical reported cases [5,7,8,15,16]. Several mechanisms may be incriminated. Firstly, the hypertrophy of the hepatic cysts can be the cause of compression of the inferior vena cava, or of the supra-hepatic veins responsible for chronic Budd Chiari syndrome, as is the case in our patient [5,7]. Secondly, some cysts developed in the hepatic hilum can compress the portal bifurcation responsible for portal hypertension (PH) which can lead to transudative ascites, especially if the patient is malnourished [17-19]. Finally, at advanced stages of the disease, the evolution can be done towards the development of fibrosis, see hepatic cirrhosis, with all its complications including ascites.

Therapeutic management depends on the symptomatology, extent, distribution and anatomy of the cysts. Percutaneous aspiration with alcoholic sclerosis can be offered to patients with dominant, large and symptomatic cysts [13]. Laparoscopic fenestration is particularly effective if there are a few large localized cysts. Its effectiveness is limited when the cysts are difficult to access or when they sit at the level of the dome of the liver [20]. When the cystic involvement is diffuse and disseminated, preserving little or no healthy parenchyma, liver transplantation becomes the best therapeutic choice [16], especially in patients with severe associated renal impairment requiring a renal transplant [21-23].

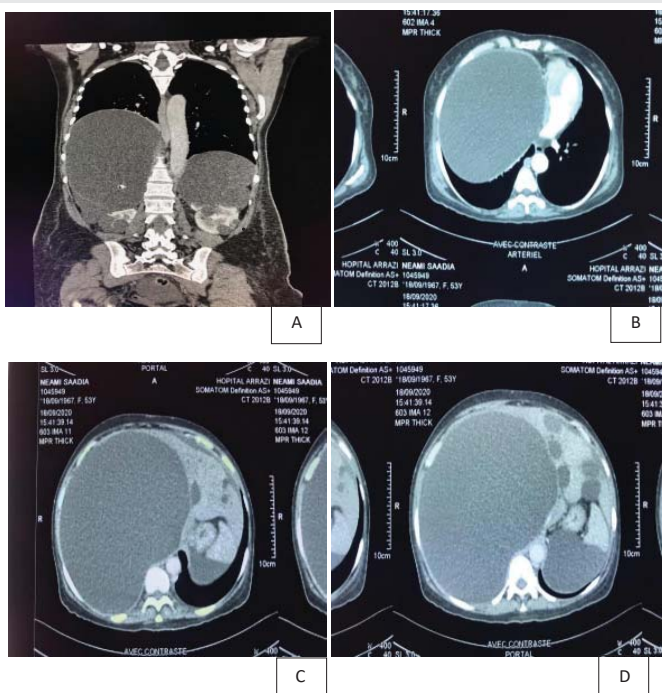


Figure 1: Abdominal computed tomography of the patient's abdomen revealing multiple liver cysts and, as a result, massive hepatomegaly severe (A), compression of hepatics veins (B), stomach (C,D).



Conclusion

In conclusion, ascites is an unusual complication of hepatorenal polycystosis. Several mechanisms can be incriminated, A Budd-Chiari syndrome could occur following the compression of the supra-hepatic veins by the cysts. An early diagnosis can be useful in a number of cases allowing a therapeutic management in order to avoid the evolution towards complications, which can engage the vital and functional prognosis of the patient.

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