Portal Hypertension Due to Multiple Hepatic Cysts in Autosomal Dominant Polycystic Kidney Disease: Case Report

Otozomal Dominant Polikistik Böbrek Hastalığında Karaciğer Kistlerine Bağlı Portal Hipertansiyon

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ABSTRACT Liver cysts are the most frequent extrarenal manifestation of autosomal dominant polycystic kidney disease. Usually, there are no symptoms related to liver cysts and no treatment is required. Symptoms, when they occur, include abdominal distention and pain, dyspnea, low back pain, and those caused by the development of complications such us cyst infection, hemorrhage, rupture, extrinsic compression of the portal vein and intrahepatic radicles, inferior vena cava, hepatic veins, or bile ducts. Extrinsic compression of the hepatic veins, portal vein and intrahepatic venules by extensive hepatic cysts is the most likely mechanism of portal hypertension at the polycystic liver disease associated with polycystic kidney disease. We are reporting a 67 year-old woman who had portal hypertension due to severe hepatic cystic disease associated with autosomal dominant polycystic kidney disease.

Key Words: Portal hypertension; autosomal dominant polycystic kidney

ÖZET Karaciğer kistleri otozomal dominant böbrek hastalığına en sık eşlik eden ekstrarenal bulgudur. Kistler genellikle semptoma neden olmaz ve tedavi gerektirmez. Kist enfeksiyonu, kist içine kanama, rüptür, portal ven ve intrahepatik dalları, hepatik ven, inferior vena kava ya da safra kanallarına dıştan kist basısı gibi komplikasyonlar nedeniyle abdominal distansiyon, ağrı, dispne ve bel ağrısı gibi semptomlar izlenebilir. Hepatik venlere, portal vene ve intrahepatik venüllere genişlemiş hepatik kistler tarafından oluşturulan bası polikistik böbrek hastalığına eşlik eden karaciğerin kistik hastalığında portal hipertasiyon gelişiminin olası mekanizmasını açıklar. Bu yazıda otozomal dominant polikistik böbrek hastalığı ile ilişkili ciddi kistik karaciğer hastalığı olan 67 yaşındaki hastada gelişen portal hipertansiyonu bildiriyoruz.

Anahtar Kelimeler: Portal hipertansiyon, otozomal dominant polikistik böbrek hastalığı

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Autosomal dominant polycystic kidney disease (ADPKD) is a hereditary, multisystemic disorder. The most common extrarenal feature of ADPKD is liver cysts originating from biliary ductal microhamartomas. Usually, there are no symptoms related to liver cysts and no treatment is required. Symptoms, when they occur, are caused by the mass effect of the cysts. They include abdominal distention and pain, dyspnea, low back pain, and those caused by the development of complications such us cyst infection, hemorrhage, rupture, extrinsic compression of the portal vein and intrahepatic radicles, inferior vena cava, hepatic veins, or bile ducts.

Portal hypertension may develop at these conditions: First congenital hepatic fibrosis or liver cirrhosis may lead to portal hypertension.[1] Second, the obstruction of portal venous flow due to mechanical compression of portal vein or intrahepatic portal radicles by liver cysts.[1,2] And the third disorder that may lead to portal hypertension is hepatic venous outflow obstruction due to compression of hepatic veins, or the inferior vena cava, or both by one huge cyst or multiple liver cysts that can be demonstrated by imaging techniques.[1,2]

The diagnosis of portal hypertension at the presence of liver cysts can be established with imaging techniques. Congenital hepatic fibrosis and liver cirrhosis can be diagnosed by liver biopsy. Computerized tomography scanning and ultrasound of the abdomen is useful in showing kidney cysts, liver cysts, extrinsic compression of portal vein, hepatic veins, inferior vena cava and in detecting ascites.[3] Color Doppler Ultrasound and MRI is useful in establishing the patency of flow in the inferior vena cava, hepatic and portal veins and in detecting abnormal collateral veins.

CASE REPORT

A 67 year old woman had been admitted to our hospital because of abdominal discomfort, bloating and pain one year ago. She had been diagnosed with autosomal dominant polycystic kidney disease, polycystic liver disease, nephrolithiasis and splenomegaly by abdomen CT. There were multiple cystic lesions with the largest 50 mm in diameter at the right kidney and 40 mm in diameter at the left kidney. An a stone was visible at the left kidney. The liver was enlarged (200 mm in length) and there were multiple cystic lesions with the largest 60 mm in diameter. The length of the spleen was 150 mm. The liver enzymes of the patient were found normal. The BUN value was 72 mg/dL (normal:17-43 mg/dL); serum creatinin 2.2 mg/dL (normal: 0.8 – 1.4 mg/dL), hemoglobin 10.9 g/dL (normal: 12.6 – 17.4 g/dL), hematocrit 31.2% (normal: 37-51%), white blood cell count 3200/μL (normal: 4.500-11.000 /μL) , and platelet count 159.000/μL (normal: 150.000-400.000/μL) one year earlier.

The patient applied repeatedly with the rising complaints of abdominal pain, discomfort, bloating and constipation. Two months ago she underwent right percutaneous nephrolithotomy and was treated for complicated urinary tract infection at another hospital.

She was admitted to the hospital for further evaluation. Hepatomegaly, splenomegaly and swelling at the left side of the abdomen -under left costal arc- were established on physical examination. The AST, ALT and alkaline phosphatase levels were found normal. BUN was 65 mg/dL, creatinine was 2.3 mg/dL, and serum albumin was 2.9 g/dL (normal: 3.5-5.0 g/dL). Hemoglobin was 7.1 g/dL, hemotocrit was 21.2%, white blood cell count 3800/μL, and platelet count 126 000/μL. INR was normal. Hepatitis B and C serologies were negative. Transferrin saturation, ferritin, and serum ceruloplasmin, were within normal limits.

The abdominal CT revealed enlarged bilateral kidneys including multiple cysts in different dimensions showing typical appearance of adult polycystic kidney disease (Figure 1). The liver is

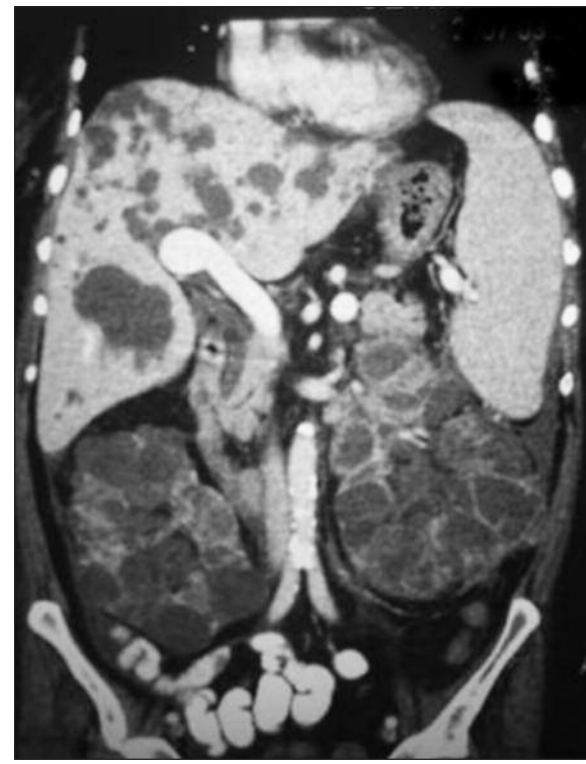

FIGURE 1: Bilateral enlarged kidneys containing multiple cysts in different dimensions having typical appearance of adult polycystic kidney disease.

containing multiple cysts with the largest 60 mm in diameter, the right lobe length is found 230 mm, the remaining hepatic parenchyma is found normal and the portal vein is dilated (20 mm in diameter) (Figure 2). The spleen's length is 170 mm and the splenic vein is found dilated too (15 mm in diameter). Additionally, CT revealed minimal ascites. No esophageal varices were established on endoscopi.

Doppler ultrasound examination showed increased volume flow at portal, splenic and superior mesenteric veins. Portal vein (21 mm) and splenic vein (15 mm) calibres were enlarged correlated with portal hypertension. There was no thrombosis within the portal vein.

DISCUSSION

ADPKD is associated with extrarenal manifestations such as liver cysts, intracranial aneurysms, colonic diverticula and cardiovascular abnormalities like mitral-valve prolapse, mitral regurgitation, aortic insufficiency, aortic aneurysms and tricuspid regurgitation. Cysts may also occur occasionally in the pancreas, ovaries and spleen.[4]

Development of portal hypertension in autosomal polycystic kidney disease is not frequent. It appears with splenomegaly, ascites, and variceal bleeding; established. If congenital hepatic fibrosis or liver cirrhosis is suspected liver biopsy should be performed.

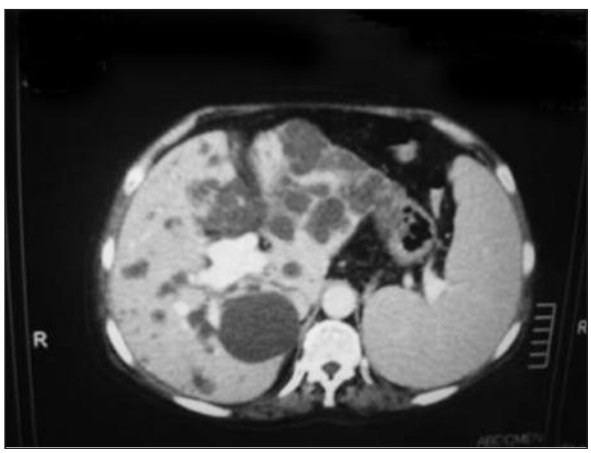

FIGURE 2: Multiple cysts in liver with the largest 60 mm in diameter around the portal vein. Portal vein is widened (20 mm in diameter).

In our patient we excluded liver cirrhosis on the basis of ultrasonographic evidence of normal liver parenchyma and on the lack of etiological factors such as hepatitis B or C virus infections, alcoholism, hemochromatosis, Wilson's disease, primary biliary cirrhosis.

Congenital hepatic fibrosis is an invariable feature in autosomal recessive polycystic kidney disease. Portal hypertension associated with congenital hepatic fibrosis at autosomal dominant polycystic kidney disease patient recognize before 25 years of age.[1] Usually there are non or just a few cysts in the liver. It is established by liver biopsy and cystic enlargement of the segmental bile ducts may be observed on CT or ultrasound imaging. We did not suspect Congenital Hepatic Fibrosis because of her advanced age, normal liver enzymes and the presence of multiple cysts. Because of the large multiple cysts in her liver we avoided biopsy which may cause cyst rupture, hemorrhage and cyst infection.

Hepatic venous outflow obstruction, due to compression of hepatic veins or the inferior vena cava or both, should be suspected if there are huge cysts in liver. Our patient has multiple cysts as large as 60 mm in diameter. CT scanning did not show compression to the inferior vena cava but hepatic vein compression by multiple cysts could be responsible for portal hypertension. CT scanning revealed multiple large cysts around the portal vein also. We couldn't demonstrate an obvious obstruction of portal venous flow due to mechanical compression on CT imaging, but compression on intrahepatic portal radicles leading to portal hypertension is also possible.

As a rare complication, portal hypertension in autosomal dominant polycystic kidney disease due to compression of portal vein, venules and hepatic veins by extensive liver cysts has been reported previously, to the best of our knowledge, in 12 cases.[1,2,5-14] We are reporting this case to manifest that the liver cysts may lead to portal hypertension in ADPKD patients so that in pursuit of the patients, the relation between liver cysts and hepatic and/or portal veins must be evaluated in detail with imaging techniques. Besides, symptoms or evidences suggesting portal hypertension should be taken into consideration during evaluation of ADPKD patients.

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