

A Case of Patent Ductus Venosus Complicated with Tumor-Like Lesions of the Liver

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Patent ductus venosus (PDV) is a very rare congenital vascular anomaly of the liver. We report a 17-year-old male patient presenting with elevated liver enzymes and multiple liver masses. Physical examination was normal except for pectus excavatum deformity. Ultrasonography revealed multiple tumor-like lesions of the liver and a dilated portal vein with mild splenomegaly. Liver biopsy obtained from liver parenchyma revealed minimal hepatocellular damage. PDV was diagnosed by CT (computed tomography) portography. The radiologic findings of chronic liver disease and multiple liver masses were attributed to presence of PDV. Although it is very rare, PDV might be a diagnostic possibility in patients with mass lesions in the liver.

Key word: liver

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INTRODUCTION

The ductus venosus is an embryological porto-systemic shunt that connects umbilical vein and inferior vena cava. Ductus venosus allows oxygenated blood to flow from the placenta to the right atrium, bypassing the liver during fetal life. It closes after birth with postpartum hemodynamic changes. Persistent patency of ductus venosus is extremely rare. It is usually considered as a congenital deformity or an acquired condition resulting from liver biopsy or cirrhosis. Familial cases have been reported in the literature, suggesting that patent ductus venosus (PDV) has a recessive genetic trait.¹ PDV is diagnosed incidentally or in subjects presenting with hepatic encephalopathy, hypoxemia or hypoglycemia.² It can be also diagnosed with cardiac defects or hypoxia due to pulmonary arteriovenous shunting in the infantile period characterized with intrapulmonary arterial dilation. Elevated liver enzymes and level of ammonia or impaired liver func-

tion tests may be found. Anomalous vascular anatomy and mass lesions resembling focal nodular hyperplasia may be found radiologically. Hyperplastic response of the liver parenchyma to differential blood flow was put forward to explain the arising of nodular appearances.³

We report a patient with PDV complicated with tumor-like lesions in the liver and associated with pectus excavatum deformity.

CASE REPORT

A 17-year-old male patient was referred to our medical center for further evaluation of his chronic liver disease and liver mass lesions. There was a basically mild elevation of liver injury enzymes for two years; and hepatomegaly, dilated portal vein and tumor-like mass lesions were found upon sonographic examination.

On admission to our hospital, he had no complaints. He had no history of alcohol abuse, hepatotoxic drug use or herbal medicine intake. There was no family history of hepatic disease. He was a thin, slender male patient (height 195 cm, weight 85 kg). There was not any other positive finding upon physical examination, including the peripheral signs of chronic liver disease, except for pectus excavatum deformity. Laboratory parameters, including viral serology, and autoimmune and metabolic tests are shown in Table 1. Because of the increased 24-hour urinary copper excretion and minimally decreased serum ceruloplasmin levels, hepatic copper concentration was determined by atomic absorption spectrophotometry, which was normal. Slit lamp examination revealed no Kayser-Fleischer ring. Antinuclear antibody was positive at 1/80 dilution, and serum immunoglobulin levels for G and M were normal. A liver biopsy revealed minimal hepatocellular damage with normal liver architecture. In addition, there was no any other findings reported such as interface hepatitis or plasma cell infiltration.

Hepatobiliary ultrasonography showed nodular appearance, heterogeneity and multiple hypoechoic areas in liver parenchyma. CT portography was performed for evaluation of enlarged portal vein and splenomegaly. Nodular appearance of the liver was consid-

ered, so magnetic resonance imaging (MRI) of the liver was planned. These imaging procedures revealed an enlarged left lobe, multiple nodular areas—the biggest of which measured 4.6x3.3 cm in size (Figure 1)—and patent ductus venosus between portal vein and inferior

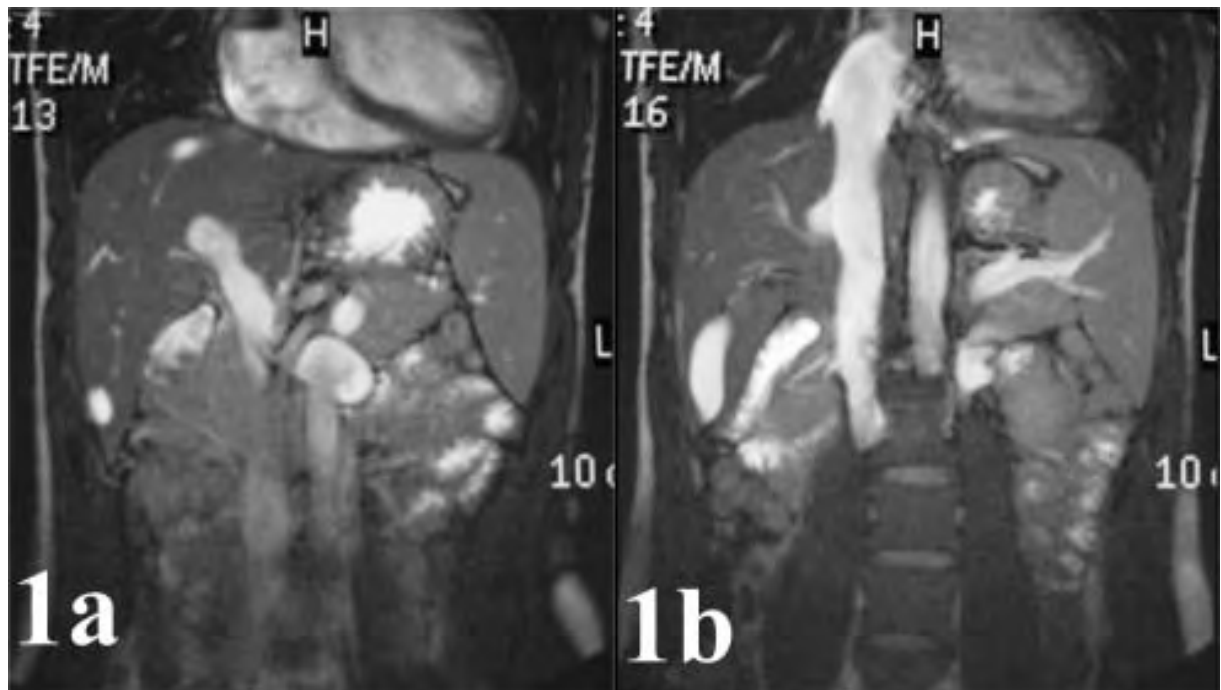
vena cava (Figure 2). The ultrasonographic appearances of chronic liver disease and suspected liver masses were attributed to presence of PDV.

The patient has no evidence of liver dysfunction. No surgical or vascular interventional procedure was per-

Table 1. The laboratory parameters of the case

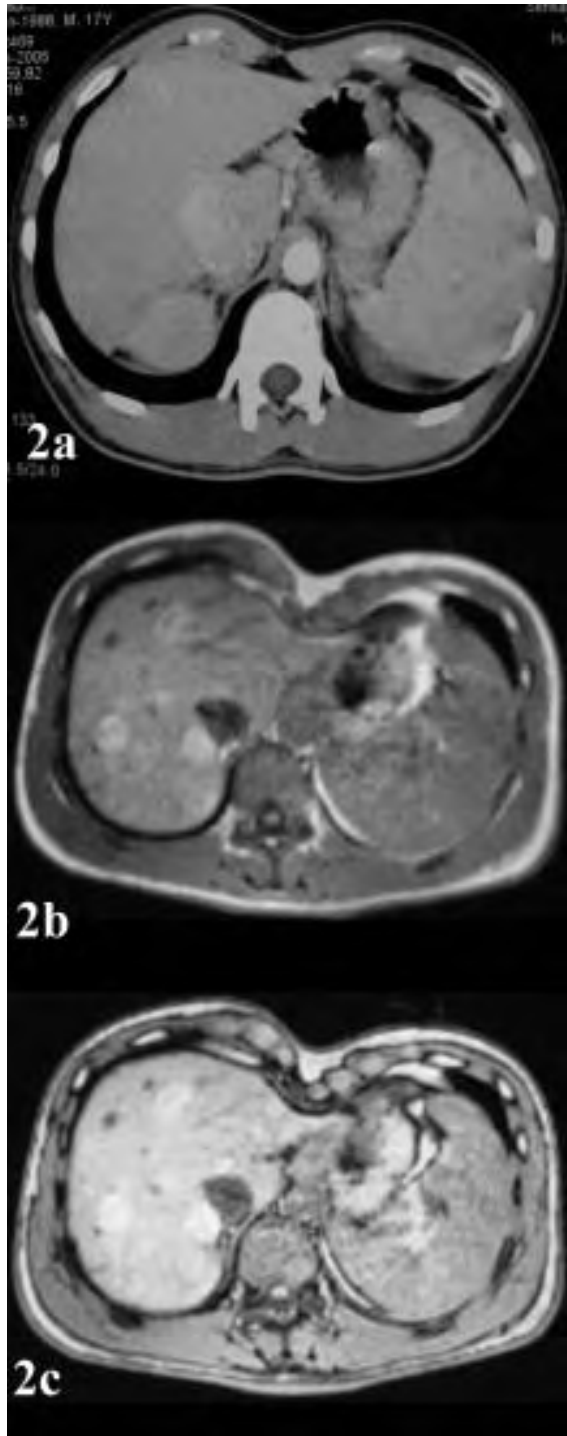
Parameter	Result
Hemoglobin (12–18 gr/dL)	14.9
WBC (3.6–10 x10 ³ /uL)	5.9
Platelet (150–450 x 10 ³ /uL)	190
Alanine transaminase (5–40 U/L)	68
Aspartate aminotransferase (8–33 U/L)	60
Gamma-glutamyl transpeptidase (5–40 U/L)	56
Alkaline phosphatase (35–129 U/L)	120
Total bilirubin (0.1–1.2 mg/dl)	1.82
Conjugated bilirubin (0.0–0.3 mg/dl)	0.59
Total protein (6–8.7 mg/dl)	6.7
Albumin (3.2–4.8 mg/dl)	3.8
Prothrombin time (sec)	15.5
HBs Ag	Negative
Anti-HBs	Positive
Anti-HCV	Negative
Antinuclear antibody	Positive
Antimitochondrial antibody	Negative
Anti-smooth-muscle antibody	Negative
Anti-liver-kidney microsomal Ab	Negative
24-hour urinary copper excretion (2–80 mg/day)	611
Ceruloplasmin (22–58 mg/dl)	21.2
Hepatic copper (1.7–32.4 mcg/gr)	18.2 mcg/gr
Alpha fetoprotein (0–5.8 IU/ml)	1.75

Figure 1. Coronal magnetic resonance angiography planes of liver showing the portal vein (1a), ductus venosus and inferior vena cava (1b)



formed for PDV in this phase. He was an active sportsman and exercise tolerance was normal. So therapy was not needed for the pectus excavatum deformity. He was discharged to be followed up and is doing well.

Figure 2. Axial computed tomography planes (2a) and magnetic resonance imaging (2b, 2c) showing multiple tumor-like lesions



DISCUSSION

Ductus venosus is normally closed two weeks after birth. As the pressure of the umbilical vein falls in the first few minutes of life, functional closure occurs. True obliteration of ductus venosus is completed in 15–20 days.⁴ Ductus venosus may remain patent because of increased vascular resistance caused by a poorly developing intrahepatic portal system.⁵

Our patient was referred to our department with the suspected liver mass and possible hepatocellular carcinoma. Viral etiological factors for hepatocellular carcinoma were negative, and alpha fetoprotein level was normal. Hepatocellular carcinoma generally arises from cirrhotic conditions. Cirrhosis was excluded with the normality of hepatic architecture in liver biopsy and liver functions. Unless there is confirmation of cirrhosis with histopathological examination, the provisional diagnosis of cirrhosis may be misleading. In this case, we emphasized that histological examination is a “sine qua none” in the diagnosis of liver cirrhosis, and patent ductus venosus may exceptionally cause a false image of chronic liver disease.

PDV may mimic the liver masses. In the literature, cases of tumor-like lesions of liver associated with portosystemic venous shunt and congenital absence of portal vein have been reported very rarely.^{6–9} The mechanism of nodularity and heterogeneity with tumor-like appearance were not understood clearly. In theory, differential blood flow, hyperplasia of well-perfused areas, atrophy of ischemic areas, vascular anomalies or thromboses were hypothesized.³

The diagnosis of patent ductus venosus can be made with ultrasonography. But in our case, chronic parenchymal liver disease was diagnosed by more than one radiologist in other centers upon ultrasonographic examinations performed. CT portography and MRI clearly visualized the patent ductus venosus in our patient.

The management and treatment of PDV is controversial. Treatment of patients with preserved liver function is generally supportive.¹⁰ Our case was in this group, thus any therapeutic procedure was not instituted. He is being followed up for one year and is doing well; he needs no supportive treatment. In cases with hepatic deterioration, therapeutic interventional or surgical approaches are required such as coil embolization of channels and surgical ligation of shunt vessel or resection of involved area of liver. In congenital PDV, an intrahepatic portal system is accepted to be hypoplastic, so liver transplantation may be the first choice of treatment.¹⁰

Although it is very rare, when a mass lesion in the liver is encountered, PDV might be a cause of this lesion. Association with other congenital deformities reflects the genetic basis of disease.

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