Idiopathic extensive peliosis hepatis treated with liver transplantation

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Abstract A 50-year-old Danish man, who neither had wasting disease nor was taking steroid-containing drugs, complained of abdominal distension, due to a markedly enlarged liver. Percutaneous needle biopsies were taken from the liver, and the findings gave suspicion of a neoplastic tumor. Because of reduced liver function and treatment-resistant ascites, he underwent liver transplantation without a definite preoperative diagnosis. The resected liver weighed 2900 g, and almost all of the parenchyma was destroyed and replaced by multicystic blood-filled spaces, diagnosed as extensive peliosis hepatis complicating liver cirrhosis. Extensive peliosis with liver cirrhosis is a rare condition. Only two cases, caused by contraceptives and treated by liver transplantation, are reported in the English-language literature. We could find no cause other than alcohol abuse lasting several years in this patient, and classified the present case as idiopathic extensive peliosis hepatis. Although scarce subjective findings and misleading liver biopsies made an exact diagnosis difficult, an orthotopic liver transplantation was the only treatment for such complicated peliosis hepatis.

Key words Peliosis hepatis · Liver cirrhosis · Liver transplantation

Introduction

Peliosis hepatis is a rare condition, showing multiple cystic blood-filled spaces through the liver parenchyma. It is found in conjunction with wasting conditions; for example, tuberculosis, disseminated malignancies, hematological disorders, and human immunodeficiency viral infection, and in renal transplant recipients. The cause is obscure; however, the administration of drugs, especially androgenic-anabolic steroids and contraceptives, is worth noting. The clinical presentation is variable, ranging from asymptomatic cases discovered at autopsy to progressive cases with liver failure or intraabdominal hemorrhage. In such advanced cases, patients have the risk of progressing to fatal status without appropriate diagnosis and adequate procedures. In this report, we describe a patient who underwent orthotopic liver transplantation due to extensive peliosis hepatis complicating liver cirrhosis.

Case report

A 50-year-old Danish man visited a public hospital with complaints of abdominal distension, although he had no history of the disorder. He had taken neither steroid-containing nor immunosuppressive drugs, but had been addicted to alcohol for several years. Abdominal ultrasonography and computed tomography (CT) showed massive ascites and multiple cystic processes in the enlarged liver, from which percutaneous needle biopsies were taken. The histopathological findings gave suspicion of endocrine tumor or epithelioid hemangioendothelioma.

He was referred to our hospital 3 months after his first consultation for additional examination and treatment. On physical examination, jaundice in conjunctiva and moderate ascites were recognized. His enlarged liver was palpable and multinodular, and extended to the umbilicus. The blood count showed anemia with hemoglobin (Hb) 6.3 mmol/l (range, 8.0–11.0 mmol/l), and no abnormality in differential white blood count. Decreased albumin 24.4 g/l (36.6–48.2 g/l), elevated total bilirubin 293 mmol/l (4–22 mmol/l), and slightly elevated liver parameters were recognized. Monoclonal gammopathy, diabetes mellitus, and renal dysfunction were not observed. There was no evidence of hepatitis B or C, human immunodeficiency, or cytomegalovirus infection, or of any bacterial infection. Systemic check-ups did not detect other malignancies. An abdominal
CT with contrast showed that the enlarged and mottled stained liver occupied the right upper abdomen, and was compressing adjacent organs (Fig. 1a). Massive ascites was also recognized (Fig. 1b).

Due to progressing liver failure and continued ascites, even after intensive treatment with diuretic drugs, the patient was registered on a liver transplant waiting list without a definitive diagnosis. He underwent orthotopic liver transplantation 2 months after registration.

The enormously enlarged liver occupied the upper abdomen and weighed 2900g. Irregular multiple nodularity was found to be due to blood-filled cysts of varying size (Fig. 2a).

On macroscopic examination, all of the liver parenchyma, except for minor subcapsular areas, was destroyed and had been replaced by spongy, hemorrhagic tissue; the individual cysts measured up to 15mm, and several were thrombosed.

Microscopically, severe dilatation of the vessels and sinuses was found, and large blood-filled cystic spaces and lakes lined with hyperplastic endothelium had formed. Some areas were hemorrhagic, and signs of new ruptures were found (Fig. 2b, 2c). The surrounding tissue showed a marked reactive change in all components, bile duct proliferation being especially dominant. The remaining liver parenchyma was cirrhotic, with no finding of alcoholic hepatitis. No tumor was identified.

Discussion

Peliosis hepatis is a rare and specific entity of liver disease characterized by sinusoidal dilatation and the existence of multiple blood-filled cystic spaces. Cases of peliosis hepatis before the 1970s were mainly reported as found at autopsy. Later, the described cases conventionally were found in conjunction with tuberculosis, disseminated malignancies, and hematological disorders. Recently, the entity has been found in connection with human immunodeficiency virus infection, after kidney transplantation, and associated with the administration of oral contraceptives. The present patient had no clinical evidence of chronic wasting disease, there was no history of ingestion of steroid-containing drugs or immunosuppressive drugs, and there were no signs of hematological disorders, viral or bacterial infections, or any malignancies. Because the histopathological findings of the cirrhotic liver did not show alcoholic liver damage, in spite of the patient's alcohol addiction, we recognized this case as idiopathic peliosis hepatis.

The clinical manifestations of peliosis hepatis are various, ranging from asymptomatic cases recognized coincidentally during liver checkups to fatal outcomes, such as complications of liver cirrhosis or rupture in the peliosis. Hepatomegaly, icterus, and fever are the main and characteristic symptoms in extensive peliosis hepatis. Our patient presented with hepatomegaly and icterus, but not fever. The clinical course also has variations; cases induced by drugs may become alleviated or disappear when the medication is discontinued, while cases associated with infection and liver abscesses can be healed by the intensive administration of antibiotics. Regardless of the treatment, the liver condition may get worse, progressing to liver cirrhosis or rupturing, with severe intraabdominal bleeding. In such cases, peliosis hepatis is life-threatening, and appropriate actions should be taken.