**Case Report**

**Peliosis hepatis complicating pregnancy: A rare entity**

Muhammad Osama Butt¹, Nasir Hassan Luck¹, Syed Mujahid Hassan¹, Zaigham Abbas¹, Muhammed Mubarak²

¹Departments of Hepatogastroenterology, Sindh Institute of Urology and Transplantation (SIUT), Karachi - 74200, Pakistan
²Departments of Histopathology, Sindh Institute of Urology and Transplantation (SIUT), Karachi - 74200, Pakistan

**ABSTRACT**

Peliosis hepatis (PH) is a rare, benign condition of the liver characterized by the presence of blood-filled lacunar spaces in the parenchyma. It usually has a chronic presentation and is a rare cause of portal hypertension reported in adult patients. Its etiology is diverse and ranges from infectious agents to tumors to toxic substances and anabolic steroids; however, the cause remains unclear in 25–50% of patients. Similarly, the symptomatology and imaging findings are diverse. Biopsy is the definitive test to diagnose the condition. Herein, we present a case of a young female presenting in her seventh month of gestational amenorrhea with signs of portal hypertension and subsequently diagnosed to have PH. She was managed conservatively and delivered her baby normally. Later, she presented with spontaneous bacterial peritonitis and hepatic encephalopathy and developed hepatorenal syndrome. She later succumbed to her illness. The condition should be kept in the differential diagnosis of the atypical liver masses and liver diseases causing portal hypertension.

**Key words**: biopsy, liver, peliosis hepatis, portal hypertension, pregnancy

**INTRODUCTION**

Peliosis hepatis (PH) is a rare, benign condition characterized by the presence of blood-filled lacunar spaces in the liver. It usually has a chronic presentation and is a rare cause of portal hypertension reported in adult patients. There are diverse etiologies of this condition; however, the cause remains unclear in 25–50% of patients.[1-5] The pathogenesis and management are still controversial. Prognosis is largely determined by the underlying cause and is generally guarded.[6-10]

Herein, we present a case of a young female presenting in her seventh month of gestational amenorrhea with signs of portal hypertension and subsequently diagnosed to have PH.

**CASE REPORT**

A 20-year-old female, primigravida, presented in the seventh month of her gestational amenorrhea with complaints of fever, abdominal pain and distention for 1 month. Pain was dull, aching and continuous, mostly in the upper abdomen. She had no past medical history of any illness and no previous surgical or blood transfusion history. There was no history of oral contraceptive pill usage or use of any illicit drugs.

Her blood picture showed hemoglobin (Hb) of 7.5 g/dL and platelets of 97,000/mm³. Her total serum bilirubin was 2.9 mg/dL with a direct component of 1.5 mg/dL. Alkaline phosphatase was 510 IU/L with alanine aminotransferase of 316 IU/L. Ultrasound of the abdomen revealed liver of 19.5 cm with coarse echotexture. Portal vein (PV) was of 1.1 cm in diameter and spleen was 15 cm with the presence of mild ascites. Left lobe of the liver was normal. Ascitic fluid analysis showed low protein ascites. Renal function was normal. She was further worked up with computerized tomography (CT) abdomen, which showed marked hepatomegaly with heterogeneous enhancement of liver parenchyma. A large
hypodense lesion was identified along the dome of the liver with multiple linear and branching structures. Multiple tiny enhancing nodules were identified in the right lobe of the liver measuring between 1 and 2 mm. The hepatic veins were attenuated and were not clearly seen and the intrahepatic portion of the inferior vena cava (IVC) was also compressed; however, it was showing normal enhancement. No evidence of IVC thrombosis was noted. The spleen appeared normal with mild ascites.

The serum alfa fetoprotein (AFP) was 64.47 ng/mL. Thrombophilia profile showed low levels of lupus anticoagulant and proteins C and S. Upper gastrointestinal (GI) endoscopy was performed, which showed two columns of small varices. Her echocardiography revealed normal-sized left ventricle (LV) with normal function and ejection fraction of 74%.

Liver biopsy was performed that showed a slightly disturbed architecture. Sinusoids were dilated at places. The dilation was marked at places, with large spaces containing blood being noted (Figure 1). There was scanty fibrosis in between the hepatocytes. The features were suggestive of early PH.

She was managed conservatively and advised for regular follow-ups. She delivered a baby at full term without any complications. Following delivery, she was soon lost to follow-up and presented again 2.5 years later in hepatic encephalopathy and spontaneous bacterial peritonitis. During admission, she also developed hepatorenal syndrome. Despite extensive treatment and later ventilatory support, the patient died.

**DISCUSSION**

PH is a rare disorder, characterized by the presence of cystic, blood-filled spaces of variable sizes in the liver. The endothelial cells lining the internal surface of the cavities and covering the neighboring hepatocytes can be normal or show degenerative changes.

Two forms of PH, a microscopic form and a macroscopic form, have been described. In the microscopic form, ultrasound and CT do not show any changes in the liver. It is mostly seen in adult patients in association with diseases such as acquired immunodeficiency syndrome, malnutrition, tuberculosis, leprosy, vasculitis, hematological neoplasms, hepatocellular adenoma and carcinoma, androgenic anabolic steroids and estrogens, immunosuppressive drugs, tamoxifene, toxic substances (arsenic, vinyl chloride), infections (Bartonella henselae) and in renal transplant recipients receiving immunosuppressive treatment.

The pathophysiology of PH is still largely speculative. It has been attributed to an increased sinusoidal pressure because of difficulties in blood outflow from the liver, leading to parenchymal necrosis of liver cells and sinusoidal wall weakness. As the disease is very rare, data about its natural history are scanty and the clinical spectrum varies from asymptomatic cases to severe complications, such as hemoperitoneum.

The clinical features are attributed to portal hypertension and its complications by the development of esophageal varices, ascites, portopulmonary hypertension or hepatopulmonary syndrome. A variety of other clinical features have been described including abdominal pain, jaundice, hepatomegaly and liver failure requiring liver transplantation. Our patient presented with abdominal pain and distention and had signs of portal hypertension including the presence of esophageal varices and low protein ascites.

Various imaging modalities are helpful in the diagnosis of this entity such as CT scan, where the lesions appear as heterogeneous densities that become hypodense on late arterial and venous phase. Peripheral ring-like enhancement is infrequently seen. A quite characteristic finding is the absence of mass effect on adjacent vasculature. In the case of hemorrhage, PH lesions appear as hyperattenuating structures on unenhanced CT. Hepatic angiography discloses a typical appearance of PH lesions that accumulate contrast in the late arterial or parenchymal phase and retain it through the early venous phase.

In our patient, an abdomen CT scan showed hepatomegaly with heterogeneous enhancement of liver parenchyma. A
large hypodense lesion was identified with multiple linear and branching structures. Multiple tiny enhancing nodules were identified in the right lobe of the liver measuring between 1 and 2 mm. The intrahepatic portion of the IVC was also compressed; however, it showed normal enhancement. No evidence of IVC thrombosis was noted. The spleen appeared normal along with the presence of mild ascites.

Liver biopsy is the most rewarding tool in diagnosing PH. It reveals round or oval intralobular cavities that are randomly distributed between areas of normal hepatic parenchyma. The cavities communicate with sinusoids that are sometimes associated with dilated space of Disse. Red blood cells can be seen in the peliotic cysts, dilated sinusoids and space of Disse. These lesions are randomly distributed and not restricted to zone 3. There is no hepatocyte dropout, nor is there any cell plate collapse with the intact reticulin framework.

Our patient’s liver biopsy showed a slightly disturbed architecture. Sinusoids were dilated at places. The dilation was marked at places with large spaces containing blood. There was mild fibrosis in between the hepatocytes. The findings were thus consistent with a diagnosis of PH.

In conclusion, this case highlights the need to keep PH in the differential diagnosis of focal liver masses on imaging studies and liver diseases presenting with portal hypertension.

Conflict of Interest

None declared.

REFERENCES


How to cite this article: Butt MO, Luck NH, Hassan SM, Abbas Z, Mubarak M. Peliosis hepatis complicating pregnancy: A rare entity. J Transl Intern Med 2017; 5: 132-134.