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Extrahepatic Portosystemic Shunt and Intrahepatic Gallbladder: A Case Report

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Abstract Case Report

Congenital extrahepatic portosystemic shunt is not that common. However, non-congenital extrahepatic portosystemic shunt can be caused by various etiologies, for instance, post-trauma, surgical intervention and portal hypertension. We're reporting a case of asymptomatic adult female and the incidental imaging findings related to this condition. **Keywords:** Congenital, extrahepatic, portosystemic, shunt.

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Introduction

Macroscopic congenital portosystemic shunts outside the hepatic parenchyma are not so common as an incidental finding but have not been well described in the literature. Most adult patients with portosystemic shunts present with bleeding or hepatic encephalopathy as a first presentation. It has been described in few papers that those patients will not be able to develop tolerance to high ammonia levels with advancing age, eventually presenting with hepatic encephalopathy.

We present a case of a lady with congenital extrahepatic portosystemic shunt discovered as an incidental finding in a 24-year old female with nonspecific diffuse abdominal pain.

CASE REPORT

24- year old lady with previous history of left cystectomy and oophorectomy for endometrioma was investigated for diffuse nonspecific abdominal pain. She has a remote history of left cystectomy and oophorectomy one year back for an endometrioma. No history of liver disease, liver biopsy, abdominal trauma, cirrhosis, mental status changes or bleeding diathesis. Physical examination was negative for hepatomegaly and jaundice, positive for splenomegaly. Vital signs were stable. All laboratory values were within normal limits. Selected images of axial and sagittal CT scan of the abdomen and pelvis at the level of the liver was performed and incidentally revealed dilated left portal vein, draining to the right atrium and intrahepatic gallbladder (fig. 1). 3-D volume rendering reconstruction of a complementary CT scan of the chest revealed the previously mentioned abnormality (fig. 2).

DISCUSSION

Congenital extrahepatic portosystemic shunt (CEPS) is a condition in which the portal blood drains into a systemic vein, bypassing the liver through a complete or partial shunt. CEPS have been described in a variety of animals [2, 3]. However, it is extremely rare in humans. CEPS was reported in 1793 by John Abernethy, who described a postmortem examination of a 10-month-old girl that revealed termination of the portal vein (PV) in the inferior vena cava (IVC) at the level of the renal veins [4].

The clinical importance of this entity is in its associations, particularly with nodular liver lesions and congenital heart defects; what is more, it may cause liver cirrhosis and hepatic encephalopathy. Two variants of CEPS have been described according to the presence or absence of intrahepatic portal vein supply. Differentiation between these two variants plays an essential role in the management plan.

Morgan and Superina [5] classified CEPS into two types. In type 1 CEPS, there is complete diversion of portal blood into the systemic circulation (end-to-side shunt), with absent intrahepatic portal branches. Type 1 shunts are further classified into those in which the splenic vein (SV) and superior mesenteric vein (SMV) drain separately into a systemic vein (type 1a) and those in which the SV and SMV drain together after joining to form a common trunk (type 1b). In type 2 CEPS, the intrahepatic PV is intact, but some of the portal flow is diverted into a systemic vein through a side-to-side shunt.

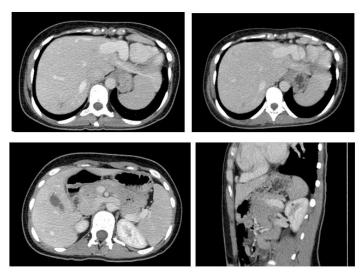


Fig-1: Multiple axial enhanced CT slices showing dilated left portal vein and intrahepatic gallbladder. Sagittal plane shows the left portal vein draining into the right atrium

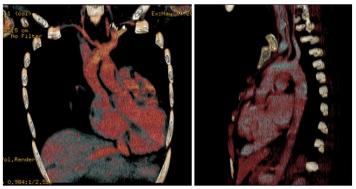


Fig-2, 3-D: volume rendering reconstruction of a complementary CT scan

However, no prior publications were found in the literature discussing the presentation of our case, which was, patient found to have an incidental finding of left portal vein draining to the right atrium through an accessory vein.

Only one other case in the literature describes an asymptomatic portacaval shunt with morphology such as seen in our patient [6]. As our patient was asymptomatic from this incidental finding, no intervention was performed or recommended. It is hypothesized that hepatic encephalopathic symptoms may develop with increasing age [7, 8].

We hope that further follow-up may clarify the natural evolution of this portosystemic shunt.

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