CASE REPORT

A naturally occurring TIPS?

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Abstract

Portal-systemic encephalopathy in both cirrhosis and non-cirrhotic patients can occur when a large portal-systemic shunt forms within the venous system. Though most commonly found in patients with portal hypertension, after trauma, surgical intervention, and liver biopsy, one can also be idiopathic. A 78-year-old Latino male developed worsening confusion and lethargy during a hospitalization for bridging of his anticoagulation. An extensive laboratory and radiologic workup did not initially reveal the etiology of his delirium and decreased state of consciousness. On exam, he had tremulousness and fetor hepaticus without corresponding liver function abnormalities in his laboratory testing. An ultrasound of the liver demonstrated a large veno-venous shunt between the right portal vein and the right hepatic vein. After multiple episodes of encephalopathy and progressive heart failure from significant right ventricular overload, the shunt was closed by interventional radiology. This treatable cause of encephalopathy may not be evident without radiologic studies.

Keywords

Portal-systemic shunt, Encephalopathy, Delirium, Heart failure

1 Introduction

Portal-systemic encephalopathy in both cirrhotic and non-cirrhotic patients can occur when a large portal-systemic shunt forms within the venous system. A macroscopic intrahepatic portal-systemic venous shunt is defined by at least a 1 mm communication between the portal and systemic venous circulation located within the liver [1]. A portal-systemic shunt is most commonly found in patients with portal hypertension, after trauma, post-surgical intervention, and post-liver biopsy, but it can also be idiopathic [1].

2 Case presentation

A 78-year-old Latino male developed worsening confusion and lethargy during his hospitalization. He had initially been admitted for bridging of heparin and monitoring of a large upper arm hematoma, as he had a mechanical aortic valve. His mental status progressively worsened during the hospitalization without obvious cause able to be identified. Initially, the patient had tremors of his bilateral hands, but he answered questions appropriately and had a non-focal neurologic exam. This progressed to lethargy with disorientation, being difficult to arouse, and unable to follow commands routinely. A CT of the head without contrast revealed no evidence of acute hemorrhage or ischemic stroke. A greater than 50 percent stenosis of the bilateral terminal ICA was noted, but no evidence of critical stenosis was seen on a CTA of the head and

neck. An infectious workup was performed, which demonstrated a urinary tract infection on urinalysis; therefore, antibiotics were initiated. The patient's mental status continued to decline despite no significant abnormalities on laboratory, including liver function, bilirubin and coagulation testing. As the patient was noted to have an odor of fetor hepaticus, a trial of lactulose was performed. With lactulose administration, his mental status greatly improved back to his baseline. A Doppler ultrasound was performed, which demonstrated a portal-systemic shunt between the right portal vein and the right hepatic vein and mild heterogeneity of the liver (see Figure 1 & 2). A CT of his abdomen demonstrated the portal-systemic shunt between the posterior division of the right portal vein into enlarged right and middle hepatic veins (see Figure 3). The communication, at its narrowest, measured 1.0 cm in diameter. As the encephalopathy was improving with conservative management, it was decided not to pursue closure of the shunt initially. He had chronic diarrhea at his nursing home and once he was admitted to the hospital, he developed constipation, which likely precipitated his encephalopathy.

The patient was readmitted for cellulitis of his right foot two months later. His stay was complicated by recurrent episodes of pulmonary edema, requiring treatment with mechanical ventilation and diuresis. An echocardiogram was performed, which demonstrated a severely dilated right ventricle and severely dilated right atrium, as well as severe tricuspid regurgitation. One year prior, his echocardiogram demonstrated moderate right ventricular enlargement and moderate to severe right atrial enlargement. The patient's mental status began to deteriorate again, much like his encephalopathy during his previous admission. An ammonia level was significantly elevated at 255 mcmol/L. Because the portal-systemic shunt was large enough to cause significant right heart failure and was causing his recurrent encephalopathy despite maximal medical treatment on lactulose, rifaximin, and zinc, the shunt was embolized by interventional radiology (see Figure 4 & 5). His encephalopathy resolved with closure of the shunt.

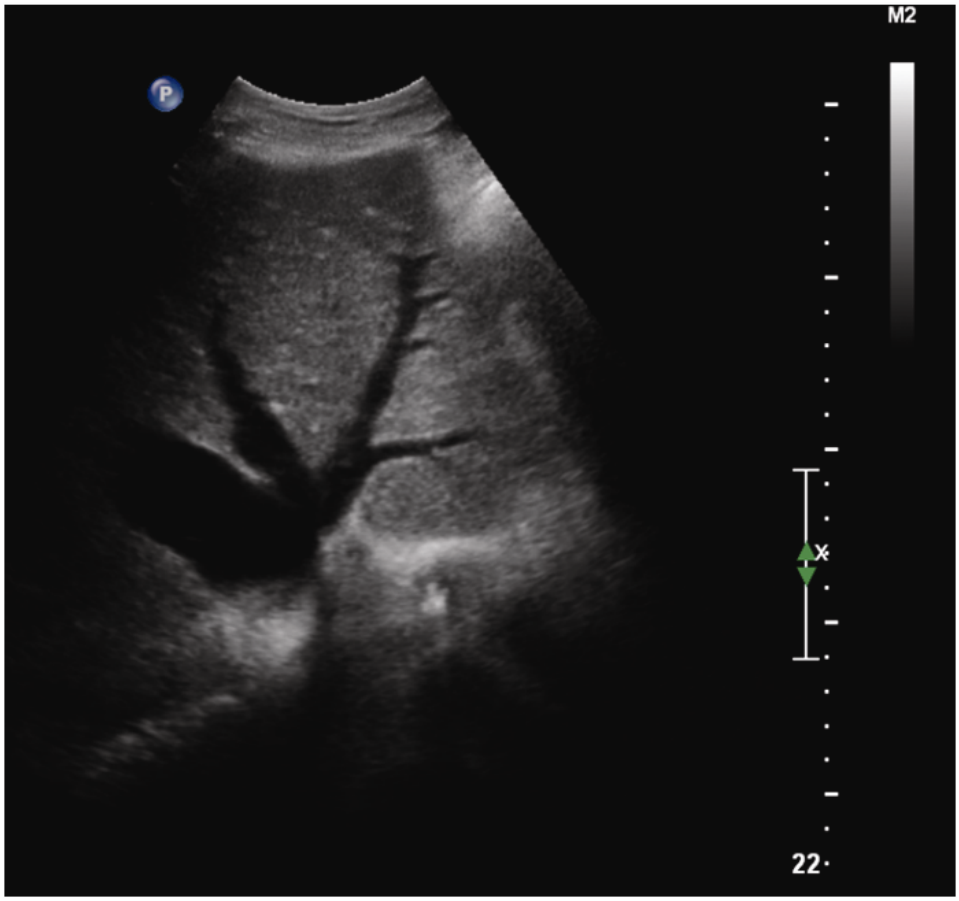

Figure 1. Ultrasound images with dilatation of the right hepatic vein

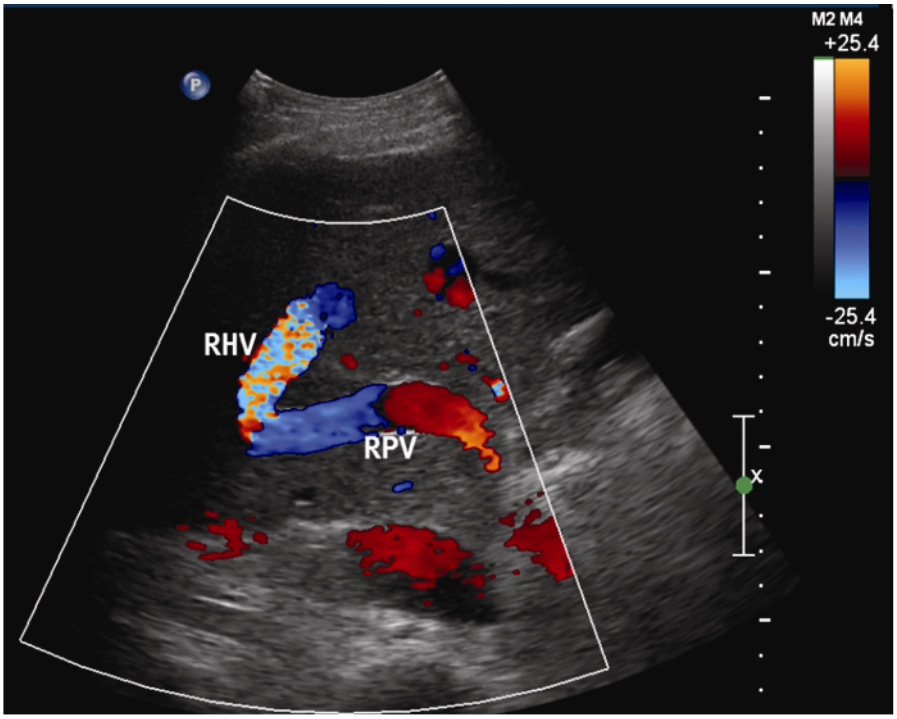

Figure 2. Doppler ultrasound demonstrated marked dilatation of the right hepatic vein with increased pulsatility in the right and middle hepatic veins.

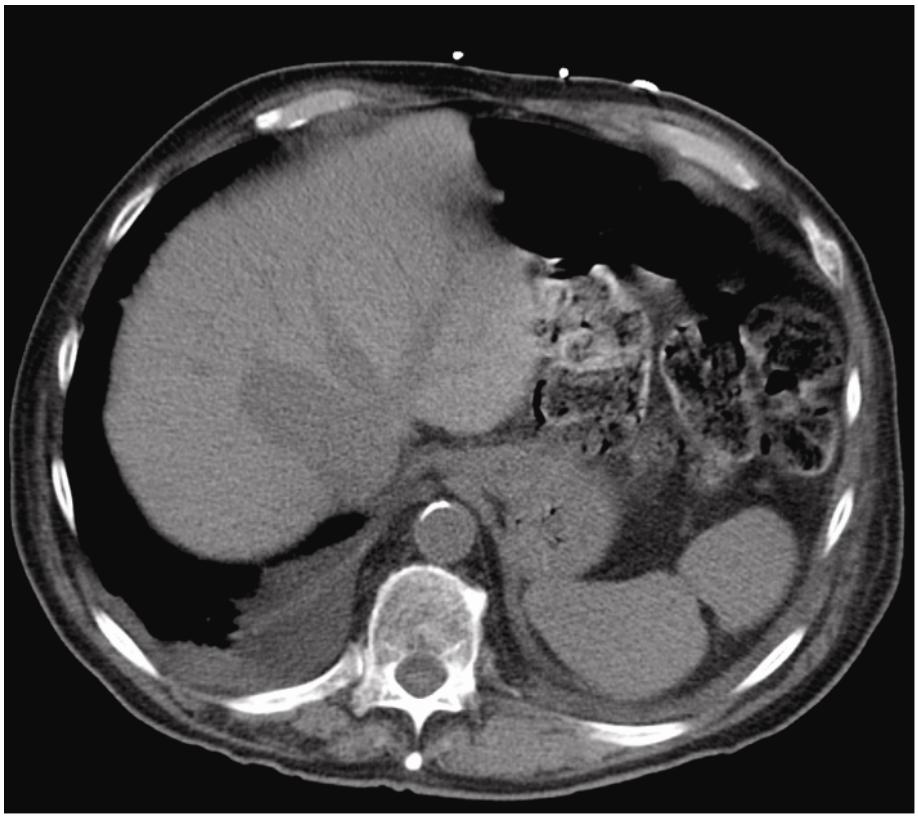

Figure 3. CT scan demonstrating large intrahepatic portal-hepatic venous shunt

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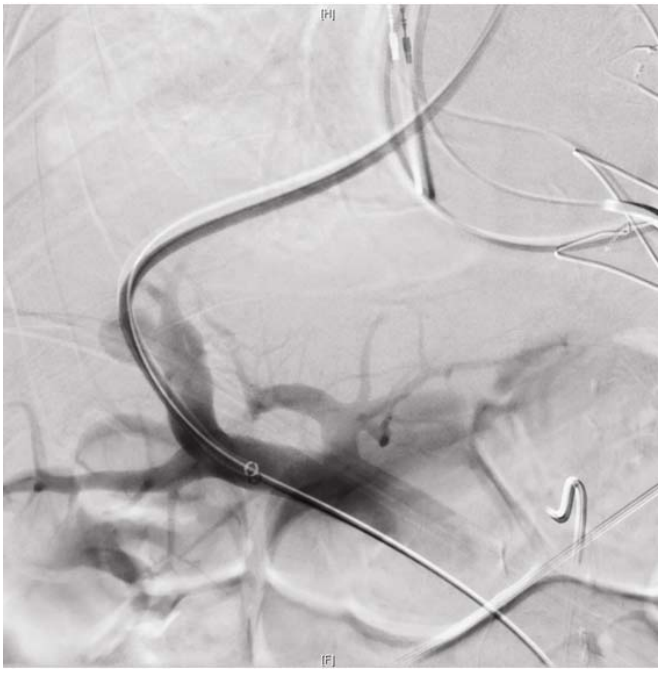

Figure 4. Interventional fluoroscopic image of large intrahepatic shunt.

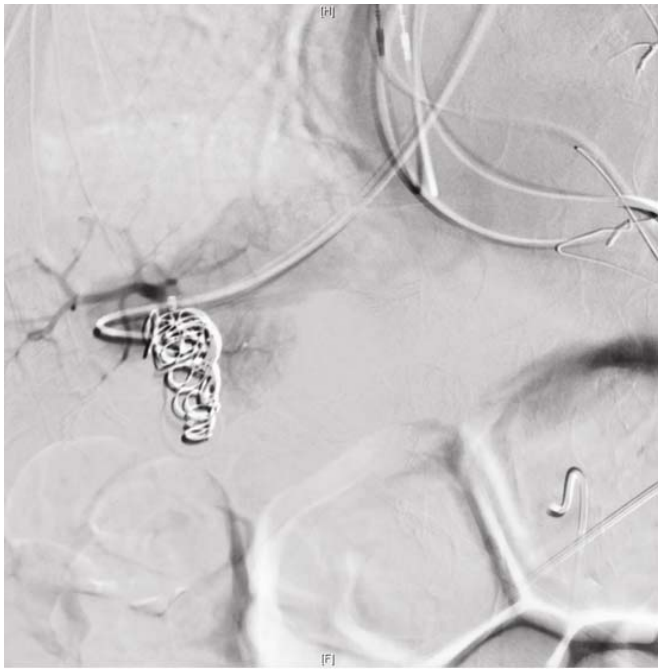

Figure 5. Interventional fluoroscopic image status post coiling of the shunt.

3 Discussion

Portal-systemic encephalopathy most commonly occurs in patients with cirrhosis and portal hypertension with spontaneous portal-systemic shunts [2]. The incidence of hepatic encephalopathy increases after intentional TIPS placement, but these shunts can also occur idiopathically or as a complication of a procedure. Congenital portal-systemic

veno-venous malformations are rare and often undiagnosed. These malformations are thought to develop from abnormal patterns of involution of the vitelline system during embryonic development [2]. Intrahepatic portal-systemic shunts are defined as a communication between the portal and systemic circulation that measures at last 1 mm in diameter, and are at least partially located in the liver [3]. When encephalopathy is diagnosed, it is often without other findings of liver disease, so it has been described as portal-systemic encephalopathy instead of hepatic encephalopathy [4]. It has been suggested that the degree of encephalopathy is worse in the elderly population, due to the older brain becoming more susceptible to symptoms from high ammonia levels [5]. The clinical signs of encephalopathy are caused by the failure of blood to pass through the hepatic sinusoids before reentering circulation [2]. This causes the degradation of ammonia to slow, thus increasing ammonia levels in the blood. Treatment of encephalopathy, both in hepatic and portal-systemic etiologies, involves decreasing dietary protein and increasing stool formation to eliminate ammonia from the body. In the case of a portal-systemic shunt, closure of the shunt can be performed by interventional radiology. These patients may often be misdiagnosed as having dementia and psychotic disorders with repeated episodes [6]. These intrahepatic portal-systemic shunts may also cause pulmonary hypertension and right-sided heart failure [7].

Acute heart failure may complicate and occur occasionally following a TIPS procedure [8]. Following TIPS, there is initial RV overload with increased RA pressures in the first 9 hours, which usually resolves after that short period of time [8]. The large portal-systemic shunt present likely contributed to the severity of our patient's heart failure.

Encephalopathy in the setting of liver disease most often stems from the inability of the liver to process ammonia appropriately, but a portal-systemic shunt without functional liver compromise can also cause a similar clinical scenario. When a patient has physical exam findings of asterixis and fetor hepaticus, even without serologic markers of liver failure, an idiopathic portal-systemic venous shunt should be considered.

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