Case Report

A case report of Spontaneous Intrahepatic Portosystemic Venous Shunt

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ABSTRACT: Intrahepatic portosystemic venous shunts are rare vascular anomalies. Only a total of 42 cases have been reported in literature. This is a case of a 56-year old female who had no history of liver disease, trauma or surgery. The patient presented with altered mental status at the emergency room and was subsequently admitted with findings of hyperammonemia. With the utilization of ultrasound with Doppler studies and CT scan, a large intrahepatic portosystemic venous shunt was detected. Surgical correction was immediately employed to avoid further complications. Ultrasonography with Doppler studies plays an important role in diagnosing this condition and guiding treatment options.
Keywords: *congenital, intrahepatic portosystemic shunt, spontaneous*

INTRODUCTION

Hepatic vascular shunts are classified as: (1) arterioportal shunts, (2) portosystemic shunts, (3) arteriosystemic shunts, (4) systemic-venous shunts and (5) portal to portal shunts.[1] According to Papamichail et al., congenital portosystemic shunts are rare vascular anomalies caused by abnormal development of fetal vasculature.[2] With these shunts, intestinal blood bypasses the liver and reaches the systemic circulation, thus producing symptoms and complications.

Portal to systemic venous shunts are classified as extrahepatic or intrahepatic. Extrahepatic communications are usually seen in the setting of chronic hepatic dysfunction, i.e. cirrhosis with portal hypertension and are commonly through the coronary vein, esophageal varices, or retroperitoneal collaterals. These shunts were first reported by John Abernethy in 1793, and are known as "Abernethy malformation.[3]" Takahashi et al. states that extrahepatic portosystemic shunts have already been greatly researched on and a number of examples are available in literature.[4] On the other hand, intrahepatic communications are located between intrahepatic portal veins and systemic veins.[3] These shunts are rare and were first reported only in 1964 by Raskin et al.[4]

Park et al. presented a classification of intrahepatic portosystemic venous shunts based on the morphological varieties of the shunt vessels in the liver: type I, a single, large tubular vessel of constant diameter that connects the right portal vein to the inferior vena cava; type II, a peripheral shunt, single or multiple communications between peripheral branches of portal vein and hepatic vein in one hepatic segment; type III, aneurysmal communication between the peripheral portal vein and hepatic vein; type IV, multiple, diffuse communications between the peripheral portal vein and hepatic vein, in both liver lobes. Type I is noted to be the most common among the four categories, while type IV is the least common.[1]

According to Naidoo et al., although intrahepatic portosystemic shunts are usually incidental findings, recognition of these anomalies is important due to possible complications such as hepatic encephalopathy, liver cirrhosis, liver failure, pulmonary arterial hypertension and metabolic abnormalities, e.g. hypergalactosemia and hyperammonemia.[5]

This is a case of type II intrahepatic portosystemic venous shunt, presenting with hepatic encephalopathy and hyperammonemia. The patient was first treated conservatively but subsequently underwent surgical correction. This case report highlights the different imaging modalities that can be utilized to detect and evaluate these rare vascular anomalies.

CASE REPORT

A 56-year-old female presented at the emergency room with a chief complaint of disorientation. The patient had no history of liver trauma or biopsy, cirrhosis, or abdominal surgery. Laboratory examinations showed increased serum ammonia. A plain cranial computed tomography (CT) scan was done which showed unremarkable results. Initial impression at this time was metabolic encephalopathy, and the patient was subsequently admitted. A contrast-enhanced CT scan of the whole abdomen was requested, which showed a large portosystemic shunt between the right portal vein and middle hepatic vein. No CT evidence of cirrhosis was noted. After conservative treatment with lactulose enema, her ammonia levels normalized and her mental status recovered. Two days after admission, the patient was cleared for discharge and was advised correction of the shunt through surgery or embolization. The patient's final diagnosis was hepatic encephalopathy secondary to a portal vein-hepatic vein shunt.

Two weeks after admission, she then consulted with the Center for Liver Disease Management and Transplantation (CLDMT) of The Medical City where further work-up, including a liver Doppler ultrasound (Figure 1), was done. Findings showed a communication between a dilated right

main portal vein and a dilated middle hepatic vein. No evidence of cirrhosis was noted.

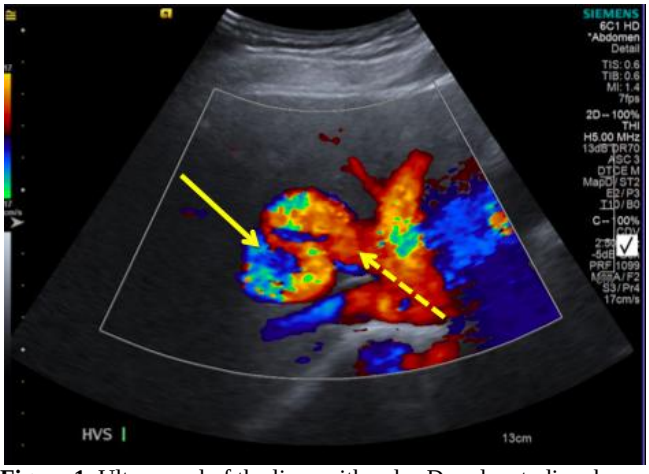

Figure 1. Ultrasound of the liver with color Doppler studies showing a shunt from the right portal vein (arrow) to the middle hepatic vein (dashed arrow)

Five months after admission, a repeat contrast-enhanced CT scan (Figure 2) of the upper abdomen was then done, which exhibited a stable communication between the right main portal vein and the middle hepatic vein. The patient was advised that the best treatment option was surgical correction of the shunt, but she decided to observe her condition. The patient had no subjective complaints at this time, including altered mental status.

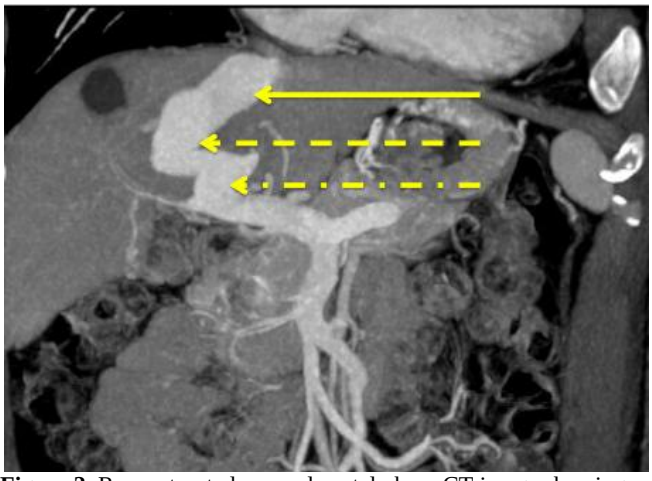

Figure 2. Reconstructed coronal portal phase CT image showing a shunt (dashed arrow) between the dilated and tortuous right main portal vein (dot dashed arrow) and dilated middle hepatic vein (arrow)

Seven months after admission, the patient then decided to undergo definitive surgical treatment. She underwent a hepatotomy, dissection and closure of the portovenous-hepatic vein shunt. Intraoperative findings included a dilated middle hepatic vein with direct communication to the right anterior portal vein. The patient tolerated the procedure well with no reportable events.

On the 6th post-operative day, a repeat liver Doppler ultrasound showed non-visualization of the previously seen communication between the right main portal vein and middle hepatic vein. The patient was subsequently discharged. Follow-up consultation revealed no recurrence of altered mental status, with no late complications following surgery.

DISCUSSION

According to Takahashi et al., an intrahepatic portosystemic venous shunt (IPSVS) is defined as a communication, measuring more than 1 mm in diameter, between an intrahepatic portal vein and a systemic vein via an anomalous intrahepatic venous channel.[4] Only 42 cases of intrahepatic portosystemic venous shunt have been reported in the English-language literature.[6]

An IPSVS can be classified as either acquired or congenital. Most cases of IPSVS develop in the setting of chronic hepatic dysfunction, such as hepatic cirrhosis, as well as following hepatic surgery or trauma. When a patient presents with an IPSVS without a history of liver disease, trauma or surgery, it is presumed to be congenital or spontaneous in origin.[7] This patient denied any history of liver disease, trauma or surgery that may have predispose her to developing this vascular abnormality. As stated by Torigoe et al., congenital IPSVS is a rare condition with a reported prevalence of 0.0235% in the general adult population.[8]

Embryologically, the development of the hepatic venous system starts by the 5th gestational week, with three major paired veins draining into the sinus venosus. These three paired veins are: the vitelline veins, umbilical veins and cardinal veins. The vitelline veins anastomose with each other around the developing duodenum and pass through the septum transversum (primitive liver) to the sinus venosus. A persistent communication between the vitelline veins and the sinus venosus is the presumed basis for intrahepatic portosystemic shunts. [6,9-10]

According to Naidoo et al., IPSVS are typically incidental findings on imaging studies, or on presentation of complications such as hepatic encephalopathy, liver failure, cirrhosis, pulmonary arterial hypertension and metabolic abnormalities such as hypergalactosemia and hyperammonemia.[5] The patient first presented with altered mental status and hyperammonemia. With the help of the CT scan, the intrahepatic shunt was detected. Unlike other congenital diseases, the presence of this anomaly may not be recognized early due to the time it takes to develop hepatic encephalopathy. Patients with congenital IPSVS, whether extrahepatic or intrahepatic, usually develop hepatic encephalopathy during mid adult life, just like in this

patient. Whether these patients develop symptoms depends on the calculated shunt ratio, the sensitivity of the brain to ammonia and the liver function of the patient. The onset of altered mental status in mid adult life may be explained by the fact that the risk for developing hepatic encephalopathy increases with age as the cerebral tolerance for hepatotoxic substances decrease.[4]

To confirm the diagnosis of IPSVS, Doppler sonography, contrast-enhanced CT, MRI and angiography can be used.[11] Gallego et al. states that ultrasound with Doppler studies is the single most important imaging for the diagnosis of IPSVSs.[12] A B mode real-time sonography study usually shows abnormal cystic or tubular, anechoic, serpiginous vascular structures communicating between portal venous structures and the systemic circulation.[5] With the utilization of Doppler ultrasound, the vascular nature of these structures can be confirmed, as well as calculation of blood flow volumes and shunt ratio. Blood flow volume is calculated by multiplying the cross sectional area by the mean velocity. According to Singh et al., shunt ratio is derived by dividing the total blood flow volume in the shunt divided by the blood flow in the portal vein.[13] Low shunt ratios (<30%) have been found to not cause hepatic encephalopathy, even in patient with cirrhosis. Regardless of age, shunt ratios that are greater than 60% should be corrected due to the risk of encephalopathy and liver dysfunction. The patient has a calculated shunt ratio of 70%, which explains her symptoms and risk for complications. CT scan and MRI both help confirm the diagnosis, but their roles have yet to be identified.[9] MRI provides a similar picture with that of CT, with the advantage of the use of non-ionizing radiation, as well as MR venography. Aside from liver Doppler studies, nuclear medicine can also be utilized to calculate the shunt ratio by portal scintigraphy with the administration of submucosal rectal injection of iodine-123 iodoamphetamine.[5] The advantages of liver Doppler studies over these other imaging modalities include: greater availability and accessibility, lower cost and non-invasiveness.

Several factors are considered to establish a treatment plan for patients with IPSVS. These include, type of shunt, location, degree of hepatic function, patient age, symptoms and complications. The therapeutic goal is to cut off the abnormal communication between the portal and systemic circulation, and restore portal flow to the liver. In the pediatric population, it has been concluded that all persisting shunts after the first year of life should be corrected, without waiting for symptoms to develop. On the other hand, asymptomatic adult patients with low flow shunts (<30% shunt ratio) can be observed and monitored with arterial ammonia levels and serial Doppler studies.[2] As stated by Naidoo et al., treatment options include surgical correction, transcatheter embolization or liver transplantation, as a last resort.[5]

Conclusion

IPSVS are rare vascular abnormalities that may be detected incidentally or on presentation of complications. Several imaging modalities can be used for diagnosis, however, ultrasonography with Doppler studies is the single most important tool due to its availability, accessibility, low cost and non-invasiveness. In addition, the shunt ratio, which is greatly used for guiding treatment options, can be derived using ultrasonography with Doppler studies.

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