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Case Report *Vascular Interventions*

Successful embolization of symptomatic intrahepatic portosystemic venous shunts through trans-splenic access – A case report

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ABSTRACT

Intrahepatic portosystemic venous shunts (IPSVS) are rare vascular disorders that are often missed or misdiagnosed in clinical practice. These shunts involve abnormal connections between the portal and systemic venous systems and can cause a range of symptoms, with encephalopathy being the most common symptom. We present a unique case of treating IPSVS in a patient with encephalopathy but no history of liver or vascular disease. The treatment successfully used trans-splenic access (TSA) for embolization to address seven IPSVS.

Keywords: Hepatic encephalopathy, Intrahepatic portosystemic venous shunts, Trans-splenic embolization, Veno-venous malformations

INTRODUCTION

Vascular anomalies cover a range of conditions that can affect any part of the body. These abnormalities are classified as acquired or congenital. Congenital vascular anomalies occur during embryonic development and may present early in life. Vascular anomalies may be sporadic, however a few cases demonstrate discrete signs of inheritance. Examples of congenital vascular malformations include capillary malformation, arteriovenous malformations, cutaneomucosal venous malformations, and hereditary hemorrhagic telangiectasia; some of which can be symptomatic or asymptomatic depending on location.[1] Acquired vascular anomalies, such as arteriovenous fistulas, develop in response to stress such as trauma and present with varied clinical presentations.[2] In contrast, acquired vascular anomalies develop in response to chronic stress on the body and often do not cause many symptoms. As a result, vascular anomalies frequently go unnoticed by patients and healthcare providers. While congenital vascular malformations may resolve early in life without causing lasting symptoms, persistent abnormal vascular connections can lead to various clinical issues, making diagnosis difficult.

Intrahepatic portosystemic venous shunts (IPSVS) are vascular anomalies that connect the portal and systemic venous systems. Clinical presentation of IPSVS may vary as these shunts may be asymptomatic or can include symptoms such as hypergalactosemia, hyperammonemia, hyperglycemia, hepatic encephalopathy, liver failure, and heart failure.[3-5] IPSVS have been

categorized into four types.[6] Type 1 involves a single large vessel connecting the right portal vein to the inferior vena cava. Type 2 consists of local shunts with one or more connections in a single hepatic segment. Type 3 features peripheral portal and hepatic veins connected through an aneurysm, and Type 4 involves multiple segments of connection between the portal and hepatic veins. The case presented here was consistent with IPSVS type 4, as there were multiple vascular connections in various hepatic segments.

We present a unique case of multiple intrahepatic vascular anomalies between the portal venous and hepatic venous systems, accompanied by hyperammonemia and subsequent encephalopathy. Medical treatment was unsuccessful in addressing the hyperammonemia and altered mental status. Interventional radiology was consulted, and the patient successfully underwent embolization via a novel TSA for embolization, addressing seven IPSVS.

CASE REPORT

A 72-year-old woman with a reported medical history of heart attack, stroke, and hepatic steatosis presented to the emergency department (ED) with acute encephalopathy that gradually worsened. The patient's encephalopathy had previously improved with lactulose, but her symptoms worsened, prompting her to visit the ED. The patient reported history of intermittent encephalopathy requiring lactulose due to an existing diagnosis of cirrhosis by gastroenterology; however, imaging upon admission did not support the diagnosis of cirrhosis or related liver damage. Initial physical exam showed moderate scleral icterus, acute encephalopathy, and right upper quadrant abdominal tenderness. Initial laboratory studies showed an albumin of 2.2 g/dL, alkaline phosphatase of 132 U/L, total bilirubin of 2.5 mg/dL, prothrombin time of 16.3 s, partial thromboplastin time of 32 s, a negative hepatitis panel, and ammonia of 122 μmol/L. CT head without contrast and brain magnetic resonance imaging showed no acute pathology. Based on her clinical presentation, hyperammonemia, and negative CT of the head, hepatic encephalopathy was diagnosed. Abdominal Doppler ultrasound of the abdomen showed normal hepatic vascular flow pattern with no suggestive findings of arteriovenous defects or fistula. Subsequent CT imaging of the abdomen revealed evidence of hepatic steatosis and a dilated portal vein measuring 13 mm, suggesting possible intrahepatic lesions versus intrahepatic arteriovenous shunt or portal shunt [Figure 1]. An esophagogastroduodenoscopy was performed, and no evidence of gastric varices was found.

On day 7 of admission, the risks, benefits, and alternatives were discussed with the patient's next of kin, and informed consent was obtained. Using sterile ultrasound guidance, a 21-gauge micropuncture needle was used to access a branch of the splenic vein through the splenic parenchyma. A 0.018 in guidewire was then inserted through the lumen of the micropuncture needle, a transitional dilator was placed over the guidewire, and contrast was injected to confirm appropriate splenic vein positioning. The guidewire was then advanced into the splenic and main portal vein. A 6 French sheath was placed into the splenic vein. A 5 French vert catheter was placed over the guidewire, and venography of the main portal vein, right intrahepatic portal vein, and left intrahepatic portal vein were performed. Venography revealed multiple veno-venous malformations involving the intrahepatic portal venous system and hepatic veins [Figure 2a]. Next, super selection of the seven IPSVS was achieved using a Progreat microcatheter and double-angle guidewire. Interventional radiology successfully treated seven vascular anomalies with coil embolization using a series of detachable embolization coils. Post-embolization venography showed successful closure of accessible vascular anomalies [Figure 2b]. The 6 French sheath was retracted into the splenic parenchyma, and coil embolization of the splenic access tract was performed using multiple detachable embolization coils. All catheters and guidewires were removed at the end of the procedure.

On postoperative day 1, the patient showed significant improvement in mental status and hyperammonemia (<10 uMOL/L) following successful embolization. Postoperatively, without hyperammonemia medications, the patient regained a completed return of mentation. Upon discharge, the patient was scheduled for a follow-up outpatient visit to monitor the progression of encephalopathy and ammonia levels. The patient was contacted at 4 weeks and 9 months post-procedure and reported no longer requiring lactulose and continued improvement regarding encephalopathy.

DISCUSSION

We are presenting a unique case of multiple veno-venous anomalies in a patient without liver disease, recent trauma,

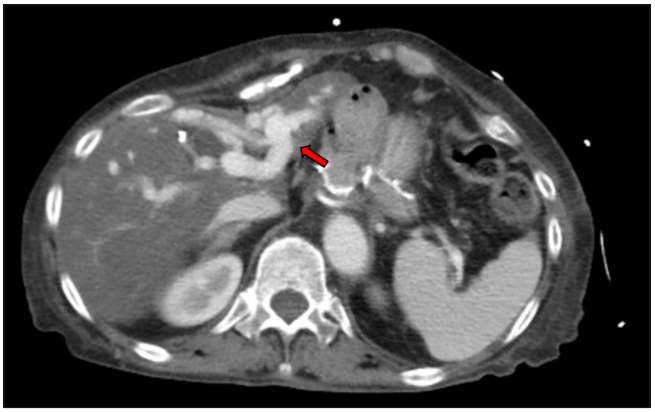

Figure 1: A 72-year-old female presenting with acute hepatic encephalopathy. Portal venous phase contrast-enhanced computed tomography image demonstrating communications between the intrahepatic portal venous branch and hepatic veins (red arrow).

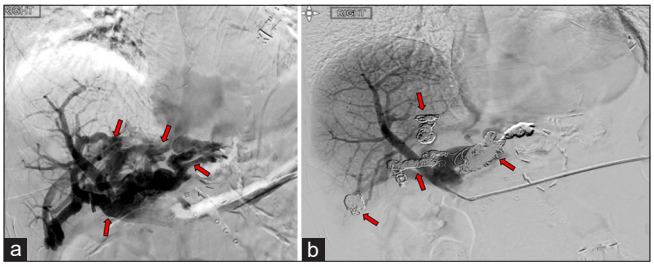

Figure 2: A 72-year-old female presenting with acute hepatic encephalopathy. (a) Percutaneous venography demonstrates multiple intrahepatic portosystemic shunts (red arrows). (b) Post-embolization of multiple intrahepatic portosystemic venous shunts with Penumbra detachable coils and decreased shunt flow (red arrows).

or surgery. Diagnosing these vascular anomalies is complex as they are often asymptomatic and have a wide range of clinical presentations.[5,7] The cause of the multiple veno-venous anomalies in this case remains unclear. Current literature suggests two possible explanations for abnormal connections between the portal and systemic vasculature: Acquired and congenital malformations.[3-5] Given this patient's presentation of seven IPSVS without evidence of liver disease or trauma, it is difficult to determine the exact cause. It is possible these shunts existed at birth and persisted into adulthood, although congenital IPSVS are usually evident in the early stages of life.[4,5,7]

There have been reports of patients with similar symptoms, such as encephalopathy, who were found to have portal-hepatic venous shunts. However these patients had a history of cirrhosis or recent trauma, which may have contributed to the shunt formation. Additionally, case reports exist for suspected spontaneous intrahepatic shunts in patients without cirrhosis or trauma; however, these patients only had 1-2 IPSVS.[3] Due to the unusual nature of our patient's presentation and lack of risk factors for an acquired vascular anomalies, we suspect she may have had asymptomatic congenital IPSVS that enlarged over time, ultimately causing her symptoms.

As previously discussed, IPSVS may present with varied clinical presentations making diagnosis difficult. However, once diagnosed, treatment depends on symptom severity. Increasing literature favors endovascular embolization for symptomatic IPSVS vascular anomalies, but it becomes more challenging to address whether to prophylactically treat asymptomatic IPSVS.[8] Endovascular interventions do pose potential complications; thus, it is imperative to involve patients in making the decision to treat asymptomatic vascular anomalies.[8,9] Conservative treatment includes lifestyle modifications and medical management for asymptomatic IPSVS, and additional interventions are available including the previously mentioned endovascular embolization and surgical interventions such as portal vein ligation, hepatic lobectomy and liver transplantation for symptomatic IPSVS.

Current literature outlines three different IPSVS endovascular treatments: trans-ileocolic, percutaneous trans-hepatic, and retrograde trans-caval embolization.[8] Choosing the best approach depends on factors such as the location, size, and number of shunts. Trans-ileocolic obliteration allows for easy catheter access but is the most invasive of the three endovascular treatments requiring an abdominal incision and general anesthesia. Percutaneous transhepatic approach offers direct portal venous access without an abdominal incision or general anesthesia, but it is best for shunts contralateral to the punctured side and carries some risks. Retrograde transcaval obliteration is the least invasive, not requiring an abdominal incision, organ puncture, or general anesthesia, but is only suitable for treatment of 1 to 2 large shunts.[10]

In a unique case involving a patient with seven veno-venous shunts across various hepatic segments, , a novel TSA for embolization was chosen as the optimal treatment. This approach did not require general anesthesia or an abdominal incision, unlike the trans-ileocolic approach. Additionally, since the patient had shunts in various hepatic segments, the transhepatic approach would have been challenging given limited access to multiple hepatic segments. Additionally, TSA requires only one access site, further reducing complication risk.

It's important to note that TSA for embolization is not currently documented in the literature as treatment for IPSVS in various hepatic segments. However, due to the observed improvement in the patient's clinical presentation and ammonia blood levels, it warrants further investigation as a novel IPSVS treatment approach.

CONCLUSION

Diagnosis and management of intrahepatic portosystemic venous shunts (IPSVS) require nuanced understanding of patient presentations and a tailored treatment approach. Recognizing the ways IPSVS can manifest, especially in asymptomatic patients without prior liver disease, underscores the importance of thorough imaging studies for accurate identification. In complex cases, as seen here, alternative approaches such as TSA for embolization highlight the evolving landscape of IPSVS treatment, stressing the importance of individualized care and further research into these advanced techniques to optimize outcomes for each patient.

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