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CASE REPORT

Extra-Hepatic Portosystemic Shunt Leading to Encephalopathy in a Noncirrhotic Liver: Successful Treatment with Endovascular Occlusion

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ABSTRACT

BACKGROUND

Hepatic encephalopathy in the western world is primarily associated with liver cirrhosis, however, it has also been reported in noncirrhotic patients with large congenital portosystemic shunts. This case reports a successful treatment with endovascular occlusion.

CASE PRESENTATION

A 60-year-old male presented with a two-year history of neurologic and psychiatric symptoms requiring multiple hospital admissions and serum hyperammonemia. Abdominal imaging was remarkable for the presence of a large vascular shunt communicating the portal vein to the left renal vein, consistent with a congenital portosystemic shunt causing hepatic encephalopathy in a noncirrhotic liver. Endovascular shunt closure was performed with Amplatzer II vascular plug with clinical improvement and reduction of serum ammonia to normal levels within two weeks.

CONCLUSION

Adult-on-set of noncirrhotic hepatic encephalopathy is a very uncommon condition and few reports have described endovascular treatment for these patients. Our work corroborates that the endovascular approach is a safe, minimal invasive technic with proven clinical benefits.

KEYWORDS

Interventional radiology; Congenital portosystemic shunt; Noncirrhotic encephalopathy

ABBREVIATION

CEPS: Congenital Extra-Hepatic Portosystemic Shunts; MRI: Magnetic Resonance Imaging

INTRODUCTION

In healthy individuals, ammonia (NH3) enters the portal circulation from the gastrointestinal tract and is subsequently converted to urea inside the hepatocytes through the urea cycle. Urea is then physiologically excreted via the colon and kidneys. However, in cases of severe hepatic dysfunction, such as in cirrhotic liver or in presence of portosystemic shunts, ammonia is not metabolized in the liver and consequently enters and accumulates in the systemic circulation. By penetrating in the blood-brain barrier, ammonia act as a strong neurotoxin implicated in the development of hepatic encephalopathy, which produces a wide spectrum of neurologic and psychiatric abnormalities [1].

Hepatic Encephalopathy in Noncirrhotic Liver

Hepatic encephalopathy in the western world is primarily associated with liver cirrhosis, which is a consequence of both hepatocyte damage and the development of portosystemic shunts. Hepatic encephalopathy has also been reported in noncirrhotic patients with large congenital portosystemic shunts, suggesting that reduction of ammonia detoxification could lead to alteration of central nervous system even in the absence of liver damage [2].

Congenital portosystemic shunt is a rare condition usually identified in pediatric patients [3]. The majority of shunts are intrahepatic, involving one or more communications between the portal vein and hepatic veins or the inferior vena cava. Congenital extra-hepatic portosystemic shunts (CEPS) are classified accordingly to the system proposed by Morgan and Superina in 1994 [4] in 2 subtypes, based on the presence or absence of the intrahepatic portal vein. In type 1 CEPS (also known as Abernethy malformation), characterized by the absence of the portal vein, the shunt represents the only mesenteric and splenic outflow. Therefore, cases are diagnosed early in childhood and liver transplant is the only therapeutic option [5]. In type 2 CEPS, the portal vein is normal or slight hypoplastic and some portal flow is diverted to the inferior vena cava through a vascular anomaly. Type 2 CEPS can be diagnosed during infancy or adulthood [6].

Adult on-set hepatic encephalopathy in noncirrhotic patients due to CEPS is extremely rare, with the exact incidence is unknown likely due to a combination of under detection and/or underreporting [7,8].

CASE PRESENTATION

A 60-year-old male presented with a two-year history of episodic confusion, depression, and hallucination, requiring multiple hospital admissions and psychiatric follow-up. Past medical history was significant for Parkinson's syndrome, which remained asymptomatic with medical therapy. The patient reported current tobacco smoking and social alcohol intake. Upon admission, serum ammonia level of 361,6 ug/dl was notable. The patient had no other relevant risk factors for hepatic disease. Extended laboratory analysis was performed, including (viral serology and autoimmune markers, with no positive findings.

An abdominal magnetic resonance imaging (MRI) was requested revealing a normal-sized and homogeneous liver, with smooth contours, without splenomegaly, perigastric or splenorenal varices or ascites, or other stigmata of portal hypertension. The portal vein was hypoplastic, with reduced caliber (Figure 1A and Figure 1B), and a

large vascular structure (approximately 16 mm caliber) was seen communicating the hypoplastic portal vein to the left renal vein (Figure 1C and Figure 1D). These findings were consistent with type 2 CEPS.

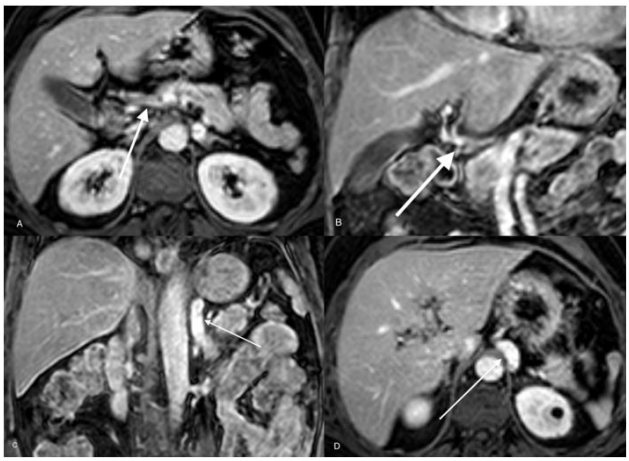

Figure 1: Abdominal MRI performed during the initial workup. T1-weighted imaging post-contrast shows a hypoplastic portal vein (A and B, arrows). Vascular structure is shown communicating the portal vein to the left renal vein (C and D arrows).

Medical treatment with a combination of antibiotics and lactulose was attempted to reduce gut toxins without significant clinical benefit.

An interventional radiology consultation was requested to proceed with the closure of the endovascular shunt, which is considered the less invasive therapeutic option with high probability of therapeutic and clinical success [9].

The right common femoral vein was punctured and a 5-French Cobra-2 catheter was introduced until the left renal vein. After a few venograms, the portosystemic shunt communicating the portal vein with the left renal vein was identified (Figure 2), confirming the 16 mm caliber near the confluence with the renal vein. A 7-French sheath was introduced in the selected shunt. A 22 mm Amplatzer II vascular plug (Abbott®) was placed within the 7-French sheath and deployed through 18 mm of the shunt near the confluence with the renal vein. The plug dimensions were 30%-50% larger than the diameter of the target vessel, following the manufacturer's recommendations. Pos-deployment angiography confirmed shunt exclusion, with contrast stasis downstream of the vascular plug (Figure 3).

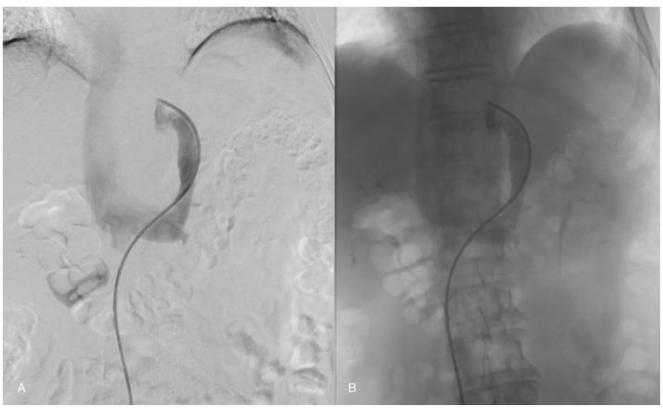

Figure 2: Angiography imaging after contrast injection in the portosystemic shunt shows communication between the portal vein and left renal vein (with (A) and without (B) digital subtraction).

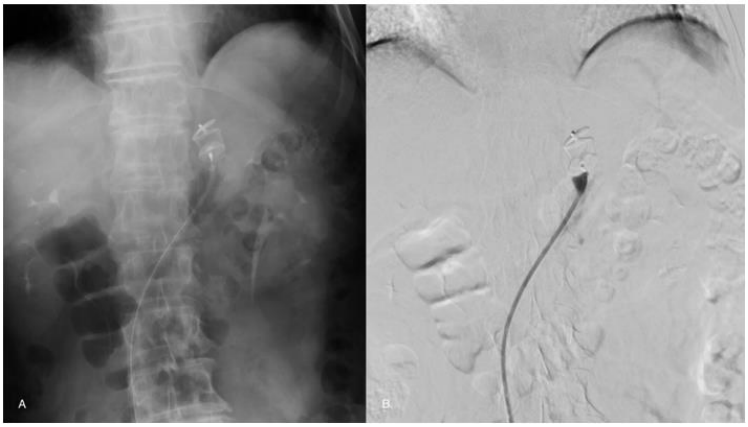

Figure 3: (A) Angiography imaging acquisition after Amplatzer II vascular plug deployment. **(B)** Digital subtraction angiography is shown confirming shunt exclusion with downstream contrast stasis.

The patient's serum ammonia levels normalized within two weeks of the procedure (17.9 ug/dl) and substantial clinical improvement of both psychiatric symptoms and cognitive function was reported by the medical assistant. A follow-up CT scan performed 1 month later (Figure 4) showed successful shunt embolization without complications related to the procedure/vascular plug.

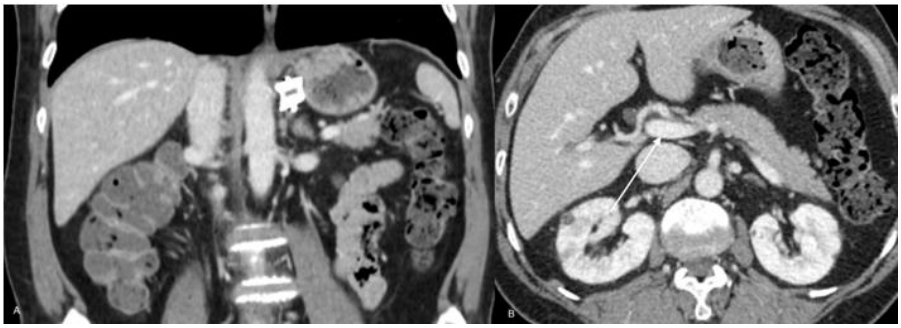

Figure 4: (A) CT scan performed 1 month after the procedure confirming shunt exclusion with correct vascular plug placement. **(B)** Portal vein with regular diameter is shown upon restoration of blood flow.

CONCLUSION

Adult on-set of noncirrhotic hepatic encephalopathy is a very uncommon condition. In this report, we describe a case of an adult man with a two-years history of psychiatric symptoms and hyperammonemia caused by a portosystemic shunt between the portal vein and the left renal vein. To our knowledge few reports have described endovascular treatment for these patients. Our work corroborates that the endovascular approach is a safe, minimal invasive technic with proven clinical benefits.

CONSENT FOR PUBLICATION

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

COMPETING INTERESTS

The authors declare that they have no competing interest.

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