

Idiopathic Portocaval Shunt Presenting with Hepatic Encephalopathy in a Non-Cirrhotic Patient: A Case Report and Literature Review

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1. Abstract

Idiopathic portocaval shunt (IPCS) is an uncommon vascular condition characterized by the diversion of portal venous blood into the systemic circulation without evidence of cirrhosis or portal hypertension. This anomaly can result in elevated systemic ammonia levels and subsequent hepatic encephalopathy (HE), even in patients with preserved liver function. We present a case of a 40-year-old man with end-stage renal disease who developed seizures and hyperammonaemia. Imaging identified a type II Abernethy malformation. The diagnosis was confirmed by Doppler ultrasonography, CT angiography, and liver biopsy. The patient showed a favorable outcome after intensification of haemodialysis. This case underscores the need to consider IPCS in the diagnostic workup of unexplained neuropsychiatric symptoms and highlights the importance of comprehensive imaging and individualized management.

2. Introduction

Idiopathic portocaval shunt (IPCS) refers to a spontaneous portosystemic venous connection occurring in the absence of cirrhosis, portal hypertension, prior abdominal surgery, or trauma [1,2]. This condition allows portal blood to bypass the hepatic parenchyma, reducing the clearance of neurotoxic substances such as ammonia, which can accumulate and lead to hepatic encephalopathy (HE) despite normal hepatic structure [3]. Morgan and Superina proposed a classification for congenital portosystemic shunts, distinguishing type I (complete portal diversion) from type II (partial shunting with preserved intrahepatic portal flow) [4]. Type I anomalies are often diagnosed in early childhood, while type II variants may remain undetected until adulthood and can manifest with neuropsychiatric features such as confusion, behavioral changes, or seizures [5,6].

3. Case Presentation

A 40-year-old man under going hemodialysis for end-stage renal disease secondary to chronic glomerulonephritis was admitted with generalized tonic-clonic seizures and right upper quadrant abdominal discomfort. Upon examination, the patient was conscious with psychomotor slowing, but exhibited no signs of chronic liver disease or portal hypertension. Laboratory findings were unremarkable, including liver function tests, serum electrolytes, fasting glucose, viral serologies, autoimmune workup, ceruloplasmin, and serum copper. Notably, plasma ammonia was elevated at 161 $\mu\text{mol/L}$. Brain MRI and electroencephalogram were within normal limits. Doppler ultrasound of the abdomen revealed a dilated portal bifurcation with turbulent flow, and a 12.4 mm vascular connection between the right portal vein and retrohepatic inferior vena cava. CT angiography confirmed a type II Abernethy malformation. Liver biopsy showed mild hepatocellular changes and nonspecific portal inflammation without fibrosis or steatosis. The patient was managed with intensified hemodialysis, leading to normalization of plasma ammonia levels and complete resolution of symptoms. He remained asymptomatic during a six-month follow-up period.

4. Discussion

Congenital portosystemic shunts, though rare, are increasingly recognized as a potential cause of hepatic encephalopathy in patients without underlying hepatic disease. When portal blood bypasses hepatic filtration, neurotoxins such as ammonia can accumulate and reach the central nervous system. Astrocytic metabolism converts ammonia into glutamine, promoting cerebral edema and dysfunction [7,8]. Clinical presentation may vary widely, with seizures being an uncommon but reported manifestation linked to hyperammonemic encephalopathy [9]. In this case, the absence of structural brain abnormalities and rapid improvement with ammonia-lowering treatment supported the diagnosis. Several reports in the literature describe adult cases of type II Abernethy malformation. Ohno et al. [10] reported a woman with recurrent HE successfully treated by shunt embolization. Lautz et al. [11] emphasized a patient-specific approach, balancing shunt anatomy and liver function. Zanetto et al. [12] showed that stable patients without hepatic dysfunction may respond well to conservative strategies, such as in our case. Franchi-Abella et al. noted that delayed diagnosis is common due to nonspecific clinical presentations [5]. Palumbo et al. highlighted the utility of CT or MR angiography for anatomical delineation and treatment planning [13]. Doppler ultrasound is a useful non-invasive screening tool [14], though cross-sectional imaging is often needed to delineate vascular anatomy [15]. In particular, recent imaging advances like 3D reconstructions and dynamic contrast-enhanced sequences improve characterization

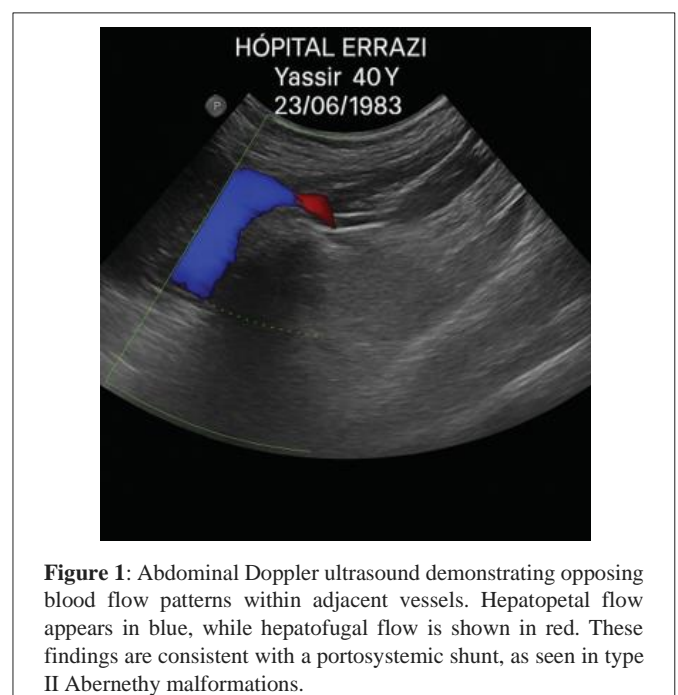


Figure 1: Abdominal Doppler ultrasound demonstrating opposing blood flow patterns within adjacent vessels. Hepatopetal flow appears in blue, while hepatofugal flow is shown in red. These findings are consistent with a portosystemic shunt, as seen in type II Abernethy malformations.

and therapeutic planning [16]. Early recognition is critical, especially in pediatric populations where unexplained neurocognitive symptoms may signal underlying shunts [17]. Treatment varies depending on severity. Initial management often includes ammonia-lowering agents like lactulose and rifaximin [18]. In refractory cases, endovascular closure using plugs or coils may be considered, provided liver histology excludes advanced fibrosis to avoid inducing portal hypertension [11,13]. In severe cases with significant hepatic dysfunction, liver transplantation may be required. According to current international guidelines, the management of hepatic encephalopathy should be individualized based on the underlying cause and symptom severity, with close follow-up to prevent recurrence [4,19,20].

5. Conclusion

This case highlights the importance of considering IPCS in the differential diagnosis of hepatic encephalopathy in non-cirrhotic patients. Comprehensive imaging and histological evaluation are essential for accurate diagnosis and treatment planning. In selected patients with stable comorbid conditions, such as chronic kidney disease, conservative therapy may yield favorable outcomes. Early identification and individualized management are crucial to prevent irreversible neurological consequences.

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