Title: Persistent encephalopathy in a non-cirrhotic- Don't shun this shunt

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Introduction

A portosystemic venous shunt is the formation of an abnormal connection between the portal vein and a systemic vein, allowing blood to bypass the liver. A large shunt can decrease hepatic reserve and liver's ability to detoxify blood. Spontaneous formation of a portosystemic shunt is rare. We report a case of large, spontaneous intrahepatic portosystemic shunt in a non-cirrhotic patient contributing to recurrent hepatic encephalopathy (HE).

Case

A 71-year-old female with a past medical history of non-alcoholic steatohepatitis, multiple episodes of gastrointestinal bleeding attributed to diverticular disease and stercoral ulceration , deep venous thrombosis and pulmonary embolism requiring an inferior vena cava filter, and atrial fibrillation presented with recurrent episodes of altered mental status for over 2 years. Initially, her encephalopathy was attributed to repeated infectious insults and dehydration. Due to persistent encephalopathy despite resolution of infection, an extended workup was performed including but not limited to thyroid function tests, vitamin levels, and dedicated cranial imaging – all of which were unrevealing. Ammonia levels were persistently elevated with a peak level of 310 µmol/L. Thus, HE was entertained as the presumptive diagnosis. This was further substantiated by her positive response to treatment with lactulose and rifaximin.

Further diagnostic imaging was pursued as she did not have clinical features consistent with cirrhosis to contextualize HE. Dedicated computed tomography (CT) imaging revealed two large (12-14 mm) intrahepatic venous malformations manifesting as a porto-venous shunt. On prior retrospective comparison imaging several years ago this was appreciated as an incidental finding, but less prominent. In addition, the magnitude of the shunt appeared to have contributed to capsular retraction resulting in nodularity of the liver.

Due to repeated episodes of HE from portovenous shunting through her intrinsic hepatic venous malformation, decision was made to perform coil embolization to close the anastomosis. On follow up the patient had no further episodes of HE and ammonia levels were normalized to 32 µmol/L.

Discussion

This is a unique case of a large spontaneous portovenous hepatic shunt without underlying cirrhosis contributing to persistent portal-systemic encephalopathy. Hepatic encephalopathy associated with porto-systemic shunting is known as Type B encephalopathy and occlusion of the shunt by endovascular management is the preferred treatment of choice (1,2). Providers should consider type B HE in patients with intractable or recurrent presentations of HE.

This abstract has not been presented elsewhere.

References

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