Confusion for Fifteen Years; A Case of Abernethy Malformation

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Kaartik Soota - literature review, drafting case, preparation of the manuscript.
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A 38-year-old male with previously undiagnosed Marfan’s Syndrome was admitted with confusion which had occurred episodically for almost 15 years. Serum ammonia was 180 (N:11-51) mmol/L. CT scan of abdomen revealed Abernethy Malformation. The superior mesenteric vein (arrow in A) and splenic vein (block-arrow in A) merged, draining directly into the left renal vein (arrow in B) causing a massively dilated inferior vena cava (IVC, star in A/B/C). His symptoms resolved with lactulose and he is being evaluated for liver transplant.

Abernethy Malformation is a rare anomaly of the splanchnic venous system and consists of a congenital porto-systemic shunt due to persistent embryonic vessels. There are two main types-Type 1 with an absence of the portal vein and diversion of portal blood into the systemic vasculature, and Type 2 with a hypoplastic portal vein and side to side portal blood diversion into the IVC.

Patients can be asymptomatic or present with confusion, hypoxia due to hepatopulmonary syndrome, or other vascular anomalies. It may also lead to focal nodular hyperplasia, hepatocellular carcinoma, or hepatoblastoma. Treatment options include liver transplantation, balloon retrograde transvenous obliteration, and surgical correction of shunts. Ours is the first report of concomitant Abernethy Malformation with Marfan’s Syndrome.