AN UNUSUAL CAUSE OF HEPATIC ENCEPHALOPATHY

Resident(s): Rajesh Bhavsar, MD

Program/Dept(s): Montefiore Medical Center, Albert Einstein College of Medicine
CHIEF COMPLAINT & HPI

- **Chief Complaint**
  - 71 year-old female presents with persistent lethargy and failure to thrive.

- **History of Present Illness**
  - 71 year-old female with history of mitral insufficiency had mitral valve repair about one month prior to presentation. She was transferred from rehabilitation facility to hospital with persistent lethargy and failure to thrive after the surgery.
RELEVANT HISTORY

- **Past Medical History**
  - multiple episodes of mitral valve endocarditis resulting in mitral valvular insufficiency
  - bicuspid aortic valve with aortic valvular stenosis
  - multiple infections and pneumonias of unclear etiology
  - incidentally found “liver abnormality” which was deemed asymptomatic on a CT from a few years ago.
  - no reported prior episodes of encephalopathy

- **Past Surgical History**
  - aortic valve repair
  - mitral valve repair one month before presentation
DIAGNOSTIC WORKUP

- Physical Exam
  - Asterixis

- Laboratory Data
  - Elevated ammonia level of 162 umol/L

- Non-Invasive Imaging
  - CT
The portal vein is marked by the letter:

A. a  
B. b  
C. c  
D. No portal vein is present.
The portal vein is marked by the letter:

A. a—this is an anomalous vessel
B. b—this is the portal vein, which is severely atretic
C. c—this is the IVC
D. No portal vein is present. — a portal vein is present.

CONTINUE WITH CASE
SORRY, THAT’S INCORRECT.

The portal vein is marked by the letter:
A. a—this is an anomalous vessel
B. b—this is the portal vein, which is severely atretic
C. c—this is the IVC
D. No portal vein is present.

CONTINUE WITH CASE
In this patient, the superior mesenteric vein drains into:

A. A normal caliber portal vein
B. The inferior vena cava
C. The left renal vein
D. A splenorenal shunt
In this patient, the superior mesenteric vein drains into:
A. A normal caliber portal vein
B. The inferior vena cava
C. The left renal vein
D. A splenorenal shunt
In this patient, the superior mesenteric vein drains into:
A. A normal caliber portal vein
B. The inferior vena cava
C. The left renal vein
D. A splenorenal shunt
Hepatic encephalopathy in a patient with congenital portosystemic shunt (Abernathy Malformation) onset by mitral valve annuloplasty.

The patient had a congenital portosystemic shunt which was compensated before heart surgery and essentially asymptomatic. The right heart failure served as a point of resistance for the portosystemic shunt forcing more blood into the diminutive portal vein. Following mitral valve repair, the right heart pressures decreased, allowing more blood flow through the shunt resulting in increased portosystemic shunting and symptoms of encephalopathy.
INTERVENTION

• Image A: Venography through the SMV demonstrates preferential flow through the shunt with little antegrade flow through a diminutive portal vein (PV) (arrow). Intravascular US showed the smallest caliber of the shunt vein to be 14mm in diameter.

• Image B: It was felt that if the shunt was occluded abruptly, the small PV would not be adequate to take all of the splanchnic flow and reducing flow in the shunt would be more prudent. A 16 x 60mm Wallstent which was constricted with a suture (arrow) was deployed within the shunt to make a landing zone for a PTFE covered device that will restrict flow.
INTERVENTION

- Images A and B: a 10mm x 38mm PTFE covered balloon-mounted stent (arrows) was positioned at the narrowed portion of the Wallstent and deployed in an hourglass shape using strategic balloon inflations.

- A second balloon (*) was inflated to prevent stent migration during deployment (Image B).
INTERVENTION

- Yellow arrow indicates the inner 10 x 38mm PTFE covered balloon-mounted stent.
- White arrow indicates the outer 16 x 60mm Wallstent.
Before (image A) and after (image B) venograms. Blood flow through the splenorenal shunt is reduced post-intervention.
CLINICAL FOLLOW UP

- Following the reduction of flow in the shunt, the patient’s ammonia levels decreased to 43 umol/L (from 162 umol/L), and the patient became alert with resolution of symptoms.
Abernathy malformation is a rare congenital anomaly of the splanchnic vasculature secondary to defects in vitelline vein formation. It is common in dogs (Yorkshire terriers) but extremely uncommon in humans.

Two types are described:

- Type I (females): absent or atretic portal vein with diversion of portal blood into systemic circulation; end to side shunts. Often associated with other abnormalities such as VSD and aortic arch defects.
  - Type Ia: separate drainage of the superior mesenteric and splenic veins into systemic veins.
  - Type Ib: superior mesenteric and splenic veins join to form a short extra-hepatic portal vein which drains into a systemic vein.
- Type II (males): hypoplastic portal vein with portal blood diversion into the vena cava through a side-to-side, extrahepatic communication.
REFERENCES & FURTHER READING
