CIRA Case of the Week
January 2017

Case Courtesy of Dr. Gillian Shiau
Chief Complaint & HPI

- **CC:**
  - 73 year old male with suspected hepatic encephalopathy

- **HPI:**
  - Recurrent hospital visits over the course of a year with vague neurological symptoms (weakness, confusion) originally thought to be related to TIAs
  - One ER visit, patient also c/o abdominal “cramp” pain diffusely throughout abdomen with no relation to meals, position, or bowel movements without N/V or change in BMs – further investigation revealed disturbance of his liver enzymes and elevated ammonia levels (> 200)
  - Treated with lactulose with symptoms clearing but treatment found to be difficult for the patient
Medical History

• Past Medical History:
  – Significant cardiac history (hypertension; 3 MI’s; atrial fibrillation with pacemaker/defibrillator insertion and subsequent lead infection requiring laser lead extraction; CHF with orthopnea)
  – NIDDM
  – Parkinson’s Disease
  – No significant history of trauma
  – No history of liver interventions including biopsy

• Social History:
  – Social drinker
  – Non-smoker

• Family History:
  – Non-contributory

• Allergies:
  – Bactrim
  – Morphine

• Home Medications:
  – Furosemide 80 mg po OD
  – Warfarin 5 mg po OD
  – Nitroglycerin patch transdermal 0.6 mg daily
  – Pantoprazole 40 mg po OD
  – Glicazide 30 mg po OD (in AM)
  – ECASA 81 mg po OD
  – Potassium chloride 8 mEq po BID
  – Levocarb 100/25 one tab po TID
  – Tamsulosin 0.4 mg po OD
  – Lactulose 60 cc po TID – patient’s symptoms rapidly recur if he misses a dose
Diagnostic Workup – Labs

AST: 27
ALT: 22
ALP: 120
Bilirubin (total): 50.0
Bilirubin (direct): 19.5
Ammonia: 176 (previously > 200)
Imaging
CTA – Coronals (ant to post)

- Right portal vein
- Fistulous communication
- Right hepatic vein
Intervention

- R IJV access
- Catheterization of shunt and across into the portal system
- Venogram
Intervention

- Pressures were measured
- Attempted occlusion balloon inflation (balloon would not stay seated)
- Procedure terminated with plans for alternative approach
Intervention

• Percutaneous access into a left peripheral portal venous branch was obtained

• Venogram showed rapid filling of the venous fistula
Intervention

- Multiple feeding vessels were identified, progressively selected and embolized using 4th generation Amplatzer vascular plugs.
Intervention

• Completion venogram confirmed absent flow through the venous fistula

Fistula excluded by AVPs

Incidental left lobe lesion

Right

RPV

MPV
Clinical Follow-up

• Post-embolization in-hospital course complicated with bouts of shortness of breath at night attributed to worsening CHF – resolved with increased Lasix dose
• Patient transferred to home hospital
• Patient doing well – hepatic encephalopathy symptoms have resolved 3 months out
Discussion

• Etiology/pathogenesis is controversial with multiple theories:
  – Congenital (multiple pediatric cases) due to a persistent embryonic venous anastomosis or in patients without history of liver disease or abdominal trauma
  – Acquired due to portal hypertension from cirrhosis/hepatitis; iatrogenic; post-traumatic; rupture of a portal venous aneurysm into a hepatic vein
• Internal type thought more likely congenital as low prevalence of coexisting cirrhosis
• Clinical significance of this entity: potential for hepatic encephalopathy (can consider it as a “natural” TIPS)
  – Postulated that there is increased risk correlated with increasing age (brain may be more susceptible to toxic metabolites) and larger shunts
Management

• Asymptomatic – watchful waiting
• Symptomatic – first step is dietary management limiting protein intake and lactulose supplementation; if fail dietary modifications, consider intervention to shut down the shunt
• Multiple case studies reporting successful embolization of IPSVS have been detailed in the literature
• Only about 50 cases had been reported in the literature as of 2003
• 47 publications in the literature describe types II-IV (Chevalier classification – those usually not associated with portal hypertension)
• Multiple different approaches including transjugular; transileocolic; percutaneous transhepatic; and retrograde transcaval
References