

Portal Hypertension – What Baveno Does Not Tell Us ...

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Case Report

- 44 year-old woman
- Born in Kazakhstan; Immigrated to Israel in 2000
- Past Medical History:
 - Hepatitis C – Successfully treated with standard Interferon- α - 2001
 - Upper GI bleeding; Gastric operation? – 1991
 - Pancreatic cyst (Echinoccocal?) removal - 1994

Case Report

- Admitted for hematemesis and syncope – 24.9.15
- Gastroscopy:
 - Stomach:
 - Large amount of blood clots and “coffee ground” blood residue
 - Small fundic varices –
No stigmata of recent bleeding
- Treated with:
 - IV Terlipressin, Ceftriaxone, Pantoprazole

Case Report

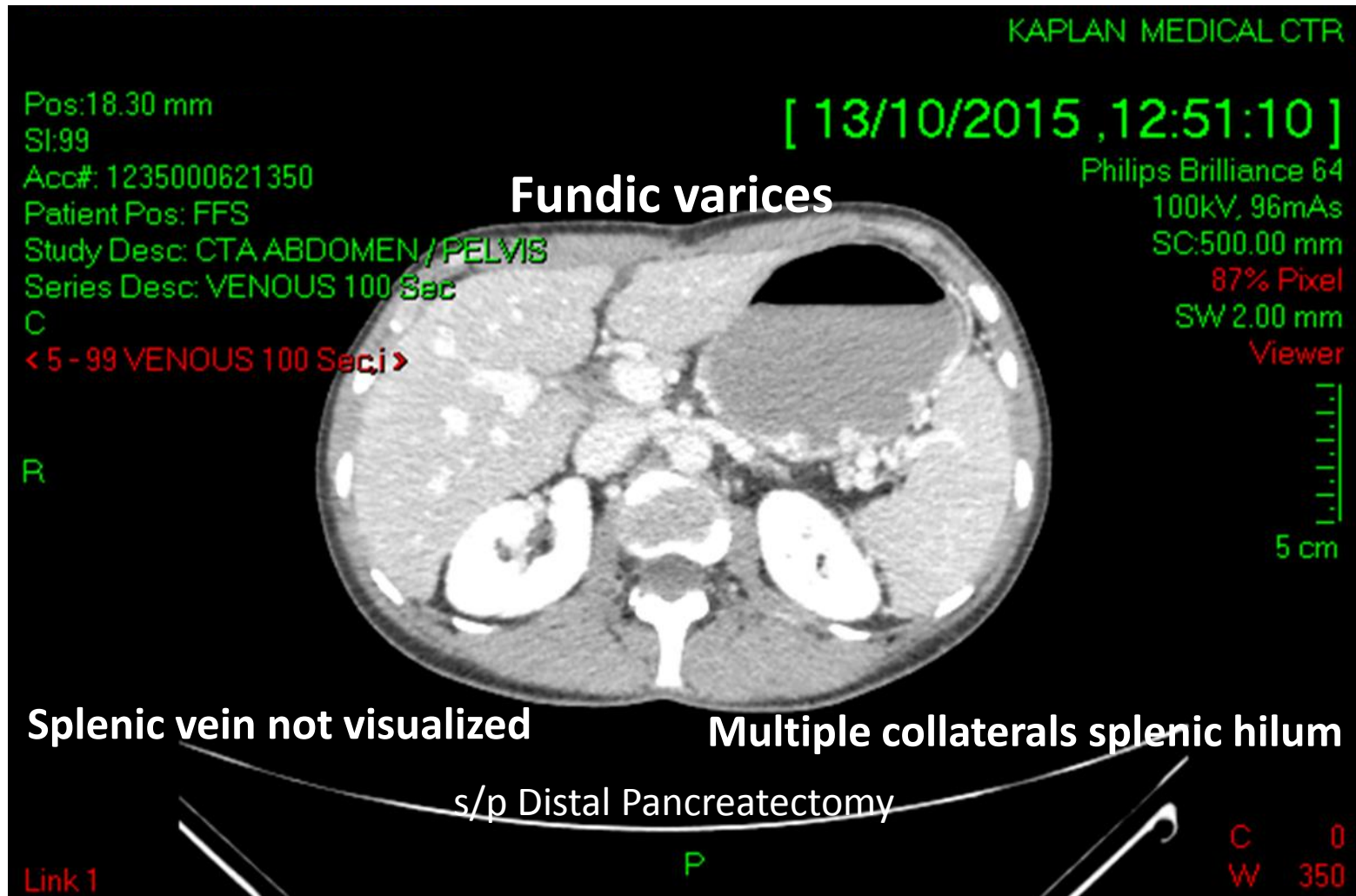
- 4 days following admission (28.9.15) - Early re-bleeding:
 - Transferred for consideration of TIPS
- Doppler US:
 - Heterogeneous liver texture
 - Hepato-petal portal vein flow
 - Normal hepatic veins
- Ligation of a fundic varix
- Discharged with β -blockers

Case Report

- 10 days following discharge (9.10.15) -
Admitted with re-bleeding
- Gastroscopy:
 - No fundic varices !

What is the Next Step ?

CT Angiography



?

Splenic Artery Embolization



Has not bled since

Left-sided - Sinistral Portal Hypertension (LSPH)

- Usually occurs as a result of isolated obstruction of the splenic vein
- Accounts for less than 5% of all patients with portal hypertension
- Flow through the splenic vein may be blocked secondary to:
 - Thrombosis - Most common
 - Neighboring mass effect - Less frequent

Etiology of LSPH

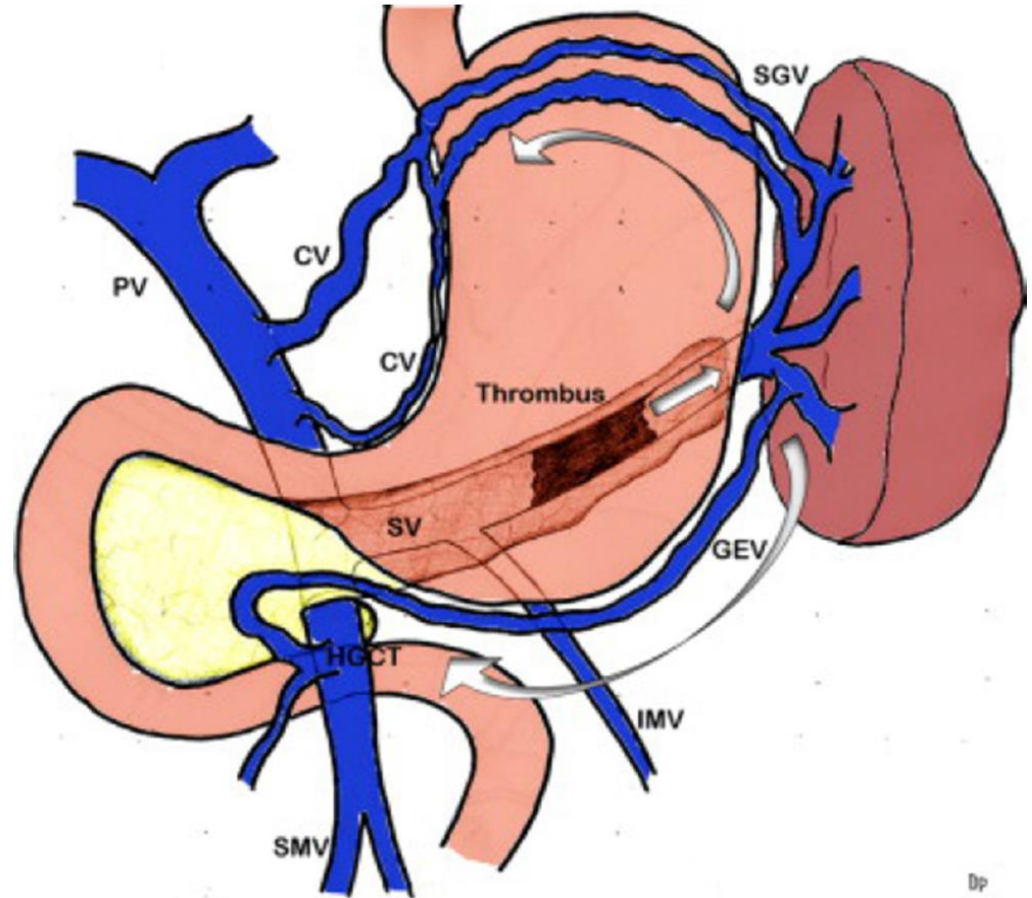
- Most common etiologies:
 - Chronic pancreatitis
 - Pancreatic pseudocysts
 - Pancreatic neoplasms
- Pseudocyst in chronic pancreatitis is associated with higher incidence of splenic vein thrombosis
- Rare etiologies:
 - Iatrogenic splenic vein injury
 - Infiltration by colonic tumor
 - Spontaneous splenic vein thrombosis
 - Peri-renal abscess

Clinical Manifestations

- Most commonly LSPH is **asymptomatic** and is found incidentally
- The first clinical manifestation is generally acute or chronic GI bleeding from esophageal/gastric varices
- Bleeding is usually serious
- The incidence of gastric bleeding in LSPH varies:
4% - 72%

Pathogenesis of LSPH

- Collateral vessels develop to shunt blood around the occluded splenic vein



Pathogenesis of LSPH

The two most common collateral pathways use the **short gastric veins**

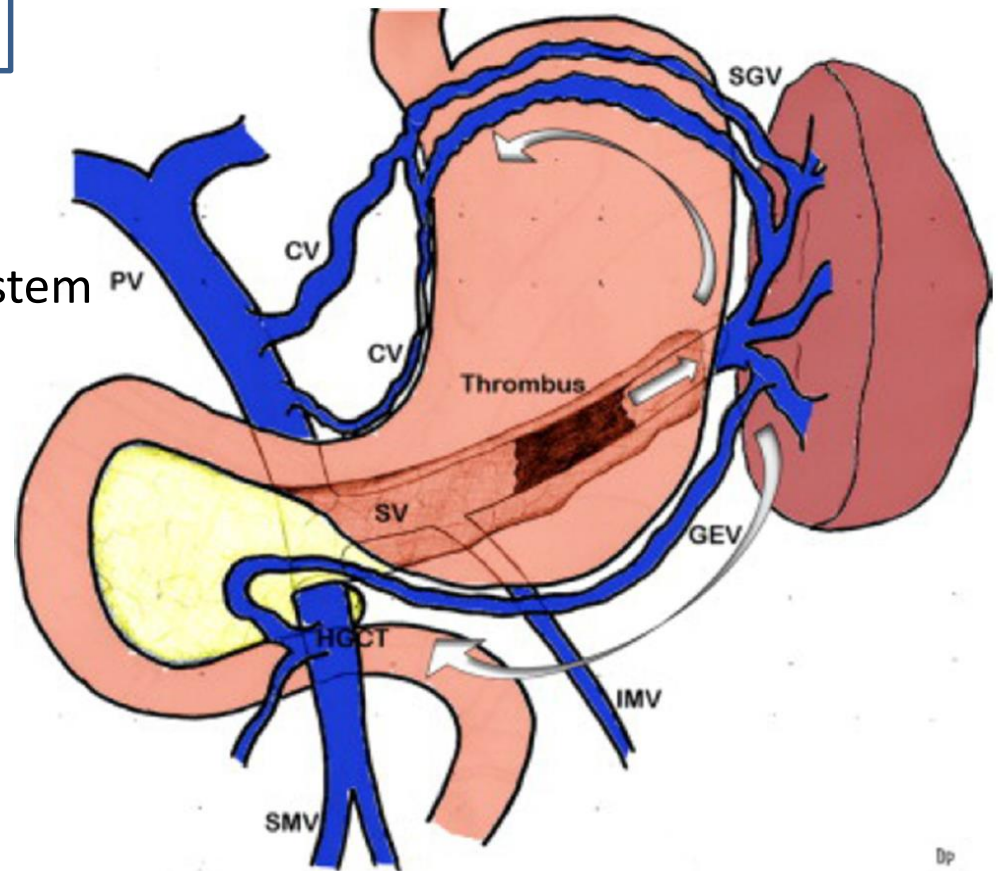
Porto-systemic collaterals:

Short gastric veins into the azygous system in the distal esophagus

Spleno-portal collaterals:

Decompress the short gastric veins through:

- The coronary vein into the portal vein
- The gastroepiploic arcade into the superior mesenteric vein



Pathogenesis of LSPH

↑Flow and ↑pressure in the short gastric veins

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graph TD; A[↑Flow and ↑pressure in the short gastric veins] --> B[↑Pressure within submucosal veins of the gastric fundus]; B --> C[Veins dilate]; C --> D[Formation of varices];
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↑Pressure within submucosal veins of the gastric fundus

Veins dilate

Formation of varices

Management

- The first-line treatment for bleeding gastric varices is endoscopic, preferably Cyanoacrylate injection
- Patient with active bleeding unresponsive to endoscopic management should be submitted to an emergent splenectomy
- Splenectomy decreases the venous outflow through the collateral circulation and decompresses the associated varices

Management

- Embolization of the splenic artery may be an option in patients who are not medically fit for splenectomy
- Embolization as a pre-operative adjunct to splenectomy has also been advocated
- Splenic infarction and abscess formation may occur after the embolization procedure

Splenic Arterial Embolization for Bleeding Gastric Varices and LSPH

- The one-step method – 3 patients with massive gastric bleeding
 - The bleeding was relieved after embolization
- The two-step method - 11 patients with chronic gastric variceal bleeding or gastric varices only
 - Gastric varices gradually disappeared on enhanced CT scan
- There was no gastric bleeding during the 4-months follow-up

Splenic Arterial Embolization for Bleeding Gastric Varices and LSPH

- Complications:
 - Post-embolization syndrome - LUQ abdominal pain and fever - Most frequent complication
 - Infection - 1 patient after the 1-step method – Probably caused by spleen necrosis
 - Platelet count $> 10^6/\text{L}$ - 3 patients
- Splenic abscess and rupture of spleen did not occur in any of the patients

While hospitalized ...and Prior to splenic artery embolization

- The patient presented with:
 - Involuntary spasms of both hands, change of speech tone and difficulties with walking
 - Developing over a 2 week period

Neurologic Examination

- Remarkable for parkinsonism:
 - Hyponymy, hypo phony, bradykinesia
 - Marked symmetric rigidity more prominent in the upper limbs
 - Dystonic posture of both hands and the right feet
 - **No signs of overt hepatic encephalopathy**

Work-up

- Ceruloplasmin levels and urinary excretion of copper - Normal
- No evident of Kayser Fleischer ring
- Ammonia blood level - Normal
- CSF analysis - Normal
- EEG findings - Unremarkable
- Serology testing for VDRL and TPHA and HIV - Negative

MRI – T1-Weighted

Bilateral and symmetrical
hyper-intensities
Globus Pallidus
(part of the Basal Ganglia)



Outcome

- Treatment with anti-parkinsonian medication Amantadine and low dosage of Clonazepam were initiated
- Progressive recovery of the parkinsonism was observed

What is the Diagnosis ?

Acquired Hepato-cerebral Degeneration (AHCD)

- A disorder characterized by parkinsonism, ataxia and other movement disorders in patients with advanced liver disease
- In case series of cirrhotic patients variable degrees of parkinsonism were manifested in 4% - 22%
- The course of AHCD is generally progressive but highly variable
- Symptoms may not parallel other features of liver disease **including hepatic encephalopathy**

Acquired Hepato-cerebral Degeneration (AHCD)

- The presence of significant porto-systemic shunts is considered a precondition for the development of AHCD
- It has been proposed that increased manganese deposition in the basal ganglia has a major role in the pathogenesis of AHCD

Acquired Hepato-cerebral Degeneration (AHCD)

- The hallmark MRI findings include:
 - Symmetric hyper-intensities on T1-weighted imaging in the basal ganglia
- Hyper-intense signals in the basal ganglia were observed on MRI in ~70%-80% of asymptomatic cirrhotic patients

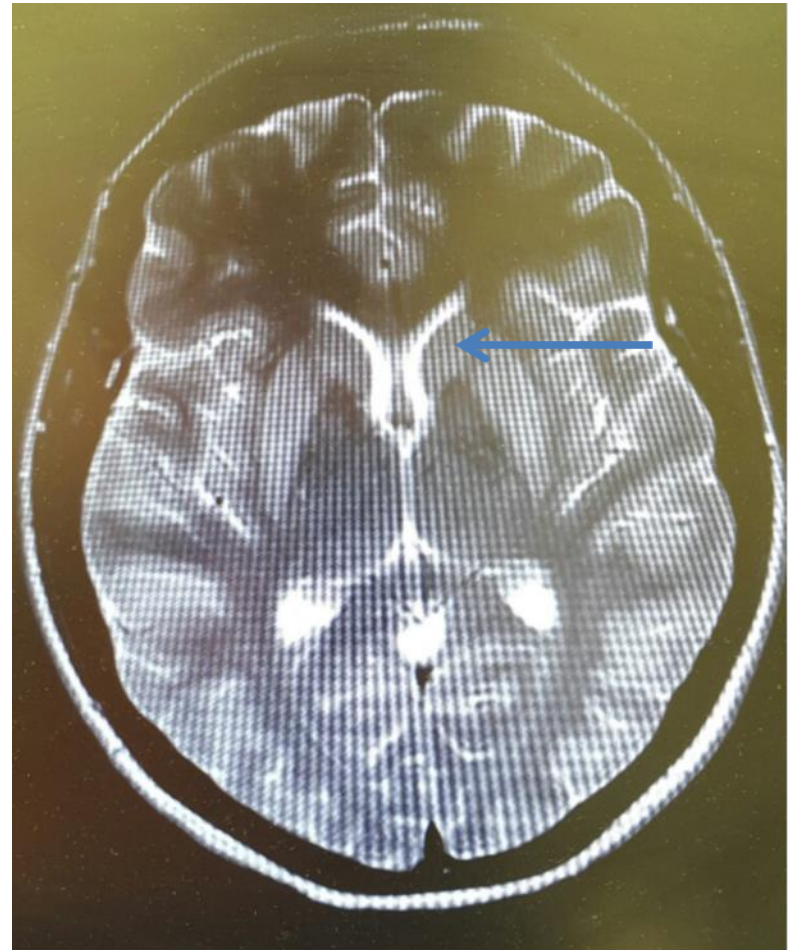
Acquired Hepato-cerebral Degeneration (AHCD)

- These MRI changes were found in 5/6 patients with complete portal vein thrombosis without cirrhosis
- Case reports:
 - Progressive extrapyramidal symptoms and typical MRI lesions in patients with portal vein thrombosis and marked collateral circulation
 - Reversal of symptoms following recanalization of portal vein and less evidence collateralization

Thank You

MRI – T2-Weighted

Bilaterally, symmetric swelling
and hyper-intensities -
Caudate Heads



MRI – T1 Weighted

- Bilateral and symmetrical hyper-intensities in the:
 - Substantia nigra
 - Cerebral peduncle
 - Sub-thalamic region
 - Hemispheric white matter
 - Globus pallidus
 - Putamen