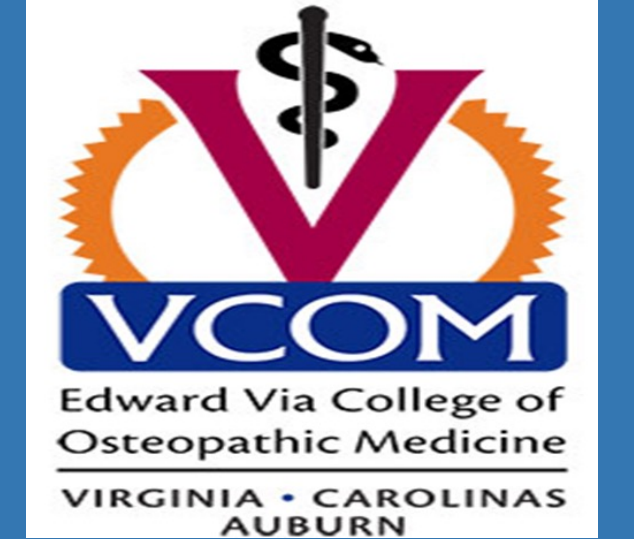




NODULAR REGENERATIVE HYPERPLASIA (NRH) AS A CAUSE FOR NON-CIRRHOTIC PORTAL HYPERTENSION



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Initial Presentation

Consult Request for management of “cirrhosis”.

HPI:

Patient is a 42-year-old-male with a twelve-year history of Common Variable Immune Deficiency (CVID). His only complaint at initial visit was mild, non-specific abdominal pain. He reported no history of jaundice, weight loss, or constitutional symptoms. Patient denied significant history of alcohol intake. He was receiving immune globulin (Gamunex-C®) every 28 days.

Objective Findings

Physical Exam

- Middle aged male in no acute distress with stable vital signs.
- Mild abdominal tenderness and palpable “fullness”.
- Examination otherwise unremarkable without ascites or clinical signs of chronic liver disease.

Imaging:

- Abdominal CT revealed massive splenomegaly (Figure 1), portal vein enlargement with no ascites.

Labs:

- Elevated Alkaline Phosphatase
- Otherwise normal

Esophagogastroduodenoscopy (EGD):

- Small esophageal varices

Transjugular Liver Biopsy (Figure 2):

- Minimal fibrosis but with features of nodular regenerative hyperplasia (NRH)

Hemodynamic Measurements:

- FHVP: 10 mmHg
- WHVP: 22 mmHg
- HVP: 12 mmHg (nl 1-5 mmHg)

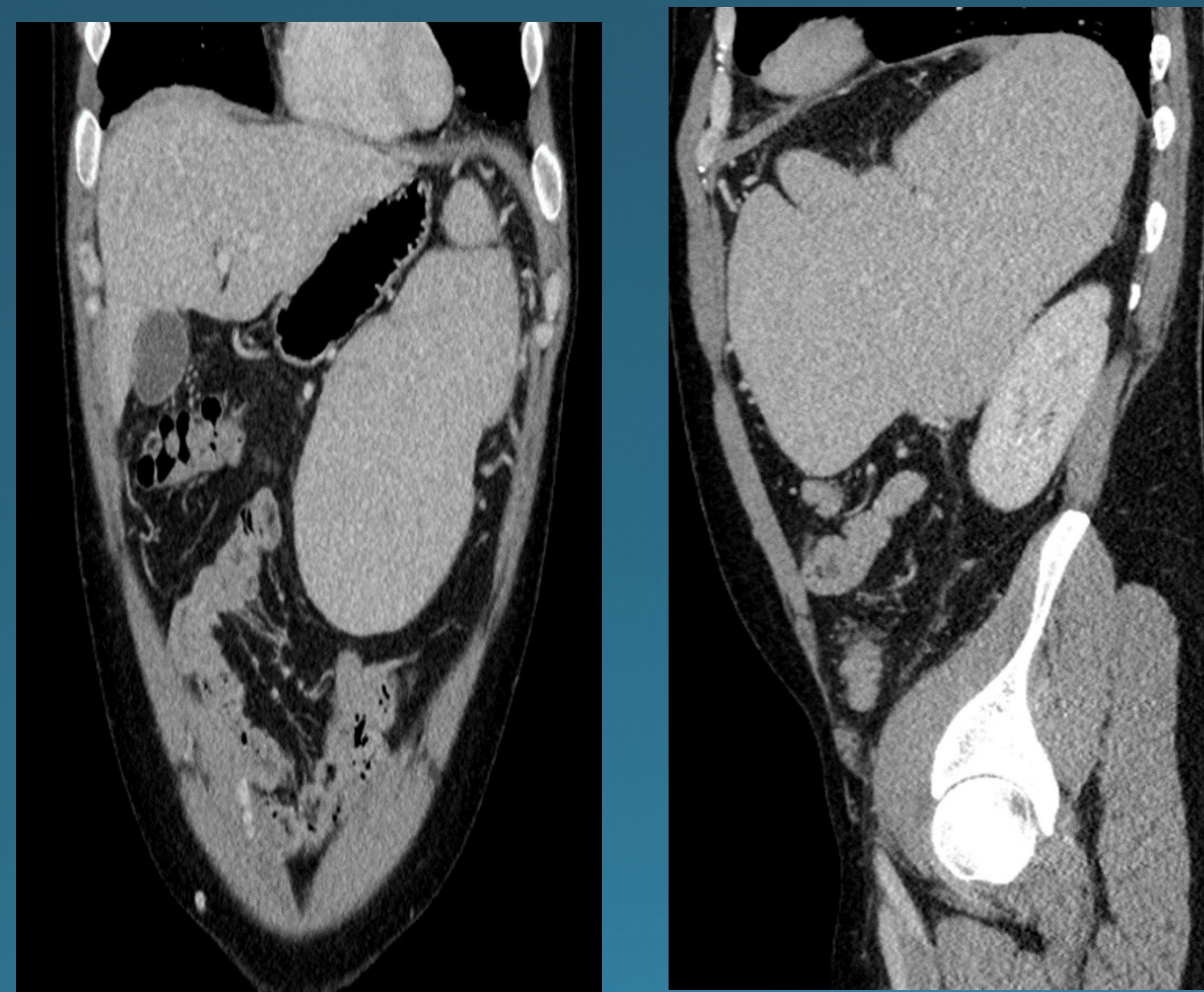
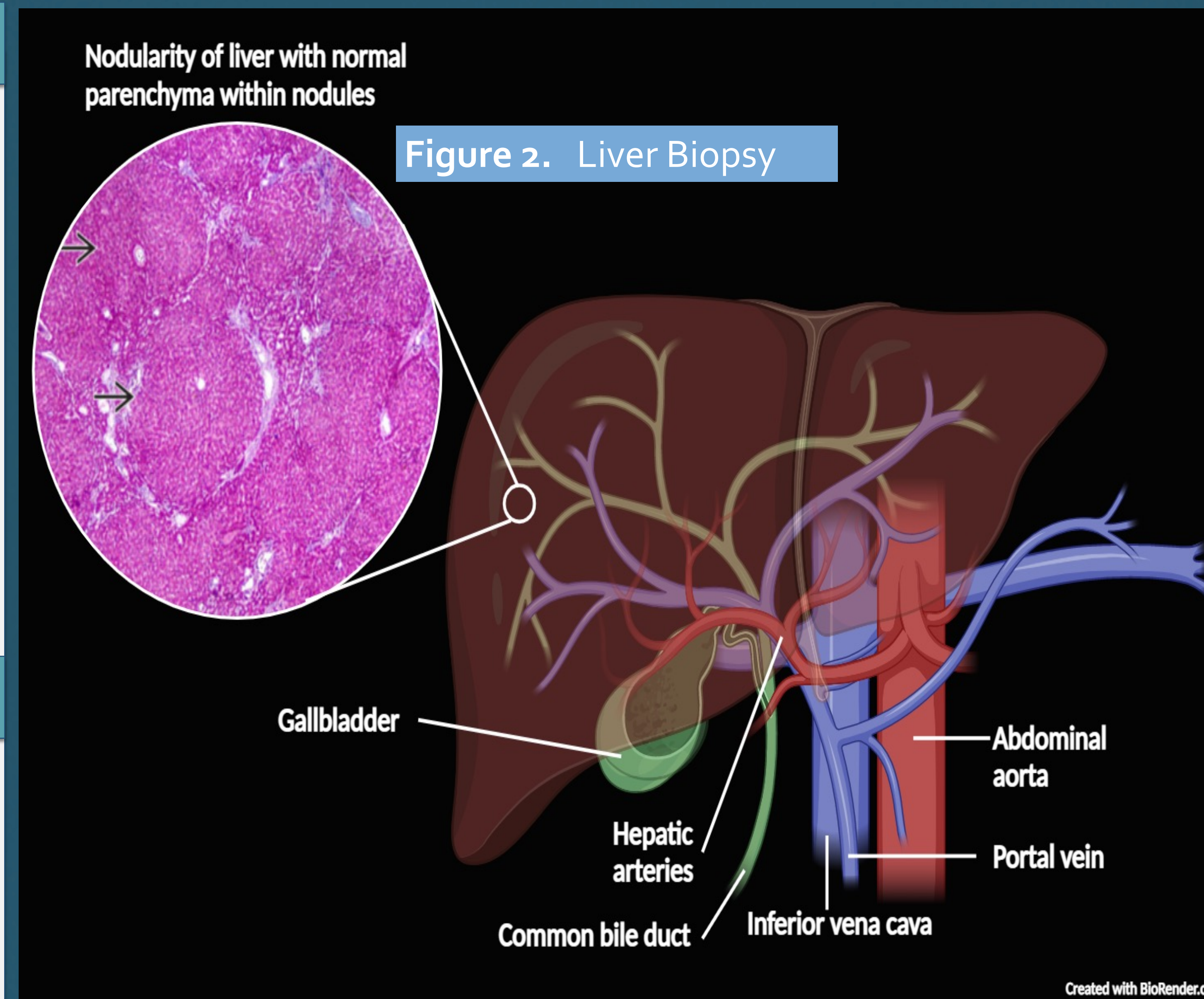


Figure 1. Splenomegaly

Discussion

- CVID is the most common immunodeficiency disease with a prevalence of 1 in 25,000.
- It is the primary B-cell disorder with hypogammaglobulinemia.
- NRH can occur in 5-10% of CVID patients and the most common hepatic dysfunction is CVID is nodular regenerative hyperplasia.
- Mixed:
 - Pre-sinusoidal portal hypertension (PHT) from obliterative portal venopathy.
 - Sinusoidal PHT from obstruction of compression of regenerative nodules.

Treatments

- Non-selective Beta Blockers
- Variceal Ligation
- Mesenteric-Caval Shunts
- Transjugular Intrahepatic Portosystemic Shunt (TIPS)

Acknowledgements & Citations

