Leaking Curiosity: Post-Operative Drainage Leading to Revelation of Noncirrhotic Portal Hypertension in a Patient With Suspected Wilson's Disease

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INTRODUCTION

In patients with an elevated serum ascites albumin gradient and low ascitic protein, the presumption is often that ascites is secondary to cirrhosis. Here, we present an interesting case of portal hypertension without significant fibrosis.

CASE DESCRIPTION

A 61 year old male with history of "chronic encephalopathy," severe agitation, dysphagia, Roux-en-Y gastric bypass, anemia, and chronic pain presented with 2 days of confusion, nausea, emesis, and abdominal pain accompanied by distention. Imaging was notable for massive pneumoperitoneum as well as abdominopelvic ascites. He was emergently taken to surgery, undergoing repair of a perforated marginal ulcer and subsequent placement of a JP drain near the gastrojejunal anastomosis at the right upper quadrant. He was downgraded from the surgical ICU to a floor medicine team with significant output from the JP drain (500-1500 mL daily), which surgery attributed to expected leakage in the context of peritonitis and severe malnutrition; nutritional deficiencies included zinc, vitamin A, and vitamin D. On further investigation, ascitic fluid studies were consistent with portal hypertension presumed secondary to cirrhosis. Duplex abdominal ultrasound was notable for ascites and slightly nodular liver surface, prompting further evaluation.

Chronic liver disease workup was notable for low ceruloplasmin at 12 mg/dL and elevated 24 hour urine copper at 165 mcg. Considering patient's overall history and concerns that his neuropsychiatric disturbances, ocular complaints, anemia, arthralgias, and presumed cirrhosis were due to Wilson's disease, Hepatology was consulted and recommended liver biopsy with quantitative copper in addition to evaluation by Neurology and Ophthalmology to add to the clinical picture. Biopsy was read as noncirrhotic portal hypertension with patchy sinusoidal dilatation and portal vein changes, suggestive of hepatoportal sclerosis/sclerosing portal venopathy; no evidence of significant fibrosis, and copper was normal at 16 ug/g. Patient was discharged from the hospital and later passed away without further evaluation.

DISCUSSION

This patient's clinical picture and laboratory results were suspicious for late diagnosis of Wilson's disease, though lack of assessment for Kayser-Fleischer rings renders it challenging to establish this as the case. That hepatoportal sclerosis was the suggested etiology for this patient's noncirrhotic portal hypertension is curious and would require exclusion of chronic liver disease.