Cavernous Transformation of the Portal Vein in the Setting of Protein C Deficiency

Abigail Carroll, D.O. and Jennifer Cox, D.O.
Oklahoma State University Medical Center Department of Internal Medicine – Tulsa, Oklahoma

Abstract

In this report, we present a case of a young female in the postpartum period that presented with liver cirrhosis and severe portal hypertension that resulted in acute esophageal variceal bleed. Imaging discovered the presence of cavernous transformation of the portal vein due to chronic thrombosis from presumed protein C deficiency. Her case was complicated by co-existing cirrhosis of unknown cause and brings into question the chronologic pathogenesis of the diagnosis.

Introduction

Chronic portal vein thrombosis can result in cavernous transformation of the vasculature, which is characterized by the formation of collaterals in the hepatic hilum to bypass the obstruction.[1] The presence of this in a patient, especially in a young female, warrants a workup for an underlying hypercoagulability, such as protein C deficiency.

Protein C deficiency is a form of inherited hypercoagulability that can lead to portal vein thrombosis.[2] Most patients with protein C deficiency have a risk factor that led to VTE: sepsis, immobilization, or OCP use. [3]

Identification of an underlying hypercoagulability helps understand both the pathogenesis and further treatment options we have for any given patient.

Case Presentation

A G3P3 23-year-old female with no past medical history presented for hematemesis. Three months prior to presentation, she had delivered her third child. Since delivery, she had required multiple iron and blood transfusions. Imaging showed cirrhosis with thrombosis of the main and right portal vein, splenic vein, superior mesenteric vein and notable ascites, esophageal/gastric varices, and splenomegaly. Further workup noted severe pancytopenia. She was transfused with two units of packed red blood cells and initiated on proton pump inhibitors plus octreotide.

Further laboratory workup for inherited hypercoagulability and cirrhosis initially proved unremarkable; however, protein C activity was notably decreased at 36%. Her hemoglobin (Hgb) continued to drop, so she was taken for endoscopy that revealed grade III esophageal varices that required extensive banding.

Figure 1: varices noted in lower 1/3 of esophagus

Her Hgb remained stable after banding, and she was discharged with recommendation to follow up outpatient with transplant hepatology.

Discussion

Cavernous transformation occurs in longstanding chronic portal vein thrombosis. In our patient, it was found that she had decreased protein C levels, which may have been a factor that predisposed her to developing this.

She was also found to have cirrhosis of the liver due to unknown cause with portal hypertension.

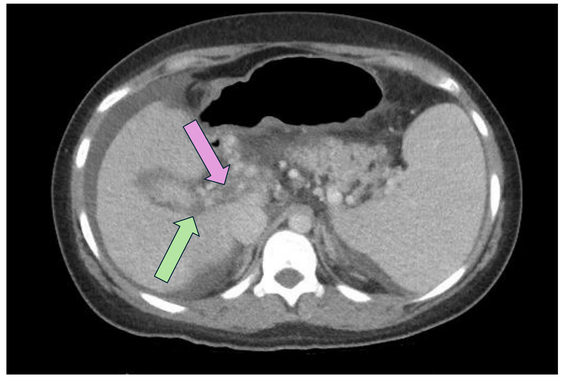

Figure 2: Portal vein thrombus (purple arrow) with surrounding cavernous changes (green arrow)

It is difficult to assess whether this patient's cirrhosis or portal thrombosis occurred first as she had no significant risk factors in her personal or family history for either diagnosis. She ultimately will require further investigation including liver biopsy, genetic testing, and continued following with a hepatologist for consideration of transplant due to her advanced disease state.

Conclusion

This patient's portal hypertension was accelerated by the presence of portal vein thrombosis that ultimately caused cavernous transformation. This case outlines that evaluation for protein C deficiency in those with portal vein thrombosis could aid in earlier identification and improved morbidity.

References

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