Image of the Month

Esophageal Variceal Hemorrhage Secondary to Post-Polycythemic Myelofibrosis

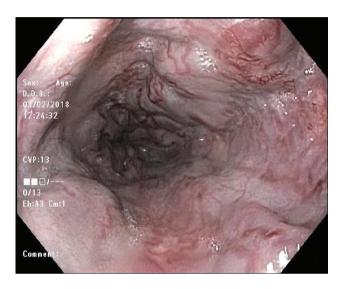


Figure 1. Upper endoscopy demonstrating medium esophageal varices with high-risk stigmata. A fibrin clot was present at the distal aspect of one column (not shown). He also had nonbleeding gastroesophageal varices (GOV1 and GOV2) (not shown).

A 71-year-old male presented to the emergency department with hematochezia and a hemoglobin drop from 90 to 70 g/L. His medical history was significant for polycythemia rubra vera that transformed to myelofibrosis (MF), but there was no known liver disease. On examination, he was hemodynamically stable with marked splenomegaly, but he had no other signs of portal hypertension (PH). Bloodwork demonstrated normal liver enzymes and function. Upper endoscopy showed varices (Figure 1). CT venogram revealed patent portal and hepatic veins with no thrombus. Massive splenomegaly (>30 cm) was noted. A severely dilated splenic vein (2.5 cm) communicated with an enlarged portal vein (1.8 cm) (Figure 2). Endoscopic variceal ligation (EVL) was performed and nonselective beta-blocker (NSBB) therapy was initiated before discharge.

As a complication of MF, PH can occur without development of portal vein thrombus (1). PH has rarely been documented in post-polycythemic MF (2). The mechanism of PH in absence of thrombosis is proposed to be extramedullary hematopoiesis in the liver and infiltration of sinusoids with

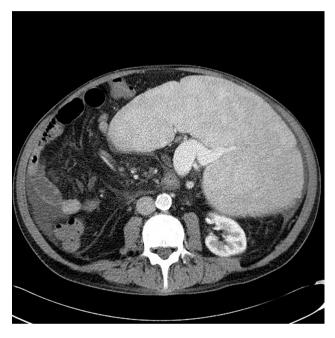


Figure 2. Axial CT sections demonstrating massive splenomegaly with a severely dilated splenic vein.

myeloid cells or increased portal flows secondary to splenomegaly (3, 4).

Small studies have shown PH develops in approximately 11% of MF individuals; however, the National Comprehensive Cancer Network provides no guidance on screening for portal hypertension (2,4). Traditional strategies to treat complications of cirrhotic PH have been utilized for MF, including EVL, NSBB and transjugular intrahepatic portosystemic shunting (1, 2). Given the frequency of PH in MF and potential catastrophic complications, further study should explore potential merits of endoscopic screening.

ACKNOWLEDGEMENTS

DM described the case. DM and TK conducted the literature review and wrote the manuscript. TK conceived the study. IE supervised the project.

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