Esophageal Varices in Myelofibrosis Associated Non-Cirrhotic Portal Hypertension: A Call to Screening

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BACKGROUND

Myelofibrosis is characterized by bone marrow hematopoietic failure, leading to extramedullary hematopoiesis and can be due to various causes such as primary myelofibrosis or chronic proliferative neoplasms i.e. polycythemia vera myelofibrosis and essential thrombocythemia.

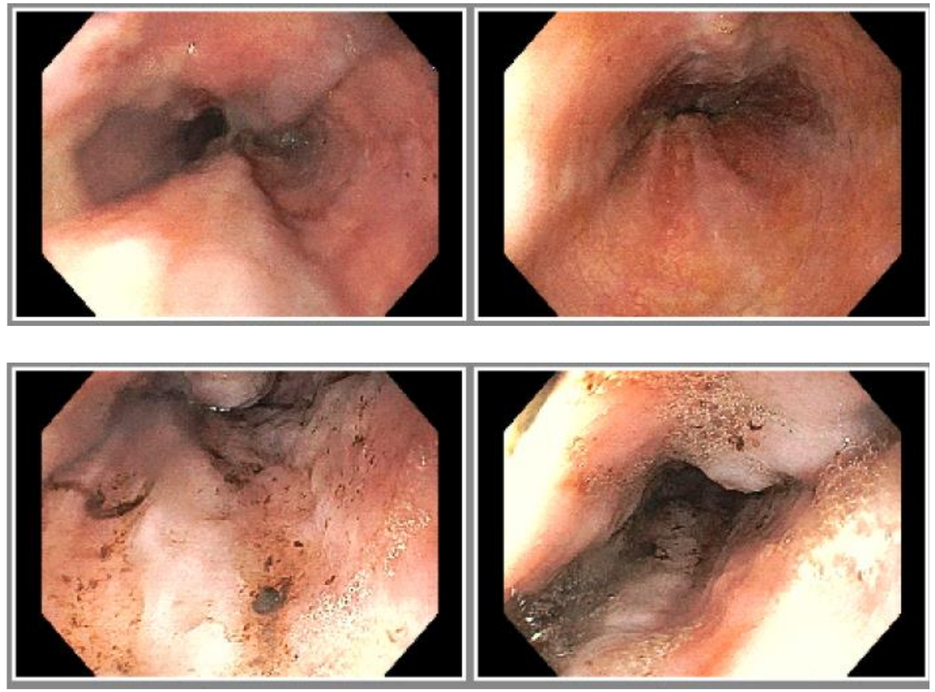
In the setting of primary polycythemia, Myelofibrosis can lead to non-cirrhotic portal hypertension with esophageal varices as demonstrated in this clinical presentation. Although esophageal varices development is a relatively uncommon complication in this setting, given its increased mortality, it is recommended these patients undergo screening and surveillance upper endoscopy.

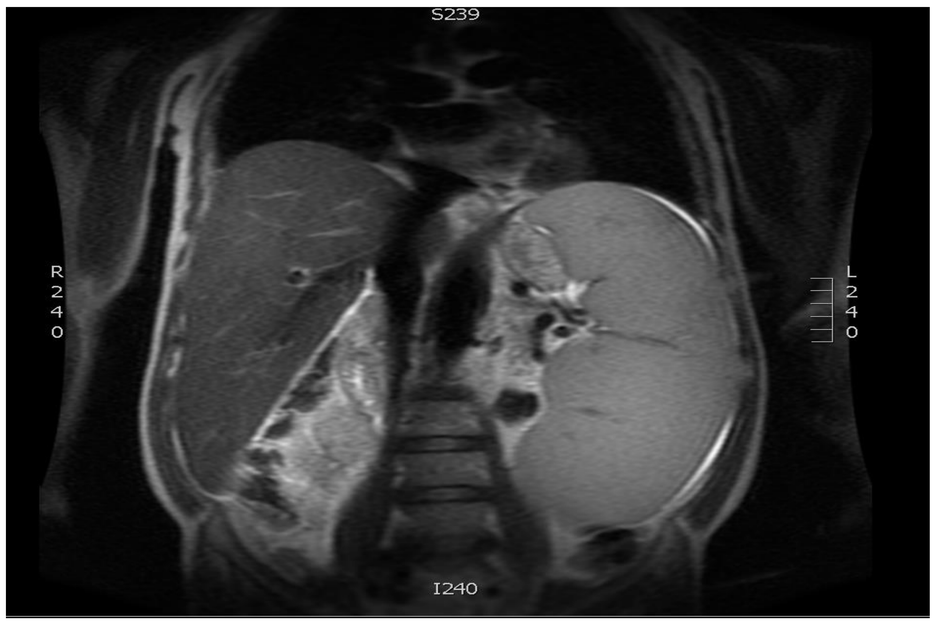
CASE PRESENTATION

A 62-year-old man presented to emergency department for several episodes of acute hematemesis and pre-syncope. Past medical history consisted of myelofibrosis secondary to primary JAK2+ polycythemia vera on daily aspirin and ruxolitinib and primary adrenal insufficiency treated with hydrocortisone and fludrocortisone. Physical examination was pertinent for splenomegaly and hepatomegaly without ascites or edema.

Initial laboratory evaluation was unremarkable with a WBC of 2.8 x 10^3/mcL, hemoglobin 7.0 g/dL, platelets 250 x 10^3/mcL. Upper endoscopy (EGD) revealed diffuse gastric erosions and small esophageal varices with no high-risk stigmata or active source of bleeding.

An MRI of his liver showed massive splenomegaly up to 27.4 cm with multiple accessory spleens, hepatomegaly up to 27.6 cm, esophageal varices, and an otherwise normal appearing liver. Previous splenic length was 19.1 cm from an ultrasound performed in 2019. He had an unremarkable doppler ultrasound evaluation of upper abdominal vasculature with normal phasic flow in hepatic and hepatopetal flow in portal venous system. Fibroscan was obtained with kPa of 12. Chronic liver disease workup including infectious, autoimmune, and metabolic causes was unremarkable. The patient declined liver biopsy and the plan was made for follow-up EGD in 1 year to survey the small varices seen on the original EGD.





DISCUSSION

While the exact mechanism of portal hypertension secondary to myelofibrosis is still under investigation, several pathophysiologic mechanisms have been proposed including: hepatic infiltration of hematopoietic stem cells, sinusoidal fibrosis, increased pre-hepatic flow as a result of profound splenomegaly, and portal venous insults.[2] As portal hypertension has been observed in 10% to 17% of patients with idiopathic myelofibrosis[3] , secondary myelofibrosis is thought to be a risk factor as well.

Given the increased risk of developing esophageal varices, patients in this population may benefit from variceal screening to proactively treat and prevent variceal hemorrhage. This is especially important as variceal hemorrhages have been associated with a mortality of at least 20% at 6 weeks. [4]

Additional imaging modalities such as utilization of Fibroscan in risk stratifying these patients remains to be established with most studies limited to small retrospective case series.

CONCLUSION

Myelofibrosis can lead to the formation of non-cirrhotic portal hypertension and development of esophageal varices.

This patient population would benefit from routine EGDs for variceal screening.

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