Extensive Portomesenteric Venous Thrombosis due to JAK2 V617F Mutation as an Indication for Multivisceral Transplantation

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Introduction

- Cavernous transformation of the portal vein (CTPV) involving the periportal or intrahepatic venous collateral network is a sequelae of chronic portal vein thrombosis (PVT).
- This adaptation increases the risk of complications and poor outcomes from revascularization procedures.
- In cases with diffuse portomesenteric venous thrombosis, multivisceral transplantation (MT) may be indicated.
- **Presented is a rare case of chronic PVT in a patient with a JAK2 mutation leading to CTPV refractory to anticoagulation and endovascular intervention, eventually requiring MT.**

Case Presentation

- **Age 43**: An otherwise healthy female was diagnosed with Janus kinase 2 V617F (JAK2) mutation after developing PVT of unknown etiology.
  - Her disease progressed with bleeding esophageal varices, portal hypertensive gastropathy, and splenomegaly.
  - Liver biopsy was negative for cirrhosis. Bone marrow biopsy was negative for leukemia or fibrosis.
- **Age 45**: She developed mesenteric vein thrombosis (MVT) and started rivaroxaban. She was transitioned to high dose enoxaparin due to worsening thrombus progression.
- **Age 47**: She underwent thrombolysis for new MVT.
- **Four months later (current presentation):** She endorsed worsening abdominal pain.

Clinical Evaluation & Treatment

- Physical exam demonstrated ascites and pitting lower extremity edema.
- **Abdominal ultrasound with venous duplex** demonstrated absent flow in the portal vein.
- **Computed tomography angiography (CTA)** revealed progressive portal, superior mesenteric, and splenic vein thromboses despite anticoagulation compliance.
- **Transjugular intrahepatic portosystemic shunt (TIPS)** procedure was unsuccessful, likely due to CTPV (*figure 1a*).
- **Post-procedure CTA** demonstrated an enlarging hepatic hematoma treated by embolization (*figure 1b*).
- Anticoagulation was discontinued resulting in worsening clot burden and hepatic ischemia.
- Given the extent of clot burden, the patient was transferred to another facility for a MT.

Imaging

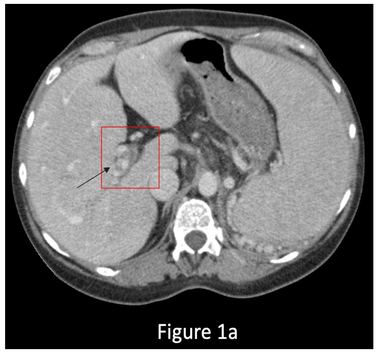

Figure 1a

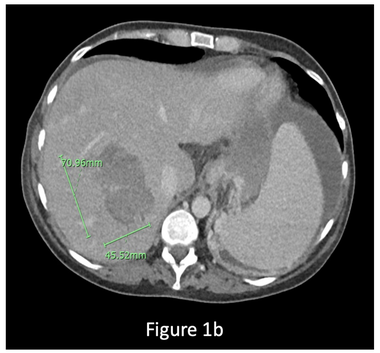

Figure 1b

Figure 1a. Cavernous transformation of the portal vein, as indicated by the arrow.
Figure 1b. Computed tomography angiography of hematoma in liver segments VII and VIII, demarcated by green lines. A complication from attempted portal vein access.

Discussion & Conclusions

- Though JAK2 mutation is associated with myeloproliferative disorders, it is an independent risk factor for the development of PVT.
- With worsening thrombus chronicity and burden, CTPV may occur thereby decreasing the chance of successful TIPS.
- When organ ischemia develops due to extensive portomesenteric venous thrombosis, orthotopic liver transplantation is no longer an option.
- **Multivisceral transplantation** replaces the liver, small bowel, and other abdominal organs.
- Replacement of the thrombosed portomesenteric system may be the only recourse to reverse portal hypertension and address the primary disease.

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