JAK2 Positive Budd-Chiari Syndrome and Risk in Liver Transplant

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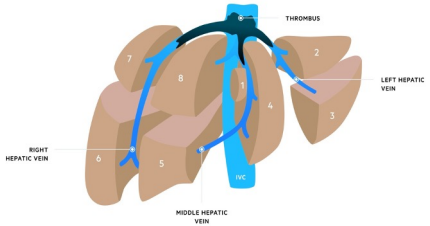
BACKGROUND

- Budd-Chiari Syndrome (BCS) describes the narrowing and occlusion of the hepatic veins in the liver.
- Primary myeloproliferative diseases leading to high clot burden are the leading cause of BCS.[1]
- Patients that have underlying myeloproliferative neoplasms (MPN) are often transplanted and long-term prognosis is often considered.
- This case highlights the rapid decline of a complex patient with BCS secondary to myeloproliferative disease and the current data available to guide decision making in the transplant evaluation process.

Figure 1: Occlusion of hepatic veins[2]



CASE

- 63-year-old male presented with 3 days of abdominal pain/distention and diarrhea found to have new onset cirrhosis complicated by ascites and hepatic encephalopathy.
- He denied any new medications, personal or family history of liver or autoimmune diseases, or history of alcohol use.
- Initial laboratory evaluation included MELD-Na 15, undetectable acetaminophen, negative viral hepatitis serologies, paracentesis was negative for SBP with SAAG of 1.9.
- CT showed cirrhotic morphology, splenomegaly, and ascites. Ultrasound with doppler and CT venogram showed findings consistent with BCS.
- He was started on therapeutic heparin. Patient decompensated further requiring pressor support along with worsening MELD-Na and renal failure ultimately requiring renal replacement therapy.
- He was found to have positive JAK2 V617F mutation. With leukocytosis and JAK2 mutation, bone marrow biopsy was obtained showing myeloproliferative neoplasm (MPN) consistent with myelofibrosis (MF).
- He was listed for transplant as MF was not deemed an absolute contraindication. Unfortunately, he decompensated further with variceal bleed and passed prior to transplant.

Temp 97.2, RR 16, HR 99, BP 94/72

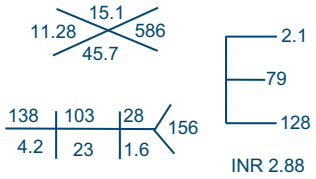
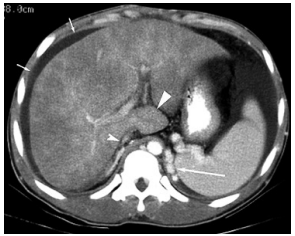


Photo 1: CT with mottled appearance of an underperfused liver, collapsed portal veins and a small, collapsed IVC[3]



Model for End Stage Liver Disease (MELD)

MELD score= 10x[0.957x log e (creatinine) + log e (bilirubin) + 1.12 x log e (INR)] + 6.43

3 month mortality according to MELD score

MELD score	<=9	10-19	20-29	30-39	>=40
Hospitalized pt.	4%	27%	76%	83%	100%
Outpatient cirrhotic	2%	6%	50%		

DISCUSSION

- Listing the patient for transplant was controversial due to increased risk of leukemic transformation in with immunosuppression as well as increased thrombotic risk in the peri/post-operative setting.
- Hydroxyurea was initiated to mitigate these risks and bridge to transplant in this otherwise healthy, young, and high functioning patient.
- Prior studies identified portal vein thrombosis as an independent risk factor for graft loss in transplant recipients, independent of other factors.[4]
- One study evaluated the prognosis of liver transplant with BCS and MPN as well as effect of immunosuppression- there was no difference in survival in BCS patients with or without MPN. Progression of MPN was not noted after transplant.[5]

REFERENCES

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