



EHPVO SECONDARY TO MYELOPROLIFERATIVE NEOPLASMS: A CASE SERIES

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Introduction

Extra-hepatic portal vein obstruction (EHPVO) is a vascular disease characterized by thrombotic obstruction and cavernomatous transformation of portal vein. In India, EHPVO is responsible for one-third cases of portal hypertension in adults and 50–60% of cases in children[1]. Among children, the major causes of EHPVO are sepsis, umbilical catheterization, and developmental anomalies. However, in adults, cirrhosis, surgery, pancreatitis, and malignancies are important local causes of obstruction[2,4]. In a study on 58 patients with intra-abdominal venous thrombosis, Deepak *et al* reported 14% positivity for JAK2V617F allele with Portal vein thrombosis[5]. Another study by Priyanka Mishra *et al* showed a JAK2 positivity of 10.6% (13/122) in EHPVO[3]. Myeloproliferative neoplasms (MPN) are a known but under-recognized cause of thrombosis leading to EHPVO. JAK2 mutations play a pivotal role in the pathogenesis by promoting hypercoagulability. This case series highlights eight cases of EHPVO secondary to MPNs.

Methodology

We analyzed patients admitted with EHPVO from Jan 2023, till Dec 2024. Total of 44 patients were diagnosed as EHPVO. All patients underwent detailed clinical examination,

laboratory investigations including complete hemogram, liver function tests, Renal function tests, Homocysteine levels, PT, aPTT, thrombophilia profile (Protein C, Protein S, antithrombin III, factor V Leiden mutation), APLA Panel, JAK2 mutation testing. Endoscopic evaluation was performed to assess the presence and grading of esophageal varices. Bone marrow biopsy was done to establish the type of underlying MPN.

Results

This case series included eight patients diagnosed with extrahepatic portal vein obstruction (EHPVO) secondary to underlying myeloproliferative neoplasms (MPNs). The mean age at presentation was 56.5 years (range: 45–67 years). There was a male predominance, with six males (75%) and two females (25%).

Among the 44 patients evaluated with EHPVO, eight patients (18.18%) were found to be JAK2 V617F mutation-positive. All eight JAK2-positive patients presented with upper gastrointestinal bleeding (100%) at initial evaluation. Six patients (75%) presented with hematemesis, while two patients (25%) presented with melena. Bone marrow examination revealed myelofibrosis in four patients (50%), polycythemia vera in two patients (25%), and essential thrombocytosis in two patients (25%).

Clinical Profile

Splenomegaly was a universal finding, present in all eight patients (100%), with massive splenomegaly noted in three patients (37.5%). Pallor was observed in five patients (62.5%). None of the patients had jaundice or ascites, suggesting preserved hepatic synthetic function. Upper

gastrointestinal endoscopy demonstrated large esophageal varices in all patients (100%). Imaging studies, including ultrasonography and contrast-enhanced computed tomography (CECT) of the abdomen, confirmed chronic portal vein thrombosis with cavernomatous transformation of the portal vein in all cases.

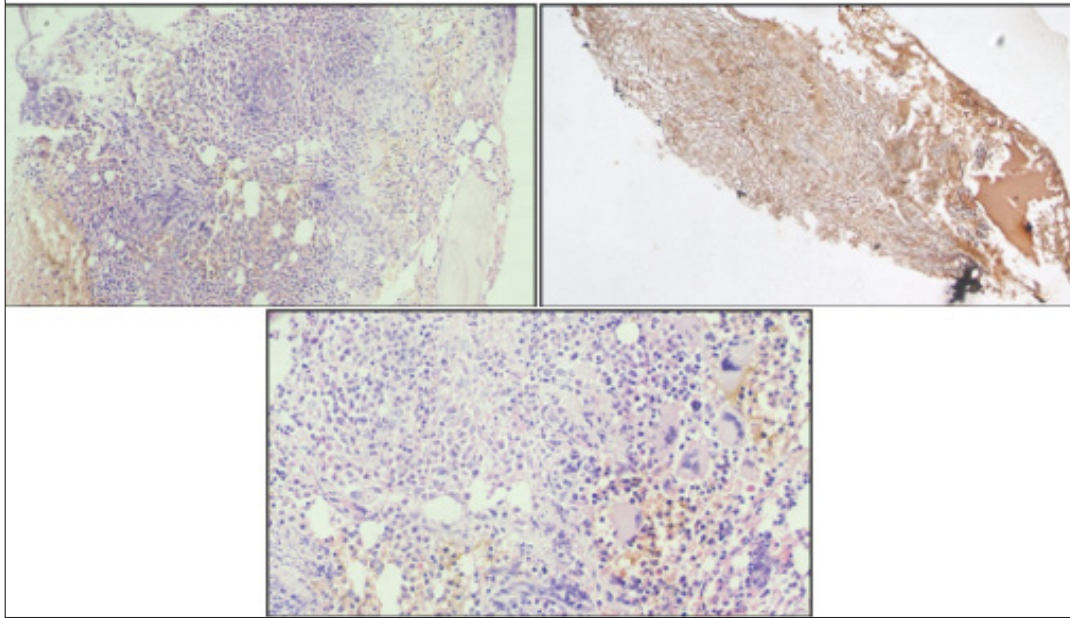


Figure 1. Bone marrow aspiration and biopsy findings

Table 1: Clinical Features of JAK2-Positive EHPVO Patients (n = 8)

Case	Symptoms	Pallor	Splenomegaly	Ascites	Jaundice
1.	Hematemesis	Yes	Yes	No	No
2.	Hematemesis	Yes	Yes	No	No
3.	Melena	Yes	Massive	No	No
4.	Hematemesis	No	Massive	No	No
5.	Hematemesis	Yes	Yes	No	No
6.	Melena	Yes	Massive	No	No
7.	Hematemesis	No	Yes	No	No
8.	Hematemesis	No	Yes	No	No

Table 2: Clinical Features (n = 8)

Feature	Number of Patients	Percentage
Upper GI bleed	8	(100%)
Hematemesis	6	(75%)
Melena	2	(25%)
Splenomegaly	8	(100%)
Massive splenomegaly	3	(37.5%)
Ascites	0	
Jaundice	0	

Table 3: Laboratory and Bone Marrow Findings

Case	Hb (g/dL)	WBC (cells/mm ³)	Platelets (×10 ⁵ /mm ³)	JAK2 Mutation	Bone Marrow Diagnosis
1.	9.4	18,460	4.18	Positive	Myelofibrosis
2.	6.2	3,300	1.2	Positive	Myelofibrosis
3.	9.4	6,020	5.1	Positive	Essential thrombocythemia
4.	9.7	14,000	5.4	Positive	Myelofibrosis
5.	7.2	2,340	1.1	Positive	Myelofibrosis
6.	12	7,000	4.9	Positive	Essential thrombocythemia
7.	16.8	6,030	6	Positive	Polycythemia vera
8.	16.0	5,450	5.0	Positive	Polycythemia vera

Table 4: Hematological and Bone Marrow Findings

Parameter	Findings	Percentages
Anemia	5 patients	62.5%
Leukocytosis	2 patients	25%
Leukopenia	2 Patients	25%
Thrombocytosis	5 patients	62.5%
Thrombocytopenia	2 patients	25%
Pancytopenia	2 patients	25%
JAK2 mutation	8/44 patients	18.8%
Myelofibrosis	4/8 Patients	50%
Essential thrombocythemia	2/8 patients	25%
Polycythemia vera	2/8 patients	25%

Discussion

EHPVO is an important cause of portal hypertension. MPNs like myelofibrosis, essential thrombocytosis, and polycythemia vera contribute to a hypercoagulable state leading to chronic portal vein thrombosis and subsequent cavernoma formation. The JAK2 V617F mutation plays a critical pathogenic role in promoting thrombosis in MPNs. In the present study, 8 of 44 patients with EHPVO (18.18%) were found to be JAK2 V617F–positive, indicating a substantial contribution of underlying myeloproliferative neoplasms (MPNs) to the pathogenesis of EHPVO. This prevalence is higher than that reported by Mishra et al., who observed JAK2 positivity in 12 of 64 patients (10.6%), and is comparable to findings from Deepak et al., who reported a prevalence of approximately 14% (3,5). Our results therefore fall within the range reported in Indian studies and support the growing recognition of occult MPNs in adult EHPVO. Previous literature has shown that JAK2 V617F mutation was detected in 17–35% of patients with portal vein thrombosis, particularly in those without cirrhosis or malignancy, highlighting the strong association between this mutation and splanchnic venous thrombosis[6,7].

Jain et al. reported a male predominance and splenomegaly in all patients with EHPVO, findings similar to our study, in which 75% of patients were male and splenomegaly was

present in 100% of cases [4]. Upper gastrointestinal bleeding was seen in all patients (100%) in our series, whereas Jain et al. reported bleeding in 22 of 30 patients (73.3%), indicating a lower frequency of bleeding at presentation in their cohort[4]. In our series, anemia was seen in 62.5% of patients, splenomegaly in all patients (100%), and thrombocytosis in 62.5%, findings that are similar to the hematological patterns reported by Mishra et al.

In our case series, 8 of 44 patients (18.18%) with EHPVO were diagnosed with bone marrow–proven myeloproliferative neoplasms (MPNs). Myelofibrosis was the predominant subtype (50%), followed by polycythemia vera (25%) and essential thrombocytosis (25%). In contrast, Mishra et al. reported polycythemia vera in 4 out of 12(33.3%)[3]. These findings further highlight the clinical importance of systematically evaluating adult EHPVO patients for the pharmacy in charge’s underlying hematological disorders. Early identification of MPNs allows initiation of disease-specific therapy, including cytoreductive treatment and anticoagulation, which may help prevent recurrent thrombosis and modify the natural course of portal hypertension in this subset of patients.

Conclusion

Myeloproliferative neoplasms should be considered as a potential underlying etiology in patients with EHPVO. Hence

all patients with EHPVO should undergo comprehensive evaluation for underlying hematological abnormalities including tests for myeloproliferative neoplasms in the given clinical scenario.

Conflict of interest

None declared

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