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ORIGINAL ARTICLE

Predictors of myeloproliferative neoplasm in non-cirrhotic portal vein thrombosis: a comparative analysis with hereditary thrombophilia-related cases

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ABSTRACT

Objective: Non-cirrhotic portal vein thrombosis (PVT) is rare in patients without cirrhosis or intra-abdominal malignancy, commonly associated with thrombophilia or myeloproliferative neoplasms (MPNs). Comparative studies on clinical and laboratory features of MPN- versus thrombophilia-related PVT are limited. This study aimed to examine etiological differences and identify parameters that may aid differential diagnosis.

Materials and Methods: In this retrospective cross-sectional study, 73 adult patients with non-cirrhotic PVT due to MPNs or hereditary thrombophilia were included. Clinical, laboratory, imaging, and endoscopic data were collected from records. Continuous variables were analyzed using t-test or Mann-Whitney U test, and categorical variables using chi-square or Fisher's exact test. Multivariable logistic regression and ROC analysis identified predictors of MPN-associated PVT.

Results: Platelet counts were significantly higher in the MPN group than in the thrombophilia group ($p<0.001$). Hepatomegaly and portal double ductopathy (PDD) were more frequent in MPN, with the difference for PDD being significant ($p=0.032$). Splenic/superior mesenteric vein involvement occurred in 41.7% versus 26.5%, and portal vein cavernous transformation (PVCT) in 79.2% versus 57.1%; these differences were not statistically significant. In multivariable analysis, platelet count was the only independent predictor of MPN ($p=0.003$). ROC analysis showed an AUC of 0.79, with a cutoff $\geq 161\times10^3/\mu L$ yielding 85% sensitivity and 61% specificity.

Conclusion: Platelet count is a strong, independent marker for distinguishing MPN-related non-cirrhotic PVT. Although PDD and PVCT are more frequent in MPN, platelet level offers a rapid, practical parameter for differential diagnosis. These findings provide valuable guidance for clinical practice and patient selection for advanced genetic testing.

Keywords: non-cirrhotic portal vein thrombosis, myeloproliferative neoplasms, hereditary thrombophilia, platelet count, portal vein cavernous transformation

INTRODUCTION

Non-cirrhotic PVT is a rare clinical condition characterized by thrombosis of the portal vein, and sometimes the mesenteric veins, in the absence of cirrhosis or intra-abdominal malignancy [1]. In the general population, the lifetime risk of PVT from all causes is approximately 1% [2]. Although PVT is common in patients with cirrhosis (5-24%; 6-64% in autopsy studies), its prevalence in individuals without chronic liver disease is not well established [3].

The major recognized risk factors for the disease include inherited or acquired prothrombotic conditions, hepatobiliary malignancies, intra-abdominal infections or inflammatory processes, and MPNs [3]. According to previous studies, an underlying etiology can be identified in nearly 70% of non-cirrhotic PVT cases [4]. In our previous study, an underlying etiology was identified in 78.8% of cases [5]. In European series, the most common risk factors are thrombophilia (42-50%) and MPNs, with the JAK2 V617F mutation particularly prevalent among the latter [4]. Thrombophilic conditions that can lead to PVT include paroxysmal nocturnal hemoglobinuria, antiphospholipid syndrome, hyperhomocysteinemia, deficiencies of protein C, protein S, or antithrombin III, mutations in Factor V Leiden or Factor II, methylenetetrahydrofolate reductase (MTHFR) gene mutation, among other causes [6]. MPNs are identified as the underlying cause in approximately 20-30% of patients with non-cirrhotic, non-malignant PVT [7]. Consistent with this, in our internal medicine specialization thesis, this proportion was found to be 20.2% [5].

The diagnostic approach to non-cirrhotic PVT is based on thrombophilia assessment and appropriate imaging studies. The American Association for the Study of Liver Diseases (AASLD) and the Baveno VII consensus recommend testing for inherited and acquired thrombophilias, while low-risk genetic tests are not routinely advised. The exclusion of cirrhosis is a crucial step in the diagnostic process and may be supported by contrast-enhanced computed tomography (CT), magnetic resonance imaging (MRI), liver stiffness assessment, and, when indicated, liver biopsy. Ultrasound, CT, and MRI are the main modalities for diagnosing PVT, with contrast-enhanced studies providing information on thrombus extent, complications, and differentiation from neoplastic thrombi [8]. Diagnosis of MPN in non-cirrhotic PVT can be challenging, as peripheral blood counts may be normal or low in the presence of portal hypertension and hypersplenism. Therefore, testing for the JAK2 V617F mutation, found in ~95% of patients with polycythemia vera (PV) and ~50% of those with essential thrombocythemia (ET), is recommended. Additionally, CALR mutation analysis should be considered in cases with platelet counts $>200\times10^3/\mu L$ and spleen size ≥16 cm, and bone marrow biopsy may be warranted if necessary [1].

Although most studies have examined the association between MPN and PVT, analyses comparing the clinical and laboratory features of MPN-related versus hereditary thrombophilia-related PVT are limited. Moreover, advanced diagnostic tests, including genetic testing, JAK2 mutation analysis, and bone marrow examination, are available only at select centers, and the time required to obtain results means that the availability of rapid and reliable differentiating parameters remains uncertain. The differential diagnosis between MPN-related and thrombophilia-related PVT is important for short-term management, including treatment strategies and thrombotic complication control, and provides guidance for long-term considerations such as disease prognosis, follow-up planning, genetic counseling, and lifelong thrombotic risk management. This highlights the need for further research to identify rapid and reliable differentiating parameters. The aim of this study was to compare the clinical and laboratory features of MPN- and thrombophilia-related etiologies in patients with non-cirrhotic PVT and to identify parameters that may aid in differential diagnosis.

MATERIALS AND METHODS

Study Design: This retrospective, analytical cross-sectional study included 74 patients with non-cirrhotic PVT, whose etiology was attributed to MPN or inherited thrombophilia, selected from 119 patients in the archives of Hacettepe University Faculty of Medicine between 1970 and 2011. The study reporting followed the STROBE (Strengthening the Reporting of Observational Studies in Epidemiology) checklist.

Patient Population: Patients were classified into two etiological groups: MPN (n=25) and those with at least one inherited thrombophilia (n=50). One patient with concomitant MPN and inherited thrombophilia was excluded. The MPN group included 24 patients and the thrombophilia group 49 patients.

Data Collection Process: Demographic data, presenting symptoms, physical examination findings, laboratory results, abdominal ultrasonography (US), endoscopic evaluations, and endoscopic retrograde cholangiopancreatography (ERCP) findings were retrospectively collected from patient records.

Variables and Definitions: The independent variables included both continuous and categorical parameters. Continuous variables included age, age at diagnosis, liver enzyme tests [alanine aminotransferase (ALT), aspartate aminotransferase (AST), gamma-glutamyl transferase (GGT), alkaline phosphatase (ALP)], total bilirubin, albumin, international normalized ratio (INR), hemoglobin, and platelet count. Since platelet levels may be influenced in patients who underwent splenectomy, their values were excluded from analysis.

Categorical variables included demographic and clinical variables [sex; abdominal pain, distension, fatigue, melena, hematemesis, hematochesia, nausea, vomiting; physical examination: pallor, splenomegaly, hepatomegaly, jaundice, venous abdominal collaterals, ascites, splenectomy scar], imaging and interventional findings [splenic and superior mesenteric vein involvement, PVCT, mild chronic hepatic changes (ultrasound-assessed)], endoscopic and ERCP findings [PDD, sclerosing cholangitis (SC), normal or abnormal endoscopic findings], comorbid conditions (diabetes, cardiovascular diseases, thyroid disorders, rheumatologic diseases, additional hematologic abnormalities), and other clinical conditions (cholelithiasis).

The dependent variable was the etiology of non-cirrhotic PVT, into two categories: MPN and thrombophilia.

Ethical Approval: Ethical approval for the internal medicine specialization thesis titled "Etiologic Distribution of Chronic Noncirrhotic Portal Vein Thrombosis in Adult Patients" (completed August 1, 2011; YÖKSİS Thesis No: 282140) was obtained from the Ethics Committee of Hacettepe University Faculty of Medicine before the study commenced. This study used only archival data from the thesis, involved no new interventions, addressed entirely new research questions, and no duplicate publication (salami slicing) occurred.

Statistical Analysis: All analyses were performed using IBM SPSS Statistics version 27. The distribution of continuous variables was assessed by the Shapiro-Wilk test. Means and 95% confidence intervals (CI) were calculated. Comparisons between groups were conducted using Student's t-test for normally distributed variables and the Mann-Whitney U test for non-normally distributed variables.

For categorical variables, frequencies and percentages were calculated, and differences were analyzed using the Pearson chi-square test or Fisher's exact test (2-sided) when the expected cell frequency was less than 5. To identify predictors of PVT etiology, multivariate logistic regression (MLR) analysis was performed. Platelet count, PVCT, and hepatomegaly were included in the model. Results were reported as B coefficients, standard errors (S.E.), Wald statistics, odds ratios (OR), and 95% CI.

The overall model significance was assessed using the Omnibus Tests of Model Coefficients, and model fit was evaluated with the Hosmer-Lemeshow test. Statistical significance was set at $p < 0.05$.

Power Analysis: Due to the retrospective design and the use of a fixed archival dataset, no prior power analysis was performed; all eligible cases were included in the study. The precision of the findings was assessed using 95% CI.

RESULTS

The mean age in the MPN group was 44.5 years, while it was 46.9 years in the thrombophilia group; no statistically significant difference was observed between the groups (p=0.630). The mean age at diagnosis was 38.6 years (95% CI: 33.7-43.4) in the MPN group and 37.1 years (95% CI: 32.2-42.0) in the thrombophilia group, with no significant difference between the groups (p=0.526) (Table 1).

Table 1. Continuous variables of patients with myeloproliferative neoplasm– and thrombophilia-related portal vein thrombosis

Variables	Etiology of PVT	n	Mean [95% CI]	p-value
Age (years)	MPN	24	44.5 (38.8-50.3)	0.630
	Thrombophilia	49	46.9 (42.5-51.2)	
Age at diagnosis (years)	MPN	24	38.6 (33.7-43.4)	0.526
	Thrombophilia	49	37.1 (32.2-42.0)	
INR	MPN	24	1.30 (1.24-1.35)	0.262
	Thrombophilia	49	1.30 (1.23-1.36)	
ALT (U/L)	MPN	24	34.1 (23.8-44.5)	0.599
	Thrombophilia	48	33.6 (25.1-42.1)	
AST (U/L)	MPN	24	37.6 (29.0-46.3)	0.473
	Thrombophilia	48	38.0 (29.7-46.4)	
GGT (U/L)	MPN	23	95.0 (48.4-141.6)	0.125
	Thrombophilia	48	57.6 (33.5-81.8)	
ALP (U/L)	MPN	23	183.0 (139.0-226.9)	0.672
	Thrombophilia	48	205.7 (149.8-261.7)	
Total bilirubin (mg/dL)	MPN	23	1.25 (0.82-1.67)	0.606
	Thrombophilia	48	1.60 (1.16-2.03)	
Albumin (g/L)	MPN	23	3.9 (3.6-4.1)	0.900
	Thrombophilia	47	3.9 (3.7-4.0)	
Hemoglobin (g/dL)	MPN	24	12.7 (11.3-14.1)	0.171
	Thrombophilia	49	11.7 (11.0-12.5)	
*Platelet (10^3/μL)	MPN	20	430 (273-587)	<0.001
	Thrombophilia	41	172 (126-218)	

PVT: Portal Vein Thrombosis

n: Number of patients

CI: Confidence Interval

MPN: Myeloproliferative Neoplasm

*Platelet counts of patients with prior splenectomy were excluded from the analysis.

INR values were similar between patients with MPN- and thrombophilia-associated PVT (1.30 in both groups; p=0.262). No significant differences were observed in liver function tests; ALT, AST, and total bilirubin levels were comparable between groups. Although not statistically significant, GGT levels were higher in the MPN group [95.0 U/L (48.4-141.6) vs. 57.6 U/L (33.5-81.8); p=0.125], whereas ALP levels were slightly higher in the thrombophilia group [205.7 U/L (149.8-261.7) vs. 183.0 U/L (139.0-226.9); p=0.672]. Platelet counts were significantly higher in the MPN group [430 (95% CI: 273-587) vs. 172 (126-218); p<0.001]. Hemoglobin levels were slightly higher in the MPN group but did not reach statistical significance, while albumin levels were comparable between the two groups (p>0.05) (Table 1).

In the MPN group, 41.7% of patients were female and 58.3% were male, whereas in the thrombophilia group the corresponding proportions were 49.0% and 51.0%. There was no significant difference in sex distribution between the groups (p=0.556). In the MPN group, 79.2% of patients were symptomatic, compared to 85.7% in the thrombophilia group, with no significant difference observed (p=0.478) (Table 2).

Physical examination was present in 95.8% of MPN patients and 87.8% of the thrombophilia group, with no statistically significant difference (p=0.271). Splenomegaly was observed in 83.3% of MPN patients and 75.5% of the thrombophilia group (p=0.448). Hepatomegaly was detected in 29.2% of the MPN group and 12.2% of the thrombophilia group; although not statistically significant, a trend was noted (χ^2=3.152, p=0.076) (Table 2).

Based on ERCP findings, PDD was observed in 55.6% of patients in the MPN group and 14.3% in the thrombophilia group, with a significant difference

Table 2. Distribution of categorical variables in patients with MPN and thrombophilia

Variable	Category	MPN (n)	Thrombophilia (n)	Test statistic	p-value
Gender	Female	10 (41.7%)	24 (49.0%)	$\chi^2 = 0.346$	0.556
	Male	14 (58.3%)	25 (51.0%)		
*Complaint	Present	19 (79.2%)	42 (85.7%)	$\chi^2 = 0.503$	0.478
	Absent	5 (20.8%)	7 (14.3%)		
** Physical findings	Present	23 (95.8%)	43 (87.8%)	$\chi^2 = 1.213$	0.271 (‡0.414)
	Absent	1 (4.2%)	6 (12.2%)		
Splenomegaly	Present	20 (83.3%)	37 (75.5%)	$\chi^2 = 0.576$	0.448 (‡0.555)
	Absent	4 (16.7%)	12 (24.5%)		
Hepatomegaly	Present	7 (%29.2)	6 (12.2%)	$\chi^2 = 3.152$	0.076
	Absent	17 (70.8%)	43 (87.8%)		
PDD (ERCP)	Present	5 (55.6%)	3 (14.3%)	$\chi^2 = 5.487$	0.019 (‡0.032)
	Absent	4 (44.4%)	18 (85.7%)		
SC (ERCP)	Present	3 (33.3%)	12 (57.1%)	$\chi^2 = 1.429$	0.232 (‡0.427)
	Absent	6 (66.7%)	9 (42.9%)		
Splenic vein involvement	Present	10 (41.7%)	13 (26.5%)	$\chi^2 = 1.710$	0.191
	Absent	14 (58.3%)	36 (73.5%)		
SMV involvement	Present	10 (41.7%)	13 (26.5%)	$\chi^2 = 1.710$	0.191
	Absent	14 (58.3%)	36 (73.5%)		
ˣMild chronic hepatic changes	Present	9 (37.5%)	25 (52.1%)	$\chi^2 = 1.365$	0.243
	Absent	15 (62.5%)	23 (47.9%)		
ˣˣEndoscopic findings	Normal	6 (30.0%)	10 (25.0%)	$\chi^2 = 0.170$	0.680
	Abnormal	14 (70.0%)	30 (75.0%)		
ʰAdditional comorbidities	Present	5 (20.8%)	16 (32.7%)	$\chi^2 = 1.098$	0.295
	Absent	19 (79.2%)	33 (67.3%)		
Cholelithiasis	Present	5 (20.8%)	10 (21.3%)	$\chi^2 = 0.002$	0.965
	Absent	19 (79.2%)	37 (78.7%)		
PVCT	Present	19 (79.2%)	28 (57.1%)	$\chi^2 = 3.408$	0.065
	Absent	5 (20.8%)	21 (42.9%)		

*Abdominal pain, abdominal distension, fatigue/weakness, melena, hematemesis, hematochezia, nausea and vomiting, etc.

**Pallor, splenomegaly, hepatomegaly, jaundice, venous abdominal collateral, ascites, splenectomy scar, etc.

***Hepatic vein, inferior vena cava, femoral vein, jugular vein, popliteal vein, etc.

ˣThe evaluation was conducted using liver ultrasound.

ˣˣEsophageal varices, gastric varices, portal hypertensive gastropathy

ʰDiabetes mellitus, cardiovascular disease, thyroid disorders, rheumatologic diseases, additional hematologic abnormalities, etc.

PDD: Portal double ductopathy

ERCP: Endoscopic retrograde cholangiopancreatography

SC: Sclerosing cholangitis

SMV: Superior mesenteric vein

PVCT: Portal vein cavernous transformation

‡Fisher's Exact (2-sided)

between groups (Fisher's Exact test, p=0.032). SC was detected in 33.3% of the MPN group and 57.1% of the thrombophilia group; this difference was not statistically significant (p=0.232) (Table 2).

In addition to portal vein involvement, splenic vein and superior mesenteric vein (SMV) involvement was observed in 41.7% of the MPN group and 26.5% of the thrombophilia group; the difference was not statistically significant (p=0.191), although a higher frequency in MPN patients was noted (Table 2).

On liver ultrasound evaluation, mild chronic changes were detected in 37.5% of the MPN group and 52.1% of the thrombophilia group; the difference was not statistically significant (p=0.243).

The prevalence of cholelithiasis was approximately 21% in both groups, with no significant difference observed (p=0.965) (Table 2).

Abnormal findings on endoscopic evaluation were observed in 70% of the MPN group and 75% of the thrombophilia group; the difference was not statistically significant (p=0.680). The prevalence of PVCT was 79.2% in the MPN group and 57.1% in the thrombophilia group; the difference approached statistical significance (χ^2=3.408, p=0.065) and was higher in the MPN group (Table 2).

Comorbidities, including diabetes mellitus, cardiovascular, thyroid, rheumatologic, and additional hematologic disorders, were observed in 20.8% of the MPN group and 32.7% of the thrombophilia group; the difference was not statistically significant (p=0.295) (Table 2).

In MLR analysis, the relationships between platelet count, PVCT, hepatomegaly, and the etiology of PVT (MPN vs. thrombophilia) were assessed. Platelet count strongly predicted MPN etiology; higher platelet values were significantly associated with increased likelihood of MPN (OR: 0.99 [95% CI: 0.99-1.00], p=0.003). The presence of PVCT showed a trend toward increased MPN probability (OR: 2.75 [95% CI: 0.59-12.84], p=0.198), whereas hepatomegaly was not a significant predictor (OR: 0.75 [95% CI: 0.12-4.91], p=0.765). The Hosmer-Lemeshow test confirmed good model fit, and the Omnibus Test indicated overall model significance (Table 3).

ROC analysis showed that platelet count predicted MPN-associated PVT with an AUC of 0.79 (95% CI: 0.66-0.91, p<0.001), indicating good discriminative ability for distinguishing MPN etiology. Using the maximum Youden index, the optimal cutoff was PLT ≥161×10^3/μL, yielding 85% sensitivity and 61% specificity (Figure 1).

DISCUSSION

In this study, we compared the clinical and laboratory characteristics of patients with non-cirrhotic PVT associated with MPNs or thrombophilia. Our aim was to identify distinguishing features that could aid in the etiological differentiation of PVT.

No significant difference in current age or age at diagnosis was observed between patients with MPN- and thrombophilia-related PVT. Although literature suggests MPN tends to cause PVT at older ages and thrombophilia in younger patients, the similarity in our cohort implies MPN may also

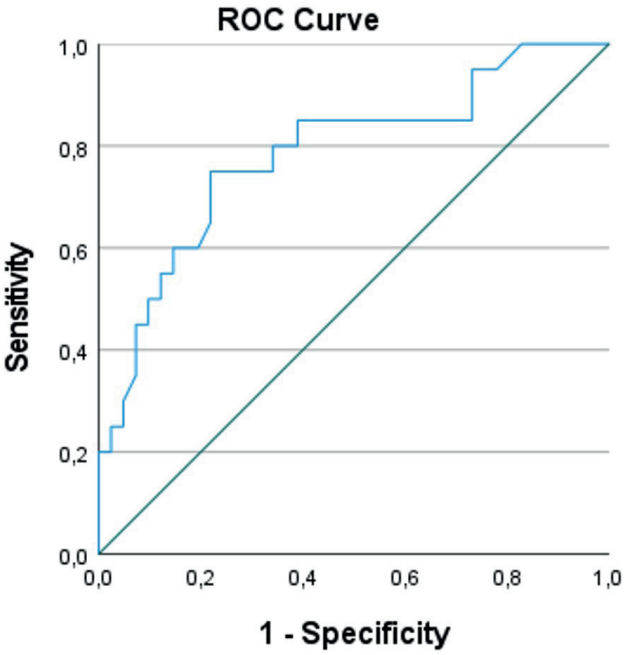

Figure 1. ROC curve for platelet count predicting MPN-associated PVT

Table 3. Multivariate logistic regression analysis for predictors of portal vein thrombosis etiology

Variable	B	S.E.	Wald	OR (95% CI)	p-value
Platelet	-0.006	0.002	8.654	0.99 (0.99-1.00)	0.003
PVCT Reference category: Absent	1.012	0.786	1.657	2.75 (0.59-12.84)	0.198
Hepatomegaly Reference category: Absent	-0.286	0.958	0.089	0.75 (0.12-4.91)	0.765

OR: Odds ratio

CI: Confidence interval

PVCT: Portal vein cavernous transformation

Omnibus Tests of Model Coefficients: χ^2=17.376, df=3, p=0.001

Hosmer-Lemeshow Test: χ^2=12.68, df=8, p=0.123

present earlier. In a study by How et al., 134 patients with MPN were compared to 52 with splanchnic vein thrombosis (SVT)-MPN, showing a significantly lower median age in the SVT-MPN group (47 vs. 56.5 years; p=0.003) [9]. Similarly, Hoekstra et al. reported a median age of 48 years in 44 patients with MPN-related PVT [7]. Overall, these results suggest neither age at diagnosis nor current age distinguishes the two groups.

In the literature, gender distribution in the general PVT population is variable. One study reported 59% of patients with PVT were male and 41% female, with this distribution remaining stable over time [10]. Another study of MPN-associated SVT patients reported a female predominance (65.2%) [11]. In our study, gender distribution was similar in both groups: MPN, 41.7% female and 58.3% male; thrombophilia, 49.0% female and 51.0% male (p=0.556). These findings suggest that gender does not significantly affect PVT etiology or clinical presentation. No studies have directly compared MPN- and thrombophilia-associated PVT regarding gender.

In non-cirrhotic PVT, the most common presenting symptoms are abdominal pain, distension, gastrointestinal bleeding, fever, and ascites. Disease severity depends on etiology and thrombosis extent [12]. In our study, most patients in both groups were symptomatic (MPN 79.2%; thrombophilia 85.7%; p=0.478), and physical examination findings were frequently observed (MPN 95.8%; thrombophilia 87.8%; p=0.271), indicating that non-cirrhotic PVT is generally symptomatic regardless of etiology.

Splenomegaly is common in PVT, typically presenting as left upper quadrant discomfort, pain from splenic infarction, or, occasionally, symptomatic hypersplenism [13]. In our study, splenomegaly was slightly more frequent in the MPN group (MPN 83.3%; thrombophilia 75.5%; p=0.448), but the difference was not statistically significant. These findings indicate that splenomegaly is common in both groups and is not a reliable standalone criterion for etiological differentiation.

Hepatomegaly was more frequent in the MPN group than in the thrombophilia group (29.2% vs. 12.2%; p=0.076), but the difference did not reach statistical significance, likely due to the limited sample size. A previous study reported a hepatomegaly rate of 18% in non-cirrhotic PVT patients with MPN [7]. This may be explained by the increased risk of hepatomegaly in MPN due to portal hypertension and extramedullary hematopoiesis, making the observed trend pathophysiologically plausible. In MLR analysis, hepatomegaly did not significantly predict MPN (OR 0.75, 95% CI 0.12–4.91; p=0.765), indicating it is not a reliable standalone marker for etiological differentiation. Larger prospective studies are needed to clarify hepatomegaly distribution in MPN- and thrombophilia-associated PVT.

In patients with chronic PVT without underlying liver disease, liver function tests are generally normal [14,15]. Marked elevation of cholestatic enzymes may indicate portal cholangiopathy [14]. In our study, ALT and AST levels were similar in both groups, supporting largely preserved liver function in non-cirrhotic PVT. GGT levels were higher in the MPN group, although not statistically significant, which may reflect intrahepatic circulatory disturbances, microthrombosis, or concomitant cholestatic processes. Conversely, higher ALP and total bilirubin levels in the thrombophilia group may result from long-standing PVT affecting the biliary system. Clinically, these findings are relevant for potential complications of portal hypertension. Although not significant, the results suggest a trend toward elevated GGT in MPN and higher ALP and bilirubin in thrombophilia. Larger studies are needed to clarify the clinical relevance of these subtle differences.

Hemoglobin levels were similar between groups, with a mean value of 12.7 g/dL in the MPN group and 11.7 g/dL in the thrombophilia group (p=0.171). This finding indicates that hemoglobin alone has limited value in the etiological differentiation of PVT. Previous studies have reported that hemoglobin levels may appear normal (masked) in patients with MPN-associated (particularly PV) SVT [16]. Our findings are consistent with the characteristic hematological features of MPNs reported in the literature and may contribute to the evaluation of PVT etiology in clinical practice. Larger prospective studies are needed to validate these results.

In our study, platelet counts were markedly elevated in patients with MPN-related PVT, consistent with the characteristic hematological profile of this group. Platelet count also emerged as an independent predictor of etiology, suggesting

its potential utility in differential diagnosis. ROC analysis demonstrated good diagnostic performance in distinguishing MPN-related cases from thrombophilia-associated PVT, with high sensitivity (85%). However, limited specificity indicates that platelet count alone may not suffice as a diagnostic marker and should be interpreted alongside other clinical and laboratory findings. Similarly, ROC analyses in patients with SVT have shown that platelet counts above 190,000/mm^3 are significantly associated with an increased likelihood of JAK2 V617F mutation and MPN. The retrospective, single-center design represents a limitation regarding moderate specificity. Future studies combining hematological parameters with genetic testing are recommended to develop more robust scoring systems for distinguishing MPN-related PVT [17].

Clinical manifestations in patients with PVT include obstructive jaundice, cholangitis, and, in the later stages, choledocholithiasis secondary to pseudosclerosing cholangitis or portal hypertensive biliopathy [13]. Portal cholangiopathy is a complication that may develop in patients with chronic PVT or portal cavernoma. In such cases, porto-portal collaterals can compress the bile ducts, leading to ischemic injury, chronic inflammation, and fibrosis [8]. In our study, PDD was observed more frequently in patients with MPN than in those with thrombophilia, suggesting that it may serve as a potential clinical marker for etiological differentiation. However, the limited number of patients and restricted availability of ERCP in all centers should be considered when interpreting the generalizability of these findings. Portal cavernoma cholangiopathy refers to abnormalities of the bile ducts that develop in patients with chronic non-cirrhotic PVT due to external compression by a portal cavernoma or altered portal blood flow [1]. Supporting this observation, PVCT was more common in patients with MPN compared to those with thrombophilia (79.2% vs. 57.1%). Interestingly, although not statistically significant, sclerosing cholangitis (SC) findings on ERCP were more frequently observed in thrombophilia patients than in MPN patients (57.1% vs. 33.3%), in contrast to PDD. This indicates that SC can also occur in non-cirrhotic PVT etiologies and is not a reliable standalone marker for etiological differentiation. The small sample size may have limited the power to detect significant differences, highlighting the need for studies with larger cohorts. Additionally, the similar rates of choledocholithiasis in both groups suggest that this condition does not differ between MPN- and thrombophilia-associated PVT.

In our study, vessels involved in addition to the portal vein were evaluated separately. Involvement of the splenic vein and SMV was observed in 41.7% of patients in the MPN group and 26.5% in the thrombophilia group (p=0.191). Although these differences were not statistically significant, the higher rates in the MPN group may suggest a tendency for more extensive involvement of the portal system in these patients. In the literature, splenic vein involvement has been reported in approximately 20% and SMV involvement in approximately 14% of MPN-related SVT cases [18]. Comparative data across different etiologies remain limited.

The literature reports chronic parenchymal changes in the liver even in late-stage non-cirrhotic PVT [8]. In our study, mild chronic changes assessed by ultrasonography were more frequent in the thrombophilia group than in the MPN group (52.1% vs. 37.5%), although not statistically significant. This may be explained by the more isolated and clinically silent progression of PVT in the thrombophilia group and by the sensitivity of ultrasonography. Larger studies are needed to clarify these findings.

Abnormal endoscopic findings, including esophageal varices, gastric varices, and portal gastropathy, were common in both groups (MPN 70.0%; thrombophilia 75.0%), likely reflecting portal hypertension secondary to PVT. This indicates that endoscopic manifestations can appear similarly in MPN- and thrombophilia-related PVT. In one study, esophageal and gastric varices were detected in 79% of MPN-related PVT patients at diagnosis [7], comparable to our findings. However, no comparative studies by etiology have been reported.

The presence of PVCT in patients with PVT suggests chronicity of the disease and is typically accompanied by splenomegaly and collateral formation associated with the portal venous system [13]. PVCT is a compensatory mechanism that develops within a few days following vascular occlusion and becomes fully established within 3-5 weeks [19]. In our study, PVCT was observed in 79.2% of patients in the MPN group and

57.1% in the thrombophilia group (p=0.065). The higher prevalence of PVCT in the MPN group is noteworthy, suggesting that MPN may exert a stronger procoagulant effect in the development of PVT, leading to more frequent obstruction-related cavernous transformation. Clinically, PVCT is an important indicator of portal hypertension and its associated complications; therefore, its presence in patients with MPN may warrant closer monitoring and, if necessary, endoscopic or radiologic intervention. Although PVCT was not statistically significant in MLR analysis, it was associated with MPN and showed a tendency to increase the likelihood of MPN. These findings support the notion that PVCT may be a clinically relevant parameter for distinguishing non-cirrhotic PVT due to MPN from that due to thrombophilia.

Strengths and Limitations: This study is among the few directly comparing clinical and laboratory differences between MPN and hereditary thrombophilia in non-cirrhotic PVT. MLR and ROC analysis demonstrated the high discriminative ability of platelet count in predicting MPN and identified a clinically applicable cut-off, providing a practical parameter for differentiating PVT due to MPN from thrombophilia and guiding further genetic evaluation and treatment planning.

However, the study is retrospective and single-center, carrying a risk of selection bias. The limited sample size, especially in the MPN group, may have prevented some variables from reaching statistical significance. Findings are generalizable only to similar centers and populations; confirmatory studies with larger cohorts in different regions are warranted.

CONCLUSION

Non-cirrhotic PVT in patients with MPN and thrombophilia shows largely similar clinical and laboratory features. However, elevated platelet count emerged as an independent and strong predictor of MPN-associated PVT. Additionally, the higher prevalence of portal cavernous transformation and biliary complications in the MPN group is noteworthy. In patients with thrombocytosis, the possibility of MPN should be considered and supported by other laboratory and genetic tests. Our findings warrant validation in larger prospective studies.

Author contribution

Study conception and design: YSH and YB; data collection: YSH and YB; analysis and interpretation of results: YSH and YB; draft manuscript preparation: YSH and YB. All authors reviewed the results and approved the final version of the manuscript.

Ethical approval

Ethical approval for the specialty thesis entitled "Etiologic Distribution of Chronic Noncirrhotic Portal Vein Thrombosis in Adult Patients" (completed August 1, 2011; YÖKSİS Thesis No: 282140) was obtained from the Ethics Committee of Hacettepe University Faculty of Medicine prior to its initiation.

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Conflict of interest

The authors declare that there is no conflict of interest.

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