ORIGINAL ARTICLE

DOI: 10.4274/csmedj.galenos.2026.2026-2-5

Cam and Sakura Med J

CSMJ

Non-Thrombotic Portal Venous Pressure Increase at Diagnosis in Myeloproliferative Neoplasms

Veysel Bilgehan Bezirhan[1], Osman Yokuş[2]

[1]Dokuz Eylül University Faculty of Medicine, Department of Geriatrics, İzmir, Türkiye
[2]University of Health Sciences Türkiye, İstanbul Training and Research Hospital, Clinic of Hematology, İstanbul, Türkiye

What is known on this subject?

Chronic myeloproliferative neoplasms (MPNs) are associated with thrombotic complications, particularly involving the splanchnic venous system. Portal hypertension in these patients is most commonly linked to portal vein thrombosis. However, data regarding portal venous hemodynamics at the time of diagnosis, in the absence of thrombosis, are limited.

What this study adds?

In this study, patients with newly diagnosed MPNs showed higher portal venous congestion index values compared to controls, despite the absence of portal vein thrombosis. These findings suggest that alterations in portal venous hemodynamics may be present at diagnosis; however, their clinical significance requires further investigation.

ABSTRACT

Objective: Chronic myeloproliferative neoplasms (MPNs) are associated with an increased risk of thrombosis, including involvement of the portosplenic venous system. Portal hypertension is usually considered a consequence of portal vein thrombosis. This study aimed to investigate whether portal venous pressure is increased at diagnosis in patients with newly diagnosed MPNs, before the development of portal vein thrombosis.

Material and Methods: This single-center, non-interventional prospective cohort study included 62 newly diagnosed patients with chronic MPNs and 30 healthy controls. All patients met the 2016 World Health Organization diagnostic criteria and had no conditions that could independently cause portal hypertension. On the day of diagnosis, portal venous Doppler ultrasonography was performed by the same operator. Portal vein diameter, portal vein flow velocity, and congestion index were measured. Statistical analyses were conducted using SPSS version 24.0. A p value <0.05 was considered statistically significant.

Results: The mean congestion index was significantly higher in the patient group compared with the control group (0.06 ± 0.02 vs. 0.05 ± 0.01, $p=0.026$). No statistically significant differences were observed between groups in terms of portal vein flow velocity, portal vein diameter, age, or sex distribution ($p>0.05$).

Conclusion: Patients with newly diagnosed chronic MPNs may have increased portal venous pressure at the time of diagnosis, even in the absence of portal vein thrombosis. Early assessment of portal venous hemodynamics may be useful for the management and follow-up of these patients.

Keywords: Myeloproliferative neoplasms, portal hypertension, Doppler ultrasonography, congestion index

Introduction

Myeloproliferative neoplasms (MPNs) are clonal hematopoietic stem-cell disorders characterized by sustained proliferation of one or more myeloid lineages. The expansion of mature blood cells contributes to a prothrombotic state and vascular complications related to the dominant cell type (1). When ineffective or immature progenitors predominate, progressive marrow fibrosis and peripheral cytopenias may occur as part of disease evolution (2). Excluding chronic myeloid

Corresponding Author: Veysel Bilgehan Bezirhan, MD, Dokuz Eylül University Faculty of Medicine, Department of Geriatrics, İzmir, Türkiye
E-mail: veyselbezirhan@gmail.com **ORCID ID:** orcid.org/0000-0002-6311-8108

Received: 16.02.2026 **Accepted:** 29.04.2026 **Epub:** 07.05.2026

Cite this article as: Bezirhan VB, Yokuş O. Non-thrombotic portal venous pressure increase at diagnosis in myeloproliferative neoplasms. Cam and Sakura Med J. [Epub Ahead of Print]

leukemia (CML), which typically presents at older ages, the mean age at diagnosis for MPN occurs approximately during the sixth decade of life. A considerable proportion of patients are over 60 years old, and outcomes in this population are often influenced by comorbidities, reduced performance status, and age-related physiological decline (3).

MPN are traditionally classified into distinct clinical entities, including polycythemia vera (PV), essential thrombocythemia (ET), primary myelofibrosis (MF), CML, and other less well-defined subtypes (4). At the molecular level, the majority of classical MPNs are driven by mutations affecting key components of the Janus Kinase–Signal Transducer and Activator of Transcription (JAK–STAT) signaling pathway. The JAK2 *V617F* mutation represents the most prevalent driver alteration, whereas CALR and MPL mutations account for a substantial proportion of the remaining cases (5,6).

Thrombotic complications constitute the principal source of morbidity and mortality in patients with MPN (7). Although arterial events are more frequently encountered, venous thromboses represent a substantial clinical burden and may involve atypical vascular territories, particularly the splanchnic circulation (8,9). MPNs are among the most commonly identified underlying disorders in patients presenting with Budd–Chiari syndrome or non-cirrhotic portal vein thrombosis (9).

MPNs may influence portal venous hemodynamics through several pathophysiological mechanisms even before the occurrence of overt thrombosis. Increased blood cell mass, particularly in PV and ET, leads to increased blood viscosity and altered rheological properties of the circulation. In addition, chronic inflammatory activation and endothelial dysfunction, associated with aberrant JAK–STAT signaling, may contribute to microvascular disturbances within the splanchnic circulation. Splenomegaly, which is frequently observed in patients with MPNs, may further modify portal venous inflow and intrahepatic vascular resistance. Together, these mechanisms may result in subtle alterations in portal venous pressure and flow dynamics that precede clinically detectable portal vein thrombosis (10).

Assessment of portal venous hemodynamics using Doppler ultrasonography has enabled the evaluation of non-invasive parameters that may reflect portal pressure. The congestion index, defined as the ratio of the cross-sectional area of the portal vein to mean blood flow velocity, has been proposed as a surrogate marker of portal venous pressure and has been shown to correlate with invasively measured portal pressure values (11). These findings suggest that the congestion index may reflect hemodynamic alterations within the portal venous system.

Although portal hypertension in patients with MPN is commonly attributed to portal vein thrombosis, the possibility that subtle hemodynamic alterations may precede overt thrombotic obstruction has not been clearly defined. It remains uncertain whether portal venous pressure is already increased at the time of diagnosis in patients without radiologically detectable thrombosis. The present study was designed to evaluate, using Doppler ultrasonography, portal venous hemodynamics at diagnosis in patients with newly identified MPN. By assessing parameters such as portal vein diameter, flow velocity, and congestion index, we aimed to determine whether early alterations in portal venous pressure could be detected before the development of portal vein thrombosis.

Material and Methods

This single-center, prospective, non-interventional cohort study investigated whether portal venous pressure is elevated at the time of diagnosis in patients with newly diagnosed MPN without portal vein thrombosis.

The study was carried out at the Hematology Outpatient Clinic of University of Health Sciences Türkiye, İstanbul Training and Research Hospital between March 2021 and December 2021. Patients were eligible if they were newly diagnosed with MPN according to the 2016 World Health Organization diagnostic criteria (12,13). Exclusion criteria included an age under 18 years, advanced right-sided heart failure, known chronic liver disease, portal vein thrombosis, Budd-Chiari syndrome, or any other condition that could independently cause portal hypertension. Initially, 73 patients were screened for eligibility. Eleven individuals were excluded due to comorbid conditions that could interfere with portal venous hemodynamic assessment. The final analysis included 62 patients. The control group consisted of 30 healthy individuals comparable in age and sex, with no known hepatic, hematologic, or cardiovascular disease. All Doppler ultrasonographic examinations were performed on the day of diagnosis by a single experienced operator using the same ultrasound device to minimize interobserver variability. The following parameters were recorded: portal vein diameter (mm); portal vein flow velocity (cm/s); and the congestion index, calculated as the ratio of portal vein cross-sectional area (cm^2) to mean flow velocity (cm/s). The primary outcome variable was the congestion index, which has been shown to correlate with portal venous pressure.

The sample size was calculated a priori based on detecting a minimum expected difference in congestion index values between groups, with 80% statistical power and a two-sided

alpha level of 0.05. Based on the anticipated effect size derived from previous literature, a minimum of 30 patients per group was required. The final sample exceeded this minimum requirement. All participants provided written informed consent prior to enrollment. The study was approved by the University of Health Sciences Türkiye, İstanbul Training and Research Hospital Ethics Committee (decision number: 2780, date: 19.03.2021).

Statistical Analysis

Statistical analyses were performed using SPSS version 24.0 (IBM Corp., Armonk, NY, USA). Continuous variables are reported as mean ± standard deviation, and categorical variables as frequencies with percentages. The normality of the data distribution was assessed using the Kolmogorov–Smirnov test. For comparisons between the patient and control groups, the independent samples t-test was applied to normally distributed continuous variables, while the chi-square test was used for categorical variables. Correlations between continuous parameters were evaluated using Pearson's correlation coefficient. As baseline demographic characteristics were comparable between groups, no multivariable adjustment was considered necessary. All statistical tests were two-sided, and a p value <0.05 was accepted as statistically significant.

Results

In our study, the patient group comprised 62 individuals (45 men and 17 women), and the control group comprised 30 individuals (19 men and 11 women) (Table 1).

Among the patients, 54.8% were diagnosed with PV, 25.8% with ET, 3.2% with MF, 11.3% with CML, and 4.8% with an unclassifiable (undifferentiated) disease. 72.6% of the patients and 63.3% of the control group were male. There was no statistically significant association between group and sex ($p>0.05$) (Table 2).

The comparison of portal venous parameters between the patient and control groups is presented in Table 3. The mean congestion index was significantly higher in patients with MPN than in healthy controls ($p=0.026$). In contrast, no statistically significant differences were observed between the groups in portal vein flow velocity, portal vein diameter, or age.

Table 1. Total number of patients and control group members included in the study

		n	%
Group	Patient	62	67.4
	Control	30	32.6
	Overall	92	100.0

Correlation analyses performed within the patient group revealed a moderate positive association between congestion index and portal vein diameter. In addition, JAK2 levels showed a moderate positive correlation with platelet count, whereas erythropoietin levels were inversely correlated with platelet count. Lactate dehydrogenase demonstrated a weak positive association with age and a moderate negative association with hemoglobin levels. Neutrophil and platelet counts were both negatively correlated with hemoglobin. Detailed correlation coefficients and significance levels are presented in Tables 4 and 5.

Discussion

In the present study, we demonstrated that patients with newly diagnosed MPNs exhibited a significantly higher congestion index at the time of diagnosis compared with healthy controls, despite the absence of portal vein thrombosis. This finding suggests that alterations in portal venous hemodynamics may already be present in the early phase of the disease. Thrombotic risk in MPNs is multifactorial and involves quantitative and qualitative abnormalities of blood cells, endothelial dysfunction, and chronic inflammatory activation mediated through aberrant JAK–STAT signaling (14). Although the incidence and pattern of thrombotic events vary among disease subtypes, vascular complications remain a major determinant of morbidity across the spectrum. In MF, the risk of thrombosis is increased but appears to be less pronounced compared with PV and ET (15). In contrast, thrombotic complications are considered one of the leading causes of morbidity and mortality in ET (16). Even in the absence of macroscopic thrombosis, these mechanisms may contribute to subtle changes in portal circulation by increasing blood viscosity, promoting microvascular dysfunction, or altering intrahepatic vascular resistance.

Non-cirrhotic portal hypertension represents a heterogeneous clinical entity in which portal venous pressure increases in the absence of cirrhotic liver architecture (17). Prothrombotic conditions, including MPNs, have been implicated in its pathogenesis. In many reported cases, portal hypertension becomes clinically evident only after the development of portal vein thrombosis or complications such as ascites and variceal bleeding (18,19). Our findings suggest that portal venous hemodynamic alterations may precede these overt manifestations. In patients diagnosed with MF, the probability of developing portal hypertension ranges between 10–17% (20). In patients with ET, the rate of thrombosis has been reported to increase up to 24% during a 27-month follow-up period (21). In a study conducted in our

Table 2. Distribution of patients included in the study according to disease subgroups and sex

		Patient		Control		p value
		n	%	n	%	
Diagnosis	PV	34	54.8	0	0.0	
	ET	16	25.8	0	0.0	
	MF	2	3.2	0	0.0	
	CML	7	11.3	0	0.0	
	Undifferentiated	3	4.8	0	0.0	
Sex	Male	45	72.6	19	63.3	0.508
	Female	17	27.4	11	36.7	

CML: Chronic myeloid leukemia, ET: essential thrombocythemia, MF: myelofibrosis, PV: Polycythemia vera

Table 3. Comparison of patient and control groups according to the parameters used in the study

		n	Average	SD	t	p
Congestion index	Patient	62	0.0622	0.0197	-2.277	0.026
	Control	30	0.0536	0.0141		
Portal vein flow velocity (cm/s)	Patient	62	17.12	3.94	0.795	0.429
	Control	30	16.47	3.01		
Portal vein diameter (mm)	Patient	62	10.26	1.96	0.378	0.707
	Control	30	10.10	1.63		
Age	Patient	62	46.82	16.43	0.527	0.600
	Control	30	44.87	17.24		

SD: Standard deviation

country by Toros et al. (22), the relationship between portal hypertension and myeloproliferative diseases was investigated in 29 patients diagnosed with Philadelphia chromosome–negative PV, ET, and PMF. Of these 29 patients, 20 were followed with a diagnosis of ET, 7 with PV, and 2 with PMF. All patients underwent portal venous Doppler ultrasonography and esophagogastroduodenoscopy. Portal hypertension was detected in 4 patients, 2 with PV, 1 with PMF, and 1 with ET; however, no statistically significant association was established ($p>0.05$). In the same study, the congestion index was evaluated, with a median value of 0.11 cm × s. In our study, this value was 0.06 cm × s. A possible explanation for this difference is the presence of severe portal hypertension due to portal vein thrombosis and splenic thrombosis in 2 of the 29 patients. The mean portal vein diameter in their study was 10.2 mm, like that observed in our study (22).

Among the 62 patients with MPN included in our study, 54.8% were diagnosed with PV, 25.8% with ET, 3.2% with PMF, 11.3% with CML, and 4.8% with unclassifiable myeloproliferative disease. In a study by Georgii et al. (23), in which 2,901 bone marrow samples were retrospectively evaluated, 19.8% of patients had ET, 18.2% PV, 28.8% CML, and 12% unclassifiable myeloproliferative disease. In another study conducted by Chebrek et al. (24), the distribution was reported as 32% CML, 27% ET, 15% PMF, and 26% PV. In a study by Bari et al. (25), evaluating 380 patients with MPN in Northern Italy, 41% had ET, 30% PV, 20% PMF, and 8% had unclassifiable myeloproliferative disease. Although the results of studies in the literature vary, all studies were evaluated retrospectively. Our study included patients newly diagnosed with MPN, which may explain the lower percentage of PMF observed in the early period. Transformation to PMF can occur, particularly in PV and ET. In addition, although the annual incidence of ET has been reported as 1–2.5 per 100,000 (26), because life expectancy is normal, the true prevalence is thought to be 9–24 per 100,000 (27).

In our cohort, male patients predominated, which may largely reflect the high proportion of PV cases. Previous studies have reported sex-related differences across MPN subtypes, with PV occurring more frequently in men, whereas ET is more commonly observed in women (23). The distribution observed in our study is therefore consistent with a predominance of PV in our patient population. The congestion index is defined as the ratio of the cross-sectional area of the portal vein (cm^2) to blood flow velocity (cm/s). In portal hypertension, the congestion index has been

suggested to reflect the pathophysiological hemodynamics of the portal venous system. Previous Doppler studies have reported congestion index values in healthy individuals, typically ranging from approximately 0.04 to 0.10, reflecting the physiological variability of portal venous circulation. In our study, the values observed in both the control and MPN groups fall within the expected physiological range, although the significantly higher values in the MPN group may suggest subtle early alterations in portal venous hemodynamics (10). When we evaluated the congestion index in all MPN patients at the time of diagnosis, in the absence of thrombosis, we found it to be significantly higher than in the control group (p=0.026). This finding suggests that patients with MPN may already have increased portal venous pressure, which is one of the complications of the disease, at the time of diagnosis. Because the number of patients with certain MPN subtypes was limited in our cohort, meaningful subgroup comparisons between individual disease categories were not feasible. Future studies that include larger and more balanced patient populations may help clarify whether portal

Table 4. Relationship between the findings detected in patients

Group-patients	1	2	3	4	5	6	7	8	9	10	11	12
Congestion index	1.000											
JAK-2 levels	0.178	1.000										
Portal vein flow velocity (cm/s)	0.036	0.021	1.000									
Portal vein diameter (mm)	0.505	0.023	0.313	1.000								
Erythropoietin	-0.156	0.015	-0.038	-0.181	1.000							
Age	0.052	0.172	-0.174	-0.077	-0.293	1.000						
Age at diagnosis	0.064	0.168	-0.187	-0.080	-0.249	0.988	1.000					
Lactate dehydrogenase	0.034	0.069	0.033	-0.080	-0.146	0.281	0.290	1.000				
White blood cell	0.161	0.070	0.034	-0.056	-0.117	0.010	0.047	0.402	1.000			
Neutrophil	0.166	0.146	0.046	-0.031	-0.141	-0.013	0.013	0.481	0.939	1.000		
Platelets	0.095	0.450	0.155	-0.143	-0.425	0.036	0.023	0.127	0.164	0.213	1.000	
Hemoglobin	0.146	-0.141	-0.190	0.136	-0.099	-0.122	-0.132	-0.431	-0.431	-0.391	-0.372	1.000

JAK-2: Januse Kinase-2

Table 5. Detailed values of the study parameters in the patient and control groups

	Patient					Control				
	Min.	Max.	Average	SD	Median	Min.	Max.	Average	SD	Median
Congestion index	0.0130	0.0830	0.06	0.01	0.0515	0.029	0.117	0.05	0.02	0.0590
JAK-2 levels	0.00	60.00	6.57	15.33	0.00					
Portal vein flow velocity (cm/s)	10.00	35.00	17.12	3.94	17.00	11.00	24.00	16.47	3.01	16.00
Portal vein diameter (mm)	7.00	15.00	10.26	1.96	10.00	5.00	13.00	10.10	1.63	10.00
Erythropoietin	1.20	17.60	6.74	4.47	6.15					
Age	18.00	78.00	46.82	16.43	48.00	25.00	75.00	44.8	17.2	39.50
Age at diagnosis	18.00	78.00	45.27	16.58	44.50					
Lactate dehydrogenase	123.00	1322.00	303.69	235.34	230.00					
White blood cell	4330.0	387050.0	24,236.9	56,760.8	9230.0					
Neutrophil	2370.0	163360.0	14,120.0	28,817,7	5490.0					
Platelets	92.00	1837.00	421.29	328.48	271.00					
Hemoglobin	8.50	21.50	15.80	3.04	16.35					

JAK-2: Januse Kinase-2, SD: Standard deviation, Min.: Minimum, Max.: Maximum

venous hemodynamic alterations differ among specific MPN subtypes, such as PV and ET.

Arterial and venous thrombosis are among the leading causes of mortality and morbidity in MPN, and atypically located thrombotic events are also common in this patient group. Our findings suggest that even in the absence of portal system thrombosis, patients with newly diagnosed MPN may already have increased portal venous pressure at the time of diagnosis (p=0.026). The mean age at diagnosis in our cohort was 46.8 years, indicating an earlier age at diagnosis compared with values reported in the literature. Sex distribution varied across disease subtypes, and the predominance of male patients in our study was primarily due to the high proportion of PV cases. PV constituted the majority of our cohort (54.8%), while ET accounted for 25.8. The relatively lower ET rate compared with that reported in the literature may be attributable to the frequently asymptomatic course of ET and to limitations in outpatient hematology referrals.

The identification of increased congestion index values at the time of diagnosis may have important clinical implications. If portal venous hemodynamic alterations precede overt portal vein thrombosis, early Doppler assessment may contribute to risk stratification in patients with MPN. Detecting such subclinical changes might allow closer monitoring and potentially earlier intervention in individuals at higher vascular risk. Although the clinical consequences of mildly elevated congestion index values remain to be clarified, our findings suggest that portal circulation may be affected earlier in the disease course than previously recognized.

In our cohort, the difference in congestion index between patients and controls was modest in absolute terms; however, even small alterations in portal venous hemodynamics may reflect early pathophysiological changes in the portal circulation. The congestion index has been proposed as a non-invasive indicator of portal venous pressure, and increases in this parameter may suggest subtle elevations in intrahepatic vascular resistance or alterations in portal venous inflow (10,11). Therefore, the slightly higher congestion index observed in patients with newly diagnosed MPNs may represent an early hemodynamic signal preceding clinically apparent portal hypertension or portal vein thrombosis. From a clinical perspective, the detection of such subclinical changes could be relevant for identifying patients who may benefit from closer surveillance of the portal venous system.

In the present study, multivariable adjustment was not performed. The primary comparison focused on newly diagnosed MPN patients and healthy controls with comparable baseline demographic characteristics; no statistically significant differences were observed between the groups in age or sex distribution. Portal vein diameter and flow velocity were similar between the groups. Given the relatively limited sample size and the exploratory nature of the study, multivariable modeling was not considered appropriate, as the inclusion of multiple covariates could lead to model instability. Nevertheless, future studies with larger cohorts may benefit from multivariable analyses to further explore potential independent determinants of alterations in portal venous hemodynamics in patients with MPN.

Study Limitations

Several limitations should be acknowledged. First, this was a single-center study with a relatively small sample size, which may limit generalizability. Second, portal venous pressure was not measured invasively; instead, the congestion index derived from Doppler ultrasonography was used as a surrogate marker. Third, subgroup analyses according to individual MPN subtypes were not feasible due to the small number of patients in certain categories. Finally, portal venous parameters were assessed at a single time point at diagnosis, and longitudinal follow-up data were not available. Therefore, it remains unclear whether the observed increase in the congestion index persists over time or predicts future development of portal vein thrombosis or clinically significant portal hypertension. Longitudinal studies evaluating the evolution of portal venous hemodynamics during disease follow-up would provide valuable insights into the temporal relationship between MPN progression and portal circulation alterations.

Conclusion

In conclusion, patients with newly diagnosed MPNs exhibited higher portal venous congestion index values compared to healthy controls, despite the absence of portal vein thrombosis. These findings suggest that alterations in portal venous hemodynamics may occur early in the disease course. However, the clinical relevance of these changes remains unclear, and further studies are needed to clarify their potential role in disease assessment and management.

Ethics

Ethics Committee Approval: The study was approved by the University of Health Sciences Türkiye, İstanbul Training and Research Hospital Ethics Committee (decision number: 2780, date: 19.03.2021).

Informed Consent: All participants provided written informed consent prior to enrollment.

Footnotes

Authorship Contributions

Surgical and Medical Practices: V.B.B., O.Y., Concept: V.B.B., O.Y., Design: V.B.B., O.Y., Data Collection or Processing: V.B.B., Analysis or Interpretation: V.B.B., Literature Search: V.B.B., Writing: V.B.B.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study received no financial support.

REFERENCES

1. Burkhardt R, Bartl R, Jäger K, et al. Chronic myeloproliferative disorders (CMPD). Pathol Res Pract. 1984;179:131-186.
2. Fayemi AO, Gerber MA, Cohen I, Davis S, Rubin AD. Myeloid sarcoma. Review of the literature and report of a case. Cancer. 1973;32:253-258.
3. Atilla FD, Şahin F, Saydam G. Chronic myeloproliferative disorders other than chronic myeloid leukemia in elderly patients. In: Nalçacı M, editor. Geriatric Approach in Hematology. Ankara: Türkiye Klinikleri; 2018. p. 31-37.
4. Marin Oyarzún CP, Heller PG. Platelets as mediators of thromboinflammation in chronic myeloproliferative neoplasms. Front Immunol. 2019;10:1373.
5. Rumi E, Cazzola M. Diagnosis, risk stratification, and response evaluation in classical myeloproliferative neoplasms. Blood. 2017;129:680-692.
6. Vainchenker W, Kralovics R. Genetic basis and molecular pathophysiology of classical myeloproliferative neoplasms. Blood. 2017;129:667-679.
7. Chachoua I, Pecquet C, El-Khoury M, et al. Thrombopoietin receptor activation by myeloproliferative neoplasm associated calreticulin mutants. Blood. 2016;127:1325-1335.
8. Barbui T, Finazzi G, Falanga A. Myeloproliferative neoplasms and thrombosis. Blood. 2013;122:2176-2184.
9. Falanga A, Marchetti M. Thrombosis in myeloproliferative neoplasms. Semin Thromb Hemost. 2014;40:348-358.
10. Dubois A, Dauzat M, Pignodel C, et al. Portal hypertension in lymphoproliferative and myeloproliferative disorders: hemodynamic and histological correlations. Hepatology. 1993;17:246-250.
11. Moriyasu F, Nishida O, Ban N, et al. "Congestion index" of the portal vein. AJR Am J Roentgenol. 1986;146:735-739.
12. Barbui T, Thiele J, Gisslinger H, et al. The 2016 WHO classification and diagnostic criteria for myeloproliferative neoplasms: document summary and in-depth discussion. Blood Cancer J. 2018;8:15.
13. Arber DA, Orazi A, Hasserjian R, et al. The 2016 revision to the World Health Organization classification of myeloid neoplasms and acute leukemia. Blood. 2016;127:2391-2405.
14. Arachchillage DR, Laffan M. Pathogenesis and management of thrombotic disease in myeloproliferative neoplasms. Semin Thromb Hemost. 2019;45:604-611.
15. Cervantes F, Alvarez-Larrán A, Arellano-Rodrigo E, Granell M, Domingo A, Montserrat E. Frequency and risk factors for thrombosis in idiopathic myelofibrosis: analysis in a series of 155 patients from a single institution. Leukemia. 2006;20:55-60.
16. Besses C, Cervantes F, Pereira A, et al. Major vascular complications in essential thrombocythemia: a study of the predictive factors in a series of 148 patients. Leukemia. 1999;13:150-154.
17. Gentilucci UV, Gallo P, Perrone G, et al. Non-cirrhotic portal hypertension with large regenerative nodules: a diagnostic challenge. World J Gastroenterol. 2011;17:2580-2584.
18. Goh BK, Chen JJ, Tan HK, Yong WS, Chan WH. Acute variceal bleed in a patient with idiopathic myelofibrosis successfully treated with endoscopic variceal band ligation. Dig Dis Sci. 2007;52:173-175.
19. Abu-Hilal M, Tawaker J. Portal hypertension secondary to myelofibrosis with myeloid metaplasia: a study of 13 cases. World J Gastroenterol. 2009;15(25):3128-3133.
20. Nakai GS, Craddock CG, Figueroa WG. Agnogenic myeloid metaplasia. A survey of twenty-nine cases and a review of the literature. Ann Intern Med. 1962;57:419-440.
21. Cortelazzo S, Finazzi G, Ruggeri M, et al. Hydroxyurea for patients with essential thrombocythemia and a high risk of thrombosis. N Engl J Med. 1995;332(17):1132-1136.
22. Toros AB, Gokcay S, Cetin G, Ar MC, Karagoz Y, Kesici B. Portal hypertension and myeloproliferative neoplasms: a relationship revealed. ISRN Hematol. 2013;2013:673781.
23. Georgii A, Buhr T, Buesche G, Kreft A, Choritz H. Classification and staging of Ph-negative myeloproliferative disorders by histopathology from bone marrow biopsies. Leuk Lymphoma. 1996;22:15-29.
24. Chebrek S, Aïssi K, Francès Y, et al. Pulmonary hypertension in patients with chronic myeloproliferative neoplasms. Leuk Lymphoma. 2014;55:223-225.
25. Bari A, Marcheselli R, Rashid I, et al. Incidence and outcome of chronic myeloproliferative disorders: a population-based study from a cancer registry in Northern Italy. Blood. 2007;110:4650.
26. Srour SA, Devesa SS, Morton LM, et al. Incidence and patient survival of myeloproliferative neoplasms and myelodysplastic/myeloproliferative neoplasms in the United States, 2001-12. Br J Haematol. 2016;174:382-396. Erratum in: Br J Haematol. 2017;177:331.
27. Ma X, Vanasse G, Cartmel B, Wang Y, Selinger HA. Prevalence of polycythemia vera and essential thrombocythemia. Am J Hematol. 2008;83:359-362.