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Characteristic and Complication Patterns of Polycythaemia Vera: A Single Centre Experience

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Abstract

Background and objectives: In addition to being at risk for thrombosis and disease progression, patients with polycythemia vera frequently have a wide variety of clinical, demographic and laboratory presentations which vary according to the geographical area and ethnic groups. This study aimed at defining these variations in our locality and comparing it to other international studies.

Methods: This cross-sectional study was carried out from January 2022 to January 2023 at Hiwa Cancer Hospital, Sulaymaniyah City, with a total of 200 adult cases of already diagnosed and registered cases of polycythemia vera according to the 2016 WHO criteria. The overview of demographic profiles: the age distribution, gender prevalence, laboratory characteristics, and complications patterns of these cases were studied.

Results: From the total of 200 cases, 118 (59%) were males and 82 (41%) of them were females , the mean age (60.4 ± 12.4) year, with a range of (25–88), 47 (23.5%) patients were having complications, either thrombosis 19 (40.4%), acute myeloid leukemia progression, 8 (4%), or myelofibrosis 20 (42.6%), and 153 (76.5%) of patients did not show any complication, 104 (52%) had splenomegaly. There were significant associations between development of complications and certain parameters like: age, disease duration, leukocyte count, platelets, and hemoglobin levels, in a concordance way i.e., the higher the value of these parameters, the higher the risk of developing complications.

Conclusions: distinctive patterns of these parameters were identified in our locality and correlated to other geographical areas. Our findings showed the significance of understanding different patterns of patient characteristics and complications of polycythemia vera in different regions.

Keywords: Characteristics, Complications, Polycythemia Vera, Single Centre

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Introduction

Polycythemia Vera (PV) is ~~a~~ part of myeloproliferative neoplasms characterized by JAK2 mutation, clonal erythrocytosis, also including leukocytosis, raised platelets, palpable spleen, constitutional symptoms and risk of thrombosis as well as myelofibrotic progression (post-PV MF) or leukemic transformation (post-PV AML). The average age, from diagnosis, of patients with PV in some studies is 61 years, 10% of whom are under the age of 40 years.[1] Nearly (36%) had enlarged spleen, and 25% with a thrombotic event or bleeding (4%) .[2] In a study involving 1545 cases with PV, after a median of seven years of follow up, outcomes included death (23%), transformation to leukemia (3%), progression to fibrosis (9%), thrombosis of both arterial (12%) and venous (9%), and major bleeding (4%). Major causes of death were leukemic transformation, secondary malignancy, and thromosis.[1,3,4] Regarding thrombosis occurrence, patients were categorized into two groups, age below 60 years without thrombotic events regarded as low-risk, on the other hand, patients aged over 60 years with thrombotic events regarded as high-risk group, The mortality rate of PV patients (4.1%) over 60 years of age was higher than the general population of the same age group (2.8%).[5] Although previous studies have showed valuable insights into several aspects of polycythemia vera, there are still some knowledge gaps and wide geographical variations regarding the spectrum of the patient's demographic, clinical, laboratory characteristics, and rate of complications associated with this hematological disorder, especially when considering our locality. The objectives_of this cross-sectional study were to analyze the demographic, clinical, laboratory and complication patterns of patients with polycythemia vera at Hiwa Hospital and comparing it with other studies reported in the literature. We anticipate it could improve our understanding of polycythemia in this locality by exploring this group of individuals. Our findings should give relevant information that can guide future research, and development and influence therapeutic practice.

Patients and methods

This is a single-center, cross-sectional, observational, retrospective study, which included 200 adults (> 18 years) patients with polycythemia vera (PV) cases who had been diagnosed according to the 2016 WHO criteria for the MPN classification and diagnosis.[6] The information was collected at Hiwa Hospital, a specialized tertiary center for cancer care, through its data recording system whom were registered through the period 2015–2022, only those who were permanent residents of Sulaymaniyah City were included, other Kurdistan regional government or Iraqi cities were excluded. The demographic, some clinical, and laboratory data at the time of the initial diagnosis of PV were collected, including age at diagnosis, duration of the disease, gender, presence of splenomegaly, original hemoglobin, WBC, platelet counts, serum erythropoietin, JAK2 mutation, bone marrow study, and presence of disease-related complications, like thrombosis, myelofibrotic transformation (post-PV MF), and leukemic transformation (post-PV AML).Data was entered and analyzed using Statistical Package for Social Sciences (SPSS) version 25 software. Categorical variables data (e.g., gender and presence of complications) are expressed as frequencies and percentages. Chi-square test for categorical variables was used for group comparisons. A significance level of ≤ 0.05 was considered to be statistically significant.
Ethical approval was obtained from the ethics committee of the Kurdistan Higher Council for Medical Specialties. The current analysis was exempted from the patient consent agreement, as no personal

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identifying information was collected or reported.

Results

A total of 200 diagnosed cases of polycythemia vera were enrolled. Cases had a mean age of (60.4 ± 12.4) years, with a range of (25–88), the majority being male (118, 59%) as compared to female (82, 41%). The mean duration of the disease was (4.9 ± 1.6) years, with a range of (2.0-9.0). The laboratory results of Hb, WBC, and platelet count are explained in Table (1).

Table (1): Frequency of some parameters in polycythemia vera cases

Categories	Age/year	Original Hb g/dL	WBC $\times 10^9$/L	Platelet counts $\times 10^9$/L	Duration since diagnosis/year
Mean.	60.4	17.9	16.51	525.4	4.99
(Range)	(25–88)	(16.5-20.8)	(12.8-23.0)	(209–988)	(2.0-9.0)
SD	12.4	0.9	2.26	179	1.96

Other categorical data such as the presence of enlarged spleen, JAK 2 mutation, erythropoietin level, and bone marrow finding, are explained in Table (2)) and Figure (1).

Table (2): Some clinical and laboratory characteristics of patients in Polycythemia Vera

Categories	Frequency n (%)
Gender	
Female	82 (41)
Male	118 (59)
Splenomegaly	
No	96 (48)
Yes	104 (52)
Bone marrow	
N/A*	32 (16)
Pan-myelosis	168 (84)
EPO (erythropoietin)	
N/A*	156 (78)
Subnormal	44 (22)
JAK2	
Negative	12 (6)
Positive	188 (94)
Complications	
AML	8 (17)
MF	20 (43)
Thrombosis	19 (40)
Total Complications	
Yes	50 (25)
No	150 (75)

* Not available

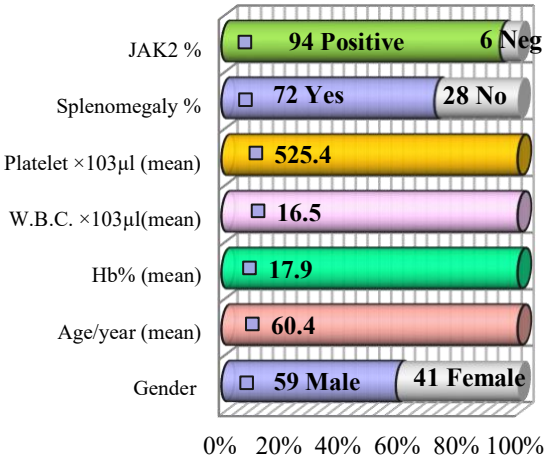

Figure (1): Frequency of various parameters in the participants

Regarding complications development during the study period, such as thrombosis both arterial and venous, progression to acute leukemia and bone-marrow fibrosis, 47 (23.5%) patients were having complications, either thrombosis 19 (40.4%), acute myeloid leukemia AML progression, 8 (17%), or MF 20 (42.6%), and 153 (76.5%) of patients did not show any complication, as shown in figure (2).

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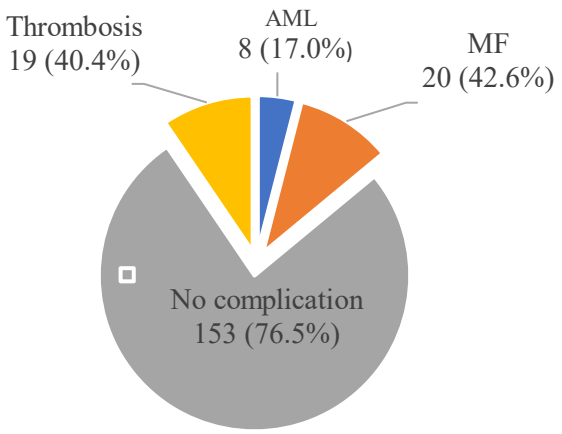

Figure (2): Frequency and percentage of PV complications

When the development of total complications (acute leukemia, post PV-MF, and thrombosis) was collectively compared to other parameters like; age, Hb, WBC, platelet count at diagnosis and duration of the disease it revealed the statistical significance in each parameter between patients who experienced complications and those who did not, as general, in a way that the higher these parameters value, the higher the likelihood of complications. For instance, if we take the age of patients who developed complications, 12 of the 47 (25%) were younger than 60 years and 35 of the 47 (74.5%) were older than 60 years and the P-value was less than ≤ 0.05, suggesting that there is a significant correlation between PV patients' ages and complications, and so on for other parameters, as shown in more detail in Table (3). According to the table, older patients with PV who have higher hemoglobin levels, longer disease duration, higher platelet counts, and higher white blood cell counts are more likely to experience complications.

Table (3): Correlation between total complications and some parameters and their significance

Parameters	Complications		Total n (%)	p value
	Absent n (%)	Present n (%)		
Age at diagnosis (year)				
≤ 60	86(87.8)	12(12.2)	98(49)	≤ 0.05
> 60	67(65.7)	35(34.3)	102(51)	
WBC count × 10^9/L				
≤ 16	148(87.6)	21(12.4)	169(85)	≤ 0.05
> 16	5(16.1)	26(83.9)	31(15)	
Platelet count× 10^9/L				
≤ 350	135(95.1)	7(4.9)	142(71)	≤ 0.05
> 350	18(31.0)	40(69.0)	58(29)	
Duration of disease/Year				
< 4.5	80(96.4)	3(3.6)	83(42)	≤ 0.05
≥ 4.5	73(62.4)	44(37.6)	117(56)	
Hemoglobulin level (g/dl)				
<1 7.5	70(95.6)	3(4.1)	73(37)	0.576
17.5-18.5	59(78.7)	16(21.3)	75(38)	≤ 0.05
> 18.5	24(46.2)	28(53.8)	52(25)	
Total	153(76.5)	47(23.5)	200(100)	

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When considering individual complications separately (AML, post PV-MF, and thrombosis) and by correlating them with some parameters like age, gender, splenomegaly, Hb, WBC, platelet count at diagnosis and duration of the disease, significant correlations between parameters as platelet count at diagnosis, duration of the disease and splenomegaly were found to be associated with development of individual complications, as shown in Table (4).

Table (4): Correlation between individual complications and some parameters and their significance

Parameters	Complications			Total n (%)	p value
	AML n (%)	MF n (%)	Thrombosis n (%)		
Gender					
Female	5(20.0)	7(28.0)	13(52.0)	25(53)	0.09
Male	3(13.6)	13(59.1)	6(27.3)	22(47)	
Age at diagnosis (year)					
≤60	2(16.7)	4(33.3)	6(50.0)	12(26)	≤ 0.05
>60	6(17.1)	16(45.7)	13(37.1)	35(74)	
Duration of disease/Year					
<4.5	0 (00.0)	2 (10.0)	1 (5.3)	3(6)	≤ 0.05
≥4.5	8 (100.0)	18 (90.0)	18 (94.7)	44(94)	
WBC count × 10^9/L					
≤16	1(4.8)	10(47.6)	10(47.6)	21(45)	0.13
>16	7(26.9)	10(38.5)	9(34.6)	26(55)	
Platelet count× 10^9/L					
≤ 350 n (%)	1(14.3)	6(85.7)	0(0.0)	7(15)	≤ 0.05
> 350 n (%)	7(17.5)	14(35.0)	19(47.5)	40(85)	
Hemoglobulin level (g/dl)					
<17.5	0(0.0)	2(66.7)	1(33.3)	3(6)	0.087
17.5-18.5	3(18.8)	6(37.5)	7(43.8)	16(34)	
>18.5	5(17.90	12(42.9)	11(39.3)	28(60)	
Splenomegaly					
No	0(0.0)	0(0.0)	5(100.0)	5(11)	≤ 0.05
Yes	8(19.0)	20(47.6)	14(33.3)	42(89)	
Total	8(17)	20(43)	19(40)	47(100)	

Patients with any complications are typically older than patients without complications; seventy-four percent of them were 60 years of age or older. Patients with thrombotic events have the highest platelet counts; 19 (47.5%) of them have counts more than 350 × 10^9/L, none of them had counts less than 350 × 10^9/L. This implies that the platelet count is a strong risk factor for thrombosis in polycythemia vera patients, and likewise for other parameters.

Discussion

The study revealed; male predominance, mean age of presentation of about 60 years, mean disease duration of 5 years, more than half had splenomegaly, majority were JAK2 positive, and mean Hb, WBC, and platelet counts of 17.9, 16.5, and 425.4 respectively,

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regarding complications about a quarter of them developed complications including; AML, thrombosis, and post PV-MF with increasing frequency. These characteristics ae consistent with other study[6] findings in patients with PV; with significant proportion of patients were over 60 years old and are categorized as high-risk due to their age or presence of thrombotic events. It is established that these variables raise the patients' risk of thrombotic events.[6]The demographics of this study were compared to those reported in other studies. The mean age at diagnosis (60.4) years was similar to that was found the European study done by Marchioli R [7] in 2005 which was (60.4) years and very close to that shown in a study done in Germany by Moulard O. in 2014 (64.3).[8] However, it was older than the (53.4) in Taiwan done by Tsai TH. in 2023. [5,9] and younger than 71.5 years in a study done by Jackson NL in 2018 in Canada,[10] and 73 years in the UK.[10] In a study done by Mehta J in 2014 in the USA broad range of age groups (54.5–77) were documented. [11] A worldwide study done by Barbui T. in 2014 including some European countries and the USA that involved 1545 cases reported 61 years as mean age.[12] A study done by Varghese.in New Zealand between 2010 and 2017 compared the age among different ethnic groups revealed these results: Caucasian (71.7), Māori (59.5), and Pacific Islanders (56.8) years.[13]As in majority of the studies, we found that male patients were more affected than females (59% males, 41% females). This difference was near to those done by Byun (60:40) ,[14] by Lim Y, (57:43) ,[15] by Bai J (54:46) ,[16] by Varghese (52:48) ,[13] and by Mehta (58:42) .[11] Although such trend was not reported in Norway (50:50).[13] On the other hand, another research conducted in China , showed that there were more female patients (48:52).[7] One factor for the male patient predominance in our research may be the mislabeling of secondary polycythemia as PV, as secondary polycythemia is more common in males due to more prevalence of smoking. Splenomegaly was found in 104 (52%) of patients in our study. Enlarged spleen has been found in (30–40%) and it is more common with progressive disease. In a meta-analysis that was done on 1545 from USA and some European countries, Tedderi found that 36% have splenomegaly.[2] Another study done by Mesa in the USA that included 587 patients also reported that 31% of the patients had splenomegaly .[9] Although lower splenomegaly cases were found in other studies. A multi-focal study by Grunwald in the USA which enrolled 1601 cases documented 17.6%.[18] Although more advanced and delayed disease presentation may be one factor behind the higher prevalence of palpable spleen in our study, also there may be some genetic factors causing diverse phenotypic types in different ethnic groups. Regarding development of complications, thrombotic events are relatively common sequelae in these patients, with ominous effect on patient's outcome. This study showed that the percentage of thrombosis was (9%) , close to those reported in a study done by Tefferi internationally including cases from Austria, Italy and the USA, where the rates were 9-12 %.[2] The percentage are much higher in other studies, some may reach as high as 34 to 39% of cases.[19] Some of the well-established factors that are associated with this complication are: age, duration of the disease, previous thrombotic events, hemoglobin level, leukocytosis, and JAK2V617F mutation.[20] In In this study there was a significant relationship of thrombosis with some of these parameters. As related to the fibrotic complications, a study done in 2015 by Cerquozzi and Tefferi revealed that the rate of the complication was <10% for a follow up period of 10 years and less.[20] The current study however, showed this to be 12%,

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throughout the study mean duration of about five years. A study done by Bai et al. in 2015 showed higher incidence of fibrosis among patients in China which was 27.4% at 10,39.9% at 15, and 61.1% at 20 years.[21,22] These discrepancies may be due to the duration of the disease, late presentation, and treatment received. Some established factors that have been associated with this complication are: age at diagnosis, leukocytosis, and JAK2V617F mutational status. [23] As revealed by this study, a significant relationship between some of these parameters and the development of post PV-MF were shown. In regards to leukemic transformation this study showed that (4%) of cases developed the condition in the mean of 5 years' disease duration. When compared to other studies, the following findings were revealed: In a multicenter research data, the rate was higher; 2.3%, 5.5%, and 10% at 10,15, and 20 years respectively.[2] Still higher rates have been reported in smaller studies; 8–14%, 14–19%, and up to 24% at 10,15 and 18 years respectively.[24] This complication usually happens between 4.6–19 years from the disease diagnosis.[25] This may explain the relatively lower rate of this complication in this locality. The rate of this complication is usually related to several factors including age, high WBC count, disease duration, and JAK2 status.[20]

Conclusions

This study revealed a better understanding regarding demographics, risk profiles, laboratory presentation, and clinical outcomes of PV cases in our locality. Furthermore, we reported several patient characteristics that were found to be different from those of other populations reported in previous studies, which may provide some evidence that requires further investigation into the landscape of PV among other geographic and ethnic groups. There were various shortcomings in this study. First, it was a single-centre study and less likely to be representative of the whole our regional population, Second, the short duration of the follow-up makes the incidence of the outcome to be under-reported. Third sample size, a larger population size would be more representative.

For the future we recommend the study to be repeated in other centers of Kurdistan region to be more representative of our region.

Conflict of interest

All authors declare that they have no conflicts of interest to disclose.

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