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OPIS PRZYPADKU
CASE REPORT

Chronic lymphocytic leukemia coexisting with polycythemia vera in a 67-year-old man: A case report

Przewlekła białaczka limfocytowa współistniejąca z czerwienicą prawdziwą u 67-letniego mężczyzny – opis przypadku

Grzegorz K. Jakubiak[1,2], Jerzy Pruszyński[2], Ewa Ziółko[2]

[1]Department of Pharmacology, Faculty of Medical Sciences in Zabrze, Medical University of Silesia, Katowice, Poland

[2]Oddział Hematologii, Szpital Specjalistyczny Nr 1 w Bytomiu / Ward of Hematology, Specialist Hospital No. 1 in Bytom, Poland

ABSTRACT

Polycythemia vera (PV) is a chronic myeloproliferative neoplasm characterized by the absence of the Philadelphia chromosome, increased hemoglobin concentration, and increased erythrocyte mass in peripheral blood. Chronic lymphocytic leukemia (CLL) is a lymphoproliferative neoplasm characterized by the chronic presence in the peripheral blood of a population of clonal B lymphocytes with a typical immunophenotype, usually accompanied by an invasion of the lymph nodes, spleen, and bone marrow. The purpose of this paper is to present a case report of a 67-year-old man diagnosed with PV with coexisting CLL. The patient was admitted to the Ward of Hematology for planned diagnostic workup for polycythemia. The presence of the V617F mutation of the *JAK2* gene was confirmed. Due to the elevated lymphocytosis, the diagnostic workup was expanded to include immunophenotyping of the peripheral blood cells, which revealed the presence of a clone with an immunophenotype typical of CLL. Histopathological examination of the bone marrow, including immunohistochemical staining, confirmed PV with concomitant CLL infiltration.

KEYWORDS

polycythemia vera, chronic lymphocytic leukemia, trephine biopsy

STRESZCZENIE

Czerwienica prawdziwa (*polycythemia vera* – PV) to przewlekły nowotwór mieloproliferacyjny bez obecności chromosomu Philadelphia, charakteryzujący się zwiększonym stężeniem hemoglobiny i zwiększoną masą erytrocytarną we krwi obwodowej. Przewlekła białaczka limfocytowa (*chronic lymphocytic leukemia* – CLL) to nowotwór limfoproliferacyjny, charakteryzujący się przewlekłą obecnością we krwi obwodowej populacji klonalnych limfocytów B o typowym immunofenotypie, zwykle z naciekiem węzłów chłonnych, śledziony i szpiku kostnego. Celem pracy jest

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Address for correspondence: dr n. med. Grzegorz Jakubiak, Katedra i Zakład Farmakologii, ul. Jordana 38, 41-808 Zabrze, tel. +48 32 272 26 83, e-mail: grzegorz.jakubiak@sum.edu.pl

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przedstawienie opisu przypadku 67-letniego mężczyzny, u którego rozpoznano PV ze współistniejącą CLL. Pacjent został przyjęty na Oddział Hematologii w celu planowej diagnostyki nadkrwistości. Potwierdzono obecność mutacji V617F genu *JAK2*. Ze względu na podwyższoną limfocytozę diagnostykę rozszerzono o badanie immunofenotypowe komórek krwi obwodowej, które ujawniło obecność klonu o immunofenotypie typowym dla CLL. Badanie histopatologiczne szpiku kostnego, obejmujące barwienie immunohistochemiczne, potwierdziło PV z towarzyszącym naciekiem CLL.

SŁOWA KLUCZOWE

czerwienica prawdziwa, przewlekła białaczka limfocytowa, trepanobiopsja

INTRODUCTION

Polycythemia vera (PV) is a chronic myeloproliferative neoplasm characterized by the absence of the Philadelphia chromosome. More than 95% of patients have the V617F driver mutation in the *JAK2* gene [1]. PV is characterized by elevated hemoglobin concentration and elevated erythrocyte mass in the peripheral blood which is often accompanied by leukocytosis and thrombocytosis because, in the course of PV – unlike secondary polycythemia – there is proliferation of all myelopoietic lines and not only of the erythroid line [2]. Patients with PV often have a low serum erythropoietin concentration [3]. One of the most important issues in the care of patients with PV is the prevention of cardiovascular events [4].

Chronic lymphocytic leukemia (CLL) is a lymphoproliferative neoplasm characterized by the chronic presence in the peripheral blood of a population of clonal B lymphocytes with a typical immunophenotype (coexistence of CD5, CD19, CD20, and CD23) in numbers exceeding 5,000 cells/μL, often with concomitant infiltration of the lymph nodes, spleen, bone marrow, and other tissues [5]. The incidence of the disease increases with age (median age of onset is 72 years) [6]. CLL is most often diagnosed incidentally, as a result of observing increased lymphocytosis in a routine complete blood count or for other indications. CLL usually follows a slow clinical course, often remaining asymptomatic for years, although the disease can also be aggressive from the outset [7].

The purpose of this paper was to present the case report of a 67-year-old man diagnosed with PV and coexisting CLL.

CASE REPORT

Anamnesis and physical examination

A 67-year-old patient was admitted to the Ward of Hematology for a planned evaluation for PV. The patient had a history of night sweats for approximately 5 years, with less severe episodes in the last 2 months, and had lost approximately 6 kg of body weight over a 4-month period. He reported no other symptoms. He denied any tendency toward infection, fever or low-grade fever, weakness, tendency to bruise, or bleeding symptoms. Due to the presence of polycythemia, the patient had recently undergone several phlebotomies. Before admission to the hospital, the attending hematologist initiated cytoreductive therapy with hydroxycarbamide, after which the patient reported feeling better. Acetylsalicylic acid was also administered. Until that point, the patient had been treated chronically for hypertension and gout (bisoprolol, amlodipine, perindopril, and allopurinol).

Physical examination revealed lower limb edema accompanied by varicose veins and trophic changes. Other than that, no significant abnormalities were found, including obvious organomegaly, lymphadenopathy in the lymph nodes visible on physical examination, or cutaneous and mucosal signs of bleeding diathesis.

Complete blood count

The peripheral blood morphology parameters confirmed the features of polycythemia (red blood cell mass: 7.19×10^6/μL; hemoglobin concentration: 17.2 g/dL; hematocrit: 55.6%). Features of microcytosis (medium cell volume: 77.3 fL) and hypochromic blood cells were observed (mean cell hemoglobin: 23.9 pg; mean cellular hemoglobin concentration: 30.9 g/dL). Leukocytosis (16.49×10^3/μL) with a predominance of lymphocytes (10.80×10^3/μL) was also noted. The number of thrombocytes was normal (188×10^3/μL) and they were characterized by a slightly reduced volume (medium platelet volume: 9.1 fL). When interpreting the results of the complete blood count, the cytoreductive treatment already in use with hydroxyurea should be taken into account.

Microscopic evaluation of the peripheral blood smear confirmed a predominance of lymphocytes in the leukogram (approximately 71%). Most lymphocytes were characterized by a sparse cytoplasmic layer and condensed chromatin within the cell nucleus. Figure 1 shows the microscope images of a peripheral blood smear.

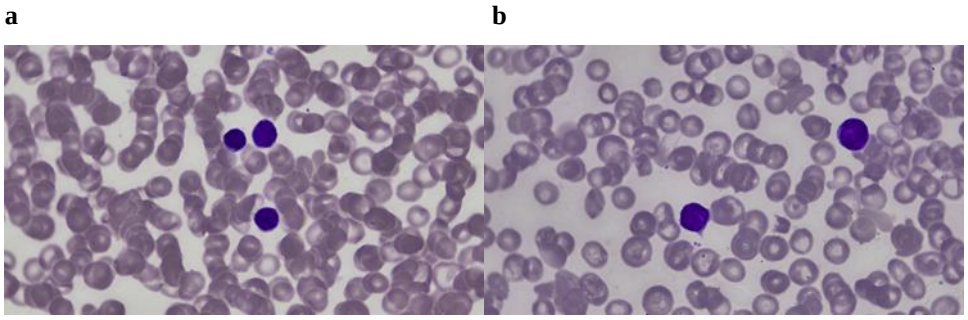

Fig. 1a and b. Lymphocytes in a microscope image of a peripheral blood smear

Other laboratory tests

Importantly, the serum erythropoietin level was very low (<1.0 mIU/mL). No significant abnormalities were found in the proteinogram parameters or in the concentration of immunoglobulins of the IgG, IgM, and IgA classes. Lactate dehydrogenase (LDH) activity and serum β2-microglobulin concentrations were normal. Serum calcium and phosphorus concentrations were normal. Ferritin and vitamin B12 levels were within normal range.

Features of impaired fasting glucose were found (112 mg/dL). The mild hyperbilirubinemia (1.62 mg/dL) and elevated gamma-glutamyl transferase activity (105 U/L) were noteworthy, with normal aminotransferase activity, alkaline phosphatase, and prothrombin time. A slightly elevated C-reactive protein level was found (7.2 mg/L). Serum creatinine, uric acid, and urea levels were normal. Sodium and potassium levels were within normal limits. The thyrotropin level and lipid profile parameters were within normal limits. Virological screening tests were negative.

Diagnostic imaging

To further assess the lymphadenopathy, an ultrasound examination of the peripheral lymph nodes was performed. In the groin area, lymph nodes with a maximum size of 14.3 × 9.0 mm were found, with a tendency to form packets with a maximum size of 27.1 × 10.3 mm. In the axillae, lymph nodes with a maximum size of 12.5 × 9.3 mm were found, with a tendency to form packets with a maximum size of 33.4 × 14.8 mm. The abdominal ultrasound examination revealed periaortic lymph nodes with a maximum size of 9 × 6 mm.

The chest radiograph was not repeated (the patient presented a recent scan, the results of which were within normal limits).

Specialized hematological diagnostics

Due to the suspicion of PV, a test for the presence of the V617F mutation of the *JAK2* gene in peripheral blood cells was performed; the result was positive.

A trephine biopsy was performed. The results of the histopathological examination, including immunohistochemical staining, confirmed the features of PV. Furthermore, the lymphocytic infiltrate was found to have an immunophenotype typical of CLL/small cell lymphoma (SLL).

Lymphocytes in the cytological image of the bone marrow presented a similar morphology to peripheral blood lymphocytes. Figure 2 shows microscope images of a bone marrow smear.

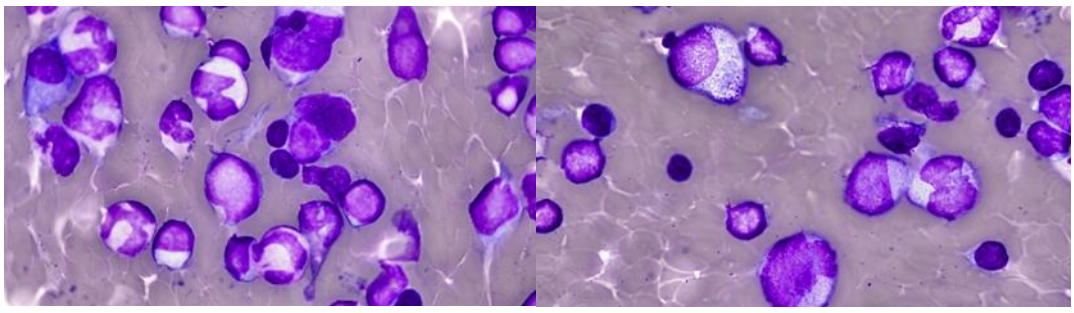

Fig. 2a and b. Lymphocytes in a microscope image of a bone marrow smear

Immunophenotypic analysis of peripheral blood cells was performed, revealing the presence of a clonal population of CD19+, CD5+ B cells (approximately 35.6%) with the following dominant phenotype: CD20+, CD200+, CD22+, CD23+, CD43+, CD79b+dim, CD10-, CD11c-, FMC-7-, SmIg-, sKappa+dim, sLambda-.

Following the diagnosis of CLL, testing for unfavorable prognostic cytogenetic and molecular factors was performed. Fluorescence in situ

hybridization (FISH) excluded the presence of a 17p deletion and an 11q deletion. An unmutated segment within the *IGHV* gene was identified.

Cardiovascular system assessment

Due to the diagnosis of chronic myeloproliferative neoplasm, the patient's diagnostic workup was also extended to include an assessment of his cardiovascular system. Transthoracic echocardiography revealed dilatation of the left ventricle (end diastolic diameter: 68 mm; end systolic diameter: 49 mm) and the left atrium (left atrium volume index: 43.1 mL/m^2). Systolic function of both ventricles was normal. No significant valvular abnormalities were identified.
Ultrasound evaluation revealed atherosclerotic plaques in the aorta and carotid arteries, without hemodynamically significant stenoses.

Treatment

Considering the overall clinical picture and the additional tests performed, the patient was kept on cytoreductive therapy with hydroxyurea for the PV, under the continued supervision of a hematologist at the outpatient clinic. Currently, there is no indication to begin CLL treatment – only the need to observe the patient.

DISCUSSION

Cases of coexisting CLL and PV have already been described in the literature, but this phenomenon is rarely encountered in clinical practice, which is why the current state of knowledge is based mainly on case reports. Li and Zhang [8] recently reported a case report of a 35-year-old man with concomitant PV and CLL. It is worth noting that, unlike in that case, our patient is an older man and that the incidence of PV and CLL increases with age. The patient presented in that publication also had a much higher leukocytosis (39.8 × 10^9/L), while in contrast to our case, lymphocytes constituted a smaller percentage of white blood cells (38%). Korkmaz et al. [9] presented the case of a 56-year-old man diagnosed with PV with the V617F *JAK2* mutation and additionally diagnosed with CLL. As in our case, the patient was treated with hydroxyurea and monitored for CLL progression. Carulli et al. [10] described a case of a patient diagnosed with CLL 15 months after a diagnosis of PV, who had been treated only with phlebotomy. Interestingly, cytogenetic analysis performed after the diagnosis of CLL revealed an atypical karyotype change (18p+).
Literature data show that the clinical course of PV in the presence of CLL may also be characterized by progression. A group of researchers from Poland described a case of PV transforming into acute myeloid leukemia (AML) after 4 years of the disease in a 67-year-old patient diagnosed with PV and concomitant CLL [11]. Another team of researchers described post-PV myelofibrosis with myelodysplastic-like progression in a patient who was also diagnosed with CLL [12].
The molecular basis explaining the simultaneous development of PV and CLL remains unclear. Tian et al. [13] presented a case report in which the V617F mutation of the *JAK2* gene and the *P53* gene mutation occurring at two sites were identified using next-generation sequencing. Chu et al. [14] published a case report identifying a new isolate (BIS8-17) of torque tenovirus (TTV) in a patient with concomitant CLL and PV. However, CLL cells do not harbor BIS8-17. Previous research suggests that the phenomena responsible for the development of PV and CLL are independent. According to the results obtained by Swierczek et al. [15] on material from three women with coexisting PV and CLL, X-chromosome inactivation patterns of the neoplastic cells revealed that granulocytes/platelets and B-lymphocytes used different X-chromosome alleles.

CONCLUSIONS

The case report presented in this article demonstrates that PV can coexist with CLL. This is important to keep in mind when planning diagnostic workup in patients with suspected hematologic malignancies. It remains unclear whether the simultaneous development of these two diseases is independent and there is no causal relationship between them, or whether there are some pathogenetic phenomena that predispose a patient to the development of both diseases.

Authors' contribution

Study design – G.K. Jakubiak, J. Pruszyński, E. Ziółko
Manuscript preparation – G.K. Jakubiak, E. Ziółko
Literature research – G.K. Jakubiak
Final approval of the version to be published – G.K. Jakubiak, J. Pruszyński, E. Ziółko

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