International Case Reports Journal
Case Report (ISSN: 2770-9647)

Polycythaemia Vera Spent phase with Highest TLC, Blast Crisis Following Mycoplasma Infection and Autoimmune Haemolysis: A Rare Case Report

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Abstract

Polycythemia Vera (PV), a chronic myeloproliferative neoplasm, may evolve into myelofibrosis and may rarely transform into acute leukemia after a prolonged course of 10-15 years or even longer. Early blast crisis is uncommon, and its precipitation by an unusual infection with overlapping features of autoimmunity is extremely rare. In addition, it is unusual for PV to enter spent phase very early after diagnosis and having highest ever reported total leukocyte count of 2.57 lakhs/μL (257 × 10^9/L) during spent phase, which on peripheral smear was reported erroneously as CML chronic phase. We had to do BCR- ABL and a repeat JAK 2 mutation analysis to differentiate between myelofibrosis and CML. We have noticed autoimmune disorders to coexist with myeloproliferative disorders but in this case, after a mycoplasma pneumonia infection, there was a heightened autoimmune manifestations and blast transformation of PV. There was DCT positive autoimmune hemolysis with very low platelet count and falling Hb and the peripheral blood did not show blasts, but the bone marrow only confirmed leukemic transformation.

Introduction

Polycythemia Vera (PV) is a chronic myeloproliferative neoplasm characterized by JAK2 mutation, erythrocytosis, leukocytosis, and thrombocytosis. It may evolve into post-polycythemia myelofibrosis or acute leukemia, the latter occurring in 5%-10% of cases after 10-15 years, usually happening in elderly patients or after exposure to cytotoxic therapy. Transformation within a short course of the disease is rare. Mycoplasma pneumoniae infection can induce autoimmune manifestations, including Autoimmune Hemolytic Anemia (AIHA), through molecular mimicry and immune activation. The overlap of infection, autoimmunity, myeloproliferative disorder and malignant progression poses a diagnostic challenge.

Citation: *Azmil K, Sasidharan PK. Polycythaemia Vera Spent phase with Highest TLC, Blast Crisis Following Mycoplasma Infection and Autoimmune Haemolysis: A Rare Case Report. Int Case Rep Jour. 2026;5(1):1-5.*

Received Date: 29 December, 2025; **Accepted Date:** 10 January, 2026; **Published Date:** 13 January, 2026

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We report a rare case of PV initially presenting as essential thrombocythemia, then progressing into spent phase in a short period with highest total leucocyte count ever reported and subsequently undergoing early blast transformation following Mycoplasma infection with Autoimmune Hemolytic Anemia (AIHA).

Case History

A 66-year-old male, chronic smoker, first presented in 2019 with a non-healing ulcer of the great toe. Laboratory evaluation revealed severe anemia (Hb 5.7 g/dl), mild leukocytosis (12,600/μL), and marked thrombocytosis 10.03 lakhs/μL (103 × 10^9/L). Secondary causes of thrombocytosis were excluded after clinical and laboratory evaluation, and a diagnosis of essential thrombocythemia was made. Peripheral smear and bone marrow trephine did not show any abnormalities then. Autoimmune markers were negative at this presentation. He was started on hydroxyurea and aspirin. The Hb improved, platelet count remained at around 5-6 lakhs/μL (560 × 10^9/L), total leukocyte count remained at 20 × 10^3/μL - 30 × 10^3/μL range, he was asymptomatic, active and doing well for three years. On follow-up, within a period of three years, he demonstrated rising leukocyte counts went up to 2.58 lakhs/μL (258/10^9/L) with moderate splenomegaly in 2022. Peripheral smear showed myelocytes and metamyelocytes, since such high counts are usually not seen in myelofibrosis, provisional diagnosis of CML was considered. However, BCR-ABL was negative and JAK2 V617F mutation was positive, confirming evolution of polycythemia Vera into spent phase. Bone marrow confirmed myelofibrosis too. He remained asymptomatic for three years on hydroxyurea, with TLC between 20-3000/μL again; another curious phenomenon observed was he never required venesection. In June 2025, he presented with dry cough, low-grade fever, hemoptysis, and purpura after a holiday trip to a hill station. On examination, he had ecchymoses and wet purpura but stable vitals. CBC showed Hb 7.1 g/dl, TLC 12,030/μL, DLC N64L29 and platelets 9,000/μL. Direct Coombs test was positive, and ANA profile showed Ro-52 antibody positivity. Chest X-ray suggested atypical pneumonia; Mycoplasma IgM was positive. His symptoms responded promptly to doxycycline. But the pancytopenia never recovered even after high doses of dexamethasone. That made us suspect blast transformation but peripheral smear was inconclusive, but bone marrow showed acute leukemia.

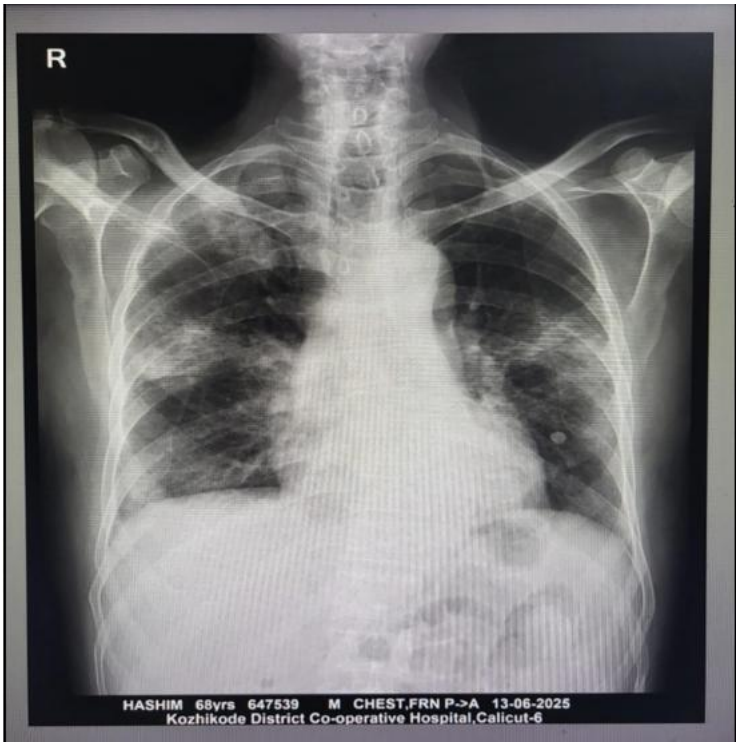

Figure 1: Chest x-ray showing atypical pneumonia.

Table 1: Serial Complete Blood Count (CBC) during hospitalization

Date	Hb (g/dL)	TLC (/mm³)	Platelets (/mm³)	Notes
10-06	7.3	12030	9000	DCT positive
11-06	7.1	9350	3000	Mycoplasma IgM positive
12-06	7.8	11870	8000	Steroids started
13-06	6.9	7530	3000	
14-06	6.1	7020	3000	
15-06	6.4	6240	5000	
16-06	6.9	7550	2000	Platelet transfusion given
17-06	6.6	10210	6000	Persistent cytopenias

Investigations

- Direct Coombs Test: Positive
- ANA profile: Ro-52 positive, DFS70 positive, others negative
- Mycoplasma IgM: Positive (11.89, normal <9 U/mL)
- CRP: 108 mg/L
- D-dimer: 1.75 µg/mL FEU
- Electrolytes: Na 129 mmol/L, 4.39 mmol/L
- LFT: Bilirubin (Total/Direct)1/0.34 mg/dL, AST/ALT 41/37 U/L, ALP 164 U/L, Albumin 2.84 g/dL
- Coagulation: PT/INR 1.29, APTT 32.6s
- Peripheral smear: Normocytic normochromic anemia with occasional monoblasts, mild neutrophilic leukocytosis, severe thrombocytopenia
- Bone marrow biopsy: Acute leukemia, consistent with AML (blast crisis of PV)

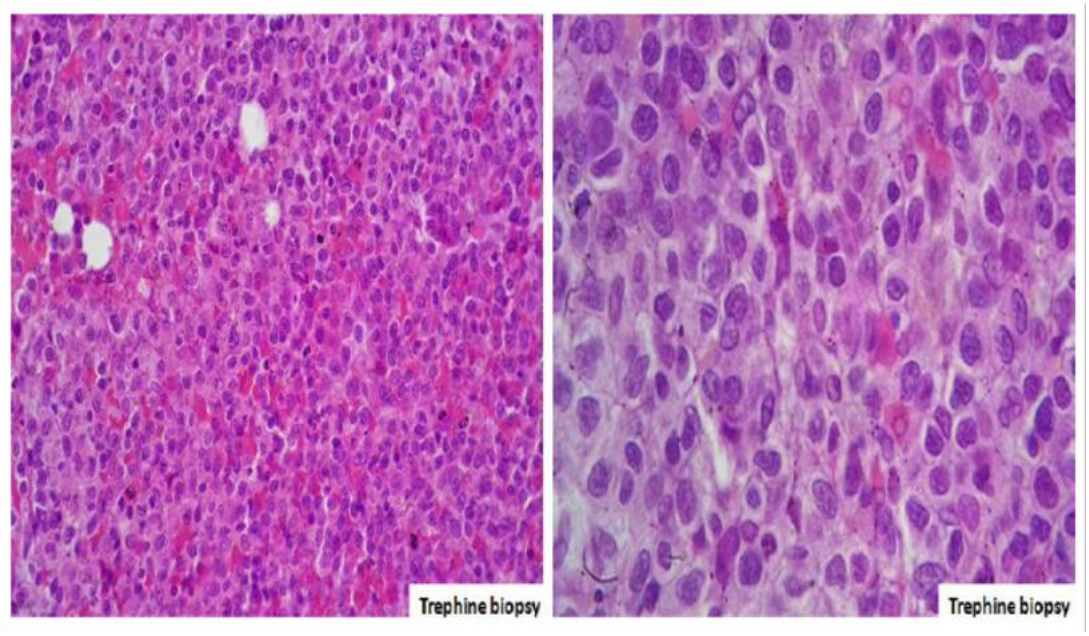

Figure 2: Bone marrow showing acute leukaemia.

Discussion

Leukemic transformation occurs only in a minority of PV patients, typically after long disease duration and in older age groups. Risk factors include advanced age, cytotoxic therapy, and high leukocyte counts. Transformation is associated with poor prognosis, with median survival less than six months. This case is unique for several reasons.

1. The disease course began with JAK 2 positive Essential Thrombocythemia-like phenotype, progressed into spent phase very early and culminated in AML transformation within a period of six years
2. The trigger for leukemic transformation was a Mycoplasma pneumonia infection, complicated by autoimmune hemolysis, which initially masked the underlying leukemic transformation.
3. Diagnostic confusion occurred with CML at one stage and autoimmune disease at another, highlighting the overlap of MPNs, infection, and immune activation.
4. Highest ever reported TLC in spent phase of polycythemia - 2.57 Lakhs/μL

Few case reports exist of infection-associated leukemic transformation in PV, making this report a valuable addition to the literature.

Conclusion

This case illustrates an atypical and accelerated progression of PV, beginning as ET, entering spent phase, and transforming into AML, possibly precipitated by infection and autoimmune activation. Clinicians should maintain vigilance for blast crisis when cytopenia persist despite infection control and immunosuppression in patients with underlying MPNs. Positive autoimmune markers suggest that a possibility of autoimmune mechanism for the diseases initiation and progression. Ruxolitinib (Jakavi) could not be used due to financial constraints.

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