Clinical Dilemma: "Atypical Presentation of Acute Coronary Syndrome in a Patient with Polycythemia Vera"

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ABSTRACT:

A 29-year-old male presented to the emergency department with acute chest pain, elevated blood pressure, and mild dyspnea. Initial laboratory investigations revealed significant abnormalities, including leukocytosis, markedly elevated hemoglobin (20.5 g/dL), and increased hematocrit (59.9%). Cardiac biomarkers showed a substantial rise in troponin-I levels, peaking at 16,176 ng/L. Electrocardiogram demonstrated T-wave inversion in anterolateral leads, suggestive of myocardial ischemia or NSTEMI. Further investigation, including bone marrow biopsy and genetic testing, confirmed a diagnosis of polycythemia vera (PV) with a positive JAK2V617F mutation. Treatment was initiated with hydroxyurea and interferon to manage PV, along with ticagrelor for thrombosis prevention. This case highlights the importance of considering hematological disorders such as PV in young patients presenting with atypical cardiovascular events, emphasizing the need for evaluation in such cases.

INTRODUCTION:

Polycythemia vera (PV) is a myeloproliferative neoplasm characterized by increased red blood cell production resulting in increased blood viscosity. This condition is known to increase the risk of thrombotic events, including acute coronary syndrome (ACS), more commonly in the elderly (>60 years) and less commonly in young populations [1]. This case report presents an unusual presentation of a patient with PV who developed ACS with atypical laboratory findings. This case report will discuss the potential association between PV and ACS.

CASE:

A 29-year-old man presented to the emergency department complaining of chest pain that had persisted for the past 3 hours. He described the pain as sharp, constant, non-radiating, predominantly localized to the left side of his chest, and accompanied by mild shortness of breath and nausea. The patient denied any prior history of cardiovascular issues but had quit smoking 8 years ago and occasionally consumed alcohol. Upon examination, the patient appeared awake and comfortable, with a regular pulse of 62 beats per minute, blood pressure of 183/125 mmHg, respiratory rate of 17 bpm, and oxygen saturation of 93% on room air. Systemic examination revealed no abnormalities, normal peripheral pulses, and no signs of elevated jugular venous distention, ankle, or sacral edema. Upon admission, comprehensive laboratory investigations showed significant hematological findings. His white blood cell count was elevated at 11.4 × 10^3/uL (normal range: 4.0–10 × 10^3/uL), indicative of neutrophilic leukocytosis. Hemoglobin levels were elevated at 20.5 g/dL (normal: 12–16 g/dL), along with an increased hematocrit of 59.9% (normal: 40–54%). Mean Corpuscular

Volume was measured at 87.8 fL (normal: 80–100 fL), with Red Blood Cells numbering 6.55 × 10^6/uL (normal: 4.0–5.5 × 10^6/uL), and platelets at 283 × 10^3/uL (normal: 100–300 × 10^3/uL). His highly sensitive troponin-I level upon arrival was markedly elevated at 2085 ng/L (typical upper limit: 29.9 ng/L) and increased to 5453 ng/L; the next day, the troponin-I level increased further to 16,176 ng/L within a few hours. Despite these hematological and cardiac abnormalities, his serum electrolytes were within normal limits: Sodium (Na): 137 mmol/L (normal range: 135–145 mmol/L), Potassium (K): 4.6 mmol/L (normal range: 3.5–5.0 mmol/L), Chloride (Cl): 97 mmol/L (normal range: 98–106 mmol/L), and creatinine: 0.97 mg/dL (normal range: 0.7–1.3 mg/dL). The lipid profile showed Total Cholesterol at 203 mg/dL (normal: <200 mg/dL), Triglycerides at 110 mg/dL (normal: <150 mg/dL), Low-Density Lipoprotein Cholesterol at 145 mg/dL (normal: <100 mg/dL), and High-Density Lipoprotein Cholesterol at 36 mg/dL (normal: 40–60 mg/dL). An electrocardiogram at admission revealed T-wave inversion in the anterolateral leads. CT scan of the chest did not reveal any acute findings. The patient's elevated hematocrit and hemoglobin levels, along with findings of bone marrow biopsy showing proliferation of all three blood cell lineages, strongly suggested a diagnosis of polycythemia vera (PV). Confirmation came through genetic testing which revealed a positive JAK2V617F mutation. The patient initially treated with Hydralazine 20 mg IV bolus to control blood pressure, received intravenous hydration for elevated hemoglobin levels, and was closely monitored. After bone marrow biopsy and genetic testing confirmation, the patient received hydroxyurea (500 mg twice daily) and interferon (30 μg once daily) with gradual tapering dose with reduction in hemoglobin value. Additionally, due to ongoing thrombosis prevention needs, the patient was prescribed ticagrelor every 12 hours. Following three months of this regimen, the patient's blood counts normalized, indicating a positive response to treatment.

DISCUSSION:

Polycythemia Vera (PV) is classified as a chronic myeloproliferative neoplasm characterized by clonal proliferation of pluripotent hematopoietic stem cells. This aberrant proliferation primarily manifests as erythrocytosis, an abnormal increase in red blood cells. Concurrently, there is an absolute rise in other hematopoietic cell lineages, resulting in leukocytosis and thrombocytosis. PV often presents with splenomegaly due to enhanced hematopoiesis and may progress to involve bone marrow fibrosis, characterized by excessive deposition of fibrous tissue within the marrow space [2]. These pathological changes can lead to clinical complications such as thromboembolic events, hemorrhage, and rarely, transformation to myelofibrosis or acute leukemia. As for the treatment, this case presents a scenario of acute

coronary syndrome (ACS) in a young patient with previously undiagnosed polycythemia vera (PV). Cytoreductive therapy is a cornerstone in PV management [3]. In this case, the initial management with hydralazine for blood pressure control and intravenous hydration for elevated hemoglobin levels was appropriate for the acute presentation. The combination of hydroxyurea and interferon was appropriately initiated; the dose of hydroxyurea was gradually reduced with improvement in hemoglobin but also resulted in decreased WBC and platelet count, as seen in figure 3. Hydroxyurea effectively reduces the risk of thrombotic events in PV patients, while interferon can induce hematological remission and potentially modify the disease course [3]. While the European Collaboration on Low-Dose Aspirin in Polycythemia Vera (ECLAP) study demonstrated a significant reduction in cardiovascular events with low-dose aspirin in PV patients [4,5], the use of ticagrelor in this case aligns with current guidelines for ACS management. Ticagrelor offers more potent platelet inhibition compared to aspirin, which may be particularly beneficial in the setting of both ACS and PV. However, the long-term use of ticagrelor in PV patients with ACS lacks extensive study and may require careful monitoring. In this case, given the presentation of ACS, the addition of a DOAC could be considered in the future, especially if there are recurrent thrombotic events despite anti-platelet therapy [6]. The initial management with hydralazine for blood pressure control and intravenous hydration for elevated hemoglobin levels was appropriate for the acute presentation. However, long-term management of hypertension in PV patients requires careful consideration, as both the disease and its treatments can impact cardiovascular risk factors. This case highlights the importance of considering myeloproliferative neoplasms in young patients presenting with ACS, especially when accompanied by abnormal troponin-I findings. The diagnosis of PV through bone marrow biopsy and JAK2V617F mutation testing was crucial in guiding appropriate management [7]. Moving forward, regular monitoring of blood counts, JAK2V617F allele burden, and cardiovascular risk factors will be essential [8]. The management plan may need adjustments based on the patient's response and any complications that may arise. There is no established treatment protocol for PV patients presenting with ACS. Future research should focus on optimizing combination therapies for such cases, including the potential role of newer agents like JAK inhibitors in managing both the hematological and cardiovascular aspects of PV. In conclusion, the initial outcome was favorable; however, long-term management remains challenging and requires a personalized approach balancing thrombosis and bleeding risks while addressing the underlying myeloproliferative disorder.

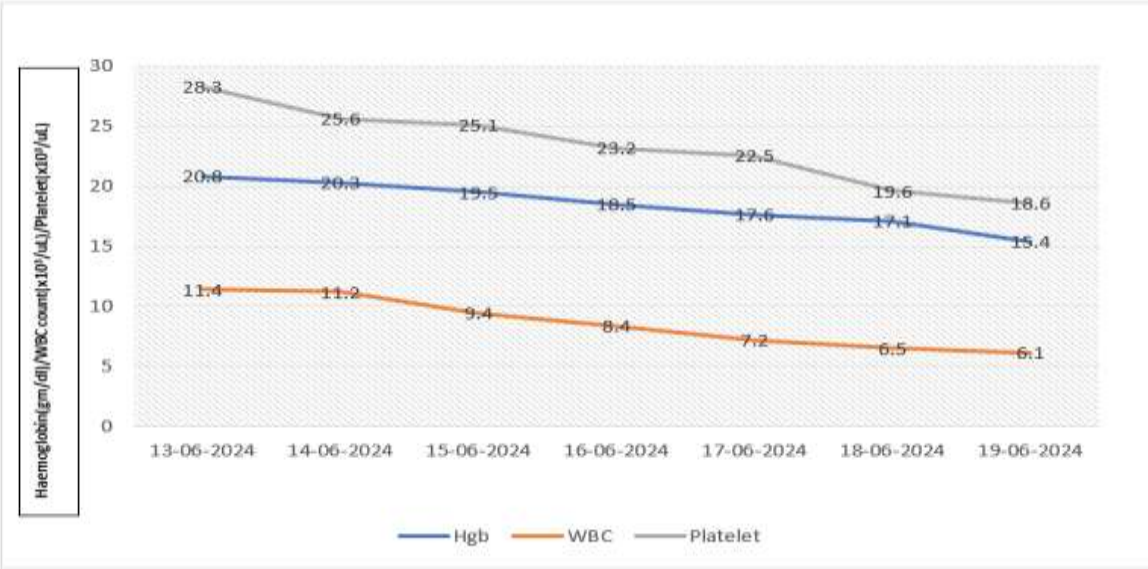

Fig: 3 Trends of blood count after initiation of treatment. Patient was started on hydroxyurea immediately after admission, and the dose was adjusted according to response. Note that hemoglobin, platelet and white blood cell counts drastically reduces after treatment

CONCLUSION:

This case illustrates the relationship between polycythemia vera (PV) and acute coronary syndrome in a young patient. The initial cardiac symptoms and abnormal test results led to a broader investigation, ultimately revealing PV as the underlying condition. This case also highlights the importance of considering hematological disorders in atypical cardiovascular presentations, especially in younger patients. The successful treatment outcome emphasizes the value of accurate diagnosis and targeted therapy in managing PV and its associated cardiovascular risks.

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