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A Rare Cause For Acute Myocardial Infarction

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Abstract

Acute myocardial infarction (AMI) is usually seen in the setting of atherosclerosis and its associated risk factors. Myocardial infarction in the young poses a particular challenge, as the disease is less likely, due to atherosclerosis. We report a case of 35-year-old male patient who presented with ST segment elevation anterolateral MI. The only abnormality on routine blood investigation was raised hemoglobin and hematocrit. After further testing, he was diagnosed as polycythemia vera. This case illustrates the importance of recognizing polycythemia vera as an important cause of thrombosis, which can present initially as AMI, and to emphasize the early recognition of the disease in order to initiate appropriate management strategies.

Keywords: Polycythemia rubra vera, Myocardial infarction

Introduction

Polycythemia rubra vera (PV) is a chronic myeloproliferative disorder characterized by elevated red cell mass with an increased risk of both thrombosis and bleeding. Thromboembolic events are the most significant and life-threatening complications associated with PV [1].Here, we report a rare case of PV presenting as acute myocardial infarction (MI) in a young man and briefly discuss the applicability of current management strategies for MI in the context of Polycythemia rubra vera.

Case Report

A 35 years old male, painter by occupation came with complaints of acute onset anginal chest pain and ECG showed evidence of acute myocardial infarction. He was a non smoker and non alcoholic. He had no previous history of hypertension, diabetes mellitus, drug abuse, hyperlipidemia. He did not have family history of cardiac disease. On physical examination, he looked plethoric with conjunctival congestion. His temperature was normal, pulse 90/min (regular), blood pressure (BP) 110/70 mmHg, respiratory rate 18/min and saturation in room air 99%. Abdominal examination revealed moderate splenomegaly. The peripheral pulses were equally felt. The ECG on admission (Fig. 1) showed ST segment-elevation in anterolateral leads (V2–V6 and I-AVL), so the impression was acute ST segment elevation myocardial infarction. Hence he was thrombolysed after ruling out contraindications.

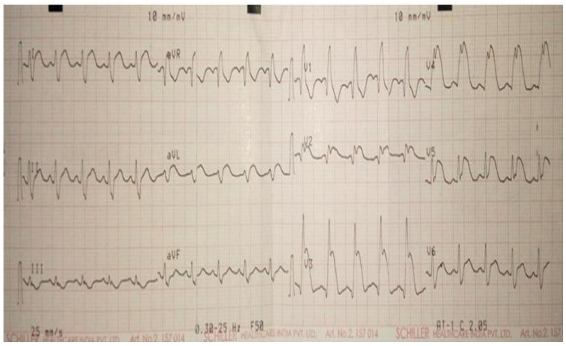

Fig.1:ECG showing ST elevation in V2 to V6, LEAD I and aVL

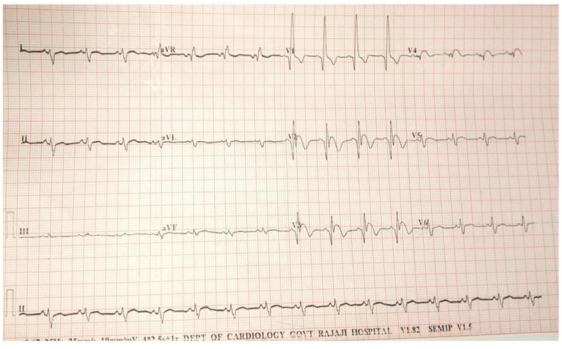

Fig 2: ECG after thrombolysis; showing HR -100/min, qRBBB and T wave inversion(v2-v4)

Then he was started on injection heparin 5000 U IV QID, T.Aspirin 150mg 1 OD, T.Clopidogrel 75mg 1 OD, T.Atorvastatin 10mg 8 HS, T.Enalapril 2.5mg 1 BD, T.Carvedilol 3.125mg 1 BD, T.Alprazolam 0.5mg HS.(Fig 2)

Laboratory investigations on admission were as follows

Complete blood count: Hemoglobin : 21.8 gm/dl , RBC count: 9.8 million cells, PCV : 63%, Platelets : 7.27 lakhs, MCV :62 pg, TC: 35,000 , DC : Neutrophils 85%, Lymphocytes 8%, Monocytes7% . His peripheral smear showed polymorphonuclear leukocytosis with reactive increase in platelets and microcytic erythrocytosis. His coagulation profile, renal function test, liver function test, and electrolytes were all within normal ranges. His fasting lipid profile was as follows: Serum cholesterol : 170 Mg/dl , TGL : 200 mgs/dl, S.HDL : 39 mg/dl, S.VLDL : 45 mg/dl, S. LDL : 86 mg/dl. Viral markers were negative.

In pursuit of the cause for his polycythemia, further tests were done. The arterial blood gas showed no hypoxia, the erythropoietin level was 4 mU/ml (4.30–29 mU/ml), and the JAK-2 V617F mutation was positive. Cytogenetic study for BCR/ABL1 rearrangement was negative. The tests for ANA, lupus anticoagulant, anticardiolipin antibody and anti beta2 glycoprotein were negative.

The abdominal ultrasound showed moderate splenomegaly with splenic vein thrombosis. CT Abdomen confirmed moderate splenomegaly with splenic infarct and perigastric splenolateral collaterals. OGD revealed diffuse fundal varices. The patient's echocardiography showed ejection fraction of 40% with hypokinesia of anterior wall and no intracardiac thrombus. The bone marrow aspirate and biopsy showed increase in trilineage hematopoiesis with no atypical cells. These findings were consistent with myeloproliferative disorder. Based on the clinical picture and laboratory results, the patient was diagnosed with polycythemia vera (PV) fulfilling the WHO diagnostic criteria (Table 1), and started on hydroxyurea 15 mg/kg, and scheduled for regular phlebotomy to keep hematocrit less than 45% .Four weeks later, the patient underwent Coronary angiogram which showed recanalised LAD.(Fig 3)

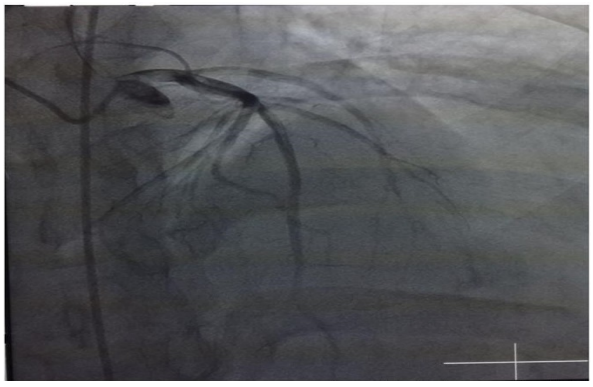

Fig 3: CAG showing recanalised lad

WHO 2016 Diagnostic Criteria for PV

Stratification of Criteria	Clinical and Laboratory Features
Major criteria	1. HGB >16.5 g/dL (men), >16.0 g/dL (women) ***OR*** HCT >49% (men), >48% (women) ***OR*** increased red cell mass 2. BM biopsy showing hypercellularity for age with trilineage growth, including prominent erythroid, granulocytic, and megakaryocytic proliferation with pleomorphic, mature megakaryocytes 3. Presence of *JAK2 V617F* or *JAK2* exon 12 mutation
Minor criterion	• Subnormal serum EPO level

• Patients must meet either all 3 major criteria ***or*** the first 2 major criteria and the minor criterion

Table 1 : WHO 2016 DIAGNOSTIC CRITERIA FOR POLYCYTHEMIA VERA

Discussion

Polycythemia Vera (PV) is a chronic myeloproliferative disorder, involving a multipotent hematopoietic progenitor cell, which causes in general, an increased production of red cells, granulocytes and platelets, but most significantly in erythrocytes, which leads to hyperviscosity and an increased risk of thrombosis. The median age of diagnosis is 60 years, though the disorder can occur in all age groups . It can be discovered incidentally after routine blood investigation or may present with signs and symptoms related mainly to thrombosis and/or hemorrhage.

Bleeding and thrombotic(venous and arterial) complications are the major causes of morbidity and mortality in PV, occurring in 40 to 60% of the patients [1,2].The pathophysiology of thromboembolic events in polycythemia Vera has not been elucidated, but many factors are involved: increases in hematocrit and blood hyperviscosity, stimulation of platelet aggregation and thrombogenesis, the presence of leukocytosis, rigidity of the membrane and intimal proliferation [3,4].Advanced age and a prior history of thrombosis are the two most important risk factors for vascular complications.Hypercholesterolemia, hypertension, smoking, and diabetes have been recognized as predictors of thrombosis [5].

Myocardial infarction and sudden death are complications of newly diagnosed or untreated PV; they occur most often in elderly people (≥ 65 years) with underlying coronary artery disease [6]. However, younger patients with PV who are free from coronary artery disease can also be affected, and sometimes the outcome is death . Benita reported a 30-year-old male patient who died due to myocardial infarction as initial manifestation of PV. Rossi et al. followed 149 patient diagnosed with PV for 10 years and found that 11.4% had myocardial infarction .Despite the association of PV with coronary artery disease, the presentation of PV as AMI is very rare

Cytoreductive treatment of blood hyperviscosity by phlebotomy or chemotherapy and antiplatelet therapy with low-dose aspirin have dramatically reduced the number of thrombotic

complications and substantially improved survival [7]. The major goal of treatment is to prevent thrombotic events. Our patient was scheduled for regular phlebotomies to keep hematocrit less than 45%. In addition, he was commenced on hydroxyurea 15 mg/kg and continued other drugs except clopidogrel and statins. Our patient showed dramatic response with hydroxyurea on follow up. The use of angioplasty and stenting is challenged by the development of stent thrombosis. Hydroxyurea is an important component of the treatment of such patients.

Evidence for the prevention and treatment of specific cardiovascular complications in PV is too scarce.There is no standard consensus on management of acute MI in patients with PV. In one study, standard anti platelet therapy combined with recurrent phlebotomy reduced the risk of re-infarction by 70% in patients with acute coronary syndrome [6].Few studies suggested the use of anticoagulants such as warfarin along with antiplatelet agents will help prevent recurrent thrombosis .However, the effectiveness of clopidogrel, glycoprotein IIb-IIIa inhibitors and intervention with coronary stents or CABG has not been rigorously studied in patients with PV. In general, the treatment of acute coronary syndromes secondary to chronic myeloproliferative disorders require special attention in maintaining the delicate balance between the risk of bleeding and clotting tendency.

Conclusion

There is no doubt that MI accounts for a substantial percentage of morbidity and mortality in patients with PV. Although relatively rare, AMI can be the initial manifestation of myeloproliferative disease. Attending physicians should keep a high index of suspicion for PV, especially when a young person presents with myocardial infarction and raised hematocrit in the absence of atherosclerotic risk factors, as initiation of early management will alter patient prognosis. Nevertheless, the search for new strategies for management the cardiovascular events in PV patients has undoubtedly become a priority for future research. The management of patients with PV presenting with acute MI is complex. It is important to consider adequate use of myelosuppressive agents such as hydroxyurea or ruxolitinib in high risk patients to prevent future thrombotic events.

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