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Acute ST-Segment Elevation Myocardial Infarction during Stress Testing: First Presentation of an Undiagnosed Case of Essential Thrombocytemia

Running Title: Essential Thrombocytemia complication

Juan Carrillo Navas, MD, Jonathan Rodríguez, MD, Edgardo I Santiago Guzman, MD, Angel López-Candales, MD, FACC, FASE

Cardiovascular Medicine Division, University of Puerto Rico School of Medicine, San Juan, Puerto Rico

Abstract: - Essential thrombocythemia (ET), a myeloid stem cell clonal disorder, can clinically present with variable manifestations including vasomotor symptoms, hemorrhage or thrombotic events. Acute coronary syndromes specifically ST-segment elevation myocardial infarction (STEMI) is an uncommon event in ET patients. We report the case of a 43-year-old male presenting with a STEMI while performing a routine exercise stress test requested for atypical chest pain evaluation. Urgent coronary angiography demonstrated total occlusion of proximal right coronary artery that was successfully treated with percutaneous coronary angioplasty and stent deployment without adverse events or complications.

Key words: - Acute myocardial infarction, coronary angiography, essential thrombocytemia, platelet disorders, thrombosis, stress testing.

Introduction:

Essential thrombocythemia (ET) is a myeloproliferative disorder clinically characterized by an increased platelet count, megakaryocytic hyperplasia, and a hemorrhagic or thrombotic tendency [1].

Although published epidemiology data is scarce, overall prevalence is low with approximately 2.5 per 1 million with an approximate mean age of presentation of 60 years or older [2, 3]. In the United States, ET is more commonly diagnosed among Blacks when compared to non-Hispanic Whites. In sharp contrast, ET is less commonly seen in both Hispanic Whites and Asian Pacific Islanders [4].

ET patients carry a JAK2 (V617F), an MPL substitution, or a calreticulin gene (CALR) mutation that determines both its biological and clinical manifestations [5].

Although coronary artery disease (CAD) involvement has been described in up to 7% of ET cases; certainly, this incidence increases with the presence of traditional risk factors such as age greater than 60-year-old, tobacco use, hypertension, and diabetes mellitus. Furthermore, a higher incidence of CAD is also seen in ET patients with a prior history of thrombosis, documented JAK2 V617F mutation or white blood cell counts in excess of 11,000/microL [6].

We report the case of a young male patient presenting with an acute ST-segment elevation myocardial infarction (STEMI) during stress testing procedure for evaluation of atypical chest pain.

Case Report:

A 43-year-old hypertensive male exposed to second hand smoke presents with two months of mid epigastric burning sensation. An upper endoscopy was unremarkable. An exercise electrocardiogram was then requested using a standard Bruce protocol. The patient achieved a total of 13.5 METS with otherwise unremarkable peak exercise stress test electrocardiogram, normal blood pressure response, no arrhythmias or chest pain. However, 5 minutes into recovery the patient started complaining of diaphoresis, near syncope, and hypotension and became acutely ill (blood pressure 106/72 mmHg and heart rate of 100 bpm). An electrocardiogram (Figure 1) showed extensive ST-segment elevations in the inferior (III, II, and aVF) and lateral (V4-V6) leads.

On physical examination, muffled first and second heart sounds were present with no murmurs, rubs, or gallops. There was no jugular venous distention and lungs fields were clear.

An emergent coronary angiogram demonstrated total occlusion of the proximal right coronary artery. A wire was immediately introduced promptly restoring flow (Figure 2). Percutaneous transluminal coronary angioplasty was then performed with a pressure of 8 atmospheres for 14 seconds and a drug eluding stent (Xience alpine 4.0 x 18 mm (diameter × length) was successfully deployed demonstrating excellent TIMI grade 3 flow as shown in Figure 3. Medical treatment with aspirin, Ticagrelor, Metoprolol succinate and atorvastatin was then initiated.

A follow-up 2D echocardiogram showed adequate left ventricular systolic function with no residual wall motion abnormalities or mitral as insufficiency.

Laboratory results with regards to electrolytes, renal and liver function tests were all unremarkable. Total serum cholesterol was 146 mg/dL, triglyceride was 200 mg/dL and high-density lipoprotein was 22 mg/dL. Prothrombin time and activated partial thromboplastin time were both within the normal range. However, his complete blood count demonstrated mildly elevated white blood cell count (12.6 k/μL), hemoglobin 15.1 g/dL and hematocrit 47.9% with a markedly elevated platelet count of (674 k/μL). A hematology-Oncology consult was requested and JAK 2 (V617F) was found positive by PCR sequence assay. In view of these results, Hydroxyurea was initiated and after a normal submaximal stress test the patient was discharged home with no follow-up complications.

Discussion:

Essential thrombocythemia is a clonal disorder of myeloid stem cells causing abnormal proliferation of megakaryocytic elements in the bone marrow. The resulting elevation in the platelet count can cause arterial and venous thrombosis secondary to platelet dysfunction.

It is well known that both unstable angina and acute myocardial infarction are characterized by the formation of thrombi within the coronary artery and systemic platelet secretion with increased aggregability [7, 8, 9]. In fact, persistent platelet hyperactivity not only is a well-recognized marker for mortality and further cardiac events in survivors of acute myocardial infarctions, but also of subsequent unstable angina [10,11].

Although coronary artery involvement leading to acute coronary syndromes remains a rare ET complication; controversies regarding the best recommended treatment approach continue to plague management of these patients. Particularly, when most of these patients not only have a non-contributory coronary artery disease risk factor profile, but also are most likely to have insignificant coronary artery stenosis on coronary angiography either during the acute event or when they experience recurrent coronary artery thrombosis post the initial treatment considerations [12]. These initial interventions might include a combination of the following including, the use of glycoprotein IIb/IIIa inhibitor, early angioplasty with stent deployment and institution of immediate dual anti-platelet therapy combined with cytoreduction therapy are the cornerstone of recommended therapies, having satisfying outcomes [13].

Evidence based medicine suggests that hydroxyurea is effective in preventing thrombus formation in ET patients at high risk for systemic artery thrombosis. The latter might occur in the absence of a significantly elevation in the platelet count. In sharp contrast, paradoxical bleeding can occur with excessively high platelet counts (> 100000000 k/ml) due to qualitative and quantitative alterations in platelet function [13]. The basic aim of cytoreduction therapy is to lower platelet counts below 400×10^9/L to achieve a complete response

[14]. Finally, aggressive therapy must also be implemented in individuals with identifiable atherosclerotic risk factors.

In summary, though significant coronary artery atherosclerosis among ET patients leading to an acute coronary artery event is rare occurrence, several cases have been reported; however, our case is the first, to our knowledge, in which an acute myocardial infarction occurs following the completion of an otherwise unremarkable stress test. We speculate that acute vasomotion mechanisms occurring in the recovery period after the patient attained maximal workload precipitated total occlusion of the proximal right coronary artery leading to the ST-segment elevation myocardial infarction.

Figure Legends:

Figure 1: Marked and diffuse ST-segment elevations were noted in leads II, III, avf, and V4-V6.

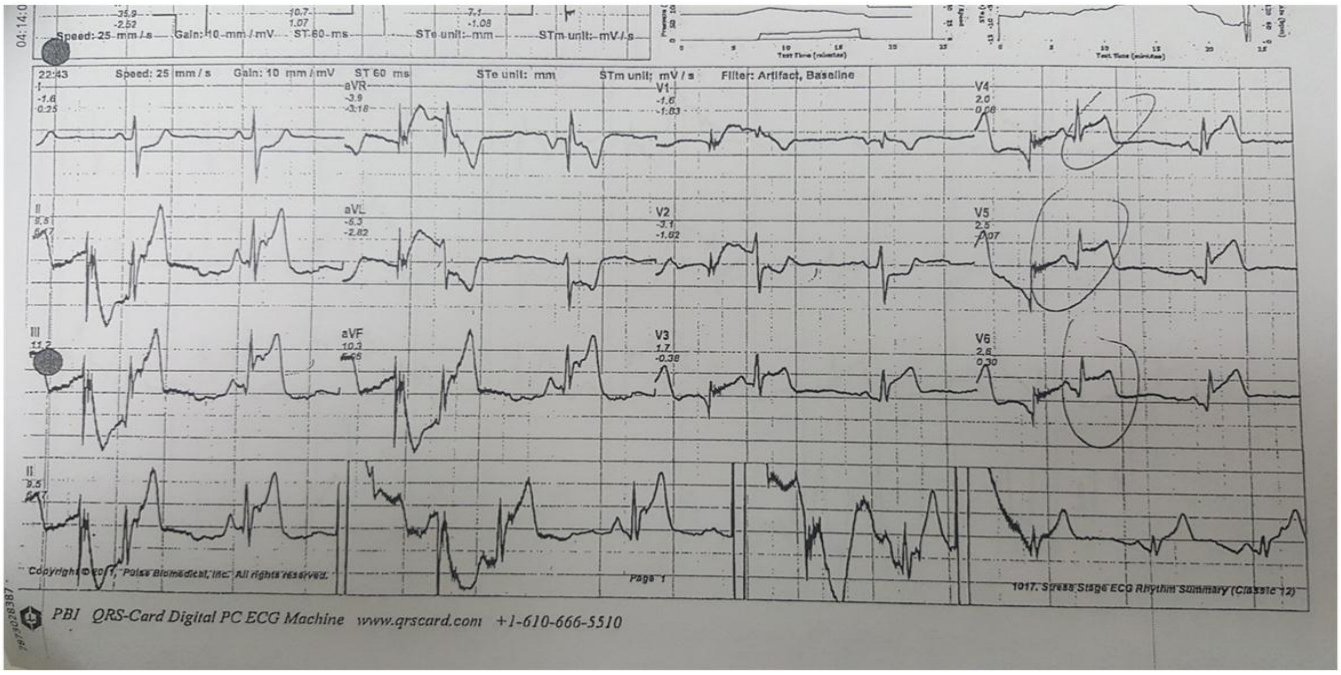


Figure 2: Coronary angiography (Left anterior oblique cranial views) depicting a proximal total occlusion of Right coronary artery.

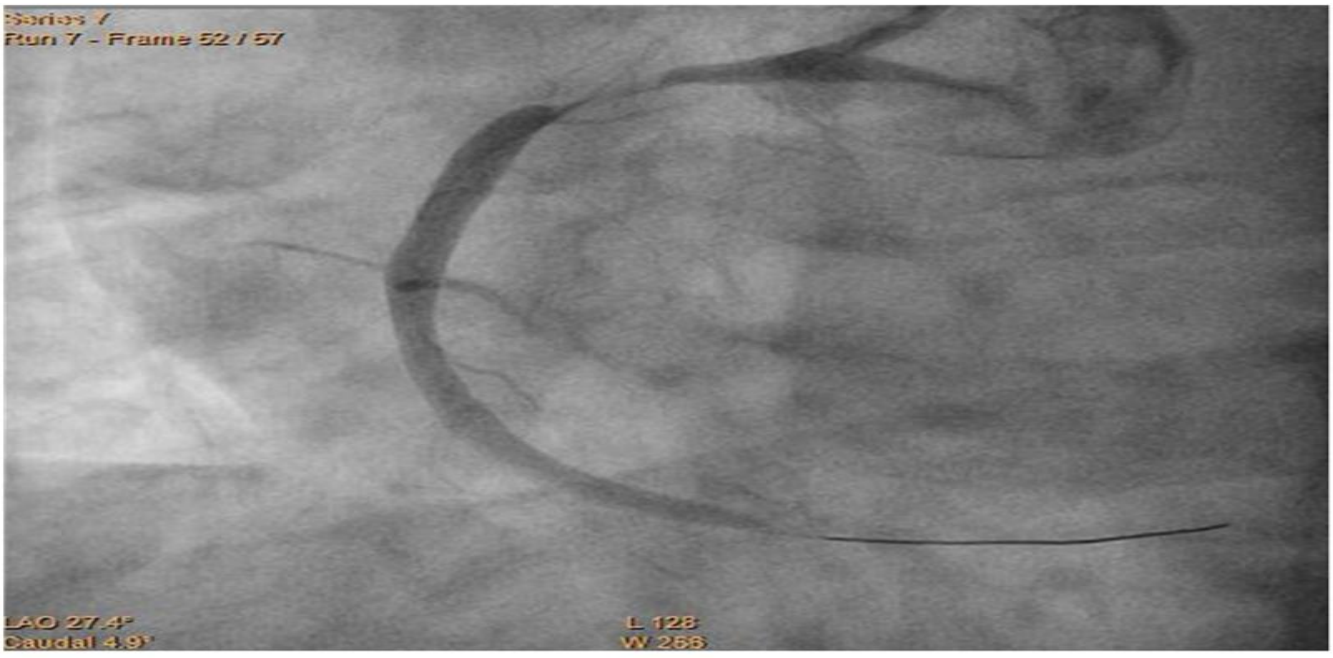
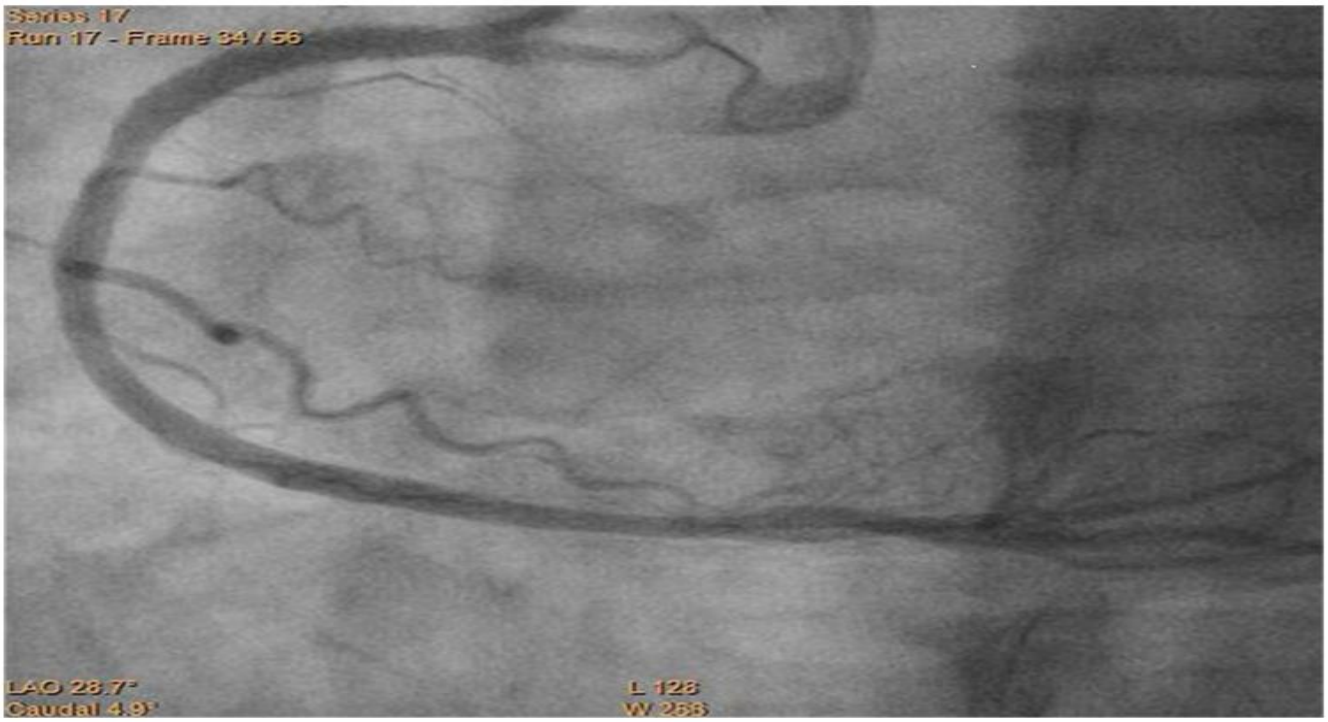


Figure 3. Restoration of TIMI flow grade 3 after successful percutaneous balloon angioplasty with a drug eluting stent deployment.



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