Infarctus du myocarde comme première manifestation clinique de la thrombocytémie essentielle

Acute myocardial infarction as first clinical manifestation of essential thrombocythemia

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Résumé

La thrombocytémie essentielle (TE) est associée à un risque élevé de thrombose. Nous rapportons un cas rare d'un patient de sexe masculin âgé de 30 ans qui a présenté un infarctus du myocarde (IM) secondaire à un thrombus intra-coronaire. Devant l'absence d'athérosclérose associée et la thrombocytose, des investigations plus poussées ont révélé une mutation positive du gène Janus kinase 2 V617F sur la biopsie médullaire et le diagnostic de TE a été établi. Notre cas met en évidence l'importance de poursuivre les recherches sur la TE chez les adultes atteints d'IM et de thrombocytose.

Mots-clés
Thrombocytémie essentielle (TE) ; Infarctus du myocarde (IM) ; Thrombose

Summary

Essential thrombocythemia (ET) is associated with an elevated risk of thrombosis. We describe a rare case of 30-year-old male patient who presented to the emergency room for an acute myocardial infarction (MI) due to a severe intra coronary thrombus. Further investigations for intracoronary thrombus with no atherosclerotic disease and increased platelet count revealed positive Janus kinase 2 V617F gene mutation on bone marrow biopsy and the diagnosis of ET was established. Our case highlights the importance of further investigation for ET in adults with MI and thrombocytosis.

Keywords
Essential thrombocythemia (ET) ; myocardial infarction (MI) ; Thrombose

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INTRODUCTION

Essential thrombocythemia (ET) is a myeloproliferative neoplasm characterized by abnormal proliferation of the megakaryocytes in the bone marrow and sustained elevation of platelet count in the peripheral blood [1]. The estimated incidence of ET is 0.2-2.5/100,000 people per year with a prevalence of 38- 57/100,000 people [2]. The median age at diagnosis is 60 years but seems that in the current years age is slightly younger [3]. Thrombosis in the cerebral, coronary, and peripheral arteries is the major cause of morbidity and mortality in patients with ET [4]. The incidence of acute coronary disease is about 9.4% [5]. The JAK2-V617F mutation is involved in approximately 50% of patients and recent studies have demonstrated that it contributes to the rise of both venous and arterial thrombosis [6]. In the medical literature, just few cases of acute myocardial infraction (AMI) have been reported in association with ET. Actually, we present an infrequent case of ST-segment elevation myocardial infarction (STEMI) as the first clinical presentation of previously undiagnosed ET, with positive Janus kinase 2 (JAK2) V617F gene mutation.

CASE REPORT

A 30-year-old man with no past medical history presented to the emergency room reporting chest pain. His symptoms of severe, crushing, non-radiating, and retrosternal chest pain and associated with multiple episodes of vomiting, which started half an hour before he came to the emergency department. He reported no history of cardiac problems, was not taking any cardiac medications, had no family history suggesting any cardiac diseases and he denied smoking and alcohol or illicit drug use. On physical examination the blood pressure was 104/70 mmHg and pulse were 100 beats per minute, respiration was 25 breaths per minute, and temperature was 37.2°.

The lungs were clear to auscultation, there were no organomegaly and focal neurology deficits. A 12-lead electrocardiogram revealed sinus tachycardia associated with ST-segment elevation and necrosis Q waves in the anterior and lateral leads.

Atrial and ventricular extrasystoles were also noted [figure1]. An acute STEMI was diagnosed, so a loading dose of aspirin (250mg intravenous), clopidogrel (600mg orally) and 35 mg intravenous bolus of enoxaparin were given.

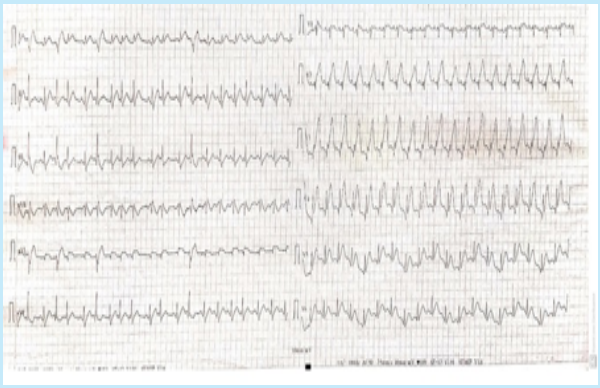

Figure 1 : Electrocardiogram: sinus tachycardia with ST waves elevation and pathologic Q waves in the anterior and lateral leads. Note the frequent atrial and ventricular extrasystoles.

5 minutes later the patient presented a cardiac arrest (ventricular fibrillation) which was resuscitated by an external electric shock. He was transferred immediately to the cardiac catheterization laboratory for primary percutaneous coronary intervention. the patient underwent urgent coronary angiography, revealing an acute occlusion by an intramural thrombus of the proximal LADA with TIMI 0 flow. There was no other significant lesion [figure2]. A Bare metal stent was successfully implanted in the proximal LAD, this was because we did not have a drug eluting stent at the time of angioplasty [figure3]. Anti GP IIb IIIa and in situ thrombolysis were not used given that the patient was not previously diagnosed with ET.

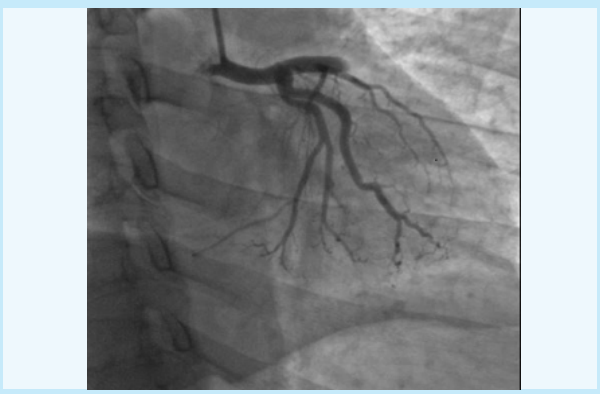

Figure 2 : Left coronary angiogram showing thrombotic occlusion of the proximal left anterior descending artery with TIMI 0 flow.

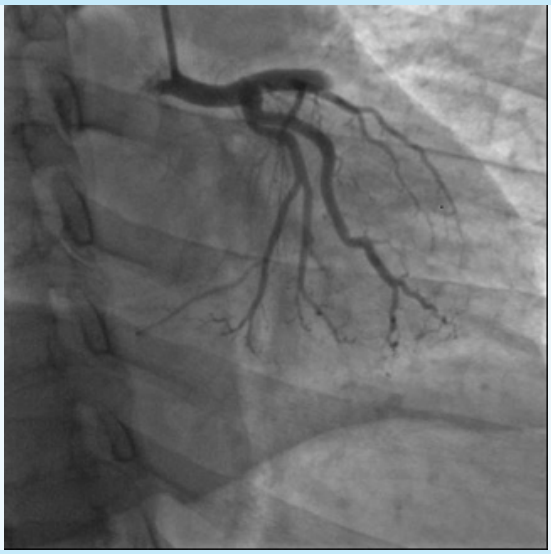

Figure 3 : A 4.5 × 20 mm bare metal stent was placed at the proximal left anterior descending artery to restore normal TIMI flow grade 3.

Regardless of the underlying mechanism, acute myocardial infarctions are currently treated in a standardized fashion, simply because they all look alike in coronary angiography. The primary treatment objective in patients with STEMI is rapid restoration of TIMI III flow; usually achieved with stenting. Coronary angiography cannot only identify the underlying pathology but also deprives us from understanding the natural history of different types of plaques, that's why intra coronary imaging would be the ideal solution to define the mode of disruption at the time of the acute coronary syndrome. There was regional wall motion abnormality with hypokinesia of the anteroapical wall with estimated left ventricular ejection fraction of 35-40%. Blood tests revealed isolated but severe thrombocytosis with 1220 × 10^9 platelets /l, the rest of the hemogram was normal. Liver function test results were normal.

The high platelet count persisted after 3 months of control so He was evaluated by the hematology team and had a bone marrow aspiration and biopsy and ET was confirmed by positive JAK2-V617F mutation, negative BCR-ABL1 translocation, and compatible bone marrow biopsy [7].As our patient had high risk factors for thrombotic complications [8], treatment with cytoreductive therapy using hydroxyurea was instituted to lower the platelet count [9] in addition to Aspirin and Clopidogrel. Three months after the prescription of Hydroxyurea, patient continues to do well with no further thrombotic or hemorrhagic complications. Current platelet count is 650 ×109 /l.

DISCUSSION

Numerous pathological processes other than atherosclerosis can involve the coronary arteries and result in myocardial infarction such as coronary spasm, coagulation disorders, collagen vascular diseases, embolization, and oral contraceptive use [10]. In our case, ET with JAK2 V617F mutation initially presented with acute STEMI secondary to intracoronary thrombus formation, otherwise normal coronary arteries, and required percutaneous intervention with successful revascularization. MI in patients with ET often results from in situ thrombosis without associated coronary atherosclerosis. Thrombectomy in similar situations may be discussed but taking into account the risk of stroke linked to the procedure, the limited success of our catheters at retrieving effective thrombus, and the special attention that needs to be paid to avoid a risk of embolization during removal of thrombotic material, the place of thrombo-aspiration is now considerably limited [11]. The LAD artery was occluded in most of the patients presented with MI and ET [12]. The pathophysiological mechanisms remain unclear but appear to be related to qualitative rather than quantitative platelet abnormalities. Vianello et al, [13] demonstrated that ET patients have endothelial dysfunction and lower coronary flow reserve. multiple risk factors have been identified to predict ET-related thrombotic complications. Age more than 60, previous history of thrombotic complications, and platelet count 1500 109 /l were reported as high-risk factors for thrombotic complications [14]. ET treatment is a difficult compromise between balancing the necessity of preventing thrombotic and/or hemorrhagic complications with the drug toxicity. No writing regulations of the treatment of acute STEMI with ET were found in the literature. there are cases reported where patients have undergone angioplasty or coronary artery bypass surgery. The initial drug of choice is hydroxyurea to prevent thrombotic events in ET for high-risk individuals. De Stefano et al. [15] demonstrated a significant reduction of 70% in recurrent thrombotic events in patients with ET who had acute coronary

syndrome. the contemporary use of an antiplatelet agent with cytoreductive therapy showed enhanced efficacy in preventing re-thrombosis.

For patients with ACS, prolonged Dual Antiplatelet Therapy (DAPT) (≥12 months) is beneficial and therefore reasonable as long as the patient is tolerating the therapy and having low bleeding risk. The management of DAPT must be seen as a dynamic prescription with regular re-evaluations of the benefit- risk to the patient according to changes in his/her clinical profile [16].

CONCLUSION

Myocardial infarction as first clinical manifestation leading to the diagnosis of ET is infrequent. Our case emphasizes the importance of pursuing the investigations in young patients presenting with an acute STEMI and high platelet counts. We strongly recommend to keep these patients under double antiplatelet therapy for a long period (for life if bleeding risk is low).

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