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Case Report

Acute Coronary Syndrome and Essential Thrombocytemia: a Case Report

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Abstract

Essential thrombocythemia is a myeloproliferative syndrome that rarely complicates myocardial infarction. We report the observation of a 32-year-old man with no particular history admitted to the cardiology department for an acute coronary syndrome without ST-segment elevation from V3-V6 with elevated troponin. Subsequent coronary angiography revealed a thrombosis of the IVA between IVA 2 and IVA 3, with slight recurrence in the right network. The blood count showed an isolated thrombocytosis of 600,000/mm3.

The evolution was favorable under BASIC treatment and hydrea-based treatment of hyperplaquettosis.

Keywords: acute coronary syndrome; essential thrombocytemia; cardiovascular surgery

Introduction

Essential thrombocythemia is a group of relatively chronic myeloproliferative disorders. It is characterized by an abnormal proliferation of megakaryocytes in the bone marrow and a significant increase in the number of platelets in the peripheral blood. The main clinical manifestations are an increased incidence of thromboembolic and hemorrhagic events [1].

It is currently defined according to WHO criteria, revised in 2008. It is often asymptomatic and is characterized by a chronic primary elevation of platelet count ≥ 600,000/mm3 on two successive examinations separated by 1 month.

Bone marrow biopsy confirms normal or cell-rich marrow, with little or no bone marrow fibrosis. Megakaryocytic hyperplasia is the main finding. The dystrophic megakaryocytes are large, giant, in clusters, with multi-segmented, multi-lobed "stag's horn" nuclei. The diagnosis can only be made after other etiologies have been ruled out.

The main complications are arterial thrombosis of the microcirculation (cutaneous microthrombosis, regressive TIA, mesenteric angina) and the macrocirculation (coronary, cerebral and peripheral arteries), or even venous thrombosis (hepatic veins, portal thrombosis).

Contrary to popular belief, the risk of bleeding is much rarer (< 10%). It is increased in cases of extreme thrombocytosis (platelets ≥ 1,000,000 to 1,500,000/mm3), especially when antiaggregants are taken. This is due to acquired von Willebrand disease, with platelets consuming high-polymer von Willebrand factors [2-3].

The incidence of acute coronary heart disease in patients with thrombocythemia was 9.4% according to the literature. [4]

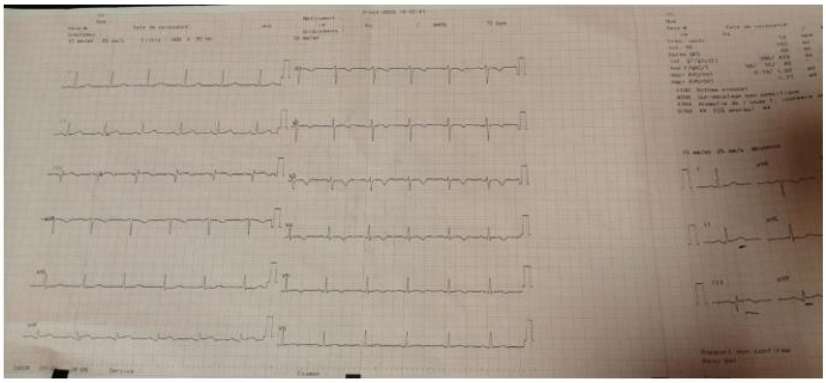
Observation:

We report the case of a 32-year-old man with no particular history and not known to have hyperplateletosis who was admitted to our hospital as an emergency patient with severe chest and back pain evolving for more than 12 hours.

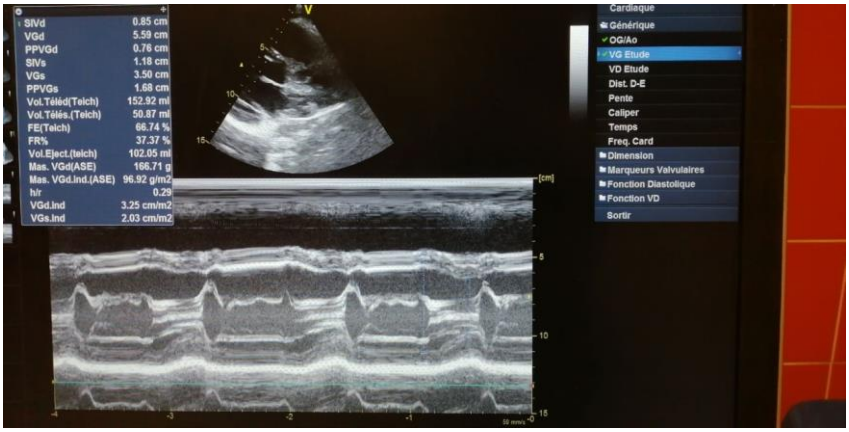
The patient had no specific cardiovascular risk factors, but reported dyspnea and fatigue for the past 2 months.

On his arrival, the patient reported typical angina pains that had been evolving since 5 a.m., prompting his consultation.

The electrocardiogram on admission showed negative t-waves from v3 to v6 with no ST-segment shift. Hemodynamic constants were correct.



The echocardiogram showed a slight reduction in cardiac tip motion with good systolic and diastolic function, both of which remained preserved; there were no valvular defects or pericardial effusion.

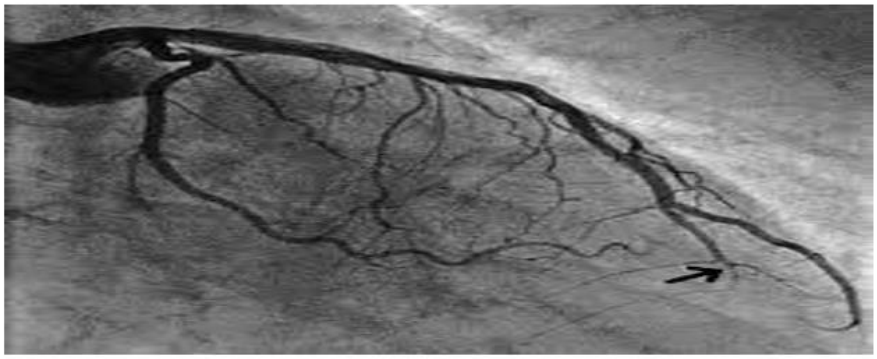


The emergency biological workup after thrombolysis revealed hyperplateletosis with 600,000/mm^3 platelets.

A hematology consult was requested urgently, suggesting that the patient be put on hydration therapy and seen again in consultation after stabilization.

Coronary angioplasty revealed thrombosis between the IVA 3 and the IVA 2, the latter being taken over by the contralateral network; the operator's attempt to unblock it failed.

On discharge, the patient received BASIC treatment and hypo-platelet therapy using hydroxyurea (1000 mg/day) was initiated.



Two weeks after starting hydroxyurea, the platelet count was reduced to 380,000/mm 3 with the onset of excruciating pain at the radial puncture site; vascular Doppler revealed total occlusion with thrombus in the right brachial artery and asymmetry of brachial pulses and blood pressure.

Anticoagulant therapy with acenoucoumarol was instituted in addition to antiplatelet therapy and hydroxyurea to prevent further thrombotic complications.

Discussion:

Essential thrombocytosis/thrombocythaemia (ET) has been considered a rare underlying aetiology for acute coronary syndromes (ACS) [5, 6]. However, we recently showed a prevalence of at least 2.1% in a cohort of patients under 40 years that underwent coronary angiography (CAG) in the setting of their first ACS [7]. More importantly, as shown previously [8], this diagnosis was either missed/severely delayed (average 6 years) despite the presence of elevated thrombocytes (i.e. > 450 × 109/l) upon presentation. This observation might suggest that ET is insufficiently known among cardiologists. The present review aims at summarising the available literature on ET that is relevant, e.g. with regard to diagnosis and treatment, for the cardiologist.

Both thrombotic and haemorrhagic complications have been observed in the setting of ET. Interestingly, many ET patients that present with an ACS have

a normal CAG without signs of atherosclerosis [10, 11–15], which supports the hypothesis that vascular events can be a direct result of the haematological problem, i.e. be unrelated to pre-existing atherosclerosis [16]. In support of this, an > 80% incidence of spontaneous platelet aggregation was shown in patients with ET [17] and large thrombus burden is often described in cases of ACS in the setting of ET (e.g. [7, 8]). Platelet function tests such as prothrombin time, partial thromboplastin time and bleeding time are usually within reference ranges [19]. In those cases where no obstructive coronary artery disease is observed during CAG, it could be argued that thrombus may have resolved after initiation of antiplatelet/ anticoagulation therapy combined with delayed (i.e. after several days or months) CAG. On the other hand, spasm could be provoked by provocation testing in at least two cases [14, 15], suggesting that besides hyperviscosity, endothelial dysfunction or the release of certain platelet-derived vasospasm-promoting substances, such as serotonin and thromboxane A2, might also contribute to the aetiology of ACS in the setting of ET. In support of this, Cheng and Hung

[14] described a patient that experienced recurrent anginal symptoms after discontinuation of diltiazem, but no recurrence of symptoms after discontinuation of cytoreductive therapy despite platelets counts > 900 × 109/l. Both mechanisms for acute occlusion, i.e. thrombotic and vasospastic, and their possible underlying aetiologies are depicted in [10]. Likewise, alterations in platelet function and composition have been implied in long-term complications of ET. For example, organised fibrin, which will replace aggregates of platelets that have become attached to the endothelial surface, may result in extensive intraluminal narrowing of coronary arteries, causing anginal symptoms [16]. Alternatively, the production of proinflammatory eicosanoids [12] and cytokines in the setting of MPNs is thought to explain the development of premature atherosclerosis (and malignancies, see below) secondary to a state of chronic low-grade (endothelial) inflammation in these Altered platelet surface glycoproteins Altered platelet eicosanoid activity Proinflammatory substances induce chronic (endothelial) inflammaon Endothelial surface platelet aggregates are replaced by fibrin Smooth muscle cell proliferaon Fibromuscular inma proliferaon Platelet activation due to endothelial injury Increased platelet procoagulant activity

Possible explanations for this phenomenon include:

1) platelet activation resulting from endothelial damage;

2) prolonged arterial spasm followed by thrombosis;

3) increased platelet activity;

4) changes in platelet granule glycoprotein in ET patients

5) possible selective lipoxygenase deficiency in individuals with myeloproliferative disorders[21]. General treatment of ET includes control of the usual cardiovascular risk factors and cytoreductive therapy such as hydroxyurea in high-risk patients. Patients are considered high-risk if they meet any of the following criteria: age ≥60 years, history of thrombosis or major bleeding and platelet count

≥1500×10(9)/L. In addition, the presence of cardiovascular risk factors or a positive JAK2 mutation confers a significant risk of thrombosis. Low-dose acetylsalicylic acid is the mainstay of primary and secondary thrombosis prevention.22]

Cytoreductive therapy should be added in high-risk patients, including all those who have suffered a cardiac thrombotic event. Hydroxyurea is the first-line choice in the majority of patients due to its favorable side-effect profile and effective reduction in thrombotic events. [22]

However, hydroxyurea has long-term irreversible gonadal toxicity and is contraindicated in the first trimester of pregnancy. Interferon-alpha may be the preferred option in young patients with childbearing intentions, but is associated with more frequent side effects such as hyperthermia, general malaise and gastrointestinal disturbances. The aim of treatment is to maintain a platelet count of less than 400×10 9 /L and, in the case of concomitant leukocytosis, a white blood cell count of less than 11×10.9 /L.[23]

Conclusion:

Essential thrombocythemia is a myeloproliferative disorder manifested clinically by overproduction of platelets in the absence of a defined cause, leading to thrombus formation in systemic arteries, including the coronary artery, resulting in acute coronary syndromes that are sometimes difficult to treat given the high thrombotic load.

The presence of a thrombotic coronary artery with minimal or no cardiovascular risk factors and an elevated platelet count should be considered as potential warning signals for ET. [24]

Even if acute myocardial infarction is caused by ET, emergency interventional treatment is still necessary and should be performed as soon as possible. [25]

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