Acute Coronary Syndrome in Essential Thrombocythemia: Usefulness of Optical Coherence Tomography

Essential thrombocythemia is a rare myeloproliferative neoplasm, characterized by platelet proliferation with quantitative and qualitative alterations. Patients suffering from this condition are more likely to have thrombosis and hemorrhages. It has an incidence of 1 to 2.5 new cases per 100,000 inhabitants per year, (1, 2) and its frequency increases with age, with a 2:1 ratio in favor of women. (3)

One of the complications of essential thrombocythemia is coronary thrombosis, which can be potentially fatal. The incidence of acute coronary events with this hematology-oncology disease is 9.4%, with a rate of fatal and non-fatal thrombotic events of 1.9 per 100 patients/year. (4)

We report the case of a 35-year old female patient with thrombocythemia under study and no cardiovascular history, who presented with moderate, oppressive chest pain lasting 20 minutes, in functional class IV.

Physical examination showed the patient was hemodynamically stable, with blood pressure of 110/60, heart rate of 60 bpm, and no signs of heart failure.

The electrocardiogram revealed sinus rhythm without conduction disorders, narrow-QRS with 0.5 mm transient ST segment elevation (lasting < 20 minutes) from V1 to V3.

Lab tests reported hematocrit 37%, hemoglobin 13 mg/dl, white blood cells 5500/mm^3, and platelets 1,200,000/mm^3. Ultrasensitive troponin was requested, with negative value: 5 pg/ml (normal <14 pg/ml). JAK2 was negative, Leyden factor V was negative, lupus anticoagulants were negative, and bone marrow puncture revealed megakaryocytic hyperplasia consistent with chronic myeloproliferative neoplasm.

Coronary CT angiography showed lack of filling in proximal anterior descending artery causing 80% luminal obstruction, possibly indicative of thrombus or soft plaque (Figure 1A). The patient was started on anticoagulation with unfractionated heparin and aspirin 100 mg/day.

The patient coursed asymptomatic for angina, and a coronary angiography was performed at 72 hours of admission, which showed no significant obstructions (Figure 1B); the study was completed with an optical coherence tomography (OCT) that revealed atherosclerotic fibrolipid plaques in the anterior descending artery at the proximal and middle-third levels (Figure 2A), and an image consistent with plaque rupture (fissure) at the proximal third level in the origin site of the first diagonal branch (Figure 2B).

Due to findings in the catheterization study, atorvastatin 80 mg/day was added to the previous treatment.

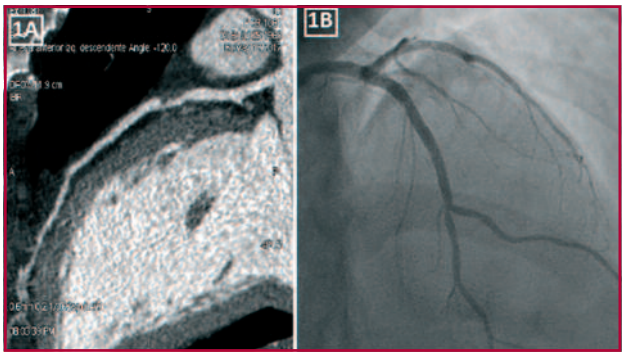

Fig. 1. A. Multislice CT-scan image showing 80% obstruction in the proximal third of the anterior descending artery. **B.** Coronary angiography image showing anterior descending artery with no significant lesions.

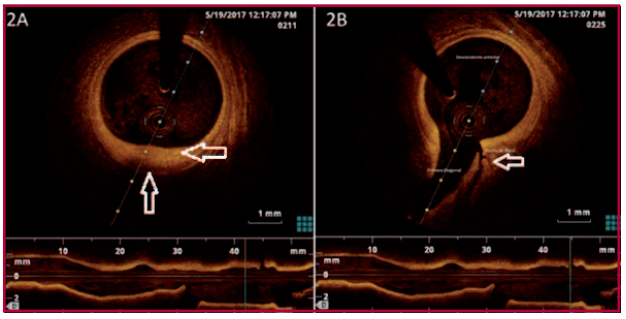

Fig. 2. A. Optical coherence tomography image showing at 6 clock the presence of fibrolipid plaque in the anterior descending artery. The lipid core is observed as an area of low refringence (*arrow*), and the fibrous capsule covering it as a more refringent layer in contact with the arterial lumen (*arrow*). **B.** Optical coherence tomography image showing plaque rupture at the origin of the first diagonal artery. Notice the fissure of the fibrous capsule causing the lipid core to come into direct contact with the arterial lumen.

Echocardiography showed no motility disorders. Left ventricular systolic function was normal, with no relevant valve diseases.

The patient made good progress with no further chest pain, platelet reduction (450,000/mm^3 on discharge), and no changes suggestive of necrosis in the electrocardiogram. The patient was discharged 7 days after admission, continuing with aspirin 100 mg/day, anticoagulation with enoxaparin 80 mg/12 hours, atorvastatin 80 mg/day, and hydroxyurea 2000 mg/day.

Essential thrombocythemia is a condition that may present as an acute coronary syndrome, due to thrombotic events that affect the epicardial coronary arteries. It is a life-threatening complication. (5)

No cases have been described in the literature in which the pathophysiological mechanism of the acute coronary syndrome (in young patients with low risk

coronary pretest, and essential thrombocythemia) is the combination of plaque rupture and superimposed thrombosis.

Our patient was young and had no risk factors for coronary disease; however, the OCT revealed mild atherosclerosis and plaque rupture, accounting for the mechanism of the coronary event. Furthermore, this finding allowed treatment optimization at discharge, since it supported the continuation of high-dose statin therapy together with anticoagulation therapy.

Optical coherence tomography is a high-resolution intravascular diagnostic technique. Initially, it was developed for identifying plaque instability, but nowadays it is also used to identify periprocedural complications, correct stent implantation and, as in our case, plaque rupture detection. (6)

This case is reported to consider the possibility of using OCT in this type of patients, not only to get information about the mechanism of thrombosis but also to guide treatment on discharge.

Conflicts of interest

None declared.

(See authors' conflicts of interest forms on the website/Supplementary material).

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Controversial Management of Severe Thrombocytopenia Induced by Abciximab

The three glycoprotein-IIb/IIIa inhibitors currently in clinical use, abciximab, eptifibatide and tirofiban, all share the same therapeutic target, namely blockade of the final common pathway of platelet aggregation and management of acute coronary syndromes. (1) Thrombocytopenia associated with glycoprotein-IIb/IIIa inhibitors occurs in about 1-2% of the patients exposed to this type of drugs. (2)

We report the case of a 50-year old, hypertensive, obese female patient, with type 2 diabetes. She was admitted with non ST-segment elevation acute coronary syndrome and maximum TnI of 0.55 ng/ml. Dual antiplatelet therapy was initiated with aspirin and clopidogrel, and anticoagulation with subcutaneous enoxaparin 1 mg/kg every 12 hours; a coronary angiography was performed. A drug-eluting stent was implanted in the mid-right coronary artery (RCA) with transient ST-segment elevation by microembilization of the acute marginal branch of the RCA. Intracoronary abciximab bolus application resulted in subsequent recanalization of the branch. Another drug-eluting stent was implanted in the most caudal branch of the obtuse marginal artery. Petechiae and ecchymosis in the upper limbs and body were targeted 24 hours after catheterization, progressing to the lower limbs. Control blood count revealed severe thrombocytopenia with 6,000 platelets/μl (199,000 platelets/μl before catheterization). Hematologists ruled out pseudothrombocytopenia, and recommended platelet transfusion if thrombocytopenia was suspected secondary to intracoronary abciximab administration.

The patient was transferred to the Intensive Care Unit (ICU) due to hypotension with SBP of 80 mmHg and anemia (hemoglobin 8.1 g/dl) (on admission, hemoglobin 12.8 g/dl), even though exteriorization of bleeding was not observed. During her stay in ICU, the patient had episodes of self-limited melena and increased cutaneous ecchymoses. Discontinuation of dual antiplatelet therapy was decided, informing her family of the high risk of severe hemorrhage and stent thrombosis. Three platelet concentrates were transfused while in ICU. Stabilization of platelets was achieved 4 days later. Antiplatelet therapy with aspirin was initiated on the 4th day after catheterization (46,000 platelets/μl), and on the 5th day, in view of hemoglobin stabilization and increased platelets (75,000/μl), clopidogrel therapy was restarted and was well tolerated, with no chest pain episodes or ECG abnormalities during follow-up. On discharge, 9 days after catheterization, the patient had platelet count of 230,000 platelets/μl and hemoglobin of 9.7 g/dl.

Patients with drug-induced thrombocytopenia (DIT) typically present with petechiae, ecchymosis and epistaxis caused by acute, and often severe, decrease of platelet production. When thrombocytopenia