



Difficult Management of Coronary Artery Disease in a Patient with Thrombotic Thrombocytopenic Purpura

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Abstract

Thrombotic thrombocytopenic purpura (TTP) is a rare syndrome affecting multiple organs. There is no sufficient evidence regarding the clinical cardiac manifestations of TTP. Nonetheless, pathologic cardiac involvement is quite frequent in acute TTP, which is predominantly manifested as myocardial necrosis due to coronary arteriolar microthrombosis. The present case report describes a 43-year-old man with long-standing remitted TTP, who suffered from a sequence of refractory thrombotic epicardial coronary events. Aggressive medical and interventional therapies, including long-term dual antiplatelets and coronary angioplasty, were finally successful in remitting the thrombotic events. During his two-year follow up, he has been asymptomatic.

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Introduction

Thrombotic thrombocytopenic purpura (TTP) is a rare and acute syndrome which involves multiple organ systems. In spite of the consistent findings of cardiac involvement in the autopsy studies, clinical evidence for cardiac involvement is surprisingly limited.^{1,2} We report a patient with seemingly remitted TTP, who presented with recurrent episodes of myocardial infarction (MI) and refractory coronary artery thrombosis.

Case Report

A 43-year-old man with recent anterior wall ST-segment

elevation MI (STEMI), which had been previously treated with Streptokinase, referred to our hospital. His past medical history included smoking and a history of TTP, which was in clinical remission after splenectomy 8 years previously. His physical examination was unremarkable, and there was no clinical or laboratory evidence of TTP.

The patient had a platelet count of 367,000 in mL, hemoglobin concentration of 14 g/dL, and serum lactate dehydrogenase (LDH) of 287 IU/mL. In addition, there was an acceptable number of fragmented red cells on the peripheral blood smear.

Echocardiography revealed left ventricular ejection fraction (LVEF) of 35%, and selective coronary angiography showed a fully recanalized left anterior descending artery

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(LAD) with an insignificant ostial lesion. Because the patient was asymptomatic, he was discharged on anti-ischemic medications - including Aspirin (80 mg/d), Plavix (75 mg/d), Atorvastatin (40 mg/d), Metoprolol, and Captopril. Three weeks later, the patient presented again with prolonged resting chest pain, ST elevation in the precordial leads, and new right bundle branch block on the electrocardiogram. Emergent coronary angiography was done, which showed a thrombotically occluded LAD in the ostial portion, accompanied by a large clot in the non-dominant right coronary artery (RCA) (Figure 1). As a result, primary angioplasty was performed using manual thrombectomy, followed by bare-metal stenting (Liberte 3.5×16), with resultant Thrombolysis in Myocardial Infarction (TIMI) flow of 2 (Figure 2). On account of the small caliber, we opted for non-intervention vis-à-vis the RCA and mere reliance on glycoprotein IIb/IIIa inhibitor therapy. The patient spent an uncomplicated hospital course and was discharged 7 days later with LVEF of 20%. Of note, Plavix (75 mg/d) was continued and the doses of Aspirin and Atorvastatin were raised to 325 mg/d and 80 mg/d, respectively.

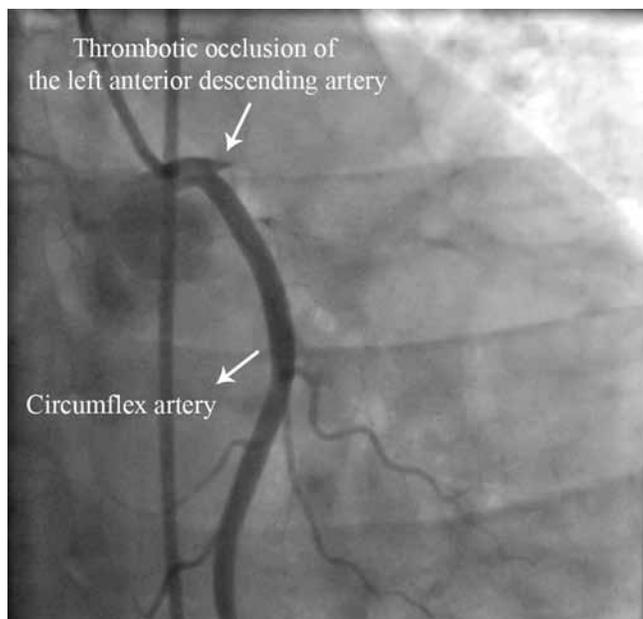


Figure 1. Right anterior oblique-caudal coronary angiographic view, indicating thrombotic occlusion of the ostial left anterior descending coronary artery

The patient frequently reported chest pain during the next 3 months, which finally led to re-admission with the diagnosis of non-STEMI. Nevertheless, laboratory findings were still within normal limits. Refractory symptoms prompted us to perform the third coronary angiography. Apart from slow-flow coronary arteries, the previous LAD stent was patent; however, there was a small globular intraluminal filling defect (possibly thrombosis) at the distal aspect of the stent, alongside the segmental thrombosis of the RCA (Figure 3).

In light of these findings, Plavix was changed to Prasugrel and the patient was discharged. For the past two years since, he has been followed up on medication and has experienced no events.

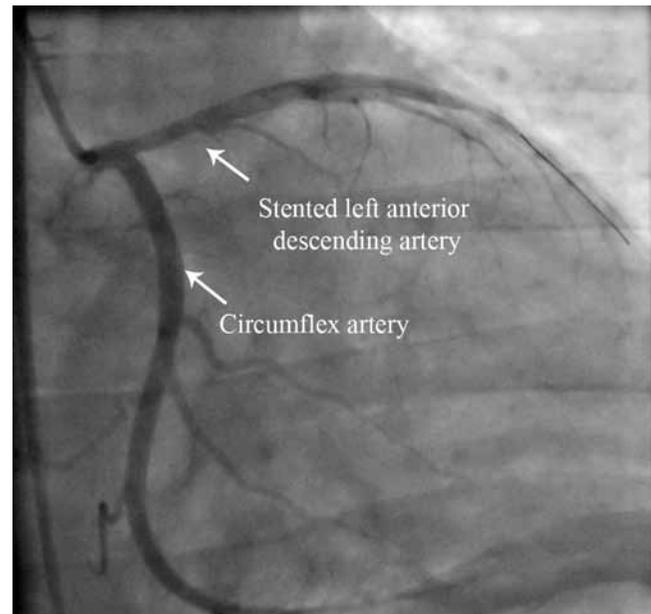


Figure 2. Final right anterior oblique-caudal coronary angiogram after primary angioplasty of the left anterior descending coronary artery

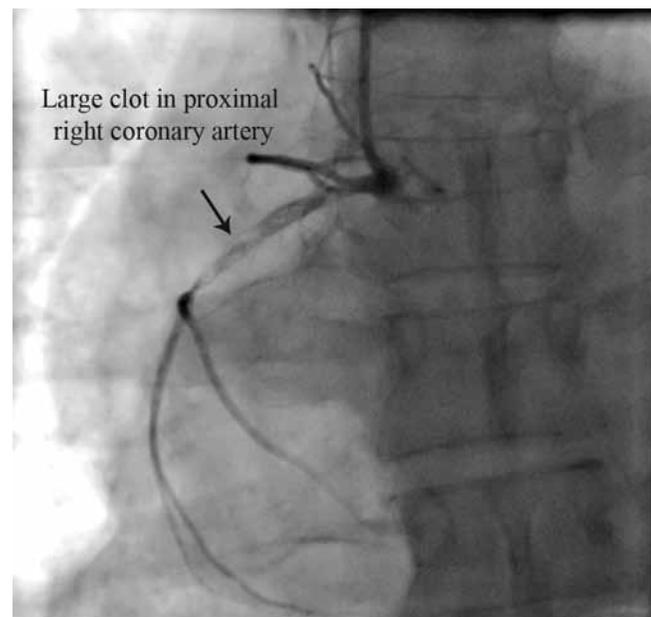


Figure 3. Large thrombosis in the right coronary artery evident in the left anterior oblique view of coronary angiogram

Discussion

This case demonstrates recurrent acute coronary events in a young male with a history of prior manifestations of acute TTP.

TTP is deemed one of the rare causes of non-atherosclerotic MI or a differential diagnosis in some cardiac-involving diseases such as infective endocarditis, systemic lupus erythematosus, and antiphospholipid antibody syndrome. Autopsy studies have shown that the heart is among the most frequently involved organs in patients with TTP, whether before or after the advent of modern therapies. Be that as it may, cardiac symptoms are rarely reported in the literature.³ In addition, it remains unclear whether cardiac involvement may persist after recovery from TTP. It seems that cardiac symptoms are predominantly manifested in the acute phase and are most commonly manifested as MI, congestive heart failure, and arrhythmias.⁴ Hawkins et al.⁴ conducted a systematic review on thirty articles which reported a total of 111 TTP patients: cardiac symptoms were reported in 24 patients in thirteen articles; 13 patients in eleven articles had symptoms described as chest or substernal pain; 10 patients in two articles had symptoms consistent with congestive heart failure; one patient had syncope attributed to a cardiac origin; MI was reported in 26 patients (including STEMI in 7 patients and non-STEMI in one patient; the type of MI was not described in the remaining 18 patients); congestive heart failure was reported in 17 patients; and arrhythmias were reported in 10 patients.

MI could be an early and severe complication of TTP, especially in those who have significantly elevated LDH and cardiac troponin I levels.⁵ Previous studies have concluded that the myocardial damage is most probably due to hemorrhagic necrosis from hyalinized arteriolar microthrombi rather than large-vessel coronary thrombosis, although STEMI has also been reported.⁶

There is paucity of data regarding suitable therapeutic options because of the limited number of involved patients and the absence of uniform recommendations. It seems that the current recommendations on non-TTP patients might be attributable to TTP cases. The case presented herein poses the question whether TTP could play a role in the acceleration of coronary artery disease or recurrent thrombotic coronary events of large epicardial vessels even years after the acute phase.

Conclusion

As TTP is quite rare, studies in this regard are very limited, and a broad follow-up of patients afflicted with this syndrome is required.

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