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ISCHEMIC HEART DISEASE

CASE REPORT: CLINICAL CASE

Coronary Artery Aneurysm Thrombosis Causing Acute Myocardial Infarction in an Adolescent With Neurofibromatosis Type 1

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ABSTRACT

Acute myocardial infarction is a rare though possible occurrence in young patients, especially with predisposing conditions, and the treatment is still debated. We present the case of a teenager known to have neurofibromatosis type 1 with acute coronary syndrome caused by coronary aneurysms and thrombotic occlusion of the left circumflex artery treated with a stent-sparing strategy. (JACC Case Rep. 2024;29:102488) © 2024 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

A 13-year-old teenaged boy was admitted to the emergency department because of acute chest pain at rest in the previous 3 hours.

On admission, the patient's vital signs were normal and the results of cardiovascular physical examination were unremarkable, without any sign of acute heart failure or friction rubs. Skin fold freckling and few café-au-lait spots were noted.

LEARNING OBJECTIVES

- To consider the whole spectrum of causes in myocardial ischemia–like frameworks in the young.
- To investigate possible links between syndromic conditions and heart diseases.
- To prefer a conservative strategy in the presence of coronary aneurysms thrombosis.

PAST MEDICAL HISTORY

The patient's family history revealed that both the mother and the twin brother were affected by neurofibromatosis type 1 (NF-1). The patient had received that diagnosis at the age of 6 years following the onset of dermatologic manifestations.

DIFFERENTIAL DIAGNOSIS

There was no history of fever or recent infection, so suspicion of myocardial or pericardial involvement was unlikely.

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ABBREVIATIONS AND ACRONYMS

ECG = electrocardiogram

LCX = left circumflex artery

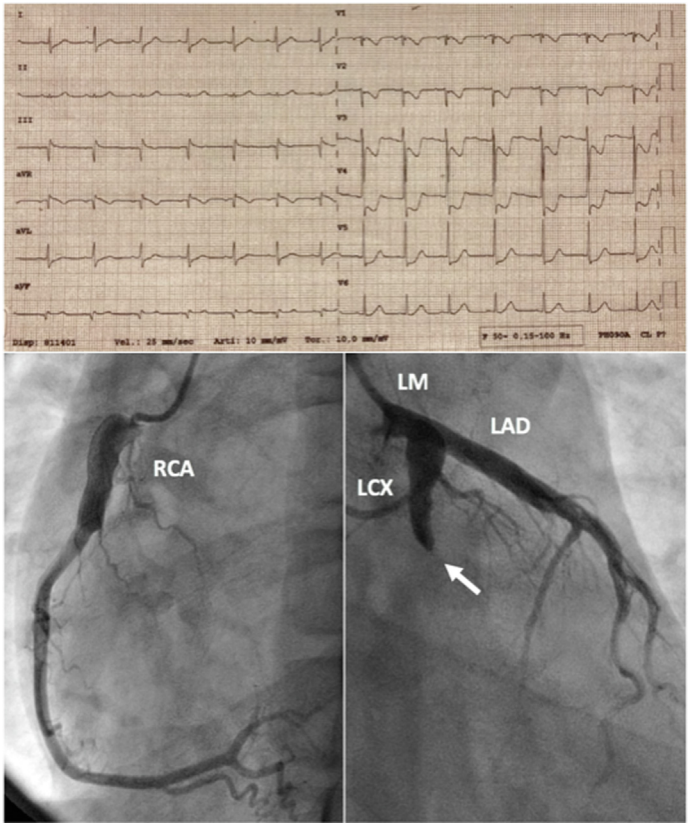
NF-1 = neurofibromatosis type 1

INVESTIGATIONS

The patient underwent cardiovascular assessment through physical examination, electrocardiogram (ECG), and transthoracic echocardiography, without any pathologic finding. No other relevant comorbidities were reported, and the patient was not taking any medication. His weight at admission was 48 kg.

The ECG showed sinus tachycardia and significant ST-segment depression in precordial leads from V_1 to V_5 (**Figure 1**, Video 1). Transthoracic echocardiography showed a moderate reduction in left ventricular ejection fraction (40%), with inferior and posterior wall akinesia and mild functional mitral regurgitation; there was no pericardial effusion; right ventricular function was normal; and congenital heart defects were excluded. Blood tests showed an initial rise in high-sensitive troponin I, which was 0.36 ng/mL (normal value: <0.034 ng/mL), blood count was normal, and there was no rise in C-reactive protein level.

FIGURE 1 Clinical Presentation and Baseline Coronary Angiography



At hospital admission, electrocardiogram shows posterior myocardial infarction. Coronary angiography demonstrates acute thrombotic occlusion of a giant aneurysm of the proximal LCX (arrow) and a further giant aneurysm of the proximal right coronary artery.

MANAGEMENT

Considering the patient's clinical presentation, and given the known association between NF-1 and coronary abnormalities, the diagnostic workup of myocardial infarction was made, and the patient was referred to the catheterization laboratory. He underwent urgent coronary angiography by a right radial approach, which showed proximal left circumflex artery (LCX) acute thrombotic occlusion at the site of a giant aneurysm; the left anterior descending artery was normal, and a giant aneurysm of the proximal right coronary artery was also identified without signs of thrombosis (**Figure 1**, Video 1). Thrombectomy using the 6-F Eliminate (Terumo, Hatagaya) thrombus aspiration catheter was performed with several passages, obtaining prompt vessel recanalization with TIMI flow grade 2 (**Figure 2**, Video 1). To further reduce the residual thrombotic burden, intracoronary thrombolysis (alteplase 5-mg bolus) was administered followed by downstream glycoprotein IIb/IIIa inhibitor infusion (tirofiban 6 mL/h for 18 h, without bolus) and parenteral anticoagulation (unfractionated heparin with target-activated partial thromboplastin time of 50-70 s), with prompt complete ST-segment resolution shown by the ECG and regression of symptoms. Dual antiplatelet therapy (aspirin 100 mg daily and ticlopidine 250 mg twice daily) was started, and the patient was monitored in the hospital. The hospitalization was uneventful.

DISCUSSION

Across the wide spectrum of NF-1 phenotypical manifestations, the development of coronary artery

aneurysms is rarely described.[1] This complication, commonly recognized in childhood or early adulthood, may cause acute myocardial infarction and is associated with a high risk of recurrent thrombotic events.[2] Nevertheless, the management of coronary artery aneurysm thrombosis in pediatric patients is still a matter of debate.[3] We used in the immediate postoperative management a dual antiplatelet therapy, in a setting of acute coronary syndrome. Inasmuch as the patient's weight was <50 kg, treatment with prasugrel and ticagrelor was excluded because of poor safety data.[4,5] Moreover, a resistance test for clopidogrel was performed, showing poor response to the active substance. Therefore, ticlopidine was the drug of choice in addition to aspirin.

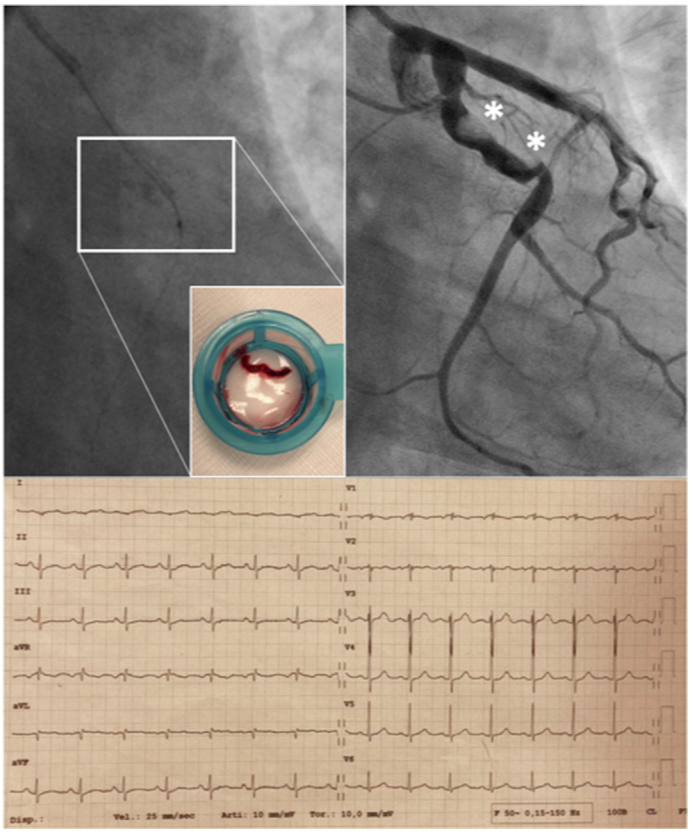
FOLLOW-UP

Follow-up coronary angiography at 7 days showed LCX patency with complete thrombus clearance and TIMI flow grade 3 without residual stenosis (**Figure 3**, Video 2). The patient was discharged asymptomatic and in good clinical condition to take single antiplatelet therapy (aspirin 100 mg daily) and oral anticoagulation therapy (warfarin with target INR of 2.5). The necessity for anticoagulant therapy in coronary aneurysms to prevent the progression of intracoronary thrombosis and recurrence of myocardial infarction has already been established inn the literature.[6] At the moment, there are no available data about the efficacy and safety of direct oral anticoagulants in this specific population; therefore, we opted for a vitamin K antagonist. Echocardiography at discharge showed left ventricular ejection fraction recovery to 50% with residual posterior wall hypokinesia. At his 6-month follow-up visit, the patient had experienced no adverse events.

CONCLUSIONS

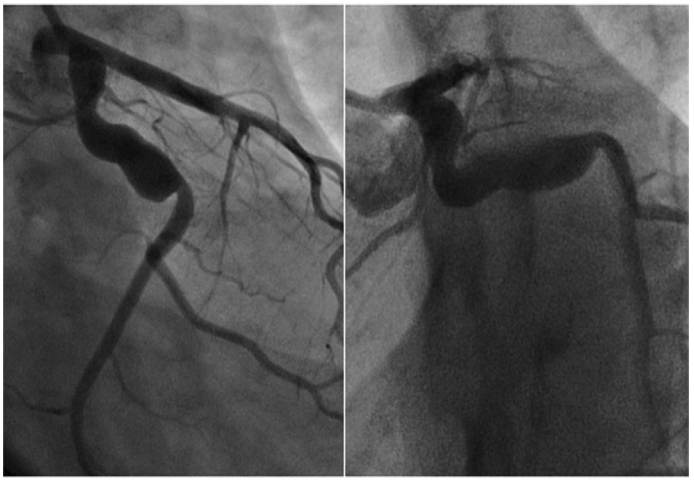
In this population, conservative strategies to restore blood flow should be pursued, because stent implantation could be technically challenging and limited by the lack of long-term data. Optimal antithrombotic therapy regimen is controversial. Anticoagulation may reduce recurrent events, although the patient's compliance and bleeding risk should be carefully assessed in this setting.

FIGURE 2 Conservative Management of Coronary Artery Aneurysm Thrombosis



Thrombectomy using manual thrombus aspiration catheter (box) with removal of thrombus is shown. Post-thrombectomy angiogram demonstrates flow restoration and residual thrombotic burden (asterisks) in the LCX.

FIGURE 3 Follow-Up Coronary Angiography



Follow-up angiography after 7 days of antithrombotic therapy showing complete thrombus resolution in the LCX.

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KEY WORDS coronary artery aneurysm, myocardial infarction, neurofibromatosis, pediatric cardiology

APPENDIX For supplemental videos, please see the online version of this paper.