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Case Report

Progressive spontaneous coronary artery dissection secondary to fibromuscular dysplasia requiring mechanical circulatory support

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ABSTRACT

Spontaneous coronary artery dissection (SCAD) usually appears as an acute coronary syndrome with good prognosis. We present the case of a 39-year-old woman with progressive SCAD secondary to fibromuscular dysplasia with catastrophic course. The patient required several mechanical circulatory support systems including a left ventricular assist device (CentriMag®, Thoratec, Pleasanton, CA, USA) as bridge to recovery.

<**Learning objective:** The beneficial use of mechanical circulatory support devices as a bridge to recovery or heart transplant in the setting of refractory cardiogenic shock in spontaneous coronary artery dissection.>

Introduction

Spontaneous coronary artery dissection (SCAD) is a vessel wall lesion characterized by the acute development of an intramural hematoma (i.e. false lumen) that flattens the lumen (i.e. true lumen) through the shift of the inner media against the opposite wall, which by definition requires the absence of an iatrogenic, non-coronary, or traumatic etiology [1]. This clinical entity supposes an uncommon cause of sudden death and acute coronary syndrome (ACS) and its etiology and pathophysiology are not fully understood [2]. SCAD has been linked to multiple diseases, among them, fibromuscular dysplasia (FMD), another rare arterial disease with equally uncertain etiology [3].

We present the case of SCAD in a young woman, presenting as ACS and cardiogenic shock that required implantation of several mechanical circulatory support devices and was finally diagnosed as FMD.

Case report

A 39-year-old female with hypertension, who was lactating during the seventh month postpartum, was admitted to the emergency department with chest pain and ST-segment elevation on the inferior wall. An emergent coronary angiography showed intra luminal filling defects involving the distal circumflex artery and in intermedius branch, which suggested a spontaneous dissection with thrombolysis in myocardial infarction 3 flow, which was verified with optical coherence tomography (OCT) (Fig. 1A, B). According to the angiographic findings and the clinical stability, we chose a conservative approach. The echocardiogram showed normal biventricular systolic function.

Two hours later, the chest pain worsened associating electrocardiography (ECG) signs of diffuse ischemia and she evolved in a few minutes into cardiogenic shock and cardiac arrest with electrical storm. Despite 20 min of advanced cardiovascular life support, refractory ventricular fibrillation (VF) was noted and we decided to perform extracorporeal life support (ELS) with peripheral veno-arterial extracorporeal membrane oxygenation (ECMO). A new coronary angiography showed progression of the dissection and intramural hematoma toward the left main artery but maintaining adequate distal flow. The echocardiographic control revealed a worsening of biventricular function (left ventricular ejection fraction: 20%) with diffuse hypokinesis. Due to this situation an early surgical revascularization was decided.

However, before the patient could reach the operating room, she experienced a new episode of hemodynamic instability with diffuse ischemia in the ECG, and a third coronary angiography was performed, which confirmed a progression of the dissection with flow compromise in left main, anterior descending, and first diagonal arteries (Fig. 1C). Due to this situation of instability, a

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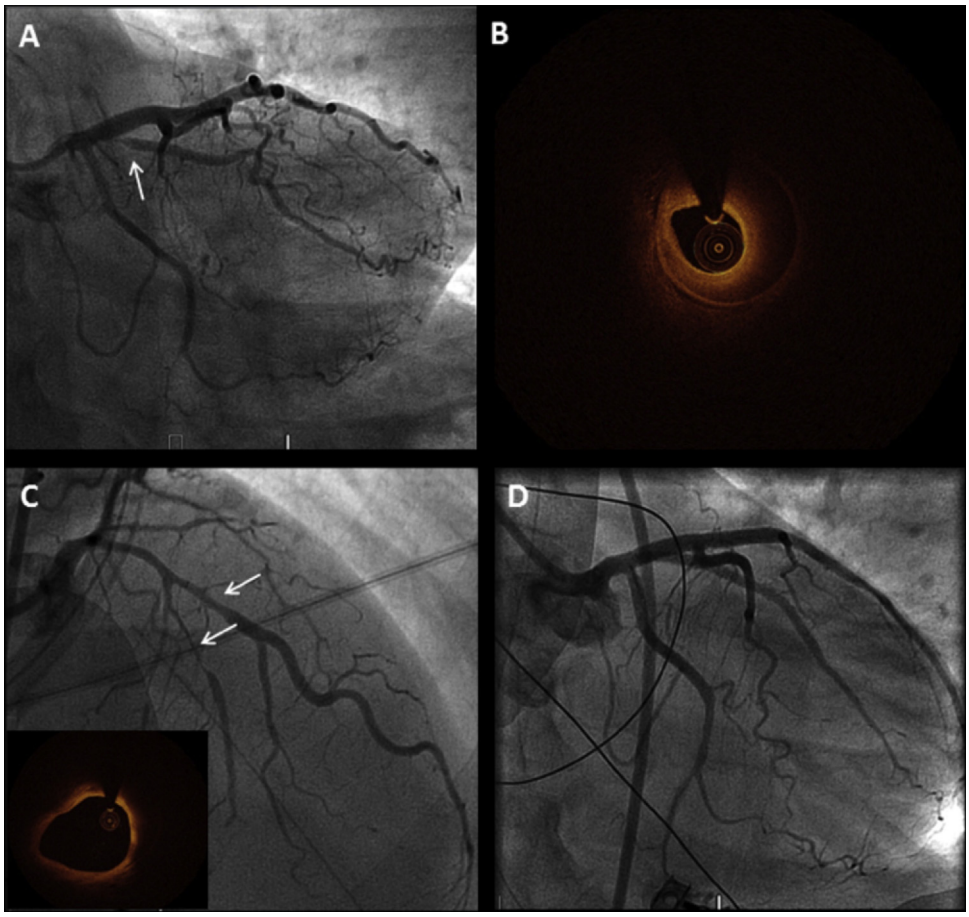

Fig 1. Coronary angiography. (A) First coronary angiography with critical stenosis on the start of the intermedius branch and involvement of the distal circumflex artery. (B) Intermedius branch optical coherence tomography (OCT) with spontaneous dissection and intramural hematoma. (C) Progression of the dissection to left main (OCT), anterior descending and first diagonal (coronary angiography). (D) Coronary angiography after the percutaneous revascularization.

complete percutaneous revascularization was applied with drug-eluting stents, with good angiographic and hemodynamic results (Fig. 1D).

The patient remained stable under the ECMO support but still presented severe ventricular dysfunction and required mechanical support without successful weaning. Given the possibility that the cardiogenic shock became irreversible, the patient was transferred to a heart transplant center, where the mechanical support was replaced by a longer lasting support system: a left ventricular assist device (CentriMag®, Thoratec, Pleasanton, CA, USA) and right central veno-arterial ECMO.

The improvement in her hemodynamic condition and biventricular contractility made it possible to reduce the mechanical support and weaning was achieved after 11 days, being discharged approximately two months after her admission with normal biventricular function. A carotid arteriogram revealed the classical appearance of "string of beads" on the right carotid artery, which suggests the diagnosis of FMD (Fig. 2).

Discussion

SCAD is a relatively uncommon entity. Its prevalence is reported to be 0.07–0.28%; it affects primarily women and usually appears as an ACS with good prognosis [2].

The angiographic appearance may be indistinguishable from arteriosclerosis; the use of intravascular ultrasound (IVUS) and OCT is useful to visualize the vascular wall and differentiate it, to quantify the extension of the lesion, and to guide treatment [4], although intracoronary manipulation may lead to risk for expansion of dissection. Several factors have been related to the etiology of this condition. One of the most important factors involved is FMD [3].

FMD is an idiopathic, segmental, nonatherosclerotic and non-inflammatory disease of the musculature of arterial walls, leading to occlusion, stenosis, aneurysms, and dissection of small and medium-sized arteries [5]. A strong association has been observed between SCAD and FMD, and ACS may be the first manifestation of this disease [3].

Although the ultimate diagnosis is histopathological, angiography is the choice technique to rule out involvement in most locations [5]. The characteristic angiographic pattern of medial fibroplasia is that of alternating dilated and stenosed areas, forming what is called a "string of beads" appearance. FMD can also be associated with aneurysms and dissections, which may form a pathophysiological link with SCAD. Anatomicropathologically, this image corresponds with alternating zones of thickening and thinning of the arterial medial layer. These findings have recently been visualized in vivo using OCT [6].

Despite SCAD being a serious condition with high risk of death, there are no standardized management plans. Medical treatments are frequently considered in hemodynamically stable patients and while the coronary flow is maintained. Aspiration thrombectomy [7] or primary intracoronary stenting is usually provided in case of recurrent angina, hemodynamic instability, or mono-vessel

Fig. 2. Right carotid arteriogram showing the "string of beads" appearance.

disease. Despite the use of drug-eluting stents being controversial, they could be implanted when there is left main involvement to prevent the high risk of restenosis and due to the length of the region to be treated. Surgical revascularization is the treatment of choice when the main stump or many coronary arteries are involved. Unfortunately, this procedure can fail when the largest surface of myocardium is under ischemia for many hours [8].

Extracorporeal life support (ECL) as an adjunct to cardiac resuscitation has shown encouraging outcomes in patients with cardiac arrest [9]. In our case, ECMO was not enough, due to severe ventricular dysfunction, pulmonary congestion and the need for a longer circulatory support as a bridge to decision (transplant vs. recovery), and we decided make a switch for a left ventricular assist device (CentriMag) and right central veno-arterial ECMO.

The low complication rate makes the CentriMag safe to use for patients who need additional time for evaluation or for sufficient recovery for purposes of being considered suitable for a heart transplant or a long-term device [10].

Our case report appears to be the first published use of several mechanical circulatory support devices in a patient with SCAD and FMD. This case demonstrates the beneficial use of mechanical circulatory support devices as a bridge to recovery or heart transplant in the setting of refractory cardiogenic shock in this pathology.

Conflict of interest

None declared.

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