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Case Report

A Rare Case of Fibromuscular Dysplastic Coronary Artery Disease: A Multi-Disciplinary Approach to Management

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Abstract

Fibromuscular Dysplasia of the Coronary Arteries (FMD-CAD) is a rare finding and may present with an acute coronary syndrome requiring medical, interventional, procedural and/or surgical interventions. The case described in this report illustrates a case in which the acute presentation required a multi-disciplinary approach to treatment. Rare coronary angiographic imaging of the pathology is provided.

Keywords: Coronary Artery Disease; Fibromuscular Dysplasia; Coronary Artery Bypass Grafting

Case Presentation

A 59-year old man with no known past medical history presented to the emergency department with exertional chest pressure. There were no other associated symptoms. An electrocardiogram was consistent with an acute ST-elevation anterior wall myocardial infarction. The patient was loaded with aspirin and clopidogrel followed by emergency cardiac catheterization. This study showed FMD-CAD with acute thrombotic occlusion of the Left Anterior Descending (LAD) and its diagonal branch (Figure 1A and B). A percutaneous coronary intervention was performed to the LAD with resultant perforation of the artery and development of a hemopericardium with cardiac tamponade. A covered stent was placed within the artery and a pericardial drain deployed. A brief period of pulseless electrical activity occurred requiring Cardiopulmonary Resuscitation (CPR) and placement of an Impella® device (Abiomed Inc., Danvers MA). Once vital signs were restored, a repeat echocardiogram showed a persistent pericardial effusion accompanied by moderate to severe Left Ventricular (LV) dysfunction. Cardiac surgery was consulted when the patient's hemodynamics deteriorated upon arrival to the coronary care unit.

The patient was taken emergently to the operating room where a median sternotomy was performed. The pericardium was found to be tense. Upon opening the pericardium an immediate rush of bloody fluid was observed with mprovement in the blood pressure. Inspection of the anterior epicardial surface showed extensive ecchymosis with no visible LAD and no obvious bleeding point. The decision was made to apply topical sealants: Evicel® (Ethicon, Somerville NJ), SurgiFlo® (Ethicon, Sommerville NJ), and Surgicel® (Ethicon, Somerville NJ) to the area of injury in the LAD territory. The procedure concluded and the patient recovered uneventfully. He was discharged home on the fifteenth postoperative day.

The patient was followed in the cardiology and cardiac surgery clinics with occasional periods of exertional angina over the course of six months. Despite medical management, the symptoms persisted.

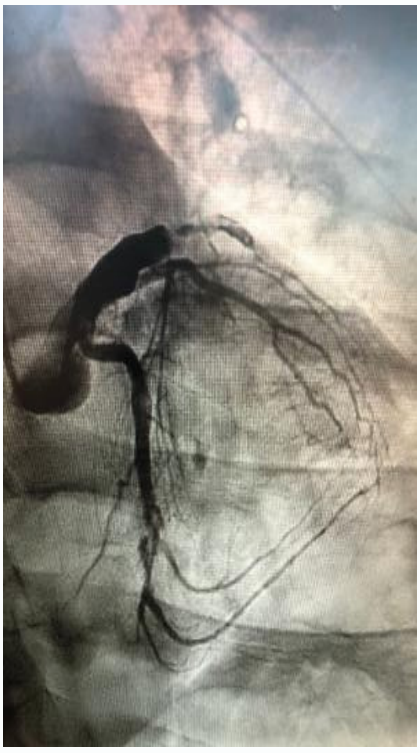

Figure 1A: Left coronary artery.

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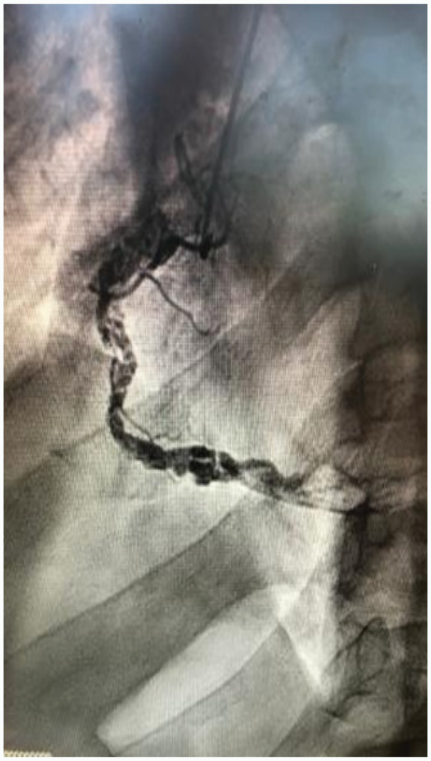

Figure 1B: Right coronary artery.

Repeat cardiac catheterization was performed showing similar coronary pathology with faint visualization of the LAD. The patient was offered redo sternotomy with Coronary Artery Bypass Grafting (CABG). Echocardiography showed significant LV dysfunction with an estimated Ejection Fraction (EF) of 30% - 35%. Intraoperatively there were extensive adhesions making coronary artery identification challenging, but possible to the LAD and Posterior Descending Artery (PDA). There were no suitable targets for grafting of the obtuse marginal branches of the left circumflex. The patient was discharged home on the sixth postoperative day.

The patient continued to follow up with cardiology and cardiac surgery with no further anginal symptoms. CT angiograms of the chest, abdomen, pelvis, and neck were performed to determine if any other vascular territory showed evidence of FMD. Although the renal and carotid arteries did not reveal evidence of FMD, the V3 segment of the right vertebral artery was positive for FMD changes.

Serial echocardiograms over the course of several months failed to show any improvement in the LVEF. An electrophysiology consultation was obtained and the recommendation for placement of an Implantable Cardiac Defibrillator (ICD) was made. This device was successfully deployed without complication. At present, the patient remains symptom-free.

Discussion

The first reported pathology of coronary FMD was described by Hill in 1965 [1]. Subsequent reports of FMD-CAD have been described in mini-series and autopsy reports [2]. However, the diagnosis of FMD-CAD is sometimes problematic since some cases are assigned this label because of an associated condition-Spontaneous Coronary Artery Dissection (SCAD). Although FMD-CAD may result in SCAD, SCAD itself may not always be a manifestation of FMD-CAD. As such, it is thus far impossible to determine the prevalence of this specific disorder. From a strict definition standpoint, FMD must be identified in at least one other noncoronary arterial territory to attribute coronary findings to FMD [3]. Although the classic angiographic "string of beads" observed in renal artery FMD has not been seen in FMD-CAD, the angiographic images of this case would suggest otherwise.

The treatment of FMD-CAD is challenging and can be considered in two broad categories: a) those with stable asymptomatic disease and b) those with unstable symptomatic disease. A conservative approach is recommended for stable patients without active ischemic symptoms. Although the concern for Spontaneous Coronary Artery Dissection (SCAD) is real, this event will often heal on its own reserving percutaneous or surgical intervention for those cases in which symptoms persist despite optimal medical management. Despite the absence of formal management guidelines, medical therapeutics includes dual anti-platelet therapy and beta-blockade [4].There is no evidence to support the role of statin therapy in patients with FMD-CAD. For symptomatic patients, percutaneous or surgical intervention is appropriate. When an interventional approach is chosen, care must be taken to avoid stent or vessel overdilation-avoiding high balloon inflation pressures and using smaller-sized stents is advised to avoid complications [5]. Coronary artery bypass grafting should be reserved for patients with multivessel CAD with adequate target vessels. Based on this case, additional surgical indications to consider include complication(s) from percutaneous intervention and/or persistent of symptoms despite medical and interventional treatments.

The case described in this report was noteworthy for several reasons: the angiographic images, the management of the interventional complication, and the eventual need for cardiac surgical bypass grafting and subsequent placement of an ICD at a later date. The purpose of this case is to describe this rare entity, adding to the limited knowledge base of this unusual disorder.

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