Coronary Vasospasm Attributable to Fibromuscular Dysplasia
The Long Bridge to Transplant

Jose M. Castellano, MD, PhD; Prashant Vaishnava, MD; Javier G. Castillo, MD; Anelechi C. Anyanwu, MD; Valentin Fuster, MD, PhD

Case
A 32-year-old woman with a 6-year medical history of peri-menstrual chest pain and left ventricular systolic impairment after a presumed episode of idiopathic myocarditis presented to the emergency department with 2 hours of severe chest pain and an abnormal ECG with anterolateral ST-segment elevations (Figure 1A). During medical interview, the patient reported the use of cigarettes and cocaine in the past (not for the last 2 years) and very similar symptoms just 2 months earlier. At that time, troponin-I was elevated (12 ng/mL), and subsequent coronary angiogram revealed a dissection of the left circumflex artery, but percutaneous coronary intervention (PCI) could not be performed because of technical complexity, and the patient was commenced on aspirin and clopidogrel.

Cardiovascular examination was unremarkable. Laboratory testing revealed elevated creatinine-kinase muscle and brain and troponin-I (95.5 ng/mL and 12.6 ng/mL, respectively). Urine toxicology testing for the presence of cocaine and serum β-human chorionic gonadotropin was negative. Urgent coronary angiography showed a thrombotic subtotal occlusion of the mid-left anterior descending artery with probable vasospasm (Figure 1B). In addition, angiographically, the left circumflex artery had a moth-eaten appearance (Figure 1C). Successful PCI was performed with deployment of a drug-eluting stent. Thrombophilia testing was negative as was genetic interrogation for familial vasculopathies (TAAD1 [thoracic aortic aneurysm and dissection], TAAD2, and FAA [familial aortic aneurysm]), Marfan’s (FBN1), Loeys-Dietz (TGFBR1 and TGFBR2), and Ehlers-Danlos type 4 (COL3A1). Further surveillance of other vascular territories demonstrated an occluded left internal carotid artery from a previous dissection.
commonly affects the renal and carotid arteries, and epicardial vascular disease that disproportionately affects women. It most tissue diseases.1 FMD is a nonatherosclerotic noninflammatory process with abnormal connective tissue, rendering the vessel wall vulnerable to repetitive vasospasm. Coronary spastic angina has been demonstrated among patients with connective tissue diseases.1 FMD is a nonatherosclerotic noninflammatory vascular disease that disproportionally affects women. It most commonly affects the renal and carotid arteries, and epicardial coronary FMD is an uncommon entity.2 Data regarding epicardial coronary artery involvement in FMD are scarce. One case series describes 7 premenopausal or perimenopausal women with acute coronary syndromes and epicardial coronary artery FMD, in which all patients presented with mono or bilateral renal artery involvement as well.1 Recently, Olin et al2 presented the results of the United States registry for FMD. The registry included 447 cases of FMD, 88 of which presented with arterial dissections and aneurysms. Out of these, only 3 cases (3.4%) presented dissections on coronary arteries. The natural history, prognosis, and outcomes in FMD vary according to the vascular site that it involves. Disease progression is thought to take place in about one third of patients with renal FMD, whereas cerebrovascular disease is reported to progress infrequently.1 Data regarding outcomes on other arterial territories are scarce, and arterial dissection has been proposed as the most likely mechanism by which FMD causes acute coronary syndrome.3

Our patient demonstrated recurrent coronary arterial vasospasm, superimposed thrombosis, and dissection, leading to refractory symptoms with extensive myocardial necrosis and left ventricular dysfunction. Multiple trials of medical therapy were attempted, along with multiple PCIs, mechanical circulatory support, and eventual heart transplantation. On the basis of the histopathologic finding of FMD and cystic medial degeneration, we suspected a pathological process with abnormal connective tissue, rendering the vessel wall vulnerable to repetitive vasospasm. Coronary spastic angina has been demonstrated among patients with connective tissue diseases.1 FMD is a nonatherosclerotic noninflammatory vascular disease that disproportionately affects women. It most commonly affects the renal and carotid arteries, and epicardial coronary FMD is an uncommon entity.2 Data regarding epicardial coronary artery involvement in FMD are scarce. One case series describes 7 premenopausal or perimenopausal women with acute coronary syndromes and epicardial coronary artery FMD, in which all patients presented with mono or bilateral renal artery involvement as well.1 Recently, Olin et al2 presented the results of the United States registry for FMD. The registry included 447 cases of FMD, 88 of which presented with arterial dissections and aneurysms. Out of these, only 3 cases (3.4%) presented dissections on coronary arteries. The natural history, prognosis, and outcomes in FMD vary according to the vascular site that it involves. Disease progression is thought to take place in about one third of patients with renal FMD, whereas cerebrovascular disease is reported to progress infrequently.1 Data regarding outcomes on other arterial territories are scarce, and arterial dissection has been proposed as the most likely mechanism by which FMD causes acute coronary syndrome.3

This case highlights the devastating consequences of a vessel wall disrupted by FMD with coronary vasospasm refractory to traditional therapy. Unconventional, but hypothesis-driven medical and percutaneous treatments were unable to halt this unremitting disease process, with mortality averted only by aggressive surgical intervention and ultimate heart transplantation. To our knowledge, this is the first report of a patient affected by FMD in the epicardial coronary arteries requiring heart transplantation attributable to the aggressive nature of the vasospastic phenomenon despite all available medical and percutaneous management.

Discussion

Our patient demonstrated recurrent coronary arterial vasospasm, superimposed thrombosis, and dissection, leading to refractory symptoms with extensive myocardial necrosis and left ventricular dysfunction. Multiple trials of medical therapy were attempted, along with multiple PCIs, mechanical circulatory support, and eventual heart transplantation. On the basis of the histopathologic finding of FMD and cystic medial degeneration, we suspected a pathological process with abnormal connective tissue, rendering the vessel wall vulnerable to repetitive vasospasm. Coronary spastic angina has been demonstrated among patients with connective tissue diseases.1 FMD is a nonatherosclerotic noninflammatory vascular disease that disproportionately affects women. It most commonly affects the renal and carotid arteries, and epicardial coronary FMD is an uncommon entity.2 Data regarding epicardial coronary artery involvement in FMD are scarce. One case series describes 7 premenopausal or perimenopausal women with acute coronary syndromes and epicardial coronary artery FMD, in which all patients presented with mono or bilateral renal artery involvement as well.1 Recently, Olin et al2 presented the results of the United States registry for FMD. The registry included 447 cases of FMD, 88 of which presented with arterial dissections and aneurysms. Out of these, only 3 cases (3.4%) presented dissections on coronary arteries. The natural history, prognosis, and outcomes in FMD vary according to the vascular site that it involves. Disease progression is thought to take place in about one third of patients with renal FMD, whereas cerebrovascular disease is reported to progress infrequently.1 Data regarding outcomes on other arterial territories are scarce, and arterial dissection has been proposed as the most likely mechanism by which FMD causes acute coronary syndrome.3

This case highlights the devastating consequences of a vessel wall disrupted by FMD with coronary vasospasm refractory to traditional therapy. Unconventional, but hypothesis-driven medical and percutaneous treatments were unable to halt this unremitting disease process, with mortality averted only by aggressive surgical intervention and ultimate heart transplantation. To our knowledge, this is the first report of a patient affected by FMD in the epicardial coronary arteries requiring heart transplantation attributable to the aggressive nature of the vasospastic phenomenon despite all available medical and percutaneous management.

Disclosures

None.

References


Key Words: fibromuscular dysplasia
Coronary Vasospasm Attributable to Fibromuscular Dysplasia: The Long Bridge to Transplant
Jose M. Castellano, Prashant Vaishnava, Javier G. Castillo, Anelechi C. Anyanwu and Valentin Fuster

Circ Heart Fail. 2013;6:e31-e32
doi: 10.1161/CIRCHEARTFAILURE.112.000090
Circulation: Heart Failure is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 2013 American Heart Association, Inc. All rights reserved.
Print ISSN: 1941-3289. Online ISSN: 1941-3297

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circheartfailure.ahajournals.org/content/6/3/e31

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation: Heart Failure can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation: Heart Failure is online at:
http://circheartfailure.ahajournals.org//subscriptions/