Recurrent Right Coronary Artery Occlusion Caused by Cardiac Fibroelastoma Attached to the Aortic Valve

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A 60-year-old church minister was referred to our department for a stress-perfusion cardiac MRI with a clinical presentation of central chest pain 6 weeks previously with a positive troponin result and dynamic inferoposterior ST elevation on ECG. The patient was a nonsmoker, nondiabetic, otherwise fit and well gentleman on no regular medications. Emergency angiography at the time of his initial presentation revealed unobstructed coronary arteries, by which time the ECG changes had returned to normal. Troponin was very mildly elevated (131 ng/L; normal range, 0–13 ng/L) and the patient was given a likely clinical diagnosis of severe coronary spasm. He had no further symptoms until he returned for the outpatient cardiac MRI (CMR).

The CMR was initially unremarkable with normal volumes, function, and wall thickness. T2 images were normal, identifying no acute myocardial edema. The patient had a normal response to adenosine infusion (140 μg/kg body weight per min for 3 minutes) with an appropriate increase in his heart rate and symptoms of chest tightness and flushing that are typical for adenosine; 3 standard short-axis slices were acquired during the first passage of a contrast bolus (gadobutrol, 0.07 mmol/kg body weight).

However, once the study was completed, the patient continued to report severe persisting central chest pain, nausea, and clamminess, despite terminating adenosine, which required patient evacuation from the scanner for further assessment. A subsequent 12-lead ECG demonstrated inferior ST-segment elevation and anterior ST depression consistent with an inferoposterior ST-elevation myocardial infarction (Figure 1A). Following reconstruction, the stress-perfusion images demonstrated marked inferoseptal and inferolateral hypoperfusion with virtually no gadolinium seen in the right coronary artery territory during the acquisition sequence (Figure 1B through 1D, and Movie I in the online-only Data Supplement).

Before emergency coronary angiography, which revealed unobstructed coronary arteries, the patient’s symptoms had spontaneously resolved (Movie II in the online-only Data Supplement). On this occasion, peak troponin was significantly raised (4059 ng/L; normal range, 0–13 ng/L).

The patient was seen in clinic 3 months later where he underwent a transthoracic echocardiogram and functional MRI (Figure I in the online-only Data Supplement). He had reported feeling well, but had experienced 2 further episodes of chest pain, both occurring on a Sunday, the most stressful day of the week for him as a church minister. The CMR revealed extensive inferior scar in the territory that was hypoperfused during the previous CMR. The transthoracic echocardiogram suggested that there was a mass attached to the aortic valve (Movie III in the online-only Data Supplement), which was further quantified by transesophageal echocardiogram (Figure 2A through 2C, and Movie IV in the online-only Data Supplement). The patient underwent urgent removal of the mass (Figure 2D), which was attached to the right coronary leaflet of the aortic valve. The surgery was uncomplicated, and the patient has since made a good recovery. Histological analysis confirmed the appearance of a papillary fibroelastoma and no evidence of malignancy or infection.

Primary cardiac tumors are extremely rare. A series of >12 000 autopsies identified only 7 cases,1 and an echocardiographic series identified 54 cases of all patients who had echocardiographic evaluation over 15 years (109 502 patients had echocardiograms recorded in their database),2 an incidence of <0.03%. Papillary fibroelastomas are said to be the third most frequent primary cardiac tumor (after cardiac myxomas and cardiac lipomas), arising most frequently from the cardiac valves, in particular, the aortic valve (29%), followed by mitral valve (25%), tricuspid valve (17%), and pulmonary valves (13%).3 The clinical presentation of aortic tumors depends on the anatomic position of the mass and may affect the coronary blood flow by a number of mechanisms, including (1) the embolization of tumor mass or platelet thrombi on the tumor into the coronary arteries and (2) the intermittent obstruction of the coronary ostium by the mobile tumor mass. This is the first reported case of a fibroelastoma causing a myocardial infarction during an adenosine stress perfusion CMR.
We suggest that the unusual presentation of acute myocardial infarction in our patient was probably attributable to 1 of 2 possible mechanisms—mechanical obstruction of the right coronary artery ostium in the aortic root owing to the variable mobility of the tumor or the tumor acting as a source of embolic material. The spontaneous and complete resolution of his symptoms on each occasion would make coronary embolism less likely, because this may be expected to cause distal microvascular obstruction and persisting pain.

Intermittent coronary obstruction is a likely explanation. Conditions of stress, as occurs during delivering a Sunday sermon or adenosine infusion, lead to microvascular dilatation and a fall in distal coronary artery pressure. This increased pressure gradient between the coronary ostium and coronary microcirculation augments coronary blood flow via a suction wave effect. This suction wave increases dramatically with pharmacological vasodilation and exercise and has recently been recognized as the main impetus for coronary blood flow. This suction effect may have caused the fibroelastoma to obstruct the right coronary artery ostium causing transient obstruction to the right coronary artery.

Disclosures
None.

References

Figure 1. A, ECG showing inferoposterior ST elevation. B through D, Basal, mid, and apical perfusion images demonstrating pronounced hypoperfusion of gadolinium to the RCA territory seen as black, representing areas not perfused by gadolinium, which has the effect of shortening T1 values. RCA indicates right coronary artery.
Figure 2. A through C, Transesophageal echocardiogram showing a 1.5-cm homogenous pedunculated mass attached to aortic valve. D, Papillary fibroelastoma removed from the aortic valve.
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Supplemental Figure 1. LGE scar imaging during repeat CMR at 3 months. Extensive subendocardial scar is noted in the inferior territory, corresponding to the area of severe hypoperfusion during the initial prior stress CMR. Top left: 2-chamber view, top right: 3-chamber view, bottom left: short axis near base, bottom right: short axis near apex. Of note, retrospectively the subsequent mass is visible in the left ventricular outflow tract in the 3-chamber view.