Calcified Bicuspid Aortic Valve Mass Prolapsing Into the Left Main Coronary Artery

TAUQIR Y. GORAYA, MD, PhD; FAROUK MOOKADAM, MD; ANDRE C. LAPEYRE III, MD; RICHARD C. DALY, MD; HENRY D. TAZELAAR, MD; AND KYLE W. Klarich, MD

We report a case of a mobile calcific mass on the aortic valve that prolapsed into the left main coronary artery of a 51-year-old man. This case and a review of the literature suggest that calcific embolization to coronary arteries is a rare but possibly underrecognized complication of calcified degenerative or bicuspid aortic valves. This potentially catastrophic complication of calcified aortic valves needs to be suspected and recognized in clinical practice.


Embolism of calcareous material is an uncommon clinical event. Sources of calcareous emboli include calcified mural cardiac thrombi and aortic plaques. Bicuspid or degenerative calcified aortic valves are a rare source of calcareous emboli. Autopsy series of calcific aortic stenosis have identified calcareous embolic material in retinal, renal, intracerebral, and coronary arteries. Although embolic stroke, blindness, and limb ischemia have been reported frequently in patients with calcified aortic valves, clinical reports that document coronary calcareous emboli are rare.

We report an unusual case of a mobile calcified mass on a bicuspid aortic valve that prolapsed into the ostium of the left main coronary artery, threatening complete occlusion and sudden death. Emergent cardiac surgery retrieved the mass from the left main coronary artery while the mass was still attached to the aortic valve. Herein, we review the literature to highlight the need to recognize calcareous coronary emboli as a potential complication of calcific bicuspid aortic stenosis and, furthermore, point out that such lesions rarely resemble papillary fibroelastomas on echocardiography.

REPORT OF A CASE

A 51-year-old asymptomatic man was evaluated because of a known, mildly stenotic bicuspid aortic valve. He had a history of type 2 diabetes mellitus and a strong family history of coronary artery disease. His general physician had recently performed a screening exercise treadmill test, the result of which was markedly positive for ischemia at a low workload. Subsequently, an exercise sestamibi scan also showed an ischemic electrocardiographic (ECG) response at 7 minutes on the Bruce protocol, with a peak heart rate of 133 beats/min. Perfusion images showed a large area of apical, anterior, anteroapical, inferior, and inferoseptal ischemia. The patient was then admitted to Saint Marys Hospital, Rochester, Minn, for elective cardiac catheterization and transthoracic echocardiography to reevaluate the bicuspid aortic valve. On physical examination, the blood pressure was 130/80 mm Hg, and the heart rate was 66 beats/min. A grade II/VI mid-peaking ejection systolic murmur was noted. No diastolic murmur or carotid shudder was described. A resting ECG was normal.

Transthoracic echocardiography demonstrated normal left ventricular size and function (ejection fraction, 65%). A bicuspid aortic valve with mild stenosis (mean gradient, 23 mm Hg; aortic valve area, 1.3-1.5 cm²) and trivial aortic regurgitation were observed. Also noted was a highly mobile linear mass, approximately 2 cm long, arising from the aortic side of the aortic valve. Transesophageal echocardiography confirmed the presence of a pedunculated mass with a head that was almost 1 cm in diameter, arising from the aortic surface of the left coronary cusp of the aortic valve (Figure 1). The motion of the valve carried the mass to the vicinity of the sinotubular junction and the left main coronary artery. The differential diagnosis included papillary fibroelastoma, thrombus, vegetation, and calcification, the lattermost suggested by increased echocardiographic density. Because of the possible diagnosis of papillary fibroelastoma, surgical excision of the mass was planned following coronary angiography. At coronary angiography,
special precaution was taken because of the presence of the mobile mass in the vicinity of the left main coronary artery ostium. The initial injections were made in the aortic root. The left coronary cine-pictures showed a filling defect in the left main coronary artery, occupying 70% of the lumen (Figure 2). The filling defect was calcified and appeared to be attached to a stalk leading out of the proximal left main coronary artery into the aorta. The proximal left anterior descending coronary artery was obstructed 60% by multiple discrete lesions, and the distal part of the artery by a single 80% lesion. The other coronary arteries had only 30% to 40% discrete lesions.

The patient had no chest pain, and the ECG was normal both during and after coronary angiography. Prolapse of the pedunculated mass into the left main coronary artery was thought to pose a risk for distal embolization and complete occlusion of the distal left main coronary artery or one of its major branches. Therefore, an emergent cardiac operation was performed.

Intraoperative transesophageal echocardiography confirmed that the previously mobile mass, while still attached to the raphe of the conjoined left and noncoronary cusps of the aortic valve by its stalk, had prolapsed into the left main coronary artery (Figure 3). During the operation, the mass was removed successfully from the left main coronary artery by gently grasping the stalk, which was friable and broke easily near the aortic cusp (Figure 4). The aortic valve was replaced successfully with a 25-mm mechanical prosthesis (Sulzer Carbomedics, Austin, Tex). The patient also underwent grafting of the left internal mammary artery to the left anterior descending coronary artery.

Pathologic examination confirmed the diagnosis of a congenitally calcified bicuspid aortic valve. A firm, white tissue fragment measuring 1.5 × 0.4 × 0.2 cm (representing the prolapsing mass) was also submitted. Histologically, this was composed of elongate strips of calcium surrounded by recent bland thrombus and degenerating fibrin. There was no evidence of an underlying papillary fibroelastoma (Figure 5).

The patient had an uneventful postoperative course. Six months postoperatively, he was asymptomatic.

**DISCUSSION**

Embolic occlusion of the coronary arteries was first reported by Virchow in 1856. In 1933, Saphir noted that the commonest cause of coronary embolization was bacterial
vegetations on the mitral or aortic valve; other frequent sources included thrombi on an atherosclerotic or syphilitic lesion of the ascending aorta or intracardiac mural thrombi. Thrombi in peripheral veins (paradoxical emboli), papillary fibroelastomas, and neoplasms have since been recognized as rare causes of coronary embolism. In more recent years, prosthetic heart valves, dilated cardiomyopathy, and invasive procedures like coronary angiography have increasingly been associated with embolic occlusion of coronary arteries.

Calcific Coronary Emboli

Initial observations of calcific embolization to coronary arteries were made in autopsy series. Soulie et al described epicardial coronary embolization in 18 (20%) of 81 autopsy cases with calcific aortic stenosis. Holley and colleagues reported autopsy findings from a series of 163 patients with calcific aortic stenosis. Of these patients, 31 (19%) had a total of 45 embolic events. Ten patients (6%) had embolization of a major coronary artery, and 18 (11%) had embolization of minor coronary arteries. Histologic evidence of myocardial infarction was noted in only 6 of these 28 cases, all of which were thought to have been clinically silent. Although reports of clinical events resulting from calcific embolic occlusion of coronary arteries have been rare, there are numerous descriptions of neurologic, retinal, or peripheral arterial occlusion from calcific embolism in patients with degenerative or bicuspid calcific aortic stenosis.

In 1950, Moragues et al first described a patient with an anterior myocardial infarction due to an embolus from a calcified aortic valve. Mansur et al reported a case of acute inferolateral myocardial infarction in the setting of calcific aortic stenosis. Embolic occlusion of a distal branch of the left circumflex coronary artery was demonstrated by coronary angiography. The calcific nature of the embolus (and the aortic valve as the source of the embolus) was assumed but could not be confirmed. Salka and colleagues were the first to report a well-documented case of calcific embolization to a coronary artery in a patient with a calcified bicuspid aortic valve. They retrieved a calcific embolus from the second marginal branch of the left circumflex coronary artery during aortic valve surgery. Pathologic examination of the excised aortic valve showed a raw surface, which was assumed to be the site of origin of the calcific embolus.

To our knowledge, this is the first documentation of a calcified intracoronary mass that was still attached to the aortic valve at the time of diagnosis and surgery. The intracoronary mass was identified as a calcified tissue fragment arising from the conjoined raphe of the bicuspid aortic valve. Therefore, this case provides direct confirmation of the previously assumed origin of calcareous emboli from calcified aortic valves.

Differentiation From Papillary Fibroelastoma

In our patient, the echocardiographic appearance of the mobile pedunculated mass arising from the aorta was reminiscent of a papillary fibroelastoma. Indeed, the decision for surgical excision was made on the basis of a provisional diagnosis of papillary fibroelastoma. These benign tumors are a well-documented cause of cerebral, retinal, and coronary embolization. Papillary fibroelastomas arising from the aortic valve have been implicated in sudden death in otherwise healthy young persons by causing transient or complete obstruction of the ostium of the right coronary artery.

Papillary fibroelastoma is the third most common type of primary cardiac tumor, with more than 80% arising from
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Coronary Angiography and Embolization

While air and thrombotic emboli to the coronary arteries are well-recognized complications of coronary angiography, reports of calcareous coronary embolization as a complication of this procedure have been rare. In 1962, Arvidsson et al reported a case of coronary embolization with presumed calcareous material in a patient with heavily calcified aortic stenosis undergoing retrograde left ventricular angiography. Coronary embolization with calcareous material has also been reported as a rare complication of percutaneous aortic balloon valvuloplasty for severe calcific aortic stenosis. Deligonul and colleagues reported a case of acute lateral myocardial infarction during such a procedure. Subsequent coronary angiography revealed the appearance of an abrupt cutoff of the obtuse marginal branch of the left circumflex coronary artery.

In our case, the decision to proceed to surgical resection of the aortic mass had already been made, and angiography was performed only to define coronary anatomy. The angiographers had been apprised of the mass on the aortic valve and its proximity to the ostium of the left main coronary artery. Despite all possible precautions, the mass was noted, on the initial test injection in the aortic root, to have prolapsed into the left main artery. It is not possible to determine if this prolapse was caused by coronary angiography. An intermittent spontaneous occlusion of the left main coronary artery by the lesion could potentially explain the observed discordance between the severity of the underlying atherosclerotic coronary artery disease and the extent of perfusion defect noted on the previous exercise sestamibi scan. Nevertheless, this and other reports of calcific coronary embolization in patients with calcified aortic valves highlight the need to exercise extreme caution during invasive catheter-based procedures in this setting.

Unanswered Questions

Our report raises several questions. Angina pectoris is a frequent symptom in patients with severe aortic stenosis. Angina in the absence of coronary artery disease is assumed to result from severe left ventricular hypertrophy.
and the consequent supply-demand mismatch. In view of our case findings and those of Salka et al., calcific emboli should be considered as a cause of angina in this setting. Intermittent prolapse of the calcific mass into the ostium of the left main coronary artery of our patient may have produced the transient ischemia observed on the treadmill and the exercise nuclear scans. The incidence of sudden death reportedly is increased in patients with severe aortic stenosis. Although such deaths generally are assumed to have an arrhythmic or vasodepressor cause, embolic occlusion of a major coronary artery may explain some of these sudden catastrophic events.

REFERENCES