Papillary fibroelastoma on the aortic valve presenting as multiple cardiac arrests from electrical storm due to ischemia in patient without previous cardiac history

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Introduction

The prevalence of primary cardiac tumors varies from 0.02% to 0.45%.[1] Cardiac papillary fibroelastoma (PFE) is a benign endocardial papilloma. It is considered the second most prevalent primary cardiac tumor after myxoma and the most prevalent valvular tumor.[2]

The typical clinical presentation is either cardiac or neurologic, including transient ischemic attack, stroke, ischemic heart disease, sudden death, ventricular arrhythmia, heart failure, syncope, and blindness.[3]

We report a rare case of PFE on the aortic valve in a patient with no previous cardiac history who presented with multiple cardiac arrests from electrical storm despite intravenous antiarrhythmic drugs that resolved after surgical removal of the tumor.

Case report

The patient was a 64-year-old woman with no previous cardiac history and no family history of sudden cardiac death who had out-of-hospital cardiac arrests from polymorphic ventricular tachycardia (VT) requiring shock as noted by emergency medical services. She had return of spontaneous circulation (ROSC). En route to a community hospital she continued to experience multiple cardiac arrests that required multiple shocks. Each of these episodes was followed by ROSC between episodes. These episodes continued in the emergency room (ER) despite continuous infusion of intravenous amiodarone and lidocaine. Electrocardiography after ROSC showed atrial fibrillation, ST elevation in leads aVR and aVL, and diffuse ST depression in most of the other leads (Figure 1). The patient had another episode of cardiac arrest from VT that degenerated to ventricular fibrillation, which required another shock (Figure 2). Serum electrolytes were normal. Emergent point-of-care ultrasound showed normal left ventricular ejection fraction and no wall-motion abnormality at rest. Because of the emergency nature of the patient's condition, only a subcostal view on ultrasound was performed and did not show any mass. Emergency cardiac catheterization revealed normal coronaries and normal left ventriculogram. The patient continued to have intermittent VT, and the interventionist decided to perform aortic root angiography, which showed a filling defect in the aortic root with a masslike structure intermittently occluding the left main coronary artery and then coming out of the ostium of the left main coronary artery to allow blood flow (Figure 3). Cardiac catheterization film video showed the PFE occluding and then coming out of the ostium of the left main coronary artery (Supplemental Video 1). This was thought to be PFE on the aortic valve.

The patient was transferred to our facility for cardiothoracic evaluation and surgical removal of the suspected mass. She experienced another cardiac arrest en route to our facility and was again successfully resuscitated. The mass was removed emergently by cardiothoracic surgery. The mass measured 2 cm × 2 cm and was attached to the commissure of the left and noncoronary cusps. The VT episodes completely resolved after surgery, with no recurrences. Postoperative transthoracic echocardiography showed normal ejection fraction, trileaflet aortic valve of normal thickness, and normal aortic valve leaflet excursion with evidence of mild aortic regurgitation. Histopathologic report confirmed the mass to be PFE. Hematoxylin and eosin stain showed mucopolysaccharide matrix with typical filiform projections (Supplemental Figure 1). The macroscopic picture of the mass is shown in Supplemental Figure 2. The patient was

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KEY TEACHING POINTS

- Never give up. Keep searching for the etiology even when your normal search did not yield any result.
- When you see patients, always have a broad differential in mind, then narrow it down with history, physical examination, and other investigations.
- Common diagnoses are common, but also know that uncommon ones, though uncommon, do exist.
- The decision to perform a root aortogram when the initial coronary angiogram and left ventriculogram did not reveal a culprit is instructive.
- Transfer the patient to a more specialized center when you do not have the expertise or required setup to care for the patient in your facility.
- Papillary fibroelastoma is increasingly being diagnosed because of use of more imaging in clinical practice. This tumor, if found, should be removed to prevent serious complications, as occurred in our patient. Even though the tumor is benign, it can cause fatal complications.

discharged in stable condition with recommendation for outpatient follow-up with a cardiologist.

Discussion

Primary cardiac tumors are rare and have a very low prevalence based on 22 large autopsy studies.[4] They are divided into primary metastatic tumors and intracardiac tumors originating from infradiaphragmatic organs.[5] Cardiac PFE is a benign endocardial papilloma that is common in the sixth to eighth decades of life.[3] The aortic valve is most commonly involved (prevalence between 35% and 63%), followed by the mitral valve (9%–35%), tricuspid valve (6%–15%), and pulmonary valve (0.5%–8%).[3]

Although PFE is considered histologically benign, it can cause life-threatening complications such as embolic phenomena, which range from transient ischemic attack and amaurosis fugax to acute ischemic stroke, valvular dysfunction, ventricular fibrillation, and sudden death.[6] The tumor can occlude the coronary ostia or embolize into the coronary vessels, resulting in myocardial infarction, atypical angina, or VT.[7] The diagnosis is usually made incidentally in asymptomatic patients who undergo echocardiography for other reasons, or the tumor can present with any of the complications previously mentioned.[7] Although transthoracic echocardiography can adequately screen for PFE, transesophageal echocardiography is currently the preferred method because of its high resolution and optimal imaging capabilities.[7]

A study on a series of patients with PFE found that PFE can be identified by echocardiography with a high degree of certainty; is associated with embolic events in a high percentage of patients; does not cause significant valvular dysfunction despite its tendency to be located on the valvular endocardium; tends to occur in areas of endocardial damage; and is not associated with recurrence of embolic phenomena after surgical removal.[8] On echocardiography, PFE usually appears pedunculated and mobile, with a homogeneous speckled pattern and characteristic stippling along its edges that correlates with the papillary projections on the surface of the tumor.[9] A retrospective study on patients with cardiac PFE found typical echocardiographic features of PFE: the tumor is round, oval, or irregular in appearance, with well-demarcated borders and a homogeneous texture; most PFEs are small (99% were <20 mm in the largest

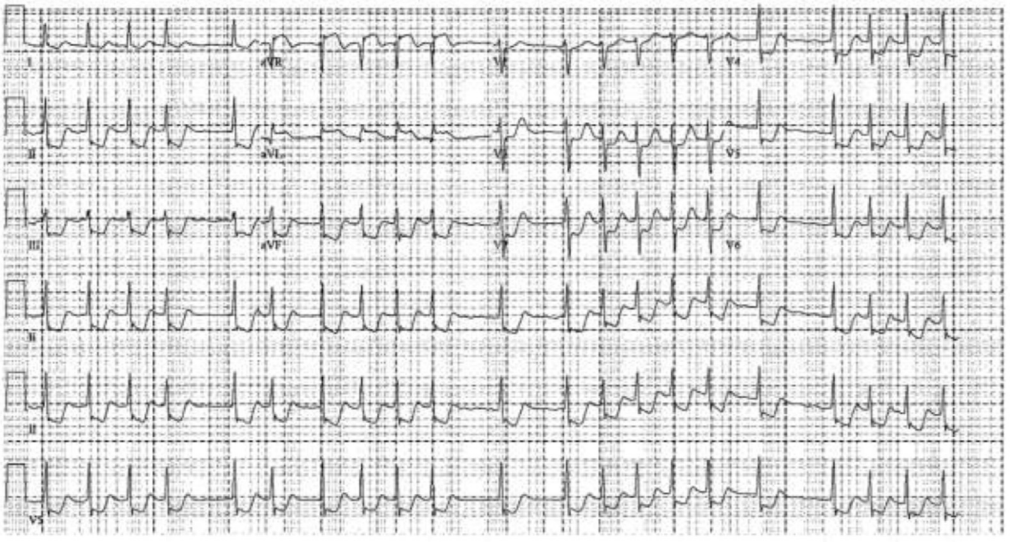

Figure 1 Twelve-lead electrocardiogram showing ST elevation in leads aVR and aVL with diffuse ST depression in the other leads.

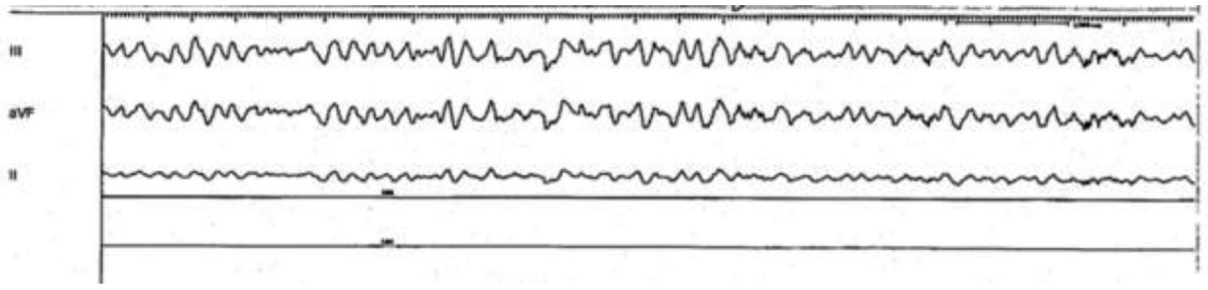

Figure 2 Strip showing ventricular fibrillation that degenerated from polymorphic ventricular tachycardia.

dimension); nearly half of PFEs had small stalks, and those with stalks were mobile; and PFEs may be single or multiple lesions and are most often associated with cardiac valvular disease.[10] In our case, aortic root angiography showed a filling defect in the aortic root with a masslike structure intermittently occluding the left main coronary artery.

Management of PFE depends on its clinical presentation. Patients who are asymptomatic with a small tumor (diameter <1 cm) that is sessile, fixed, and has no evidence of impingement on the coronary ostia usually require regular follow-up with serial imaging studies. Surgery should be offered only if the tumor increases in size or if the tumor is mobile or pedunculated.[11] Patients who are asymptomatic in whom the tumor was discovered incidentally during cardiac surgery for other reasons should be offered surgical resection because there will be no additional surgical risk.[12] Patients who are experiencing serious complications such as embolic events, myocardial infarction, or VT should undergo surgical resection.[12] Medical management with oral anticoagulation has been suggested for patients with smaller, nonmobile tumors and for those in whom valve-sparing surgery is not possible. This approach was associated with 50% tumor-related mortality in 1 study and has not been universally accepted.[9] It has been recommended that once the diagnosis of PFE is established, prophylactic anticoagulation therapy should be initiated to prevent the occurrence of any thrombi until surgical resection is accomplished.[12] More than 80% of cardiac PFEs can be treated with shave excision. A study conducted on patients who underwent shave excision noted no recurrence after the procedure.[13]

Ikegami and colleagues[14] analyzed 21 cases of PFE on the aortic valve. In their study, the 1 patient who presented with cardiac arrest had a known history of PFE, unlike our own patient, who did not have a previous diagnosis of PFE. Their patient had only 1 episode of cardiac arrest, unlike our patient, who had multiple episodes. Bussani and Silvestri[15] reported a case of sudden death in a woman with PFE on the aortic valve that chronically occluded the right coronary ostium. The patient had nonspecific ST-T wave changes on electrocardiography and was asymptomatic before the event. The PFE was later discovered at autopsy.

The mechanism of ventricular arrhythmia in our patient is thought to be ischemia from the tumor blocking the left main coronary artery and preventing normal coronary blood flow. Because the tumor intermittently occluded the left main coronary artery and was not a permanent occlusion, the patient had an arrhythmia-free period and ROSC between episodes, with the arrhythmia-free period coinciding with normal coronary blood flow in the left main coronary artery. The size of the tumor is such that it could have been seen on full echocardiogram. However, the mass was not seen on the subcostal point-of-care ultrasound. It is important to note potential complications from placing a pigtail catheter for aortography include embolization of a fragment or movement of the mass, which did not occur in our patient. It would have been important to rule out acute type A aortic dissection, which also could cause coronary occlusion; however, this was unlikely in our patient because she was stable between cardiac arrests. Causes of ventricular arrhythmia other than ischemia should always be included in the differential diagnosis. Of note, in this case the

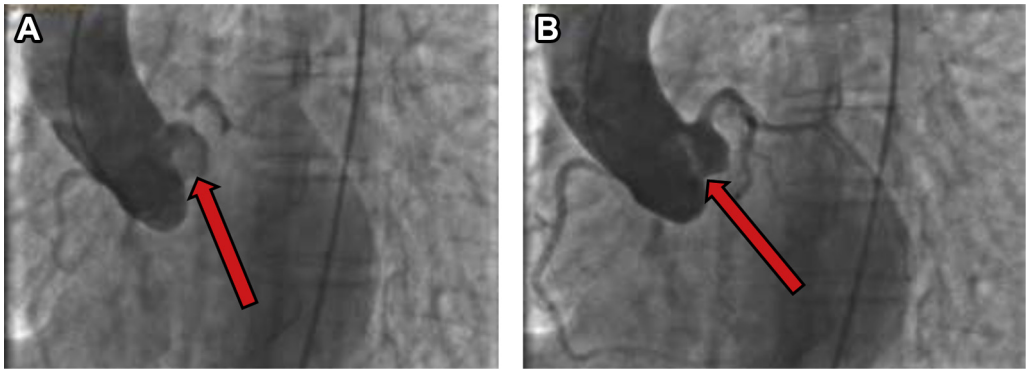

Figure 3 Aortic root angiogram showing the papillary fibroelastoma obstructing the left main coronary artery (**A**) but intermittently not obstructing the artery (**B**).

decision to perform root aortography after the initial coronary angiogram did not reveal a culprit was instructive.

Conclusion

Although PFE is considered a benign tumor, surgery is increasingly being considered a form of treatment because of its potential serious complications. Once PFE is suspected, we recommend performing transthoracic echocardiography and early referral for consideration of surgical removal. Asymptomatic patients do not necessarily require surgical removal unless the tumor is mobile. The recommendations regarding anticoagulation and surgery are not based on randomized controlled trials.

Our case emphasizes the need to continue searching for the diagnosis in a patient with multiple cardiac arrests from electrical storm when the electrocardiogram recorded between cardiac arrests shows evidence of significant ischemia but cardiac catheterization of the coronary arteries shows no obstructive disease to explain the arrhythmia. Other causes of ventricular storm should also be considered. Other primary electrical disease resulting in electrical storm can be considered in the absence of coronary artery disease and structural heart disease (eg, long QT syndrome, catecholaminergic polymorphic VT, Brugada syndrome).

Appendix

Supplementary data

Supplementary data associated with this article can be found in the online version at https://doi.org/10.1016/j.hrcr.2018.11.013.

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