A Case of Submitral Aneurysm Presenting with Severe Mitral Regurgitation and Shock in an African Male Patient with Rheumatic Heart Disease

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INTRODUCTION

Submitral aneurysm (SMA) is a rare cardiac pathology. Etiology is secondary to either an inflammatory process or congenital weakness in the fibrous annulus of the valve. Known secondary inflammatory causes include infective endocarditis (IE), rheumatic carditis, HIV infection, tuberculosis, and Takayasu arteritis. It has been described predominantly in the African population. Presentation varies among patients. The most common presentation is mitral regurgitation and heart failure. Other reported presentations include systemic thromboembolism, ventricular wall rupture, cardiogenic shock, ischemia in the left circumflex coronary artery territory, and ventricular arrhythmias.

CASE PRESENTATION

We present a case of a 50-year-old male patient who had a remote diagnosis of rheumatic heart disease. The patient presented to our institute with progressive shortness of breath. His temperature and blood pressure were within normal limits, but his oxygen saturation SpO2 was 89%. The laboratory investigations revealed microcytic anemia, mildly abnormal kidney function test, and elevated pro-brain natriuretic peptide of 694 pg/mL. Transthoracic echocardiography (TTE) showed mild to moderate rheumatic aortic regurgitation and mildly reduced left ventricle (LV) systolic function with ejection fraction of 50%. Anti-failure treatments were initiated. The patient’s clinical condition has improved, and the patient was discharged with a follow-up plan in the cardiology clinic after 1 month. Unfortunately, the patient was lost to follow-up and presented to our emergency department 3 months later with fever and progressive heart failure symptoms.

Laboratory investigations were performed and were significant for leukocytosis and neutrophilia. Transthoracic echocardiography was performed and revealed severe mitral and aortic valve regurgitation, thickening of the mitral valve with a small bright fixed mass attached to the mitral annulus (Figure 1), and severely reduced left ventricular (LV) ejection fraction of 20%. A multidisciplinary team meeting was conducted, and the consensus was reached to treat the possibility of IE based on the clinical presentation and the mass that was noted in the echo study. Empirical antibiotics were started and were to be adjusted after the blood culture result became available. All blood culture results were negative for bacterial growth. During his hospital stay, his condition continued to deteriorate and he became more hypoxic, requiring mechanical ventilation. The patient was also tested for COVID-19, MERS COV, and H1N1, and all results were negative. Upon a second review of the TTE, a perforation in the mitral annulus was suspected with the formation of a pouch that opens into the LV cavity (Figure 2, Video 1). A transesophageal echocardiogram (TEE) was then performed and showed a perforated noncoronary cusp with severe aortic regurgitation; in addition, a pulsatile pouch was noted above the posterior mitral leaflet (Figure 3, Videos 2 and 3). Submitral aneurysm was also identified in the midesophageal view in a nonstandard off-axis rotation with to-and-from flow noted with color Doppler (Figure 4, Videos 4 and 5). Three-dimensional TEE confirmed the presence of SMA at the inferomedial part of the mitral annulus (Figure 5, Videos 6 and 7). A multidisciplinary decision was made to accept the high surgical risk and to surgically replace both the aortic and the mitral valves, as well as repair the mitral annulus. A coronary angiogram was done before the surgery, which showed no significant coronary artery disease.

During surgery, a standard aortotomy was done to expose the aortic valve. The aortic valve was thickened and retracted with no evidence of vegetation or abscess. The aortic valve was replaced with a size 23 mm mechanical valve. The mitral valve was exposed using a left atriotomy approach. Both mitral valve leaflets were thickened and retracted. The mitral valve annulus was found to have a separation from the LV musculature at the level of P3 with the presence of the SMA at the annulus separation site. The opening of the aneurysm was thickened, indicating chronicity. When the mitral valve leaflets were excised, deep pledgeted sutures were taken to repair the separation site. The mitral valve was replaced with a size 29 mm mechanical valve. Postoperatively, the patient was severely coagulopathic and vasoplegic and was transferred to the intensive care unit with an open chest. Histopathology specimens of both mitral and aortic tissue revealed degenerative valvopathy.

Unfortunately, the postoperative course was complicated, and eventually the patient died due to multiorgan failure.

DISCUSSION

In our case, the patient was African in origin and presented initially with heart failure symptoms. Given the patient’s remote history of
rheumatic heart disease and his aortic and mitral valve degeneration, the etiology of SMA in this patient is most likely rheumatic inflammation. Submitral aneurysm was most likely present in our patient during his initial presentation, which was not picked up due to the rarity of the disease and low index of suspicion, as well as the limited visualization of the TTE compared to the TEE. The finding of a bright mass on the anterior mitral leaflet was presumed to be a vegetation; however, other differential diagnoses include healed vegetation, leaflet perforation, and calcified secondary chord. In addition, the presentation of heart failure symptoms that were not explained by the degree of the initial valve disease should have triggered a TEE study.

**VIDEO HIGHLIGHTS**

**Video 1:** Apical two-chamber view illustrating an opening in the mitral annulus, which opens into a pouch.

**Video 2:** Midesophageal view showing the aortic valve in the long axis and showing a dropout in the noncoronary cusp suggestive of a perforation. There is a pulsatile pouch noted above the posterior mitral leaflet.

**Video 3:** Midesophageal view with color Doppler showing the aortic valve in the long axis with aortic valve perforation at the noncoronary cusp and severe aortic regurgitation.

**Video 4:** Midesophageal view with off-axis rotation illustrating SMA originating behind the posterior mitral leaflet.

**Video 5:** Midesophageal view with off-axis rotation illustrating SMA with to-and-from flow identified by color Doppler.

**Video 6:** Three-dimensional clip of the mitral valve en face view from the LV perspective illustrating the SMA relation to the adjacent structure.

**Video 7:** Three-dimensional clip of the mitral valve en face view from the left atrial perspective illustrating the SMA relation to the adjacent structure.

*View the video content online at www.cvcasejournal.com.*
The basic pathology of SMA is a separation of the posterior mitral annulus from the LV musculature creating an outpouching in the LV wall. This process will distort the mitral valve and subvalvular supporting apparatus and results in restriction of the mitral leaflets and malcoaptation, resulting in a significant mitral regurgitation. Genetic predisposition has been suggested based on the observation that SMA occurs predominantly in the African race. Also, Nayak et al. have found that the membranous submitral curtain of the mitral valve in 45 out of 75 normal heart autopsies is a potentially weak area through which congenital SMA can occur. Submitral aneurysm occurs solely near the posterior mitral leaflet. Du Toit et al. have classified the aneurysm into three types: type I, single localized neck; type II, multiple necks; and type III, involvement of the entire posterior mitral annulus. The most important differential diagnosis of the SMA is a left atrium pseudoaneurysm (LAP), a rare cardiac pathology that shares the same management plan by surgical intervention with SMA. The most important differentiating feature between LAP and SMA is that LAP communicates with the left atrium, while SMA communicates with the LV. Surgical repair is considered the main treatment option and was first reported in 1963 by Shrire and Barnard. There is no clear timing for surgery; however, if mitral regurgitation is present, the standard guidelines recommendations for mitral regurgitation intervention should apply. Earlier intervention might be a wise strategy, since we know that many reported cases, including ours, had a fast progression to cardiogenic shock and poor outcome.

CONCLUSION

Submitral aneurysm can be congenital or secondary to an inflammatory process. It is commonly described among the African population. The most common presentation is moderate to severe mitral valve regurgitation that an early diagnosis and treatment may prevent. However, a high index of suspicion is required, particularly if the clinical condition does not match the initial cardiac imaging result. Transthoracic echocardiogram, TEE, and computed tomography scans are important tools for the diagnosis of SMA and the identification of its complications. A consensus of a multidisciplinary team for the appropriate surgical planning is the mainstay treatment. We believe that surgical treatment should be performed once the defect is identified to prevent complications and improve outcomes.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2021.04.001.

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