Pseudoaneurysm of the Mitral-Aortic Intervalvular Fibrosa: A Case Series with Literature Review

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INTRODUCTION

Pseudoaneurysm of the mitral-aortic intervalvular fibrosa (P-MAIVF) was traditionally considered a rare, life-threatening sequela of endocarditis or valve surgery.

Pseudoaneurysm is an outpouching where the aortic and mitral valves meet and forms between the aorta and the left atrium with superior and posterior extension. It is contiguous with the left ventricular outflow tract, which differentiates it from an abscess.[1,2] In this article we present three consecutive cases and one highly suspicious case of P-MAIVF diagnosed at our institution using transesophageal echocardiography and discusses the causes, presentation, diagnosis, and management of that entity.

CASE 1 PRESENTATION

A 56-year-old white man presented with fevers and blood cultures positive for β-hemolytic *Streptococcus* group B.

Ten months prior, he was diagnosed with aortic valve endocarditis and an aortic root abscess, due to *Streptococcus agalactiae* infection. He was managed with aortic valve replacement with a 19-mm St. Jude mechanical prosthesis and patch reconstruction of the aortic root abscess. Transthoracic echocardiography upon discharge demonstrated a well-seated mechanical valve and a normal aortic root.

On his current presentation, initial transthoracic echocardiography showed mild aortic prosthesis regurgitation with no vegetation or evidence of perivalvular infection. Subsequently, transesophageal echocardiography revealed the presence of P-MAIVF, approximately 1.2 cm at its largest dimension (Figure 1). The pseudoaneurysm extended superiorly and posteriorly between the aorta, the left atrium, and the pulmonary artery and exhibited systolic expansion and diastolic collapse (Videos 1 and 2). There was no communication with the left atrium or the aorta and no signs of prosthetic valve dysfunction. Because of hemodynamic and electrical stability, the patient was treated with penicillin G and gentamycin for 1 month and subsequently underwent repair of the pseudoaneurysm with a bovine patch and received a new 19-mm mechanical aortic valve. He had an uneventful recovery and was discharged home 1 week later.

CASE 2 PRESENTATION

An 84-year-old Caucasian man with diabetes and chronic stage IV kidney disease was referred for transesophageal echocardiography for evaluation of endocarditis.

Eighteen months previously, he had undergone single vessel (left internal mammary artery to left anterior descending coronary artery) coronary artery bypass graft surgery combined with aortic valve replacement with a 23-mm Magna bovine pericardial valve for severe aortic stenosis. Results of postoperative transthoracic echocardiography were unremarkable.

One month before his current presentation, he developed fevers and malaise. He was found to have transient staphylococcal bacteremia and was treated with 2 weeks of antibiotics. The fevers resolved, but the patient developed dyspnea on exertion and lower extremity edema. He was then referred for transesophageal echocardiography, which showed a well-seated aortic bioprosthesis with markedly thickened cusps, no vegetation, and mild transvalvular regurgitation. The images were highly suggestive of P-MAIVF without fistula formation or compression of adjacent structures (Figures 2 and 3, Videos 3–6). After discussion of the risks and benefits of complex surgical intervention in an elderly patient, he and his family opted against it. He was managed with chronic suppressive intravenous antibiotics with close clinical and imaging follow-up. He was alive 15 months after the diagnosis.

CASE 3 PRESENTATION

A 67-year-old Caucasian man was referred for evaluation of dyspnea on exertion and weight gain of 1 week's duration. He had a history of two-vessel bypass surgery in 2007.

Five months before the current presentation, he was admitted with fevers and disorientation. His blood cultures grew β-hemolytic *Streptococcus* group G. Transesophageal echocardiography at that time revealed a large vegetation on the noncoronary cusp of the aortic valve (1.7 $\times$ 0.2 cm) and an aortic root abscess. He underwent aortic valve replacement with a 23-mm bovine pericardial valve and patch repair of the aortic abscess. He was discharged 1 week later and completed a 2-week course of intravenous ceftriaxone.

On his current presentation, transesophageal echocardiography revealed P-MAIVF with characteristic systolic expansion and diastolic collapse, which measured 3.5 cm at its largest dimension. The pseudoaneurysm demonstrated a fistula to the left atrium (Figures 4–6, Videos 7–11). Multiple blood cultures at that time were negative, and the patient had no fevers, but he was restarted prophylactically

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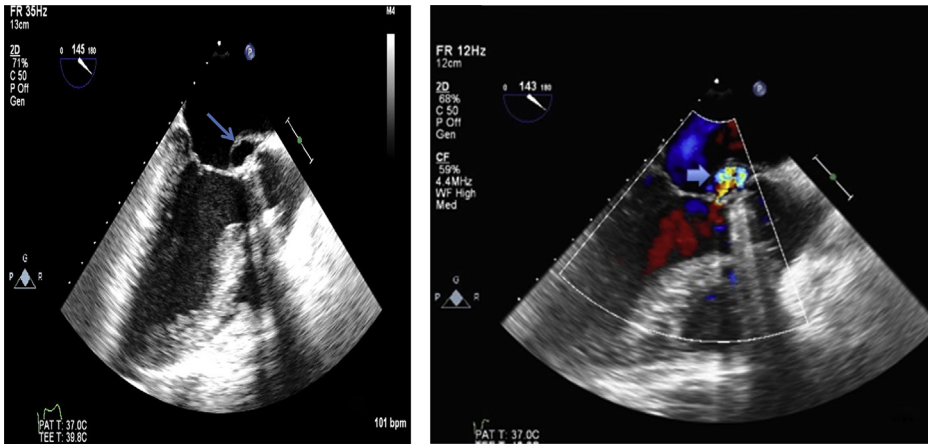

Figure 1 Transesophageal echocardiogram, midesophageal long-axis views without color (*left*) and with color (*right*), demonstrating P-MAIVF. Both images were obtained during systole and show the characteristic systolic expansion of the pseudoaneurysm (*arrows*). The color Doppler image demonstrates the pseudoaneurysm's communication with the left ventricular outflow tract (*thick arrow*). A mechanical prosthesis in the aortic position is present.

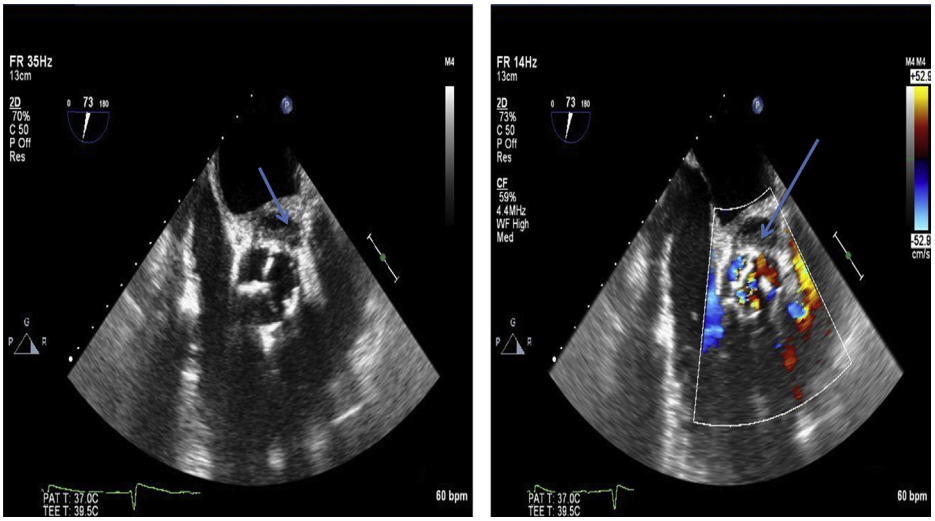

Figure 2 Transesophageal echocardiogram, midesophageal short-axis views showing the pseudoaneurysm (*arrows*) at the level of the aortic valve without color (*left*) and with color (*right*).

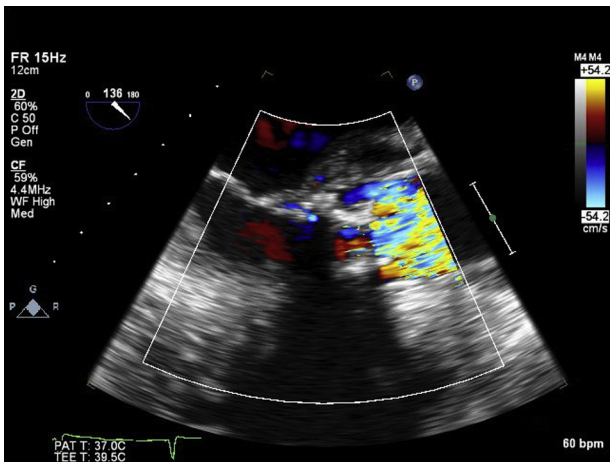

Figure 3 Image was obtained at the end of systole and demonstrates a thickened bioprosthesis in the aortic position with a possible pseudoaneurysm between the left atrium, the aorta, and the pulmonary artery.

on ceftriaxone. He underwent a third cardiac surgical procedure at an outside institution per the family's wishes, which by report involved repeat aortic valve replacement with a tissue valve and repair of the pseudoaneurysm with an unremarkable postoperative course. After discharge, the patient had alleviation of his heart failure symptoms and was enrolled in cardiac rehabilitation. A follow-up transesophageal study 4 months later showed persistent P-MAIVF. Because of his disease complexity and multiple prior sternotomies, he was deemed at very high risk for a repeat intervention. He was managed with close clinical and echocardiographic follow-up. He has remained symptom free to date.

CASE 4 PRESENTATION

A 43-year-old African American woman presented with right upper extremity pain and was found to have a thrombus in the right axillary/brachial artery, for which she underwent thrombectomy. The patient had a history significant for end-stage renal disease requiring hemodialysis three times per week through an arteriovenous fistula

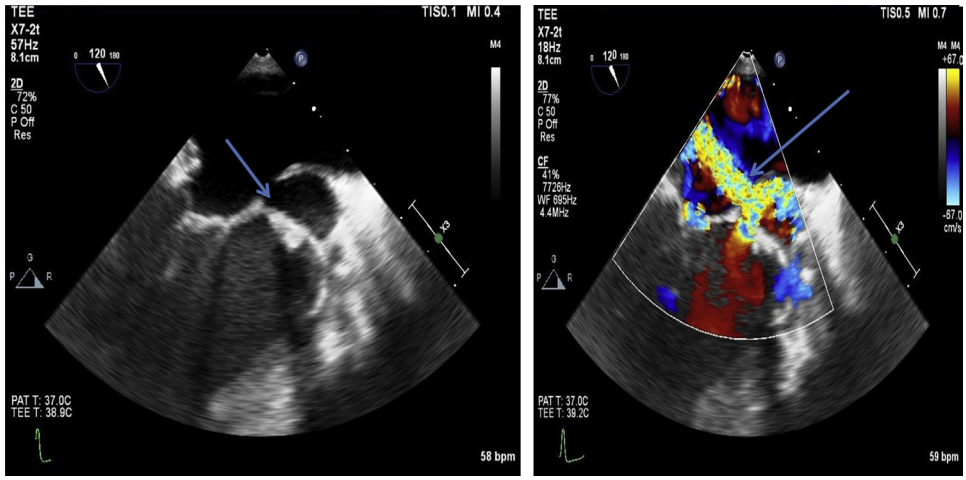

Figure 4 Transesophageal echocardiogram, midesophageal zoomed long-axis views showing the pseudoaneurysm (*arrows*) without color (*left*) and with color (*right*) in midsystole. The image on the *left* is a two-dimensional picture of the large P-MAIVF. On color Doppler, a fistula between the pseudoaneurysm and the left atrium was demonstrated (*arrow*). During systole, the blood moved from the left ventricle into the pseudoaneurysm and then the left atrium. There was no diastolic flow, as the pseudoaneurysm was not pressurized in diastole.

Figure 5 Three-dimensional representation of the pseudoaneurysm in systole in the short-axis view.

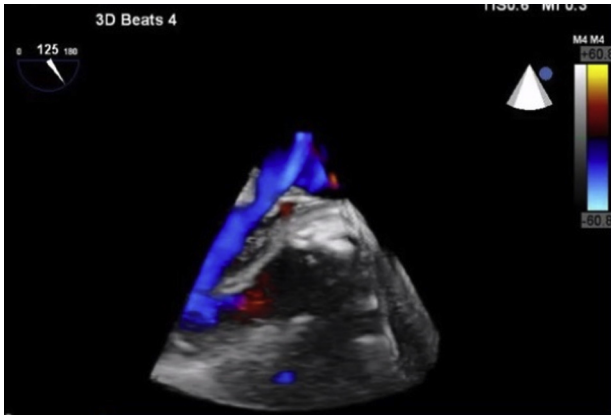

Figure 6 Three-dimensional long-axis view with color demonstrating the communication of the pseudoaneurysm with the left ventricular outflow tract obstruction.

in the right arm and two failed renal transplantations. Upon presentation she was afebrile, and blood cultures grew *Streptococcus epidermidis*. The same bacterium grew from the excised right upper arm thrombus. Transthoracic echocardiography showed normal biventricular systolic function and an 11.7 × 8 mm vegetation in the aortic valve with associated severe aortic regurgitation. Electrocardiography showed new first-degree atrioventricular block. Transesophageal echocardiography confirmed the presence of a large vegetation in the aortic valve with significant regurgitation. The mitral-aortic intervalvular fibrosa appeared thickened, consistent with an aortic root abscess, and demonstrated what appeared to be the beginning of pseudoaneurysm formation (Figure 7), which had a small communication to the aorta (Figure 8). The pseudoaneurysm demonstrated minimal pulsatility and minimal systolic expansion and diastolic collapse (Videos 12 and 13). The patient underwent surgery, which involved extensive debridement of the intervalvular fibrosa and aortic valve replacement with a 19-mm St. Jude's mechanical valve. After separation from cardioplegia, severe mitral regurgitation was noted, which was managed with a 26-mm Cosgrove annuloplasty ring, but moderate to severe mitral regurgitation persisted at the conclusion of the case. Unfortunately, 24 hours after surgery, the patient became unresponsive and apneic and developed pulseless electrical activity arrest. Despite prolonged resuscitation efforts, the patient expired.

DISCUSSION

Definitions

The mitral-aortic intervalvular fibrosa or aortomitral curtain is the fibrous, avascular region between the anterior leaflet of the mitral valve and the noncoronary cusp of the aortic valve.[3] It is prone to infection and injury, and as a result a pseudoaneurysm may develop. Eighty-nine cases were reported in the literature from 1966 to 2009.[1] Advances in imaging techniques and the widespread use of

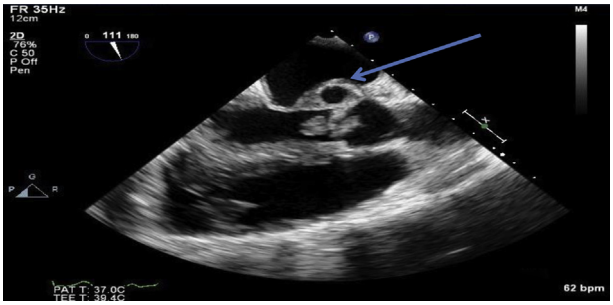

Figure 7 Transesophageal echocardiogram, midesophageal zoomed long-axis view without color. A large vegetation in the aortic valve prolapsing in the left ventricular outflow tract and P-MAIVF are visualized (*arrow*).

transesophageal echocardiography have led to its prompt recognition. As a result, 166 cases were reported by 2014, almost double the number in a span of 5 years.[4]

Etiology

Infection and surgical trauma are the most common causes of P-MAIVF.[1,2,5] History or active endocarditis, most commonly involving the aortic valve, is the number one association with pseudoaneurysm formation.[1,2] *Staphylococcus* and *Streptococcus* spp are the most common causative microorganisms.[1,2,5] Trauma from valve surgery without evidence of infection is the second most common cause of pseudoaneurysm formation.[1,2,5] That is usually a sequela of aortic valve replacement[6] but has been reported after ablation, repair of ventricular septal defects, and even cardiac catheterization.[5] The presence of aortic regurgitation appears to contribute to pseudoaneurysm formation.[7] A few cases of idiopathic pseudoaneurysms, with no identifiable causes, have been reported.[8]

Our four cases are in line with the discussed etiology of pseudoaneurysms. The first patient had active endocarditis of the aortic valve. Patients 2 and 4 had bacteremia without valve vegetation or destruction visualized by echocardiogram. Patient 3 initially presented with endocarditis of the aortic valve requiring aortic valve replacement. The pseudoaneurysm was found 3 months later with repeated negative blood cultures, but it was unclear if the pseudoaneurysm was present at the time of surgery or formed later secondary to infection. *Streptococcus* infection was present in two patients and *Staphylococcus* infection in one patient (Table 1).

Clinical Presentation

In the current case series, two patients presented with symptoms of endocarditis (fevers, embolic events, positive blood cultures), and two presented with heart failure symptoms (Table 1).

Symptoms and signs of active infection are the most common manifestation of that entity (40%), followed by heart failure symptoms (16%).[1] Cerebrovascular accidents and embolic events occur in 12% of cases and chest pain in 10%.[1] Approximately 10% of cases are asymptomatic, especially in the absence of complications and are incidentally discovered on routine imaging. Rare presentations include the development of a chest wall mass[1,2] and palpitations due to ventricular ectopy.[9]

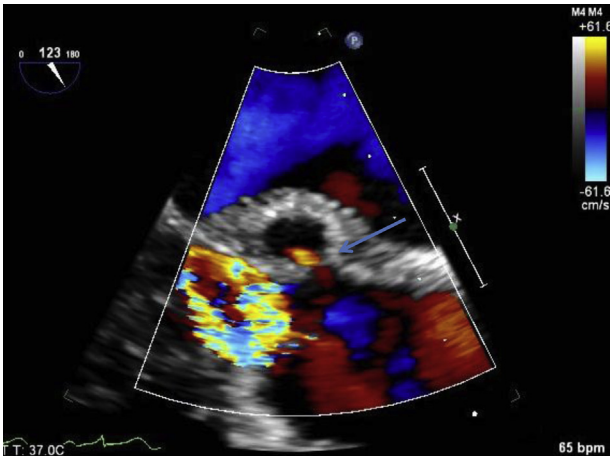

Figure 8 Transesophageal echocardiogram, midesophageal zoomed long-axis view with color demonstrating the pseudoaneurysm communicating with the aorta (*arrow*).

Complications

Compression of Adjacent Structures. The pseudoaneurysm can compress the coronary arteries, most commonly the left circumflex artery, causing angina.[1,2,5] It may compress the pulmonary artery, causing pulmonary hypertension.[1,2] Rarely, compression of the anterior mitral valve leaflet leads to mitral regurgitation.[1,2,5]

Fistula Formation. A fistula can develop between the left ventricular outflow tract and the left atrium or the aorta.[10] Patient 3 in our series demonstrated a fistula to the left atrium, and patient 4 demonstrated a communication to the aorta.

Rupture. The pseudoaneurysm may rupture into the pericardium, which is usually a fatal complication because of tamponade.[11] Rupture in the anterior wall has been reported and presented with a pulsatile chest wall mass.[1,2]

Thromboembolic Events. Cerebrovascular accidents and systemic embolization may follow clot formation in the pseudoaneurysm cavity.[1,2]

Diagnosis

Echocardiography is the major imaging modality for diagnosis and evaluation of complications of P-MAIVF.[12] Transesophageal echocardiography has superior sensitivity in the diagnosis of that entity compared with transthoracic echocardiography.[10]

Pseudoaneurysm presents as an echo-free pouch located between the anterior mitral valve leaflet and the posterior aortic root, exhibits systolic expansion and diastolic collapse, and communicates with the left ventricular outflow tract.[7,10] The blood during systole moves from the left ventricle into the pseudoaneurysm. There is no diastolic flow in the pseudoaneurysm, because during diastole, the blood flows back into the left ventricular outflow tract. Color Doppler demonstrates the to-and-fro (pulsatile) flow in the pseudoaneurysm and may also demonstrate the development of complications.[10,12] When a fistula develops between the left ventricular outflow tract and the aorta, the pulsatility of the pseudoaneurysm is less pronounced.[10] Pericardial effusion with or without tamponade may be demonstrated in case of pseudoaneurysm rupture into the pericardium. Three-dimensional echocardiography can clearly demonstrate the exact

Table 1 Summary of clinical and imaging characteristics and management strategies for the four patients

Patient	Gender	Age (y)	Diagnostic method	Aortic valve	Vegetation	Causative organism	Associated AR	Fistula	Surgery
1	Male	56	TEE	AVR, mechanical prosthesis	No vegetation	*Streptococcus* group G	Mild	—	AVR + P-MAIVF repair with a patch
2	Male	84	TEE	AVR, bioprosthesis	No vegetation	Reported *Staphylococcus* bacteremia	Mild	—	Refused surgery
3	Male	67	TEE	AVR, bioprosthesis	No vegetation	—	—	LA	AVR + P-MAIVF repair
4	Female	43	TEE	Native	Vegetation of the aortic valve	*Streptococcus epidermidis*	Severe	Aorta	AVR + extensive debridement of MAIVF

AR, Aortic regurgitation; *AVR*, aortic valve replacement; *LA*, left atrium; *MAIVF*, mitral-aortic interventricular fibrosa; *TEE*, transesophageal echocardiography.

location and size of the pseudoaneurysm, its relationship to adjacent structures, and the presence of complications.[13] Cardiac computed tomography and magnetic resonance imaging appear to be useful complementary imaging modalities to determine the pseudoaneurysm extent and its relationship with neighboring structures.[1,4] Transesophageal echocardiography diagnosed the pathologic condition in all four patients in this series (Table 1) because of technical limitations of transthoracic imaging.

Natural History

The natural history of uncomplicated P-MAIVF is not well known, because the diagnosis of the entity usually leads to surgery for treatment of complications or in asymptomatic patients to prevent complications.[1] Over the past few years, some cases of uncomplicated pseudoaneurysms were managed with watchful surveillance. Gin *et al.*[14] described three patients who had minimal change in pseudoaneurysm dimensions and were alive without surgery at the end of a 5-year follow-up period, except one patient who died at the age of 92 years of noncardiac causes. Similarly, Hasin *et al.*[15] presented two cases of large, uncomplicated pseudoaneurysms that were managed with watchful follow-up and had benign long-term clinical courses.

Management

Aortic valve replacement is performed in the majority of patients in conjunction with some type of pseudoaneurysm repair, which varies from simple closure of the aneurysm neck to closure using pericardial or synthetic graft material and even complete removal of the pseudoaneurysm from the left ventricular outflow tract.[1,2,5] Some centers advocate extensive debridement and replacement of the aortic valve, aortic root, and mitral-aortic intervalvular fibrosa with a homograft. In our case series, three patients underwent aortic valve replacement with pseudoaneurysm repair. Homografts were not used, because of surgeon's preference.

The presence of a complication and the rapid expansion of the pseudoaneurysm mandate surgical intervention. In our series, three patients underwent surgery, and one was managed conservatively because of advanced age and multiple comorbidities (Table 1).

Successful percutaneous and transapical closure cases have been reported, but this should be considered only when patients are not operative candidates given the risks of introducing foreign material at the site of a current infection.[2]

Clinical and echocardiographic monitoring of uncomplicated P-MAIVF may be an alternative in the management of high-risk surgical patients.[14,15]

CONCLUSION

P-MAIVF usually follows endocarditis or aortic valve surgery, and it has been increasingly recognized with the use of echocardiography, especially transesophageal echocardiography. It may cause symptoms or important complications that warrant timely diagnosis and surgical intervention. In this case series, two of four cases of asymptomatic P-MAIVF were managed without surgery, but surgical intervention is still the recommended management.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at http://dx.doi.org/10.1016/j.case.2017.07.001.

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