Infective Endocarditis of the Aortic Valve Complicated by Aorta–To–Pulmonary Artery Fistula

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INTRODUCTION

Infective endocarditis (IE) is a common entity in clinical practice, and echocardiography plays a key role in diagnosis, prognostication, and surveillance. In addition to determining the presence of vegetations, echocardiography is crucial in assessment for structural complications caused by IE and in decision making for surgical intervention. Intractable heart failure, embolization, and signs of uncontrolled infection such as abscess, false aneurysm, and fistula formation are indications for urgent cardiac surgery.[1] Although complications of uncontrolled infection are relatively uncommon, they can cause rapid clinical deterioration and increased morbidity. Additional hemodynamic data such as pulmonary artery pressure, cardiac output, and parameters of valve function obtained by echocardiography also assist in determining whether clinical deterioration in critically ill patients is due to complications from IE, with serial echocardiography aiding in the determination of prognosis and response to medical therapy. We present a case of IE with the rare complication of an ascending aorta–to–pulmonary artery fistula diagnosed by echocardiography and compare this with other causes of aortopulmonary connections.

CASE PRESENTATION

A 43-year-old Burundian man presented to a community hospital with a 1-month history of fevers, night sweats, and weight loss. He had a medical history of malaria, schistosomiasis, *Strongyloides* infections, α-thalassemia minor, chronic splenomegaly, and chronic pancytopenia. The schistosomiasis and *Strongyloides* infections were treated upon immigration to the United States. He was previously tested and found to be negative for tuberculosis and was treated before presentation at an urgent care facility for influenza, with no alleviation of his symptoms.

On initial laboratory findings at the community hospital, the patient's white blood cell count was 5.4×10^9/L, platelet count was 12×10^9/L, and hemoglobin was 4.7 g/dL. Blood cultures grew group B *Streptococcus*, and therapy with ceftriaxone was initiated. Transthoracic echocardiography (TTE) was performed and showed a vegetation measuring 8 mm on the aortic valve, mild aortic regurgitation, and thickening of the aortic root suggestive of an abscess. A workup for other infectious causes, including human immunodeficiency virus, atypical bacteria, and parasitic infection, was negative, and he was transferred to a tertiary care hospital for further management.

Physical examination on presentation to the tertiary care center revealed a heart rate of 83 beats/min, a blood pressure of 95/47 mm Hg, a temperature of 37.8°C, and oxygen saturation of 93% on room air. He was noted to be cachectic, with a jugular venous pressure at 8 cm above the sternal angle. On cardiopulmonary auscultation, there was a 4/5 early diastolic murmur at the left sternal border, a 2/6 holosystolic murmur at the apex radiating to the axilla, and bibasilar lung crackles. He was noted to have a distended abdomen with shifting dullness and palpable splenomegaly and 3 + pitting edema of both lower extremities, without peripheral stigmata of IE. Initial chest radiography showed mild interstitial edema (Figure 1).

The PR interval on electrocardiography at the tertiary care center was 188 msec (Figure 2), which had increased from 149 msec on initial community hospital electrocardiography.

An abdominal computed tomographic scan was performed, which showed hepatic cirrhosis, massive splenomegaly measuring 20 cm, and large-volume ascites with multiple esophageal varices. Abdominal paracentesis was negative for bacteria. No source of streptococcal infection was identified, and the patient did not use intravenous drugs, but his splenomegaly was thought to reflect splenic dysfunction, which in turn may have contributed to his susceptibility to infection with encapsulated bacteria such as *Streptococcus*.

Repeat TTE was performed on arrival at the tertiary care hospital 8 days after initial imaging to assess for progression of aortic root thickening given the change in duration of the PR interval. TTE showed multiple vegetations on the aortic valve with severe aortic regurgitation and perforation of the right coronary cusp (Videos 1 and 2). There was evidence of holodiastolic flow reversal in the thoracic and abdominal aorta (Figure 3). There was continuous flow by color Doppler interrogation from the aortic valve commissure between the right and left coronary cusps to the main pulmonary artery consistent with an aorta–to–pulmonary artery fistula (Figure 4, Videos 3 and 4). There was also a 16×10 mm vegetation attached to the wall of the main pulmonary artery (Videos 5 and 6). There was no clear evidence of an aortic root abscess. Left and right ventricular systolic function was normal. There was moderate mitral and tricuspid regurgitation, without vegetations on either valve. Transesophageal echocardiography (TEE) was considered to further assess valvular structures but deferred because of the patient's severe thrombocytopenia (repeat platelet counts ranged from 11×10^9/L to 29×10^9/L), esophageal varices, and clear demonstration of an indication for cardiac surgery on TTE.

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VIDEO HIGHLIGHTS

Video 1: Parasternal long-axis view demonstrating layered and mobile vegetations on multiple aortic leaflets, a coaptation gap between the leaflets in diastole, and diastolic fluttering of the anterior mitral leaflet suggestive of significant aortic regurgitation.

Video 2: Color Doppler on the aortic valve demonstrates a posteriorly directed jet of aortic regurgitation through the central coaptation point. There is a jet of aortic regurgitation anterior to the central coaptation point, which represents perforation of the right coronary cusp.

Video 3: Parasternal short-axis view demonstrating systolic and diastolic flow through a fistula located at the right and left coronary cusp commissure to the main pulmonary artery.

Video 4: Parasternal short-axis view demonstrating vegetations on the aortic valve and in the main pulmonary artery. In systole, a gap in the posterior wall at the base of the main pulmonary artery hints at the location of the fistula. A pericardial effusion is seen adjacent to the right atrium.

Video 5: Modified parasternal short-axis view better demonstrating the size of the vegetation attached to the wall of the main pulmonary artery.

Video 6: Parasternal long-axis view with focus on the pulmonic valve and demonstration of the vegetation in the main pulmonary artery.

Video 7: Short-axis view of the aortic valve on intraoperative TEE demonstrating vegetations on the aortic valve and continuous flow across the aortopulmonary fistula.

Video 8: Short-axis view of the aortic valve on intraoperative TEE after repair showing interval placement of a bioprosthetic aortic valve and resolution of the aortopulmonary communication.

Video 9: A PDA with flow from the descending thoracic aorta into the left pulmonary artery toward the pulmonic valve.

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Because of the patient's severe thrombocytopenia and liver cirrhosis, he was initially deemed too high risk for cardiac surgery. However, because of progressive clinical decompensation and heart failure, a multidisciplinary team discussion was undertaken with the patient, and a decision was made for high-risk cardiac surgical intervention after platelet transfusion. The patient underwent aortic valve replacement with a 21-mm bioprosthetic St. Jude Trifecta valve (St. Jude Medical, St. Paul, MN) and aortic root enlargement with bovine pericardium with intraoperative transesophageal echocardiographic guidance. A bioprosthesis was selected because of the patient's high bleeding risk due to severe thrombocytopenia and liver cirrhosis. The pulmonary artery was opened and debrided, and the fistula between the aorta and pulmonary artery was closed. Videos 7 and 8 demonstrate resolution of aortopulmonary flow after surgical repair.

Gross surgical findings included large vegetations on all leaflets of the trileaflet aortic valve, destruction of the right coronary sinus and adjacent annulus, and a vegetation in the main pulmonary artery originating from the site of the fistula. The patient's postoperative course was complicated by complete heart block requiring a permanent pacemaker but was otherwise uneventful. He was treated with a full 6-week course of ceftriaxone as an outpatient. Subsequent pathology identified abundant Gram-positive cocci in pairs and short chains on Gram stain of the aortic valve tissue, but there was no growth.

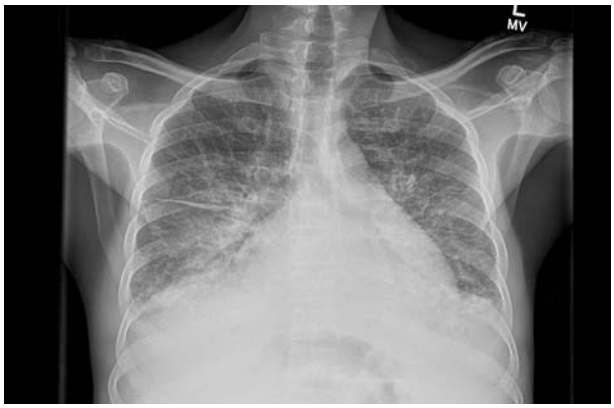

Figure 1 Anterior-posterior chest radiograph showing interstitial edema.

DISCUSSION

Acquired aorta–to–pulmonary artery communications are rare but can occur as fistulization from IE, as in the present case; secondary to rupture of a thoracic aortic aneurysm into the pulmonary artery[2]; as a complication of aortic dissection; as a complication of procedures performed on the aorta or pulmonic valve[3-5]; or as a ruptured congenital sinus of Valsalva aneurysm.[6] Balloon dilation of pulmonary arteries has also been reported to cause aortopulmonary fistulas, possibly due to contact between an anatomically abnormal or dilated pulmonary artery and aorta in patients with histories of pulmonary artery stents or suture lines from previous Ross or arterial switch procedures.[5] Physical examination findings of aortopulmonary fistula formation include a widened pulse pressure, a continuous cardiac murmur that has been described as "machinery-like" and sometimes localized to the left second and third intercostal spaces, and clinical signs of right ventricular volume overload due to the large degree of left-to-right shunting.[7-9] Similar to congenital aortopulmonary windows, patients with aortopulmonary fistulas can develop left ventricular dilation, pulmonary hypertension, and heart failure due to unrestricted shunting. TTE is commonly the first diagnostic modality, particularly using color flow Doppler interrogation to assess for continuous (systolic and diastolic) high-velocity flow.[2] Live three-dimensional TTE performed with full-volume and color Doppler in multiplanar and nonmultiplanar modes can assist in locating and measuring the orifices of fistulas.[10] TEE has higher sensitivity and specificity for diagnosis of aortic root abscesses[11] and can be used if the quality of transthoracic imaging is suboptimal. Cardiac computed tomography has similar sensitivity and specificity to TEE for detecting valvular abnormalities in IE and is superior to TEE in the detection of pseudoaneurysm formation. Computed tomography is also useful in the evaluation of coronary atherosclerosis in preparation for cardiac surgical intervention and can detect extracardiac complications of endocarditis such as septic emboli. Although cardiac magnetic resonance imaging can identify valvular vegetations and structural complications of endocarditis, its role in addition to computed tomography and TEE is uncertain.[12]

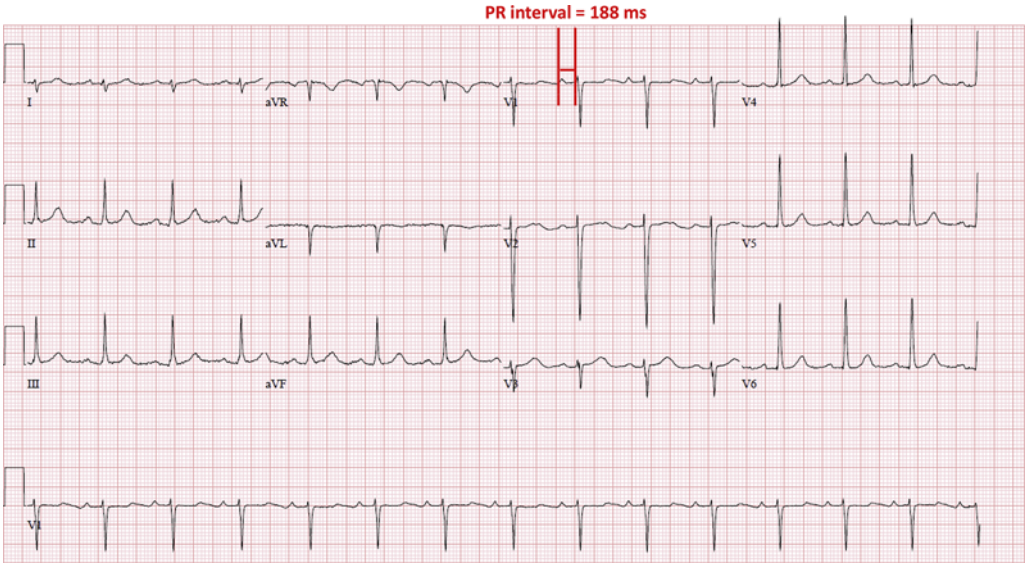

Figure 2 Standard 12-lead electrocardiogram showing normal sinus rhythm and PR interval of 188 msec.

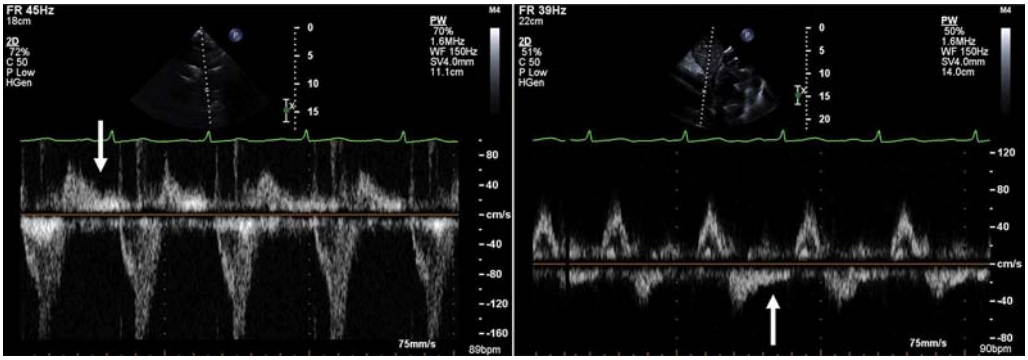

Figure 3 Holodiastolic flow reversal was seen in the proximal descending thoracic aorta (*left, white arrow*) and abdominal aorta (*right, white arrow*).

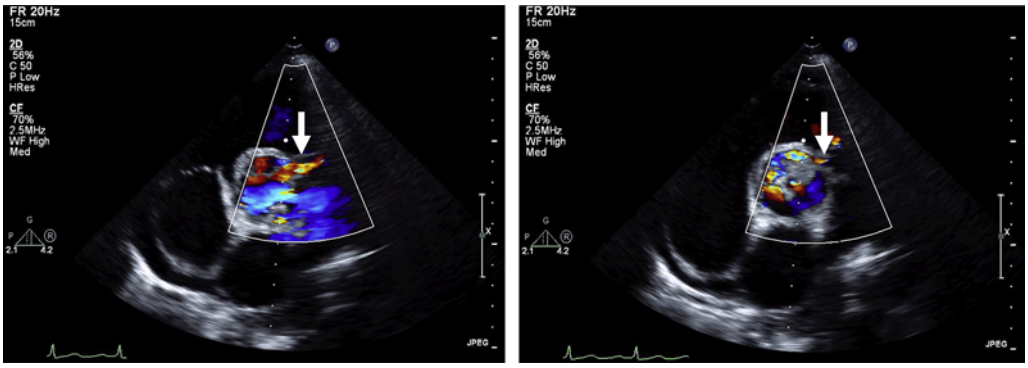

Figure 4 Parasternal short-axis views in systole (*left*) and diastole (*right*) showing fistula flow from the aortic root to the pulmonary artery throughout the cardiac cycle (*white arrows*).

The most common causative organisms implicated in the formation of any type of fistula in IE are *Staphylococcus* species, followed by various strains of *Streptococci,* including the viridans group.[13] Retrospective studies have shown that *Staphylococcus* endocarditis carries a higher mortality and worse postoperative outcomes compared with streptococcal endocarditis.[14,15] Intravenous drug use in patients with native aortic valve endocarditis is a risk factor for periannular extension of their infection.[16]

Fistula formation between cardiac chambers is a known, albeit uncommon, complication of IE. In a case series of 76 patients with

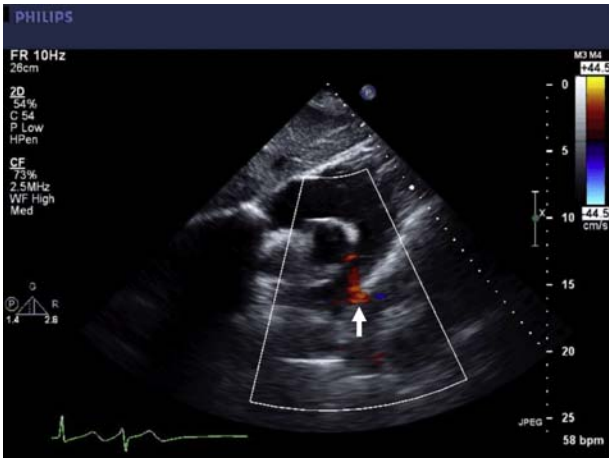

Figure 5 Subcostal view of the right ventricular outflow tract demonstrating flow from a PDA entering the left pulmonary artery and directed toward the pulmonic valve (*arrow*).

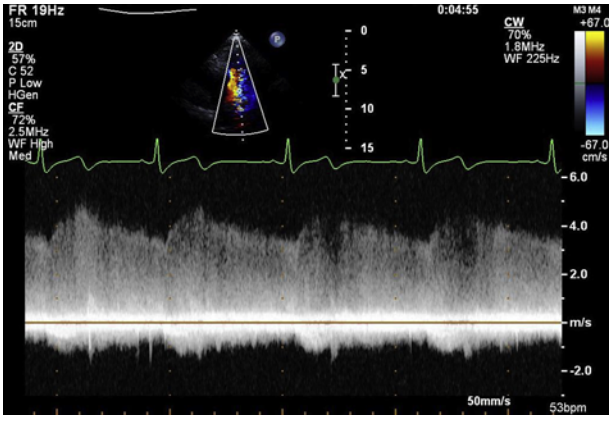

Figure 6 Continuous-wave Doppler interrogation of a PDA demonstrating systolic and diastolic flow toward the pulmonic valve.

aortocavitary fistula formation due to IE,[13] an accurate diagnosis was achieved with TTE in 53% of cases compared with 97% with TEE. In this series, all patients had aortic valve involvement, and the majority (78%) had periannular abscesses. Fistula formation was equally distributed between the three aortic sinuses of Valsalva and affected all four cardiac chambers equally, although none of the patients in this series had fistula formation to the pulmonary artery. In this series, fistula formation was more common in prosthetic valve IE compared with native valve IE.[13]

To fully evaluate aortopulmonary fistulas on two-dimensional TTE, modified parasternal views must be used, as was done in our case, to fully examine the abnormal flow seen on color flow Doppler. A modified parasternal long-axis view may show turbulent flow into the main or right pulmonary artery.[10] Modified parasternal short-axis views of the aortic root and pulmonary artery with color Doppler may demonstrate turbulent flows between the two structures, and continuous-wave Doppler interrogation (if proper Doppler alignment with the flow can be achieved) will reveal continuous aorta–to–pulmonary artery flow due to higher aortic pressures throughout the cardiac cycle. Fistula formation between the left ventricular outflow tract and pulmonary artery due to native aortic valve IE has previously been reported.[17] Subcostal modified right ventricular outflow views can be helpful in demonstrating high-velocity flow away from the pulmonic valve, which helps distinguish an aortopulmonary fistula from a patent ductus arteriosus (PDA) in which flow travels toward the pulmonic valve[2] (Figure 5). Other imaging features, such as location and associated findings, can also be helpful in differentiating aortopulmonary fistula from PDA. A PDA by definition arises from the proximal descending thoracic aorta distal to the subclavian artery, so flow is best seen in parasternal long-axis views into the proximal left pulmonary artery (Video 9) and demonstrates continuous flow on continuous-wave Doppler interrogation (Figure 6). PDA can be associated with ventricular septal defects, coarctation of the aorta, right-sided aortic arch, and complex congenital heart defects involving the great vessels.[18]

In contrast to fistulization from IE, ruptured sinus of Valsalva aneurysms most often occur at the right coronary cusp and drain into the right ventricle.[6] Rupture of a sinus of Valsalva aneurysm into the pulmonary artery is very rare, though several isolated case reports have been published.[19] The finding of a dilated aortic sinus helps differentiate between a ruptured sinus of Valsalva aneurysm and another cause of aortopulmonary communication. Sinus size can be assessed by multiple imaging modalities, and aortic insufficiency is often associated with sinus of Valsalva aneurysms.[6]

Surgical closure of an aortopulmonary fistula secondary to IE can be performed using pericardial or Gore-Tex patches or simple suture closure depending on the size and location of the fistulous tract.[13] Amplatzer occluder devices (St. Jude Medical) have been used with a transcatheter approach in fistulas not caused by IE.[5] Prognosis is poor in patients with structural complications of endocarditis, and in-hospital postoperative mortality has been reported to be as high as 42%.[13]

CONCLUSION

Echocardiography plays a crucial role in the assessment of structural complications caused by IE, such as the rare acquired aorta–to–pulmonary artery fistula presented in our case. TTE was valuable in demonstrating continuous left-to-right shunting, as well as evaluating the size, location, and extent of the hemodynamic burden imparted by the fistula. Careful evaluation for structural complications should always be undertaken in IE, particularly in cases affecting the aortic valve.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2019.01.004.

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