A Shortcut to Death: Aorto–Left Atrial Fistula in the Setting of MRSA Infective Endocarditis

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INTRODUCTION

The development of an aorto-atrial fistula (AAF) is a rare but problematic complication of endocarditis. We present an exceptional case of a 60-year-old man with infectious endocarditis (IE) leading to aorto–left atrial fistula.

CASE PRESENTATION

Our patient was a 60-year-old gentleman with comorbidities including stroke, chronic obstructive pulmonary disease, intravenous drug use, and hepatitis C who presented to the emergency department after being found down and disheveled by his roommate. He initially presented to a nearby hospital, where he was found to have splenic and left renal infarcts on imaging; he was then transferred to our hospital for further management. On initial presentation, he was febrile to 102.6°F and was started empirically on cefepime and metronidazole. Blood cultures were positive for methicillin-resistant *Staphylococcus aureus*, and he was transitioned to daptomycin and ceftaroline. Workup also revealed a urinary tract infection and a urine drug screen positive for cocaine and opioids. Initial transthoracic echocardiography (TTE) demonstrated an ejection fraction of 50% to 55%. TTE demonstrated no evidence of vegetations, but the parasternal short-axis view was difficult to visualize. Transesophageal echocardiography (TEE) revealed a bicuspid aortic valve (BAV) with severe thickening of the aortic leaflets with a flail noncoronary cusp, leading to severe aortic regurgitation (see Videos 1 and 2). There was also thickening of the aortic root with multiple echo-lucencies extending to the base of the anterior mitral leaflet, findings suggestive of an aortic root abscess (see Video 3). Furthermore, TEE demonstrated an aorto–left atrial fistula with continuous shunt on color flow imaging (see Figures 1 and 2, Video 4). Cardiothoracic surgery was consulted, but the patient was deemed at too high a risk for surgery. Given his worsening hemodynamic state, he was started on a norepinephrine drip to maintain adequate perfusion. Nevertheless, his overall clinical state continued to deteriorate. Despite treatment with daptomycin and ceftaroline, he had refractory bacteremia with methicillin-resistant *S aureus* continuing to grow on multiple blood cultures. Furthermore, he had worsening renal failure with subsequent extreme electrolyte derangements and a lactic acid level of 4.7, requiring continuous renal replacement therapy. The next day, however, his wife requested for continuous renal replacement therapy to be discontinued with no further escalation of care, opting for comfort measures only. His respiratory status continued to decline, and the patient passed away later that evening. His splenic and left renal lesions are now thought to have been septic emboli. He developed acute decompensated heart failure, most likely due to a left-left shunt and regurgitation caused by the aorto–left atrial fistula. Chest computed tomography demonstrated no evidence of emboli.

DISCUSSION

An AAF is a rare but potentially life-threatening condition in which an anomalous connection forms between the aortic structures and cardiac atria.[1] The etiology can widely differ, including congenital causes, secondary to conditions such as IE or valve replacement, or iatrogenic.[1] In the case of IE, when the infection spreads beyond vascular structures, it can lead to periannular complications, such as AAFs.[1] In the literature, the prevalence of AAF in the setting of IE is 1% to 2%.[2] The pathogenesis of AAFs related to infection is caused by extension and infiltration of abscesses related to the endocarditis. As the infection spreads to surrounding tissue, it inflames and weakens the myocardium, slowly eroding a path in the form of a fistula into a nearby space, such as the atrium.[3] Our literature suggests that 51.7% of AAF formations open into the right atrium.[4] Interestingly, the junctional zone between the mitral and aortic valve annulus, known as the mitral-aortic intervalvular fibrosa, is, unfortunately, ideal for abscess formation given its relative hypovascularity.[5,6] This is thought to be a result of the decreased ability to mount an immune response.[1] Multiple reviews have demonstrated a roughly similar distribution of causative organisms with *Staphylococcus* species being found in 58% of cases,[1] followed by *Streptococcus* species in 28%, *Enterococcus* species in 7%, and 7% of cases being polymicrobial.[7] Prognosis, however, did not seem to differ on the basis of the organism involved.[1]

Echocardiography is a core component in the diagnosis and management of valvular pathologies.[8] Although TTE is a great tool for the initial assessment of suspected structural pathologies, TEE is superior in providing a detailed assessment of structure and function. Anguera *et al.*[4] demonstrated that the detection rates of AAF using TTE and TEE are 53% and 97%, respectively. Additionally, TEE is superior to TTE in assessing valve function and morphology as well as delineating intracardiac pathology, such as complications of endocarditis, namely, root abscess and fistulas.[8] Furthermore, because most aorto–left atrial fistulas usually occur from the posterior aspect of the aorta, the anatomy is naturally better assessed using TEE.[8] Naturally, this leads to a better signal-to-noise ratio on TEE, resulting in higher quality images and less attenuation.[8] TEE is the superior imaging technique and has better resolution in the evaluation of cardiac pathology because of the proximity of the probe to the diseased segment of the aorta.

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Keywords: Aorta, Left atrium, Fistula, Infective endocarditis, Abscess

Conflicts of interest: The authors reported no actual or potential conflicts of interest relative to this document.

2468-6441

https://doi.org/10.1016/j.case.2020.03.006

VIDEO HIGHLIGHTS

Video 1: Thickening of the aortic root with multiple echo-lucencies extending to the base of the anterior mitral leaflet, suggestive of an aortic root abscess.

Video 2: Aorto–left atrial fistula with continuous shunt on color flow imaging; color comparison.

Video 3: Bicuspid aortic valve with severe thickening of the aortic leaflets. Flail noncoronary cusp and poor mobility of the opposing cusp.

Video 4: Color Doppler demonstrating severe aortic regurgitation caused by the flail noncoronary cusp of the patient's BAV.

View the video content online at www.cvcasejournal.com.

Given its course through a dense and anatomically delicate area, the development of an AAF is often associated with life-threatening complications. The literature reports very high rates of these complications, with >60% of patients developing significant heart failure and >40% ending in death.[1] Unfortunately, surgical mortality itself is also very high at about 40%.[4] Factors associated with adverse outcomes, in the literature, include septic shock, hemodynamic instability, and congestive heart failure. All of these factors were present in our patient. With severe thickening of the aortic leaflets and a flail noncoronary cusp leading to severe aortic regurgitation, our patient struggled to maintain adequate perfusion. Despite pressor support and proper antibiotic therapy, his clinical course continued to decline. Systolic failure and underperfusion ultimately led to renal failure, requiring the use of continuous renal replacement therapy. Although not manifested in our patient, AAF can also be associated with an array of conduction abnormalities, from bundle branch blocks to first-, second-, and third-degree heart blocks in up to 10% of cases.[7] Unsurprisingly, AAF is a rare but very problematic complication of IE with high mortality despite adequate treatment.[7] For this reason, high suspicion, timely diagnosis, and early treatment with antibiotics and/or surgery are of utmost importance.[7]

Surgery remains the preferred choice for management of aortocavitary fistulae.[7] Ergin *et al.*[9] stressed the importance of removing all infected elements, followed by reconstruction of the aortic annulus for safe anchoring of a valve conduit. Fierro *et al.*[1] summarized currently available conduit options for reconstruction to include conventional aortic valve replacement (using a mechanical or stented biological valve), aortic valve replacement with translocation, aortic root replacement using a homograft, pulmonary autograft (Ross procedure), a stentless biological valve, or a composite graft. With this approach,

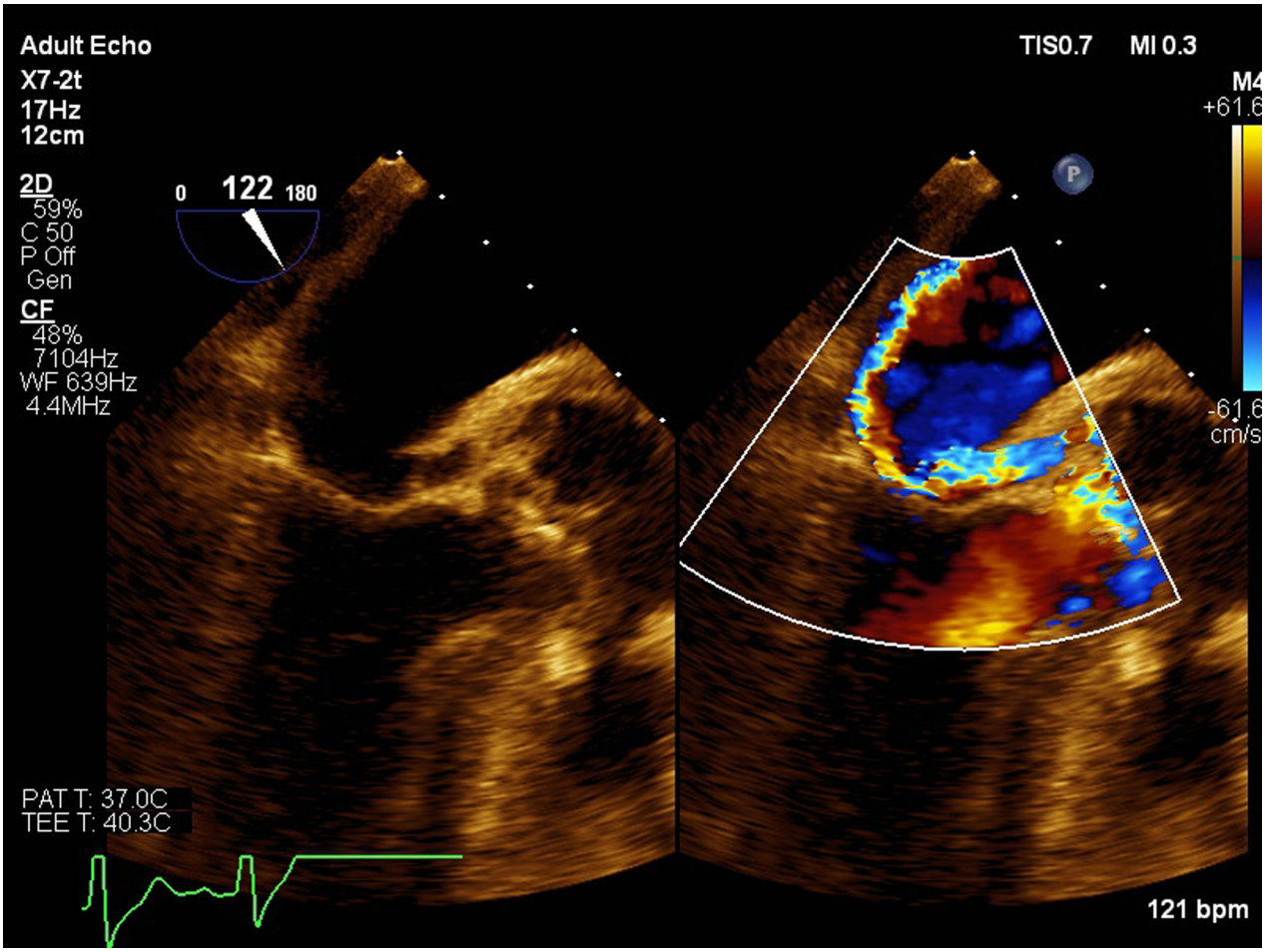

Figure 1 A color compare of an aorto–left atrial fistula with continuous shunt on color flow imaging.

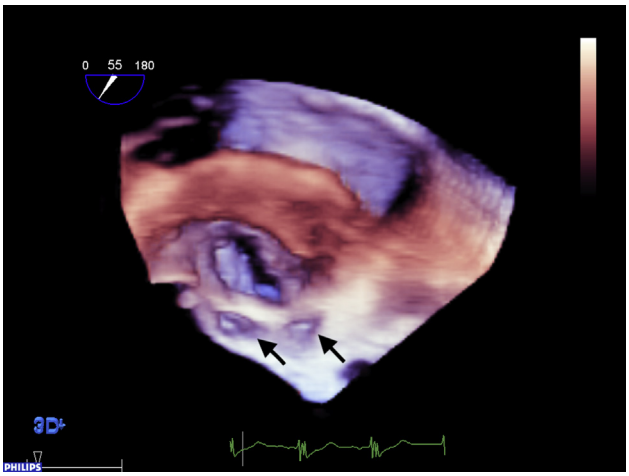

Figure 2 Three-dimensional picture of BAV with thickened leaflets. The *arrows* point to a thickened root with tracts suggestive of aortic fistula.

Fierro *et al.* reported freedom from residual or recurrent infection and paravalvular leaks of 92% and freedom from reoperation at 17 years of 75%.[1] Nevertheless, the surgical approach is still associated with significant morbidity and mortality. Given surgery's limitations, however, the literature includes several reports of successful attempts at transcatheter closure of AAFs using the Amplatzer plug technique.[10,11] Alkhouli *et al.*[10] were able to completely seal a post–valve replacement AAF immediately after deployment of the ADO-II, a multilayered mesh delivered through a small catheter without impingement of adjacent aortic valve or coronary arteries.[10] Per Alkhouli *et al.*, this approach can be applied in patients with bioprosthetic or mechanical valves. Potential complications associated with this approach include impingement of the valve, device embolization, stroke, and coronary artery obstruction; nevertheless, none of these complications have yet been reported, to our knowledge.[10] Although short-term outcomes seem promising, there have been only a handful of transcatheter attempts reported, and long-term data are not yet available.

AAF was first discovered as an incidental finding during an autopsy in 1924,[12] and it remains a very rare and deadly complication today. Although timely diagnosis is absolutely crucial for successful management, clinical diagnosis can be exceedingly difficult. We present an exceptional case of AAF with rapid clinical deterioration. Our patient possessed all poor prognostic factors, as listed in the literature, including septic shock, hemodynamic instability, and congestive heart failure. Furthermore, his case highlights the importance of high suspicion for AAF in patients who present similarly, as his transthoracic echocardiographic study was technically difficult, with short-axis views that did not permit full assessment of the aortic valve, demonstrating only heavily calcified leaflets and thickened aortic root with mild to moderate aortic regurgitation. TEE was not only the key in successfully diagnosing the case, but also provided very remarkable, classic imaging findings that can serve as a reference for future cases.

BAV is the most common congenital cardiac abnormality, affecting up to 0.5% to 2% of the population.[13-15] Tzemos *et al.*[16] report that aortic valve endocarditis occurs in 2% of patients with BAV or 0.3% per year. Despite steady improvement in medical and surgical therapies, IE remains a deadly disease, especially in patients with BAV.[17] Findings by Chen *et al.*[17] suggest that BAV is the only independent predictor associated with an increased risk for aortic perivalvular abscess, though the underlying mechanism for this remains unclear. Histopathologic analyses show that reduction in fibrillin-1 and upregulation of matrix metalloproteinases lead to apoptosis and disruption of the aortic media.[18,19] Such vascular matrix remodeling predisposes these patients to infection extension.[17] Our patient had known BAV since at least 2016. Unfortunately, he had an extensive history of polysubstance abuse and had been nonadherent to follow-up through the years.

CONCLUSION

The development of an AAF is a rare but life-threatening complication of IE that is often associated with acute decompensated heart failure, heart blocks, and death.[7] The acuity and severity of this disease process, as well as its high mortality rate, highlight the importance of high suspicion and early management to provide the patient the best chance for survival.

ACKNOWLEDGMENT

We acknowledge the contribution of Teryn Nix, RDCS, the sonographer who acquired the images.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2020.03.006.

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