Aorto-left atrial fistula: Rare cause of acute cardiac failure in a previously healthy individual

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Complications of valvular infective endocarditis involving the peri-annular region puts the patient at a significantly high risk of adverse outcomes including heart failure and death. The “mitral–aortic intervalvular fibrosa” is relatively avascular and offers little resistance to the spread of abscesses, aneurysm, and fistula formation. Aorto-cavitary fistulous tract formation in the setting of native valve infective endocarditis is associated with higher rates of heart failure, ventricular septal defect, and atrioventricular block than nonruptured abscesses. Thus, a high index of suspicion is needed for rapid and accurate diagnosis, which can guide further management. A transesophageal echocardiogram is the preferred modality of investigation in such cases. Staphylococci are reported to be the most common pathogen involved. In addition to early antibiotic therapy, prompt surgical intervention might save lives in these scenarios.

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Introduction

When infective endocarditis (IE) is suspected a transesophageal echocardiogram (TEE) should be performed promptly. Moreover, when the peri-valvular annulus region is involved with abscess formation, surgical intervention should be emergently initiated.

Case report

A 57-year-old Asian man without any significant medical history presented to the Emergency Department (Western Michigan University Homer Stryker School of Medicine) with fever, dyspnea, diffuse abdominal pain, and chest pain ongoing for 5 days. His chest pain was left-sided, dull, constant, nonradiating, and 4/10 in intensity. Vital signs were significant for blood pressure...
which was 197/66 mmHg, he had a heart rate of 137 bpm, a respiratory rate of 37/min, temperature of 37°C, and oxygen saturating 86% on room air. Physical examination was remarkable for Grade 3/6 pan-systolic murmur heard all over the precordium and bilateral crackles. He was emergently intubated due to hemodynamic instability. Chest radiograph was consistent with acute pulmonary edema (Fig. 1). Electrocardiogram showed infero-lateral infarct. Laboratory results were significant for leukocytosis of 31,600/cmm, bicarbonate 10 mmol/L, lactic acid 12.9 mmol/L, pH of 7.00, and troponin T 0.22 ng/mL. He was transferred to the intensive care unit and placed on a nitroglycerin drip and started on empiric antibiotics. Emergent transesophageal echocardiogram revealed distortion of the left coronary cusp with possible vegetation on the aortic valve with mild aortic regurgitation. An aortic root abscess perforating in the intervalvular fibrosa region with left atrial shunting was also appreciated (Fig. 2; Video 1). Blood cultures were positive for methicillin sensitive Staphylococcus aureus.

The patient went into cardiac arrest over the next few hours and passed away despite vigorous cardiopulmonary resuscitation efforts. Autopsy confirmed the findings of an abscess at the junction of the noncoronary and left coronary cusps of the aortic valve and fenestration of these valves. Perforation of the abscess from the subaortic left ventricle into the left atrium was also appreciated. Microscopy of the abscess revealed numerous neutrophils and large colonies of gram-positive bacteria in clusters, morphologically consistent with S. aureus (Fig. 3).

**Discussion**

Spread of infection in IE from valvular structures to the surrounding tissue results in perianular complications, which can cause adverse outcomes such as heart failure, arrhythmias, and death. An aorto-cavitary fistula is one of the rare and dreadful complications. Various etiologies
for aorto-cavitary fistulas have been described such as IE, complication of aortic valve replacement, congenital, trauma, Behcet’s disease, rupture of the sinus Valsalva, and aortic dissection. Aortic valve IE complicated with abscesses and pseudo aneurysms involving the sinuses of Valsalva may rupture internally with the subsequent development of an aorta-cavitary fistula. These communications create intracardiac shunts, which may result in further clinical deterioration and hemodynamic instability [1]. The junctional zone between the mitral and aortic valve annulus, also known as the “mitral–aortic intervalvular fibrosa,” is relatively avascular and offers little resistance to the spread of abscesses, aneurysm, and fistula formation. In a retrospective multicenter study of 2055 native valves with IE, 201 patients (9.8%) with peri-annular complications in aortic valve IE were identified. Of the 201 cases, 46 had aorto-cavitary fistulization and 155 had nonruptured abscesses. The most common organism identified were staphylococcal and streptococcus species, accounting for 70% of cases with aorto-cavitary fistulae [1]. Aorto-cavitary fistulous tract formation in the setting of native valve IE is associated with higher rates of heart failure, ventricular septal defect, and atrioventricular block than nonruptured abscesses. Despite these higher rates of complications, fistulous tract formation in the current era of IE is not an independent risk factor for mortality [1]. The dilated pouches corresponding to each leaflet of the aortic valve are divided into three spaces: noncoronary, left aortic sinus, and right aortic sinuses. The right and noncoronary aortic sinuses are anatomically close to right atrium and tricuspid valve. The left atrium is close to left coronary aortic sinus and noncoronary aortic sinus but for the most part is separated from aortic sinuses by the trunk of the pulmonary artery. Aorto-left atrial fistulae are hence less common than aorto-right atrial fistulae, at least in theory and per anatomy [2]. In a case series of 24 patients with complicated subaortic IE including abscess, aneurysm formation, and perforation of mitral valve, all patients had some form of predisposing factor like unicuspid, bicuspid, prosthetic aortic valve, or were an intravenous drug user [3]. In our patient, there was no significant medical, surgical, or social history making it even more interesting. Aorto-atrial fistulas are better visualized using TEE, especially those involving the left atrium because of the proximity of the transducer to the structures of interest. The emphasis on early TEE cannot be undermined due to the rapidity and fatality of the condition. In our scenario, the patient expired in a span of 3–4 hours from being apparently hemodynamically stable. Multiple case reports have reported the high sensitivity and specificity of three-dimensional TEE in making the diagnosis and delineating the anatomy more accurately [4,5]. The different surgical modalities for valve replacement included bioprosthesis, mechanical prosthesis, and homograft. The majority of patients with aorto-cavitary fistulae underwent surgery urgently at Day 3 after diagnosis. The mortality among the ruptured and nonruptured abscesses was 35% versus 26% which was insignificant signifying the unpredictable course of the disease. Patients who were severely hemodynamically unstable or with multiple comorbidities could not have surgery performed [1]. A percutaneous approach with Amplatzer devices can be utilized in chronic stable conditions where anatomically feasible cases [6]. In acute cases it is not feasible, due to the friable nature of the tissue, to hold the device in place. Aorto-cavitary fistulae formation with multiple fenestrations in the aortic valves in a setting of fulminant IE causing flash pulmonary edema and acute heart failure in a previous healthy patient has rarely been reported.

Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at http://dx.doi.org/10.1016/j.jsha.2016.02.005.
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