

CASE REPORT

Aortocavitary fistula as a complication of infective endocarditis and subsequent complete heart block in a patient with severe anemia

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Infective endocarditis has different presentations depending on the involvement of valvular and perivalvular structures, and it is associated with high morbidity and mortality. Aortocavitary fistula is a rare complication. We introduce the case of a 48-year-old female with native valve endocarditis, complicated by aortocavitary fistula to the right atrium, and consequently presented with syncope.

Keywords: *endocarditis; aortocavitary fistula; complete heart block*

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Infective endocarditis (IE) presents in different clinical forms and may be associated with high morbidity and mortality. The involvement of valvular and perivalvular structures increases the risk of adverse outcomes. Abscesses and pseudoaneurysms involving the sinuses of Valsalva may rupture with ensuing development of aortocavitary fistula (ACF). This abnormal communication between the aortic root and heart chambers creates an intracardiac shunt, which further jeopardizes hemodynamics. We report the case of a patient who presented with IE, complicated by ACF, leading to congestive heart failure and complete heart block.

Case report

A 48-year-old female presented to the emergency department (ED) with syncope. Her past medical history was significant for mild mental retardation, former tobacco use, and hernia repair. She was not on any home medications.

The patient stated that while cooking she felt lightheaded, subsequently lost consciousness, and fell to the ground recovering after a few minutes. After recovering, she noticed her exercise tolerance declined, feeling shortness of breath after climbing just one flight of stairs. On review of systems, she reported 2 weeks of menorrhagia. She denied chest pain, palpitations, diaphoresis, lower extremity edema, orthopnea, and paroxysmal nocturnal dyspnea.

On physical examination, her heart rate was 88 beats/min, blood pressure 113/48 mmHg, oxygen saturation 98% on room air, and a temperature of 98.4°C. Cardiac examination revealed a 3/6 decrescendo diastolic murmur best heard on the right sternal border with radiation to the carotids. The physical exam was also remarkable for poor dentition with evidence of decay in multiple teeth as well as pallor in mucous membranes, palms, and soles.

Laboratory workup was remarkable for profound iron deficiency anemia (Hgb 2.9 g/dl, Hct. 10.7%, mean corpuscular volume 54 fl, ferritin 15 ng/ml, iron 261 µg/dl, transferrin 218 mg/dl). In the ED, she received three units of packed red blood cells. Her chest X-ray (CXR) showed cardiomegaly, computed tomography (CT) of the head ruled out hemorrhage, and CT of the abdomen/pelvis revealed multiple myomas which were thought to be the source of bleeding. Due to a new diastolic murmur and cardiomegaly on CXR, a transthoracic echocardiogram (TTE) was obtained, demonstrating a thickened aortic valve with severe aortic insufficiency. Left ventricular chamber size was normal with an ejection fraction of 65%. Given high suspicion for IE, a transesophageal echocardiogram (TEE) was performed revealing an irregularly shaped mobile echodensity (1.7 cm × 1.0 cm) on the aortic valve consistent with a vegetation (Fig. 1). Also, a root abscess was present with fistula formation between the right sinus of Valsalva and the right atrium (Fig. 2).

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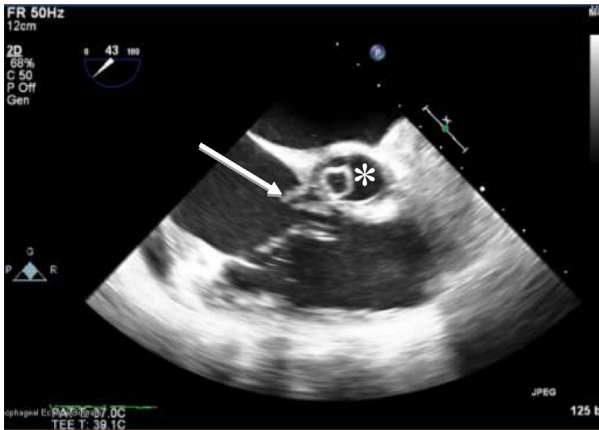


Fig. 1. TEE at the mid-esophageal level showing vegetation (*) on the aortic valve and fistula formation (arrow) between the aortic root and the right atrium.

Broad-spectrum antibiotics were started with vancomycin and ceftriaxone, and she was transferred to the cardiac care unit. Blood cultures were positive for *Streptococcus anginosus* and antibiotics were narrowed to ceftriaxone for 4 weeks. Cardiac surgery service was consulted for aortic valve repair (AVR) and repair of the fistula.

The next day the patient underwent AVR with a bioprosthetic valve and bovine patch repair of sinus of Valsalva fistula. She remained hemodynamically stable, but required permanent pacemaker implantation due to permanent complete atrioventricular block post-surgery. At a month follow-up, the patient has been stable, with normal valvular function, and preserved ejection fraction.

Discussion

ACF is a rare entity occurring in less than 2.2% of native valve endocarditis (1). The most common agents are *Staphylococcus aureus*, *Streptococcus* spp., *Enterococcus* spp., and other less common bacterial and fungal infections (1, 2). It is thought that the process leading to an ACF is the resulting inflammation from the bacterial colonization

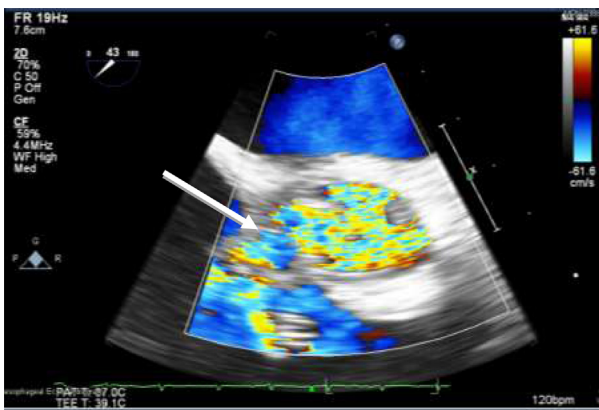


Fig. 2. Color Doppler of the fistula communicating the aortic root and the right atrium.

of the valve and surrounding tissue with subsequent abscess formation, creating erosion of the sinus of Valsalva (1–3). The intervalvular fibrosa is particularly prone to infection given its avascularity and infected regurgitation of jet striking subvalvular structures (4).

A left-to-right shunt is created from any of the aortic valve sinuses into any of the cardiac chambers without preference, which causes hemodynamic instability (2, 5). The size of the shunt determines clinical presentation of ACF. Therefore, clinical presentation can range from a non-clinically significant murmur, to a chest pain syndrome, acute coronary syndrome, refractory heart failure, or even aortic dissection (6, 7). (See Table 1 for presenting complaint of reported cases.)

As seen in our index case, ACF has been documented most commonly in cases of aortic valve IE, more in prosthetic than native aortic valves (5, 8). ACF rarely presents as a complication of right-sided IE, and when that is the case, it is usually a late presentation (9, 10).

Clinical scenarios from primary IE as in our case have also been described with cardiac surgery (11, 12), percutaneous interventions (13), chest trauma, and autoimmune process (14).

Though clinical presentation of ACF will be determined mainly by the inciting primary process and the size of the shunt, the variety of the initial clinical presentations can range from acute decompensated heart failure (6) to asymptomatic patients with only an associated murmur (15, 16). Our case would be the first case presenting as a syncopal episode. When symptomatology is poor, a high index of suspicion is required, especially in the background of recent surgery or IE. The auscultation of a murmur can be the key to diagnosis. The ACF murmur has been described as a continuous-thrill murmur (17, 18). The median time for the echocardiographic diagnosis of the fistula is usually 1 month after onset of symptoms (5).

TTE is usually the first imaging method used, although TEE is a better modality to describe valvular and paravalvular pathology (19, 20), with better delineation of function and morphology (19, 20). In both, TTE and TEE, color Doppler can detect a highly turbulent flow between the aorta and the cardiac chambers (5). Recently, three-dimensional echocardiography has also emerged as an excellent modality to delineate anatomic and unconventional views of cardiac structures detecting volume data, cropping, and slicing in various planes (21–23).

CT can be considered as a useful complement in visualizing the cardiac lesions of IE if echocardiography is inconclusive. It provides assessment of valvular and perivalvular involvement, extra-cardiac lesions, and non-invasive evaluation of the coronary artery anatomy, simultaneously (24). Moreover, comprehensive evaluation of anatomic relationships of perivalvular abscess/pseudoaneurysms may be beneficial for presurgical planning (25). This can also be accomplished with cardiac magnetic resonance

Table 1. Reported cases of aortocavitary fistula

Year	Authors	Presenting complaint	Etiology
1983	Lorenz et al.	Decreased exercise tolerance, SOB	Surgical complication (valve replacement)
1989	Chow et al.	Asymptomatic	Surgical complication (valve replacement)
1999	Roy et al.	Decreased exercise tolerance, SOB	Surgical complication (valve replacement)
2001	Chakko	Right heart failure	Trauma (stab wound)
2002	Akowuah et al.	Cellulitis, pneumonia	Native tricuspid valve endocarditis
2005	Ananthasubramaniam	Fatigue, paroxysmal nocturnal dyspnea, SOB	Surgical complication (valve replacement)
2006	Rubin et al.	Chest trauma	Crush trauma
2008	Moral et al.	Fever, edema, SOB	Prosthetic aortic valve endocarditis
2009	Amabile et al.	ACS	Surgical complication (valve replacement)
2009	Pinaud et al.	SOB	Congenital anomaly – fistula to right ventricle
2010	Patel et al.	Edema, SOB	Surgical complication (valve replacement)
2012	Candan et al.	Fever, fatigue	Prosthetic mitral valve endocarditis
2012	Maffè et al.	Fever, SOB	Prosthetic valve endocarditis
2012	Najib et al.	Palpitations, SOB	Surgical complication (valve replacement)
2013	Kalra et al.	Dizziness, fatigue, SOB	Post-surgical – myomectomy (HOCM)
2013	Dias et al.	Fatigue, fever, SOB	Bicuspid aortic valve endocarditis
2013	Gunarathne et al.	'Unwell', fever	Bicuspid aortic valve endocarditis
2013	Chandra et al.	Fever, 'symptoms of heart failure'	Native aortic valve endocarditis
2014	Frey et al.	Fatigue, fever, palpitations, malaise, weight loss, irregular tachycardia	Aortic and tricuspid valve endocarditis
2014	Rocha et al.	Pulmonary edema	Native aortic valve endocarditis
2014	Villablanca et al.	Abdominal discomfort, chest tightness, edema, SOB	Native tricuspid valve endocarditis
2015	Shakoor et al.	Found on ECHO post-TAVR	Post-TAVR

SOB, shortness of breath; ACS, acute coronary syndrome; TAVR, transcatheter aortic valve replacement; HOCM, hypertrophic obstructive cardiomyopathy.

and aortography; both of which can provide an excellent anatomical and flow dynamic description of the ACF before closure (26–29). Disadvantages of CT and angiography compared to echocardiography are the contrast-induced nephropathy and radiation exposure.

ACF carries severe morbidity and mortality, especially in patients who are elderly, have severe heart or renal failure, require emergent procedures, or are infected with *Staphylococcus* (1, 3, 5). Treatment is mainly surgical closure.

Conclusion

Acute and subacute IE can present in different forms with subtle clinical signs and symptoms. Physical examination and non-invasive imaging are essential in the diagnosis and management of IE. Paravalvular complications of IE should be treated in a timely manner to avoid further destruction of paravalvular tissue including the conduction system.

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