Aorto-Left Atrial Fistula as Sequela of Endocarditis

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Aortoatrial fistula formation is a rare complication of bacterial endocarditis. Fistulous tracts may form between the aorta and either atrium. Clinical presentation varies from an insignificant murmur to refractory congestive heart failure. Most clinically relevant fistula manifests with acute and severe symptoms. Transesophageal echocardiography (TEE) is more sensitive than transthoracic echocardiography (TTE) in diagnosing intracardiac shunts, and invaluable in guiding intraoperative surgical repair. Definitive therapy involves closure of the fistula either through an open surgical approach or percutaneously with an occluder device. (A&A Practice. 2020;14:e01233.)

GLOSSARY

2DTEE = 2-dimensional transesophageal echocardiography; **3DE** = 3-dimensional echocardiography; **AR** = aortic regurgitation; **AV** = aortic valve; **LAX** = long axis; **LCA** = left coronary artery; **LCC** = left coronary cusp; **ME** = midesophageal; **MR** = mitral regurgitation; **SAX** = short axis; **TEE** = transesophageal echocardiogram; **TTE** = transthoracic echocardiogram

CASE

A 60-year-old woman diagnosed with bacterial endocarditis presented for open surgical aortic valve replacement. Her diagnosis was based on a preprocedural transthoracic echocardiogram (TTE) and confirmatory transesophageal echocardiogram (TEE) that reported a mass resembling vegetation at the aortomitral junction, moderate mitral regurgitation (MR), and severe aortic regurgitation (AR) secondary to a prolapsed leaflet.

Baseline intraoperative 2-dimensional TEE (2DTEE) images were obtained under anesthesia, before the start of surgery. The midesophageal (ME) 5-chamber view demonstrated a prolapsed left coronary cusp (LCC) and an area of tissue disruption along the aortic root with a thin flap protruding into the left atrium (Figure 1). The addition of color Doppler over the aortic and mitral valves demonstrated severe AR, moderate MR, and an oddly positioned flow jet from the aortic root into the left atrium that was continuous throughout the cardiac cycle (Supplemental Digital Content, Video 1, <http://links.lww.com/AACR/A316>). The ME long-axis (LAX) view again demonstrated a highly mobile mass arising from the anteromedial aspect of the aortic root and protruding into the left atrium (Figure 2A). The addition of color Doppler again demonstrated MR, AR, and abnormal continuous flow from the aortic root into the left atrium through an apparent fistula tract (Supplemental Digital Content, Video 2, <http://links.lww.com/AACR/A317>). The ME short-axis (SAX) view (Figure 2B) (Supplemental Digital

Content, Video 3, <http://links.lww.com/AACR/A318>) demonstrated a cavity at the level of the LCC and again abnormal continuous flow from the aortic root into the left atrium (Figure 3A, B).

Based on this new finding of an aortoatrial fistula with intracardiac shunting, the surgical plan was altered to include patch repair of the aortic root to eliminate the fistula.

Written informed consent was obtained from the patient to publish this article.

DISCUSSION

Our patient had a rare complication of endocarditis development of a fistula between the aortic root and left atrium. Other reported causes of aortoatrial fistulae include ruptured sinus of Valsalva aneurysm, revision aortic valve replacement, aortic dissection, saphenous vein graft aneurysms after coronary artery bypass, and previous atrial septal defect closure with a device.^{1,2}

TEE is more sensitive than TTE in diagnosing valvular vegetations and perivalvular sequelae, including fistulae, because of its close proximity to the aortic root and use of higher ultrasonic frequencies.³ While TTE is rapid, noninvasive, and has excellent specificity for vegetation (98%), it has a low sensitivity (50%–60%) for detection of fistulae compared with TEE (97%).^{4–6} In recent years, 3-dimensional echocardiography (3DE) has become increasingly popular because it allows superior spatial orientation and high anatomic definition of cardiac structures, and is now emerging as the imaging modality of choice in the examination of fistulae in many centers.^{7,8}

In our patient, intraoperative 2DTEE confirmed a prolapsed LCC causing severe AR and moderate MR. However, contrary to the previous diagnosis of vegetation at the aortomitral junction, we identified a filamentous flap-like mass arising from a point in the left atrium along the anteromedial aspect of the aortic root, in close proximity to the A1/A2 segments of the mitral valve. Immediately proximal to this flap was an area of tissue disruption that we identified as a fistula from the aortic root into the left atrium. The fistula tract appeared to originate at the point of attachment of the LCC to the aortic root wall in the left sinus of Valsalva, undermining it, and likely

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Figure 1. Midesophageal 5-chamber view in 2 dimensions showing a prolapsed left coronary cusp and a juxtaposed mass in the left atrium.

Figure 2. A, ME AV LAX view in 2 dimensions showing prolapsed left coronary cusp and fistula tract with flap protruding into the left atrium. B, ME SAX AV showing an area of lucency next to the left coronary cusp, with flap from aortic root protruding into left atrium. AV indicates aortic valve; LAX, long-axis; ME, midesophageal; SAX, short axis.

Figure 3. A, ME AV LAX view in 2 dimensions with color Doppler in systole showing mitral regurgitation and flow from aorta into left atrium. B, ME LAX AV view in 2 dimensions with color Doppler in diastole showing aortic regurgitation and flow from aorta into left atrium. AV indicates aortic valve; LAX, long-axis; ME, midesophageal.

causing the prolapse. This is seen in both the ME 5-chamber (Supplemental Digital Content, Video 1, <http://links.lww.com/AACR/A316>) and ME LAX views (Supplemental Digital Content, Video 2, <http://links.lww.com/AACR/A317>).

On TEE, the ME 5-chamber view is an important, standard view, although it is often skipped during interrogation. It allows visualization of the anteromedial wall of the aortic root. As demonstrated in this case, it was crucial to arriving at the correct diagnosis.

Aortocavitary fistula formation in the setting of infective endocarditis is associated with high rates of atrioventricular conduction block and dysrhythmias.⁹ Endocarditis is one of the most common causes of acquired aortoatrial fistula. Inflammatory processes and bacterial tissue destruction cause fistula formation in 1.6% of affected patients with native valves and up to 5.8% with prosthetic valves.¹⁰ Symptoms of aortoatrial fistula depend on the shunt volume and chronicity of the defect. Flow through the fistula into the left atrium can cause acute left-sided volume overload with pulmonary edema and chronically, dilation of the affected atrium leading to arrhythmias, dyspnea, pulmonary edema, and heart failure.

The location and trajectory of an aortoatrial fistula tract are very important. The fistula in our patient was quite proximal to the ostium of the left coronary artery (LCA), although it was spared. Erosion into the LCA would have acute and catastrophic consequences.

The ME LAX view (Supplemental Digital Content, Video 2, <http://links.lww.com/AACR/A317>) shows flow through the fistula was missed in certain frames but picked up with a careful rotation of the probe. All abnormal flow should be closely interrogated by focusing on the lesion, decreasing scan depth to increase frame rate and enhance temporal resolution, and if necessary decreasing sector width to improve lateral resolution. An abnormal jet should be traced by careful probe movements—rotation of the probe to the right and left, advancement and withdrawal to identify the point of origin and direction. Each lesion should be examined from different planes to verify the accuracy of a diagnosis. It is also crucial to note the timing of flow jets with regard to the cardiac cycle. MR jets are typically seen in systole, AR jets in diastole. However, flow through an aortoatrial fistula is continuous—seen in systole and diastole.

Close interrogation of the flap-like mass and its associated jet utilizing the zoom function, freezing and scrolling through successive frames while noting the timing of flow with regard to the cardiac cycle was necessary to determine that this was a fistula tract between the aorta and the left atrium, and to avoid misdiagnosing the jet as MR, which was also present in this patient. Color M-mode could also have been employed in this situation for a clear delineation of timing of flow.

Using the techniques described above, color flow through the defect was found to be continuous throughout the cardiac cycle, a notable discerning characteristic of

aortoatrial fistulae, and was found to be originating from a point proximal to the left sinus of Valsalva. Importantly, when severe valvular regurgitation is present, a small jet through a fistula may be difficult to delineate. Freezing the image and scrolling through while noting systolic and diastolic color flow helps isolate an abnormal jet's origin and direction.

Postcardiopulmonary bypass TEE performed after surgical replacement of the aortic valve and patch repair of the aortoatrial fistula demonstrated a normal functioning aortic bioprosthetic valve and complete elimination of the fistula tract. In addition, there was marked improvement in the degree of MR from moderate to mild, underscoring its etiology as mostly functional. The patient's postoperative course proceeded smoothly without complications. ■■

DISCLOSURES

Name: Abimbola O. Faloye, MD.

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