34-year-old black man was treated for supraventricular tachycardia and subsequently found to have a 2/6 pansystolic murmur. He had been an asymptomatic healthy individual with no previous medical history of significance, including no past chest trauma.

After transthoracic and transesophageal echocardiography (TEE), the provisional diagnosis was a congenital submitral aneurysm (SMA), which originated along the lateral aspect of the posterior and anterior mitral annulus and spanned across the anterolateral commissure (Figure 1) associated with severe mitral regurgitation (Figure 2).

Three-dimensional (3D) TEE depicted the spatial extent of the aneurysm, which was bilobed and involved the basal and middle segments of the anteroseptal, anterior, anterolateral, and inferolateral left ventricular walls. Paradoxical outward systolic motion of the enlarging aneurysm was seen, and ejection fraction was 35% (Figure 3A and Movie I in the online-only Data Supplement). The neck of the aneurysm originated along the annulus of the mitral valve, and its location in relation to the left ventricular outflow tract and anterolateral commissure was elucidated from the left ventricular perspective on 3D TEE (Figure 3A and 3B and Movie II in the online-only Data Supplement).

A 128-mm gated computed tomography scan depicted the relationship of the bilobed aneurysm to the left anterior descending and circumflex arteries, respectively (Figure 4). Additionally, a small unruptured left sinus of Valsalva aneurysm was seen (Figure 4). Invasive coronary angiography revealed no significant intrinsic coronary disease; however, dynamic systolic extrinsic compression of the left anterior descending and circumflex arteries was seen (Figure 5).

This patient underwent surgery, which confirmed the spatial anatomy of the aneurysm. The neck of the aneurysm was closed and the mitral valve successfully repaired.

Discussion
Congenital SMA was first described by Abrahams et al in 1962 and is a disorder observed typically in adults of African ancestry. It is thought to be most likely attributable to a congenital defect of the atrioventricular junction. The origin of this aneurysm is usually located along the mitral annulus, most commonly involving the posterior aspect. The main clinical manifestations of SMA are mitral regurgitation, thromboembolism, arrhythmias, sudden death, and heart failure. The mechanisms of mitral regurgitation are multifactorial and include lack of support of the leaflets at the site of the annular deficiency, leaflet retraction attributable to chordal traction secondary to altered ventricular geometry, and rupture of the aneurysm into the left atrium.

The spatial extent of these aneurysms can be challenging to delineate, thus resulting in surgical failure, which is often attributable to either failure to identify additional aneurysm necks (50% of failures) or inadequate closure of the aneurysm. Preoperative assessment has previously been done using transthoracic echocardiography, 2-dimensional TEE, and invasive angiography. The addition of 3D TEE allows accurate identification of the SMA neck, improves assessment of the SMA’s spatial extent, and provides necessary information regarding mitral valve anatomy and the mechanism of mitral regurgitation. Computed tomography also provides further information on the spatial extent of SMA and its relationship to the coronary arteries.

Multimodality imaging aptly delineated the challenging spatial extent of this submitral aneurysm, which caused severe mitral regurgitation, left ventricular systolic dysfunction, and extrinsic coronary compression of both the left anterior descending and circumflex coronary arteries.

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Disclosures
None.

References


Figure 1. Short-axis view in diastole (A) depicting the neck of the aneurysm and the anterior location of the submitral aneurysm (red arrow). Apical 2-chamber view (B) depicting the anterior submitral aneurysm (red arrow).

Figure 2. Severe mitral regurgitation depicted by color Doppler echocardiogram (A) and continuous-wave Doppler echocardiogram (B).

Figure 3. The relationship of the submitral aneurysm (SMA) to the mitral valve (MV) is depicted from the left ventricular perspective using 3-dimensional transesophageal echocardiography during diastole (A) and systole (B). LVOT indicates left ventricular outflow tract.

Figure 4. The bilobed submitral aneurysm and its relationship to the left anterior descending (LAD) and circumflex (CX) arteries are depicted on computed tomography. The yellow arrow depicts a small unruptured sinus of Valsalva aneurysm originating from the left coronary sinus.

Figure 5. Dynamic systolic compression of the circumflex coronary artery (red arrow) and the left anterior descending coronary artery (yellow arrow) are depicted on angiography.
Out of Africa: Congenital Submitral Aneurysm With Extrinsic Compression of the Left Anterior Descending and Circumflex Arteries
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Movie Legends

**Movie 1.** Three-dimensional volumetric analysis of the left ventricle depicts the systolic dyskinesis of the submitral aneurysm involving the basal and midanteroseptal, anterior and anterolateral walls. Best viewed with Windows Media Player.

**Movie 2.** The relationship of the submitral aneurysm to the mitral valve is depicted from the left ventricular perspective using three-dimensional transesophageal echocardiography during diastole and systole. The unsupported area of the mitral leaflets at the posteromedial commissure can also be appreciated. Best viewed with Windows Media Player.