DIAGNOSTIC DILEMMAS

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Section Editor

Rare Cause of Chest Discomfort and Modest Exercise Intolerance in an Athlete

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A 32-YEAR-OLD, 105 kg, 188 cm previously healthy, athletic man presented to the authors' institution complaining of "deep" chest discomfort and back pain after lifting weights and running on a treadmill at his home. He denied orthopnea, paroxysmal nocturnal dyspnea, lightheadedness, syncope, and peripheral swelling, but he stated that he developed palpitations and dyspnea on exertion during more intense exercise (eg, running stairs). Considering his focus on fitness, the patient was surprised by these symptoms, which had modestly limited his admittedly intense exercise routine. The patient also complained about occasional atypical chest pain at rest. His past medical history was notable for congenital pulmonic valve stenosis. The patient underwent a balloon commissurotomy as a 3-year-old boy, but he was lost to follow-up as a teenager when he and his family moved to another state. He denied a family history of acquired or congenital heart disease or sudden cardiac death. The physical examination revealed a widely split second heart sound, a grade III of VI systolic murmur, and a grade IV of VI holodiastolic murmur heard best at the left upper sternal border. Peripheral edema, hepatosplenomegaly, and hepatojugular reflux were absent. The remainder of the physical examination was noncontributory. Right bundle-branch block was present on the electrocardiogram. An exercise stress test demonstrated normal functional capacity and no evidence of myocardial ischemia. Transesophageal echocardiography (TEE) was performed as part of the diagnostic evaluation, and the following images were obtained (Figs 1 and 2; Video clips 1 and 2). What is the diagnosis?

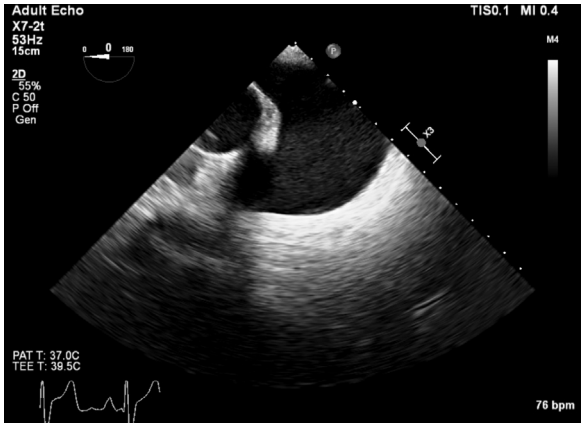

Fig 1. Modified midesophageal ascending aortic short-axis transesophageal echocardiography image.

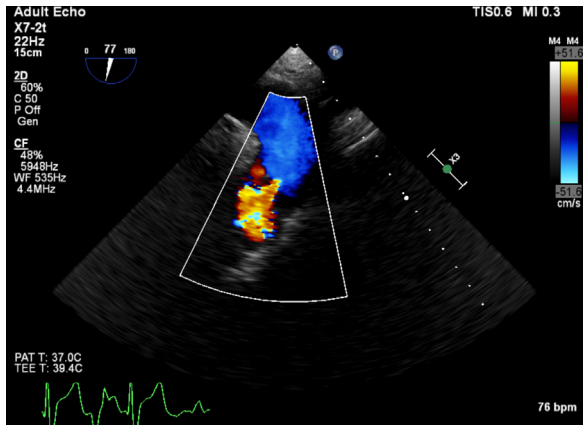

Fig 2. Color Doppler midesophageal ascending aortic short-axis transesophageal echocardiography image during late diastole.

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DIAGNOSIS: LARGE PULMONARY ARTERY ANEURYSM WITH SEVERE PULMONIC VALVE INSUFFICIENCY

The midesophageal ascending aortic short-axis TEE view demonstrated an enlarged main pulmonary artery (PA) (maximum diameter 6 cm) consistent with a PA aneurysm (Fig 1; Video clip 1). Color Doppler blood flow mapping in the same imaging plane showed severe pulmonic valve insufficiency (Fig 2; Video clip 2). The proximal right and left PA branches also were aneurysmal (Fig 3; Video clip 3). Pronounced right ventricular dilatation also was observed (Fig 4; Video clip 4). The pulmonic valve was redundant and dysplastic (Fig 5; Video clip 5) concomitant with the severe insufficiency (Fig 6; Video clip 6). PA systolic pressure was estimated to be 28 mmHg using pulse-wave Doppler interrogation of the tricuspid regurgitant jet velocity. A patent foramen ovale was noted using an agitated saline test. A computed tomography scan with angiographic contrast also identified the 6.7-cm PA aneurysm (Fig 7). The patient was transported to the operating room for repair of the PA aneurysm and pulmonic valve replacement. Direct inspection of the pulmonic valve during cardiopulmonary bypass revealed that it was malformed and had 2 dysplastic leaflets. The native valve was excised and replaced with a 25-mm bioprosthesis. The main PA was reconstructed after resection of a large amount of redundant aneurysmal tissue. The patent foramen ovale was repaired via direct suture plication. The patient separated from cardiopulmonary bypass without inotropic support. He was transferred to the cardiac intensive care unit in stable condition with satisfactory hemodynamics. The remainder of his hospital course was unremarkable.

PA aneurysms are very rare, occurring in approximately 0.007% of the population.[1] Main PA diameter greater than the upper limit of normal (2.9 cm in adults) is used most often to identify a PA aneurysm,[2] although some centers use a diameter of 4.0 cm to define this clinical pathology.[3] As observed in the current patient with a history of congenital pulmonic valve stenosis and a patent foramen ovale, PA aneurysms frequently are associated with congenital heart disease.[4-6] The vast majority (89%) of PA aneurysms involve only the main PA trunk, sparing the left and right PA branches.[7] The current patient had aneurysmal dilatation of the main PA and its proximal branches, as both the left and right proximal PA diameters exceeded 3.0 cm (normal caliber <1.7 cm). The presence or absence of elevated PA pressures often is used to classify PA aneurysms.[6,8] Patients with PA hypertension are more likely to develop PA aneurysmal dilation,[9] which correlates with an increased risk of sudden cardiac death resulting from acute right ventricular failure.[10] Abnormal vascular wall shear stress often contributes to PA aneurysm development in the presence of PA hypertension.[2] A wide range of etiologies for PA aneurysms associated with normal PA pressures have been identified including infection,[1,11,12] connective tissue disease,[6] Behçet vasculitis,[13,14] previous palliative surgery for congenital heart defects,[5] malignancy,[15] chronic pulmonary embolism,[2] and idiopathic causes.[16] As observed in the current patient, poststenotic PA dilatation is another common cause of PA aneurysm formation associated with normal PA pressures before or after remote balloon commissurotomy.[2,5] The patient underwent a balloon commissurotomy of his stenotic pulmonic valve decades before his presentation, but it is likely that the resulting dysplastic bicuspid valve contributed to the development of eccentric blood flow in the proximal PA during right ventricular ejection. This mechanism of vascular damage leading to PA aneurysm formation has been described

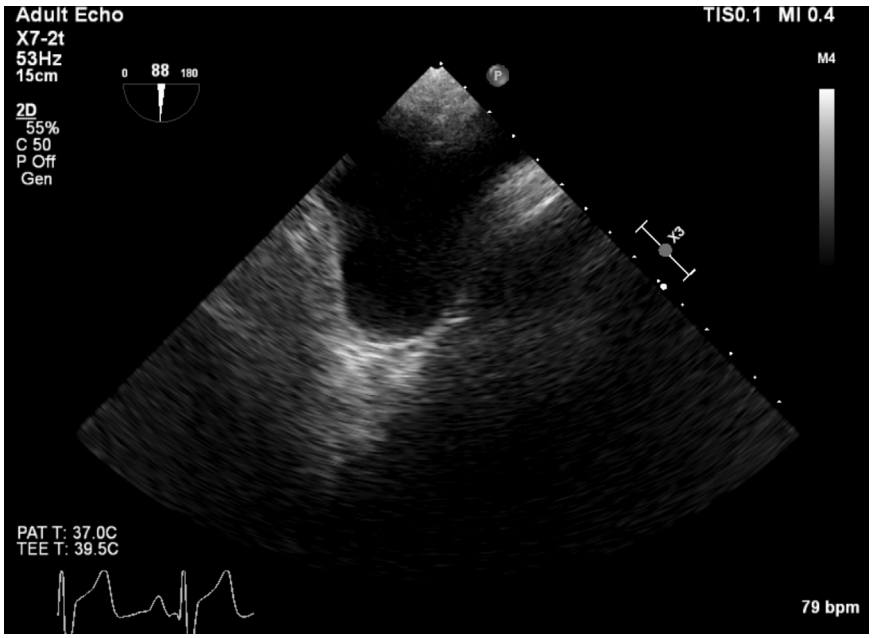

Fig 3. Midesophageal ascending aortic short-axis transesophageal echocardiography image showing aneurysmal dilatation of the main and right pulmonary arteries with relative sparing of the left pulmonary artery.