Aortic dissection masquerading as a bicuspid aortic valve

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Abstract

A 37-year-old male presented to the Emergency Department with acute worsening of back pain and new onset dyspnea. Transthoracic echocardiography revealed moderate left ventricular dysfunction and a bicuspid aortic valve (BAV). In addition, he was noted to have a dilated thoracic aorta concerning for a dissection, severe aortic insufficiency (AI), and both a pericardial and pleural effusion. Magnetic resonance imaging revealed a Type A ascending aortic dissection. He was taken emergently to the operating room for repair. An intraoperative transesophageal echocardiography examination was performed which revealed a normal trileaflet AV with a Type A aortic dissection flap masquerading as a BAV. The dissection flap interfered with both the valve's function, causing severe AI, as well as the valve's appearance, causing it to look bicuspid on echocardiography.

Key words: Bicuspid aortic valve, transesophageal echocardiography, Type A aortic dissection

Introduction

Type A aortic dissection is a potentially devastating complication that commonly presents for emergent cardiac surgery. It can be associated with aortic valve (AV) pathology such as bicuspid AV (BAV) or damaged leaflets. It becomes important to determine whether the native AV is intact and can be spared, or needs replacement. In this case, a normal trileaflet native valve was reported as a bicuspid valve on transthoracic echocardiography (TTE) and could not be adequately visualized on magnetic resonance imaging (MRI) due to motion artifact. An intraoperative transesophageal echocardiography (TEE) exam was suggestive of a normal trileaflet AV. This was eventually confirmed on surgical inspection and resulted in valve-sparing surgery. As illustrated, in this case, the presence of a dissection flap in the vicinity of the AV can complicate its evaluation on imaging. To our knowledge, a dissection flap masquerading as a bicuspid valve has not been previously reported in the literature.

Case Report

A 37-year-old male with a past medical history significant for back pain after a back injury 3 months prior, presented to the Emergency Department with acute worsening of back pain. He also reported new onset dyspnea for the past 12 h and was found to be hypoxic with an oxygen saturation of 60% on room air, which improved with supplemental oxygen. Initial arterial blood gas showed a pH-7.18, PCO$_2$-30, PO$_2$-130, bicarbonate- 11.2, and base balance- $-15.8$. Laboratory investigations were significant for elevated cardiac biomarkers (troponin I-1.89 ng/ml and CK-MB-7.8 ng/ml) and acute renal insufficiency (urea nitrogen- 40 mg/dl, creatinine-3.03 mg/dl). A TTE revealed moderate left ventricular dysfunction with an ejection fraction of 35-40% left atrial (LA) enlargement and a BAV [Video 1 and Figure 1]. In addition, he was noted to have a dilated thoracic...
aorta concerning for a dissection (3.3 cm at the annulus and 3.8 cm at the sinotubular junction), severe aortic insufficiency (AI), and both a pericardial and a pleural effusion. MRI revealed a Type A ascending aortic dissection but the AV could not be adequately visualized due to motion artifact. The patient was taken emergently to the operating room for repair. An intraoperative TEE examination was performed which revealed a normal trileaflet AV with a Type A aortic dissection flap masquerading as a BAV [Videos 2-5 and Figures 2 and 3].

After initiation of cardiopulmonary bypass (CPB), surgical inspection revealed a tear in the ascending aorta extending to the level of the sinotubular junction, sparing the aortic root. The dissection flap was occluding the AV and the left main coronary artery; however, the patient was found to have an intact trileaflet AV [Figure 4]. It became apparent that the dissection flap interfered with both the valve’s function, causing severe AI, as well as the valve’s appearance, causing it to look bicuspid on echocardiography. The patient was cooled to 17°C, a hypothermic circulatory arrest was initiated, and the head was packed in ice. The ascending aorta was repaired with a 26-mm diameter Gore-Tex graft, and the intact native AV was re-suspended. The patient was rewarmed and uneventfully weaned from CPB requiring minimal inotropic support. His post-CPB TEE examination showed adequate repair of the ascending aortic dissection flap, a trileaflet, and competent re-suspended native AV [Figure 5 and Video 6]. The hospital course was prolonged by postoperative complications, including renal insufficiency likely from the state of hypoperfusion during the initial phase of cardiogenic shock as well as the effects of circulatory arrest. The patient eventually recovered and was discharged home on postoperative day #11.

Discussion

The primary method of diagnosis for aortic dissection is either TTE or TEE with a sensitivity of 92% and specificity of 96%. TEE has the advantage of portability, real-time information, and the ability to be performed intraoperatively.

![Figure 1: Preoperative transthoracic two-dimensional echocardiographic image showing parasternal aortic valve short-axis view](image1)

![Figure 2: Transesophageal two-dimensional echocardiographic image showing mid-esophageal aortic valve short-axis view](image2)

![Figure 3: Transesophageal two-dimensional echocardiographic image showing mid-esophageal aortic valve long-axis view](image3)

![Figure 4: Intraoperative view of the aortic valve on surgical inspection after aortotomy revealing a normal trileaflet valve](image4)
in an unstable patient. When available, intraoperative three-dimensional TEE can further provide additional information in aortic dissection cases about the spatial orientation of the flap in relation to the neighboring structures. It can also help delineate coronary involvement.\(^2,3\)

While the gold standard for diagnosing a BAV is echocardiography, it does have limitations if the AV is heavily calcified. A dissection flap in the vicinity of the AV may also limit clear visualization of the cusps. Other diagnostic modalities such as MRI, magnetic resonance angiography (MRA), and computed tomography (CT) enable views of the AV without interference from calcification.\(^1\) Contrast-enhanced CT scanning has a sensitivity of 82-100\% and a specificity varying between 90\% and 100\%.\(^1\) Its disadvantages include the potential inability to localize the primary intimal tear accurately, inability to quantify the severity of AI, radiation exposure, and intravenous contrast use.\(^4\) MRI/MRA allows a better assessment of aortic pathology, particularly due to its ability evaluate the severity of dilation of the ascending aorta with sensitivity and specificity rates in the range of 95-100\%.\(^4\) Disadvantages of MRI/MRA include its limited portability and immediate availability, contraindication in patients with implanted pacemakers or defibrillators, and the relatively increased length of time necessary for imaging.\(^4\) Although the exact incidence is not known, it has been reported that the frequency of BAV in aortic dissection is approximately 4\% of all cases.\(^5\)

In this patient, an initial TEE examination confirmed the presence of an ascending aortic dissection flap, which was prolapsing into the AV orifice [Video 4]. In addition, a doming appearance and coaptation defect of the AV leaflets with severe AI was best visualized in the mid-esophageal AV long-axis view [Video 5]. In the mid-esophageal AV short-axis view [Video 2 and Figure 2], the AV orifice appeared to be elliptical rather than star shaped with a larger rightward leaflet and a smaller leftward leaflet. Although a BAV could not be ruled out, the echocardiographic appearance was atypical, representing its less common form with the fusion of the right and noncoronary cusps. Based on these findings, a congenital bicuspid valve was unlikely and a decision was made to surgically inspect the valve pathology prior to making a definitive decision about the surgical technique (and whether to replace the valve or not).

As demonstrated above, the key to quick identification and diagnosis is vigilance, knowledge about the typical echocardiographic appearance and a high index of suspicion. Communication between the surgical and anesthesia teams regarding atypical TEE findings helped guide further decision-making and surgical management with a successful outcome.

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**Conflicts of interest**

There are no conflicts of interest.

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