

Acute aortic regurgitation in a bicuspid aortic valve due to the rupture of an anomalous cord

A 64-year-old male with acute-onset dyspnea and diastolic murmur was referred to our hospital. Eight months earlier, he had developed atrial fibrillation. At that time, echocardiography showed a reduced ejection fraction of 41% and a bicuspid aortic valve (BAV) with mild aortic stenosis (Fig. 1a, b, Video 1). On admission, echocardiography showed prolapse of the conjoined cusp and severe aortic regurgitation (AR) accompanied by an eccentric jet (Fig. 1c, Video 2). Careful observation revealed a 10-mm-long, highly mobile, thread-like structure attached to the aortic valve on the ventricular surface, which mimicked valvular vegetation (Fig. 1d, Video 2). However, laboratory testing showed no inflammatory reaction. Blood cultures were negative for pathogens. Enhanced chest computed tomography showed mild dilation but not dissection of the ascending aorta. The patient's hemodynamic deterioration prompted urgent surgical intervention. The aortic valve was resected and replaced with a 22-mm ATS Medical prosthesis. Grossly, the excised aortic valve was bicuspid. The conjoined cusp had a small raphe with incomplete commissural fusion, implying a forme fruste BAV. Moreover, it contained an anomalous cord attached by one-and to the raphe near the free margin (Fig. 2). Any signs of infective endocarditis were not found. We diagnosed acute-onset AR caused by the rupture of an anomalous cord in BAV, in which the conjoined cusp had completely lost its coaptation and suspension. If acute-onset severe AR develops in BAV patients, in addition to infective endocarditis and aortic dissection, the rupture of an anomalous cord should be considered.

Video 1. TTE images during the precritical stage showing trivial AR and BAV.

Video 2. TTE images on admission showing severe AR accompanied by an eccentric jet directed toward the interventricular septum. Note the 10-mm-long, thread-like structure attached to the aortic valve.

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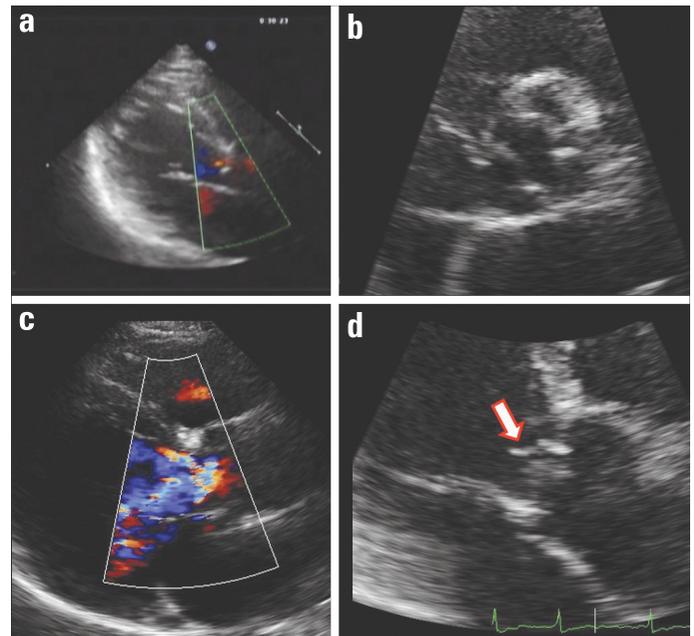


Figure 1. TTE images acquired 8 months earlier (a and b). Long-axis color Doppler view showing trivial AR (a). Short-axis view showing BAV (b). TTE images on admission (c and d). Long-axis color Doppler view showing severe AR (c). Note that severe AR was accompanied by an eccentric jet directed toward the interventricular septum (c). Close observation in the long-axis view revealed a 10-mm-long, thread-like structure attached to the aortic valve (arrow in d)

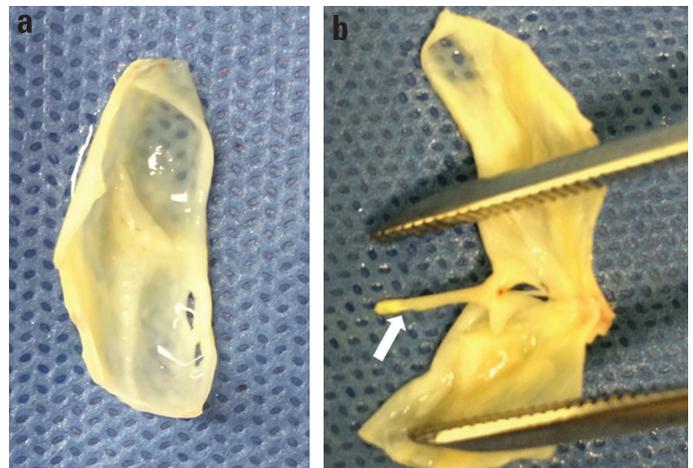


Figure 2. Macroscopic view of the resected aortic valve from the aortic side (a and b). (a) The non-coronary cusp had two fenestrations near the commissure. (a) The conjoined cusp had a small raphe with incomplete commissural fusion. Note the anomalous cord extending from the raphe near the free margin

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