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Case Report

Two rare cases of congenital aortic stenosis showing a discrepancy between preoperative imaging diagnosis, intraoperative findings, and histopathological diagnosis

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ABSTRACT

Unicuspid aortic valve (UAV) is an extremely rare congenital heart valve abnormality while bicuspid valve (BAV) has been reported as one of the most common cardiac anomalies. With a UAV usually showing similar presentations to a BAV, such as aortic regurgitation or aortic stenosis (AS), it is challenging to differentiate them from each other in clinical settings. Despite some features shared between both valve disorders, there can be a clinical significance in distinguishing UAV from BAV for the management of patients with these heart anomalies. Herein, we describe two cases where patients with hemodynamically severe AS were diagnosed with BAV and UAV, respectively based on preoperative examinations and intraoperative findings, but subsequent pathological examinations confirmed the opposite diagnosis in both cases.

<**Learning objective:** Preoperative diagnosis of congenital aortic valve diseases can often be challenging. There remains a remarkable number of misleading cases. Thus, it is strongly recommended that an accurate diagnosis should be attempted at the earliest stages of congenital aortic valve disease. Additionally, both careful follow-ups using multiple imaging modalities and confirmations via pathological diagnosis for patients undergoing surgery, if they are first found to be at an advanced stage or remain undiagnosed preoperatively are important.>

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Introduction

Unicuspid aortic valve (UAV) is a rare congenital heart anomaly with an estimated frequency of 0.02% among the adult population [1] and often can present as a form of significant aortic stenosis (AS) or aortic regurgitation (AR) in symptomatic relatively young subjects who sometimes require cardiac surgery. Bicuspid aortic valve (BAV), on the other hand, is one of the most common cardiac malformations with an estimated incidence of 0.9–2% [2], which shows similar clinical manifestations to UAVs, as related to valve dysfunction, and is often accompanied with abnormalities of the aorta (aneurysm, dilatation, and dissection) and other cardiac malformations [3].

Despite some features shared between both valve disorders, there can be a clinical importance in distinguishing UAV from BAV for the management of patients with these heart anomalies. Serial assessments using transthoracic echocardiography (TTE) have disclosed that valve dysfunctions progress more rapidly in patients with UAV than those with BAV [4] and that UAV patients are more likely to develop symptoms at an early age [1]. Regarding the anatomical aspects, Noly et al. [5] demonstrated that there are several distinct characteristics between them. The rate of aortic dilatation was significantly lower in UAV than in BAV cases, leading to a relatively low incidence of acute aortic events in UAV. In contrast, the aortic annulus was dilated (>25 mm) in most patients with UAV requiring surgery in 71%. Consequently, preoperative discrimination of the two is of significant importance in determining an appropriate surgical approach, whether it be

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aortic valve repair or replacement, with these findings supporting the concept that UAV and BAV are two discrete entities. Besides, there is a growing interest in congenital aortic valve disorders, as indications for transcatheter aortic valve implantation have been expanding to include patients with congenital aortic valve disease, particularly bicuspid aortic stenosis from those with calcified and degenerative AS [6]. Nevertheless, preoperative diagnosis of UAV is rare and sometimes misleading. A systematic review reported that only 23 cases of 231 UAV patients were identified preoperatively using TTE while an additional 16 cases were diagnosed through transesophageal echocardiography (TEE).

In this report, we describe two cases that focus on the difficulty in discriminating between UAV and BAV, both preoperatively and intraoperatively, with careful pathological examination of surgical specimens helping to reach a final diagnosis.

Case report #1

A 30-year-old woman was referred with worsening exertional dyspnea. On physical examination, grade 4/6 systolic ejection murmur was audible at the right sternal border. A TTE displayed a normal left ventricular (LV) size with preserved systolic function [LV ejection fraction (LVEF), 69%], complicated by a thickened aortic valve with spotty calcification, which was assessed as a severe AS (aortic valve area calculated by Doppler method, 0.67 cm^2), along with a trivial AR. This examination, however, did not provide a detailed observation on the leaflet morphology due to inadequate image qualities. A subsequent TEE, however, revealed that the etiology of her valve abnormality could be BAV based on clear three-dimensional live images of the two leaflets in both lateral positions and two commissures between them without any raphe structure with an eccentric orifice (Fig. 1A and B). A cardiac computed tomography (CT) revealed normal coronary arteries without any dilatation of the ascending aorta (Fig. 1C) with two cusps of almost the same size lined up with two commissures between them, which were indicative of a mildly calcified BAV. At operation, the valve was replaced with a 19-mm prosthetic valve and was reported as a BAV based on the removed valve specimen (Fig. 1D and E). A macroscopic pathological examination, however, revealed that it was an acommissural UAV. The surgical specimen exhibited an eccentric orifice with no obvious commissures confirmed at each edge. The area that was thought to be a commissure on the TEE examination was almost the same width as the other areas of the leaflets with the elastic fibers in the commissure-like area displayed continuously in "its circumferential direction" (Fig. 1F). A similar observation was obtained on the other edge, which supported the claim that the congenital valve consists of one piece of leaflet, not two leaflets with commissural structures.

Case report #2

A 50-year-old female with a history of hyperthyroidism was referred complaining of a gradual development of dyspnea. A TTE showed a severe AS (peak transaortic velocity, 4.5 m/s) with mild to moderate AR while the systolic function was preserved (LVEF, 65%) with a normal LV size. A TEE was performed because close observation on TTE was difficult due to the extremely heavy calcification of the valve. The TEE findings suggested that the valve had a single unfused commissure, or unicommissural UAV, attached to the aorta between the right coronary cusp (RCC) and left coronary cusp (LCC) positions in the short-axis view

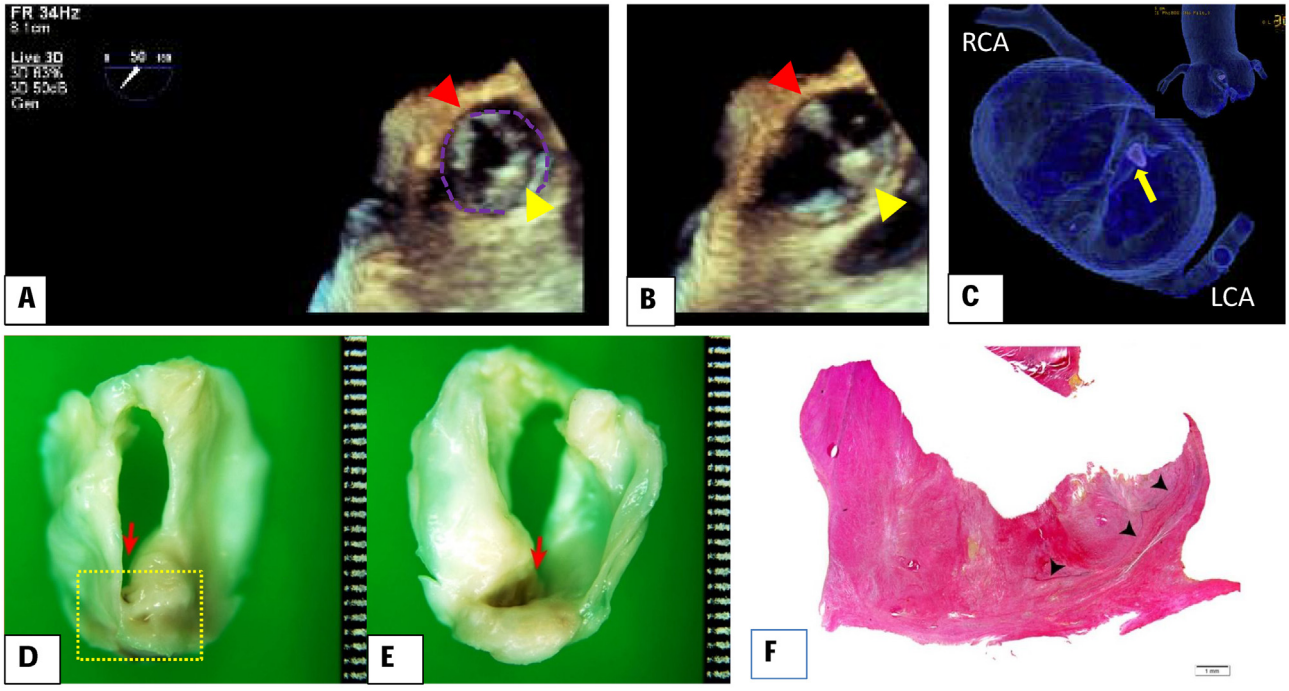

Fig. 1. Three-dimensional live transesophageal echocardiography (short-axis view) during mid-systole (A) and mid-diastole (B), showing two commissures in the anterior (yellow head) and posterior positions (red head) in the absence of raphe, and the right-left located leaflets (dotted purple line) formed an eccentric orifice opening. (C) Three-dimensional cardiac CT scan described two leaflets at almost same size were lined with two commissures, mimicking BAV with a trivial calcification (yellow arrow). Surgical specimen of the removed aortic valve from aortic side (D) and from LV side (E). The shape of orifice was eccentrically long and narrow with no visible commissures confirmed on each edge. The sharply curved area (red arrow) held a similar width to that of other sections of the leaflet, which on LV side was accompanied by the formation of a nodular calcification. (F) Microscopic findings of the curved section on hematoxylin and eosin stain. Elastic fibers in the commissure-like area displayed continuous sequence in "its circumferential direction" (black arrowheads).
RCA, right coronary artery; LCA, left coronary artery; CT, computed tomography; BAV, bicuspid aortic valve; LV, left ventricle.

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