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Case Report

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Multimodal Imaging Analysis in A Child with Aortico-Left Ventricular Tunnel and Left Ventricular Noncompaction

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1. Case Report

An 11-year-old girl was admitted to the hospital with new-onset chest tightness with exercise. Physical examination revealed a harsh systolic and diastolic flow murmur, grade 4/6, loudest between the second and third intercostal spaces along the left sternal border, with radiation throughout the precordium. The second auscultation area of the aortic valve revealed a severe breath-like murmur with extensive conduction. A chest radiograph demonstrated widening of the aortic shadow and cardiomegaly with a characteristic left ventricular (LV) contour. Transthoracic echocardiography (TTE) and transesophageal echocardiography（TEE）showed a moderately dilated LV (LV end-diastolic diameter, 56 mm) with normal LV wall motion. The parasternal long-axis view outside the aortic valve ring showed a tunnel-like structure in front of the right coronary sinus (Figure 1A, 1B ; Video S1, S2), ranging from 27 × 14 mm, with an inner diameter of 7 mm at the entrance of the aorta ascendens and 15 mm at the lateral outlet of the LV outflow tract; the tunnel was also clearly seen on the three-dimensional reconstruction of the echocardiographic image (Video S3). In addition, TTE revealed that the aortic valve had a trefoil structure and was thickened and rough, especially the right coronal valve, with moderate aortic insufficiency. The interventricular septum was aneurysmal and thin and bowed into the right ventricle.

Color Doppler echocardiography showed systolic blood flow signals entering the aorta through the tunnel from the LV outflow tract. During diastole, two clear high-speed colourful blood flows into the LV outflow tract from the tunnel and aortic valve were visualized (Figure 1C, 1D; Video S4, S5). A left-to-right shunt was seen at the fossa ovalis. The pulsed-wave Doppler mode showed a biphasic turbulent flow spectrum in the tunnel. In addition, TTE demonstrated LV hypertrabeculation from its mid portion to the apex with a ratio of nondense-to-dense myocardium of 2.3:1 (Figure 2A; Video S6), a finding also confirmed by contrast echocardiography (Figure 2B; Video S7, S8). Cardiac computed tomography (CT) and CT three-dimensional reconstruction visualized a tunnel in the anterior wall of the aorta on the right coronary valve and showed an LV outflow tract with a diameter of about 5.7 mm, protruding to the right ventricle with a calibre of 13.0 mm and a depth of 10.2 mm (Figure 3A, 3B, 3C). Cardiac magnetic resonance imaging (CMR) showed the anterolateral side of the aortic valve ring (anterolateral side of the right coronary sinus) and a tunnel connecting with the LV outflow tract (Figure 4A, 4B, 4C; Video S9). Cardiac CT and CMR also confirmed the failure of myocardial compaction, resulting in a hypertrabeculated myocardium with a noncompact/compact myocardium ratio greater than 2:1(Video S10).

Based on these findings, the diagnosis was made of congenital heart disease, aortico-left ventricular tunnel (ALVT), noncompaction of ventricular myocardium (NVM), and moderate aortic insufficiency. During this hospital admission, the patient underwent surgical intervention to repair the ALVT， the aortic valve leaflets were excised, and the patient underwent mechanical aortic valve replacement. The intraoperative findings were consistent with the diagnosis based on the preoperative imaging (Figure 5A, 5B). The

ALVT was successfully repaired during the operation, and the prosthetic aortic valve functioned well. Postoperative transesophageal echocardiography confirmed the absence of the tunnel and that the prosthetic aortic valve functioned well, with no significant perivalvular leak (Video S11, S12). Histopathology of the aortic valve and right coronary sinus revealed fibrous tissue and findings consistent with chronic inflammation (Figure 6A, 6B). Postoperative TTE showed that the LV was smaller (LV end-diastolic diameter 40 mm) than before surgical intervention (56 mm). The most striking finding in this case was the postoperative significant reduction of LV noncompaction. The patient's postoperative course was uneventful. No murmur was heard in each valve area, and she was discharged on postoperative day 17.

As a rare congenital cardiac anomaly, ALVT represents only 0.001%–0.5% of cases of congenital heart disease [1]. This condition is an abnormal tunnel between the aortic root and the left ventricle, which is associated with elastic fibre dysplasia in the aortic sinus, resulting in tunnel-like changes in the aortic root [2]. Regardless of clinical severity, surgery should be performed early upon diagnosis of ALVT because it can cause serious cardiac insufficiency, has a poor prognosis for nonsurgical patients and has a high mortality rate due to congestive heart failure. Also, diagnosed in this patient was NVM, an unclassified cardiomyopathy that is characterized by deep trabeculations in the ventricular wall and by varying degrees of ventricular systolic performance, resulting from intracardiac arrest in the normal process of myocardial compaction. Two distinct processes exist as the embryologic and pathophysiologic basis for noncompaction:(1) primary noncompaction due to failure of development of the compact layer of the myocardium and (2) secondary noncompaction due to failure of regression of embryonic myocardial trabeculations as a consequence of lesions producing volume and/or pressure overload.

Both ALVT and NVM are rare congenital cardiac anomalies with varied clinical presentations, and their co-existence is sparsely described in the medical literature. The mechanism of the relationship between these two conditions has not been well-delineated; however, their association is supported by the relationship of LV noncompaction with congenital heart block, which causes ventricular volume overload [3]. The excessive cardiac pressure load and insufficient myocardial blood supply interfere with the normal closure of embryonic myocardial trabecular space, causing secondary myocardial incompressibility. This case suggests an association of these entities, possibly related to ventricular volume overload and demonstrates the concerns and complications that require continued follow-up.

The association of ALVT with NVM is a rare combination of congenital abnormalities. This case highlights the value of multimodal imaging analysis for the detailed assessment of ALVT and NVM, and to observe the changes of LV noncompaction before and after operation.

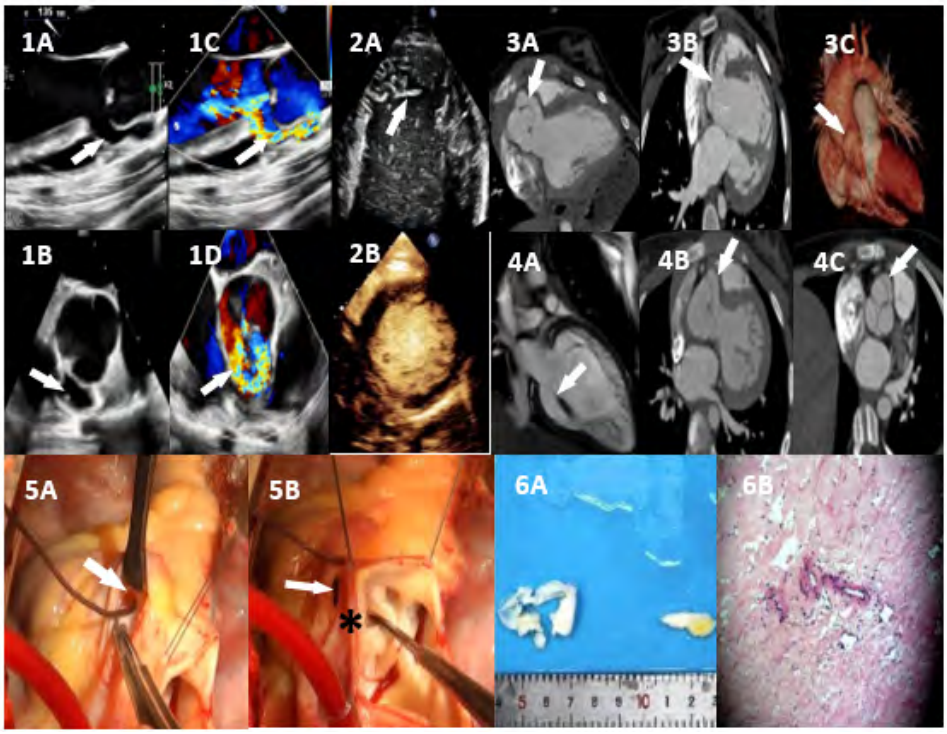

Figure 1A-1B: TTE shows outside the aortic valve ring showed a tunnel-like structure in front of the right coronary sinus. Figure1C,1D CDFI shows systolic blood flow signals entering the aorta through the tunnel from the LV outflow tract. During diastole, two clear high-speed colourful blood flows into the LV outflow tract from the tunnel and aortic valve were visualised, and the blood flow in the tunnel goes back and forth.

Figure 2: TTE and contrast echocardiography shows LV hypertrabeculation from its mid portion to the apex with a ratio of nondense-to-dense myocardium of >2.

Figure 3: CT and CT three-dimensional shows visualised a tunnel in the anterior wall of the aorta on the right coronary valve and showed an LV outflow tract with a diameter of about 5.7 mm, protruding to the right ventricle with a calibre of 13.0 mm and a depth of 10.2 mm.
Figure 4: CMR showsthe anterolateral side of the aortic valve ring (anterolateral side of the right coronary sinus) and a tunnel connecting with the LV outflow tract.
Figure 5: ALVT was probed intraoperatively (* to show the RCA ostium in Figure 5B.
Figure 6: Histopathology of the aortic valve and right coronary sinus revealed fibrous tissue and findings consistent with chronic inflammation.

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3. Disclosures

None.

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