

Case Report

A case report of fetal aortico-left ventricular tunnel complicated with endocardial fibroelastosis diagnosed by ultrasonography

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Abstract: A rare case of fetal heart malformation screened by color Doppler was reported in this study. The fetus was diagnosed with aortico-left ventricular tunnel (ALVT) at 30th-week gestation. Ultrasonography showed that the typical phenotypes of this case were as follows: The aorta was divided into two parts by the carination structure at the aortic annulus. Valve echo was not detected in the lateral cavity near the interventricular septum. Valve echo was detected on the other side and showed a square box-like shape in the systolic period. Abnormalities in the aorta were found in the fetus after induction; the carination structure was seen in the center of aortic lumen. Enlargement of the left ventricle and thickening of the endocardium were observed. Pathology showed significant hyperplasia of endocardial fibroelastosis.

Keywords: Aortico-left ventricular tunnel, endocardial fibroelastosis, ultrasonography, fetus, case report

Introduction

Aortico-left ventricular tunnel (ALVT) is a rare congenital heart disease characterized by the existence of perivalvular vice-transport between the ascending aorta and left ventricle. Most previous studies were case reports from Children's Cardiovascular Centers [1, 2]. ALVT is often accompanied by progressive left ventricular dilatation needs early correction and its incidence was lower than 0.1% of congenital heart disease. Moreover, the fetus has rarely reported to have been combined with such a malformation, and most reports are limited to infants [3, 4]. Echocardiography is the preferred method for non-invasive preoperative diagnosis of the ALVT and can accurately describe the type and involvement of the cardiac structure [5]. In the prenatal examination of pregnant women, we accidentally found a case of such a cardiac malformation by echocardiography.

Case report

The 27-year-old pregnant woman had no history of pregnancy. She was healthy, without family history of infertility, adverse exposure history

and drug use during pregnancy. Routine fetal heart ultrasonography was performed at 30 weeks of pregnancy; it was found that the heart was in the normal position with the apex pointing to the left front and the atrium in the correct position; a right ventricular loop was also observed. The four chambers of the heart were obviously abnormal. The transverse diameters of the left atrium, left ventricle, right atrium, and right ventricle were 13.0 mm, 16.8 mm, 10.6 mm, and 9.6 mm, respectively. The aortic dimension was 9.7 mm, and the pulmonary artery diameter was 6.4 mm. The left ventricular endocardium showed uneven thickening and echo enhancement, with 5.9 mm at the thickest point. Further, the motion amplitude of the ventricular wall was decreased (**Figure 1**). The echo of the papillary muscle and tendon was not detected in the left ventricle. The diameter of the oval foramen was 6.4 mm, and the echo of the foramen ovale valve was detected in the left atrium. The origins of the aorta and pulmonary artery were normal with an increased internal aortic diameter and a normal range of pulmonary artery internal diameter. The aorta was divided into two parts by the carination

Fetal aortico-left ventricular tunnel

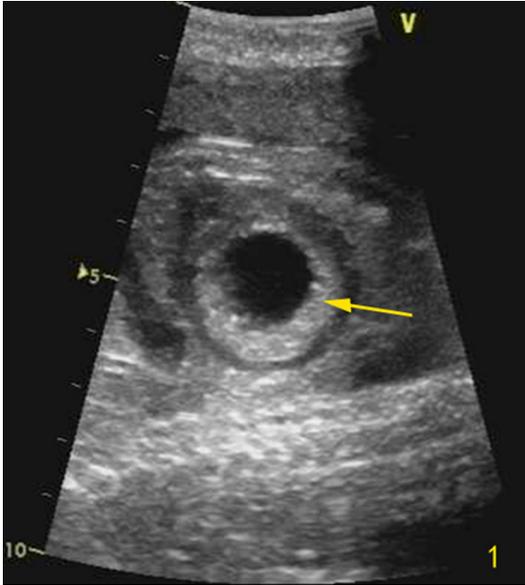


Figure 1. The left ventricular endocardium showed uneven thickening and echo enhancement.

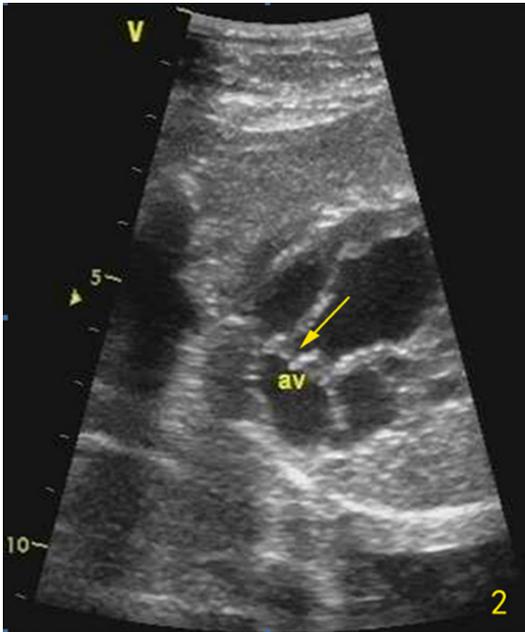


Figure 2. The aorta was divided into two parts by the carination structure at the aortic annulus, and valve echo was not detected in the lateral cavity near the interventricular septum.

structure at the aortic annulus, and valve echo was not detected in the lateral cavity near the interventricular septum (**Figure 2**). The blood flow in this cavity was “reciprocating” with the relaxation and contraction of the heart (**Figure 3**). The valve echo was detected on the other

side and showed a square box-like shape in the systolic period (**Figure 4**). The forward flow rate of the aortic valve was increased to 1.85 m/s. A large amount of regurgitation signals were detected during the relaxation period, and the regurgitation flow rate was 2.67 m/s. A small amount of regurgitation signals were detected at the mitral valve. The ultrasound indicated that the development of the fetal heart was abnormal. The considerations were as follows: 1. Abnormal development of the aortic valve and aortic valve stenosis complicated with insufficiency; 2. ALVT; 3. Mitral valve dysplasia and few regurgitations; 3. Increased thickness of the endocardium, considering the endocardial fibroelastosis; 4. Dilatation of the ascending aorta and a reversed flow in the transverse arch of the aorta; and 5. Enlarged left heart and impaired heart function.

Informed of the situation, pregnant women voluntarily accept abortion, which is permitted by Chinese law. The fetus of the induced labor was a male, and the development of the external features was normal (height: 40 cm; weight: 1500 g). Anatomy showed that the heart was in the normal position in the chest, with the atrium in the correct position; right ventricular loop and enlarged left ventricle were also observed. The origins of the aorta and pulmonary artery were normal with coarse aorta. However, the morphology of the outflow tracts of the ascending aorta and left ventricle was abnormal. Scissoring along the ascending aorta was observed, and the aortic lumen was divided into two parts by the carination structure at the center. One side of the lumen near the interventricular septum was smooth, and no valve-like structure was detected. While several short valves were observed on the other side of the lumen, the valves were thickened and contracted (**Figure 5**). The left and right ventricles were cleaved with an intact ventricular septum. Enlargement of the left ventricle and thickening of the endocardium were observed. The valve cusp of the mitral valve showed a malformation, and the number of tendons was reduced. The tendons were directly connected to the ventricular wall, and a normal papillary muscle structure was not observed (**Figure 6**). No significant abnormalities were found in the right ventricle. The pathological diagnosis included aortic valve dysplasia and stenosis, ALVT, secondary endocardial fibroelastosis, and mitral valve dysplasia. Endocardial fibroelastosis in

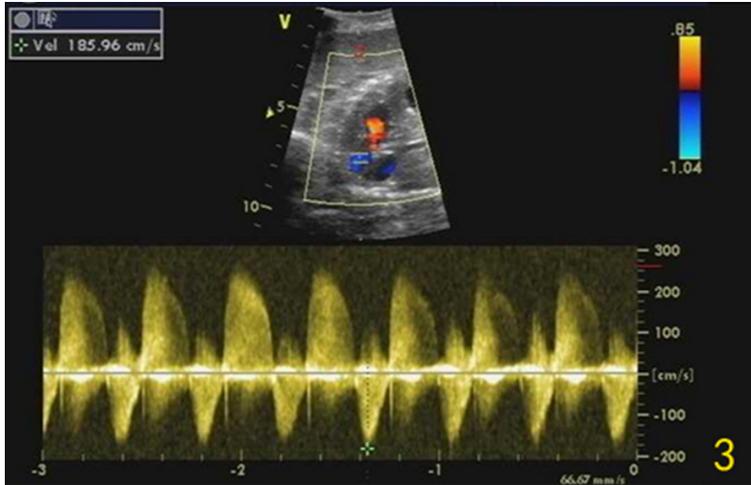


Figure 3. The blood flow in this cavity was “reciprocating” with the relaxation and contraction of the heart.

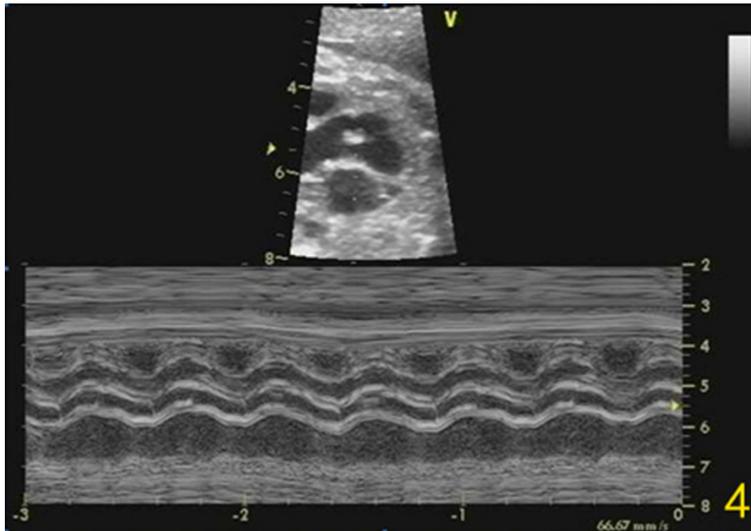


Figure 4. The valve echo was detected on the other side and showed a square box-like shape in the systolic period.

the left ventricle was confirmed by endocardium pathology (**Figure 7**).

Discussion

The embryological mechanism of ALVT is still unclear and is mostly due to the dysplasia of the elastic fibers in the aortic sinus, resulting in the tunnel-like change in the root of the aortic sinus [6, 7]. ALVT could be divided into several types and complicated with aortic valve stenosis or insufficiency, bilobular malformation of the aortic valve, patent ductus arteriosus, pul-

monary valve stenosis, coronary artery abnormalities, etc [8-10]. Limited reports suggested that the incidence of this disease was low; however, the mortality of fetuses after birth was high [11, 12]. Epidemiological data of ALVT in the Han population are lacking, and no fetal ALVT had been reported yet.

The carination structure at the aortic annulus was detected by fetal ultrasonography in this case. One side showed no valve-like structure or valve activity, and the tunnel-like structure was formed between the aorta and left ventricle. The blood in the left ventricle flowed into the aorta through the tunnel and aortic valve during the systolic period, and flowed backward through the tunnel during the relaxation period; so the overmuch blood in the aorta lead to an increased capacity load of the ascending aorta and outflow tract of the left ventricle, which further led to the enlargement of the left ventricle and dilatation of the ascending aorta. The blood flowing backward from the tunnel section of the aortic annulus to the outflow tract of the left ventricle was eccentric and not located on the side of the valve. The blood flow spec-

trum in this case was similar to that in absent pulmonary valve syndrome [13].

Pathologic biopsy confirmed endocardial fibroelastosis in the left ventricle. Research suggests mesenchymal cells of the embryonic heart expand and contribute to the majority of endocardial fibroelastosis [14].

In this case, fetal ultrasonography showed an obviously enlarged and spherical left ventricle, thin left ventricular wall, diffusely thick left ventricular endocardium, enhanced echo, and reduced ventricular wall motion, which were in

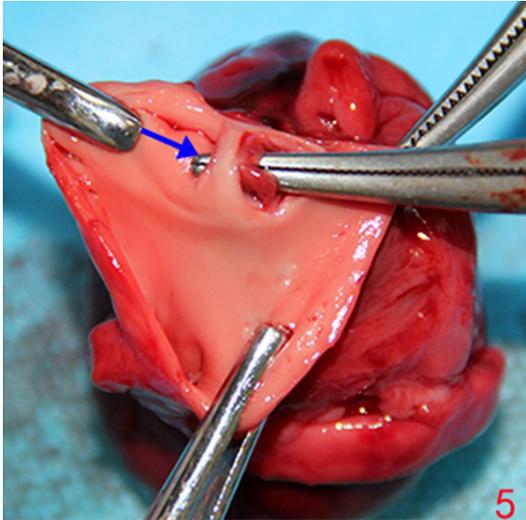


Figure 5. Several short valves were observed on the other side of the lumen, the valves were thickened and contracted.

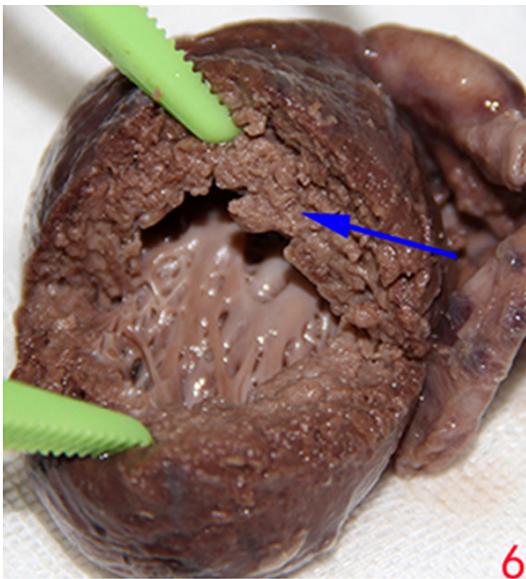


Figure 6. The tendons were directly connected to the ventricular wall, and a normal papillary muscle structure was not observed.

accordance with the ultrasonographic alterations of endocardial fibroelastosis [15].

Acknowledgements

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Disclosure of conflict of interest

None.

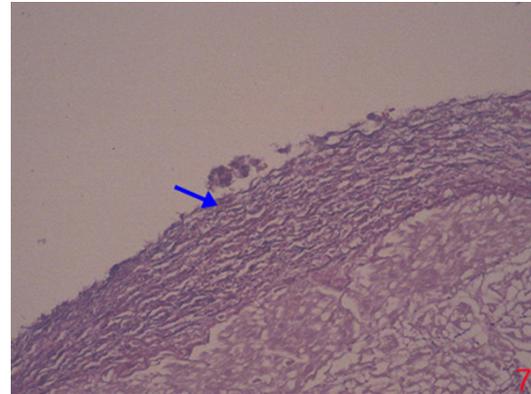


Figure 7. Endocardial fibroelastosis in the left ventricle was confirmed by endocardium pathology.

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