Scientific Research Publishing

Case Reports in Clinical Medicine, 2022, 11, 194-199
https://www.scirp.org/journal/crcm
ISSN Online: 2325-7083
ISSN Print: 2325-7075

Fetal Endocardial Fibroelastosis with Coarctation of Aortic Arch: A Case Report

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How to cite this paper: Ding, J., Liu, B.H., Sun, B.Y. and Hong, Y. (2022) Fetal Endocardial Fibroelastosis with Coarctation of Aortic Arch: A Case Report. *Case Reports in Clinical Medicine*, **11**, 194-199.
https://doi.org/10.4236/crcm.2022.115029

Received: April 3, 2022
Accepted: May 24, 2022
Published: May 27, 2022

Abstract

This study describes a 34-year-old pregnant woman who underwent prenatal echocardiography at 24 weeks of gestation and found fetal heart abnormalities. She underwent fetal echocardiography in our hospital. Echocardiography showed complex malformation of fetal heart, thickening and calcification of left ventricular myocardium and tendon, weakening or even flattening of left ventricular wall movement, very severe mitral stenosis, severe mitral regurgitation, aortic stenosis, narrowing of the aortic arch, countercurrent of the ductal arch to aortic arch, small diameter of the oval foramen and significantly increased flow velocity. The patient decided to induce labor after expert consultation, but no autopsy was performed due to his family's refusal. Color Doppler ultrasound can find the positive signs of endocardial elastofibroplasia earlier, and accurately diagnose patients with neonatal elastofibroplasia. It provides accurate diagnostic information for the clinic and can be used as the first choice. This report has obtained the informed consent of its parents.

Keywords

Endocardial Fibroelastosis, Coarctation of the Aorta, Prenatal Ultrasound Diagnosis

1. Introduction

Endocardial fibroelastosis (EFE) is also known as endocardial sclerosis. EFE was first reported in 1740 and named by Weinberg in 1943. Its pathogenesis has not been fully elucidated. Many scholars believe that it is closely related to intrauterine infection, and can also be secondary to left ventricular dysplasia syndrome, aortic stenosis, atresia, the abnormal origin of the coronary artery, and other cardiac malformations. It is a common kind of infant primary cardiomyopathy.

There is diffuse hyperplasia of endocardial collagen and elastic fiber tissue [1], resulting in thickening and hardening of the endocardium, which can eventually lead to cardiac enlargement and heart failure. In this study, the value of ultrasound in the diagnosis of a case of fetal endocardial fibroelastosis was discussed. The report is as follows.

2. Case Presentation

A pregnant woman, female, 34-year-old, 24 weeks pregnant, underwent prenatal ultrasound diagnosis in our hospital. Fetal echocardiography was performed in our hospital. Pregnant women are generally in good health. They deny the history of cold during pregnancy, and have no history of adverse pregnancy, have no history of close relative marriage and genetic diseases, and the children born by cesarean section are in good health. The fetal echocardiography showed that the left and right ventricles of the fetus were asymmetric in size (the inner diameter of the left atrium was about 8.6 mm, the inner diameter of the left ventricle was about 8.4 mm, the inner diameter of the right atrium was about 10.8 mm and the inner diameter of the right ventricle was about 9.9 mm), and the left ventricular myocardium and tendon were significantly thickened and calcified. (**Figure 1**) Left ventricular wall motion is weakened or even flat. (**Figure 2**) The mitral valve is short and small, the valve is thickened and the echo is enhanced. There is no obvious mitral valve opening and closing activity, the mitral valve opening is limited, and a large number of reflux signals can be seen. (**Figure 3**) The tricuspid valve is slightly thickened, the leaflets are slightly lengthy, the echo is enhanced and slightly stiff, and the opening range is acceptable.

The inner diameter of the aorta is thin (the inner diameter of ascending aorta is about 2.0 mm, the inner diameter of the transverse arch of the aortic arch is about 1.8 mm, and the inner diameter of the aortic isthmus is about 2.1 mm), the inner diameter of aortic valve ring is about 2.7 mm, and the inner diameter of the main pulmonary artery is about 5.0 mm. The cross-relationship between

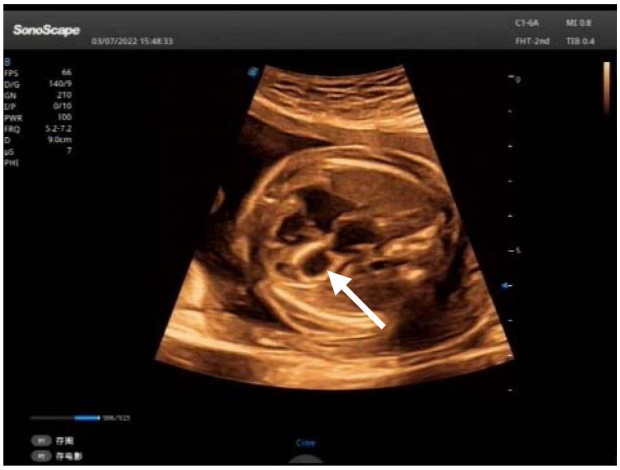

Figure 1. Left ventricular myocardial thickening and calcification in apical four-chamber view (left ventricular myocardium thickened and calcified by arrow).

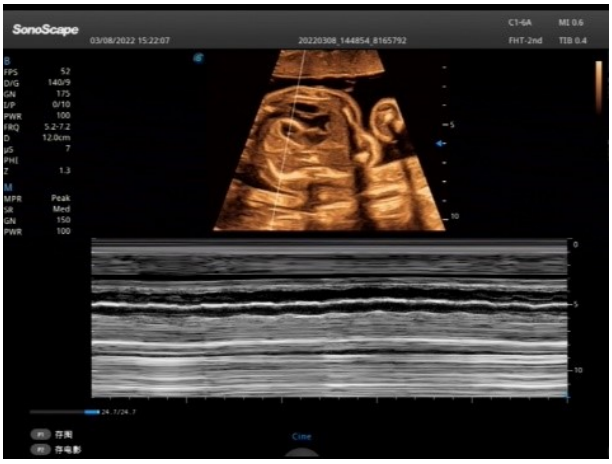

Figure 2. M-mode echocardiography showed decreased and flat left ventricular wall motion.

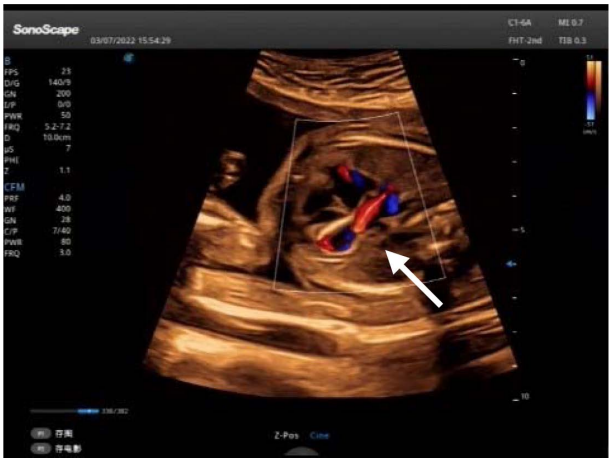

Figure 3. A large number of mitral regurgitation signals can be seen on CDFI (as the arrow shows).

the pulmonary artery and aorta is normal, and the left ventricular outflow tract and aortic arch are severely narrowed. It can be seen that the arterial catheter supplies the aortic arch in reverse, and the local blood flow of the aortic arch is in reverse. (**Figure 4**) The ratio of the pulmonary artery to aortic diameter on the three-vessel trachea section increased, and the blood flow of the pulmonary artery and aorta was reversed on CDFI. (**Figure 5**) The velocity of the aortic valve is about 78 cm/s and that of the pulmonary valve is about 82 cm/s.

The opening of the foramen ovale is to the right, with an inner diameter of about 1.9 mm. The flow velocity at the foramen ovale increases significantly, and the peak flow velocity is about 153 cm/s. (**Figure 6**)

The patient decided to induce labor after expert consultation, but no autopsy was performed due to his family's refusal.

3. Discussion

Endocardial fibroelastosis (EFE) is a rare heart disease in infancy. The pathological manifestations are excessive deposition of collagen and elastin in the

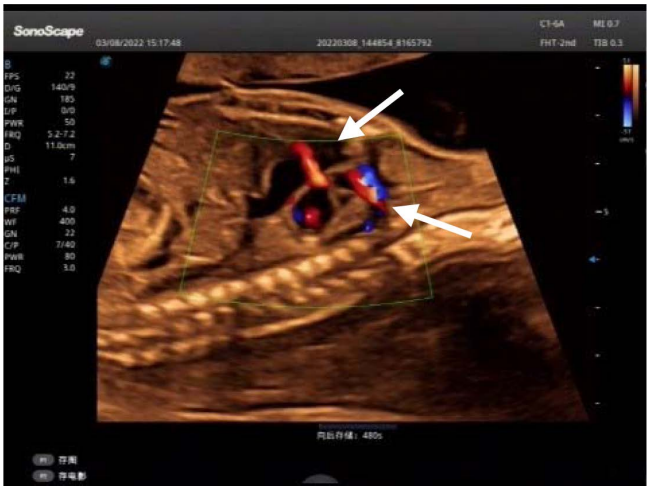

Figure 4. The reverse flow of the aortic arch in the same direction as the flow of foramen ovale (as the arrow shows).

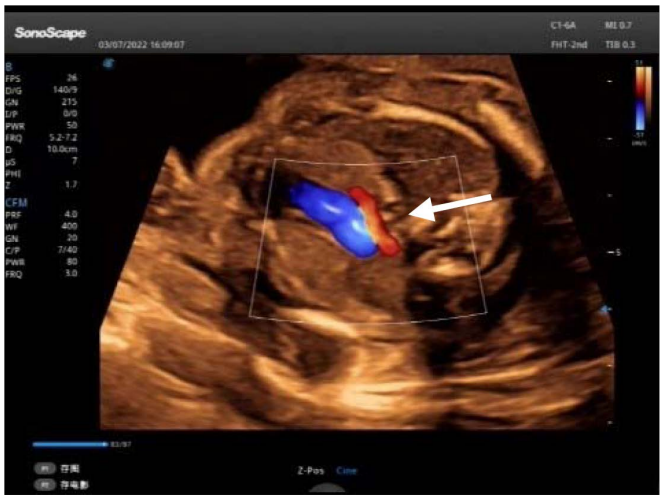

Figure 5. Three-vessel trachea section shows that the proportion of the inner diameter of the main pulmonary artery increases and the blood flow is reversed (as the arrow shows).

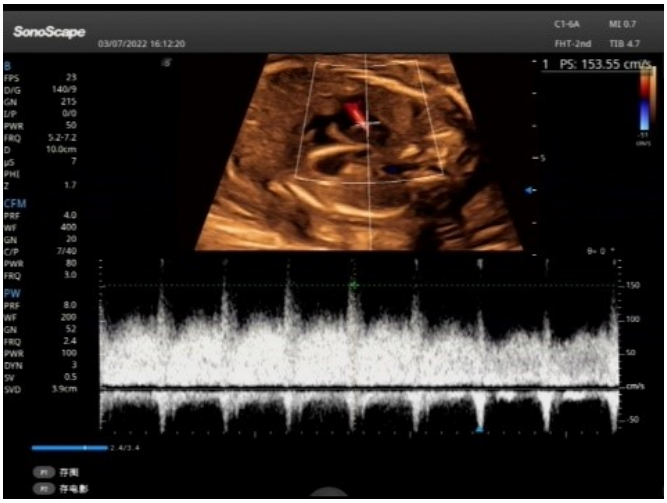

Figure 6. Peak velocity of the oval hole 153 cm/s.

endocardium and hyperplasia of fibrous tissue, which eventually changes the morphology and function of the left ventricular endocardium. A previous study by Pek *et al.* [2] suggested that abnormal endothelial-to-mesenchymal transition (EndoMT) could explain EFE pathogenesis. In the neonatal stage, the abnormal transformation of EndMT will lead to the development of fibroelastosis [3] [4] [5]. In addition, virus infection and immune response may also lead to the occurrence of diseases.

EFE has two ways: primary and secondary. Generally, primary lesions exist alone and are not combined with other congenital heart diseases, while secondary lesions are often combined with other cardiovascular malformations, such as coarctation of the aorta (COA), aortic valve stenosis, left ventricular dysplasia syndrome, and incomplete myocardial densification [6] [7] [8]. In this case, the fetal heart cavity was out of proportion. The left ventricular myocardium and tendon were diffusely thickened, and the left ventricular wall motion was weakened and flat due to calcification; Mitral valve leaflets are thickened, calcified, and restricted in the opening. Thin and narrow blood flow can be seen in diastole, but a large number of blood flow reflux signals can be seen in systole, resulting in increased left atrial volume and increased pressure, resulting in high-speed left to right septal blood flow signals (153 cm/s) in foramen ovale, resulting in limited blood flow in foramen ovale. At the same time, since the upper and lower parts of the aorta supply the left and right hearts respectively during the fetal period, the blood volume received by the left and right cardiac cavities accounts for almost 1/2 of the venous return flow of the whole body. Any situation that can cause the decrease of fetal aortic blood flow will inevitably lead to the increase of blood flow through the arterial catheter, resulting in the decrease of aortic and aortic arch blood flow, resulting in serious stenosis, resulting in the reverse perfusion of ductal arch blood flow, Therefore, on the three-vessel section, the proportion of pulmonary artery to aorta increases and the blood flow is reversed. Endocardial fibroelastosis (secondary) was diagnosed.

4. Conclusion

Ultrasound is the first choice for the diagnosis of EFE. If you encounter EFE in the fetal period, you should pay attention to observe whether it is combined with other malformations to determine whether it is secondary EFE, which can provide some ideas for clinical diagnosis and treatment. In this case, pregnant women's early fetal NT examination is normal, and fetal heart abnormality is diagnosed by prenatal ultrasound from 22 weeks to 25 weeks. There is no doubt about the importance of prenatal screening and eugenics. Therefore, it is suggested that pregnant women with high-risk factors should increase several prenatal ultrasound screenings as early as possible, and screen for some serious fatal malformations as soon as possible to reduce the psychological and physiological trauma of pregnant women.

Acknowledgments

We sincerely thank the obstetrics and Gynecology ultrasound, cardiothoracic surgery, and anesthesia team for their multidisciplinary care of this patient. We also thank our patients. Without them, this case report will not be possible.

Funding

Provincial Quality Engineering Project of colleges and universities in Anhui Province (2020xskt525); Provincial Quality Engineering Project of colleges and universities in Anhui Province (2020xsxxkc458); Scientific research fund project for introducing talents from Yijishan Hospital of Wannan Medical College (yr201511).

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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