RHYTHM
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Case Report

Echocardiographic Diagnosis and Treatment of ALCAPA Syndrome in a Pregnant Woman: A Case Report

Seyed Mohammad Hashem Montazeri[1], Somayyeh Norouzi[2*]

[1] Student Research Committee, School of Medicine, Qom University of Medical Sciences, Qom, Iran.
[2] Department of Internal Medical Sciences, Qom University of Medical Sciences, Qom, Iran.

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Highlights

- Echocardiography is a safe tool for diagnosis of ALCAPA syndrome in pregnant patients with diagnostic clues.

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* **Corresponding Author:**
Somayyeh Norouzi
Department of internal medical sciences, Beheshti Hospital, Beheshti Boulevard, Qom, Iran.
Tel:+98253719964797
Mobile:+989124338082
E-mail: snorouzimd@gmail.com

ABSTRACT

In this report, we present the case of a 21-year-old woman diagnosed with ALCAPA syndrome during pregnancy, based on echocardiographic findings. We also review the diagnostic process, pregnancy management, and treatment of the patient.

Keywords: Echocardiography; Coronary Vessel Anomalies; Syncope; Pregnancy

Introduction

Anomalous left coronary artery from the pulmonary artery (ALCAPA) syndrome is a rare congenital anomaly in which the left coronary artery arises from the pulmonary artery.[1]

It is typically an isolated defect, accounting for less than 0.5% of congenital heart anomalies. This anatomical abnormality likely originates during the fifth fetal week in the development of the aorta.[2] Prenatal blood flow in the left coronary artery remains tolerable until birth due to high pulmonary artery pressure. Nonetheless, after birth, as pulmonary artery pressure decreases, blood flow in the left coronary artery reverses, leading to inadequate perfusion of the left ventricular myocardium. Consequently, ischemia and death may occur.[3] Typically, 90% of patients die within the first few months of life. Still, in some cases, sufficient collateral artery formation allows survival. Despite this, myocardial perfusion remains inadequate, leaving patients at high risk for ventricular arrhythmias and sudden death.[4] Imaging modalities such as computed tomography (CT) angiography and magnetic resonance angiography can aid in diagnosis. Definitive treatment requires surgical intervention, which is the only curative option for surviving patients. [5,6]

This report presents the case of a young pregnant patient and details the diagnostic process, pregnancy management, and treatment.

Case Report

A 21-year-old primigravida (G1P0) woman presented to the hospital at 15 weeks of gestation with complaints of atypical chest pain and severe weakness. Her vital signs showed a heart rate of 98 bpm and blood pressure of 110/65 mm Hg.

The patient reported a long-standing history of shortness of breath and fatigue since childhood, for which no specific cause had been identified. She had never undergone a comprehensive cardiac evaluation. During the eighth week of pregnancy, she experienced an episode of hypotension and syncope at a physician's office, which was initially attributed to maternal stress. Her only medications were folic acid supplements and routine prenatal vitamins.

Due to her presenting chest pain, an ECG was obtained (Figure 1). Given her history of syncope, echocardiography was performed to evaluate for potential valvular insufficiency or structural heart defects (Figure 2).

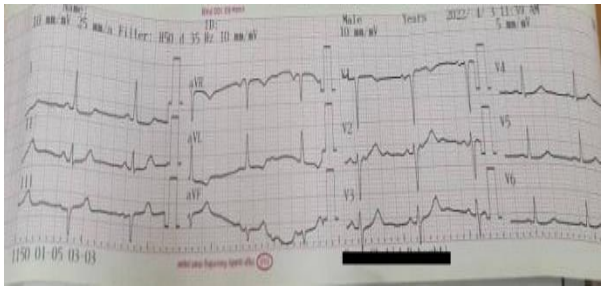

Figure 1. The image shows the first ECG of the patient.

The echocardiogram demonstrated retrograde blood flow from the coronary arteries into the pulmonary artery, along with extensive coronary collaterals throughout the ventricular septum and the lateral wall. Marked dilation of the right coronary artery (RCA) was evident. Color Doppler imaging revealed multiple coronary collaterals within the ventricular septum and the free ventricular wall.

A short-axis view at the aortic valve level showed significant RCA dilation. Further, parasternal short-axis views on color Doppler echocardiography clearly depicted the left main coronary artery originating from the main pulmonary artery, with retrograde flow signals within the pulmonary artery. These characteristic findings confirmed the diagnosis of ALCAPA syndrome (Figures 2–4).

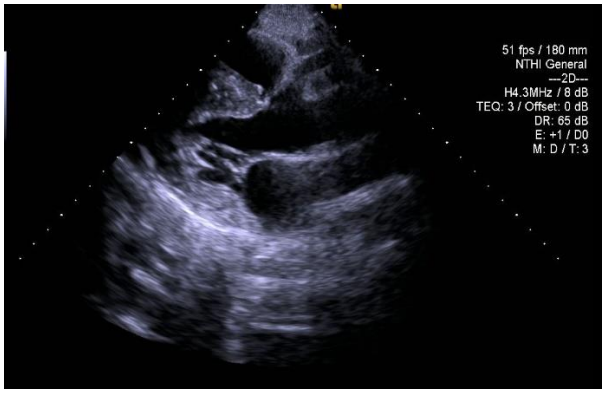

Figure 2. The 2D echocardiography, in the parasternal long-axis view, shows a dilated right coronary artery, which is a sign of anomalous left coronary artery from the pulmonary artery (ALCAPA) in echocardiography.

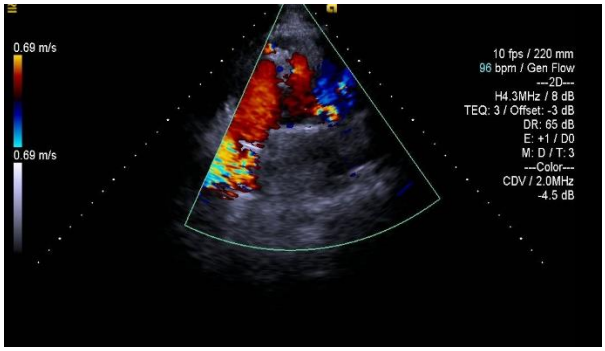

Figure 3. The 2D echocardiography with color flow mapping, in the parasternal short-axis view, demonstrates abnormal flow within the left coronary artery toward the main pulmonary artery.

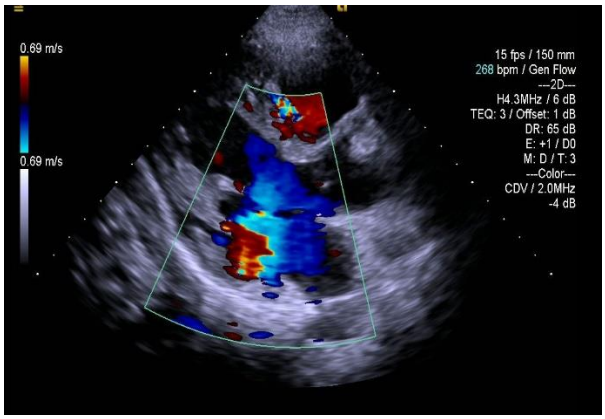

Figure 4. The parasternal short-axis view on 2D echocardiography with color flow mapping demonstrates coronary collateral vessels supplying myocardial perfusion.

The patient was initiated on medical therapy including isosorbide dinitrate (2.5 mg daily) and bisoprolol (2.5 mg daily). She was advised to maintain relative rest and minimize stress. The diagnosis of ALCAPA syndrome was confirmed, with plans for definitive diagnostic evaluation (CT angiography) and surgical intervention deferred until postpartum.

Throughout gestation, the patient underwent weekly monitoring by both cardiology and obstetrics teams. At 38 weeks' gestation, she developed acute hypertension prompting hospitalization for preeclampsia evaluation. An emergency cesarean delivery was performed, resulting in a healthy male infant weighing 3150 g with an Apgar score of 8.

As previously noted, definitive diagnostic evaluation (CT angiography) and surgical intervention were deferred until postpartum. Two weeks after delivery, the patient underwent CT angiography (Figure 5) and was subsequently referred for surgical correction.

The Takeuchi procedure was successfully performed for defect repair.[7] During one year of postoperative follow-up, the patient demonstrated complete resolution of symptoms, including exertional dyspnea and fatigue. A follow-up echocardiographic examination revealed normalized cardiac findings, and an exercise stress test (Bruce protocol) showed the absence of ST-T segment changes and achievement of 10 minutes' exercise duration without dyspnea or chest pain.

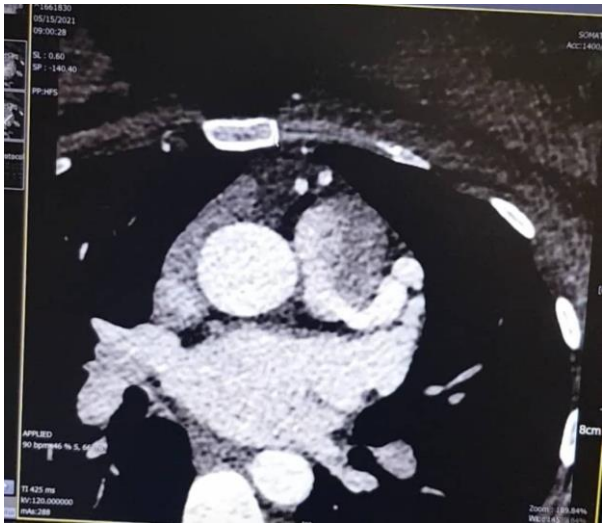

Figure 5. The postpartum computed tomography angiography demonstrates an anomalous origin of the left main coronary artery from the pulmonary artery (ALCAPA), confirming the diagnosis of ALCAPA syndrome.

Discussion

Our patient was a young woman whose condition was not diagnosed until her pregnancy. A subset of individuals with this syndrome is not diagnosed until adulthood, with a high percentage experiencing sudden cardiac death in young adulthood. Accordingly, advancements in imaging techniques are crucial for the early detection and treatment of this rare disorder, which can save lives.[8]

Nielsen et al.[9] reported several cases of ALCAPA syndrome. The patients they described, who ranged in age from 18 to 35 years, were of a similar age to our own. They experienced varying degrees of ischemia and cardiac symptoms following exercise. The diagnosis was made by angiography, not echocardiography. In one case, only left ventricular dilatation was observed.

The use of noninvasive modalities, such as echocardiography and Doppler ultrasound, to detect ALCAPA could accelerate the diagnostic process, particularly for patients with absolute or relative contraindications to angiography. This, however, depends on an accurate interpretation of the imaging findings. The limitations of these noninvasive tools, such as lower spatial resolution and operator dependence, cannot be ignored. Although our patient achieved a favorable outcome, the delayed diagnosis—resulting from the initial attribution of her syncope to stress and psychiatric causes—represented a critical clinical oversight. A prompt cardiac examination, potentially including Holter monitoring, should have been performed.

In describing an 11-year-old patient with ALCAPA syndrome, Lin et al.[10] detailed several echocardiographic manifestations of the disease. Echocardiography revealed inverted flow in the left coronary artery, a dilated left ventricle, mitral valve regurgitation, and an expanded right-to-left collateral vascular network.

Previous reports have indicated that pregnancy and childbirth are safe for patients who have undergone surgery for ALCAPA syndrome, with most deliveries being vaginal. Our case is, nevertheless, distinguished by the postponement of surgical treatment until after delivery.[11] In some instances, the diagnosis in pregnant women is made only after the patient experiences cardiac arrest.[12] Although surgical correction is the standard treatment strategy for these patients,[13] we were able to postpone the procedure in our patient using supportive therapy with beta-blockers and nitrates, which successfully prevented heart failure. Furthermore, detailed information on the exact etiology and risk factors for this disease is lacking; therefore, global data collection is recommended to facilitate future meta-analyses.

Conclusion

Echocardiography can facilitate the diagnosis of ALCAPA syndrome in pregnant patients through key findings such as numerous coronary collaterals, dilation of the RCA, and retrograde flow in the left main coronary artery originating from the main pulmonary artery. This case suggests that with appropriate medical management, corrective surgery can be safely postponed until after delivery. Nonetheless, further studies are needed to establish more precise guidelines for the safe management of pregnancy in patients with ALCAPA syndrome.

Declarations:
Ethical Approval

The patient gave her informed written consent and provided her medical records to the research team with the permission and under the supervision of the rules of the University Ethics Committee for the publication of the case report.

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Conflict of Interest

The authors report no conflict of interest.

Acknowledgment

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