Radiology Case Reports 20 (2025) 570–573

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case Report

Untreated ALCAPA diagnosed in gestational ultrasonography ☆

Lam Truong Hoai, MD[a,*], *Nguyen Van Thanh, MD*[b], *Nguyen Cong Thanh, MD*[b], *Nguyen Duc Hung, MD*[a], *Tran Duc Minh, MD*[a], *Nguyen Tuan Long, MD*[a]

[a] *Cardiovascular Department, Tam Anh Hospital, Hanoi, Viet Nam*
[b] *Vietnam National Heart Institute, Bach Mai Hospital, Hanoi, Viet Nam*

ARTICLE INFO

Article history:
Received 13 September 2024
Revised 3 October 2024
Accepted 4 October 2024

Keywords:
Anomalous coronary artery
High-risk pregnancy
Echocardiography
Congenital heart disease
ALCAPA

ABSTRACT

ALCAPA is a rare congenital anomaly that presents with left ventricular (LV) dysfunction and mitral valve regurgitation. The mortality rate is roughly 90% if the intracoronary collateral isn't significantly augmented. Malignant arrhythmias resulting in sudden death are common, affecting nearly 90% of patients with a mean age of 35 years. Especially during pregnancy, untreated ALCAPA can lead to high mortality and complications. Pregnant women with congenital heart disease have high risks for both themselves and their fetuses. Therefore, screening for congenital heart disease is very important in early diagnosis, counseling, and management. Most women born with congenital heart disease (CHD) will reach reproductive age.

We report a case of a 30-year-old woman in her second trimester of pregnancy who presents with ALCAPA, preserved ejection fraction (EF), and mild LV dilation. An uncommon feature, in this case, is the origin of the left main (LM) coronary artery from the posterior to the right of the main pulmonary artery (MPA), which is very close to the ascending aorta, mimicking the normal origin of the LM from the aortic root in 2D transthoracic echocardiography. The patient also has preserved left ventricular function with endocardial fibroelastosis. Echocardiography should be performed every 4 weeks to assess the progression of the disease during pregnancy.

Introduction

Echocardiography plays an important role in the detection of ALCAPA. Early diagnosis helps improve the prognosis of the patient through timely surgical intervention, aiming to restore the anatomy and physiology of the coronary system, preserve LV function, and avoid sudden cardiac death during exertion, especially in the gestation period. Critical echocardiographic findings include dilation of the right coronary artery, absence

☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

* Corresponding author.
E-mail address: truonglamcs@gmail.com (L.T. Hoai).

https://doi.org/10.1016/j.radcr.2024.10.031

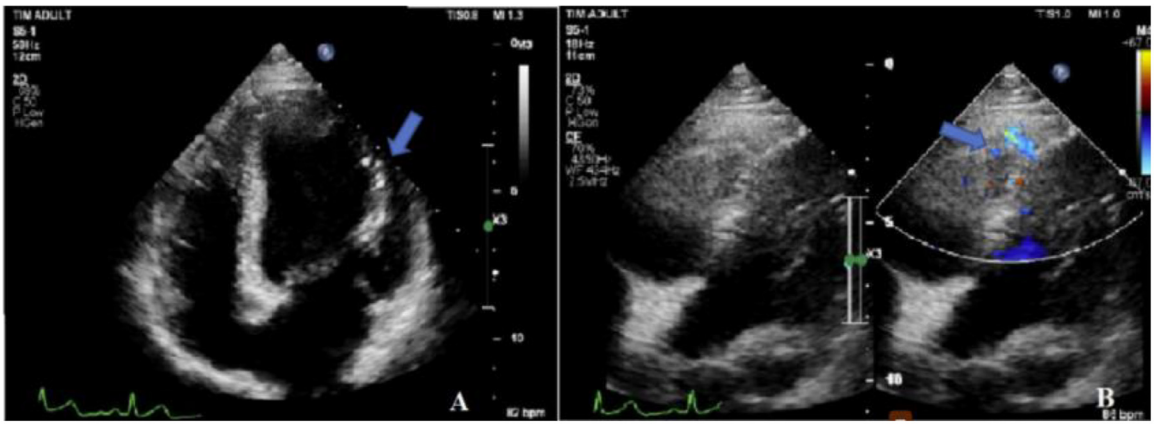

Fig. 1 – The echocardiography showed endocardial fibroelastosis in the 4-chamber view (4CV) (blue arrow) (A), and collaterals within the ventricular septum in the 5-chamber view (5CV) (blue arrow) (B).

of the left coronary artery (LCA) ostium in the aortic root, continuous color flow in the MPA above the pulmonary valve, and collaterals within the ventricular septum and free ventricular wall, along with left ventricular (LV) dysfunction and mitral valve regurgitation [1,2]. Multislice computed tomography can accurately identify the abnormal coronary artery origin but is harmful during gestation. Magnetic resonance imaging is also valuable for diagnosing ALCAPA. Various imaging modalities have been described, which help diagnose ALCAPA, but differences in echocardiographic features can lead to missed diagnoses [3].

The case report showed uncommon features, including significant dilation of the LCA and an uncommon origin of the LM from the posterior to the right of the MPA, which is very close to the ascending aorta, mimicking the normal origin of the LM from the aortic root in 2D transthoracic echocardiography. The patient also had preserved LV function with endocardial fibroelastosis. Hemodynamics change during pregnancy due to disturbances in heart structure, especially abnormal coronary vasculature. There are major increases in cardiac output and a decrease in maternal systemic vascular resistance; the renin-angiotensin-aldosterone system is significantly activated, and the heart and vasculature undergo remodeling [4], resulting in an increased risk of cardiac events and mortality.

Case Presentation

A 30-year-old woman in her second trimester (19 weeks) of pregnancy presents with chronic dyspnea on exertion. The patient has been experiencing worsening left-sided chest heaviness and was subsequently referred to our hospital. She has no medical history of coronary risk factors and no family history of premature coronary artery disease or congenital heart conditions. Physical examination was normal, and a heart murmur was not recorded. ECG showed sinus rhythm without ST or T wave changes. Obstetric ultrasound showed normal amniotic volume with a heart rate of 146 bpm, and positive fetal movement.

Echocardiography showed mild LV dilation with preserved EF, without mitral valve regurgitation, and endocardial fibroelastosis (Fig. 1A). Collaterals were observed within the ventricular septum and free ventricular wall (Fig. 1B). The origin of the LM was from the posterior to the right of the MPA, very close to the ascending aorta, mimicking the normal origin of the LM from the aortic root (Fig. 2A, B). There were no structural abnormalities of the aortic, mitral, tricuspid, or pulmonic valves, and there was very mild mitral regurgitation by Doppler. We will continue to follow the patient's pregnancy every 4 weeks to check echocardiography and obstetric Doppler.

Discussion

The adult patient presents with signs of chest discomfort and dyspnea on exertion due to increased flow from coronary steal, which occurs during stress. Because of the formation of collateral circulation, patients with ALCAPA can be asymptomatic in the early stages. However, as the disease progresses, cardiac structural changes such as mitral valve regurgitation occur. There are different proposed mechanisms, including left ventricular myocardial infarction, scarring, calcification of papillary muscles, and endocardial fibroelastosis, which result in varying degrees of fibrosis in myocardial tissue and increase the risk of fatal ventricular tachyarrhythmias. Physiological changes in the cardiovascular system during pregnancy may pose a risk for those with congenital heart disease who are not able to sufficiently adapt [5]. Hypertensive disorders are the most frequent cardiovascular disorders during pregnancy, occurring in 5%-10% of all pregnancies. Moreover, pregnancy is a hypercoagulable state associated with an increased risk of thromboembolism. This situation, combined with ALCAPA, results in an increased risk of pulmonary embolism and myocardial infarction during pregnancy. The left ventricular adaptation to pregnancy can be suboptimal in heart disease. Maternal cardiac dysfunction is related to impaired uteroplacental flow and suboptimal fetal outcomes. The risk estimation needs to be re-evaluated during each pregnancy visit, as the risk of complications may change over time.

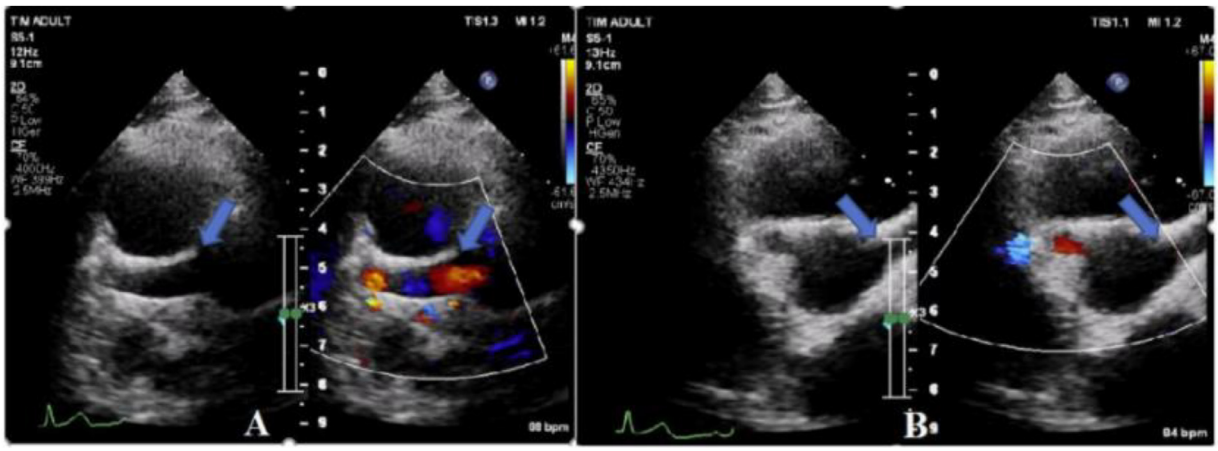

Fig. 2 – The echocardiography showed the LCA originating from the pulmonary artery (blue arrow) (A). The origin of the LM is from the posterior to the right of the MPA, very close to the ascending aorta, mimicking the normal origin of the LM from the aortic root. This cross-section appears to show the origin of the LCA, but it is a dropout artifact (blue arrow) (B).

Natriuretic peptide levels are associated with the occurrence of cardiac events, with N-terminal pro-B-type natriuretic peptide (NT-proBNP) levels greater than 128 pg/mL at 20 weeks of pregnancy being predictive of events later in the pregnancy [6]. Pre-eclampsia is associated with heart failure in women with heart disease. Transthoracic echocardiography is a method to diagnose ALCAPA, but in this case, there is an uncommon feature: dilation of both the coronary artery and the origin of the LM from the posterior to the right of the MPA, which is very close to the ascending aorta, mimicking the normal origin of the LM from the aortic root. Additionally, there is mild LV dilation, which can mimic a coronary fistula, but the right ventricle and pulmonary artery are not dilated.

In women with a moderate or high risk of complications during pregnancy, the minimum team requirements are a cardiologist, obstetrician, and anesthetist, all with expertise in managing high-risk pregnancies in women with heart disease. Additional experts may be involved depending on the individual situation. The patient should have an echocardiogram every 4 weeks to assess the progression of the disease, including left ventricular function and the mitral valve, and to develop a clear plan for labor and the early postpartum period [7]. The timing of delivery depends on the weighing of maternal, obstetric, and fetal risks. The ACOG recommends elective induction of labor for pregnant women with cardiac abnormalities between 39 and 40 weeks of gestation if they do not go into labor spontaneously [8]. SPECT has been shown to detect ischemia in adult patients with untreated ALCAPA. Preoperative demonstration of ischemia is required, given that the myocardium cannot recover from this lesion [9].

Conclusions

ALCAPA has a high mortality rate in adult patients. The uncommon feature of echo misdiagnosis results in delayed surgery. Direct transfer and reimplantation of the anomalous coronary artery into the aorta is the preferred surgical technique. Especially in pregnant women, increased cardiac events lead to higher mortality. Follow-up every 4 weeks with echocardiography is necessary to assess the progression of the disease.

Patient consent

The following information must be provided for this form to be processed accurately.

- I hereby give my consent for images or other clinical information relating to my case to be reported in a medical publication.
- I understand that my name and initials will not be published and that efforts will be made to conceal my identity, but that anonymity cannot be guaranteed.
- I understand that the material may be published in a journal, Web site or other form of publication. As a result, I understand that the material may be seen by the general public.
- I understand that the material may be included in medical books. A statement indicating: “Written informed consent was obtained from the patient to publish this report by the journal’s patient consent policy”.

Availability of data and materials

Data and materials used and/or analyzed during the current study are available from the corresponding author upon reasonable request.

Ethics approval

Our institution does not require ethical approval for reporting individual cases or case series.

REFERENCES

[1] Wesselhoeft H, Fawcett JS, Johnson AL. Anomalous origin of the left coronary artery from the pulmonary trunk. Its clinical spectrum, pathology, and pathophysiology, based on a review of 140 cases with seven further cases. Circulation 1968;38:403–25. doi:10.1161/01.cir.38.2.403.

[2] Blanco VM, Blalock SE, Ramaciotti C, et al. Oxygen supplementation is helpful for the echocardiographic detection of anomalous left coronary artery from the pulmonary artery. J Am Soc Echocardiogr 2010;23:1099–102. doi:10.1016/j.echo.2010.07.005.

[3] Jinmei Z, Yunfei L, Yue W, Yongjun Q. Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) diagnosed in children and adolescents. J Cardiothor Surg 2020;15:90. doi:10.1186/s13019-020-01116-z.

[4] Sanghavi M, Rutherford JD. Cardiovascular physiology of pregnancy. Circulation 2014;130:1003–8. doi:10.1161/circulationaha.114.009029.

[5] Cornette J, Ruys TP, Rossi A, et al. Hemodynamic adaptation to pregnancy in women with structural heart disease. Int J Cardiol 2013;168:825–31. doi:10.1016/j.ijcard.2012.10.005.

[6] Stout K. Pregnancy in women with congenital heart disease: the importance of evaluation and counselling. Heart 2005;91:713–14. doi:10.1136/hrt.2004.047886.

[7] Weinberg CR, Ahmad A, Li B, Halpern DG. Pregnancy in women with congenital heart disease: a guide for the general cardiologist. US Cardiol Rev 2020;14:e10. doi:10.15420/usc.2020.08.

[8] Mehta LS, Warnes CA, Bradley E, et al. Cardiovascular Considerations in caring for pregnant patients: a scientific statement from the American heart association. Circulation. 2020;141:e884–903. doi:10.1161/cir.0000000000000772.

[9] Crean AM, Ahmed F, Motwani M. The role of radionuclide imaging in congenital heart disease. Curr Cardiovasc Imag Rep 2017;10:38. doi:10.1007/s12410-017-9434-0.