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Case Report

Cor Triatriatum Dextrum in a Pregnant Woman Combined With an Atrial Septal Defect and Severe Pulmonary Hypertension: A Rare Case Report

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ABSTRACT

Case Presentation

Cor triatriatum, first described by Church in 1868, is an extremely rare congenital cardiac defect and accounts for between 0.1% and 0.4% of all congenital cardiac anomalies. Isolated cor triatriatum is rare and is usually associated with other cardiac anomalies, most commonly the atrial septal defect (ASD). Although the clinical presentations of cor triatriatum depend on the size of the fenestration, almost all cases are diagnosed in childhood and very few cases remain asymptomatic until adulthood. Several techniques such as transthoracic echocardiography, transesophageal echocardiography, computed tomography, and magnetic resonance imaging are used for the diagnosis of cor triatriatum; nevertheless, the definitive diagnosis and the identification of the associated anomalies can be easily made by echocardiography. The use of computed tomography is associated with the risk of radiation, and transesophageal echocardiography has the discomfort of scope intubation. [5]

We herein describe a 35-year-old pregnant woman with no previously described cardiac disorder who was diagnosed with cor triatriatum dextrum presenting with shortness of breath, tachycardia, and palpitation of 1 week's duration. She had a history of 5 uncomplicated vaginal deliveries. ***(Iranian Heart Journal 2020; 21(4): 120-123)***

KEYWORDS: Cor triatriatum, Congenital cardiac anomalies, Atrial septal defect, Pregnancy, Echocardiography

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Cor triatriatum, first described by Church in 1868, is an extremely rare congenital cardiac defect and accounts for 0.1% to 0.4% of all congenital cardiac anomalies. It is characterized by the presence of a membranous structure that divides the right atrium (cor triatriatum dextrum) or the left atrium (cor triatriatum sinistrum) into 2 chambers: a chamber with the pulmonary veins and the other with the mitral and atrial valves. The 2 chambers communicate with 1 or more openings in the

intra-atrial membrane. [1,2] Isolated cor triatriatum is rare; it is usually associated with other cardiac anomalies, most commonly the atrial septal defect (ASD). [3] Herein we describe a pregnant woman with no known cor triatriatum who presented with shortness of breath, palpitation, and hemoptysis of 1week's duration.

Case Report

A 35-year-old pregnant woman (gravida 5 para 6) who was at 8 weeks of gestation visited the emergency department of our institution with the complaint of shortness of breath, palpitation, and hemoptysis of 1 week's duration. The patient had no previous history for cardiac disease and claimed that she had no cardiac symptoms such as dyspnea, orthopnea, and chest pains in her previous pregnancies. On physical examination, she had a blood pressure of 120/70 mm Hg, a pulse rate of 130 bpm, a respiratory rate of 20 breaths per minute, and a temperature of 36.6 °C. The cardiovascular examination revealed normal heart sounds and no murmurs. The chest auscultation was normal. However, she had pedal edema. Her laboratory investigations were not remarkable. The electrocardiogram showed sinus tachycardia (Fig. 1). Transthoracic echocardiography detected cor triatriatum dextrum with ASD and pulmonary hypertension (Fig. 2). The fetal ultrasound examination revealed a healthy fetus with a heart rate compatible with the gestational age. The patient was admitted to the cardiology department, where she received medical therapy such as nasal oxygen, furosemide, digoxin, and enoxaparin. No surgical therapy was offered at the time. On day 4, her vital signs were normal, and there were no signs of heart failure, including no jugular venous distension, hepatomegaly, or peripheral edema. Additionally, echocardiography found a normal pulmonary arterial pressure and no heart failure. The patient had no uterine contraction, and her heart rate was 150 bpm. She was discharged home on post-admission day 6 with oral medications and was recommended to do monthly follow-ups by her both obstetrician and cardiologist. She continued her pregnancy. A healthy infant was delivered at term through normal vaginal delivery under noninvasive monitoring. Six months after delivery, we performed echocardiography and found cor triatriatum dextrum and primum ASD with a normal ejection fraction and no pulmonary hypertension.

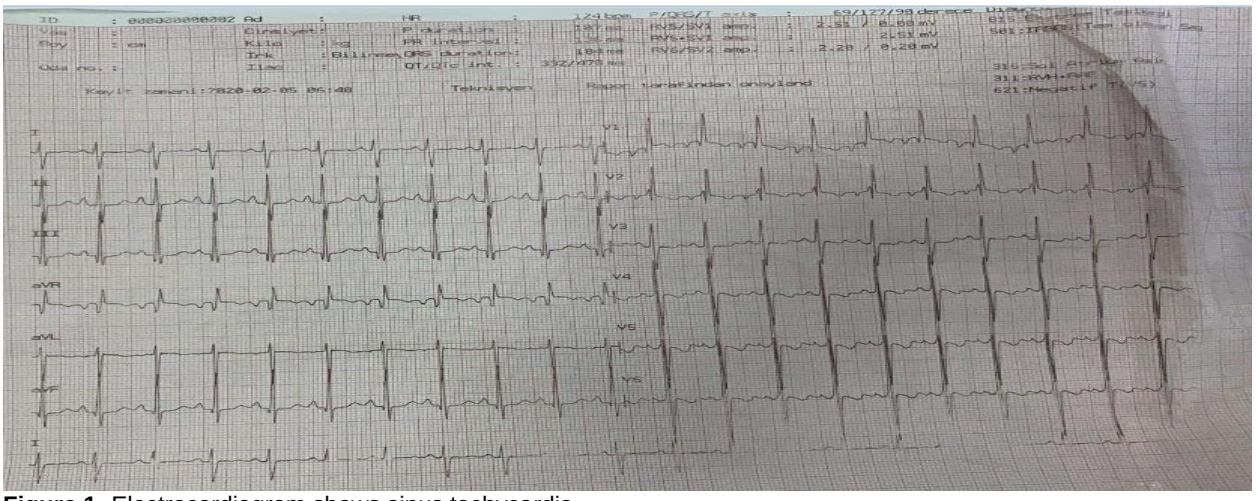

Figure 1: Electrocardiogram shows sinus tachycardia.

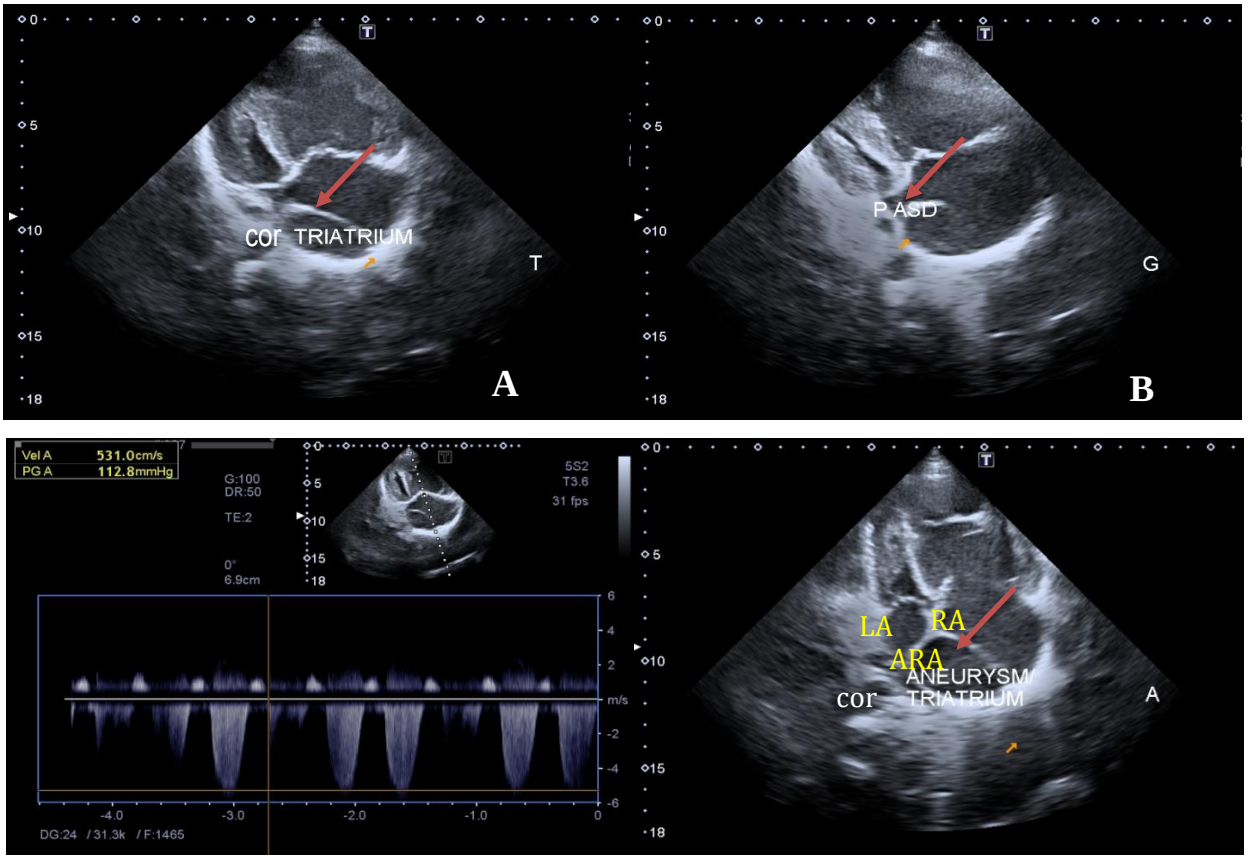

Figure 2: Transthoracic echocardiogram of the right atrial view shows a membrane (red arrow) dividing it into proximal and distal chambers. Panel A reveals cor triatriatum dextrum, Panel B demonstrates a primum ASD, Panel C indicates unusual pulmonary hypertension, and Panel D shows a fenestrated membrane (the red arrow) and a small atrial aneurysm.
LA, Left atrium; RA, Right atrium; ARA, Accessory right atrium; ASD, Atrial septal defect

DISCUSSION

Cor triatriatum, first described by Church in 1868, is an extremely rare congenital cardiac defect. It comprises 0.1% to 0.4% of all congenital cardiac anomalies and is characterized by the presence of a membranous structure that divides the right atrium (cor triatriatum dextrum) or the left atrium (cor triatriatum sinistrum) into 2 chambers: a chamber with the pulmonary veins and the other with the mitral and atrial valves. The 2 chambers communicate with 1 or more openings in the intra-atrial membrane. [1, 2]

According to the Loeffler classification in 1949, cor triatriatum is classified into 3 types based on the number and size of fenestrations in the fibromuscular membrane: Type I: no opening in the accessory membrane, Type II: 1 or a few small openings in the intra-atrial membrane (restrictive opening or fenestration), and Type III: large, single opening in the membrane through which the accessory chamber communicates with the true atrium. Similar to our patient, patients with Type III cor triatriatum either have minor symptoms or are asymptomatic and can survive into adulthood. [4]

Bai et al [5] reported that the most common presenting symptoms in pregnant women with cor triatriatum were dyspnea, hemoptysis, orthopnea as a result of the obstructive function of the intra-atrial membrane, and increased physiological

demand. We herein described a pregnant woman presenting with shortness of breath, tachycardia, and palpitation of 1 week's duration. Our case had a history of 5 uncomplicated vaginal deliveries.

Several techniques such as transthoracic echocardiography, transesophageal echocardiography, computed tomography, and magnetic resonance imaging are utilized for the diagnosis of cor triatriatum; nonetheless, the definitive diagnosis and the identification of the associated anomalies can be easily made by echocardiography. The use of computed tomography is associated with the risk of radiation, and transesophageal echocardiography poses the discomfort of scope intubation. [5] Our transthoracic echocardiography of the patient revealed a membrane dividing the right atrium into 2 chambers with primum ASD and pulmonary hypertension.

In patients with symptomatic obstructive cor triatriatum, the surgical resection of the accessory membrane is necessary; the management, however, depends on the degree of the obstruction between the right atrial chambers. [6]

Although our patient had symptomatic cor triatriatum with primum ASD and pulmonary hypertension, she refused to terminate her pregnancy. She ultimately gave birth to a healthy baby at term through normal vaginal delivery.

Conflict of Interest: None

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