

Incidental Finding of Incomplete Cor Triatriatum Dextrum During 2nd Trimester of Pregnancy

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Abstract

Background:

Cor triatriatum sinistrum is a type of congenital heart defect that occurs due to the left atrium being divided into two chambers by a fibromuscular septum. An even rarer subsection of this is cor triatriatum dextrum, in which the fibromuscular septum fully divides the right atrium.

Case presentation:

We present a patient with intermittent palpitations, chest pressure, and lightheadedness without syncope related to her cor triatriatum dextrum exacerbated by her pregnancy status. Without any prior history of congenital heart defects or pertinent family history, this seemingly healthy female prior to her pregnancy likely revealed and exacerbated her symptoms and lead to the diagnosis.

Conclusions:

Treatment in the setting of an asymptomatic presentation of a pregnant patient with history of cor triatriatum remains unclear. Many physicians elect close follow up to monitor for development of symptoms. In symptomatic cases, rate control, thromboembolic prophylaxis, and hemodynamic stabilization are mainstays of treatment. This unique presentation in a rare patient population furthers literature and gives a perspective on pregnancy and structural heart disease like cor triatriatum.

Keywords: Cor triatriatum dextrum, structure heart disease, pregnancy, echocardiography

1. Introduction

Cor triatriatum sinistrum is a type of congenital heart defect that occurs due to the left atrium being divided into two chambers by a fibromuscular septum. It is seen in 0.1% of all congenital heart disease (1). An even rarer subsection of this is cor triatriatum dextrum, in which the fibromuscular septum divides the right atrium. This occurs during cardiogenesis, when the sinus venosus is closed by the left and right venous valves. The right venous valve involutes during development, and an interruption of this process is thought to be the cause of this condition (2). Typical presentation is symptoms shortly after birth, which makes an incidental diagnosis in adulthood extremely rare.

Cor triatriatum dextrum is an exceedingly rare congenital abnormality, and it is a result of the failure of a regression process, specifically of the right sinus venosus valve (3). This malformation results in a Chiari network – filamentous, weblike structure from remnant tissue. This can result in right ventricular inflow tract obstruction (3).

2. Case Presentation

A 23-year-old female presented to the cardiologist for intermittent palpitations, chest pressure, and lightheadedness without syncope. At time of her presentation, she was 30 weeks pregnant with no prior pregnancy history. She had experienced this throughout her life, but she becomes increasingly concerned due to the increase in occurrences during her pregnancy. She had a past medical history of mild intermittent asthma and anxiety disorder. Her cardiac symptoms have no correlation with caffeine intake nor activity and endorsed sudden onset and resolution during episodes. There was no family history of Wolff-Parkinson-White (WPW) syndrome nor supraventricular tachycardia (SVT). Review of symptoms revealed no further information that has not been expanded upon above. Chart review from prior laboratory results show nothing abnormal on her complete blood count nor her comprehensive metabolic panel.

Differential diagnosis included anxiety, supraventricular tachycardia, paroxysmal/persistent atrial fibrillation, pulmonary hypertension, tricuspid stenosis, atrial myxoma, mitral stenosis, along with our final diagnosis of a

structural cardiac abnormality which workup revealed cor triatriatum dextrum.

An electrocardiogram (ECG) (Figure 1) was performed in the outpatient setting which revealed ectopic atrial activity shown by different inflections of the p-wave in multiple leads. There was no evidence of preexcitation, and QT intervals were within normal limits. At that time of the visit, an event monitor and a transthoracic echocardiogram were ordered.



Figure 1. Electrocardiogram revealing normal sinus rhythm with ectopia atrial activity.

The transthoracic echocardiogram (TTE) (Figure 2) was performed and revealed incomplete cor triatriatum dextrum. The right atrium was normal in size, and the anomaly did not seem to have a hemodynamic or anatomic effect. Her ejection fraction was estimated to be 65% and no other abnormalities were noted.

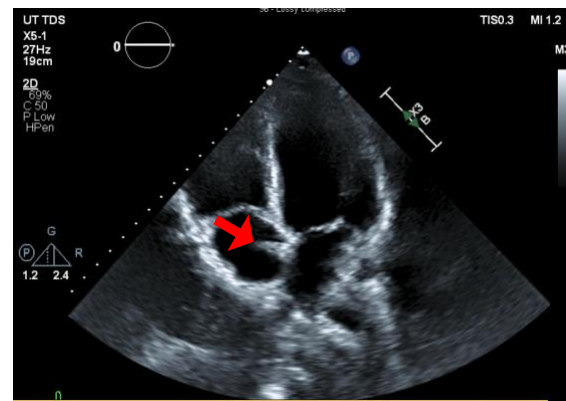


Figure 2. Transthoracic echocardiogram revealing the fibromuscular membrane of the incomplete cor triatriatum dextrum.

Her event recorder study was ten days long and revealed a range in heart rate from 80-125 bpm and only one single premature atrial contraction during her symptomatic events. No other abnormalities were noted.

The broad spectrum for our differential diagnosis led us to order different labs and tests which included the electrocardiogram, echocardiograph, complete blood count, comprehensive metabolic panel, and a ten-day event recorder. Effectively, this allowed us to rule out a majority of our original differential diagnosis and leave us with cor triatriatum dextrum.

Supportive care for her pregnancy was continued along with close follow up. Namely, care involved limiting strenuous physical exertion and monitoring blood pressure and symptoms. No other treatment post-partum was required as patient's symptoms and their severity subsided. As the patient progresses in age and if symptomatology develops, the patient may require a corrective cardiac surgery in the future.

The patient successfully delivered her child without complications. She continues to follow up cardiology outpatient, and her symptoms have continued to decrease in occurrences since her delivery.

3. Discussion

Cor triatriatum dextrum is an exceedingly rare congenital abnormality. The failure of this regression process of the right sinus venosus valve (3). This malformation results in a Chiari network. This can result in right ventricular inflow tract obstruction (3).

Similar cases published with cor triatriatum dextrum had symptoms that prompted further work up. One such example would be a patient who had recurrent strokes secondary to cor triatriatum dextrum with concomitant patent foramen ovale (4). Another example is of a patient who had persistent cyanosis and polycythemia which prompted further work up and revealed cor triatriatum dextrum (3).

The mainstay of diagnosis includes electrocardiography, echocardiogram, and left and right heart catheterization. Electrocardiography has multiple nonspecific findings which include different morphology in p-waves, right ventricular hypertrophy, and right axis deviation. Echocardiogram allows for definitive diagnosis and allows for differentiation between valvular (i.e. mitral stenosis) and structural causes (i.e. cor triatriatum dextrum) that cause similar symptomatology. While a transesophageal echocardiogram is preferred for precision, transthoracic echocardiogram is still a viable option. Left and right heart catheterization allows

for severity of obstruction and surgical correction options (5).

Treatment is dependent on severity of symptoms. In the patient case presented, the return to asymptomatic status allowed for the choice of close follow up. For symptomatic patients, treatment options involve both medical and surgical options. These options include rate and/or rhythm control if concomitant atrial fibrillation is present, thromboembolic prophylaxis if history of deep vein thrombosis or clotting disorders, and hemodynamic stabilization if applicable with antihypertensives. These all may require some combination of medicinal interventions depending on patient severity, comorbidities and rate of adherence. If these are persistent or severe enough, surgical consultation is recommended. This may involve complete surgical resection of atrial appendage or accessory membrane under cardiopulmonary bypass. The 10-year survival rate is estimated to be 83% (6,7).

This case illustrates why listening to the patient's symptoms and catching subtle cue on a test can result in a large finding. The exacerbating factor thought to have attributed to her symptoms was the increase in fluid homeostasis that accompanies pregnancy. The patient's symptoms did decrease after delivery, which is likely consistent with our hypothesis. This allowed the patient to receive vital information about her exceedingly rare abnormality that will serve valuable in the future.

List of Abbreviations:

Transesophageal Echocardiogram – TEE
Supraventricular Tachycardia – SVT
Electrocardiogram – ECG
Wolff-Parkinson-White Syndrome – WPW Syndrome

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