Cor Triatriatum Dexter: Unveiling a Rare Congenital Cardiac Anomaly

Cor Triatriatum Dexter : Révélation d'une Rare Anomalie Cardiaque Congénitale

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Abstract

Cor triatriatum dexter is a rare congenital anomaly caused by the persistence of the right valve of the sinus venosus, leading to the formation of an additional chamber in the right atrium. We present a case of a 50-year-old woman with a history of congenital heart disease, who developed acute right heart failure. Echocardiography revealed a membranous structure dividing the right atrium. Early recognition is essential for proper management, ranging from conservative monitoring to surgical intervention in symptomatic cases.

Keywords

Cor triatriatum dexter, congenital heart disease, echocardiography

Résumé

Le cor triatriatum dexter est une anomalie congénitale rare due à la persistance de la valve droite du sinus veineux, entraînant la formation d'une chambre supplémentaire dans l'oreillette droite. Nous rapportons le cas d'une patiente de 50 ans, ayant des antécédents de cardiopathie congénitale, qui a développé une insuffisance cardiaque droite. L'échocardiographie a révélé une membrane divisant l'oreillette droite. Un diagnostic précoce est essentiel pour une prise en charge adaptée, allant de la surveillance à l'intervention chirurgicale selon la symptomatologie.

Mots-clés

Cor triatriatum dexter, cardiopathie congénitale, échocardiographie

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INTRODUCTION

The incidence of cor triatriatum is approximately 0.1% of congenital heart malformations (1). Cor triatriatum dexter, or partitioning of the right atrium (RA) to form a triatrial heart, is an extremely rare congenital anomaly caused by the persistence of the right valve of the sinus venosus (2).

CASE PRESENTATION

A 50-year-old woman with a cardiac history of ventricular septal defect and pulmonary stenosis, diagnosed and treated surgically at the age of 32, presented for acute right heart failure. Since her surgery, the patient had not sought medical care until she experienced shortness of breath due to abdominal distension caused by ascites. She reported paroxysmal palpitations and dyspnea over the previous months.

She had two children with no complications during pregnancy or delivery. The patient was hypotensive (90/50 mmHg), with irregular cardiac sounds and a systolic murmur in the tricuspid region, but no signs of left heart failure. Examination revealed hepatomegaly, jugular venous distension with jugular venous pulsations, ascites, and no peripheral edema.

Electrocardiogram showed atrial fibrillation (AF) with diffuse microvoltage. Transthoracic echocardiography revealed normal left ventricular ejection fraction (60%), an enlarged left and right atrium, no mitral regurgitation, or atrial septal defect. The right atrium was divided into two chambers by an echogenic linear structure suggestive of a membrane (Figure 1). The patient had right ventricular dysfunction and torrential tricuspid regurgitation.

Pulmonary artery systolic pressure was measured at 80 mmHg through pulmonary regurgitation. The pressure gradient across the membrane fenestration was estimated at 40 mmHg. Both the superior and inferior vena cava drained into the proximal chamber of the right atrium. Transesophageal echocardiography was not performed due to patient intolerance, and MRI was impossible because of the arrhythmia.

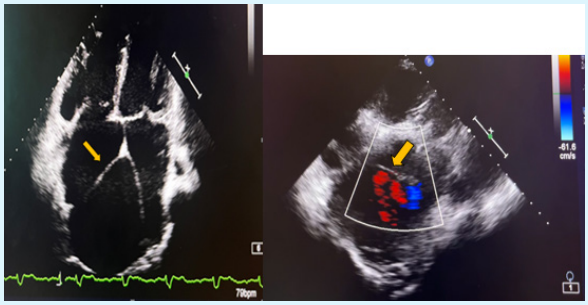

Figure 1. Right atrium divided into two chambers by an echogenic linear structure (yellow arrow)

DISCUSSION

The persistence of the right valve of the sinus venosus during embryonic development leads to various anomalies, including membranous remnants of the crista terminalis, the Chiari network, and cor triatriatum dexter. In this condition, the entire right valve remains, dividing the sinus venosus from the primitive atrium within the right atrium. Echocardiographically, the membrane runs from the inferior vena cava to the superior vena cava. Since the membrane is usually perforated, venous flow from the upstream chamber crosses the membrane into the downstream chamber (1-3).

This rare congenital anomaly is often associated with significant malformations of other right heart structures, most commonly right heart chamber hypoplasia. Historically, cor triatriatum dexter was first discovered in the 19th century and was diagnosed primarily post-mortem. However, with the advancement of medical imaging, particularly echocardiography, the condition can now be identified non-invasively during a patient's lifetime.

Clinically, the presentation of cor triatriatum dexter varies depending on the degree of atrial partitioning. Mild cases may remain asymptomatic and are often discovered incidentally during imaging studies or surgeries for other conditions. More severe cases can lead to symptoms such as right-sided heart failure, elevated central venous pressures, or cyanosis, particularly when associated with other congenital heart defects.

Transesophageal echocardiography remains the preferred method for diagnosing cor triatriatum dexter, offering superior imaging of posterior cardiac structures and defining embryonic remnants with greater precision. Incorporating three-dimensional reconstruction has proven beneficial in visualizing the anatomical features dividing the right atrium (3-5).

Due to the rarity of the diagnosis, no standardized guidelines exist regarding the optimal modality and timing for treatment. Surgical treatment is typically recommended for symptomatic patients with obstructive fenestration, with membrane resection being the primary approach. In patients who are unfit for surgery, particularly those with acute heart failure, balloon catheter dilatation of the fenestration can serve as a bridge to surgical intervention.

CONCLUSION

Cor triatriatum dexter is a rare congenital heart anomaly resulting from the incomplete regression of the right valve of the sinus venosus, leading to the formation of an additional chamber in the right atrium. While many cases remain asymptomatic and are incidentally detected, more severe presentations can mimic right heart obstruction, resulting in symptoms such as cyanosis, heart failure, or venous congestion. Early diagnosis through echocardiography and other imaging techniques is crucial for proper management. Treatment strategies range from conservative monitoring in mild cases to surgical or interventional correction in symptomatic patients. Awareness of this condition is vital for clinicians to differentiate it from other right heart pathologies, ensuring optimal patient care.

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