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CorTriatriatum: A Case Report

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Abstract

A 40-year-old female patient with history of palpitation underwent transthoracic echocardiography that revealed an extra septum that subdivided the left atrium in to proximal and distal chambers. The diagnosis of cortriatriatum sinister(CTS) was confirmed by transesophageal echocardiography. The communication between proximal and distal chambers was provided by large fenestrations in the fibromuscular membrane. CTS is rarely presents in adults and is primarily diagnosed with echocardiography. Treatment options depend on size of the fenestrations in the membrane and in some instances surgical resection of membrane may be necessary.

Keywords

CorTriatriatum, Echocardiography, Left Atrium

Introduction

Cortriatriatum is a congenital anomaly that was first reported by Church in 1868[1].CTS is a heart with three atria (triatrial heart), in which the left atrium (cortriatriatum sinister) or right atrium (cortriatriatumdexter) is divided into two compartments by a fold of tissue, a membrane, or a fibromuscular band[2-12].

In this anomaly veins drain into the upper portion of the corresponding atrium, whereas the atrial appendage and the true atrial septum are components of the inferior portion. The inferior or distal part terminates to atrioventricular valve[13]. CTS is generally diagnosed in pediatric population, with age of presentation depending on the size of membrane fenestration and the resulting obstruction to inflow[14]. There is significant variability in size and shape of membrane and size and number of fenestrations. Echocardiography can diagnose most cases of CTS, measure a gradient between the proximal and distal chambers, and estimate pulmonary artery pressure using Doppler[15].

Case Presentation

A 40-year-old female patient was referred to our hospital due to palpitation.Physical examination including cardiovascular exam revealed nothing unusual and no cardiac murmur. An electrocardiogram was normal andcardiothoracic ratio was normal in chest radiography. A transthoracic echocardiogram showed an extra septum that subdivided the left atrium. (Figures1-2)

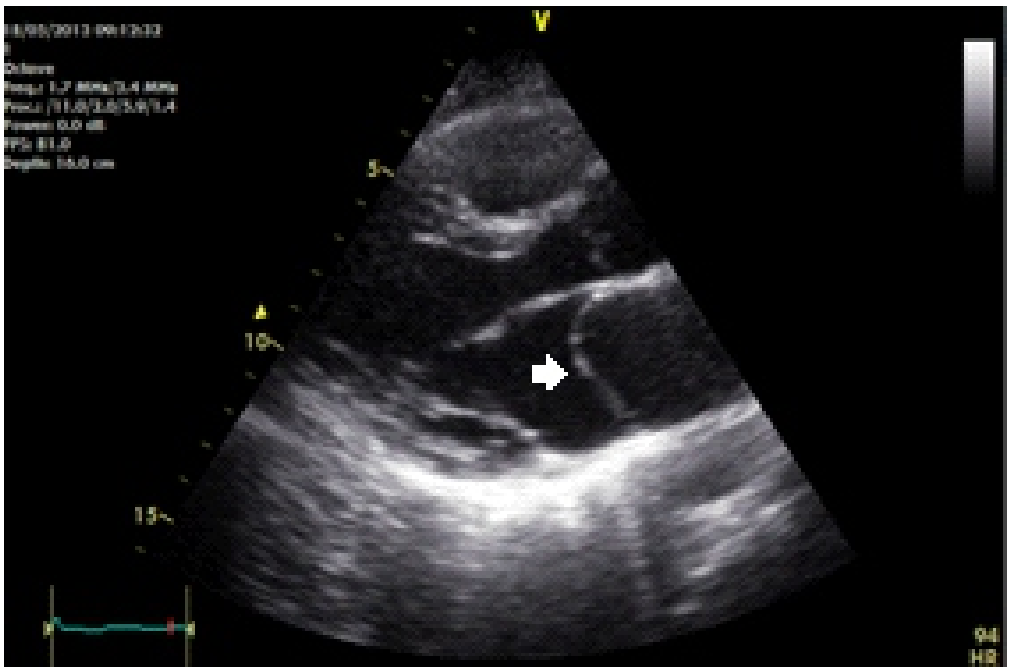

Figure 1: Transthoracic echocardiography (parasternallong axis view) shows discrete membrane (arrow) in left atrium

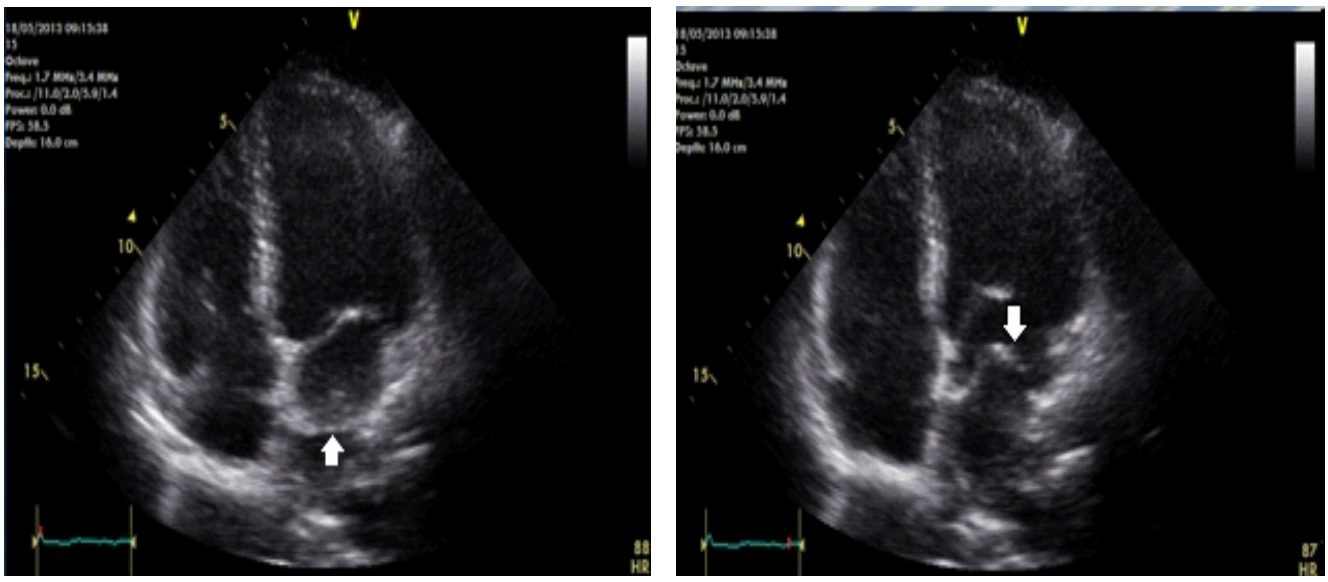

Figure 2: Transthoracic echocardiography (apical four chamber view) ofthe same patient during systole(left) and diastole (right).

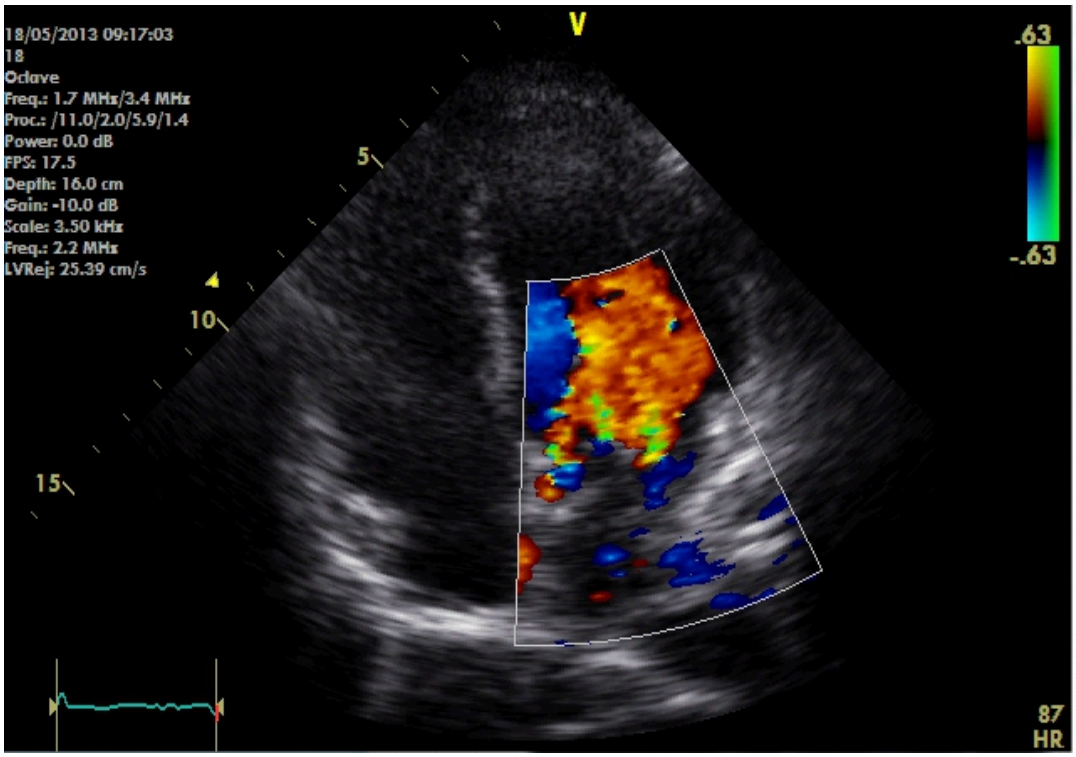

Figure 3: Color- flow Doppler transthoracic echocardiography(apical four chamber view) showing fenestrations in the membrane

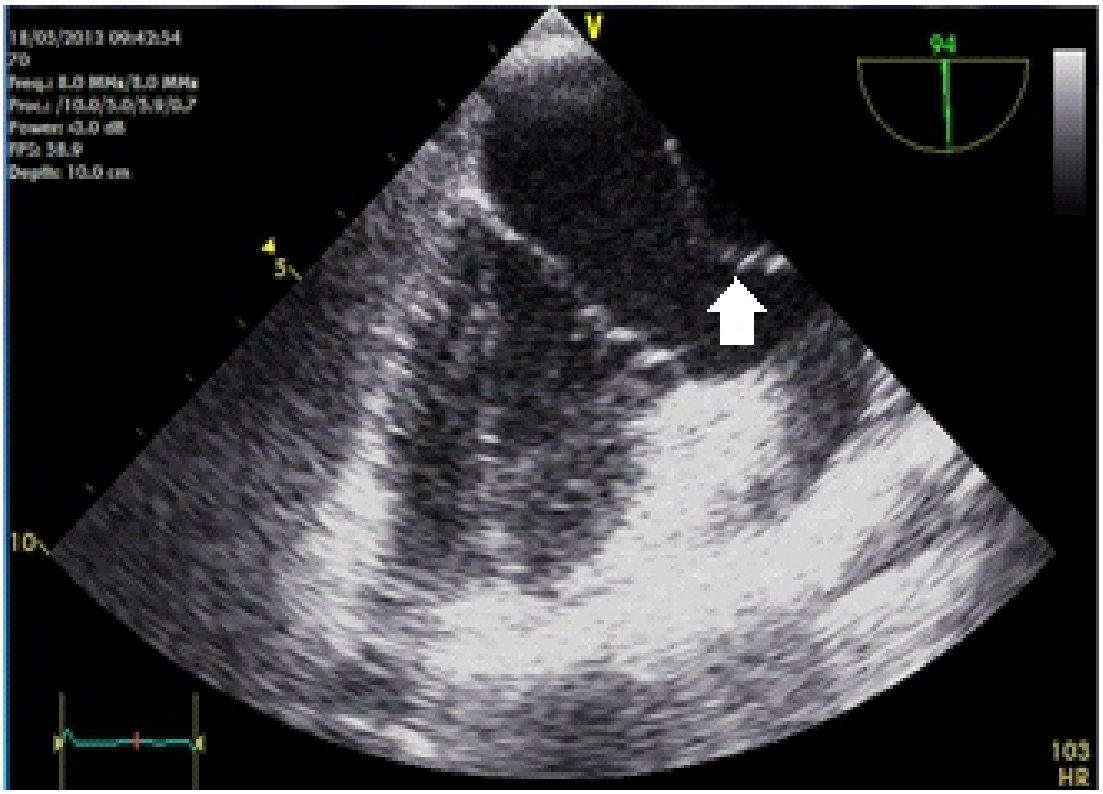

Figure 4: Transesophageal echocardiography (two chamber view) of discrete membrane (arrow) in left atrium

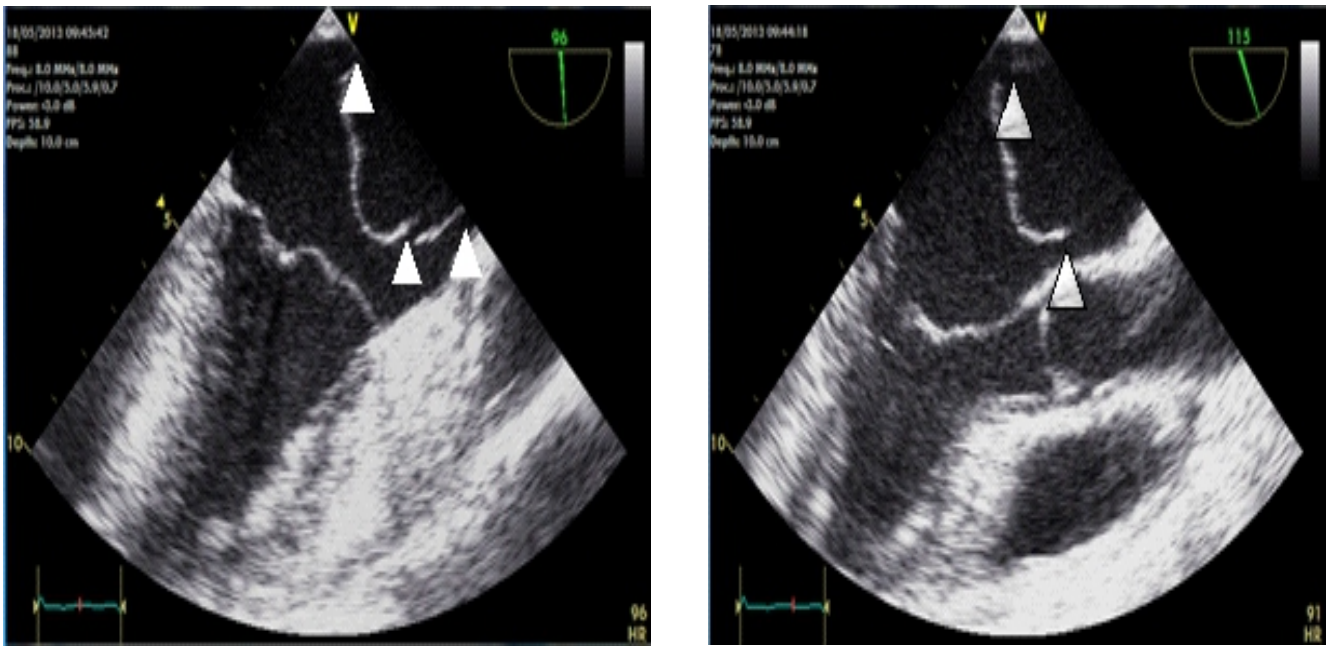

Figure 5: Transesophageal echocardiography (two chamber and long axis views) demonstrating fenestrations or openings in the membrane (arrowheads)

Subsequently, transesophagealechocardiography(TEE) confirmed the diagnosis of cortriatriatum(Figures 4-5). In TEE left atrium was divided by an abnormal fibromuscular diaphragm into a postero superior chamber receiving the pulmonary veins and an antero inferior chamber giving rise to the left atrial appendage and leading to the mitral orifice. The communication between the divided atrial chambers consisted of at least three openings or fenestrations in the diaphragm with mild obstruction to the pulmonary venous return (peak pressure gradient=7 mm Hg, mean pressure gradient=4 mm Hg).

Discussion

Cortriatriatum is a rare congenital heart disease (CHD), 0.1% of all congenital cardiac defects but a higher incidence, up to 0.4% has been reported in autopsies of patients with CHD[9]. There are two types of this anomaly: left and right. Cortriatriatum sinister is more common that dexter. Cortriatriatum dextrum is extremely rare. Fewer than 300 cases of cortriatriatum have been reported. It's a surgically correctable CHD and can occur as an isolated defect (classic) or in association with other congenital cardiac anomalies (atypical)[8]. The increase in reports of cortriatriatum in recent years most likely results from the widespread use of advanced cardiovascular imaging, particularly echocardiography.[9]

In this malformation, failure of resorption of the common pulmonary vein results in a left atrium divided by an abnormal fibromuscular diaphragm into a postero superior and antero inferior chamber.

Morphology: The communication between the divided atrial chambers may be large, small, or absent. The size of the opening or openings in the diaphragm determines the degree of obstruction to pulmonary venous return. Elevations in both pulmonary venous pressure and pulmonary vascular resistance may result in severe pulmonary artery hypertension[13].With respect to the presence of large defects of diaphragm in our patient, elevation in pulmonary artery pressure was mild (35 mm Hg).

Clinical Features: Communication between the pulmonary veins and anterior chamber is provided by fenestrations on the membrane. As a result of atrial flow obstruction, symptoms of congestive heart failure develop during infancy and childhood[11] . The majority of reported cases of CTS occur in infants with symptoms of pulmonary venous obstruction; with adult cases being rare[12].Cortriatriatum may be detected as an incidental finding in a patient in whom an echocardiogram is obtained for another reason. In general, these cases represent the unobstructed form that requires no early intervention. Patients with more severe obstruction have similar findings as those with congenital pulmonary vein stenosis[13]. Fibrosis and calcification of the orifice in the accessory membrane or development of mitral regurgitation and/or atrial fibrillation may cause symptoms (exertional dyspnea, exercise intolerance and easy fatigability, hemoptysis, rthopnea, systemic thrombo embolism and palpitation) in an asymptomatic patient. Our patient had several openings in the left atrial membrane without a significant gradient on Doppler study which probably accounted for the late diagnosis.

Laboratory Investigations

Electrocardiography: In unobstructed cases, findings on electrocardiography are normal, whereas patients with significant obstruction have right ventricular hypertrophy because of the associated pulmonary hypertension[13]. Our patient had normal ECG.

Chest Radiography: The findings may be normal in those with mild obstruction or demonstrate pulmonary edema with significant obstruction[13].Chest radiography was normal in our patient.

Echocardiography: The diagnosis is established by two dimensional echocardiography or TEE, with further insight obtained from three-dimensional reconstruction. The obstructive diaphragm is visualized on parasternal long- and short-axis and four-chamber views and can be distinguished from a supravalvular mitral ring by its position superior to the left atrial appendage, which forms part of the distal chamber. Also present is diastolic fluttering of the mitral leaflets and high-velocity flow detected by Doppler examination in the distal atrial chamber and at the mitral orifice[13]. Supravalvar mitral ring is a circumferential ridge or membrane arising from the left atrial wall overlying the mitral valve and frequently attached to the mitral valve. Variable in thickness and extent, the ring ranges from a thin

membrane to a thick discrete fibrous ridge. The membranous variety may be difficult to detect because the membrane often adheres to the anterior mitral valve leaflet while remaining just proximal to the posterior mitral leaflet. Adhesion to the valve may impair the opening of the leaflets, and this impairment may be the main mechanism of mitral valve inflow obstruction in some patients[16].

Cardiac Catheterization and Angiocardiography: This technique is usually unnecessary since the advent of echocardiography and MRI[13].

Management Options & Outcomes

Surgical resection of the membrane is the treatment of choice for patients with significant obstruction. It results in relief of symptoms and a reduction in pulmonary artery pressure. In general, the outcome following surgery is good[13]. Most patients are diagnosed with the condition in infancy or childhood. Neonates born with classic CTS have a high mortality rate: without surgical correction, only 25% survive past infancy[9]. With the advent of more routine echocardiography, a subset of patients with typical but non obstructive forms (like our patient) has been recognized. Thus far these patients appear to remain asymptomatic, with infrequent need for surgical intervention[13].

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