Cor Triatriatum Sinister Diagnosed In An Elderly Male During Work-Up For New-Onset Atrial Fibrillation

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Citation

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Abstract

Cor triatriatum sinister is a rare congenital heart anomaly that leads to a septation in the left atrium with resultant obstruction to proximal intra-atrial blood flow. The clinical manifestation depends on the presence and diameter of fenestrations in the septation with pathophysiologic consequences being similar to the effects of mitral stenosis. We present an unusual case of a 61-year-old hypertensive male who presented with sudden onset of focal neurologic deficits and new-onset atrial fibrillation. Echocardiography revealed a horizontal linear septation in the left atrium with a central ostium. This lesion effectively simulated mitral stenosis with significant obstruction to proximal intra-atrial flow and dilatation of the accessory chamber. The patient had no other structural abnormalities on echocardiography pointing to the possibility of this congenital anomaly being the underlying substrate of his atrial fibrillation. A conservative approach was adopted for his management with a good outcome on long-term follow-up.

INTRODUCTION

A diagnosis of a congenital heart anomaly in adulthood may occur as an incidental finding on cardiac imaging with no hemodynamic consequences to the patient. On the other hand, these lesions may be revealed as the cause of new-onset cardiac symptoms in adulthood. Cor triatriatum sinister is a rare congenital heart anomaly resulting in a septation in the left atrium[1]. It is rarely diagnosed in adults. Pathophysiologically, this lesion can mimic mitral stenosis in instances where the opening in the septation is significantly narrowed, resulting in obstruction to proximal intra-atrial flow. Symptomatic adults may present with atrial fibrillation and cardioembolic phenomena as seen in our patient[2]. This case demonstrates the unmasking of an unusual and possibly curable cause of atrial fibrillation using echocardiography.

CASE PRESENTATION

A 61-year-old male with a recent diagnosis of hypertension a month prior reported with sudden intense numbness in his right arm with associated inability to speak. He reported for evaluation soon after the onset of his symptoms. However, by the time he reported, his symptoms had already begun to resolve. He noted progressive improvement and subsequent complete resolution of his sympt oms within 72 hours. He had however observed new-onset exertional dyspnea and palpitations around this index event with no significant limitation in his activities of daily living. He reported compliance on his antihypertensives (oral Amlodipine 10mg daily and oral Lisinopril 10mg daily) with adequate control of his blood pressures. Physical examination was remarkable for an irregularly irregular heart rate of 114 beats per minute. An electrocardiogram (ECG) showed atrial fibrillation with a rapid ventricular response. An emergent plain head computed tomography (CT) scan done at the time of presentation was normal.

A transthoracic echocardiogram (TTE) revealed a horizontal septation, dividing the left atrium into a superior chamber into which the pulmonary veins drained, and an inferior chamber abutting the mitral valve and left atrial appendage. A transoesophageal echocardiogram revealed similar findings (Figure 1) with turbulent flow across an ostium in the septation on Colour Doppler interrogation. The peak velocity of flow across the ostium was 2.4m/s with a peak pressure gradient of 24mmHg (Figure 2). The ostium in the septation on transesophageal echocardiography (TEE) measured approximately 1.3cm in diameter (Figure 3). Colour Doppler revealed obstruction to the forward flow of blood from the superior to the inferior chamber mimicking

mitral stenosis. The jet was directed toward the anterior mitral valve leaflet causing mild deformity of the anterior leaflet. There were no other structural abnormalities on echocardiographic imaging.

Figure 1

TEE showing septation of the left atrium (LA) by a membrane (arrow). The left atrial appendage (LAA) has no visualized clot and opens into the outer chamber of the left atrium that has the mitral orifice (star). LV=left ventricle

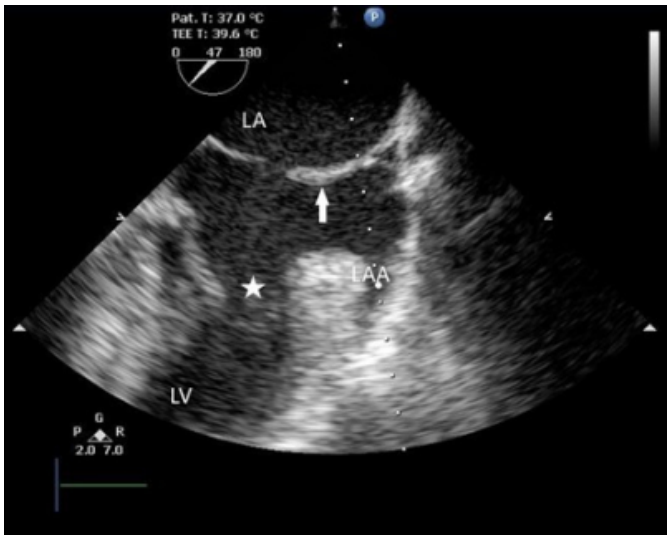


Figure 2

TTE. Apical 4 chamber view showing inlet flow jet across the membrane (arrow). LV=left ventricle

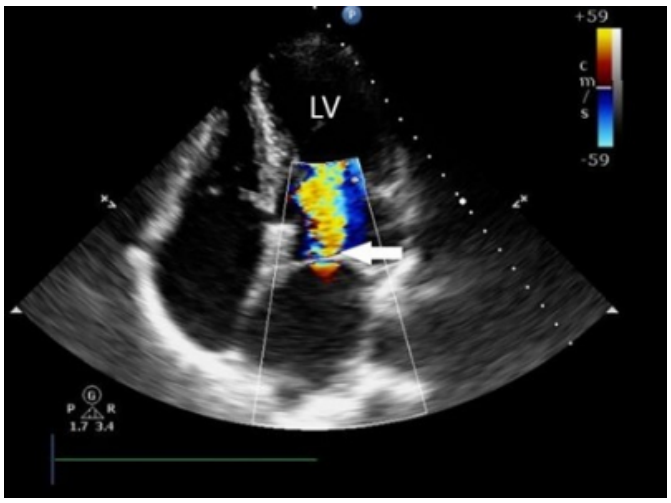
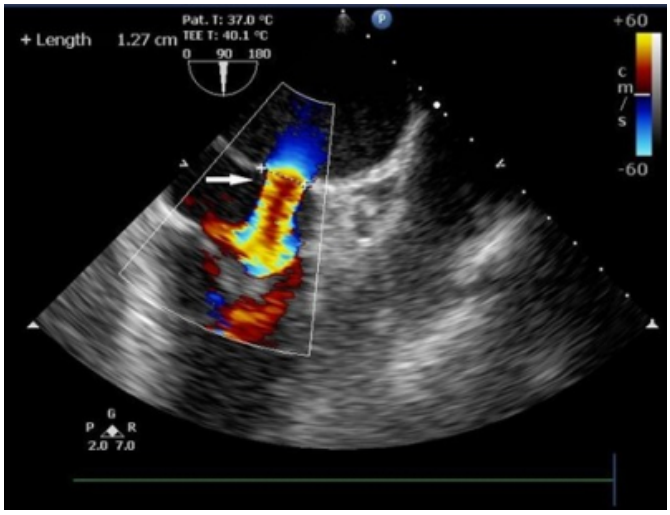


Figure 3

TEE demonstrating the diameter of the window (1.27cm) in the left intraatrial septation as well as the colour Doppler flow pattern across it (arrow).



The patient was recently diagnosed with hypertension. This put him at risk for structural heart changes which could be responsible for his atrial fibrillation. However, given the absence of significant left ventricular hypertrophy and the presence of left atrial dilatation limited to the accessory chamber, we theorized that the cause of the patient's atrial fibrillation was likely due to the pathological changes caused by the presence of the abnormal septation in the left atrium. He was managed for Cor Triatriatum Sinister complicated by atrial fibrillation, cardioembolic stroke and hypertension.

Beta-blocker therapy was added on to his antihypertensive therapy (oral bisoprolol 2.5mg daily) for rate control. Anticoagulation was initiated with Oral Warfarin 5mg and the dose was titrated to maintain his INR in a therapeutic range of 2 -3. On account of his cardioembolic phenomena, persistent atrial fibrillation and mild exertional dyspnea the patient was counseled on surgical excision of the abnormal left atrial septation as a therapeutic option. He however declined a surgical intervention and opted for a conservative management strategy. Over a period of approximately two years, the patient has remained in atrial fibrillation with adequate rate control. He has achieved optimal anticoagulation with warfarin with no recurrence of clinically overt thromboembolic phenomena. He still has mild exertional dyspnea with no significant limitation of his activities of daily living.

DISCUSSION

Cor triatriatum sinister is a rare congenital heart anomaly with an estimated incidence of 0.1% of all congenital heart

defects[1]. It is characterized by the presence of an abnormal fibromuscular septation in the left atrium, dividing it into superior (accessory atrium) and inferior (true atrium) chambers. The accessory atrium receives blood from the pulmonary veins whilst the true atrium is adjacent to the mitral valve and the left atrial appendage[3]. When present in the right atrium it is called cor triatriatrum dexter. Cor triatriatum sinister is however more common. This abnormal membrane may be horizontal or transverse in orientation and may have some fenestrations permiting flow of blood from the superior to the inferior chamber[4].

Cor triatriatrium may occur in isolation (classical) or in association with other defects (atypical), commonly atrial septal defect, patent ductus arteriosus and total anomalous pulmonary venous drainage[4]. It has also been associated with persistent left superior vena cava, ventricular septal defect, and more complex cardiac lesions such as tetralogy of Fallot, atrioventricular septal defect and double outlet right ventricle[5].

A case of cor triatriatum associated with a dilated left atrium, anomalous right pulmonary venous connection and virtual absence of the inter-atrial septum in a young girl was successfully repaired at a tertiary centre in Ghana[6]. In this case, the diagnosis was made intra-operatively as pre-operative transthoracic echocardiography was unable to demonstrate the lesion[6]. Our case represents, to the best of our knowledge, the first documented echocardiographic diagnosis of cor triatriatum sinister in adulthood, in Ghana. Globally, fewer than 250 cases had been documented by 2005[7] with limited recent data on the prevalence in adults.

The clinical presentation varies and is dependent upon the size of the window connecting the two intra-atrial compartments, the presence of associated congenital heart defects as well as other comorbid cardiac[4] and systemic conditions. Infants and children with significant obstruction due to an imperforate membrane or restrictive ostia are often symptomatic and require surgical intervention to survive[4]. Persons who survive to adulthood usually have no significant intra-atrial obstruction on account of a large window in the membrane. However with age, the membrane may become fibrosed and calcify leading to significant obstruction and consequent symptoms[4]. In late adulthood, as in our case, most persons with cor triatriatum sinister present with atrial fibrillation[8]. Atrial fibrillation has been described in 30% of published cases of adults with cor triatriatum[2].

Symptoms are due to intra-atrial inflow obstruction caused by the presence of this abnormal membrane, particularly in the presence of narrow fenestrations[4]. In the presence of significant obstruction, mimicking mitral stenosis, elevated left atrial pressures result in pulmonary venous and arterial hypertension[9] manifesting as dyspnea, orthopnea and hemoptysis[4].

Dilatation of the superior chamber may provide a substrate for atrial fibrillation[2] which may present asymptomatically or with palpitations. Thrombus formation may also occur in the dilated accessory chamber culminating in cardioembolic phenomena to the brain or systemic circulation. It has however been noted that cardioembolic phenomena may occur in the absence of atrial fibrillation[2].

Due to the rarity of this condition, there are no formal guideline directed management options for these patients[2]. However, it is agreed that patients who have become symptomatic with significant pressure gradients across the membrane should have the lesion surgically excised. For asymptomatic or minimally symptomatic adults, close follow-up with medical management when indicated is reasonable[7].

CONCLUSION

With the advent of widespread utilization of highly sensitive cardiac imaging modalities, the diagnosis of cor triatriatum sinister may become more common. The clinical presentation however varies and largely depends on the presence and size of an ostium in the abnormal septation. In asymptomatic or minimally symptomatic patients a conservative approach may be acceptable whereas surgical excision should be considered in symptomatic patients with documented complications resulting from the lesion.

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