Cor triatriatum sinister: A rare cause of atrial fibrillation in adult

Le cœur triatrial gauche : une cause rare de fibrillation auriculaire chez l'adulte.

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Résumé

Le Coeur triatrial est une anomalie cardiaque rare. Il représente 0.1 à 0.4% des cardiopathies congénitales. Son diagnostic est généralement fortuit chez l'adulte. Nous présentons le cas d'une femme âgée de 47 ans, sans antécédents cardiaques qui s'est présentée pour hernie ombilicale étranglée. L'électrocardiogramme pré opératoire a objectivée une fibrillation auriculaire et l'échographie trans thoracique a montré la division de l'oreillette gauche par une membrane fenestrée, en deux chambres antérieure et postérieure, suggestive du cœur triatrial gauche. Après une chirurgie urgente de l'hernie ombilicale étranglée, le diagnostic de cœur triatrial a été confirmé par l'échographie trans œsophagienne et le scanner cardiaque. Ces examens ont montré une oreillette gauche divisée en chambre proximale recevant le flux veineux pulmonaire et une chambre distale en contact avec la valve mitrale. Les deux chambres sont séparées par une membrane avec une large fenestration.

Mots-clés

Cœur tri atrial, fibrillation auriculaire

Summary

Cor triatriatum is a rare heart anomaly. It accounts for 0.1 to 0.4% of congenital heart diseases. It is usually an incidental diagnosis in adult. Herein we present the case of a 47-year-old woman with no cardiac history who presented with strangulated umbilical hernia. Preoperative electrocardiogram showed atrial fibrillation (AF) and transthoracic echocardiography (TTE) revealed a fenestrated membrane which divided left atrium (LA) into two chambers, anterior and posterior, suggestive of cor triatriatum sinister (CTS). After urgent surgery of strangulated umbilical hernia, the diagnosis of CTS was confirmed by transesophageal echocardiography (TEE) and cardiac computed tomography (CCT). They showed a LA divided into a proximal chamber receiving pulmonary venous flow and a distal chamber in contact with mitral valve. The two chambers were separated by a large membrane fenestration.

Keywords

Cor triatriatum, atrial fibrillation

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INTRODUCTION

Triatrial heart is a rare congenital cardiac abnormality in which left atrium (LA) is divided into two distinct chambers by a fenestrated membrane. Its incidence was reported as 0.1 to 0.4% of congenital heart disease (1, 2). This anomaly was also named "cor triatriatum" since 1905 (3). It affects usually LA leading to cor triatriatum sinister (CTS) and rarely the right atrium called cor triatriatum dexter. The classic form of CTS is characterized by the presence of a common pulmonary venous chamber named proximal or posterior chamber separated from a distal or anterior chamber containing the mitral valve and the left atrial appendage (LAA) (4). The chambers are separated by a fenestrated membrane. Other abnormalities may be associated with CTS such as mitral regurgitation, ostium secondum atrial septal defect and anomalous partial pulmonary venous connection (5). Although CTS manifests in infancy, in rare cases it appears in adulthood (6).
Herein we present a case of CTS which was diagnosed incidentally by echocardiography. Pathophysiology, diagnosis and imaging findings are briefly reviewed.

CASE PRESENTATION

A 47-year old woman with no cardiac history presented with strangulated umbilical hernia. She complained of paroxysmal palpitation and dyspnea over the previous eight months. She gave birth to four children with no complication during pregnancy and delivery. The patient was normotensive. Cardiac sounds were irregular. There were no signs of left or right heart failure. Electrocardiogram showed atrial fibrillation (AF). Preoperative transthoracic echocardiogram (TTE) showed normal left ventricular ejection fraction (60% by Simpson biplan), an enlarged LA (54 mL/m^2) and no mitral regurgitation or atrial septal defect. LA was divided into two chambers by an echogenic linear structure suggestive of a membrane (figure 1). Pulmonary artery systolic pressure was at 40 mmHg. Pressure gradient across membrane fenestration was estimated at 4 mmHg. The patient underwent an urgent surgery of strangulated hernia and after recovery, transoesophageal echocardiogram (TEE) was performed to confirm the diagnosis of CTS and estimate the size of membrane fenestration. TEE with color flow and pulse wave Doppler analysis revealed a large communication between posterior and anterior chambers (figure 2). All pulmonary veins drained into proximal LA. There was no evidence of atrial septal defect. LAA was in the distal chamber. Vacuity of LAA and LA from thrombus was confirmed but there was an important spontaneous contrast in the LA. Mitral valve was structurally normal. CCT (cardiac computed tomography) was performed, showing a LA divided by a membrane into anterior and posterior chambers (figure 3). There was a large fenestration in the membrane leading to a blood flow between the two chambers (figure 4). The patient was put on diuretics. Given the coexistence of AF, the decision was to initiate heparin anticoagulation with subsequent oral anticoagulation. After heart team discussion and given on one hand the absence of fenestration obstruction and on the other the presence of persistent AF, the decision was to not proceed to surgical membrane excision and to follow up the patient to detect fenestration obstruction.

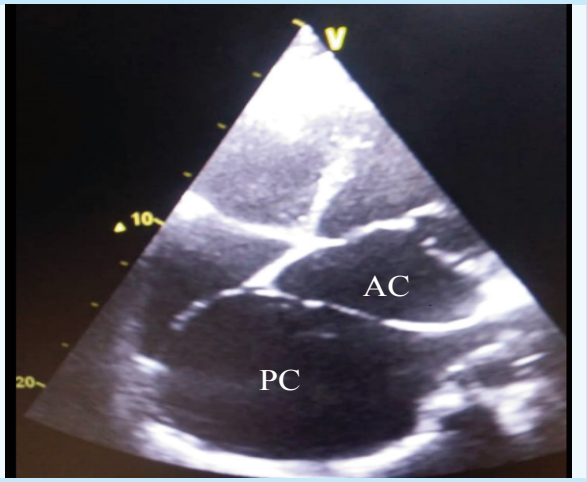

Figure 1. Transthoracic echocardiogram, four chambers view showing Cortriatriatum with anterior chamber (AC) and posterior chamber (PC).

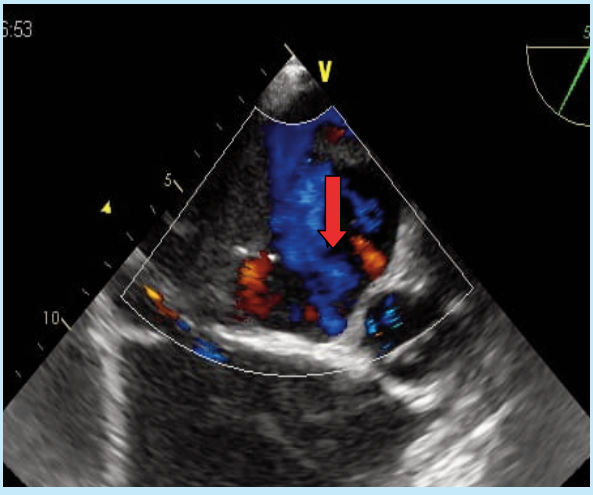

Figure 2. Multiplan transesophageal echocardiogram (at 60°) showing a large membrane fenestration (red arrow) between anterior and posterior chambers.

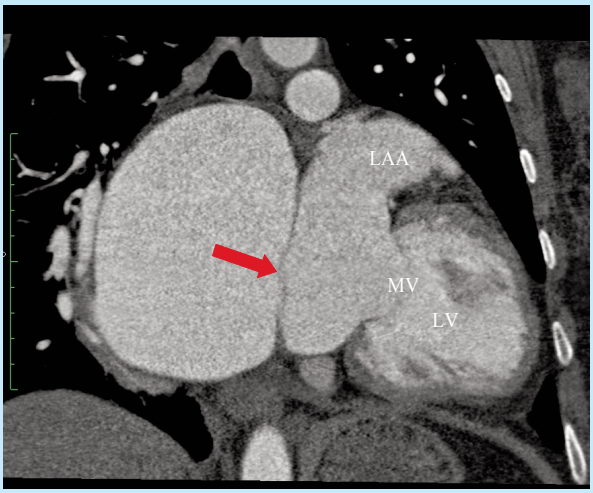

Figure 3. Cardiac computed tomography (frontal reconstruction) showing cor triatrium sinister with an enlarged left atrium divided into two chambers by a membrane (red arrow). LAA: left atrial appendage, LV: left ventricule, MV: mitral valve.

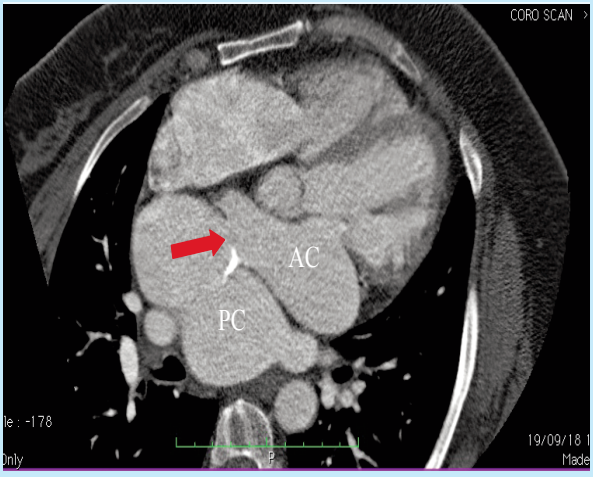

Figure 4. Cardiac computed tomography, axial cross section showing the membrane fenestration (red arrow). AC: anterior chamber, PC: posterior chamber.

DISCUSSION

CTS, first discovred by Church in 1868 [7], is a rare congenital heart malformation. It is characterized by a LA divided by a fenestrated membrane into a proximal chamber receiving inflow from the pulmonary veins and a distal chamber containing LAA [8].

The most common genesis theory is that the pulmonary vein does not incorporate normally into the LA leading to two chambers separated by a membrane (9). CTS is usually diagnosed in infancy because of early and severe symptoms caused by pulmonary venous flow obstruction and narrow fenestration or concomitant congenital anomalies (10). When symptoms appear in adulthood, they can be explained by the fenestration fibrosis or calcification, association with mitral regurgitation or AF with high ventricular rate (11). In the present case, persistent AF was the element that led to the diagnosis of CTS. AF is caused by LA enlargement and elevated filling pressure (12). The echocardiography is the imaging modality of choice for CTS diagnosis (13). It shows the membrane, determines the gradient across it and diagnoses coexisting congenital anomalies. CCT is becoming an increasingly important cardiovascular imaging modality, especially for localizing the pulmonary venous connection (14). Because of the rarity of the diagnosis, no clear recommendation exists to guide optimal modality and timing of treatment. Surgical treatment is typically intended for symptomatic patients with obstructive fenestration. It consists of membrane resection. Outcomes after surgical repair have been excellent with total resection of the membrane and repair of additional congenital abnormalities (12, 15). Balloon catheter dilatation of fenestration as a bridge to surgery could be an alternative to surgical treatment in selected patients who have acute heart failure and are unfit for surgery (16). This method was used as a definitive treatment, but long term outcomes remain to be determined. Control of concomitant diseases and adequate follow-up might be sufficient if there is no fenestration obstruction, a strategy adopted in the current case.

CONCLUSION

CTS is rare in adult and has a variety of clinical manifestations. It could be complicated by pulmonary artery hypertension, AF and thrombo-embolic events. Echocardiography is the method of choice for CTS diagnosis. Patients who have non obstructive CTS can be managed conservatively. However, routine follow-up with echocardiography is warranted because fenestration obstruction can occur due to progressive fenestration calcification or fibrosis.

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