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COR TRIATRIATUM ASSOCIATED WITH RHEUMATIC MITRAL STENOSIS: A VERY RARE ASSOCIATION

Malick Bodian, Dr. Hariniaina Ravaoavy*, Simon Antoine Sarr, Fatou Aw, Joseph Salvador Mingou, Kana Babaka, Zomana Sangaré, Ibrahima Sorry 2 Sylla, Mouhamadou Bamba Ndiaye, Adama Kane, Maboury Diao, Serigne Abdou Ba

Aristide Le Dantec Teaching Hospital, Dakar, Senegal.

***Corresponding Author: Dr. Hariniaina Ravaoavy**
Aristide Le Dantec Teaching Hospital, Dakar, Senegal.

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ABSTRACT

Cor triatriatum is a rare congenital malformation. Symptoms mimic a mitral stenosis. The combination of a cor triatriatum and mitral stenosis is less common. Some very rare cases of intraoperative findings were described. We report the case of a woman of 38 years, came to consultation for exertional dyspnea progressively worsening and palpitations during exercise. Clinical examination found mainly irregularity of the heart sounds, diastolic mitral murmur 3/6, an accentuated pulmonary component of the second heart sound. The electrocardiogram showed an atrial fibrillation and right ventricular hypertrophy. Echocardiogram concluded a cor triatriatum with a patent foramen ovale, associated with rheumatic mitral stenosis tight and severe pulmonary hypertension.

KEYWORDS: Fibromuscular membrane, rheumatic valvular disease, echocardiography, congenital heart disease, Dakar.

INTRODUCTION

The cor triatriatum is a congenital malformation representing 0.1% of congenital heart disease.[1] The atrium is subdivided into two distinct chambers by a fibromuscular membrane, one proximal, receiving the veins and the other distal chamber, in continuity with the atrioventricular valve. Classically left, it remains asymptomatic in most cases. In other cases, corresponding to groups 1 and 2 according to the Loeffler classification, the symptomatology is similar to that of mitral stenosis. The triatrial heart is often seen in association with other congenital pathologies. In 10% of cases, it is associated with atrial septal defect.[2] An association with an acquired pathology is rare. To our knowledge, a pre-therapeutic surgical discovery of a left triatrial heart associated with rheumatic mitral stenosis has been described only once. We report the case of a triatrial heart associated with mitral stenosis in a 38-year-old woman.

OBSERVATION

This is a 38-year-old woman who came to our department for dyspnea of progressing and worsening with effort and palpitations. The dyspnoea had been evolving for 2 months and was progressively worsening reaching a stage 3 of the New York Heart Association classification. It was associated with palpitations. This symptomatology occurred during the death of her husband during the period of widowhood. Clinical examination revealed irregular heart sounds, a diastolic rotation in the mitral focus, a flare, and a doubling of the second sound in the pulmonary focus. There was no sign of heart failure.

The electrocardiogram recorded complete arrhythmia with atrial fibrillation and right ventricular hypertrophy. The chest X-ray showed cardiomegaly with a 0.65 cardio-thoracic ratio, a bulge of the left middle arch. There was a redistribution of pulmonary vasculature to the apices (Figure 1).

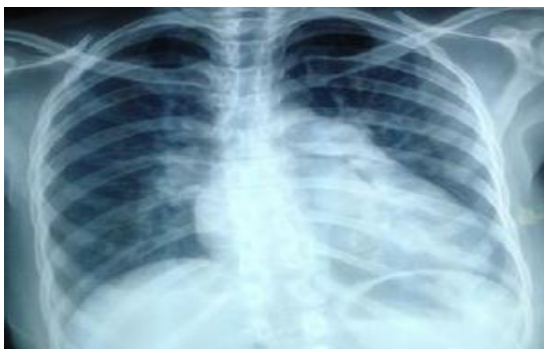

Figure 1: Thoracic chest x-ray showing global cardiomegaly, a prominent pulmonary trunk and an over-diaphragmatic tip.

Echocardiography Doppler, performed with a general electric vivid 7 ultrasound system, noted a significant dilation of the left atrium at 33 cm² surface, a left ventricle of normal size, right cavities dilated with a right ventricle at 40 mm in diameter longitudinal section and a right atrium with 22 cm² surface. Systolic function of both ventricles was normal. There was tight rheumatic mitral stenosis (Figure 2) with a mitral surface at planimetry at 0.5 cm², a mean transmitral gradient at 20 mmHg, and severe pulmonary arterial hypertension at 85 mmHg.

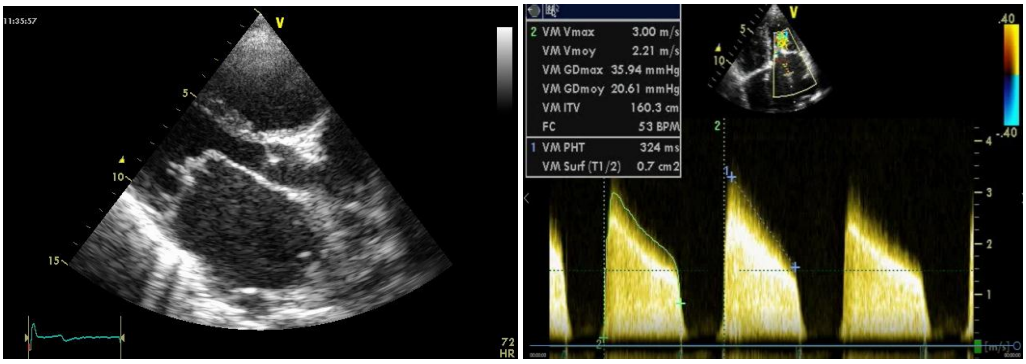

Figure 2: transthoracic echocardiography images showing the mitral stenosis with limited opening of the mitral valves and diastolic doming (hockey-stick shape) of anterior mitral valve leaflet (2A), and the continuous Doppler stenosis flow (2B).

In addition, there was a fibro-muscular membrane separating the left atrium in two chambers (Figure 3A), the proximal receiving the four pulmonary veins, the distal contact with the stenotic mitral valve and the left auricle. The fibro-muscular membrane had a single large opening of 43 mm (Figure 3B) not creating a gradient between the two chambers, corresponding to group 3 according to the Loeffler classification.

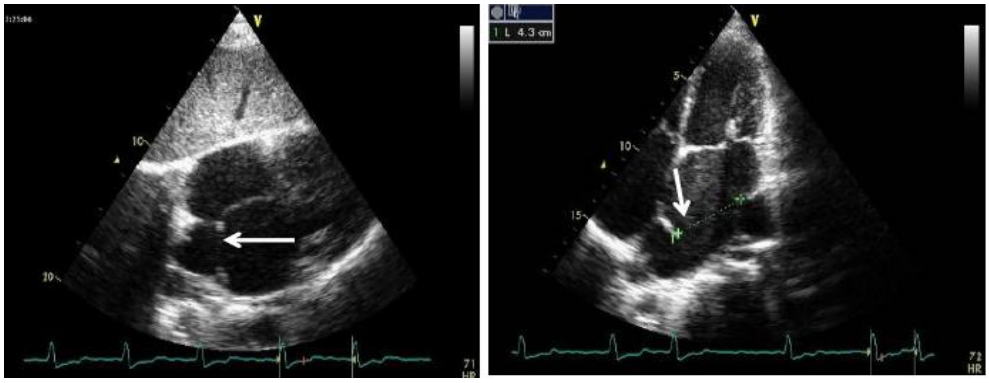

Figure 3: images of transthoracic echocardiography showing in 3A, the fibromuscular membrane subdividing the left atrium (arrow) and 3B, the wide orifice at the level of the membrane.

There was patent foramen ovale in the distal chamber, with a continuous left-right shunt (Figure 4).

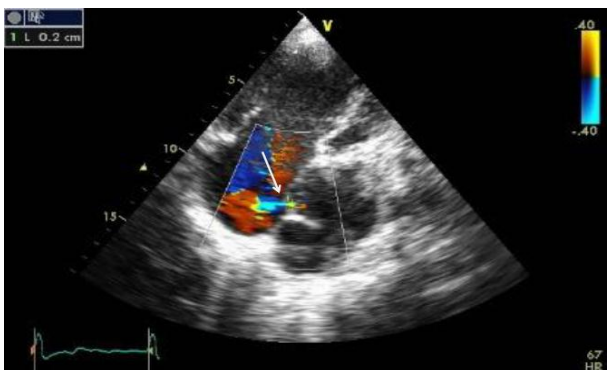

Figure 4: trans-thoracic ultrasound image showing the permeability of the foramen ovale located in the distal chamber.

The patient was under the following medical treatment: spironolactone-altizide, bisoprolol and acenocoumarol awaiting surgery for mitral stenosis as well as triatrial heart which has been indicated but not yet performed.

DISCUSSION

The cor triatriatum is a heart disease discovered by CHURCH in 1868.[3] It was named cortriatriatum by BORST in 1905. Van Praagh in 1969 argues that the triatrial heart is the result of a failure of the common pulmonary veins entering the left atrium during the fifth week of embryonic development.[4] Other authors say that the embryological origin of this pathology is still controversial. Three theories have been proposed: malseptation (the septum that divides the left atrium is the result of an abnormal proliferation of the septum primum), trapping (the left horn of the sinus tendon

entraps the common pulmonary vein and prevents its incorporation into the left atrium) and noncorporation (the incomplete incorporation of the common pulmonary vein into the left atrium).[5]

The cor triatriatum heart is a rare condition and the discovery in adulthood, such in our case, is exceptional.[6,7] The symptomatology in the triatrial heart will depend on the presence and the width of the communication between the two chambers. Indeed, the lack of communication between the two chambers is presented with a much more alarming clinical picture marked by a significant dyspnea. This dyspnea is also present in those with a high gradient between the two chambers. Wide communication at the level of the fibromuscular membrane is asymptomatic[8] apart from another pathology such as mitral stenosis. Loeffler in 1949 and Lam in 1962 proposed classifications of the triatrial heart according to the number and size of the orifices of the fibromuscular membrane.[9,10] The first group is characterized by a posterior chamber called proximal, limited by a perfectly tight membrane. It then drains systematically into the right atrium through atrial septal defect. The second group is characterized by small openings and few, responsible for a high degree of obstruction, and the third group, larger orifices with a discreet (or absence) obstructive phenomenon.

Overflow the left atrium predisposes to the onset of atrial fibrillation. A high gradient between the two chambers or tight mitral stenosis is predictive of the occurrence of atrial fibrillation as in our case. We did not find a study that reports cases of atrial fibrillation in the triatrial heart.

Concerning the association of the triatrial heart and rheumatic mitral stenosis, Fuster-Siebert in Texas in 1982 reported two cases of this association seen during the surgery of rheumatic mitral stenosis.[11] Brieger and his team reported in Australia in 1995 a similar case to our diagnosed fortuitously on the echocardiogram during the exploration of dyspnea. It was a woman, older than our patient (60 years), but with many similarities: atrial fibrillation, the location of the pulmonary veins in the proximal chamber, the permeability of the foramen ovale in the distal chamber. Nevertheless, it differs from our case by its fibro-muscular membrane having two orifices significant gradient and mitral stenosis which is moderate.

CONCLUSION

The association of cor triatriatum *and acquired heart disease* such in our case is rare. View the wide communication to the fibromuscular membrane, the symptomatology is attributable to the mitral stenosis and therefore allowed the discovery of the congenital heart disease.

CONFLICT OF INTEREST

The authors declare that they have no conflict of interest.

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