Histological impairment of the mitral valve in an adult patient affected by cor triatriatum sinistrum

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Abstract

In the case of cor triatriatum, as in other congenital abnormalities, especially if discovered in adult age, concomitant pathology of the cardiac valves should be carefully investigated. This demands an adequate pre-operative study, today possible with the modern imaging techniques. Here, we report a 19-year-old woman operated for a cor triatriatum sinister; the typical fenestrated diaphragm was excised from the left atrium. The mitral valve, characterized by a moderate-to-severe incompetence, was also excised and substituted. The histological examination revealed an altered valve structure: All its layers were completely replaced by homogeneous fibrous tissue. The atrial diaphragm showed the same histological features. About the pathogenesis of this complex disease, involving different components of the left atrium, we can suppose a unique congenital aetiology.

Introduction

Cor triatrium sinistrum is a congenital abnormality determined by the presence of a cardiac diaphragm, which divides the left atrium into two chambers. This congenital disease is well known from a diagnostic and therapeutic point of view [1,2]. The mitral valve is usually considered not involved by the disease, whose surgical repair consists only in the excision of the abnormal membrane from the left atrium [2]. Nowadays, very rare cases of cor triatriatum sinister are observed in adult age. Our histopathological finding allows to focus on a singular alteration of the mitral valve associated with this congenital abnormality, never described previously.

Case Report

A 19-year-old female came to our observation for a recent history of breathless attacks. At physical examination, a pre-systolic murmur could be heard on the left parasternal line. A transthoracic and transesophageal echocardiography demonstrated a left atrium subdivided into two chambers by a perforated membrane. A diagnosis of cor triatrium sinister, subtype A, according to the Lam classification [3], was formulated. Nevertheless, the systolic function of the left ventricle was preserved. On computed tomography, the pulmonary veins and both the venae cavae showed a regular course. The patient was subsequently operated through a median sternotomy, with the use of cardiopulmonary by-pass. The left atrium was opened, and a typical fenestrated membrane was excised. The mitral valve was thick and stiff, with evidence of moderate-to-severe incompetence on saline testing. For this reason, it was excised and replaced with a biological bio-prothesis. The pulmonary vein ostia did not display any macroscopic alteration, as well as superior and the inferior vena cava. The post-operative was uneventful, followed by a complete recovery.

We performed a detailed histopathological evaluation of the excised atrial membrane and the mitral valve. Besides to hematoxylin/eosin, histochemistry for elastic fibers (Weigert’s staining) was implemented following the standard protocols. Surprisingly, our examination revealed a completely altered mitral valve structure: All its layers (fibrosa, spongiosa, atrialis and ventricularis) were completely replaced by homogeneous fibrous tissue, containing few fibrocytes and devoid of any well-defined elastic framework. The endocardial sheet was absent. The same histological features were found in the excised diaphragm (Figure 1).
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Discussion

Rare cases of the mitral pathology associated to cor triatriatum sinistrum are reported up to here: mixomatous degeneration, hypoplasia, atresia and rheumatic calcific stenosis [4-7].

For the first time in literature, we have described in an adult patient affected by cor triatriatum sinister the same morphological abnormality in the excised atrial membrane and the mitral valve. This finding, supported by the presence of malformative fibrous tissue, suggests a complex variant of cor triatrium sinister.

From our experience, it is emerged that in every case of cor triatriatum, as in other congenital cardiac diseases, especially if discovered in adult age, careful pre-operative study of the cardiac valves should be performed, in order to rule out concomitant anomalies. This demands an adequate pre-operative study, today possible with the modern imaging techniques, such as real-time three-dimensional transesophageal echocardiography and cardiac magnetic resonance.

From a pathogenetic point of view, an abnormal or interrupted cardiac development can explain this complex pathology, interesting both the atrium and the mitral valve.

Conclusion

The presence of fibrous tissue, as the single histological component of the mitral valve, in absence of signs of superimposed degenerative or inflammatory pathology, can be considered a marker of uncorrected cardiac development, which involves both the mitral valve and the left atrium.

References