Can cor triatriatum hide mitral valve pathology?

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A 40-year-old man was admitted to our Department for paroxysmal atrial fibrillation. A transthoracic echocardiographic examination revealed a membrane attached laterally to the junction between the left upper pulmonary vein and left atrial appendage, dividing the left atrium into two chambers: the former communicating with the mitral valve and the latter with pulmonary veins. One fenestration of ~10 mm connected the two chambers and an echo colour-Doppler demonstrated a flow through this fenestration (Panel A; see Supplementary material online, Video S1). The mean pressure gradient across the membrane was 5 mmHg, while the maximum gradient was 12 mmHg. The echocardiogram also showed a dilation of the left atrial chamber (volume 59 mL/m²), a slight dilation of the pulmonary veins, a reduced mitral flow, and a mild mitral valve regurgitation due to mitral valve prolapse. The exceeding tissue of mitral valve leaflets and their systolic bellowing associated with a myxoid thickening let us to suppose a Barlow’s disease. Additional cardiac magnetic resonance imaging (MRI) evaluation depicted the membrane in the left atrium and confirmed that the pulmonary veins and the left atrial appendage were connected to the accessory chamber (Panel B). A cine gradient-echo MRI was performed to confirm the mitral valve regurgitation, and the visual assessment of the signal void area was consistent of a mild regurgitation. The patient underwent surgical intervention for excision of the membrane (Panels C and D). The post-operative course was uneventful. The post-operative transthoracic echocardiography (Day 6) showed no residual images of the membrane in the left atrium, but a significant increase in mitral regurgitation up to moderate (Panels E and F; see Supplementary material online, Video S2). At 1-month follow-up, the patient was asymptomatic, but the mitral regurgitation was still moderate and increased with respect to the preoperative echo. In our case, we suppose that the presence of the membrane hid a moderate mitral valve regurgitation. We suggest that it may be due to several factors that affect an exact echocardiographic assessment.

This thin layer is responsible for a pressure gradient in the left atrium which may reduce the volume of blood flowing through the mitral valve causing a reduction in left ventricular filling. Atrial overfilling, as it should normally appears in mitral regurgitation, is disrupted by the presence of the large membrane that occludes the passage of blood to pulmonary veins, and therefore, just the mitral chamber receives the mitral regurgitation, while the upper left atrial chamber does not. Moreover, Doppler flow studies are confused by the continuous passage of flow through the orifice in the membrane that is responsible for an aliasing effect which does not allow an accurate estimate of the regurgitation. Another aetiopathogenetic hypothesis might be explained by the lack of structural support supplied by the membrane to the shape of the left atrium because its removal could lead to a change in its tridimensional geometry. Nevertheless, the surgical procedure comprises that a small edge of the membrane should be left to its attachment site (Figure 1E) preventing therefore left atrial remodelling. The membrane was not attached to the annulus and we exclude that its resection might lead to a structural modification of the annulus itself. In conclusion, once examined this case, it would seem essential in the pre-operative period to perform a cine gradient-echo MRI to estimate the regurgitant fraction with ventricular volumetric measurements and a quantification of the regurgitant flow with phase-contrast MRI in the position of the valve, to yield a complete quantitative assessment in a cor triatriatum sinister. Furthermore, a three-dimensional study and an echocardiographic haemodynamic evaluation after exercise can be performed to precisely assess the patient’s mitral valve functionality.

Panel A Four-chamber apical view (LV, left ventricle; LA, left atrium; RV, right ventricle; RA, right atrium; white arrowhead, left atrium membrane).

Panel B Cardiac magnetic resonance imaging.

Panel C Intra-operative picture showing the membrane (asterisk, left atrium membrane).

Panel D Gross specimen of the membrane.

Panel E White arrow, residual membrane.

Panel F Post-operative echo colour-Doppler evaluation showing moderate mitral valve regurgitation.

Supplementary material is available at European Heart Journal online.