Triple-orifice congenital stenotic mitral valve: a unique finding characterized by multimodality echocardiography

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Case report

A 23-year-old woman, with mitral stenosis since childhood, presented with worsening exertional dyspnoea. She had no previous acute rheumatic fever. Two-dimensional transthoracic echocardiogram demonstrated thickened mitral leaflets, with diastolic dome-shape due to restricted posterior leaflet and a formerly non-recognized triple-orifice mitral valve (Panels A and B). Doppler revealed a mean gradient of 17 mmHg and larger orifice area was 0.6 cm². These findings were consistent with severe congenital mitral stenosis. In addition, a muscular ventricular septal defect (VSD) and severe pulmonary hypertension were noticed. In order to better characterize mitral valvular anatomy, 2D and 3D transoesophageal echocardiogram were performed, clearly confirming the presence of a triple-orifice stenotic mitral valve, with two papillary muscles of unequal size (Panels C–F; Supplementary data online, Videos S1 and S2). Because of chordae shortening, it was difficult to evaluate the pattern of chordae distribution.

At surgery, it was noticed that the two mitral leaflets were highly asymmetric and mitral valve behaved like having a funnel-shaped single leaflet with three orifices. Papillary muscles were of unequal size, closely located, and the posteromedial papillary muscle attached almost directly to the mitral leaflet as chordae tendinea were so reduced. Moreover, no clear relation between chordae coming from the anterolateral papillary muscle and the three orifices in the main anterior leaflet was discerned. The patient was submitted to bioprosthetic valve replacement, tricuspid valve repair because of annulus dilation and VSD closure.

This case highlights that the spectrum of mitral valve anomalies associated with obstruction to left ventricular inflow is complex and very assorted. Included in this spectrum is the double-orifice mitral valve, a rare congenital malformation characterized by two separate valve orifices of varying sizes associated with abnormalities of the subvalvular apparatus. To our knowledge, this is the first case of a triple-orifice mitral valve.

Figure 1 Thickened mitral leaflets with a diastolic doming shape could be observed by 2D-transthoracic echocardiography apical four-chamber view (A). Transoesophageal echocardiographic two-chamber view better defined the presence of a triple-orifice mitral valve (B). Two papillary muscles (white arrows), with shortened interpapillary muscle distance, were identified by transgastric short-axis view (C). The funnel-shape aspect of the mitral valve, the main orifice, and the shortened chordae were better appreciated by 3D Zoom Echo (D) as well as intraoperatively (E) and the three orifices (the main and two smaller sized in an anterior position) were unequivocally demonstrated by full volume 3D colour (F).

Supplementary data

Supplementary data are available at European Journal of Echocardiography online.