Systemic scleroderma with moderate to severe mitral regurgitation: Unusual three-dimensional echocardiographic features

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A 60-year-old woman was admitted with pulmonary edema and a holosystolic apical murmur. She had had Raynaud's disease for two years and scleroderma for 20 years. On two-dimensional echocardiography, the mitral valve appeared mildly thickened, the papillary muscle was much thickened, and the chordae tendineae were sclerotic and much shortened (Figure 1A). Colour Doppler imagery showed severe mitral regurgitation (Figure 1B). Three-dimensional echocardiography confirmed very thick papillary muscles and almost direct insertion of papillary muscles on the mitral leaflets due to severe

chordal shortening, which prevented proper apposition of the mitral leaflets (Figures 1C, 1D and 1E). A virtual section of the papillary muscles on three-dimensional imaging electronically showed patches of echogenic tissue (presumably fibrosis) within the ventricular myocardium, a hallmark of systemic sclerosis (1).

REFERENCE

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Figure 1 A Two-dimensional echocardiography, parasternal long-axis view, showing the papillary muscle (PM) inserted apparently directly on the anterior leaflet of the mitral valve (MV) due to very shortened chordae tendineae (CT). B Two-dimensional echocardiography, modified parasternal long-axis view, showing black and white rendering of colour Doppler. C Three-dimensional echocardiography of the interior of the left ventricle (LV). Arrow indicates very shortened CT connecting the anterior mitral leaflet (AML) to the PM. D Three-dimensional echocardiography showing LV structures: AML, CT, PM and trabeculations (TRA). E Three-dimensional echocardiography of the LV in a different orientation of the same structures. AV Noncoronary cusp of aortic valve; LA Left atrium; LVO Left ventricular outflow; MR Wide mitral regurgitant jet

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