Primary Oxalosis With Cardiac Involvement
Echocardiographic Features of an Unusual Form of Cardiomyopathy

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A 40-year-old man with primary oxalosis, nephrocalcinosis, severe renal failure, and a permanent dual-chamber pacemaker due to intermittent complete atrioventricular block was admitted to the hospital because of increasing dyspnea. His chest x-ray showed an increased cardiothoracic ratio, pulmonary venous congestion, and bilateral pleural effusions. ECG demonstrated sinus tachycardia at 97 bpm with atrioventricular sequential pacing. The transthoracic echocardiogram showed a mildly dilated left ventricle with moderately severe systolic dysfunction (Figure 1). The right ventricle was mildly dilated, with moderately impaired systolic function. There was marked concentric thickening of both left and right ventricles, with patchy echodense speckled reflections in the myocardium (Figure 2). There was moderate pericardial effusion, with no signs of cardiac tamponade. Moderate tricuspid regurgitation was present with a right ventricular systolic pressure of 48 mm Hg. Doppler mitral inflow and pulmonary venous flow indices were consistent with a restrictive left ventricular filling pattern (Figure 3). Color M-mode tissue Doppler echocardiography of the posterior wall of the left ventricle showed a reduction of both myocardial velocities and myocardial velocity gradient during the cardiac cycle and abnormally positive myocardial velocity gradient during isovolumic relaxation. This finding was consistent with the late stage of infiltrative cardiomyopathy. A right ventricular endocardial biopsy confirmed the diagnosis of cardiac oxalosis by showing extensive deposition of crystals typical of calcium oxalate, predominantly intracellularly within myocytes (Figure 4).

Primary oxalosis is a rare hereditary metabolic disorder in which an enhanced production of oxalic acid leads to hyperoxalemia and a deposition of calcium oxalate in different body organs (eg, kidney, heart). We described the echocardiographic features of cardiac oxalosis manifesting as a form of infiltrative cardiomyopathy with restrictive physiology.
Figure 2. Two-dimensional apical 2-chamber view during (A) systole and (B) diastole. Note the increased speckled myocardial reflections with neighboring echo dropouts (arrow) in the left ventricular inferior wall and the moderately reduced systolic function (ejection fraction, 38%).

Figure 3. A, Mitral inflow pattern; B, pulmonary venous flow; C, mitral annulus motion by tissue Doppler echocardiography; and D, color M-mode tissue Doppler echocardiography of the left ventricular posterior wall with the quantification of mean myocardial velocities (MMV) and myocardial velocity gradient (MVG). Dotted green lines show isovolumic relaxation time.

Figure 4. A, Right ventricular endocardial biopsy showing extensive deposits of crystalline material that was predominantly present intracellularly (hematoxylin and eosin; original magnification ×400); B, under polarized light, the biopsy demonstrates birefringence (hematoxylin and eosin; original magnification ×200). Note that there was no associated inflammation or necrosis of myocytes.
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