An atypical form of severe aortic regurgitation in a patient with ankylosing spondylitis (Morbus Bechterew)
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Ankylosing spondylitis (M. Bechterew) is an inflammatory disease that may involve the aortic root (aortitis) as well as the aortic and less often the mitral valve (valvulitis). In such cases, fibrosis may lead to thickening and dilation of the aortic wall as well as to thickening and retraction of the aortic cusps and of the anterior mitral leaflet with subsequent valvular regurgitation. We report on a patient with ankylosing spondylitis and an atypical form of severe aortic regurgitation.

Case report: A 47-year-old man presented with a 10-day history of dyspnea. For more than two decades the patient had been suffering from ankylosing spondylitis that had led to severe immobilisation of his spinal column. A prior therapy with etanercept had been stopped because of insufficient benefit. Actually, the patient took naproxen a non-steroidal antiinflammatory drug. At presentation the patient was afebrile, heart rate was 96bpm and blood pressure was 160/60mmHg. Physical examination revealed normal heart sounds but a 4/6 mitral regurgitation murmur, a 2/6 aortic regurgitation murmur and bilateral pulmonary rales. A chest X-ray showed cardiac enlargement and bilateral pleural effusions. In the ECG sinusrhythm and signs of left ventricular hypertrophy were found. Blood analysis showed no signs of inflammation and blood cultures were negative.

Transthoracic echocardiography revealed a dilated and hypertrophied left ventricle with normal systolic function and marked dilation of the left atrium, whereas right-sided cardiac chambers were normal. The aortic root was of normal size. Tricuspid aortic valve showed only slight thickening of the cusps but an isolated prolapse of the right-crownary cusp with subsequent severe aortic regurgitation. In addition, incomplete coaptation of the otherwise normal mitral leaflets resulted in moderate mitral regurgitation. No echocardiographic features typical for infective endocarditis were present. Dilation of liver veins and of the inferior vena cava with blunted respiratory changes indicated the presence of pulmonary hypertension.

Comment: Although involvement of the aortic root and/or the aortic and mitral valve can occur in a considerable proportion of patients with ankylosing spondylitis, clinically evident aortic root or valvular disease occurs in less than 20% of patients. Our case illustrates that pathomechanisms other than cusp fibrosis and retraction may also exist in patients with ankylosing spondylitis.