Slowly progressive conduction system disturbance in a patient with polymyositis

Polimiyozitli bir hastada yavaş ilerleyen ileti sistemi bozukluğu

Polymyositis (PM) is an autoimmune disease characterized by progressive weakness of the proximal skeletal muscles that can affect the heart. Cardiac manifestations usually present with minimal symptoms and the most often reported include congestive heart failure as consequence of diffuse myocarditis, arrhythmias and conduction system disturbances that can be related to secondary vascular changes and cellular infiltration. Vascular alterations in the coronary arteries have also been reported such as vasculitis, intimal proliferation, media sclerosis and microvessel disease; that has been associated with vasospastic angina. Small vessel disease characterized by narrowing of vessel lumen by smooth muscle hyperplasia with little or no intimal proliferation was also observed. This may cause clinical symptoms like palpitations and angina pectoris. Electrocardiography (ECG) abnormalities included mainly left anterior fascicular block (LAFB) and right bundle branch block (RBBB). AV blocks have been reported less frequently. Autopsy studies revealed histopathological changes in the conducting system including lymphocytic infiltration, fibrosis of the sinoatrial node and contraction band necrosis. Some of these cases evolved into complete heart block.

We report a 45-year-old woman with eight-year history of PM who presented with syncope at rest. The patient had no history of cardio-vascular disease or risk factors for coronary atherosclerotic disease (smoking, diabetes mellitus or systemic hypertension) and did not present menopause.

ECG evolution of the last eight years showed incomplete RBBB, QRS axis at 0° , and PR interval at 180 ms (Fig. 1 Panel A). Two years later, the ECG showed bifascicular block (RBBB and LAFB) with a PR prolongation

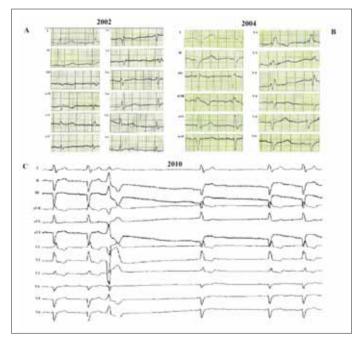


Figure 1. Electrocardiogram evolution in a patient with polymyositis
A-incomplete right bundle branch block, QRS axis at 0°, and PR interval at 180 ms,
B-bifascicular block (right bundle branch block an left anterior fascicular block) with a PR
prolongation to 200 ms, C-complete atrioventricular block

to 200 ms (Fig. 1 Panel B). During this admission (8 years after the initial presentation, she presented with complete AV block (Fig. 1 Panel C), symptomatic by syncope. A permanent pacemaker was implanted.

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A ring in the heart: an atrial septal aneurysm

Kalpte bir yüzük: Atriyal septumda bir anevrizma

An atrial septal aneurysm (ASA) is a thin, located segment of the atrial septum that bulged into the right or left atrium. They are mobile and can be seen moving between the atria during the cardiac cycle. Rarely, some ASAs mimic a right/left atrial cyst or tumor.

A 45-year-old man was admitted to our department because of dyspnea. He had history smoking. On physical examination, there were no audible murmurs, rales or rhonci. The chest radiography and electrocardiography were normal. His blood pressure was 120/85 mmHg. Echocardiography demonstrated normal left ventricular function, mild mitral regurgitation, mild tricuspid regurgitation and interatrial septal aneurysm, mimicking left atrial ring shaped cystic mass (Fig.1, Video1. See corresponding video/movie images at www.anakarder.com). Multiple parasternal long- and short-axis, apical 4-chamber, and modified echocardiographic evaluations revealed that the circular cyst-like image was a cross-section of an interatrial septal aneurysm (Video 2.

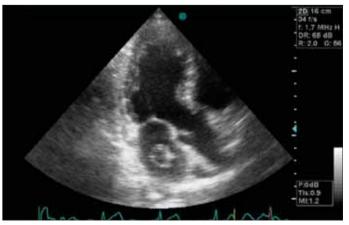


Figure 1. Transthoracic echocardiographic apical 5-chamber view of a ring shaped cystic mass in the left atrium

See corresponding video/movie images at www.anakarder.com). Further evaluation by contrast echocardiography and transesophageal echocardiography for ring shaped cystic mass confirmed the ASA and allowed to exclude a left-to right shunting (Video 3. See corresponding video/movie images at www.anakarder.com).

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Diagnosis of a caseous mitral annular calcification

Kazeöz bir mitral anülüs kalsifikasyonun tanısı

A 75-year-old woman was presented with shortness of breath and palpitations. After initial evaluation, a transthoracic echocardiographic examination was planned and showed a large, round, echodense mass with central areas of echolucencies attached to the posterior mitral

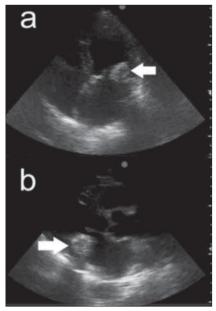


Figure 1. The apical (a) and parasternal (b) TTE views of a large, round, echodense mass with central areas of echolucencies attached to the posterior mitral annulus

TTE-transthoracic echocardiography

annulus (Fig.1, Video 1-3. See corresponding video/movie images at www.anakarder.com). Caseous calcification of the mitral annulus (CCMA) was suspected. A multidetector computed tomography (MDCT) scan without contrast agents (because of moderate chronic kidney disease) was performed to aid differential diagnosis and to establish the nature of the mass. The bone window and level settings showed a rim of peripheral calcification with central homogeneous hyperdense mass lesion (Fig. 2A). The mediastinal window and level settings showed homogeneous hyperdense mass lesion that cannot be differentiated from other calcific structures (Fig. 2B).

CCMA could be misdiagnosed as infective endocarditis, myocardial abscess, benign or malignant cardiac tumors (such as myxoma, lymphoma, sarcoma, metastatic disease), thrombus, lipomatosis of the atrioventricular groove, and enlarged lymph nodes. In cases with CCMA, misdiagnosis may lead to unnecessary cardiac surgery. In this case, a diagnosis was made according to the echocardiographic and MDCT findings. In cases of CCMA, pathologic confirmation is needed for a definitive diagnosis, but imaging findings may defer pathologic examination.

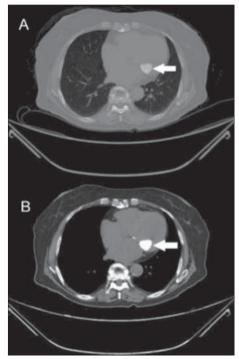


Figure 2. (A,B) MDCT views of the same image of a mass in two different windows and level settings

MDCT-multidetector computed tomography

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