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FIT Clinical Decision Making

AN UNUSUAL CASE OF CARDIAC AMYLOIDOSIS

Poster Contributions
Hall C
Sunday, March 30, 2014, 9:45 a.m.-10:30 a.m.

Session Title: FIT Clinical Decision Making: Heart Failure / Cardiomyopathies
Abstract Category: Heart Failure and Cardiomyopathies
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Background: Multiple myeloma manifests with back pain, anemia and renal failure. It is associated with AL amyloidosis in only 10%-15% of patients. Fewer than 5% of patients with AL amyloidosis involving the heart have clinically isolated cardiac disease. Recurrent pleural effusion as a manifestation of AL amyloidosis is rare.

Case: A 78-year-old male presented with a three-month history of progressive exertional dyspnea and recurrent right transudative pleural effusions that had been tapped twice. Previous medical history was unremarkable. Physical examination showed mildly distended jugular veins, mild bilateral pedal edema, and decreased air entry over the lower half of the right lung. Heart sounds were normal without murmurs or gallop. Chest radiography revealed a large right pleural effusion without cardiomegaly or pulmonary congestion. ECG was unremarkable. Hemoglobin, renal function, liver enzymes and electrolytes were all normal.

Decision-making: Echocardiography was notable for moderately thickened walls (up to 17mm) and diastolic dysfunction grade 2/4 with elevated E/e' ratio. There was preserved systolic function, mildly dilated left atrium, normal aortic valve, and mild mitral regurgitation. Serum protein electrophoresis did not detect a monoclonal band. Serum immunofixation electrophoresis was also negative. Subcutaneous abdominal fat pad aspirate stained for amyloid with Congo red was negative. In the absence of hypertension or aortic stenosis to explain the cardiac hypertrophy, AL amyloidosis was still considered a likely diagnosis, and therefore a serum free light chain (FLC) assay was performed. This showed a very low kappa/lambda FLC ratio of 0.07 (normal 0.26-1.65). Bone marrow biopsy immunohistochemical staining and Congo red stain were consistent with multiple myeloma and AL amyloidosis.

Conclusion: This is a case of restrictive cardiomyopathy with atypical clinical presentation and negative initial work-up for amyloidosis. A high index of suspicion for systemic amyloidosis warrants obtaining also a serum FLC assay as it can detect circulating FLC with 10-fold greater sensitivity than immunofixation in AL amyloidosis, as exemplified in this patient.