MULTIORGAN INVOLVEMENT OF AMYLOID: HOW TO FLY UNDER THE RADAR

Poster Contributions
Poster Hall B1
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Session Title: FIT Clinical Decision Making: Heart Failure and Cardiomyopathies
Abstract Category: Heart Failure and Cardiomyopathies
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Background: Infiltrative cardiomyopathy is an uncommon cause of systolic or diastolic dysfunction and progressive heart failure (HF). Early diagnosis and treatment can be achieved by careful clinical exam and non-invasive tests, but tissue diagnosis remains important when considering chemotherapy options.

Case: A 60 years-old gentleman was admitted for the second time in two months for dyspnea. He had a history of hypertension, idiopathic patchy motor neuropathic progress for 2 months, dysphagia and voice hoarseness for 6 months, and cryptomeningitis 10 years ago. Physical examination revealed a cachectic man with a jugular venous pressure of 8cm above the sternal angle, a normal S1 and S2, an S3, no murmurs, and decreased breath sounds to the mid right lung field. His EKG showed a normal sinus rhythm with low voltages. An echocardiogram showed significant biventricular hypertrophy and a depressed ejection fraction of 20%.

Decision Making: The constellation of heart failure symptoms, unilateral pleural effusion, and increased wall thickness with a well controlled blood pressure were highly suggestive of an infiltrative cardiomyopathy. In particular, low voltages on EKG made amyloid most suspicious. A fat aspiration biopsy was negative for amyloid deposition. The patient improved with diuresis. Infectious work-up and further neurologic imaging were non-revealing. The constellation of neuropathy and unexplained systolic dysfunction, clinical suspicion remained high for amyloid. A fat pad block biopsy was therefore planned prior to further cardiac tests; it was positive for amyloid light-chain subtype amyloidosis by Congo Red stain. A cardiac CT was done and showed subendocardial delayed gadolinium enhancement in a pattern consistent with amyloid; this was deemed enough evidence of amyloid cardiomyopathy in the right clinical setting and a right ventricular biopsy was not performed. He was started on bortezomid and prednisone. With chemotherapy, he has had improvement in his energy levels and HF symptoms.

Conclusion: This case demonstrates that sequential and sometimes repetitive testing may be required to diagnosed infiltrative cardiomyopathies, in this case amyloid.