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Cardiac Amyloidosis: Surviving the kitchen sink

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Amyloidosis refers to the abnormal deposition of proteins in tissues causing physiologic abnormalities in their function. Amyloidosis is often an overlooked cause of commonly encountered medical issues commonly affecting the heart, blood vessels and kidneys. A 61-year-old woman with a history of systolic heart failure (EF 10% 3 months PTA), ischemic CVA (3 years PTA), pericardial effusion s/p pleuro-pericardial window (nl tissue), CKD stage III, DM, HLD and HTN was admitted to the hospital with hypertensive emergency and pulmonary edema. Upon admission, the patient was chest pain free, however, admission ECG revealed a new LBBB with elevation of cardiac biomarkers (peak troponin, 9). She underwent emergent cardiac catheterization revealing a 50% lesion in the LAD with otherwise non-obstructive coronary artery disease. CT angiogram while negative for PE, revealed an incidental bilaterally enlarged cystic thyroid. The patient was treated for flash pulmonary edema and hypertensive emergency with nitroglycerin, furosemide, morphine and BiPAP. After resolution of pulmonary edema and stable hemodynamics, she was medically optimized for systolic heart failure. Blood pressure remained persistently elevated with systolic measurements in the range of 180-200 despite full doses of furosemide, carvedilol, spironolactone, hydralazine, isosorbide dinitrate and clonidine 1.8mg/day. A renal ultrasound revealed bilateral kidney enlargement with no cysts indicative of chronic kidney disease. A trans-thoracic echo revealed an EF 5-10%, mild MR, moderate TR, moderate concentric LVH, global wall thickening and a small pericardial effusion. In light of the patient's non-ischemic cardiomyopathy, thyroid disease, systemic vasculopathy, hypertension refractory to medical therapy, chronic kidney disease and echocardiogram findings an underlying etiology was sought. Workup included serum studies for autoimmune disease (ANA, ANCA, RF, etc.), a fat pad biopsy and SPEP/UPEP. The fat pad biopsy returned positive for Congo red staining; SPEP/UPEP were inconclusive. The patient was diagnosed with amyloidosis and referred for endomyocardial biopsy and fitted for a cardiac lifevest with eventual plan for CRT/ICD placement. Despite diagnosis, the patient's prognosis remains poor. She was discharged home with cardiology and primary care follow-up. This case illustrates the importance of maintaining a broad differential in patients with non-ischemic heart failure and multi-system involvement including medically-refractory hypertension. Amyloidosis is a rare but severely debilitating syndrome but with proper identification and diagnosis, important work-up and treatment can be initiated to improve quality of life. Further differentiating amyloidosis into the respective subtype via biopsy will further help guide management.

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