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Reversible Pre-Capillary Pulmonary Hypertension in a Patient with Systemic AL Amyloidosis- A Rare Association and Outcome

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Background: The presence of pulmonary hypertension associated to AL amyloidosis is rare. AL amyloidosis may deposit in the lung parenchyma and pulmonary vasculature. All previously reported cases had a rapid fatal outcome. This is a case report of a 64-yo woman previously healthy who presented with severe right sided heart failure symptoms, shortness of breath, and weight loss who was diagnosed simultaneously with advanced AL amyloidosis and pre-capillary pulmonary hypertension.

Methods: Retrospective chart review and case report.

Results: 64 yo woman who initially presents with atrial fibrillation and diastolic heart failure. She undergoes an atrial fibrillation ablation successfully. After the ablation she reports persistent shortness of breath and fatigue with evidence of resting oxygen desaturation at 89%. Repeat echo indicated evidence of a restrictive cardiomyopathy. A cardiac MRI is performed showing right ventricular dilatation with flattening of the septum in diastole and slight left ventricular dysfunction without evidence of delayed enhancement. PFT revealed a DLCO 31% predicted. Serum light chains revealed a marked elevated kappa and lambda chains with an abnormal K/L ration. Fat pad biopsy and bone marrow biopsy confirmed the diagnosis of AL amyloidosis. The patient underwent a right heart cath that revealed the following pressures: RA= 17 mm Hg, RV=80/18 mm Hg, PA= 85/30 50 mm Hg, PW=15, CI=3.91, PVR=717. The patient was treated with Bortezomib, Cyclophosphamide and Prednisone with almost normalization of Kappa chains and K/L ratio. Pulmonary hypertension was treated with combination of tadalafil and ambrisentan. After 8 months of intensive medical therapy the right ventricular function normalized without evidence of a restrictive pattern on echocardiography. Repeat right heart cath showed: RA=7 mm HG, RV=45/7 mm Hg, PA=45/15 25 mm Hg, PW=13 mm Hg, CI=3.765 and PVR=178.

Conclusions: This case is an example of a rare association between systemic AL amyloidosis and group WHO 1 pulmonary hypertension with an excellent outcome. Approaching two lethal diseases in an aggressive, multidisciplinary, and closely monitored approached allowed this patient to achieve rapid remission.



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