Successful Treatment of Idiopathic Pulmonary Vasculopathy With Targeted Pulmonary Vasodilator Therapy

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INTRODUCTION: Pulmonary vasculopathy is associated with a diverse group of diseases that manifest with various histopathological features. We describe a patient with idiopathic pulmonary arteriopathy and normal hemodynamics on right heart catheterization (RHC) who benefited markedly from bosentan and subsequent tadalafil with resolution of chronic hypoxic respiratory failure.

CASE PRESENTATION: A 44 year old African American female with a history of hypertension and fibromyalgia presented to our center in 2006 for evaluation of progressive dyspnea since 2001. She had been treated with methotrexate and prednisone without improvement. Pulmonary function tests (PFTs) revealed moderate restriction with severely reduced diffusion capacity (DLCO). No parenchymal pathology or lymphadenopathy was found on a high resolution CT scan. CT angiogram was negative for pulmonary embolism. Extensive cardiac and rheumatologic workup was negative. Patient endorsed NYHA class IV symptoms requiring >10 liters of oxygen. Open lung biopsy revealed pulmonary arteriopathy characterized by muscularization of arterioles and concentric mural thickening of muscular arteries with muscular hypertrophy and intimal expansion. No venous pathology or thromboembolic disease was identified. RHC showed normal hemodynamics. Due to progressive respiratory failure, she was trialed on bosentan with resultant rapid and sustained resolution of hypoxia. In 2009 an interruption in therapy led to clinical deterioration. She improved with reinitiation. She remained well until 2014 when she developed increased dyspnea. Due to concern for waning bosentan effect, she was transitioned to tadalafil with full recovery. She continues to do well without supplemental oxygen requirement.

DISCUSSION: Pulmonary arteriopathy is well described with cardiopulmonary diseases. It involves muscular arteries accompanying bronchioles and arterioles within alveolar septae. Etiologic considerations include pulmonary vascular disease, cardiac abnormalities, parenchymal lung disease, thromboembolic disease, drug reactions and connective tissue disease.

CONCLUSIONS: We present the first case of idiopathic pulmonary vascular disease unexplained by a systemic condition with a remarkable and sustained response to pulmonary vascular targeted therapy. Isolated pulmonary vasculopathy should be considered in unexplained hypoxic respiratory failure.


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