

Cardiac Arrest and Pulmonary Arterial Hypertension in Scurvy; Rediscovery of an old Observation.

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Objectives

- Raise awareness about a modern presentation of scurvy, and the role it plays in the development PAH.
- Identify mechanisms for the development of pulmonary arterial hypertension in vitamin C deficiency.

Pulmonary arterial hypertension (PAH) is a rare, progressive disease of the pulmonary vasculature. In a few confirmed cases, scurvy has been linked to PAH, and can present as a severe, but reversible, cause of this condition through non-hypoxic activation of hypoxia-inducible transcription factors (HIF) and low nitric oxide (NO) levels in the pulmonary vasculature, leading to subsequent pulmonary vasculopathy and an exaggerated pulmonary vasoconstrictive response. In this case, a delayed diagnosis of scurvy resulted in fatal PAH. Our patient is a 73-years-old female, 10 years post-bariatric surgery. She presented with two-month history of progressive dyspnea. Her physical exam was positive for cardiac and vascular concerns, and scattered ecchymosis bilaterally. Laboratory data uncovered mild acute kidney injury. Echocardiography (ECHO) showed normal global systolic function of left ventricle, ejection fraction of 75% with normal diastolic function. The estimated right ventricular systolic pressure (RVSP) was measured at 112. PFTS showed a mild decline in DLCO. The patient was initially diagnosed with pulmonary hypertension, then PAH diagnoses was confirmed by right heart catheterization RHC. She had negative detailed workup for PAH. Sildenafil (20 mg) was administered with no hemodynamic improvement. Inotropic therapy was initiated, but also without clinical improvement. A reviewed history revealed that her diet was significantly unbalanced, lacking fruits and vegetables, and that the patient had poor compliance to vitamins and supplements prescribed post-bariatric surgery. Given the patient's history and presentation, vitamin C deficiency was suspected, and levels were ordered. However, before these results were available, the patient developed severe cardiogenic shock that was non-responsive to maximal medical supportive therapy, resulting in her death. Post-mortem, the previously sent lab results revealed an ascorbic acid level of zero. It is reasonable to assume that administration of supplemental doses of vitamin C may have prevented this outcome as reported in few occasions.