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Case report

Pulmonary hypertension secondary to pulmonary veno-occlusive disease complicated by right heart failure, hypotension and acute kidney injury

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ABSTRACT

Pulmonary veno-occlusive disease (PVOD) is rare condition which can lead to severe pulmonary hypertension, right ventricular dysfunction, and cardiopulmonary failure. The diagnosis of PVOD can be challenging due to its nonspecific symptoms and its similarity to idiopathic pulmonary arterial hypertension and interstitial lung disease in terms of diagnostic findings. This case describes a 57 year old female patient who presented with a 5-month history of progressive dyspnea on exertion and nonproductive cough. Workup at another hospital was nonspecific and the patient underwent surgical lung biopsy due to concern for interstitial lung disease. She subsequently became hemodynamically unstable and was transferred to our hospital where she presented with severe hypoxemia, hypotension, and suprasystemic pulmonary artery pressures. Preliminary lung biopsy results suggested idiopathic pulmonary arterial hypertension and the patient was started on vasodilating agents, including continuous epoprostenol infusion. Pulmonary artery pressures decreased but remained suprasystemic and the patient did not improve. Final review of the biopsy by a specialized laboratory revealed a diagnosis of PVOD after which vasodilating therapy was immediately weaned off. Evaluation for dual heart-lung transplantation was begun. The patient's hospital course was complicated by hypotension requiring vasopressors, worsening right ventricular dysfunction, and acute kidney injury. During the transplantation evaluation, the patient decided that she did not want to undergo continued attempts at stabilization of her progressive multi-organ dysfunction and she was transitioned to comfort care. She expired hours after removing inotropic support.

1. Background

Pulmonary veno-occlusive disease (PVOD) is a rare cause of pulmonary arterial hypertension (PAH) in which the pulmonary venules and small veins undergo intimal fibrosis, leading to pulmonary hypertension, interstitial and pleural edema, and right ventricular failure [1]. PVOD has been associated with connective tissue diseases, HIV infection, bone marrow transplantation, and chemical exposures but is idiopathic in the majority of cases [2]. Definitive diagnosis requires lung biopsy for histology but acquiring such a tissue specimen is usually contraindicated in patients with respiratory and hemodynamic instability [3]. Although there are alternative diagnostic tests that are less invasive than biopsy, without histology it can be difficult to differentiate PVOD from idiopathic PAH and interstitial lung disease (ILD) [4]. Treatments for PVOD reported in the literature have included PAH-specific therapies, which are associated with an increased risk of pulmonary edema, but which may act as a bridge to lung or combined lung-heart transplantation [5].

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2. Case description

A 57-year-old female presented with a 5-month history of progressively worsening dyspnea and non-productive cough. Her symptoms were initially associated only with moderate exertion, but progressed to dyspnea at rest. Her medical history was significant for hypertension for which she had been taking a combination angiotensin receptor blocker-thiazide diuretic for 20 years; she was a lifetime non-smoker and had no other significant history of cardio-pulmonary disease. Her past surgical, family, and social history were otherwise non-contributory. On presentation, she was severely hypoxemic and hypotensive. Cardiac exam revealed a loud second heart sound, ventricular gallop (S3), parasternal heave, and tachycardia. Pulmonary exam was significant for increased work of breathing and bilateral rhonchi. Her extremities showed significant bilateral pitting edema up to her thighs. The remainder of her exam was unremarkable.

Four months prior to admission, the patient had a transthoracic echocardiogram suggestive of mild-moderate pulmonary hypertension with pulmonary arterial (PA) systolic pressure of approximately 40 mmHg as well as evidence of septal flattening. Subsequent left and right-heart catheterization revealed a PA pressure of 50/19 (mean 31) and non-obstructive coronary artery disease. Despite aggressive diuresis with furosemide and vasodilator therapy with sildenafil, the patient continued to have worsening dyspnea with associated palpitations.

Prior to admission to our hospital, she underwent an extensive pulmonary hypertension workup that revealed worsening PA systolic pressure of approximately 80 mmHg, increased septal flattening with D-shaped left ventricle, and moderate right ventricular dilatation. After diuresis, computed tomographic angiography (CTA) of the chest showed nonspecific mild reticular interstitial changes concerning for ILD with no evidence of pulmonary embolism. Pulmonary function tests were significant for a diffusing capacity of the lungs for carbon monoxide at 41% of the predicted value. Laboratory testing including thyroid function, HIV, and ANA was unremarkable.

At this point, given the concern for ILD as suggested by the patient's severe hypoxemia, CTA, and pulmonary function tests, a decision was made at the outside hospital to perform a bronchoscopy and video-assisted thoracoscopic lung biopsy, a course of action which we would not have recommended. The procedure was complicated by pulseless electrical activity arrest immediately after anesthesia induction. The patient underwent cardiopulmonary resuscitation for less than 5 minutes at which point return of spontaneous circulation was attained and the procedure was completed. Subsequently, the patient became hypotensive and required inotropic support with dopamine and milrinone. Preliminary lung biopsy results were consistent with idiopathic PAH (IPAH); biopsy slides were sent to a specialized outside laboratory (Fig. 1). The patient was then transferred to a nearby hospital for consultation and therapy. She had a Swan-Ganz catheter placed showing suprasystemic PA pressure of 104/46 (mean 67) mm/Hg; pulmonary capillary wedge pressure was 10 mmHg with an augmented cardiac output of 4.98 L/min. The patient was started on treatment for IPAH including vasodilation with inhaled nitric oxide, sildenafil, and epoprostenol. The patient had mild improvement in her PA pressure but remained hypoxemic and symptomatically dyspneic.

Two days after initiation of vasodilating agents, final review of

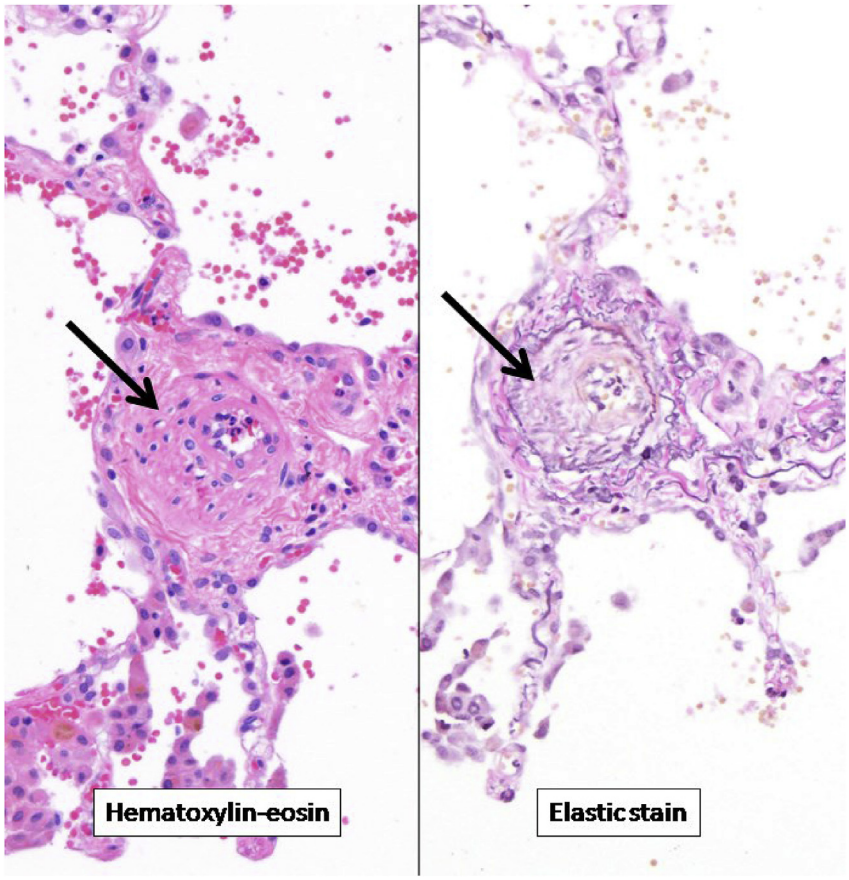

Fig. 1. Lung biopsy pathology slides. Subpleural vein in intralobular septum with luminal compromise by proliferating fibrous tissue (arrows).