INTRODUCTION: Anti-synthetase syndrome (ASS) is a heterogeneous connective tissue disease, characterized by an association with interstitial lung disease (ILD) and/or inflammatory myositis and the presence of anti-aminoacyl-tRNA-synthetase antibodies. Pulmonary hypertension (PH), a progressive pulmonary vascular disease, defined by elevation of pulmonary arterial pressure, has been reported with a prevalence of 7.9% in patients with ASS. However, ASS associated with PH due to Pulmonary veno-occlusive disease (PVOD) has never before been described in literature. We present a novel case of anti-synthetase syndrome associated with PVOD.

CASE PRESENTATION: 55 year old man with history of ILD presented with hypoxic respiratory failure and right heart failure. He was positive for ANA (1:160), and Anti-Jo (226 AU/mL) antibodies and hence was diagnosed with Anti-synthetase syndrome. Heart catheterization was done which showed, preserved LV function, and PA 100/45 mmHg with mean of 65, PAOP of 8 mmHg, PVR of 21 WU, and CO 2.7 L/min. Findings of elevated pressures out of proportion to his ILD, led to a diagnosis of pre-capillary PH and he was started on therapy with Milrinone, Macitentan and Sildenafil which he tolerated. Later in the hospitalization, IV Epoprostenol was added which resulted in acutely worsening hemodynamics and CT Chest remarkable for worsening pulmonary edema, pleural effusions and lymphadenopathy concerning for PVOD. He was deemed not a candidate for lung transplant. IV Epoprostanol was de-escalated, however at that time patient opted for comfort measures and expired. On autopsy, bilateral lungs revealed definitive evidence of PVOD, including vascular myxoid fibrosis, intimal hyperplasia, and venous occlusion in a background of pulmonary parenchyma with mature fibrosis. Changes suggestive of pulmonary arterial hypertension including arterial intimal thickening were also observed.

DISCUSSION: PVOD is a rare form of group 1 PH, that has never before been described in association with anti-synthetase syndrome. The pathologic hallmark of PVOD is the extensive and diffuse occlusion of pulmonary veins by fibrous tissue. A definite diagnosis of PVOD requires histologic analysis of a lung sample. Patients with PVOD may either be refractory to pulmonary arterial hypertension (PAH)-specific therapy or deteriorate with the start of therapy. It is difficult to clinically distinguish patients with PVOD from PAH prior to start of therapy. CT of the chest can suggest PVOD in the setting of PH when it shows nodular ground-glass opacities, thickened septal lines, and lymph node enlargement, however in patients who present with decompensated heart function such details are difficult to distinguish on imaging.

CONCLUSIONS: Anti-synthetase syndrome can be associated with PVOD, and it should be suspected in patients that have evidence of pre-capillary disease, and who deteriorate with initiation of PH therapies.

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