Refractory Pulmonary Hypertension in a Lupus Patient with Occult Pulmonary Vasculitis.

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Primary pulmonary hypertension is a disease that has become increasingly recognized in lupus patients. Pathologic findings from lupus patients usually do not differ from those who have idiopathic pulmonary hypertension. In recent years, intravenous vasodilator therapy has improved morbidity and mortality in patients with primary pulmonary hypertension. In this case report, we describe a young woman with severe pulmonary hypertension refractory to aggressive parenteral vasodilator treatment. Steroid treatment was initiated after a tentative diagnosis of lupus was made on the basis of the presence of Raynaud's phenomenon, proteinuria, pericarditis with tamponade, and a positive anti-Ro antibody. Despite treatment with vasodilators and steroids, the patient's condition rapidly deteriorated and she died. Unexpectedly, active pulmonary arteritis was demonstrated at autopsy. This case suggests that immunosuppressive therapy should be considered in lupus patients with severe pulmonary hypertension who are refractory to aggressive vasodilator therapy and high-dose parenteral steroids.

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