

## **Abstract**

Connective tissue disease-associated pulmonary hypertension is a heterogeneous, multifaceted disease. Pulmonary arterial hypertension, the most progressive form of all, is a potentially fatal complication, necessitating early detection and timely initiation of treatment to ensure better clinical outcomes. Despite recent advances, the progression shown in patients with connective tissue disease-associated pulmonary arterial hypertension treated with targeted therapy has been dismal. We describe a young female with mixed connective tissue disease presenting with worsening exertional dyspnoea and right heart failure, subsequently diagnosed with predominantly pulmonary arterial hypertension. The diagnostic and therapeutic dilemmas we encountered will be discussed together with a novel treatment approach in the form of immunosuppressive therapy with cyclophosphamide and glucocorticoids.