Introduction

Rapidly progressive-Interstitial Lung Disease (RP-ILD) is a subset of interstitial lung diseases characterized by a rapid deterioration in lung function over a short period, typically within three months. In patients with dermatomyositis (DM), anti-melanoma differentiation-associated gene 5 (anti-MDA5) has shown 40 to 70% association, with the development of RP-ILD. We present a unique case of anti-MDA5 antibody-associated RP-ILD without muscle or skin manifestation.

Case description

An 85-year-old man with type 2 diabetes and CKD stage 3B presented to emergency care with worsening exertional dyspnea, cough, and swelling over two weeks. Despite being physically active and healthy, his condition rapidly declined. He denied any history of tobacco abuse, or exposure to asbestos, silica, or beryllium. Physical exam revealed bilateral pitting edema but no classic dermatomyositis rashes. Tests showed evidence of interstitial pulmonary fibrosis with a UIP pattern, positive ANA and anti-MDA5 antibodies, confirming clinically amyopathic dermatomyositis (CADM-140). Treatment with steroids and nebulization was started, followed by inhaled corticosteroids upon discharge. Two months later, he returned with hypoxia, and imaging revealed bilateral pulmonary emboli and deep vein thrombosis. He was admitted and treated for acute hypoxic respiratory failure. Diagnosis of rapidly progressive MDA-5 positive amyopathic dermatomyositis-related interstitial lung disease (ILD) was made. He received five days of intravenous immunoglobulins (IVIG) and mycophenolate mofetil with pneumocystis pneumonia prophylaxis. However, despite aggressive treatment, the patient deteriorated over four weeks, requiring high-flow oxygen. Eventually, a decision for comfort care was made, and the patient passed away the next day.

Discussion

Anti-MDA5 antibody-related interstitial lung disease (ILD) usually links with a diagnosis of dermatomyositis accompanied by severe cutaneous manifestations. Limited reports discuss cases involving the lungs but lacking skin involvement, as seen in our patient. Literature review shows that not all Anti-MDA5 positive cases initially show classic dermatomyositis cutaneous manifestations, often developing about a month after respiratory symptoms. Our patient didn't survive beyond this timeframe. Despite its rarity, anti-MDA5 antibody-associated ILD should be suspected in cases of Rapidly Progressive-Interstitial Lung Disease (RP-ILD) even without other signs of DM or CADM as prompt and aggressive treatment could improve prognosis.