P_07 DOI: https://doi.org/10.5114/reum/219179

Unmasking the hidden cause of muscle weakness in a patient with previously amyopathic dermatomyositis: a diagnostic and therapeutic challenge

Izabella Ławińska[1], Wiktor Schmidt[2,3]

[1]Student's Research Group of Rheumatology, Systemic Connective Tissue Diseases and Immunotherapy in Rheumatic Diseases, Poznan University of Medical Sciences, Poland

[2]Department of Internal Diseases and Metabolic Disorders, Poznan University of Medical Sciences, Poland

3Department of Rheumatology, Systemic Connective Tissue Diseases and Immunotherapy of Rheumatic Diseases at Józef Struś Hospital in Poznan, Poland

Key words: dermatomyositis, muscle weakness, nonspecific interstitial pneumonia

Introduction: Dermatomyositis (DM) is a systemic autoimmune disorder characterised by symmetrical muscle weakness and pathognomic skin lesions. Interstitial lung disease frequently coexists. Notably, in up to 20% of patients, DM can present as an amyopathic form, in which characteristic skin findings occur in the absence of clinical or laboratory evidence of muscle involvement.

Case description: A 62-year-old male was admitted to the Rheumatology Ward with decreasing exercise tolerance and cough. Dermatological findings included mechanic's hands, Gottron's papules over joints, and hyperkeratosis with fissuring of the fingertips. High-resolution computed tomography (HRCT) revealed fixed interstitial lung changes consistent with fibrosing nonspecific interstitial pneumonia. Pulmonary function tests showed significant impairment of the transfer factor of the lung for carbon monoxide (TLCO), measuring 48% of the expected value. Laboratory tests showed mildly elevated aldolase and C-reactive protein, with positive antinuclear antibodies (anti-MDA5, anti-PM-Scl-100, anti-PM-Scl-75, anti-Ro-52, anti-SS-B). A diagnosis of clinically amyopathic dermatomyositis was established. The patient received intravenous methylprednisolone and cyclophosphamide (6 monthly 1,000 mg pulses). On assessment after completing the cycle, he developed new-onset, rapidly progressing proximal muscle weakness. Cervical spine magnetic resonance imaging revealed severe C3–C4 stenosis with myelopathy and oedema of the nucleus pulposus. Pulmonary changes on HRCT improved, and TLCO function was 63% of the expected value. The patient subsequently underwent a laminectomy, followed by rehabilitation and continuation of his treatment. Methotrexate and nintedanib were initiated, and at a 1.5-year follow-up, the patient demonstrated further pulmonary and functional improvement (TLCO: 81% of the expected value). The patient was enrolled in a drug program and started on, which stabilised the disease course.

Conclusions: Rheumatological conditions can exhibit atypical or nonspecific presentations, often complicating timely and accurate diagnosis. In the described case, a comprehensive diagnostic evaluation enabled the implementation of targeted therapy, resulting in disease remission.