Abstract

Background: Dermatomyositis (DM) is an idiopathic inflammatory myopathy classically presents with characteristic cutaneous manifestations and proximal myositis. Clinically amyopathic dermatomyositis (CADM) has minimal or no evidence of myositis.
Case report: 43-year-old female with diabetes and hypertension presented with a violaceous heliotrope rash and progressive shortness of breath on exertion with significant desaturation at rest. She did not have clinical evidence of a proximal myopathy. Clinical evaluation revealed evidence of an organizing pneumonia with positive Ro-52 and Mi-2 antibodies. Patient was diagnosed with clinically amyopathic dermatomyositis complicated with interstitial lung disease. She was commenced on intravenous (IV) methylprednisolone pulses and IV cyclophosphamide pulse therapy with a remarkable improvement. Patient's malignancy screening was negative.
Conclusion: Rapidly progressive ILD is a frequent manifestation of CADM and is associated with poor outcomes. Anti MDA5 antibody is highly specific for CADM and is a predictor of occurrence of ILD. It is prudent to aggressively treat CADM patients with ILD using a combination of immunomodulators with different modes of actions. High dose corticosteroids combined with ciclosporin, or cyclophosphamide are established treatment options