Diagnosis and Treatment of Dermatomyositis-Systemic Lupus Erythematosus Overlap Syndrome

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Introduction
Dermatomyositis is an autoimmune condition classically characterized by symmetric proximal muscle weakness, inflammatory muscle changes, and dermatologic abnormalities.1 Several studies have shown that the inflammatory myopathies, such as dermatomyositis, commonly overlap with other connective tissue disorders, significantly complicating the diagnosis.2 The reported incidence of overlap syndrome ranges from 11% to 40% in patients diagnosed with dermatomyositis. Overlap syndromes in dermatomyositis are most common with connective tissue diseases such as Sjögren syndrome, scleroderma, rheumatoid arthritis, systemic lupus erythematosus, and mixed connective tissue disease. A disorder can only be considered an overlap syndrome if the patient meets the separate diagnostic criteria for both conditions, however, overlap syndromes create diagnostic difficulty because the patient often presents progressively with variable symptoms from each disorder making it hard to establish a definitive disease process.3

Case Description
A 39-year-old woman with known non-alcoholic fatty liver disease, iron deficiency anemia, and peripheral venous insufficiency presented for the evaluation of a facial rash, arthralgias, and worsening lower extremity edema. The arthralgias and edema had been present intermittently for one year but worsened over the past month. She reported no history of a rash and had already tried some over the counter moisturizers without relief. Physical exam revealed 1+ bilateral edema to the mid-calves and an erythematous, maculopapular rash across the nose, forehead, and behind the ears. The rash was initially treated with hydrocortisone 2.5% and then ketoconazole 2% with no improvement and her peripheral edema persisted despite the use of compression stockings and 20 mg of daily furosemide. The patient followed up in the clinic 3 months later with worsening lower extremity edema and facial rash, along with new, tender, subcutaneous nodules in the abdomen, axillae, and groin. Initial work-up showed a c-reactive protein of 2.2, erythrocyte sedimentation rate of 87, a positive anti-nuclear antibody 1:1,280 in a speckled pattern, and microcytic anemia.

This case of overlap syndrome between dermatomyositis and systemic lupus erythematosus presents a rare but important challenge to the primary care physician. Our patient presented initially with arthralgias and fatigue, symptoms more characteristic of systemic lupus erythematosus. However, these symptoms were followed by a facial rash that involved the nasolabial folds and periorbital regions more in line with dermatomyositis. Interestingly, despite the prominence of the characteristic proximal muscle weakness, a rash is the most common presenting feature of dermatomyositis.4 This patient’s initial labs were also not significantly concerning on presentation as her muscle enzymes didn’t show marked elevation until the patient had already developed significant disease. The atypical mix of symptoms with relatively benign laboratory findings exemplifies the difficulty in identifying overlap syndromes of classic rheumatologic conditions. Systemic glucocorticoids are the mainstay of initial treatment for dermatomyositis and are often used in the treatment of SLE as well, suggesting the combination may be effective in dermatomyositis- SLE overlap.5 However, prolonged use of glucocorticoids can result in steroid-induced myopathy, which can mimic the weakness of dermatomyositis as in the case of our patient. Ultimately, the switch to tofacitinib improved the patient’s weakness and decreased the glucocorticoid dose needed.6 In our patient the combination of tofacitinib, hydroxychloroquine, and low dose prednisone has been the most effective in inducing symptom remission in dermatomyositis and lupus overlap.

References

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