A "bumpy" rash in a patient with seronegative rheumatoid arthritis

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Ackerman syndrome is a rare form of arthritis with interstitial granulomatous dermatitis that can be associated with connective tissue disease. It is associated with a pathognomonic rash but certain cases can be difficult to treat.

A 71-year-old male with past medical history of seronegative rheumatoid arthritis (RA) hypertension, and coronary artery disease presented to the rheumatology clinic to establish care. He had been diagnosed with seronegative RA by an outside rheumatologist four years prior given his history of morning stiffness, pain, and swelling in his bilateral MCPs and PIPs; he was trialed on leflunomide and prednisone at that time. After weaning off prednisone, he was doing well on leflunomide monotherapy. Three years later, he began having recurrent symptoms. He was evaluated by a new rheumatologist and was switched to methotrexate and prednisone.

At our initial visit, the patient was taking methotrexate and low-dose prednisone. To help him wean off prednisone, he was started on leflunomide. Three months later, he continued to have morning stiffness. He was started on colchicine given concerns for a potential autoinflammatory syndrome, with significant improvement in his joint pain.

Prior to his next appointment, he had developed a bumpy rash on his bilateral hands. He was taking leflunomide and colchicine; he had stopped methotrexate. The next month, he saw dermatology; his biopsy was consistent with necrobiosis granulomatous inflammation. He was started on prednisone and dapsone for his skin findings. Three months later, his rash persisted, but his arthritis was well-controlled.

At this time, his case was thoroughly reviewed and was thought to be most consistent with Ackerman syndrome, interstitial granulomatous dermatitis with arthritis (IGDA). His symptoms began to resolve with continuation of leflunomide, colchicine, dapsone, clobetasol cream, and a slow prednisone taper.

This case illustrates a patient with Ackerman syndrome who achieved resolution of symptoms on combination therapy. Utilization of leflunomide, colchicine, dapsone, and topical steroids was a successful way to reduce oral steroid usage. These agents may be applied to other difficult-to-treat cases of this rare form of arthritis with dermatitis.