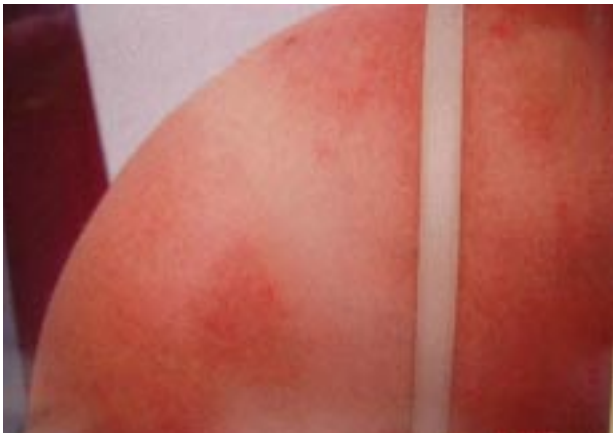
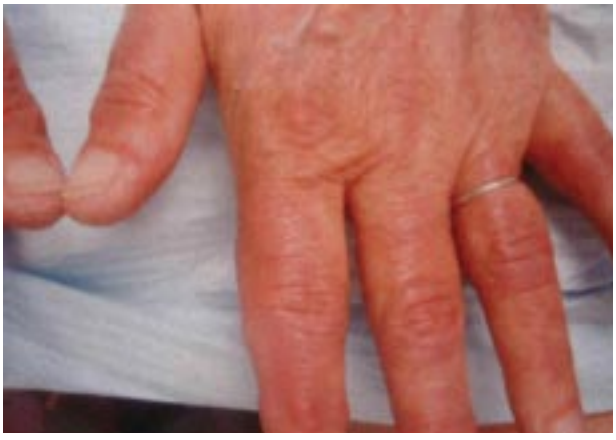
What Is Your Diagnosis?





A 55-year-old woman presented with an increasingly intense erythematous eruption involving the *V* of the chest, shoulders, dorsal fingers, and upper back of 5 months' duration. She reported pain on the soles of her feet and shoulders as well as joint aches and stiffness in her hands and wrists.

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The authors report no conflict of interest.

The Diagnosis: Multicentric Reticulohistiocytosis With Dermatomyositislike Features

Physical examination of the patient revealed erythematous papules on the upper back with sparing of the mid and lower back as well as under the bra strap, chest, and abdomen. There was prominent erythema of the V of the chest, shoulders, and upper arms (Figure 1). Multiple pink papules were noted on the distal dorsal fingers (Figure 2). A few fine pink papules were noted along the eyebrows and hairline. Dermographism and rare, faint pink papules were noted on the posterior scalp and conchae as well as tiny pink papules on the lower mucosal surface of the lip.

A biopsy specimen from the neck revealed a superficial and deep perivascular lymphocytic infiltrate as well as numerous cells with ground glass cytoplasm. An S-100 stain failed to decorate the cells, but a CD68 stain was positive, indicating that the cells were histiocytes (Figure 3). Multinucleated histiocytes were present and abnormal mitotic figures were not identified (Figure 4). A colloidal iron stain failed to reveal an increased amount of dermal mucin, and an elastic stain revealed elastic fibers within some of the histiocytic cells.

Extensive workup to date is negative for malignancy. Her skin lesions, pruritus, and joint pains have substantially improved with methotrexate sodium at a dosage of 20 mg weekly, hydroxychloroquine sulfate 200 mg twice daily, hydroxyzine hydrochloride 25 mg daily at bedtime, fexofenadine hydrochloride 180 mg daily, and intermittent use of clobetasol propionate cream 0.05% and tacrolimus ointment 0.1%.

Multicentric reticulohistiocytosis is a rare type of non–Langerhans cell histiocytosis that may involve multiple organ systems. Women are affected approximately twice as often as men, and affected individuals usually are middle aged. Papules and nodules on the hands often are associated with severe polyarthritis.[1] Biopsy of cutaneous lesions reveals histiocytes. Some histiocytes coalesce to produce multinucleated giant cells with ground glass cytoplasm. Multicentric reticulohistiocytosis may occur in association with an underlying malignancy in approximately 25% of cases[2] and has been noted to occur as a paraneoplastic phenomenon.[3-5] A few rare reports indicate that multicentric reticulohistiocytosis can present with a dermatomyositislike clinical picture.[2,6,7] Multicentric reticulohistiocytosis usually presents with the gradual development of arthritis in the majority of patients.[2] Mucocutaneous lesions comprised of erythematous papules, plaques, and nodules gradually develop. Arthritis and skin lesions may go through periods of worsening and remission but eventually lead to disfigurement and mutilating arthritis if untreated. Presentation with erythematous patches that appear to be photodistributed is a rare phenomenon. Biopsy of multicentric reticulohistiocytosis shows characteristic giant cells, while dermatomyositis shows increased mucin, thickening of the basement membrane zone, and hydropic vacuolization of the basal layer of the epidermis. Dermatomyositis responds well to corticosteroid and immunosuppressive therapy, but multicentric reticulohistiocytosis has not been found to have a consistently

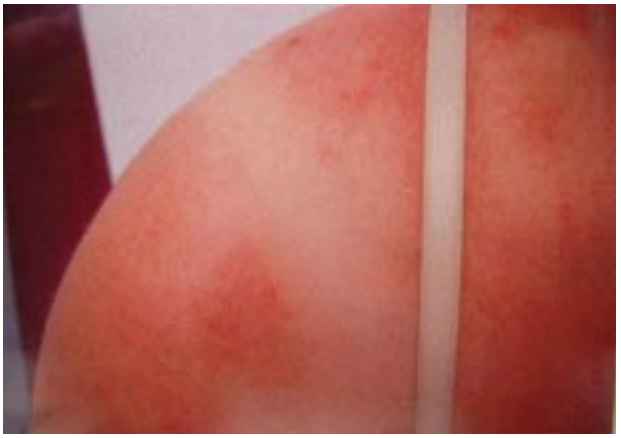

Figure 1. Erythema on the shoulder that is photodistributed.

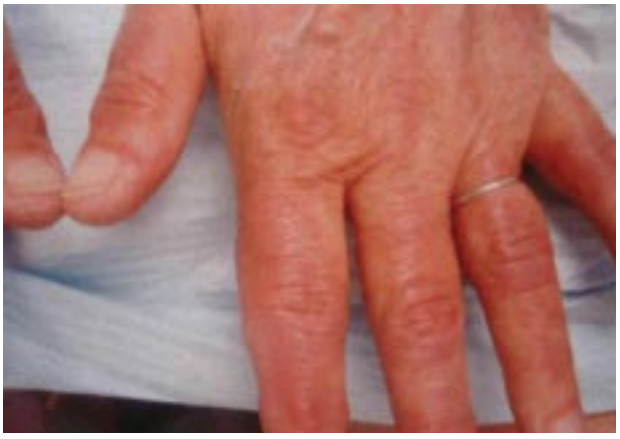

Figure 2. Erythematous papules on the distal dorsal fingers.

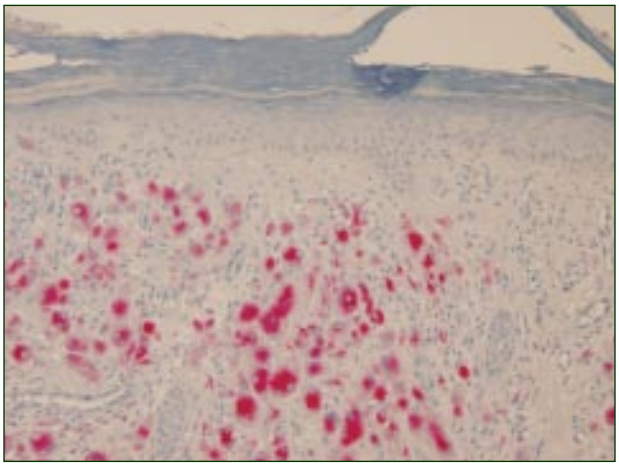

Figure 3. A nodular infiltrate of lymphocytes and histiocytes. CD68 stain highlights numerous giant cells (original magnification ×20).

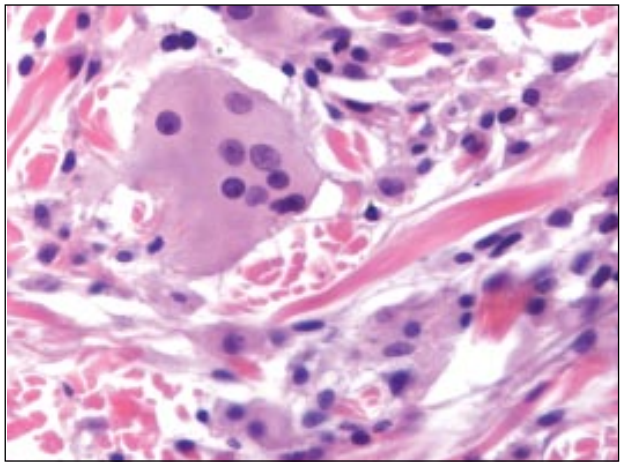

Figure 4. Multiple histiocytes with ground glass cytoplasm (H&E, original magnification ×40).

reliable effective therapy. Some reports suggest that anti–tumor necrosis factor α therapy is of value,[8,9] but a report indicates that this therapy may help the cutaneous lesions more than the arthritis.[10]

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