Multicentric reticulohistiocytosis (MCRH) is a rare dermatoarthropathy of unknown etiology characterized by infiltration of predominantly skin and synovial tissue with histiocytes and multinucleated giant cells exhibiting a microscopic eosinophilic, ground-glass appearance. MCRH presents in 60% of cases with an erosive, symmetric polyarthritis, which can easily be mistaken for a seronegative or early inflammatory arthritis such as rheumatoid or psoriatic arthritis based on predominant interphalangeal synovitis. Cutaneous findings are often described as having a distinctive “coral-bead” appearance with grouped, firm, reddish papules, typically overlying the dorsal aspect of the fingers, nailfolds, and hands. Lesions occasionally occur on the trunk and hands. We describe a rare case of an elderly woman who presented with initial clinical features of prominent proximal muscle weakness, which was subsequently accompanied by a symmetric polyarthritis and evolving cutaneous features mimicking classical dermatomyositis (DM) (Figure 1). The histologic findings on skin biopsy of several representative skin lesions were diagnostic for MCRH. Her skin and joint disease responded quickly to treatment with methotrexate and low dose prednisone. There have been documented clinical responses to similar therapies. There are limited reports describing in similar detail MCRH cases that were clinically suspected to initially have DM. All previously reported cases of MCRH confused with DM were based on isolated dermatologic features of amyopathic DM in which there was no associated muscle weakness or myalgias. Our case further emphasizes the importance for the consideration of MCRH among other common inflammatory dermatoarthropathies, such as DM and systemic lupus erythematosus, which may present with a similar constellation of clinical features. Skin biopsies of lesions should be obtained early in the evaluation since the histological findings are often diagnostic.

Figure 1. A. Coalescing erythematous papules and nodules distributed in a V-shaped pattern over the upper chest and neck in a photodistributed skin eruption. B. Dozens of asymptomatic, brawny-colored, waxy papules over the dorsum of the hands and nailfolds, and (C) peri-orificial regions. D. Extensive dermal infiltrate with homogenous-appearing mononuclear cells and multinucleated giant cells with an eosinophilic “ground-glass” cytoplasm, sparing the epidermis.
REFERENCES


