Multicentric Reticulohistiocytosis Can Mimic Rheumatoid Arthritis

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Multicentric reticulohistiocytosis (MRH) is a rare non-Langerhans histiocytosis^{1,2}. It is sometimes mistaken for rheumatoid arthritis (RA).

A 56-year-old woman with a history of diabetes mellitus type 1 presented 24 years earlier with symptoms consistent with inflammatory arthritis. She was initially diagnosed with RA and treated with hydroxychloroquine, with good response. This was ultimately discontinued. Following discontinuation, she developed worsening joint symptoms as well as a pruritic widespread nodular skin eruption (Figure 1). A skin biopsy was performed, which demonstrated mononuclear and multinucleated dermal histiocytic infiltrate with ground glass cytoplasmic inclusions (Figure 1). Radiographs of the hands showed erosive changes in

proximal interphalangeal joints, distal interphalangeal (DIP) joints, and wrist joints (Figure 2). Based on these findings, including DIP involvement and skin findings, the patient was diagnosed with MRH. She was restarted on hydroxy-chloroquine and methotrexate. A trial of adalimumab did not affect her symptoms and she was then switched to infliximab. Alendronate was also added. With this regimen the patient has experienced improvement in her skin and joint symptoms. For evaluation of potential associated malignancy, she underwent positron emission tomography imaging, which was negative.

MRH is a rare non-Langerhans histiocytosis with about 250 cases in the literature^{1,2}. Destructive arthritis can affect any joint, but it typically affects the interphalangeal joints.

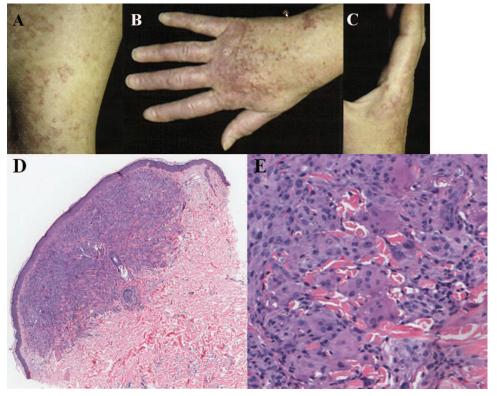


Figure 1. Skin findings included clusters of reddish-brown papules (A, elbow; B, hand; C, hand). Microscopic findings included a dense dermal aggregate of mononuclear and multinucleated histiocytes with ground-glass cytoplasm (H&E stain; D, original magnification × 4; E, original magnification × 40).

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Figure 2. Hand radiographs demonstrate erosions at proximal interphalangeal joints, distal interphalangeal (DIP) joints, and wrist joints. Insert shows marginal erosions of right second DIP.

Skin involvement includes variable-sized reddish-brown papules and nodules with classic findings surrounding the nostrils and around nailfolds termed "coral beads." Diagnosis is confirmed by biopsy, which demonstrates the presence of multinucleated histiocytes with distinctive ground-glass cytoplasm. The underlying etiology is unclear but is often associated with malignancy and autoimmune conditions³. Because of its rarity, no trials have been performed to test efficacy of treatment, but frequently methotrexate, hydroxychloroquine, and tumor necrosis factor inhibitors are used¹. More recent case reports demonstrate efficacy of bisphosphonates, which may be effective because of the osteoclast-like properties of the ground-glass histiocytes^{4,5}. MRH should be considered in patients who present with skin nodules and destructive arthritis. Erosive disease at the DIP joints can help to distinguish MRH from RA.

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