

# Multicentric Reticulohistiocytosis Associated With Burkitt Lymphoma and Adenocarcinoma

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*Multicentric reticulohistiocytosis (MRH) is a rare disease of unknown etiology characterized by cutaneous nodules and destructive, sometimes crippling, polyarthritis. The diagnosis is confirmed by histopathologic features of the cutaneous nodules or synovial tissue, including an infiltrate composed of histiocytes, many of them multinucleate, with a ground glass appearance. Multicentric reticulohistiocytosis has been associated with a number of chronic conditions and various malignancies. We report a case of MRH in a patient with Burkitt lymphoma and metastatic adenocarcinoma of the gastrointestinal tract.*

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## Case Report

A 31-year-old man presented with a papular eruption over his bilateral upper extremities. He also complained of weakness of 4 months' duration and chronic joint pain. His medical history included Burkitt lymphoma diagnosed at 7 years of age in his abdomen, brain, and spine, which eventually resulted in paraplegia. He was treated with chemotherapy and radiation. Four months prior, he also was diagnosed with adenocarcinoma of the gastrointestinal tract discovered in a hilar lymph node.

On physical examination, numerous 2.0- to 2.5-mm smooth, dome-shaped to flattopped shiny papules were noted on the bilateral upper extremities. The lesions were most prominent along the

ulnar aspect of the forearms (Figure 1), with slight extension onto the upper arms. Similar papules were noted around the fingernails in the paronychia folds in a coral-beading pattern.

The biopsy specimen showed a dome-shaped lesion with an effaced epidermis and a dermal proliferation abutting the epidermis. The dermal nodule was composed of large histiocytes, many of them multinucleate, with abundant eosinophilic cytoplasm with a ground glass appearance and a lymphocytic infiltrate (Figure 2).

Laboratory tests revealed Epstein-Barr virus viremia and an elevated erythrocyte sedimentation rate as well as the presence of antineutrophil cytoplasmic antibodies and a positive reaction for antinuclear antibody.

## Comment

Multicentric reticulohistiocytosis (MRH) is a rare granulomatous disease that appears to result from a reactive inflammatory response to an undetermined stimulus. It is associated with a number of autoimmune diseases with systemic effects, and there is evidence of tuberculosis exposure in up to 50% of cases.<sup>1,2</sup> An elevated erythrocyte sedimentation rate is a common finding in MRH; however, MRH is not known to be associated with Epstein-Barr virus viremia or the presence of antineutrophil cytoplasmic antibodies and antinuclear antibody.<sup>3</sup>

The medical literature has revealed association of MRH with several different malignancies. Multicentric reticulohistiocytosis has been associated with malignancy in approximately 25% of reported cases.<sup>3,4</sup> Some researchers and physicians argue that MRH is in fact a paraneoplastic syndrome.

The most common malignancies noted in patients with MRH are breast cancer, gastric carcinoma,

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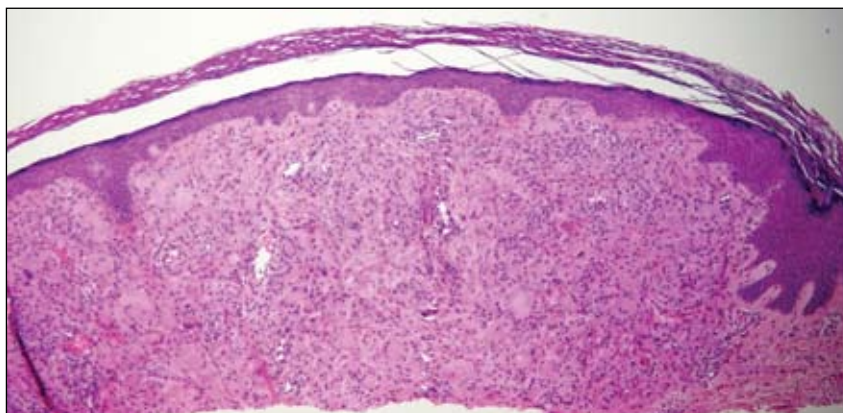
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**Figure 1.** Ulnar aspect of the patient's right forearm demonstrating numerous erythematous papules.



**Figure 2.** On histologic examination, the lesion demonstrated an abundance of mononuclear histiocytes with eosinophilic cytoplasm as well as several multinucleate giant cells and a lymphocytic infiltrate (H&E, original magnification  $\times 4$ ).

leukemia, lymphoma, melanoma, mesothelioma, and sarcoma.<sup>1,3,4</sup> Furthermore, there appears to be a temporal relationship between the two, with the onset of MRH and malignancy occurring within 2 years of each other in most cases.<sup>4</sup> In some reported cases, the initial workup of MRH led to the discovery of an underlying malignancy.<sup>4,7</sup> However, MRH does not always run a parallel course with the malignancy, leading some researchers or physicians to question the strength of the relationship. While it is not certain that there is a true association of MRH and malignancy, our case and several other case reports serve to provide more evidence of the potential paraneoplastic nature of MRH.<sup>4,7</sup>

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