Volume 19 Number 11
November 2013

Case Presentation

Granuloma annulare mimicking multicentric reticulohistiocytosis

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Dermatology Online Journal 19 (11): 6

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ABSTRACT

We describe a patient with granuloma annulare (GA) who presented with firm periungual papules mimicking “coral beads”, a characteristic sign of multicentric reticulohistiocytosis (MRH). We highlight the importance of distinguishing between GA and MRH because the prognoses differ significantly.

Case synopsis

A 72-year-old woman presented with asymptomatic diffuse erythematous patches on her trunk and extremities and firm papules on her fingers. She also noted mild joint pain. Her past medical history was significant for a benign brain tumor 16 years prior to presentation. Her medications at presentation included levetiracetam, aspirin, vitamin D, and calcium. Skin examination revealed multiple firm 5-mm skin-colored to erythematous papules on her distal fingers (Figure 1) and erythematous patches on her trunk (Figure 2) and extremities. The digital papules were arranged along the nail folds on several digits bilaterally (Figure 1). There was no conjunctival or oral mucosal involvement.

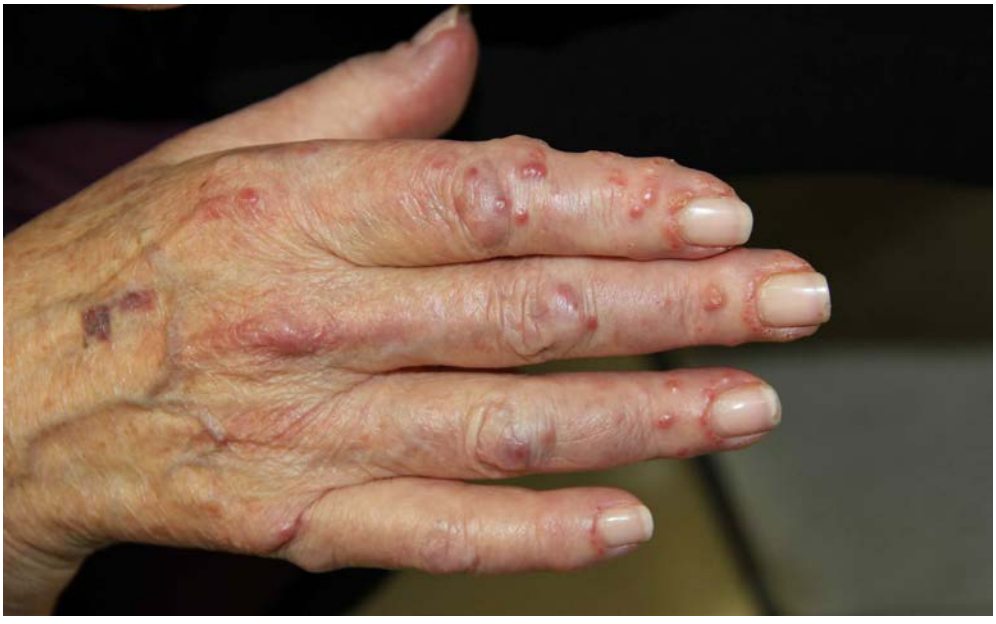

Figure 1. Multiple firm papules on dorsal hand with periungual papules arranged in a coral bead appearance

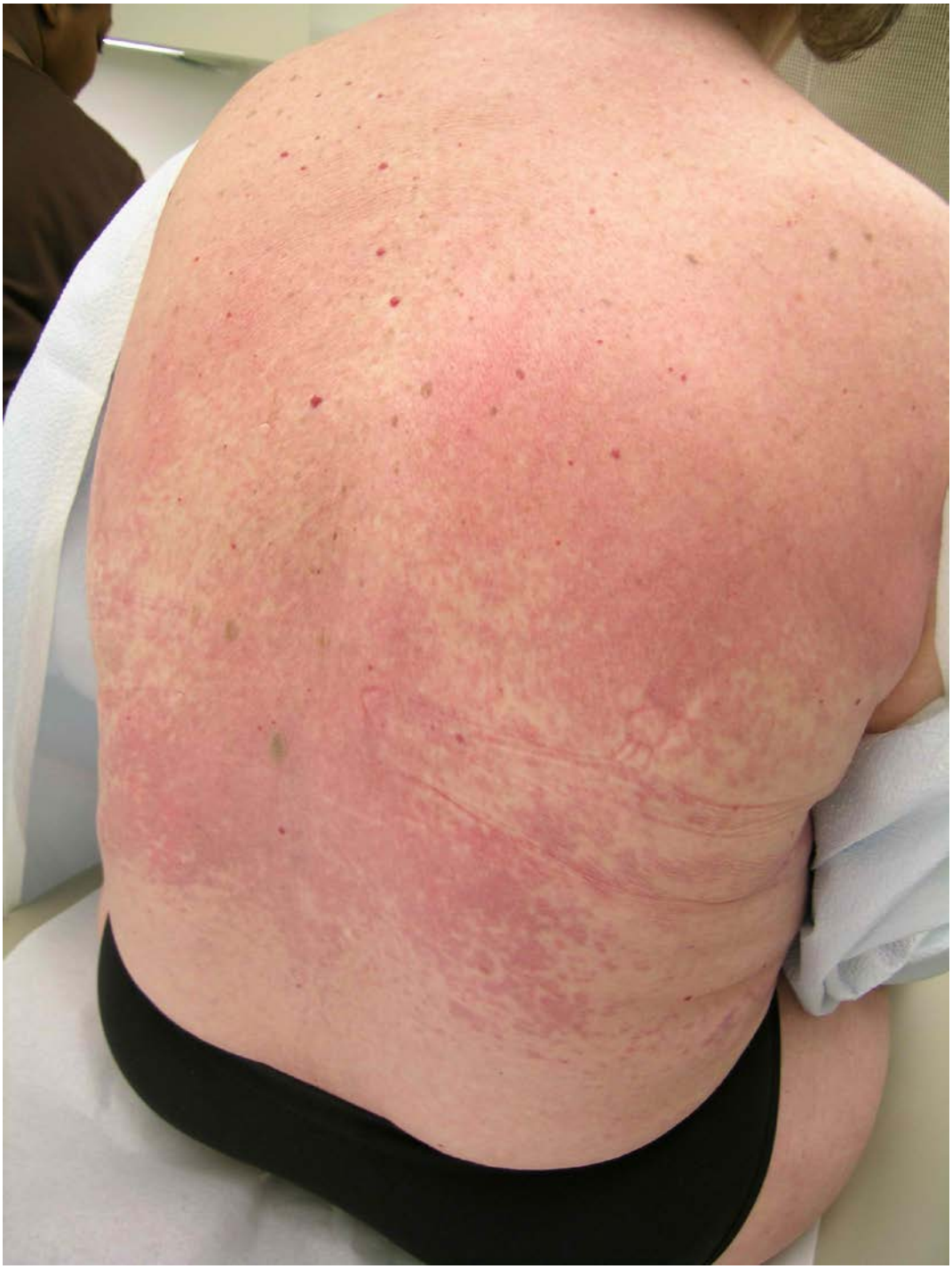

Figure 2. Erythematous patches on the trunk

Histopathology

Histopathologic features of a 4-mm punch biopsy from a chest lesion revealed changes consistent with granuloma annulare (GA) with zones of palisaded histiocytes and giant cells with necrobiosis and increased mucin. There were scattered lymphocytes. Another 4-mm punch biopsy of the thigh also revealed GA with discrete areas of palisading histiocytes surrounding collections of mucin with perivascular lymphocytes. Microscopic examination of a papule from the dorsum of the right second digit over the proximal interphalangeal joint revealed histologic changes consistent with GA/elastolytic granuloma, with abundant epithelioid histiocytes and giant cells with vague palisading in foci. (Figure 3). There were more numerous giant cells than are usually seen. Focal elastophagocytosis was also present.

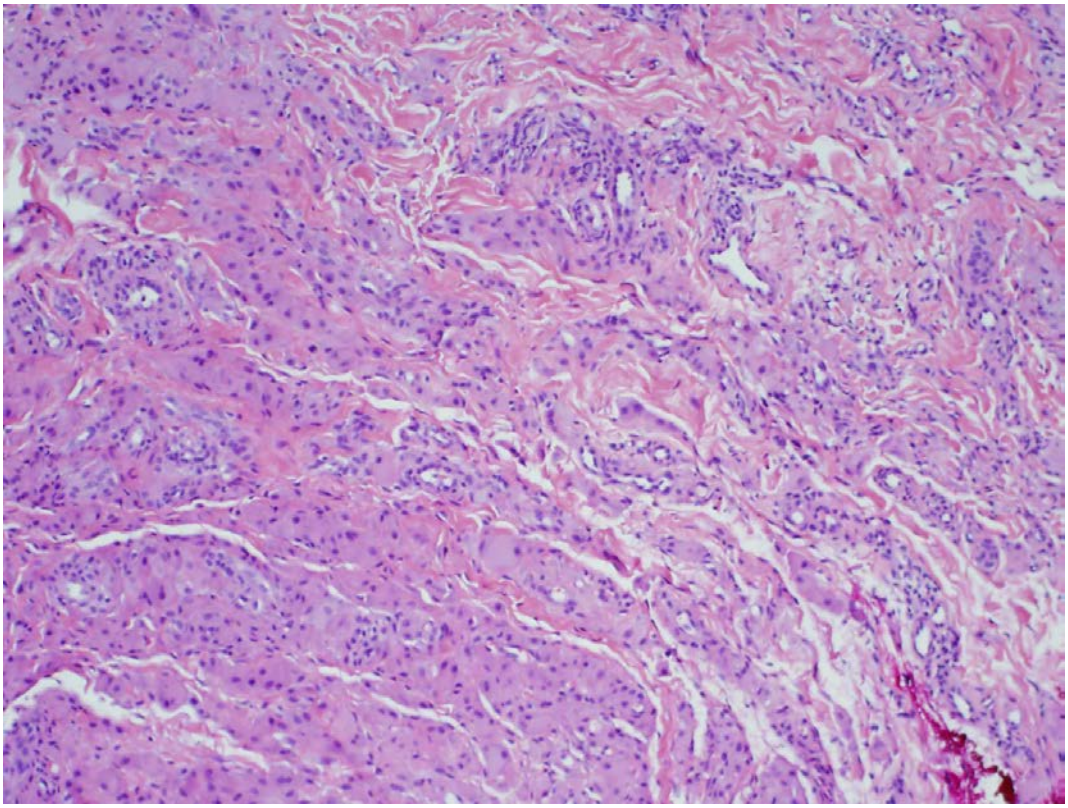

Figure 3. Abundant epithelioid histiocytes and giant cells with vague palisading in foci (H&E 100X)

Course of treatment

At the initial visit, the patient received intralesional triamcinolone (10 mg/ml) to several papules on the fingers and the papules cleared within several weeks. The intralesional triamcinolone was repeated 2 months later for additional lesions. One year later, the GA plaques on her trunk and extremities had resolved, although she continued to have the papules on her fingers. The patient elected not to have further treatment of the papules on her fingers because the lesions had recurred.

DISCUSSION

GA is a relatively common benign self-limited disorder of the dermis and subcutaneous tissues. Typical lesions of GA include a localized ring of beaded papules coalescing into arciform and annular plaques on the extremities [1].

Our patient presented with multiple, firm, round, pinkish-brown to purple, generally isolated papules and nodules scattered over her dorsal hands and digits. These were aligned along the nail folds (Figure 1), resulting in a characteristic “coral bead” appearance, which is classic for multicentric reticulohistiocytosis (MRH) and present in up to 40% of MRH cases.

MRH is a rare disorder of the non-Langerhans cell histiocytosis family that often systemically involves the skin, mucous membranes, and joints. Cutaneous lesions are 2 mm to 2 cm in diameter and are red, brown, yellow, or skin-colored; mucous membrane involvement occurs in 50%. Joint disease is a prominent feature of MRH in which symmetric, destructive polyarthritis can progress to arthritis mutilans in 45% of patients. Joint symptoms typically present earlier than or concomitantly with the cutaneous eruptions of MRH [2]. The disease can spontaneously remit in five to ten years, although patients are left with significant disability [2].

Although our patient also presented with joint pain, over the next year and half she did not experience the progressive arthritis characteristic of MRH. Presentation of joint pain along with papules along the nail folds may suggest MRH in the differential diagnosis. Our patient presented with clinical lesions of GA, which were clinically suspicious for MRH. Orkwis *et al* presented a patient with a similar appearance to our patient who had a 6-month history of annular plaques on her dorsal hands and palms with an underlying history of arthritis in her hands and feet [3]. Biopsy revealed a dermal lymphocytic infiltrate with prominent multinucleated giant cells containing abundant eosinophilic cytoplasm and a “ground glass” appearance, findings consistent with MRH [3]. Multiple biopsies of our patient, on the other hand, revealed palisading histiocytes surrounding collections of mucin

with perivascular lymphocytes and necrobiosis, classic histopathological features of GA. Given the range of possible clinical presentations, these cases illustrate the critical importance of histopathology for proper diagnosis.

Distinguishing between GA and MRH is important because prognoses differ greatly. GA is a common benign disease that usually resolves with or without treatment. In contrast, MRH is associated with destructive arthropathy that leads to permanent disability in a significant percentage of patients. Furthermore, MRH correlates significantly with other systemic diseases, including internal malignancy in 25% of patients, hyperlipidemia in 30-50% of patients, and autoimmune disease in 5-15% of patients.

We present a 72-year-old patient with an atypical presentation of GA consisting of nodules with a morphology and distribution similar to that of MRH. Although the clinical presentation was atypical, histopathologic analysis demonstrating palisading granuloma with necrobiosis and mucin confirmed the diagnosis of GA.

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