

Introduction

We describe a patient with multicentric reticulohistiocytosis (MRH), a rare inflammatory disorder with distinct physical examination and histopathologic findings. This case highlights important characteristics that differentiate this disorder from other inflammatory diseases that present similarly with rash, hand lesions, and joint involvement and describes appropriate management of the condition.

Case Description

A 79-year-old Hispanic woman presented with a 1-year history of pruritic papules and plaques on her chest, back, arms, hands, legs, and feet. The patient reported a history of hypothyroidism, arthritis, and vitiligo but denied a history of cancer. Physical examination showed pink papules coalescing into plaques on her upper chest and lower back (Figure 1) as well as lichenified plaques on her bilateral forearms and knees. Erythematous papules with a “coral bead appearance” on the proximal nail folds of her right first and second digits were noted (Figure 2). Multiple depigmented patches on her hands, wrists, arms, and lower back were also present, and deformities of bilateral hands and bulbous-appearing knees were observed. Results from a complete blood cell count and blood chemistry analyses showed mild anemia but were otherwise normal. A radiograph of her right knee showed degenerative changes and periarticular radiolucencies consistent with an inflammatory arthropathy. A 4-mm punch biopsy specimen from her back was obtained. Histopathologic examination revealed a dense infiltrate of epithelioid histiocytes with amphophilic “ground glass” cytoplasm in a nodular configuration (Figure 3). This pattern, in conjunction with the clinical features seen in this patient, was consistent with a diagnosis of multicentric reticulohistiocytosis. The cutaneous symptoms of the patient in the present case were managed with triamcinolone 0.1% ointment twice daily and oral hydroxyzine three times daily as needed for itching with moderate improvement. She was referred to Rheumatology for management of her arthritis, and initial cancer screening was negative.



Figure 1. Pink papules coalescing into plaques on the lower back.



Figure 2. Erythematous papules on the proximal nail folds of her right first and second digits

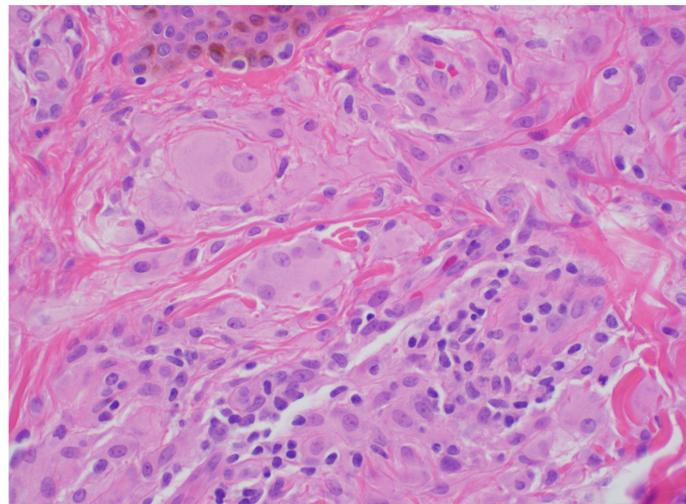


Figure 3. Lesional histopathology showing dermal histiocytic infiltration with multinucleated giant cells containing two-toned, “ground-glass” cytoplasm and prominent nucleoli (H&E, original magnification X200).

Discussion

Multicentric reticulohistiocytosis is a rare granulomatous disease characterized by papulonodular cutaneous lesions and severe erosive arthritis. It typically presents as rounded pruritic papules or nodules that may be pink, red, or brown affecting the face and distal upper extremities. A hallmark feature of MRH is the presence of multiple shiny erythematous papules along the proximal and lateral nail folds with a “coral bead” appearance. Patients with MRH often have mucosal involvement, destructive arthritis, and autoimmune conditions. Of note, MRH also has an association with underlying malignancy in up to 25% of patients and may even precede the diagnosis of cancer. Thus, it is important to distinguish MRH from other autoimmune disorders with similar cutaneous and physical examination findings, such as erythema elevatum diutinum, dermatomyositis, rheumatoid arthritis, and sarcoidosis. Skin biopsy must be performed to establish the diagnosis of MRH. In general, patients with MRH and no underlying malignancy have a good prognosis and respond to anti-inflammatory therapies such as NSAIDs and corticosteroids but may require methotrexate, cyclophosphamide, and TNF- α inhibitors in more severe cases.

Conclusions

This case highlights a key physical examination finding unique to MRH and discusses histopathologic findings that distinguish it from other inflammatory diseases that present with rash, hand lesions, and arthritis. Recognition of this disease is important so that patients can receive adequate treatment and surveillance for underlying malignancy.

References

The presented case is currently In Press at *Cutis*:

Kasitinon SY, Day JA, Vandergriff TW, and Gill JG. Periungual Papules in an Elderly Woman. *Cutis* 2020.

1. Tajirian AL, Malik MK, Robinson-Bostom L, Lally EV. Multicentric reticulohistiocytosis. *Clin Dermatol*. 2006;24(6):486-92.
2. Gold RH, Metzger AL, Mirra JM, Weinberger HJ, Killebrew K. Multicentric reticulohistiocytosis (lipoid dermatitis). An erosive polyarthritis with distinctive clinical, roentgenographic and pathologic features. *Am J Roentgenol Radium Ther Nucl Med*. 1975;124(4):610-624.
3. Luz FB, Gaspar TAP, Kalil-Gaspar N, Ramos-e-Silva M. Multicentric reticulohistiocytosis. *J Eur Acad Dermatol Venereol*. 2001;15(6):524-531.
4. Barrow MV. The nails in multicentric reticulohistiocytosis. (Lipoid dermatitis). *Arch Dermatol*. 1967;95(2):200-01.
5. Barrow MV, Holubar K. Multicentric reticulohistiocytosis. A review of 33 patients. *Medicine (Baltimore)*. 1969;48(4):287-305.
6. Snow JL, Muller SA. Malignancy-associated multicentric reticulohistiocytosis: a clinical, histological and immunophenotypic study. *Br J Dermatol*. 1995;133(1):71-76.
7. Yiannias JA, el-Azhary RA, Gibson LE. Erythema elevatum diutinum: a clinical and histopathologic study of 13 patients. *J Am Acad Dermatol*. 1992;26(1):38-44.