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# Multicentric reticulohistiocytosis in a patient with thymic carcinoma

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***Key words:*** multicentric reticulohistiocytosis; paraneoplastic syndrome; thymic carcinoma.

## INTRODUCTION

Multicentric reticulohistiocytosis (MCR) is a rare disease characterized by papulonodular skin lesions of the hands, forearms, face, and ears associated with erosive arthritis. A review of the literature shows an association with underlying malignancy in approximately 25% of cases but only 1 reported association with thymic carcinoma. Here, we describe a patient with an atypical presentation of MCR that previously had been diagnosed as thymic carcinoma. The rarity of this disease necessitates awareness of uncommon manifestations, and its potential to be a paraneoplastic syndrome warrants an evaluation for associated malignancy.

## CASE PRESENTATION

A 61-year-old woman presented to the dermatology clinic with a 1-year history of increasing pruritus intermittently controlled with oral prednisone. After 1 year of these symptoms, she noted the onset of small papules over the posterior neck and along the left flank, which prompted a visit to the dermatologist.

Shortly after the onset of pruritus a year earlier, she had developed arthralgias that were diagnosed as rheumatoid arthritis and treated with hydroxychloroquine and prednisone. Six months after the onset of arthritis, she experienced acute chest pain and was found on imaging to have a 7.5-cm mass in her anterior chest. Biopsy showed a thymic carcinoma with extensive local invasion and a metastasis to the right pelvic bone. She was treated with carboplatin/paclitaxel and radiation and presented to our dermatology clinic 5 months later.

On examination, the patient had widespread areas of pruritus without primary lesions, as well as 2 nonpruritic areas with numerous 2- to 3-mm firm violaceous papules. These areas were located over the left flank and posterior neck (Fig 1). Of note, there were no lesions over her forearms, face, or ears. Although the patient was unaware of them, several slightly larger papules were noted over her fingers bilaterally (Fig 2). Ultrasonographic examination of her bilateral hands showed significantly active and symmetric inflammatory arthritis consistent with rheumatoid arthritis; no erosive changes were present. Laboratory workup for the pruritus was unremarkable.

*Abbreviation used:*

MCR: multicentric reticulohistiocytosis

Biopsies of the posterior neck and left flank were performed, showing a well-defined dermal infiltrate composed of histiocytes and multinucleated giant cells with copious pink cytoplasm and a ground-glass appearance (Fig 3). A scattered lymphocytic infiltrate was present, but there were no eosinophils. Lesions had positive test results for CD68 and negative results for S100, p63, and CK5/6 markers, establishing a histologic diagnosis of MCR.

At the time of publication, the patient is contemplating alternative chemotherapeutic options for persistent disease and recovering from the acute effects of radiation. Her pruritus has resolved, and her skin lesions are almost clear. She denies pain, and her hands are unchanged from her initial visit.

## DISCUSSION

MCR most commonly presents as flesh- to pink-colored papulonodules over the hands, forearms,

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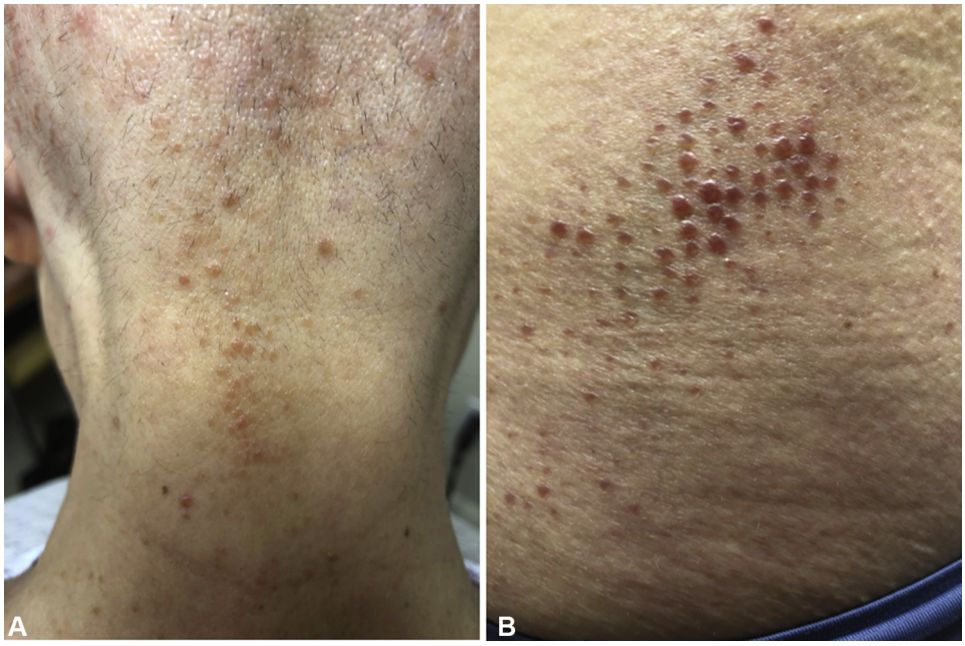


**Fig 1.** Flesh-colored and violaceous grouped papules on (**A**) the posterior neck and **(B)** the left flank.

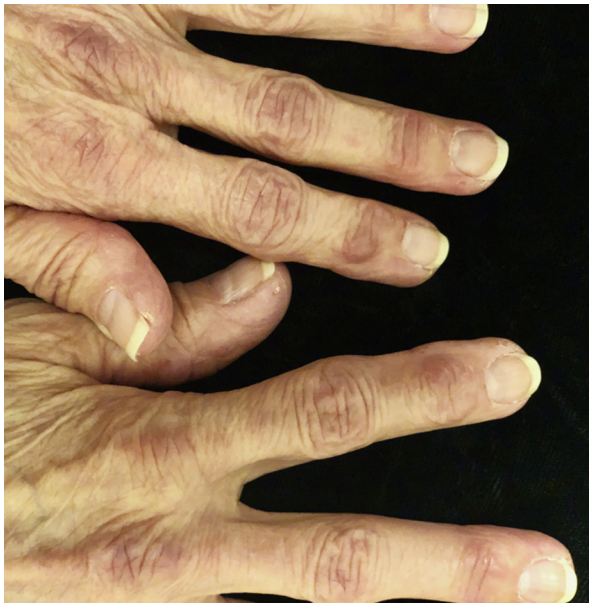

**Fig 2.** Papules on the dorsal hand.

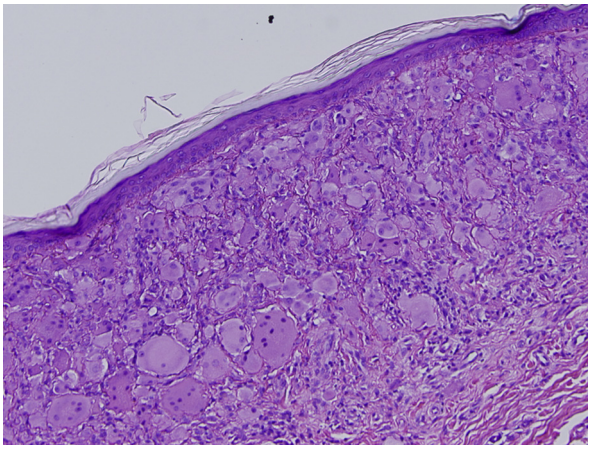

**Fig 3.** Histologic imaging of the posterior neck showing nodular aggregation of multinucleated histiocytes with ample pink cytoplasm.

face, and ears and is associated with an erosive arthritis. Roughly 300 cases have been reported worldwide, the majority of which involved middle-aged white women. Although the disease is rare, histopathologic findings are characteristic and include numerous histiocytes and multinucleated giant cells with ground-glass, eosinophilic cytoplasm. Immunostaining is characterized by monocyte-macrophage markers CD68 and CD45 and results are negative for S100, distinguishing this entity from Langerhans cell histiocytosis. Etiology of the disease has not been established but is postulated to involve stimulation of non-Langerhans cell histiocytes and osteoclast activation.[1]

Although the majority of MCR cases in the literature have no association with malignancy, an underlying cancer has been reported in approximately 25% of cases.[2] In most of these cases, cutaneous manifestations of MCR precede the diagnosis of cancer, prompting a screening for malignancy. The most common associations include

breast cancer, leukemia, and gastrointestinal malignancies, but cases have also been reported of gynecologic, lung, and skin cancers.[3] In only 1 other case was there a coexistence of MCR with thymic carcinoma.[4] In this report, a 70-year-old man diagnosed with MCR was found to have thymic carcinoma 4 months later, after a bout of chest pain.

Our patient presented with atypical features of MCR, including the location and timing. She also had pruritus, which may or may not have been a manifestation of the MCR and could have represented a separate paraneoplastic symptom or a reaction to treatment with chemotherapeutic agents. There was no significant inflammatory joint disease, often the sole presenting symptom of MCR, but it is worth noting that she was already receiving hydroxychloroquine. Finally, unlike other cases of malignancy associated with MCR, our patient had a diagnosis of thymic carcinoma before the discovery of MCR.

We have reported this case because of its rarity and characteristic histopathologic findings. MCR will often remit after several years; however, in some cases it may progress to arthritis mutilans, causing significant morbidity. Improved prognosis may be afforded by early diagnosis and treatment and recognition of the need for further testing.

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