DERMATOLOGICA SINICA 34 (2016) 42–45

Contents lists available at ScienceDirect

Dermatologica Sinica

journal homepage: http://www.derm-sinica.com

CASE REPORT

Multicentric reticulohistiocytosis in a Taiwanese woman with Sjögren syndrome

Lan-hsin Cheng, Ying-Yi Chiang*

Department of Dermatology, Wan Fang Hospital, Taipei Medical University, Wenshan District, Taipei City 116, Taiwan

ARTICLE INFO

Article history:
Received: Jun 6, 2015
Revised: Jul 31, 2015
Accepted: Aug 7, 2015

Keywords:
Multicentric reticulohistiocytosis
non-langerhans cell histiocytosis
erosive polyarthropathy
dermatomyositis
Sjögren syndrome

ABSTRACT

Multicentric reticulohistiocytosis is a rare, non-Langerhans cell histiocytosis characterized by specific skin lesions and destructive arthritis. Clinically, it generally presents with multiple reddish-brown papulonodules over the hands, face, and trunk. The associated destructive arthropathy tends to be severe, with 50% of patients developing arthritis mutilans. Approximately 25–30% of patients present with neoplasia. Coexisting autoimmune diseases are also commonly reported. Here we report the case of a 59-year-old Taiwanese woman with underlying Sjögren syndrome who presented with the typical skin manifestations of multicentric reticulohistiocytosis and early interphalangeal joint involvement. The patient later showed partial response to methotrexate treatment.

Introduction

Multicentric reticulohistiocytosis (MRH) is a rare, non-Langerhans histiocytosis of unknown etiology with a predilection for joints and skin.[1] Approximately 300 cases of MRH have been reported thus far.[2] Cutaneous manifestations typically present as multiple erythematous to brown papulonodules, possibly coalesced to plaques, over the acral regions of the face, such as the helices and antihelices of the ear and perinasal region; on the dorsal lateral aspect of the hands; around the nail folds; and across the trunk. Eruptions surrounding the periungual region give the characteristic appearance of coral beads. Mucosal involvement is common. The associated destructive polyarthritis tends to be severe, with approximately 50% of patients ending up with arthritis mutilans.[3] The arthritis mainly affects the interphalangeal joints, knees, wrists, shoulders, and elbows, but other joints may also be involved.[1] MRH can also affect other organs, including the heart, lungs, gastrointestinal tract, liver, and urogenital system. Possible association with underlying malignancy or autoimmune disease warrants careful clinical evaluation.[2] Here we report the case of a 59-year-old Taiwanese woman with underlying Sjögren syndrome who presented with the typical skin manifestations of MRH and early interphalangeal joint involvement.

Case report

A 59-year-old Taiwanese woman presented with a 3-month history of asymptomatic skin eruptions affecting the face, trunk, and limbs. Her medical history included Sjögren syndrome with intermittent treatment for approximately 4 years for a waxing and waning clinical course. On physical examination, erythematous to brown papules of extremely firm consistency were observed over the ears, perinostril area, forehead, neck, nape, and trunk and the dorsum of the hands (Figure 1A and B). Confluent plaques with a cobblestone surface were observed on the knuckles of the hands bilaterally, mainly involving the metacarpal joints. Moreover, the periungual regions were erythematous to violaceous, and some distal interphalangeal joints were swollen (Figure 1C). The cutaneous findings on the hands mimicked Gottron's papules; however, the patient had no other signs of dermatomyositis (Figure 1D). Scattered individual erythematous papules were observed over the forehead, neck, nape, and abdomen, with a focally annular arrangement over the left forearm. The mucosal areas were intact. Diascopy revealed a negative “apple jelly” sign (Figure 1E). Delicate arborizing vessels within the papules were observed through contact dermoscopy

Conflicts of interest: The authors declare that they have no financial or non-financial conflicts of interest related to the subject matter or materials discussed in this article.

* Corresponding author. Department of Dermatology, Wan Fang Hospital, Taipei Medical University, Number 111, Hsing-Long Road Section 3, Wenshan District, Taipei City 116, Taiwan.
E-mail address: ellychiang@hotmail.com (Y.-Y. Chiang).

http://dx.doi.org/10.1016/j.dsi.2015.08.001

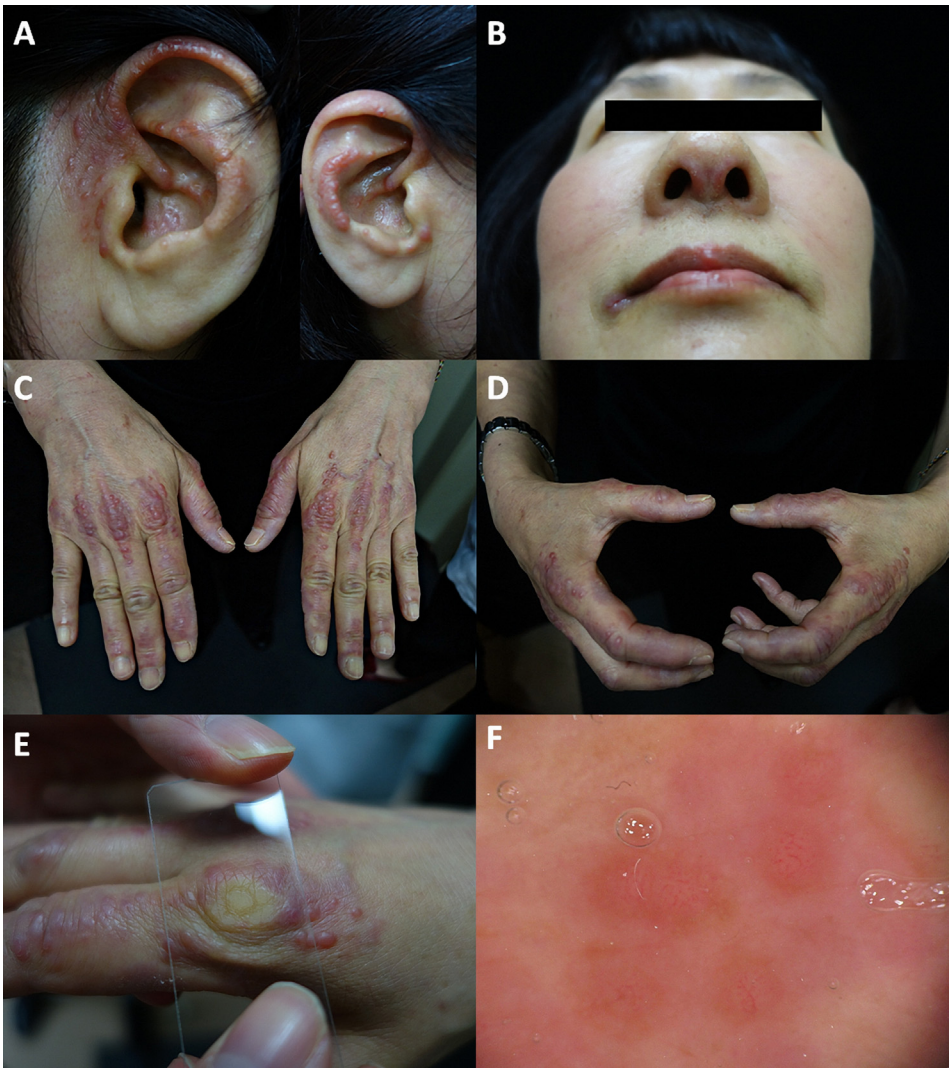

Figure 1 Clinical manifestations and dermoscopic findings of the patient with multicentric reticulohistiocytosis. (A) Closely arranged reddish-brown papules ("coral beads") were located over the helices and antihelices of the ear. (B) Similar lesions were observed over the perinostril area. (C, D) Note the confluent papules and nodules forming erythematous plaques with a cobblestone surface over the knuckles, mainly involving the metacarpal joints, as well as the presence of periungual erythema and swelling of the distal phalangeal joints. (E) Diascopy revealed a negative "apple jelly" sign. (F) Delicate thin arborizing vessels within the papules were observed via gel immersion (contact) dermoscopy.

(Figure 1F). In addition, the patient complained of finger joint stiffness and mild pain during movement of the fingers.

Laboratory tests included a complete blood count and renal and hepatic function examinations. A positive antibody to SS-A/Ro was found, compatible with a history of Sjögren syndrome. The patient's antinuclear antibody test was negative, as were her anti-dsDNA, anti–Scl-70, anti–Jo 1, anti-Smith, anti-ribonucleoprotein, and anti–SS-B/La antibody tests. Plain x-rays of both hands showed a fluffy change of cortex in the right third and fifth and left second and third middle phalanges. No underlying malignancy was detected in examinations in the oncology department.

A skin biopsy from the neck yielded dense, dome-shaped dermal infiltrates of epithelioid histiocytes with characteristic "ground-glass," eosinophilic cytoplasm and vesicular nuclei with prominent nucleoli. Binucleated and multinucleated cells were frequently observed (Figure 2A). The individual cells had two-toned cytoplasm of darker and lighter areas (Figure 2B). S100 staining was negative. Although plain radiographs of both hands revealed no obvious destruction of the articular surface, the patient complained of stiffness of the interphalangeal joints. Considering that clinical symptoms may precede detectable changes in radiographs, the patient was diagnosed with early MRH rather than diffuse cutaneous reticulohistiocytosis. The patient was then treated with methotrexate (MTX; 10 mg/wk), and the cutaneous lesions became more flattened and less obvious after 1 month of treatment (Figure 2C and D).

Discussion

MRH is a rare disease with the most cases reported among Caucasian women, although Hispanic, African, and Asian cases have also been described.[2,4,5] MRH generally begins in the 4th to 6th decades of life, and women are more commonly affected than men, in a ratio of 3:1.[2] The final diagnosis rests with tissue histology: skin, mucosa, or synovium biopsy yields multinucleated giant cells and histiocytes with eosinophilic, ground-glass cytoplasm.[6]

Clinically, approximately 60–70% of patients present with polyarthralgia as the first manifestation of the disease.[2] The differential diagnosis of MRH includes other causes of erosive polyarthritis, such as rheumatoid arthritis, psoriatic arthritis, erosive