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CASE REPORT

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Reticular erythematous mucinosis: Relationship between its dermoscopic and histopathological findings

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Key Clinical Message

In reticular erythematous mucinosis, (1) the presence of dotted vessels and (2) uniform, structureless, yellowish-white spots, and patches on dermoscopy correspond to histopathological findings of (1) vessels at the tips of the dermal papillae and (2) thickening and rupture of the collagen and fiber bundles with mucin deposition, respectively.

Abstract

Reticular erythematous mucinosis is a rare form of skin mucinosis that primarily affects middle-aged women, typically appearing as papules and plaques in the upper chest or midline of the back. Here, we report the case of a 75-year-old woman with skin papules and plaques left untreated for 8 years. A gross skin examination was followed by histopathological and dermoscopic examinations. The main dermoscopic findings were (1) dotted vessels and (2) uniform structureless yellowish-white spots and patches. The corresponding histopathological findings were (1) vessels at the tips of the dermal papillae and (2) thickening and rupturing of the collagen and fiber bundles with mucin deposition, respectively. Laboratory investigations revealed normal results, ruling out various autoimmune disorders. REM was diagnosed based on these findings. The study presents relevant evidence-based findings in dermatology and cutaneous pathology as it is the first description of REM using dermoscopy. Dermoscopic diagnosis without other unnecessary tests would benefit both the clinician and the patient.

KEYWORDS

cutaneous mucinosis, dermatopathology, dermoscopy, mucin, reticular erythematous mucinosis

1 | INTRODUCTION

Reticular erythematous mucinosis (REM) was first described by Steigleder in 1974.[1] It is a rare form of primary cutaneous mucinosis, typically affecting the upper chest or the back midline.

The exact etiology remains unknown, although familiar cases suggest a genetic predisposition. Moreover, several factors, such as ultraviolet light (UV) light, immunological disturbances, and viral infections, are thought to be associated with the development of REM. This condition is associated with autoimmune diseases, including lupus

erythematosus, hyperthyroidism, Hashimoto's disease, diabetes mellitus, thrombocytopenic purpura, tumors, myopathy, and polyneuropathy.[2] Despite being documented for several years,[3,4] no previous studies have described the dermoscopic characteristics of REM. Here, we present a case of REM visualized with dermoscopy and describe the relationship between the dermoscopic and histopathological findings.

2 | CASE PRESENTATION

The patient was a 75-year-old woman who had experienced skin redness for 8 years but left it untreated. She had no significant medical history and was generally healthy. A gross skin examination revealed red lesions on the anterior neck, chest, and back that crossed the midline of the affected areas (Figure 1A,B). Dermoscopy showed dotted vessels and uniform structureless yellowish-white spots and patches of irregular shapes and various sizes, which were diffused over a reddish background (Figure 2).

Histopathological findings revealed partial hyperkeratosis of the epidermis and vacuolar alteration of the keratinocytes with dilated blood vessels and perivascular infiltration of lymphocytes, histiocytes, and melanophages in the upper dermis. The dermis showed collagen fibers that progressed from fibrosis to sclerosis, with thickened bundles that were partially broken into clumps (Figure 3). Alcian blue staining confirmed the presence of mucin deposition in all dermal layers, with strong staining in the papillary layer (Figure 3B).

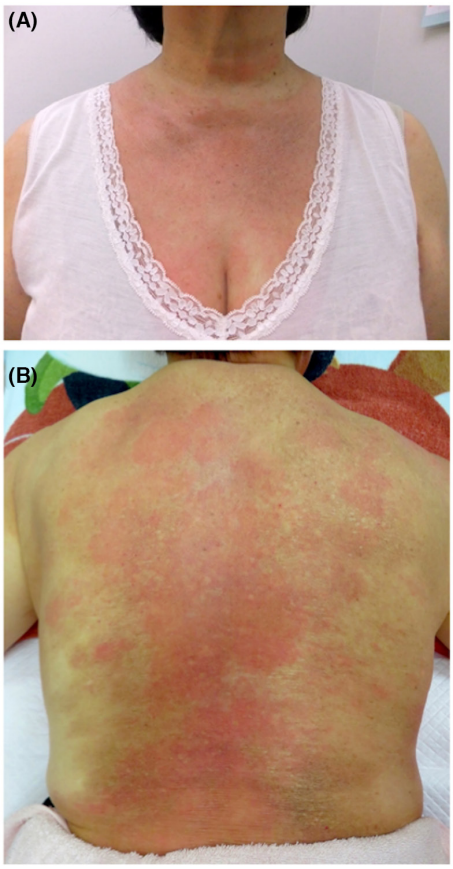

FIGURE 1 Clinical presentation. (A) Erythema is seen across the anterior neck and chest midline. (B) Erythema is seen across the midline on the back.

FIGURE 2 Dermoscopic findings. Dermoscopy shows dotted vessels, uniform, and structureless yellowish-white spots, and patches of irregular shapes and various sizes diffused on a reddish background.

Laboratory investigations for thyroid stimulating hormone (TSH), thyroxine (T4), and triiodothyronine (T3), thyroid antibodies (anti microsomal, anti peroxidase, and antithyroglobulin), antinuclear antibody (ANA), double-stranded deoxyribonucleic acid (dsDNA) antibody, Sjogren syndrome A (SSA) antibody, Ro, Sjogren syndrome B (SSB)/La antibody, Smith antibody, ribonucleic protein (RNP) antibody, scleroderma 70 (scl70) antibody, anti-Jo-1 antibodies, and complete blood count (CBC) revealed normal results, ruling out various autoimmune disorders. REM was diagnosed based on these findings. However, the patient did not approve the treatment, and follow-up was recommended.

3 | DISCUSSION/CONCLUSION

REM is a rare chronic persistent condition of unknown etiology, characterized by erythematous, papular, or plaque-like eruptions that appear on the midline of the back or chest.[1,4] REM was first described 50 years ago by Steigleder in 1974,[1] but only approximately 100 case reports have been published in English.[5] Although the etiology is unknown, fibroblasts of patients with REM have an abnormal response to exogenous IL-1beta.[6] Further,

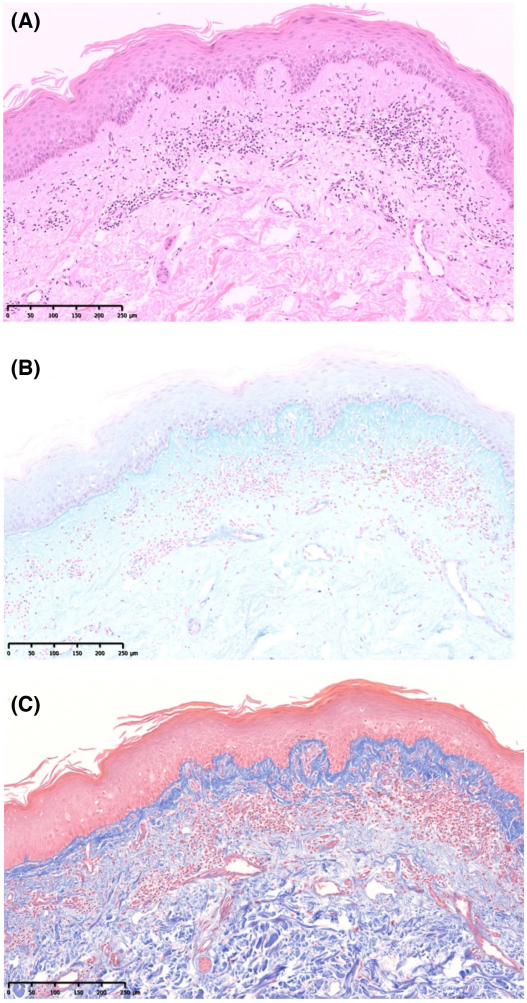

FIGURE 3 Histopathological findings. (A) The epidermis is mildly thickened, with partial hyperkeratosis and thickened granular layer. The dermo-epidermal junction is partially obscured, with vacuolar alterations in keratinocytes. Dilated blood vessels are observed in the upper dermis with perivascular lymphocyte and histiocyte infiltrate and mixed melanophages. Collagen fibers show a progression from fibrosis to sclerosis throughout the dermis. The collagen fiber bundles are thickened, homogenized, and partially broken into clumps. Hematoxylin–eosin (H&E) staining; original magnification, ×100; scale bar, 250 μm. (B) Mucin deposition is shown in blue. All layers of the dermis are stained, but the papillary layer of the dermis is particularly strongly stained. Alcian blue staining; original magnification, ×100; scale bar, 250 μm. (C). Collagen fibers are shown in blue. The papillary layer of the dermis is stained uniformly; however, below the dermal reticular layer, the staining is heterogeneous. Masson trichrome stain; original magnification, ×100; scale bar, 250 μm.

tubuloreticular inclusions have been detected in endothelial cells and pericytes within the skin lesions.[6]

Histologically, REM is associated with mild perivascular infiltrates in the superficial and mid-dermal layers with variable depth and perivascular extension. Lymphocytic perifollicular infiltration may also occur, with slight vascular dilation and sometimes mild focal hemorrhage in the papillary dermis. Separation of dermal collagen bundles and variable amounts of basophilic mucin is a hallmark of REM observed mainly in the upper and middle dermis. Mucin is the most prominent around the infiltrates, appendages, and upper dermis. A few stellate cells are also observed. The epidermis is typically normal, although mild spongiosis, and focal lichenoid inflammation have been reported. In some cases, there was slight degeneration of the basal layer and focal fragmentation of the elastic fibers. Staining reactions induced by mucins can help diagnose REM.[4] The histopathological findings of this case presented in Figure 3 were generally consistent with those previously reported.

To diagnose REM, clinicians must first rule out other conditions through clinical, histopathological, and laboratory correlation, especially lupus erythematous tumidus (LET); papular mucinosis (PM), known as the milder and more localized type; lichen myxedematosus (LM); dermatomyositis (DM); and scleredema.[3] In this case, the eruption of erythematous macules and papules in a reticulated pattern over the midline of the anterior neck and chest and back midline in sun-unexposed areas was observed (Figure 1). LET presents as erythematous to violaceous plaques or nodules, with or without annular patterns on sun-exposed areas. DM presents as erythematous to violaceous papules and plaques, most often located symmetrically on the extensor dorsal aspects of the metacarpophalangeal and interphalangeal joints, with the second most common distribution being a heliotrope eruption of the upper eyelids.

Scleredema shows firm woody plaques and diffuse symmetric non-pitting skin induration typically affecting the upper back and posterior neck. LM shows normochromic or erythematous papules ranging from 1 to 4 mm. The papules are firm, waxy, and symmetrically arranged and appear primarily on the back of the hands, fingers, and extensor surface for the arms, face, upper torso, and legs.[3] All these factors can be ruled out. Avoiding the sun and following sun protection measures are recommended as first-line therapy.

Conventional antimalarial drugs like chloroquine and hydroxychloroquine are considered second-line treatment options. Quinacrine may be used for allergic patients or patients with certain eye diseases. Other treatments with variable results include topical tacrolimus, oral antihistamines, tetracycline, cyclosporine, UVB irradiation, UVA1 irradiation, pulsed dye laser, and topical and systemic corticosteroids. It is important to note that REM is usually a self-limiting disease that may show spontaneous resolution, even after many years.[3,4] Notably, because of the similarities between REM and Tumid lupus, both diseases have been grouped together. The remarkable overlap suggests they may actually be the same disease.[4]

In the present case, the main dermoscopic findings were (1) dotted vessels and (2) uniform structureless yellowish-white spots and patches. The corresponding histopathological findings were (1) vessels at the tips of the dermal papillae and (2) thickening and rupturing of the collagen and fiber bundles with mucin deposition, respectively. We believe dermoscopy can provide useful information for the naked-eye examination and assist in the differential diagnosis by distinguishing REM from LET, PM, DM, and scleredema. However, the current study is based on a single case. Dermoscopy should be performed for a larger number of cases to validate the findings in the present case.

To our knowledge, this is the first study to report the visualization of REM with dermoscopy. Although the incidence of REM is low, it is a self-limiting disease with the best prognosis among the other diseases considered in the differential diagnosis. Therefore, dermoscopic diagnosis without other unnecessary tests would be beneficial for not only the clinician but also the patient.

AUTHOR CONTRIBUTIONS

Tomoaki Takada: Conceptualization; data curation; formal analysis; funding acquisition; investigation; methodology; project administration; resources; software; supervision; validation; visualization; writing – original draft; writing – review and editing.

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CONFLICT OF INTEREST STATEMENT

The author has no conflicts of interest to declare.

DATA AVAILABILITY STATEMENT

All data generated or analyzed during this study are included in this article. Further inquiries can be directed to the corresponding author.

ETHICS STATEMENT

All the procedures adopted in this study were in accordance with the ethical standards of the World Medical Association Declaration of Helsinki. Ethical approval was not required for this study per local and national guidelines.

CONSENT

Written informed consent was obtained from the patient for the publication of this case report and accompanying images.

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