QUIZ SECTION

Chronic Erythematous Swelling of Unilateral Eyelid: A Quiz

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A Chinese female patient in her 60s presented with a 3-month history of progressive erythematous swelling on her right upper eyelid. She reported that the lesion had been asymptomatic but the opening of the right eye had been gradually affected. Her medical history was unremarkable.

Physical examination revealed prominent erythematous swelling of the right upper eyelid, with enlarged pores and follicular plugging (Fig. 1A). Slight swelling of the right face and symmetrical erythema on the forehead and wings of the nose were also noted (Fig. 1A). Red purplish plaque and excoriation were present on the upper back (Supplementarty Material). No Gottron sign or Gottron papules were observed. Raised serum lactate dehydrogenase (271 U/L, reference 120–250U/L) and α-hydroxybutyrate dehydrogenase kinase (233.2 U/L, reference, 72–182 U/L) were noted. Anti-dsDNA, anti-Sm, and anti-MDA5 antibodies were negative. A skin biopsy showed basal layer vacuolation, basement membrane thickening, perivascular lymphocytic infiltrate, and oedema in the superficial dermis (Fig. 2A). Interstitial mucin deposition was proved by alcian blue staining (Fig. 2B), and basement membrane thickening was indicated by PAS staining (Supplementarty Material). Muscle biopsy was not conducted because the patient did not consent.

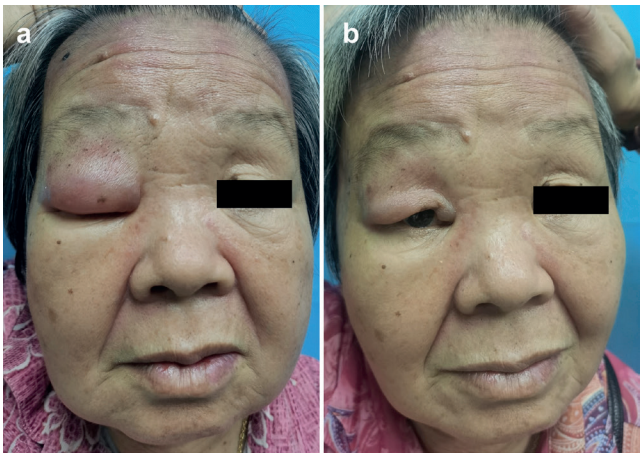

Fig. 1. Erythematous swelling with enlarged pores and follicular plugging on the right eyelid and erythema on the face (A) before and (B) after treatment. Permission given by the patient to publish these photos.

What is your diagnosis?

Differential diagnosis 1: Dermatomyositis
Differential diagnosis 2: Contact dermatitis
Differential diagnosis 3: IgG4-related disease
Differential diagnosis 4: Kimura's disease

See next page for answer

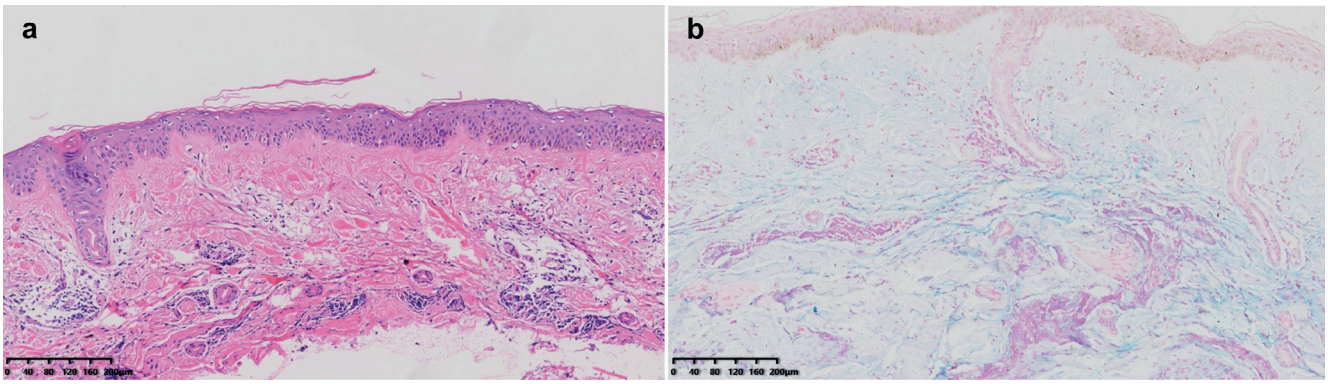

Fig. 2. (A) Haematoxylin-eosin stain (H&E) stain (magnification: 10×) and (B) alcian blue staining (magnification: 10×).

ANSWERS TO QUIZ

Chronic Erythematous Swelling of Unilateral Eyelid: A Commentary

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Diagnosis: **Dermatomyositis**

Eyelid swelling can be seen in many diseases such as contact dermatitis, angioedema, autoimmune connective tissue disease, IgG4-related disease, Kimura disease, and others (1–4). Those lesions characterized by chronic asymptomatic progression are more likely to occur in the latter 3 diseases.

Dermatomyositis is an autoimmune connective tissue disease characterized by muscle inflammation and skin lesions (5). Although dermatomyositis is relatively common, the diagnosis is often challenging as unilateral or asymptomatic lesions are easily neglected by both clinicians and patients themselves, especially in the early stage. Characteristic skin findings of dermatomyositis include photodistributed poikiloderma, heliotrope sign, Gottron papules, Gottron sign, periungual telangiectasia, and calcinosis cutis (5). Other non-specific skin manifestations such as skin ulcers, non-scarring alopecia, psoriasis-like rashes, and follicular keratoses can also be present (5). The heliotrope sign refers to erythematous to violaceous eruption on the eyelids, which may be accompanied by periorbital oedema. Although the heliotrope sign in dermatomyositis is usually symmetric, the swollen eyelid can be unilateral or bilateral, ranging from mild to marked (3). Notably, unilateral eyelid swelling was reported to be associated with anti-melanoma differentiation-associated gene 5 (Anti- MDA5)-antibody-positive dermatomyositis, which is an important risk factor for rapidly progressive interstitial lung disease (6, 7).

Pathologic changes in skin biopsy specimens from patients with dermatomyositis may include epidermal atrophy, basement membrane degeneration, and basal cell vacuolization, as well as dermal interstitial mucin deposition and sparse lymphocytic infiltration.

The myopathy of dermatomyositis often involves proximal muscles, and is characterized by myalgia and muscle weakness. Muscle enzymes and muscle biopsies can provide evidence of muscle injury. However, for patients with characteristic skin lesions but no signs of muscle injury for over half a year, the diagnosis of amyopathic dermatomyositis can be made. If there is no muscle symptom, but muscle enzyme is raised, it can be diagnosed as hypomyopathic dermatomyositis (8).

Once a diagnosis of DM is confirmed, systemic involvements should be assessed. Treatment options for dermatomyositis include systemic corticosteroids, antimalarials, mycophenolate mofetil, methotrexate, rituximab, intravenous immunoglobulin, Janus kinase inhibitors, and other (8).

In the current case, the skin lesions and histopathological changes, together with the raised enzyme levels, led to the diagnosis of dermatomyositis (probably hypomyopathic). The rash had somewhat improved after prednisone (20 mg/day) and hydroxychloroquine (0.2 g/day) treatment at the 2-week follow-up (Fig. 1B and Supplementary Material). The patient continued the therapy and adjusted the dose of corticosteroids when the condition improved. Other management of the patient included regular evaluation of overlapping autoimmune connective tissue diseases, and underlying systemic lesions and malignancy. No significant findings were made up to the last follow-up.

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