Oral lichen planus in a dermatomyositis patient that resolved after intravenous immunoglobulin therapy

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Dermatomyositis (DM) is an autoimmune muscle disorder characterized by proximal muscle weakness and skin lesions. The significance to oral health professionals includes oral manifestations and increased incidence of malignancy that might occur in the oral cavity. Reports of oral mucosal involvement in DM are scanty. When they are published, there is often a clinical description of "resembling lichen planus" or "leukoplakia-like" without histologic evaluation. This makes it difficult to establish the definitive diagnosis of these oral lesions and formulate treatment options. It is also difficult to assess the relationship between oral lesions and oral malignancy in this patient population. We report a DM patient who presented with clinical and histologic features of lichen planus of the buccal mucosa that resolved with intravenous immunoglobulin treatment for DM. (Oral Surg Oral Med Oral Pathol Oral Radiol 2014;118:e111-e114)

Dermatomyositis is a group of idiopathic inflammatory myopathies. The most recent classification divides idiopathic inflammatory myopathies into 3 categories, polymyositis, inclusion body myositis, and dermatomyositis (DM), based on the disease mechanisms. DM is subdivided on the basis of clinical manifestations into classic DM (CDM) with skin lesions and proximal muscle weakness, adermatopathic DM without skin lesions, amyopathic DM without muscle weakness, hypomyopathic DM with skin lesions and subtle muscle disease, and postmyopathic DM with previous CDM but remaining active skin lesions.[1]

The classic skin lesions in DM include heliotrope rash of the eyelid and periorbital skin with or without edema, violaceous raised rashes at the metatarsophalangeal and interphalangeal joints (Gottron's papules), rash on the anterior chest ("V" sign), rash on the back and shoulders (shawl sign), and calcium deposits under the skin or in the muscle (calcinosis cutis). A variety of oral mucosal lesions have been reported in DM including lacy white lines resembling oral lichen planus (OLP), erosions and ulcers, atrophy of mucosa, white patches, and gingival telangiectasias.[2-8] Squamous cell carcinoma of the tongue has also been reported in a DM patient.[8]

Unlike the well-established risk of internal malignancy in DM,[1] the risk of oral mucosal malignancy in DM is unknown other than the report of lingual carcinoma.[8] Difficulty in establishing the risk of oral cancer in DM can be related to the fact that various lesions in the oral mucosa reported in DM have never been histopathologically defined. Are those lesions components of DM? What is their pathogenesis? Do they simply coexist with DM? Are those lesions precancerous or do they have potential for malignant transformation? The histopathologic nature of these lesions holds promising answers to these questions as well as the management of the lesions.

We report a case of CDM with white oral mucosal lesions that clinically and histologically met the criteria of OLP. To our knowledge it is the first case report in the English literature that includes both clinical and histopathologic features of OLP for the oral lesions in DM.

CASE REPORT

A 55-year-old man came to the dental clinic complaining of a sore mouth of approximately 2 years duration. He had developed a heliotrope rash over the eyelids, forehead, and bridge of the nose (Figure 1) along with Gottron's sign on the hands almost 5 years before the dental visit. He stated that he had experienced weakness of his neck muscles that caused difficulty in raising his head. For the last 5 years he had received courses of systemic corticosteroids and other immunosuppressant therapy under the provisional diagnosis of systemic lupus erythematosus. The treatments provided little improvement of the skin lesions or muscle symptoms. His muscle weakness had progressively worsened. He was finally diagnosed with DM 1 month before his dental visit and was preparing for intravenous immunoglobulin (IVIG) therapy. His medications at the time of the dental visit included leflunomide, amlodipine besylate, citalopram, and valsartan.

Examination revealed interlacing white lines on the buccal mucosa, with focal erosive areas (Figure 2). Histopathologic examination of the buccal mucosal lesions revealed a

Statement of Clinical Relevance

To raise awareness of the possible relationship of oral lichen planus (OLP) and dermatomyositis, an autoimmune muscle disorder with or without dermatologic involvement, and possible effectiveness of IVIG in the treatment of systemic OLP.

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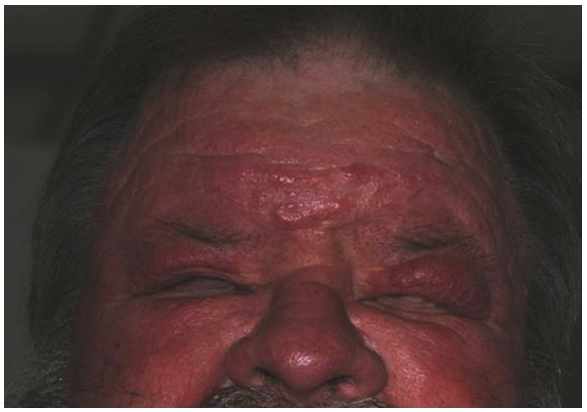

Fig. 1. Heliotrope rash over the eyelids, forehead, and bridge of the nose before the intravenous immunoglobulin (IVIG) therapy.

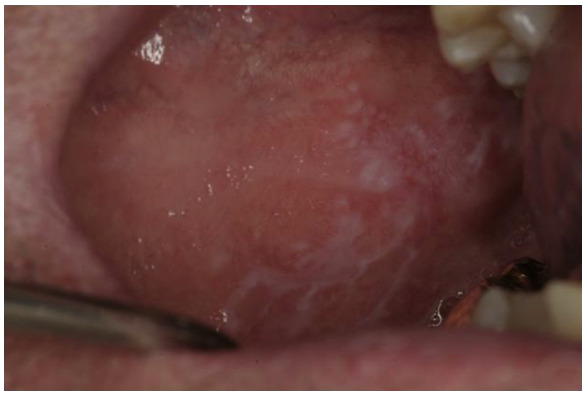

Fig. 2. Multiple white lacy lines on buccal mucosa with focal erosive areas before the intravenous immunoglobulin (IVIG) therapy.

thickened parakeratin layer, a well-defined bandlike zone of lymphocyte infiltration, Civatte bodies in the basal layer, (Figure 3), and liquefactive degeneration of the basal layer (Figure 4). Clinically and histopathologically, these features met the diagnostic criteria for OLP.

IVIG therapy (2 g/kg/month administered over 4 days every 4 weeks) was initiated shortly after the diagnosis of DM by the patient's rheumatologist. The patient reported regaining muscle strength and cessation of the sore mouth after 2 courses of IVIG.

On follow-up examination 6 months after the biopsy, the oral lesions had completely resolved (Figure 5). There was also improvement of the facial and hand rashes. The patient continues to be free of mouth soreness 18 months after the initiation of IVIG therapy.

DISCUSSION

Diagnosis of DM can be challenging, especially because the diagnostic criteria are still evolving. As knowledge of the pathogenesis expends, new biological markers continue to emerge and noninvasive diagnostic techniques and methods are increasingly available. For

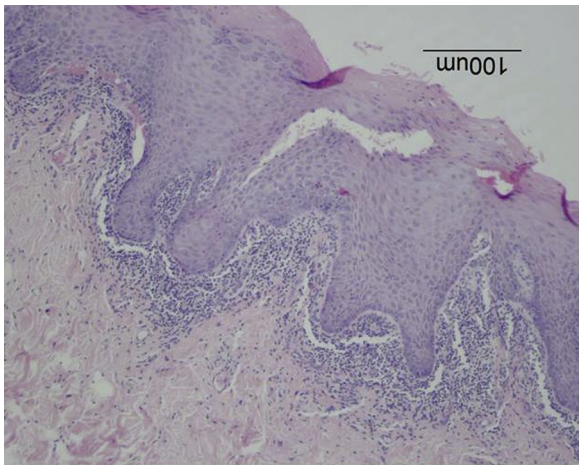

Fig. 3. A photomicrograph shows a thickened parakeratin and spinous cell layer, saw-tooth rete ridges, and Civatte bodies in the epithelium, with an intense, band-like infiltrate of lymphocytes in the superficial connective tissue. (hematoxylin-eosin, original magnification ×100).

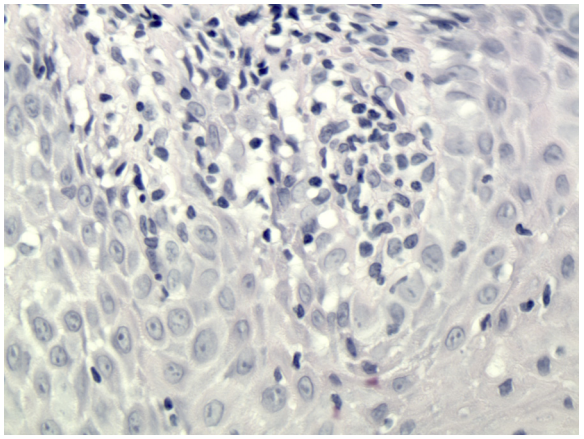

Fig. 4. A photomicrograph shows liquefactive degeneration of basal call layer. (hematoxylin-eosin, original magnification 63 × 002).

these reasons the diagnostic process has become much more complex. In general a typical case can be diagnosed based on pathognomonic Gottron's sign, a raised violaceous rash on the hand knuckles, characteristic heliotrope rash of the eyelid, and proximal muscle weakness with elevated serum creatine kinase. In an atypical case or early stage of the disease, when skin or muscle symptoms and signs are not obvious and serum creatine kinase elevation is absent, other biomarkers such as antinuclear antibodies and myositis-specific antibodies are used to aid in the diagnosis. In addition, muscle biopsy, electromyography, magnetic resonance imaging, and ultrasound are helpful. However, the results are not always consistent.[1] This complex diagnostic process can explain why our patient underwent 5