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# Dermatomyositis, clinically presenting with cutaneous ulcers, with histopathologic evidence of perforating collagenosis

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## Abstract

Dermatomyositis is a systemic, autoimmune disease with a variety of clinical features that often include myositis and characteristic cutaneous findings. A subset of patients with dermatomyositis develop cutaneous ulcers, often in the setting of vasculitis or vasculopathy. We present a case of dermatomyositis with cutaneous ulcers that show perforating collagenosis on histopathologic examination. Acquired reactive perforating collagenosis typically occurs in the setting of diabetes mellitus, chronic renal failure, and other pruritic conditions, and this case represents a rare association with dermatomyositis, which may ultimately be helpful in elucidating the pathophysiology of this perforating disorder.



**Figure 1.** Posterior aspect of the shoulder with a large, well-demarcated, oval, punched-out ulcer with violaceous borders

## Case Presentation

**PATIENT:** 48-year-old woman

**DURATION:** One year

**DISTRIBUTION:** Scalp, face, trunk, and extremities

**HISTORY:** A 48-year-old woman with a history of dermatomyositis was admitted to Tisch Hospital for evaluation of progressive shortness of breath and weight loss. The Dermatology Service was consulted to assess skin lesions that had been present for approximately nine months and that had recently become ulcers.



**Figure 2.** Right side of the back with hyperkeratotic, follicular papules.



**Figure 3.** Dorsal aspects of hands with multiple, punched-out, round ulcers with peripherally violaceous borders and central serosanguinous crust.



**Figure 4.** Palmar aspect of the fingers with multiple, scattered, resolving ulcers.

The patient initially presented in 2013 with pain in the knees and hands, swelling around the cuticles, Raynaud's phenomenon of the fingers, scale and erosions on the lateral aspects of the fingers, and difficulty breathing. She was evaluated by a rheumatologist and initially a diagnosis of systemic lupus erythematosus was made, and she was started on prednisone, hydroxychloroquine, and mycophenolate mofetil, with improvement in the joint pain and eruption on the hands. Over the next year, she developed hair loss. In April, 2015, she developed proximal muscle weakness, shortness of breath, chest pain, periorbital swelling, and a new eruption that consisted of hyperpigmentation on the forehead, which subsequently spread to involve

the chest, shoulders, and arms. She was referred to the Rheumatology Clinic in September, 2015, and found to have skin findings that were consistent with a shawl sign, V-sign, periorbital swelling, i.e., heliotrope eruption, Gottron's papules, capillary loop vasculopathy, painful and swollen fingers, symptoms of Raynaud's, and mechanics hand in the setting of proximal muscle weakness. Therefore a diagnosis of dermatomyositis was made. With the cutaneous findings in the setting of shortness of breath, there was concern for anti-synthetase syndrome. A chest computerized tomography scan showed fibrotic changes in the lower lobes and pulmonary function tests showed a low diffusing capacity.

The patient was admitted to the hospital for shortness of breath and weight loss in the setting of odynophagia. The shortness of breath was determined to be a result of muscle weakness as no evidence of interstitial lung disease or pulmonary hypertension was found on chest computerized tomography scan or transthoracic echocardiogram, respectively. Regarding her skin disease, the patient reported lesions on her hands, chest, back, shoulders, hips, and face intermittently for the prior nine months. In the two weeks prior to admission, the lesions on the hands and shoulders had developed ulcers; she also reported an ulcer on the hard palate. There was no pruritus associated with the skin lesions, but she noted appreciable pain at the sites of the cutaneous ulcers. During the hospitalization, the patient was treated with mycophenolate mofetil and prednisone.

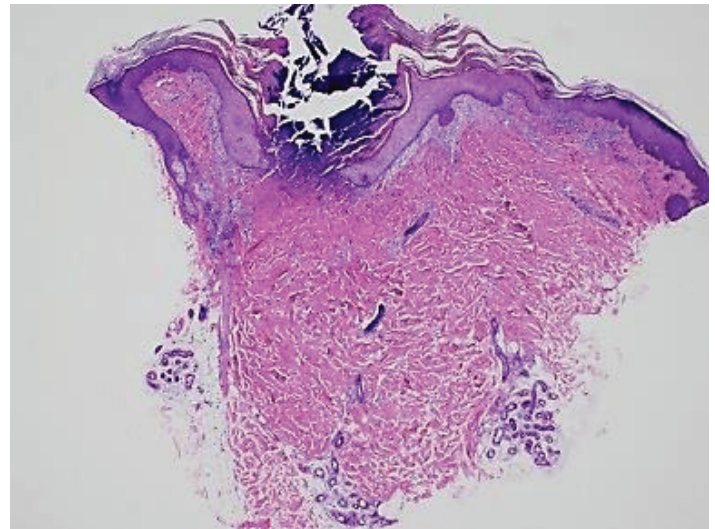
**PHYSICAL EXAMINATION:** On the scalp, there was diffusely decreased hair density and alopecic patches in the absence of erythema or scale. On the forehead and cheeks were reticular, brown patches, predominantly on the right side. In the mouth, the hard palate had an ovoid erythematous plaque. On the chest in a V-distribution, as well as on the back, upper arms, and the lateral aspects of the hips were multiple, reticulated, hyperpigmented patches. On the posterior aspects of the shoulders, there was a large, well-demarcated, oval, punched-out ulcer with violaceous borders (**Figure 1**). On the right side of the back, distributed in a linear fashion, there were four hyperkeratotic, follicular

papules, and there also was a cutaneous horn with a skin-colored papule at the base (**Figure 2**). On the elbows, there were multiple, erythematous, scaly, thin papules and plaques with scattered, punched-out, crusted ulcers. On the medial aspect of the left knee, there was a linear, thin plaque with scattered, superficially eroded papules with hemorrhagic crusts. On the dorsal aspects of the hands, there were multiple, punched-out, round ulcers with peripherally violaceous borders with central serosanguinous crust, most focused on the metacarpophalangeal joints and proximal areas of the interphalangeal joints (**Figure 3**). On the palmar aspects of the fingers were multiple, scattered, resolving ulcers (**Figure 4**). There was slightly delayed capillary refill. There was no distal finger color change. There was no ruffling or gross vessel dilation at the cuticle. There was an absence of poikiloderma, classic shawl sign, V-sign, holster sign, Gottron's papules, and heliotrope rash.

**LABORATORY DATA:** The creatine kinase level has ranged from less than 20u/L while on glucocorticoids to 208u/L (reference range: 30 to 170). The aldolase level was normal. The erythrocyte sedimentation rate ranged from 29 to 113mm/hr (reference range: 0-to-20 mm/hr). The C-reactive protein ranged from 0.2 to 25mg/L (reference range: 0 to 9). The lactate dehydrogenase level ranged from 232 to 267u/L (reference range: 135 to 214). The anti-Ro antibody was positive. The anti-double stranded DNA, anti-La, anti-Jo-1, anti-TIF1 $\gamma$ , anti-EJ, anti-Mi-2, anti-OJ, anti-PL-7, anti-PL12, anti-ribonucleoprotein, and anti-cyclic citrullinated peptide antibody levels were negative. A muscle biopsy specimen from the right thigh showed skeletal muscle with type two myofiber atrophy.

**HISTOPATHOLOGY:** There is a narrow ulcer with overlying prominent scale crust. Longitudinally oriented collagen fibers are present within the epidermis (**Figure 5**).

**DIAGNOSIS:** Dermatomyositis, clinically presenting with cutaneous ulcers, with histopathologic evidence of perforating collagenosis.



**Figure 5.** Biopsy showing longitudinally oriented collagen fibers within the epidermis

## Discussion

Dermatomyositis is a systemic, autoimmune disease that often presents with myositis with a wide spectrum of severity and variable cutaneous findings that range from erythematous macules and papules to nodules and ulcerations. Fewer than 20% of patients have been reported to present with cutaneous ulcers [1], and these ulcers typically are believed to be refractory to treatment and associated with an overall poor clinical course [2]. Ulcers often present on extensor aspects of the joints, digits, and sun-exposed skin [1]. Histopathologic sections from most of the published cases of cutaneous ulcers in adult patients with dermatomyositis show severe vasculitis [2] and other factors, such as vasculopathy, interface inflammation, and superficial trauma have been suggested to contribute to the pathogenesis [1].

Additional concerns in dermatomyositis include the development of interstitial lung disease and an association with malignant conditions [3]. Because of this, clinical, histopathologic, and serologic features of dermatomyositis have been assessed to attempt to categorize the disease and determine the likelihood of developing associated comorbidities. A subset of patients with clinically amyopathic dermatomyositis has been defined, who have a predisposition to develop progressive interstitial lung disease, which also is associated with the presence of antibodies to the melanoma

differentiation-associated gene 5 (MDA-5) RNA helicase [1, 4]. Patients with MDA-5 antibodies have been shown to possess distinct cutaneous findings, which include digital and elbow ulcers and tender palmar papules, and to have an increased likelihood of having oral findings, alopecia, joint symptoms, and hand swelling, potentially due to severe vasculopathy [5]. Our patient has several features that were consistent with the anti-MDA-5 phenotype, which include ulcers of the elbows, hand swelling, palmar lesions, alopecia, joint symptoms, oral ulcers, and pulmonary pathology. Interstitial lung disease does not appear to be the current etiology of her pulmonary symptoms.

A distinguishing feature of our patient's clinical presentation is the cutaneous ulcers with the histopathologic finding of perforating collagenosis. Acquired reactive perforating collagenosis (ARPC) is a disease with pruritic, umbilicated papules with a central keratotic plug that is characterized histopathologically by the elimination of degenerated collagen through a cup-shaped depression in the epidermis [6-8]. It most often occurs in the setting of diabetes mellitus and chronic renal failure and also is observed in liver disease, endocrine disorders, and malignant conditions [7], but, more recently, an association with inflammatory (atopic and stasis dermatitis [9]) and infectious (viral [6, 10], scabies [11], insect bites [12]) diseases has been appreciated [6]. Three cases have been published that suggest an association between ARPC and dermatomyositis. In the first case, a patient with dermatomyositis, which was characterized by typical cutaneous findings and muscle weakness, was well-controlled on oral glucocorticoids for five years when she developed severely pruritic umbilicated papules with central crusts on the back at the time that muscle weakness and tenderness returned. Histopathologic features showed findings that were consistent with ARPC. Both ARPC and muscle weakness improved with oral glucocorticoids [6]. Two patients with dermatomyositis have been described who developed ARPC over the lower back in the setting of pruritus and absence of diabetes mellitus or renal dysfunction although one had a history of hepatitis C virus infection [13], which may independently be associated with ARPC [14]. To our knowledge, the

described case is the fourth case of dermatomyositis associated with ARPC.

A relationship between superficial trauma from scratching in pruritic diseases and perforating lesions has been appreciated [8] as the lesions may improve with treatment of pruritus and the underlying disease [7, 15]. It has been suggested that scratching in the setting of certain features of the tissue environment, such as inadequate blood supply, may lead to necrosis and impair wound healing and promote the development of ARPC [6, 16]. Others have suggested that ARPC occurs more frequently in association with fibrotic diseases and that transforming growth factor- $\beta$  may be important for tissue remodeling in the lesions [15]. In the setting of malignant conditions, ARPC has been suggested to be a paraneoplastic phenomenon [16]. In the case of dermatomyositis, pruritus is often present; in our case, it is possible that prominent vasculopathy may have altered the local tissue environment and played a role in the etiology of ARPC.

ARPC may occur at the time of relapse or after prolonged lack of control of the underlying disease and will often improve with treatment of the underlying disease [6, 7], which would be the treatment of choice in our patient. Suggested management options include topical, intralesional, or systemic glucocorticoids, topical or systemic retinoids or antibiotics, antihistamines, narrow-band ultraviolet B phototherapy [9], allopurinol [17, 18], and surgical methods [16].

## References

1. Narang NS, et al. Cutaneous ulceration in dermatomyositis: association with anti-melanoma differentiation-associated gene 5 antibodies and interstitial lung disease. *Arthritis Care Res (Hoboken)* 2015; 67:667
2. Tsujimura S, et al. Complete resolution of dermatomyositis with refractory cutaneous vasculitis by intravenous cyclophosphamide pulse therapy. *Intern Med* 2008; 47:1935
3. Robinson AB, Reed AM. Clinical features, pathogenesis and treatment of juvenile and adult dermatomyositis. *Nat Rev Rheumatol* 2011; 7:664
4. Sato S, et al. RNA helicase encoded by melanoma differentiation-associated gene 5 is a major autoantigen in patients with clinically amyopathic dermatomyositis: Association with rapidly progressive interstitial lung disease. *Arthritis Rheum* 2009; 60:2193

5. Fiorentino D, et al. The mucocutaneous and systemic phenotype of dermatomyositis patients with antibodies to MDA5 (CADM-140): a retrospective study. *J Am Acad Dermatol* 2011; 65:25
6. Amano H, et al. Acquired reactive perforating collagenosis in dermatomyositis. *J Dermatol* 2011; 38:1199
7. Faver IR, et al. Acquired reactive perforating collagenosis. Report of six cases and review of the literature. *J Am Acad Dermatol* 1994; 30:575
8. Mehregan AH, et al. Reactive perforating collagenosis. *Arch Dermatol* 1967; 96:277
9. Kim SW, et al. A clinicopathologic study of thirty cases of acquired perforating dermatosis in Korea. *Ann Dermatol* 2014; 26:162
10. Bank DE, et al. Reactive perforating collagenosis in a setting of double disaster: acquired immunodeficiency syndrome and end-stage renal disease. *J Am Acad Dermatol* 1989; 21:371
11. Kurschat P, et al. Acquired reactive perforating collagenosis triggered by scabies infection. *Acta Derm Venereol* 2000; 80:384
12. Kim EJ, et al. Acquired reactive perforating collagenosis triggered by insect bite. *J Dermatol* 2007; 34:677
13. Kikuchi N, et al. Acquired reactive perforating collagenosis: a rare association with dermatomyositis. *Acta Derm Venereol* 2013; 93:735
14. Saray Y, et al. Acquired perforating dermatosis: clinicopathological features in twenty-two cases. *J Eur Acad Dermatol Venereol* 2006; 20:679
15. Tsuboi H, Katsuoka K. Characteristics of acquired reactive perforating collagenosis. *J Dermatol* 2007; 34:640
16. Karpouzis A, et al. Acquired reactive perforating collagenosis: current status. *J Dermatol* 2010; 37:585
17. Querings K, et al. Treatment of acquired reactive perforating collagenosis with allopurinol. *Br J Dermatol* 2001; 145:174
18. Hoque SR, et al. Acquired reactive perforating collagenosis: four patients with a giant variant treated with allopurinol *Br J Dermatol*. 2006; 154:759