The Weakening Rash: What Does it Mean?

By Catherine Maari, MD, FRCPC

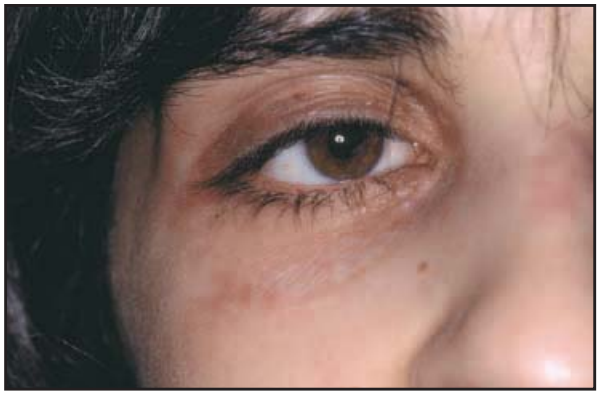

Figure 1. Heliotrope rash: periorbital macular violaceous erythema.

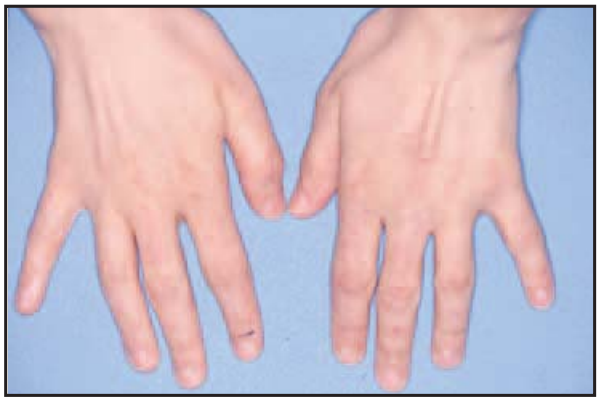

Figure 2. Gottron's sign: Confluent macular erythema most pronounced over the metacarpophalangeal/ interphalangeal joints, extending in a linear fashion over the extensor tendons.

A 27-year-old woman complains of a slightly tender rash of two months duration. It started with swelling over her eyelids and cheeks and went on to develop a red and purple color. The erythematous confluent rash extended to involve her shoulders and back, and was associated with severe pruritus.

She also reported progressive muscle weakness which severely interfered with her daily activities. She started having difficulty carrying her two-year-old child in her arms. Climbing the stairs, combing her hair and getting up from a squatting position were increasingly difficult tasks. She denied dysphagia, arthralgia and shortness of breath. Her past medical history was unremarkable and she was not taking any medications or recreational drugs.

On physical examination, erythema, swelling and minute telangiectases were seen on the eyelids (Figure 1). Diffuse erythema was noted over the upper trunk, neck and shoulders. Dilated telangiectatic vessels were seen on the proximal nail folds. A reddish purple scaling eruption was also noted symmetrically over the knuckles, knees and elbows (Figure 2).

A severe symmetric proximal muscle weakness, including the deltoid, biceps, quadriceps and triceps, was also discovered on physical exam.

What's your diagnosis?

Answer: Dermatomyositis.

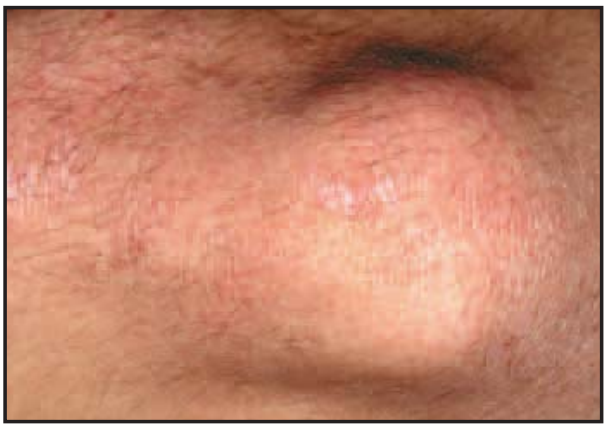

Figure 3. Confluent erythema overlying the patella.

Dermatomyositis is an inflammatory myositis that is characterized by skin involvement with muscular inflammation and degeneration. It is relatively rare and twice as prevalent in women as in men. There is a bimodal peak, one seen in children and the other seen in adults between the ages of 40 to 65. Humoral immunity is probably involved in its etiology. Muscle involvement without dermatitis is called polymyositis. Proximal muscle involvement is characteristic (Table 1).

The skin findings in dermatomyositis vary and include swollen pinkish violet eyelids (heliotrope rash), photosensitivity, pruritus, alopecia, calcification and enlarged capillaries on the proximal nail folds. A diffuse erythematous rash can be seen

Table 1

Criteria for diagnosing dermatitis

Three of the following criteria permit a diagnosis of dermatomyositis. The last two criteria make dermatomyositis probable.

1. Symmetrical weakness of limb girdles, muscles and anterior flexors of neck.
2. Elevated muscle enzymes: creatinine kinase, aldolase, transaminases and lactic dehydrogenase.
3. Abnormal electromyogram.
4. Characteristic myositis on muscle biopsy.
5. Typical dermatologic features.

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on the neck, thorax, shoulders or arms. The shawl sign was seen is this patient and represents an erythematous rash over the shoulders, neck and upper back. Gottron's papules are thought to be pathognomonic of dermatomyositis and represent flat-topped violaceous papules over the knuckles. Gottron's sign is a symmetric confluent macular violaceous erythema with or without edema overlying the dorsal aspect of the interphalangeal, metacarpophalangeal joints, olecranon processes, patellae (Figure 3) and medial malleoli.

The muscle weakness is symmetrical and most frequently involves the shoulder girdles and the pelvic region. The patient may notice difficulty in lifting objects, raising their arms, climbing up stairs or getting up from a sitting position. A difficulty in swallowing can also be seen secondary to involved muscles.

As for the laboratory findings, serum levels of creatinine kinase are elevated in most patients. Aldolase and lactic dehydrogenase are also indicators of active muscle disease. Positive antinuclear antibodies (ANAs) are seen in 60%

to 80% of patients. X-ray studies with barium swallow may show weak pharyngeal muscles and a collection of barium in the pyriform sinuses. Magnetic resonance imaging of the muscles is a way to assess muscle activity of the disease and help for site selection for the muscle biopsy.

In adults, malignant neoplastic disease can be associated with dermatomyositis. The malignant process that precedes it, occurs simultaneously as dermatomyositis or follows the disease. A search for malignancy is directed by history and physical findings. Ovarian cancer is the most common associated neoplasm.

Prednisone is the mainstay of treatment given in doses of around 1 mg/kg daily until the severity of the disease decreases and muscle enzymes are almost normal. The skin lesions may respond to systemic therapy, however, its response is unpredictable and skin disease may persist despite amelioration of muscle involvement. In these cases, hydroxychloroquine at doses of 200 mg to 400 mg per day can be added. Dx

References
1. Kovacs SO, Kovacs SC: Dermatomyositis. J Am Acad Dermatol 1998; 39(6) 899-920.
2. Dawkins MA, Jorizzo JL, Walker Fo, et al: Dermatomyositis: A dermatology-based case series. J Am Acad Dermatol 1998; 38(3):397-404.
3. Zieglschmid-Adams ME, Pandya A, Cohen SB, et al: The value of methotrexate in dermatomyositis. J Am Acad Dermatol 1998; 38(1):130-2.
4. Haftel HM, McCune WJ, Sullivan DB: Risk of cancer in dermatomyositis or polymyositis. N Engl J Med 1992; 327(3):207-8.