Dermatomyositis

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■ Cite as: *CMAJ* 2019 March 25;191:E340. doi: 10.1503/cmaj.180947

A 53-year-old otherwise healthy man presented to the rheumatology clinic with periorbital erythema and a widespread pruritic rash that was aggravated by sun exposure. Over the past year, the patient had received treatment with antibiotics and topical moisturizers to no effect. An examination of his skin showed a heliotrope rash (Figure 1A), shawl sign (Figure 1B) and Gottron papules (Figure 1C). He had proximal muscle weakness and an elevated creatine kinase level (671 [normal range < 200] IU/L. Electromyography showed short polyphasic potentials, fibrillations and repetitive discharges. Screening for malignant disease showed a 3-cm pulmonary nodule. His skin lesions improved following treatment with oral prednisone and methotrexate, and intravenous immunoglobulin, and his pulmonary nodule is being monitored.

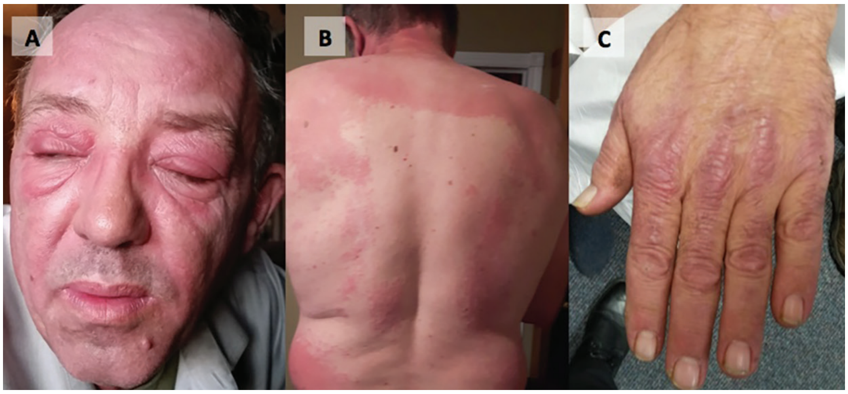

Figure 1: (A) Erythemato-violaceous changes of the eyelids (heliotrope rash), (B) confluent erythematous papules over the upper back (shawl sign) and (C) scaling over the extensor surfaces of the metacarpophalangeal and interphalangeal joints, with involvement of adjoining skin (Gottron papules) in a 53-year-old man with dermatomyositis.

Dermatomyositis is an uncommon idiopathic inflammatory myopathy with skin manifestations that vary in severity. Incidence is estimated at 1 per 100 000 population, with peaks at ages 5–15 years in children and ages 45–60 years in adults.[1] Dermatomyositis rashes are commonly photosensitive; therefore, other diagnostic considerations include subacute cutaneous lupus, contact dermatitis, drug rash and phototoxic reaction.

As proposed by Bohan and Peter in 1975,[2] a definite diagnosis requires 4 or more of the following criteria: symmetric proximal weakness, elevated levels of serum muscle enzymes, characteristic electromyography changes, characteristic muscle biopsy changes, and typical skin lesions that may include scalp dermatosis, heliotrope rash, photosensitive poikiloderma, V sign (rash on anterior neck), shawl sign, Gottron papules or Holster sign (rash over lateral hip). About 10% of patients will have amyopathic dermatomyositis with no apparent muscle involvement, frequently associated with anti-MDA5 antibody. Other manifestations may include arrhythmias, interstitial lung disease and inflammatory arthritis.[3]

Initial therapy usually involves systemic glucocorticoids plus methotrexate or azathioprine. Intravenous immunoglobulin may be beneficial in refractory disease.[4]

In adult-onset disease, risk of malignant disease increases as much as twofold within the first 5 years (especially with anti-TIF1-γ antibody).[5] Adenocarcinomas of the lung, stomach, pancreas and pelvis are most commonly implicated. Although there are no guidelines for screening, scanning using positron emission tomography–computed tomography is being used increasingly. Extensive investigation beyond age-appropriate screening is suggested.[6]

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Competing interests: None declared.

This article has been peer reviewed.

The authors have obtained patient consent.

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