ONSET OF AMYOPHATIC DERMATOMYOSITIS IN CLOSE ASSOCIATION WITH COVID-19: VIRUS TRIGGERED AUTOIMMUNITY

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BACKGROUND

Dermatomyositis (DM) is a relatively rare acquired inflammatory myopathy with an annual incidence of 2 to 9 per million. The leading theory involves an immune-mediated process triggered by external factors (e.g., malignancy, drugs, infections) in genetically predisposed individuals, targeting the skeletal muscles and skin. Dermatomyositis is characterized by the presence of distinctive rashes: the heliotrope sign and Gottron papules. A subset of DM is referred to as clinical amyopathic DM (CADM). These patients have a typical skin rash but no clinical signs of muscle involvement.

CASE REPORT

A 49-year-old white woman was diagnosed with COVID-19 after presenting with fever, dry cough, and sore throat. She had erythematous skin lesions on hands and face, swelling and rash in the periorbital area, photosensitivity, and arthritis of the knees and hands. Dyspnea was absent, and computed tomography (CT) showed minimal lung involvement. After 20 days, the infection symptoms disappeared, and a second COVID-19 RT-PCR test came back negative, although the cutaneous lesions and arthralgia persisted. Six months later, the erythematous to violaceous papules and plaques over bony prominences of the hands and elbows (Gottron papules) and the periungual erythema (Figure 1) was worse, accompanied by cutaneous erosions and ulcerations (Figure 2), erythema of the upper eyelids and edema of the lower lids (heliotrope rash) (Figure 3), oral ulcerations and prominent alopecia. Respiratory symptoms were absent. Laboratory testing showed high erythrocyte sedimentation rate and C-reactive protein, elevated d-dimers (1222 ng/mL), and normal muscle enzymes, C3, and C4. Antinuclear antibody (ANA), anti-dsDNA, anti-SSA/Ro, ANCA, anti-Jo-1, and anti-Mi-2 were negative, and anti-melanoma differentiation-associated-protein-5 (anti-MDA-5/CADM-140) was positive (45U). Chest CT showed minimal residual abnormalities. Capillaroscopy showed architectural derangement, dilated capillaries, enlarged capillary loops, and multiple hemorrhages (Figure 4). Skin biopsy showed hyperkeratosis, parakeratosis, vacuolar degeneration, thickening of the basal layer, dermal changes with interstitial mucin deposition, dilated blood vessels, and perivascular inflammatory lymphocytic infiltrate, consistent with the diagnosis of DM (Figure 5).

Figure 1. Gottron papules.

Figure 2. Cutaneous ulcerations and erosions.

Figure 3. Heliotrope rash.

Figure 4. Periungueal capillaroscopy.

Figure 5. Skin biopsy: A. HE stain; B. PAS stain.

CONCLUSION

An increase in cases of DM associated with COVID-19 has been previously suggested, as well as of other autoimmune diseases. To our knowledge, this is the first reported case of anti-MDA-5 positive CADM associated with COVID-19 that, eight months after the infection, still presents with severe cutaneous manifestations, without myositis or interstitial lung disease. Our patient provides additional evidence that SARS-CoV-2 is prone to trigger autoimmune responses.

KEYWORDS

Dermatomyositis, COVID-19, Autoimmunity.