Our Dermatology Online

Case Letter

Anti-MDA5 dermatomyositis

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Sir,

A 44-year-old male presented to emergency department at Basildon University Hospital with a two-week history of fatigue, fever, severe joint pain primarily in his jaw and shoulders, and progressive loss of muscle strength in his upper and lower body. He reported increasing respiratory distress with difficulty with swallowing. Additionally, he developed a papulovesicular rash on his knuckles, followed by a scaly rash on his hairline and both elbows. Swelling around his eyes (Figs. 1a – 1e) and oral ulceration accompanied the rash.

The patient claimed to have no recent travel history, weight loss, underlying medical conditions, or regular medication usage. During examination, the patient's skin was of Fitzpatrick Skin Type V1 with a resolving papulovesicular rash and ulcerations over the metacarpophalangeal and interphalangeal joints of both hands. He showed painful, keratotic papules on the palmar surfaces of both hands and peri-orbital swelling with a scaly rash on the frontal hairline and elbows (Figs. 1a – 1e). Muscular examination revealed upper and lower limb girdle weakness.

Investigation results showed high levels of serum ferritin, LDH, CK, ALT, AST, positive ANA test, positive MDA-5 antibodies, and low haemoglobin. MRI scan of the thigh showed subtle changes with increased signal in the vastus lateralis muscle bilaterally.

Electromyography was done and it showed evidence of inflammatory myopathy. A muscle biopsy subsequently revealed an increase in HLA-ABC's upregulation and membrane attack complex surrounding capillaries. The changes were highly suggestive of inflammatory myositis.

Pulmonary function tests showed an FVC of 79% and a TLCO of 65% of the predicted reading.

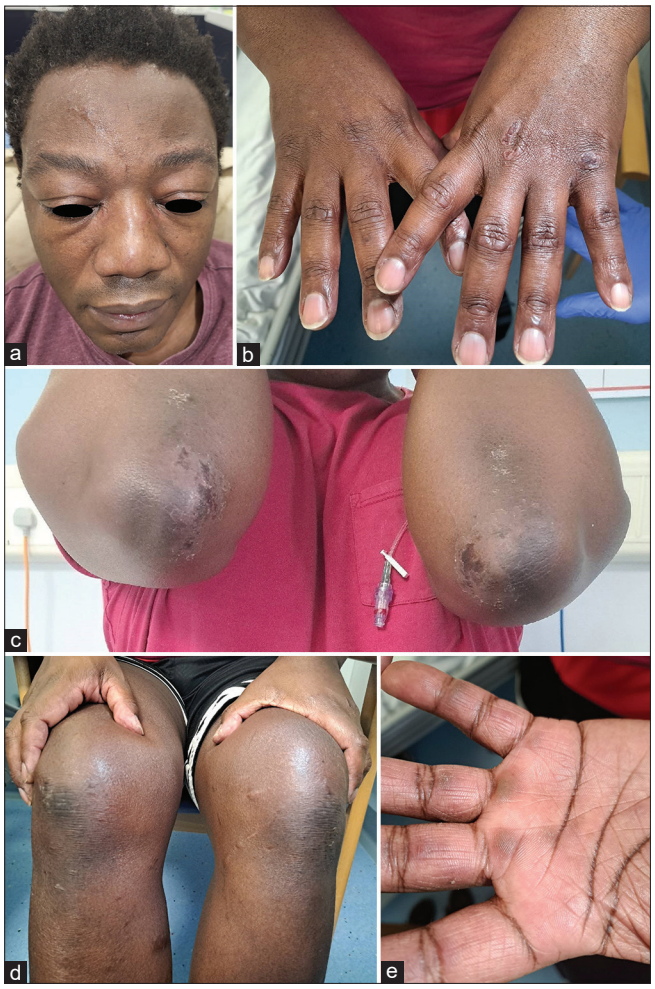

Figure 1: (a) Bilateral peri-orbital swelling (b) Red-violet, slightly raised papules (Gottron papules) on the metacarpophalangeal and interphalangeal joints with healing ulcerations, (c and d) Redness over the back of the elbows and knees (Gottron sign) (e) Painful, keratotic papules on the palmar surfaces of both hands (Mechanic's Hands).

The clinical presentation matched that of Anti-MDA-5 dermatomyositis (previously known as CADM-140). The presence of this antibody has been associated with rapidly progressive interstitial lung disease and high mortality rate especially in the Asian population.

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It also affects the prognosis for the patient who may have a lower likelihood of achieving remission [1,2]. Anti-synthetase antibodies might also be associated with interstitial lung disease, however the course of it is usually slowly progressive as opposed to rapidly progressive interstitial lung disease in Anti MDA5 dermatomyositis [3]. The clinical presentation of Anti-MDA5 dermatomyositis includes skin ulceration which usually occurs at the site of Gottron papules and might include elbows and lateral nailfolds. It is also associated with tender palmar papules (mechanics hands), oral pain and/or ulceration and arthritis [4,5].

The patient received oral prednisolone and mycophenolate mofetil treatment that resulted in a gradual improvement of symptoms, and a decrease in muscle markers including CK, ALT, and LDH. The patient was screened for associated malignancy and his chest, abdomen and pelvis CT scan was unremarkable apart from some basal atelectasis involving both lungs.

This case highlights the importance of certain antibodies in suggesting increased risk of serious complications of dermatomyositis. Patients with anti-MDA5 dermatomyositis need specific attention and monitoring of the pulmonary function tests as they carry important prognostic factors and might be associated with increased morbidity and mortality in patients with dermatomyositis.

Consent

The examination of the patient was conducted according to the principles of the Declaration of Helsinki.

The authors certify that they have obtained all appropriate patient consent forms, in which the patients gave their consent for images and other clinical information to be included in the journal. The patients understand that their names and initials will not be published and due effort will be made to conceal their identity, but that anonymity cannot be guaranteed.

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