## Sweet syndrome in anti-HMGCR immune-mediated necrotising myopathy: expanding the cutaneous spectrum

Sir,

Immune-mediated necrotising myopathy (IMNM) is typically characterised by subacute, severe proximal muscle weakness with markedly elevated creatine kinase levels. Cutaneous involvement is uncommon, and when present usually resembles dermatomyositis (1). Sweet syndrome, an acute neutrophilic dermatosis, has recognised associations with autoimmune diseases, but it has not previously been described in IMNM (2). We report what appears to be the first case of Sweet syndrome occurring in a patient with anti-3-hydroxy-3-methylglutaryl-coenzyme A reductase (HMGCR) IMNM.

A 67-year-old woman presented with a three-day history of painful, violaceous patches over the dorsal aspects of her fingers, accompanied by tender swelling of several small joints. She also reported three months of progressive truncal weakness and difficulty rising from a supine position. Her creatine kinase was 6014 U/L, and electromyography showed an irritable myopathy. Anti-HMGCR antibody levels were strongly positive. Muscle biopsy demonstrated necrosis with myophagocytosis and regeneration consistent with IMNM.

A punch biopsy of the skin lesions revealed a dense upper-dermal neutrophilic infiltrate with red cell extravasation, but without features of vasculitis or interface dermatitis. These findings supported Sweet syndrome, specifically the neutrophilic dermatosis of the dorsal hands variant. There were no medications, infections or systemic features suggestive of drug-induced or malignancy-associated Sweet syndrome. Imaging of the thorax, abdomen and pelvis, along with endoscopic evaluation, did not reveal an underlying cancer.

Treatment with pulsed methylprednisolone followed by rituximab and intravenous immunoglobulin led to resolution of the rash within several weeks and improvement in muscle strength over two months. She has remained in remission on tacrolimus and low-dose prednisolone 18 months after presentation.

To contextualise our observation, we conducted a focused literature review using both PubMed and large language models (LLMs). PubMed identified reports of dermatomyositis-like rashes in IMNM including Gottron-type lesions, periungual erythema and heliotrope changes, as well as erythema-multiforme–like lesions, tinea-versicolour-like eruptions and Jessner lymphocytic infiltrates (3-7). LLM-assisted searches (ChatGPT, Gemini and Grok) identified five additional reports, in particular papers reporting on associations of neutrophilic urticarial dermatosis, and non-specific erythema, plaques and pustulosis (8, 9). However, no published case demonstrated a neutrophilic dermatosis consistent with Sweet syndrome.

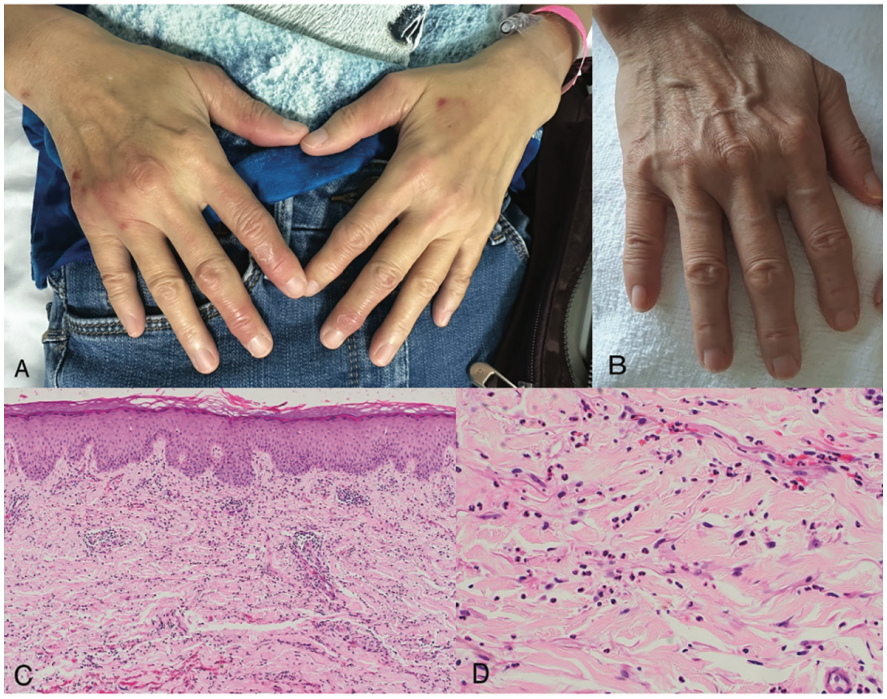


**Fig. 1.** Clinical images and skin biopsy findings. (**A**) Confluent tender violaceous patches over the extensor aspects of the thumb, index and middle fingers. (**B**) Resolution of rash after treatment. (**C, D**) diffuse dermal infiltrate and red cell extravasation without involvement of the epidermis (haematoxylin and eosin, original magnification 100x). The infiltrate is comprised mainly of neutrophils, without associated leukocytoclastic vasculitis (haematoxylin and eosin, original magnification 400x).

The acute, tender plaques in our patient were more consistent with Sweet syndrome than with any recognised IMNM-associated rash. Erythema elevatum diutinum, another neutrophil-rich dermatosis with occasional reported overlap in inflammatory myopathies, was considered but was not supported histologically due to the absence of leukocytoclastic vasculitis. Infective causes were excluded clinically and histochemically. The lack of drug triggers, the absence of malignancy and the timing of onset suggest that Sweet syndrome may represent an atypical dermatological manifestation of IMNM rather than a coincidental process. Pro-inflammatory cytokines like tumour necrosis factor (TNF-α) and interleukin-1 have been implicated in the pathogenesis of both conditions suggesting a potential shared pathway of inflammation, although leading to different inflammatory manifestations in these genetically susceptible individuals (10).

In summary, we report the first case of IMNM-associated Sweet syndrome. Increasing recognition of the diverse dermatological manifestations in IMNM highlights the importance of characterising these rashes to aid in the diagnosis of the condition and differentiating from the classical dermatomyositis phenotype. The use of LLMs has also allowed us to expand the reach of our literature search leading to a more comprehensive review on all relevant studies.

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