Vesicobullae and fever

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DESCRIPTION

A healthy man in his 40s presented with 5 days of vesiculobullous eruption with interspersed targetoid plaques (figures 1 and 2), starting over his chest before extending to the rest of his body. It was preceded by fever, sore throat and diarrhoea. The patient's general practitioner prescribed acyclovir to no effect. No other medication had been commenced and he denied anabolic steroid use. The patient is a teacher and plays waterpolo. He denied other outdoor activities, animal exposure, insect bites or recent travel. Investigations revealed a raised C reactive protein of 285 mg/L, erythrocyte sedimentation rate of 40 mm/hour and white cell count of 21.8×10^9/L, predominantly neutrophilic. He required admission and was started on piperacillin-tazobactam and clindamycin for possible streptococcal/staphylococcal infection. The dermatologists took lesional biopsies for H&E and perilesional biopsies for direct immunofluorescence. A tailing down prednisolone course was also commenced.

Blood cultures, antinuclear antibody, mycoplasma IgM and antistreptolysin O titre were negative as was the viral screen for Varicella, COVID-19, Epstein-Barr virus, cytomegalovirus, hepatitis, HIV and herpes simplex. In view of this and the rapid response to steroids, antibiotics were stopped, and he was discharged after 6 days.

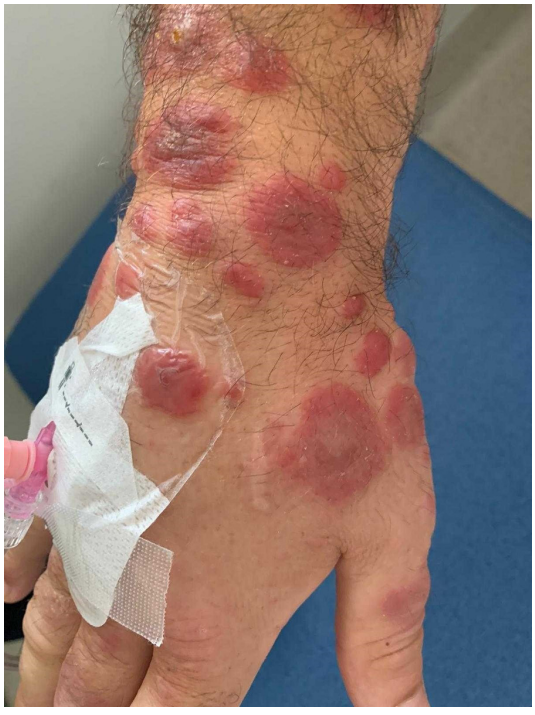

Figure 1 Annular plaques.

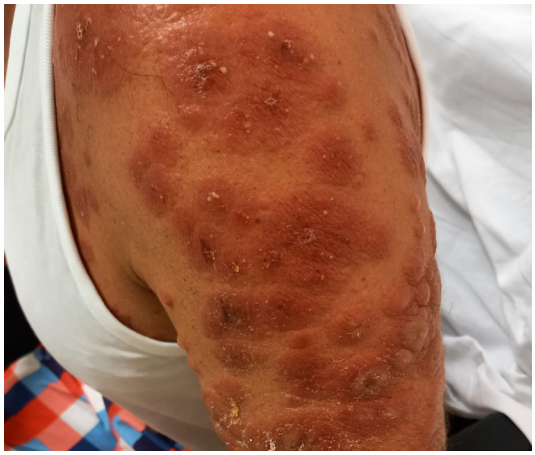

Figure 2 Bullous lesions.

Histology revealed papillary dermis oedema, focal subepidermal blister and dense neutrophilic infiltrate in the upper reticular dermis. There was no evidence of vasculitis and direct immunofluorescence was negative. This was all in keeping with bullous Sweet's syndrome, an acute febrile neutrophilic dermatosis.

Classical Sweet syndrome is characterised by fever, painful skin inflammation and mucosal lesions.[1] The latter were absent in our case and lesions were non-tender, making clinical diagnosis more difficult. However, both clinical and histopathological arms of the revised diagnostic criteria were fulfilled.[2] Differential diagnoses were excluded using the supportive sections of these criteria. These included drug eruptions, infectious disorders, neoplasia such as leukaemia cutis, reactive conditions like erythema multiforme, vasculitis and systemic inflammatory disorders such as lupus.[1]

Sweet syndrome can be associated with underlying illness as well as in previously healthy individuals.[1] It is possible that our case was triggered

Patient's perspective

I was initially very concerned and anxious as the rash was quite dramatic to look at. I was afraid that a diagnosis would not be reached and that was very stressful for me and my wife. However, a few days into my hospital stay I felt calmer as the lesions began to improve and was very happy with the thoroughness with which I was investigated. Ultimately, I was given a diagnosis and that is when I truly felt at ease. The not knowing was the worst part. Now I am undergoing follow-up at the dermatology outpatient clinic and the lesions have nearly completely resolved.

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Learning points

- The diagnosis of Sweet syndrome is difficult to make and easy to overlook as a differential, but it should be considered in all patients with bullous/targetoid lesions.
- Awareness of atypical presentations without characteristic mucosal ulcers and tender lesions, such as in this case, emphasises the above and the importance of a detailed history and examination in formulating a differential diagnosis.
- Investigating the patient for a secondary cause of the skin eruption is of paramount importance to exclude autoimmune, infective and neoplastic processes. However, one should keep in mind that this condition can occur in healthy individuals and in the absence of systemic pathology, when even stress can be a trigger.

by preceding infection, however, respiratory screen and stool culture were negative. Other triggers include pregnancy, sun exposure, vaccinations and medications such as azathioprine.[1]

The classical subtype manifests as papules, nodules or plaques which can develop a vesicular appearance.[1] The extensive nature of skin involvement in our case did not fit with neutrophilic dermatosis of the hands, a rare localised variant.[3] When secondary to malignancy, lesions are likely to be bullous and can ulcerate, mimicking conditions like pyoderma gangrenosum. Pustular and subcutaneous variants also exist. This phenotypical variety is also appreciated histologically where neutrophilic infiltration can extend into adipose tissue in the subcutaneous variant[1] or be replaced by lymphocytic and histiocytoid myeloperoxidase-positive cellular infiltration of the dermis.[4] The latter finding necessitates haematological workup as histiocytoid sweet syndrome is associated with underlying myelodysplasia.[4]

Our patient improved on follow-up. Typically lesions resolve without scarring.[1] Steroids induce faster resolution; however, these require a slow tailed down to reduce relapses which occur in around a third of patients, particularly with underlying illness.[1]

Contributors All authors were involved in the management of the patient and contributed to the writing/editing of this paper. SV, NA, DM and VAF were all involved in the planning of the paper and literature review. VAF and SV wrote up the paper which was then reviewed by NA and DM prior to submission. DM is a dermatology trainee and added further details regarding the condition, particularly the histological findings. NA also wrote up the learning points and helped in the resubmission.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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