The importance of skin biopsies: Sweet syndrome as a differential for 'acute painful red rash' in a patient with normal neutrophils and polymyalgia rheumatica on glucocorticoids

Authors: Tharshini Gerald Rajakulenthiran,[A] Roshan Amarasena,[A] Ruchir Singh,[B] Susan Kelly[B] and SSC Silva[A]

Introduction

Cutaneous lesions and skin rashes are commonly encountered on the acute take in acute medical units. Sweet syndrome (SS) is an uncommon inflammatory disorder, originally described as 'acute febrile neutrophilic dermatosis' by Sweet.[1] There are numerous aetiological associations reported and it can be classified as classical, malignancy-associated or drug-induced.[2] SS is frequently accompanied by leukocytosis, particularly neutrophilia. SS is characterised by the abrupt appearance of painful, oedematous and erythematous papules, plaques or nodules on the skin. This case emphasises the importance of utilising multidisciplinary team members and their skills such as skin biopsy, in order to achieve the right diagnosis and right treatment in a common presenting complaint.

Materials and methods

A 76-year-old woman with polymyalgia rheumatica, large vessel vasculitis, myelodysplasia and polycythaemia rubra vera presented with a painful red rash. Her current treatments included prednisolone 5 mg, hydroxychloroquine and regular venesections. The rash appeared as episodic, painful multiple red spots with a haemorrhagic and bruised appearance. She was otherwise systemically well, with no oral ulcers, joint pain or swelling. On examination she was found to have erythematous painful patches around the shin, foot, calf and a further warm painful lesion on the shoulder resembling erythema nodosum. Her cardiorespiratory and abdominal examination was normal. She was initially treated as having a possible vasculitis flare, and her prednisolone was increased to 15 mg.

Results and discussion

Initial investigations showed mild anaemia, but normal neutrophils, platelets, liver and renal function. Her erythrocyte sedimentation rate (ESR) was 5 mm/h and C-reactive protein 10 mg/L. Although her antinuclear antibodies test was negative, her extractable nuclear antigen screen was incidentally Ro-positive. Her dsDNA, anti-neutrophil cytoplasmic antibodies, cryoglobulins, anti-cyclic citrullinated peptide and rheumatoid factor were negative. Her C3, C4 and serum electrophoresis were normal.

Given the wide differentials for an acute, painful erythematous rash, the persistence of symptoms despite an initial increase in prednisolone and an essentially normal immunology screen, an urgent skin biopsy was arranged to further elucidate whether the rash was vasculitic or a mimic. Dermatologists performed the biopsy, which showed neutrophil and lymphocyte infiltrate with perivascular accentuation and focal leukocytoclasis within the dermis and subcutaneous adipose tissue. There was no evidence of frank vasculitis. These clinical and histological features were in keeping with the clinical impression of SS, a common vasculitis mimic.

She was subsequently started on high-dose prednisolone 40 mg once daily, to which she had a good response and did not relapse.

Conclusion

Acute erythematous rashes are a common in secondary care, and anecdotally result in diagnostic conundrums. This case highlights the importance of identifying the need for dermatology input and skin biopsy in diagnosing challenging cases. In this patient, a skin biopsy proved beneficial in excluding vasculitis. Furthermore, the biopsy provided evidence for the diagnosis of SS despite the absence of raised peripheral neutrophils. General physicians may benefit from greater dermatology in-reach advice and access to skin biopsy techniques in order to improve patient outcomes, as shown in this case. ■

Conflicts of interest

None declared.

References

1 Sweet RD. An acute febrile neutrophilic dermatosis. *Br J Dermatol* 1964;76:349–56.
2 Heath MS, Ortega-Loayza AG. Insights into the pathogenesis of Sweet's syndrome. *Front Immunol* 2019;10:414.

Authors: [A]The Robert Jones and Agnes Hunt Orthopaedic Hospital, Oswestry, UK; [B]Royal Shrewsbury Hospital, Shrewsbury, UK