Necrotising Vasculitis: An Aggressive Mimicker of Necrotising Fasciitis

Muhammad Azri MM[1], Idris AB[1], Mustaqim A[1], Naveen V[1], Arieff AA[1]

[1]Department of Orthopaedic, Hospital Tawau, Sabah, Malaysia

INTRODUCTION:

Necrotising vasculitis is a very rare occurrence. We describe an interesting case of a young man presented with skin lesions that mimics the presentation of necrotizing fasciitis.

REPORT:

A 33-year old male with no premorbid illness presented with acute episode of dyspnoea, accompanying by swelling with skin lesions over the both ankles and both arms for one day. He has history of consuming unknown traditional medication, and recently ventured into tropical rainforest. There was no history of trauma or penetrating wound injury.

Examination revealed multiple erythematous blisters over anterolateral aspect of both ankles, and over lateral aspect of both arms. The blisters rapidly worsening, but the underlying compartment was soft. There was no crepitus present.

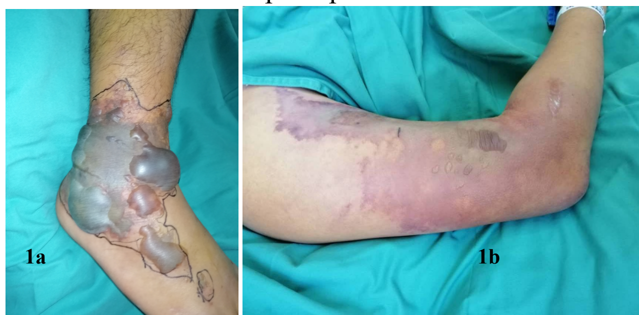

Figure 1a : Excessive blisters over the anterolateral aspect of right ankle. **1b** : Blisters formation over the anterolateral aspect of right arm. On both upper limbs and lower limbs, the underlying compartments are soft.

Laboratory investigations revealed elevated total white cell count with thrombocytopenia; deranged renal and liver function; and severe metabolic acidosis. Ultrasonogram of the right arm and ankle found only subcutaneous oedema with no focal collection or presence of abscess.

Patient's condition rapidly deteriorated as he developed jaundice and shock, and he was intubated for ventilatory support. An initial diagnosis of sepsis with necrotising fasciitis was dispelled by a negative probe test, whereby the underlying fascia was found to be healthy. Blood culture later yielded growth of Streptococcus pyogenes. However, the rapid progression and systemic manifestation rendered a suspicion of underlying autoimmune disorder.

Further work-up revealed positive classic anti-neutrophil cytoplasmic antibodies (C-ANCA) and low complement-3 (C3) level. A diagnosis of necrotising vasculitis with renal-pulmonary syndrome was established. The patient was started with hydrocortisone, and was also covered with vancomycin.

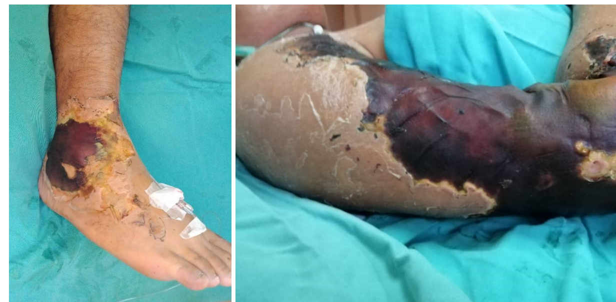

Figure 2 : All skin lesions resolved gradually, leaving only necrotic patches without the need for surgical debridement.

Without surgical debridement, commencement of both medications rapidly improved patient's condition. The skin lesions gradually resolved, leaving superficial dry necrotic patches.

CONCLUSION:

Multiple skin blisters are commonly associated with deep, aggressive infection and may typically present in necrotising fasciitis. However, it is uncommon to have necrotising fasciitis that involve multiple limbs. Streptococcal infection usually will present with scarlet rashes or erysipelas, rather than necrotising blisters. It is therefore worth to explore other possibility when facing with such an unusual presentation. Cutaneous necrotising vasculitis, including ANCA-associated vasculitis (AAV) often accompanied with multisystem involvement as shown in this case. Unlike necrotising fasciitis, avoidance or delaying surgical intervention may help in reducing mortality and morbidity in such patients.

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