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Beyond Expectations

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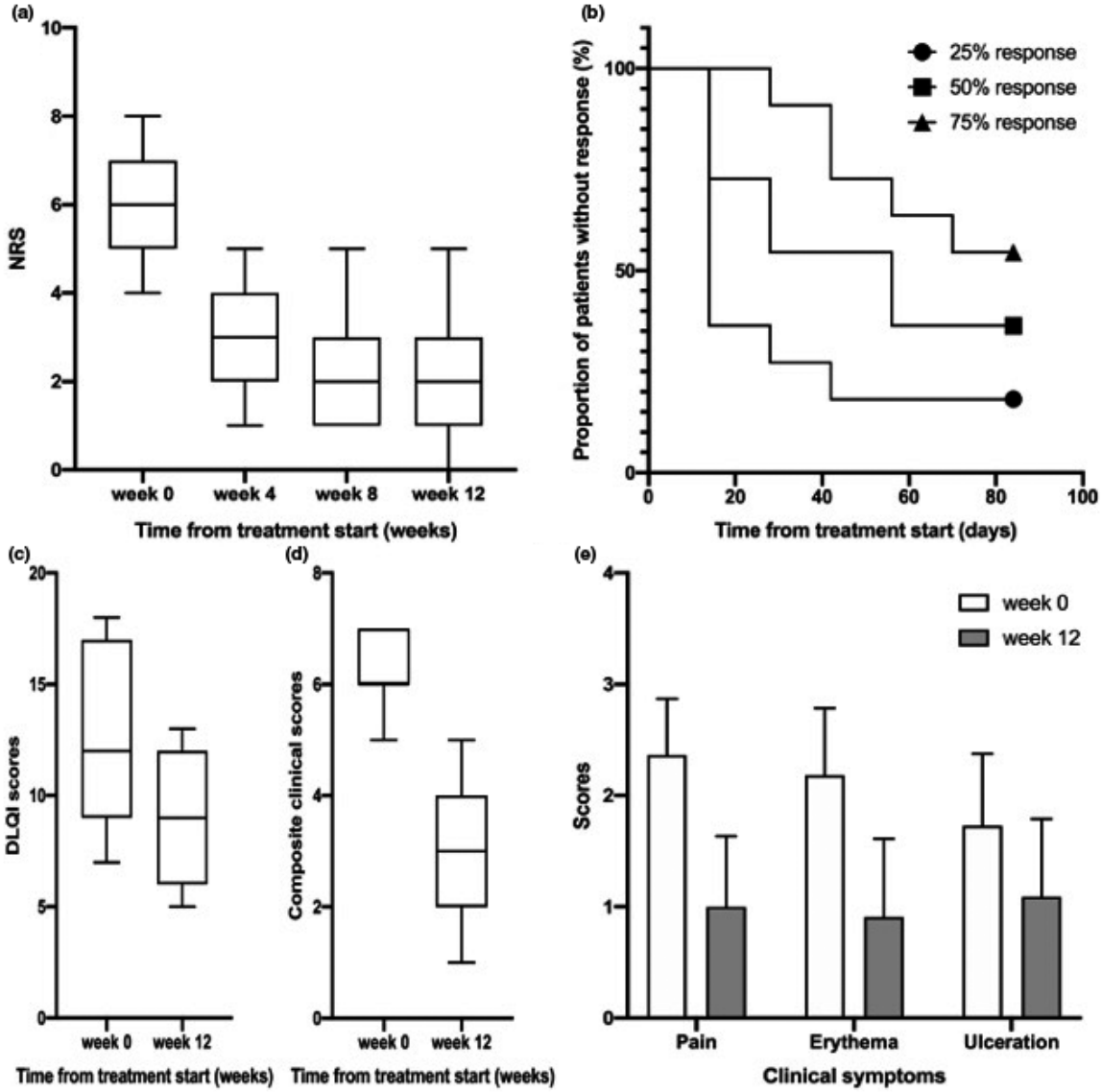

Figure 2 (a) NRS scores during the indicated weeks of anti-tumour necrosis factor (TNF) therapy; (b) responses corresponding with 25%, 50% and 75% reductions in pain according to NRS scores; (c) DLQI scores in Weeks 0 and 12 of anti-TNF therapy; (d) composite clinical scores in Weeks 0 and 12 of anti-TNF therapy; (e) distribution of pain, erythema, and ulceration scores in composite clinical scores during Weeks 0 and 12 of anti-TNF therapy.

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Acute haemorrhagic oedema-like eruption in an adult

Editor

A 49-year-old woman, with no particular medical history other than quiescent Crohn's disease, was admitted for acute gastroenteritis. Three days after admission, she developed large purpuric, ecchymotic targetoid skin lesions and oedema,

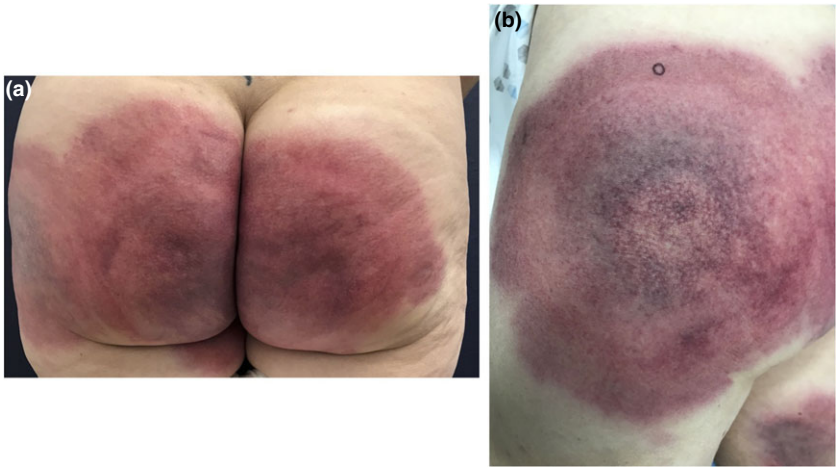

Figure 1 (a and b) Large purpuric, ecchymotic targetoid skin lesions and oedema, predominantly on the buttocks and thighs.

predominantly on the buttocks and thighs (Fig. 1a,b). The lesions were not pruritic but slightly painful at palpation. She denied any history of vaccination or injections at the sites of the skin lesions. The patient remained afebrile. Platelet counts, coagulation tests and immunoglobulin A serum levels were within normal limits. Renal function was normal, and proteinuria was excluded. Bacteriological stool samples were negative.

The gastrointestinal symptoms were attributed to viral enteritis. Skin biopsy specimens showed a dense, superficial and deep, perivascular infiltrate mainly composed of neutrophils and nuclear debris. Fibrinoid thickening of vessel walls and extravasated erythrocytes were also observed (Fig. 2a), with C3 deposits on blood vessels on direct immunofluorescence (Fig. 2b). Given the clinical presentation, the histopathological image of leukocytoclastic vasculitis and the occurrence shortly after a viral infection, a diagnosis of Acute Haemorrhagic Oedema (AHE)-like eruption in an adult was made. The enteritis resolved spontaneously within 10 days and skin lesions disappeared, without relapse, after three days of 60 mg intravenous methylprednisolone.

AHE, also known as Seidlmayer purpura, is a rare and benign leukocytoclastic vasculitis, typically described in young children and usually triggered by infections, drugs or vaccination.[1] The main clinical characteristics are symmetrically purpuric medallion- or cockade-shaped patches, associated with painful oedema and predominantly located on cephalic extremity or limbs. Although cutaneous presentation may be dramatic, in children, disease remains non-toxic, visceral involvement is rare and the disease is self-remitting within a few days.[2] In most cases, laboratory findings are non-specific.[3] Initially interpreted as a variant of Henoch–Schönlein purpura, AHE is now considered a distinct entity based on clinical, histopathological and prognostic differences.[2] Treatment of AHE remains controversial, and because it generally resolves spontaneously, a conservative approach should be considered.[4] We decided to treat our patient in view of the severe histological damage.

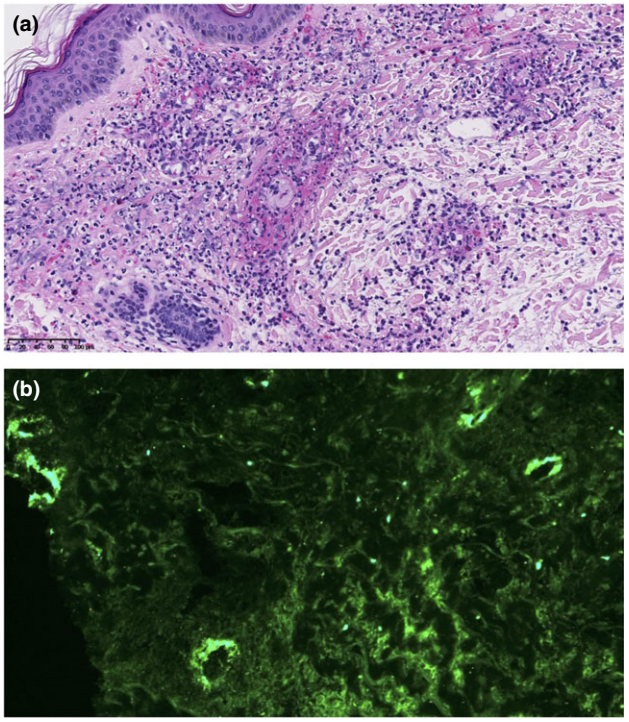

Figure 2 (a) Histopathological features showing a dense, superficial and deep, perivascular infiltrate mainly composed of neutrophils and nuclear debris. Fibrinoid thickening of vessel walls, as well as extravasated erythrocytes haematoxylin and eosin; original magnification ×20. (b) Direct immunofluorescence showing C3 deposits on blood vessels.

Ecchymotic urticaria was ruled out as lesions were non-pruritic and non-labile (persistence of lesions for more than 24 h in the same place). Even though the histopathological features are similar, urticarial vasculitis was excluded as the haemorrhagic presentation clearly predominated and is not usually typical of this affection.

To our knowledge, this is the first case describing the AHE pattern in an adult, probably precipitated by viral gastroenteritis. There is no explanation as to why this condition is only described in children.

The diagnosis of AHE was a real challenge, especially since this entity is described as a paediatric pathology, and diagnosis was only made possible through good clinical and pathological correlation.

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The patient in this manuscript has given written informed consent to publication of their case details.

Conflict of interest

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The role of genetic factors and peripheral immune cells in SAPHO syndrome

Editor

Sir, synovitis, acne, pustulosis, hyperostosis and osteitis (SAPHO) syndrome includes a spectrum of heterogeneous diseases. The pathogenesis of SAPHO syndrome is multifactorial. Our case elucidates the relevant role of genetic factors and immunity cells in pathogenesis.

An 18-year-old boy complained of acne conglobate and chest pain. One month ago, lesions initially appeared as erythema papule and infected cysts with soles were evident on face. Then, he suffered from fever (>37.5) and severe pain on the chest, neck and limb. The patient was too sore to move. High muscle tension was found in the upper and lower limbs. Grade 4 muscle strength was observed. Lumbosacral neuritis was misdiagnosed. Finally, a diagnosis of SAPHO syndrome was made. No underlying disease or family history was detected. Clinical examination revealed painful inflammatory nodules and abscesses affecting the face and central chest (Fig. 1a–d). Laboratory assays found elevation in the levels of serum leukocyte count (20.75×10^9/L), percentage of leukocyte (84%) and hypersensitive C-reactive protein (61 mg/L). Liver function was normal. Magnetic resonance imaging (MRI) of cervicale detected C3/4, C4/5 and C5/6-disc herniation, but the results of computerized tomography and MRI of thoracic vertebra were normal. The patient was administered 25 mg etanercept twice weekly for 1 month and 25 mg once weekly for 5 months, and these treatments resolved the problem (Fig. 1e–h). We encountered the following genetic variants: rs11209026, rs7517847 and rs11465804 of IL-23R; rs3819025 of IL-17A; rs763780 of IL-17F; rs324011 of STAT6; rs1800795 of IL-6; rs1805010 of IL-4R; rs2243248 of IL-4; rs6887695 of IL-12B; and rs733618 and rs5742909 of cytotoxic T-lymphocyte-associated protein 4 (Fig. 1i–t, Table 1). In our case, mass cytometry (CyTOF) determined that the proportion of monocytes, $CD4^+$ T cells and B cells decreased, while the proportion of dendritic cells and γδT cells increased in the treated phase compared with the untreated phase (Fig. 1u–x).

SAPHO syndrome has osteoarticular and dermatological manifestations. The mechanisms are unclear. Genetic factors have been investigated due to family-based investigations, but genetic studies revealed no role for HLA-B27, HLA-Cw6 or HLA-DR, PSTPIP2, LPIN2 and NOD2.[1,2] Variants rs10889677, rs2201841, rs7517847 of IL-23R and rs2243248 of IL-4 showed strong associations with SAPHO syndrome.[3] Our results for rs7517847 of IL-23R and rs2243248 of IL-4 were similar to Guo's results. However, more studies are required to further explore our findings.

Immunological dysfunction may play a crucial role. Orru *et al*. found that $CD4^+$ Th17 lymphocytes might have a relevant role in immunopathogenesis.[4] Firinu *et al*. detected the activation of Th17 axis has been found in patients affected by SAPHO syndrome with protracted course and can be observed in subjects treated with different drugs.[5,6] However, Xu revealed that the absolute counts of NK cells were significantly reduced and an imbalance of Th17/Treg cells was found in SAPHO patients compared with the controls. A reduction in peripheral NK cells may exacerbate the disease progression.[7] This is the first case in which peripheral immune cell subsets were detected by CyTOF. The role of monocytes, $CD4^+$ T cells and B cells needs to be elucidated. Dendritic cells and γδT cells may show a reactive rise.

For SAPHO patients who do not respond to conventional treatments, such as nonsteroidal anti-inflammatory drugs and

