

Bullous Sweet's syndrome in rheumatoid arthritis after streptococcal pharyngitis

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Fig. 1. Bullous Sweet's syndrome with skin and lung involvement. (a) Bullous lesions on the dorsal aspect of the left hand. (b) Bullous lesions on the oral mucosa. (c) Bullous lesions on the shoulder and scapular area. (d) Chest CT showing a 'crazy-paving' pattern consistent with organising pneumonia.

KEYWORDS: bullous Sweet's syndrome, rheumatoid arthritis, streptococcal pharyngitis, organising pneumonia

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Case presentation

A 35-year-old woman presented with a 2-week history of a painful rash, characterised by multiple well-circumscribed circular bullous lesions, with a purulent or haemorrhagic fluid, some ulcerated or leaking, affecting first hands, forearms and mouth, but rapidly spreading through her entire body surface (Fig 1a–c). This was accompanied by malaise, fever and diffuse joint pain and

swelling, and was preceded by a sore throat. She had seronegative rheumatoid arthritis, previously treated with sulfasalazine, which she had recently voluntarily stopped. Blood testing showed elevation of erythrocyte sedimentation rate (60 mm/h) and C-reactive protein (205 mg/L), and a positive antistreptolysin O. Skin biopsy revealed a neutrophilic dermatitis with oedematous bulla formation, consistent, in this clinical setting, with bullous Sweet's syndrome (SS). Chest computed tomography (CT) was performed, excluding malignancy but showing a 'crazy-paving' pattern consistent with organising pneumonia (Fig 1d). The patient started methylprednisolone and rituximab with full resolution of the bullous rash, polyarthritis and lung involvement.

Discussion

Bullous SS is a rare variant of the classical SS, or acute febrile neutrophilic dermatosis.¹ The bullous variant is most frequently described in the context malignancy, certain drugs and inflammatory bowel disease, and is rarely seen in rheumatoid

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arthritis.² This patient's currently untreated rheumatoid arthritis could have provided the clinical predisposition for this severe form of bullous SS after only a mild streptococcal pharyngitis. Furthermore, organising pneumonia is an uncommon and probably under-recognised but potentially life-threatening association with SS,³ which, in this case, fully responded to treatment with steroids and rituximab. These images remind physicians of the variable presentations and severity of SS while at the same time raising awareness for the possible triggers and underlying predisposing conditions. ■

References

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