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Skin ulcer presentation of Wegener's granulomatosis

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DESCRIPTION

A 49-year-old male presented with a 6-week history of a single ulcer (3 cm diameter) on his left leg some 8 cm above the lateral malleolus. The 4 cm ulcer had a well-demarcated edge and the base appeared mildly infected (figure 1A). His only additional complaint was of intermittent musculoskeletal pain for the previous 4 months. The pain involved the thoracic cage, cervical spine, hands, shoulders and knees. Physical examination was normal except for the presence of the ulcer. Features of leukocytoclastic vasculitis were observed in a biopsy of the ulcer. Autoantibody serology was strongly positive for cytoplasmic antineutrophil cytoplasmic antibody (C-ANCA) and anti-PR3 autoantibodies. A diagnosis of Wegener's granulomatosis was made and he responded to oral cyclophosphamide and prednisolone (figure 1B). He remained in complete remission for 4 years and then re-presented with a systemic pyrexial illness and associated ear and eye inflammation. A chest x-ray showed an opacity in the mid-zone of the right lung. A positive C-ANCA test was again noted. Based on his previous presentation, the current clinical picture and the positive

autoantibody serology, a diagnosis of Wegener's granulomatosis in relapse was made. He was treated again with immunosuppressive drugs and 4 years later (April 2010) remains well, off all therapy.

This case demonstrates how Wegener's granulomatosis can present in an atypical manner and the variable nature of this disorder.^{1 2} The value of the C-ANCA investigation is also highlighted, since in the absence of a positive autoantibody test, it is doubtful that the correct diagnosis would have been made at first presentation.

Competing interests None.

Patient consent Obtained.

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Figure 1 (A) Well-demarcated 4 cm ulcer was found above the lateral malleolus of the left leg with almost complete healing of the lesion after immunosuppressive therapy (B).

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