Immune complex vasculitis: a rash that cannot be missed

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DESCRIPTION

A 43-year-old woman with a medical history of chronic obstructive pulmonary disease, type II diabetes mellitus, hypertension, irritable bowel syndrome, osteoarthritis, epilepsy and methamphetamine use presented to the emergency department with a diffuse erythematous maculopapular rash with palpable purpura (figure 1A). Patient endorsed the rash had been present 3 days prior to presenting to the emergency department. The rash initially only involved her ankles before migrating to her thighs, buttocks, abdomen, wrists and hands. She described it as pruritic, burning and painful. The patient also described significant myalgias and arthralgias. Vital signs on arrival: temperature of 37.1°C, heart rate of 102 beats/min, respiratory rate of 20 breaths/min, blood pressure of 146/75 mm Hg and SpO₂:98% on room air. Laboratories were significant for a white cell count of $11.7 \times 10^9/L$, haemoglobin of 112 g/L, erythrocyte sedimentation rate of 38 mm/hour and a C-reactive protein level of 5.8 mg/dL. Given the patients presenting symptoms and associated rash, there was concern for immune complex vasculitis; with confirmation of leucocytoclastic vasculitis (LCV) on skin biopsy. Investigation into the aetiology of the patient's LCV was negative for HIV, causative medications, antiphospholipid syndrome and systemic lupus erythematosus (SLE). Further infectious workup only revealed positive hepatitis C antibodies but with a negative viral load. Oral glucocorticoids were initiated, and pain control was achieved with opiates and non-steroidal anti-inflammatories. The patient's systemic symptoms along with her rash

Learning points

- ▶ The rash of immune complex vasculitis most commonly presents with painful, palpable purpura starting in the lower extremities.
- ▶ Common causes of leucocytoclastic vasculitis (LCV) are infections (hepatitis C virus and HIV), autoimmune disorders (Sjogren syndrome and systemic lupus erythematosus), medications, malignancy and vascular disorders.
- ▶ When LCV is limited to the skin, management is supportive while corticosteroids and immunosuppressive agents are typically reserved for systemic involvement.

demonstrated rapid improvement 24 hours after initiation of glucocorticoids (figure 1B).

Immune complex vasculitis is a small-vessel vasculitis that typically presents with palpable, burning purpura most often found in the lower extremities. Further study of palpable purpura can be accomplished by using diascopy (pressing a glass slide against a cutaneous lesion).¹ Immune complex vasculitis can present with systemic involvement; affecting joints, muscles, the gastrointestinal tract and renal system. Immune complex vasculitis can be idiopathic (45%–55%), or secondary to autoimmune disorders, medications, infections such as HIV or hepatitis C and malignancy.² Workup for immune complex vasculitis with associated LCV should include basic laboratories in addition to inflammatory markers. Additional studies should be focused on identifying an underlying aetiology for the LCV; including workup for SLE, Sjogren's syndrome, antiphospholipid syndrome, HIV, hepatitis C and malignancy. The diagnosis of LCV is confirmed by histological evidence of perivascular and vascular damage by infiltrating leucocytes as well as fibrinoid necrosis on biopsy.^{3,4} If LCV is limited to skin involvement, management includes rest, ice, elevation of the legs and addressing the underlying cause. When there is systemic disease, treatment includes corticosteroids with consideration for other immunosuppressive therapies as well. Prognosis is more favourable for those with cutaneous-limited disease rather than those with systemic involvement, with most cases resolving in weeks to months.⁵ Clinicians should be familiar with immune complex vasculitis and its associated dermatological findings. Workup should be focused on identifying the precipitating cause, and treatment is focused around supportive management

Figure 1 (A) Lower extremities with red to purple macules and papules, some of which are coalescing into larger patches and plaques. (B) Lower extremities demonstrating marked improvement of rash after initiation of corticosteroids.

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with the addition of immunosuppressive therapies in systemic disease.

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