Purpuric lesions: what diagnosis?

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DESCRIPTION
A previously healthy 9-month-old baby was admitted to the emergency department, with a sudden onset of non-itching rash without fever. He had displayed flu-like symptoms the previous week. There was no recent ingestion of drugs and no recent vaccinations.

At admission, he was afebrile, with excellent overall health, heart rate 106 bpm, blood pressure 87/39 mm Hg, capillary refill time <2”. On skin examination, he had multiple annular purpuric lesions in the lower limbs and non-pitting oedema of the feet (figure 1). The blood count, coagulation profile, C reactive protein, renal function and urinary dipstick were normal. Furthermore, blood and urine cultures were negative. The symptoms resolved completely in 2 weeks, with symptomatic treatment. The clinical diagnosis of acute haemorrhagic oedema of infancy was established.

This is an uncommon disease (there are about 300 cases reported), which typically occurs in children <2 years. The aetiology remains unknown and the onset is sudden. It is an antineutrophil cytoplasmic antibody-negative leucocytoclastic vasculitis, characterised histologically by inflammation of the small dermal vessels, with fibrinoid necrosis and extravasation of red blood cells, and clinically by large, annular, purpuric lesions predominantly over the face, ears and extremities, often with non-pitting oedema of the extremities, ears and face. The exuberance of the lesions contrasts with a well-appearing child and with the benign course of the disease, which resolves spontaneously in 1–3 weeks. The differential diagnosis includes meningococcemia, erythema multiforme, haemorrhagic urticaria, drug eruption and child abuse. Whether or not this disease is a mild variant of Hennoch-Schönlein purpura remains controversial.1–3

Learning points
▸ Acute haemorrhagic oedema of infancy typically occurs in the first 2 years of life.
▸ The exuberance of skin lesions contrasts with a well-appearing child and with the benign course of the disease.
▸ The clinical diagnosis of this entity may avoid elaborate or invasive investigations.

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REFERENCES