Henoch-Schönlein purpura that wasn't!

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Abstract

We present an interesting case of a 13 year old girl with Wegener's granulomatosis/granulomatosis with polyangiitis, who was misdiagnosed with Henoch-Schönlein purpura. The case highlights several important learning points for paediatric practice.

Case Report

A 13 year old girl presented to the Emergency Department (ED) following a one week history of non-blanching rash to the dorsum of both feet (with subsequent spread to legs, trunk and face). Urinalysis revealed 3+ blood and 3+ protein. Henoch-Schönlein purpura (HSP) was diagnosed. The girl was discharged with a one week ward review appointment, at which repeat urinalysis remained unchanged. Blood tests were performed showing raised inflammatory markers and low haemoglobin (8.6 g/dL). An ulcer developed on the right foot (Figure 1). Three subsequent hospital reviews precipitated involvement of the community tissue viability team. Haemoglobin dropped to 7.9 g/dL; haematuria and proteinuria persisted. Prior to initial ED presentation, a General Practitioner-initiated ENT referral was made following a one year history of blocked nose, frequent epistaxis and poor appetite. Unfortunately this was not appreciated when the diagnosis of HSP was made. The girl was independently seen by the ENT team 27 days after the initial ED consultation. Nasal crusting was identified and granulomatosis with polyangiitis (Wegener's granulomatosis) diagnosed. Around the time of diagnosis blood urea and creatinine were raised (14.2 mmol/L and 133 umol/L respectively) and urine protein/creatinine ratio indicated significant proteinuria [163 mg/mmol of creatinine (<15 mg/mmol of creatinine)]. Cytoplasmic anti-neutrophil cytoplasmic antibodies (ANCA) was positive [anti-PR3 >100 U/mL (0-5), anti-MPO <5 U/mL (0-5)]. Pulmonary function testing revealed a mild restrictive defect and chest X-ray showed patchy alveolar opacification of nodular appearance in both perihilar regions. Renal biopsy performed soon after diagnosis showed 50% glomeruli obsolescence with 60% of remaining glomeruli showing crescents at various stages of development; the biopsy revealed a pauci-immune necrotising vasculitic process affecting 80% of glomeruli. A skin biopsy (of a purpuric lesion on the arm) showed non-specific dermal scarring.

Intravenous methylprednisolone and cyclophosphamide were given. The child remained well throughout, experiencing only slight shortness of breath on exertion. Renal function returned to normal over a six week period and anti-PR3 ANCA levels gradually returned to normal after 18 months.

Discussion

HSP is the commonest form of childhood vasculitis (annual incidence 20 per 100,000).[1] Granulomatosis with polyangiitis is rare with an annual incidence of around 3-6 per million (although incidence data is limited).[2] Most cases of HSP spontaneously resolve without long-term sequelae, and HSP is often therefore termed a *benign* vasculitis. However, it is important to remember that other childhood vasculitic disease is not benign, and delay in diagnosis is likely to adversely affect outcome.

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Key words: Henoch-Schönlein purpura, Wegener's granulomatosis, granulomatosis with polyangiitis.

Conflict of interests: the authors report no potential conflict of interests.

Received for publication: 3 August 2011.
Revision received: 20 April 2012.
Accepted for publication: 15 May 2012.

Rheumatology Reports 2012; 4:e8
doi:10.4081/rr.2012.e8

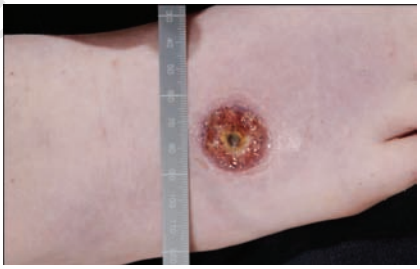

Figure 1. Ulcer to dorsum of foot.

References

1. Gardner-Medwin JM, Dolezalova P, Cummins C, et al. Incidence of Henoch-Schonlein purpura, Kawasaki disease, and rare vasculitides in children of different ethnic origins. Lancet 2002;360:1197-202.
2. Grisaru S, Yuen GW, Miettunen PM, Hamiwka LA. Incidence of Wegener's granulomatosis in children. J Rheumatol 2010;37:440-2.