Chapter 39
Henoch-Schönlein Purpura

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Definition

Henoch-Schönlein purpura (HSP) is the most common systemic vasculitis in childhood of small-sized blood vessels, resulting from an IgA-mediated inflammation. It is typically seen in school-aged children, with an estimated incidence of 13.5–18 per 100.000 children per year. HSP is more common in autumn and winter and often preceded by an upper respiratory tract infection. It mainly affects the skin, gastrointestinal tract, joints and kidney. Petechiae and palpable purpura, the typical skin lesions, involve the lower extremities and buttocks and rarely the upper extremities, face and trunk (Figs. 39.1, 39.2, 39.3, 39.4, and 39.5). The purpuric rash is a mandatory criterion to the diagnosis although may not be the presenting feature of the disease. Subcutaneous oedema over the dorsum of hands, feet, forehead and around the eyes may be also observed. Skin lesions appear on crops and may often recur. Arthralgia or arthritis may precede the purpura by 1–3 days with the ankles, knees and wrists mainly affected. Abdominal pain, gastrointestinal bleeding, haematemesis and melena develop in two-third of children and often precede the purpuric lesions. Vasculitis of the bowel may provoke intussusceptions and gut perforation. One-third of children will have glomerulonephritis but only 10 % nephritic or nephrotic syndrome, hypertension or renal failure. Isolated microhaematuria and/or proteinuria is common within the first 4–6 weeks and can continue for months though urinalysis generally normalizes in a few weeks. HSP is a benign self-limiting condition that lasts 2–4 weeks; only one-third of patients may have a recurrent disease which usually subsides within 3–6 months. Longer course has been reported in patients with renal involvement. Only 1–3 % of cases reach end-stage renal

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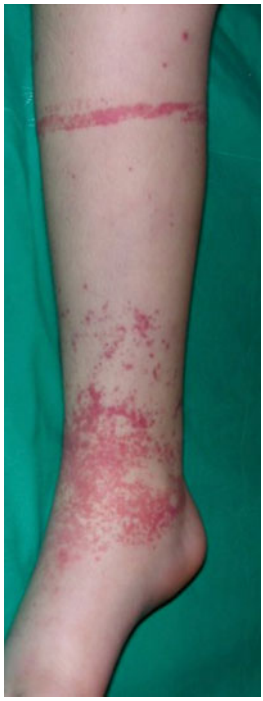


Fig. 39.1 Palpable purpura lesions involve the leg; there is also the sock's sign

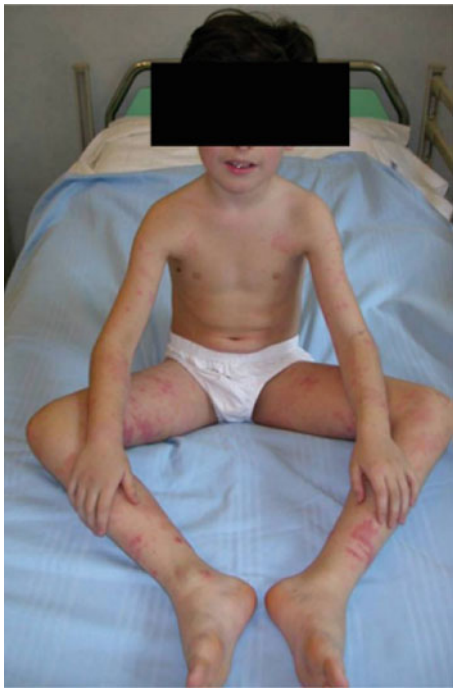


Fig. 39.2 A boy with petechiae, ecchymoses and palpable purpura involving the upper and lower extremities, arms and hands

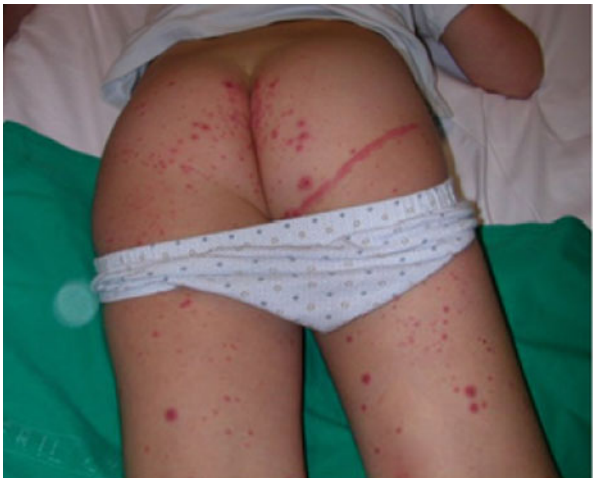


Fig. 39.3 Typical palpable purpura on the buttocks

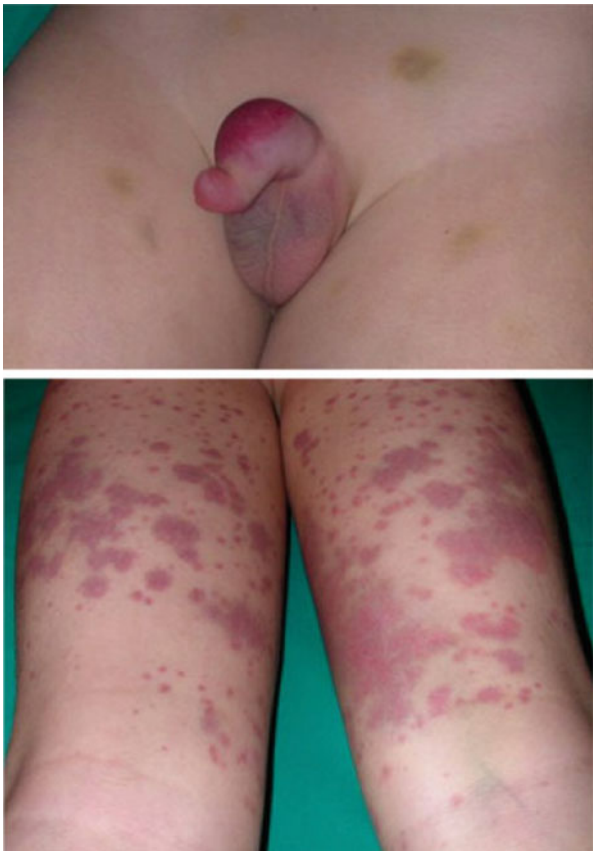


Fig. 39.4 Ecchymoses and purpura of the scrotum; the penis is swollen and ecchymotic. Different skin lesions, ranging from palpable purpura and petechiae to ecchymoses, on the legs

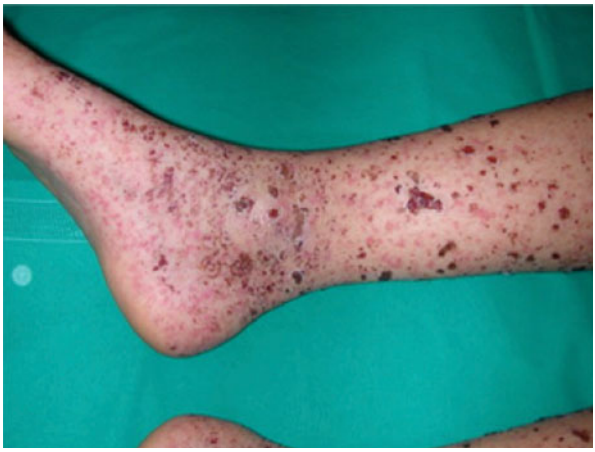


Fig. 39.5 Bullous evolution of purpuric lesions; a rare awful evolution of palpable purpura in an 8-year-old girl with HSP

disease. There is no specific treatment of HSP. No significant benefit of short-course prednisone given at HSP onset has proved to be helpful in preventing persistent renal disease. According to PRES/EULAR criteria, at least one of the following four may be present in addition to palpable purpura: (i) diffuse abdominal pain, (ii) any biopsy showing predominant IgA deposition, (iii) arthritis or arthralgia and (iv) renal involvement (any haematuria and/or proteinuria) [1–3].

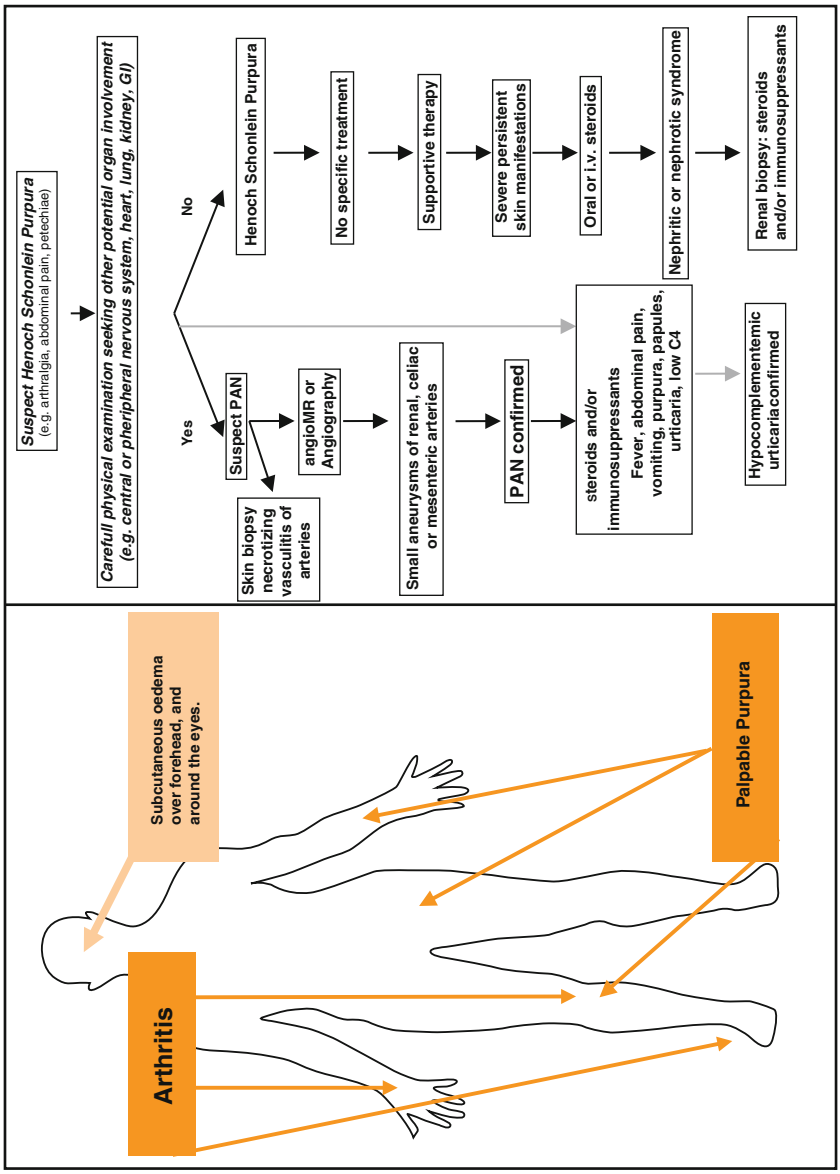
Differential Diagnosis

HSP may be differentiated from other systemic vasculitis in which purpura is the skin manifestation. They include panarteritis nodosa, hypersensitivity angiitis, hypocomplementemic urticarial vasculitis and vasculitis associated with connective tissue diseases. As purpura is the first symptom of other vasculitis, the diagnosis may be missed when the accompanying features have not yet developed. A careful evaluation of blood tests and visceral involvement may help in the correct definition of the underlying disease.

Biopsy

Skin biopsy usually shows a leukocytoclastic vasculitis in the dermal capillary and postcapillary venules with the presence of perivascular infiltrates of IgA. Immunofluorescence confirms the presence of IgA, and in addition of IgG, fibrin,

and complement around the small vessels of involved and uninvolved areas. Renal biopsy shows proliferative glomerulonephritis with features of focal and segmental lesions up to severe multiple crescents. Renal biopsy may be indistinguishable from IgA nephropathy.


Suspect Henoch Schonlein Purpura
(e.g. arthralgia, abdominal pain, petechiae)
Carefull physical examination seeking other potential organ involvement
(e.g. central or pheripheral nervous system, heart, lung, kidney, GI)
Yes
No
Suspect PAN
Skin biopsy necrotizing vasculitis of arteries
angioMR or Angiography
Small aneurysms of renal, celiac or mesenteric arteries
PAN confirmed
steroids and/or immunosuppressants
Fever, abdominal pain, vomiting, purpura, papules, urticaria, low C4
Hypocomplementemic urticariaconfirmed
Henoch Schonlein Purpura
No specific treatment
Supportive therapy
Severe persistent skin manifestations
Oral or i.v. steroids
Nephritic or nephrotic syndrome
Renal biopsy: steroids and/or immunosuppressants
Subcutaneous oedema over forehead, and around the eyes.
Arthritis
Palpable Purpura

References

1. Chartapisak W, et al. Prevention and treatment of renal disease in Henoch-Schönlein purpura: a systematic review. Arch Dis Child. 2009;94:132–7.
2. Eleftheriou D, Brogan PA. Vasculitis in children. Best Pract Res Clin Rheumatol. 2009;23: 309–23.
3. Gardner-Medwin JM, et al. Incidence of Henoch-Schönlein purpura, Kawasaki disease, and rare vasculitides in children of different ethnic origins. Lancet. 2002;360:1197–202.