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A patient with abdominal pain and a rash

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CASE REPORT

A 24-year-old male presented to the emergency room because of fever, abdominal pain nausea and vomiting. He complained of having had painful wrists, elbows, knees and shoulders for the past two days. His left ankle was swollen and red. Eight days before presentation he had noticed red, non-painful spots on both legs which increased in size during the course of the week. Six weeks before presentation he had suffered from a sore throat with fever. He was not taking any medication and had no relevant medical history.

On examination he was afebrile (temperature 37.8°C) and not in distress, blood pressure 156/94 mm Hg, pulse rate 64 beats/min. His left tonsil was enlarged. Purpuric macules were observed on both legs. (*figure 1*). Further general physical examination revealed no abnormalities. C-reactive protein was 67 mg/l, liver and kidney function were normal. Urine analysis showed 1 to 5 erythrocytes and protein 0.26 g/l. The chest X-ray was normal.

Figure 1. Rash on the left leg, consisting of purpuric macules



WHAT IS YOUR DIAGNOSIS?

See page 48 for the answer to this photo quiz.

ANSWER TO PHOTO QUIZ (ON PAGE 47)
A PATIENT WITH ABDOMINAL PAIN AND A RASH

DIAGNOSIS

The patient presented with the clinical picture of Henoch-Schönlein purpura (HSP), namely purpura, joint pain, abdominal symptoms and kidney disease. A skin biopsy of the purpura showed a histological picture consistent with leucocytoclastic vasculitis and vascular IgA deposits on direct immunofluorescence study, supporting the diagnosis of HSP.^{1,2}

HSP is much more common in children than in adults (estimated incidence 22.1 vs 1.3 per 100,000 population, respectively).³ However, it is crucial to recognise that HSP does occur in adults, as shown in our case, because complications may be severe. After the acute phase, when gastrointestinal haemorrhage can occur due to gastrointestinal vasculitis, end-stage renal failure develops in 0 to 49% of the adult patients.³ In children the risk of developing end-stage renal failure ranges from 5 to 15%. Risk factors for adults with HSP for developing end-stage renal failure include proteinuria ≥ 1 g/24h during follow-up, hypertension at presentation and during follow-up, renal impairment at presentation, age <30 years and male sex.³ In contrast with the Chapel Hill Consensus Conference criteria for HSP,¹ the American College of Rheumatology (ACR) does not require a biopsy of the skin or kidney with the presence of IgA immune deposits for the diagnosis of HSP.² However, when based on clinical signs only, the diagnosis

of HSP may be missed and patients may be inappropriately discharged from follow-up.⁴ Conversely, in IgG or IgM associated leucocytoclastic vasculitis complications are less frequent⁵ and without biopsy patients may be overdiagnosed as HSP and submitted to unnecessary follow-up.

In conclusion, although HSP is usually a paediatric disease, it can occur in adults. It is essential to make the correct diagnosis based on the clinical signs and symptoms as well as immune fluorescent staining of a skin or kidney biopsy to ensure adequate follow-up.

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