**CASE REPORT**

Primary antiphospholipid syndrome presenting as complicated Henoch–Schönlein purpura

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A 6 year old boy was admitted to hospital with a diagnosis of Henoch–Schönlein purpura. He had no previous medical problems including renal or neurological diseases, and had no family history of connective tissue disorders. Two days before admission, he developed purpuric lesions of both legs, arthralgias (especially of knees and ankles), and abdominal pain. On examination he appeared to be well but had a mild fever (38°C); blood pressure was 110/60 mm Hg. He had extensive purpuric lesions of both legs, ophthalmalgia, heart, and lung examinations were normal. Abdominal examination revealed diffuse tenderness and the limbs were normal except for knee and ankle arthralgias. Urinalysis showed mild haematuria and proteinuria. Five days later, he developed left orchitis, his abdominal pain worsened and became associated with bloody stools. Parenteral feeding was started. Three weeks later the whole of his right leg became mottled, then turned blue and cold with absent femoral pulses. Doppler flow imaging showed an iliofemoral thrombosis. An arteriogram showed extensive occlusion of the upper third of the anterior tibial, posterior tibial, and fibular arteries, and the absence of foot vascularisation (fig 1).

**DISCUSSION**

Antiphospholipid syndrome in paediatric patients has a wide clinical spectrum ranging from benign signs such as migraine or *livedo reticularis* to a “catastrophic” occlusion syndrome. It is usually primary, but sometimes associated with systemic disorders such as systemic lupus erythematosus. Our patient experienced purpura, abdominal pain with bloody diarrhoea, and joint and kidney involvement. These signs fulfil the American College of Rheumatology 1990 criteria for Henoch–Schönlein purpura. Subsequent arterial and venous occlusions proved to be caused by a primary antiphospholipid syndrome with multisystemic manifestations: purpura, vasculitic disease such as Henoch–Schönlein purpura but...
without any pathogenic events of antiphospholipid syndrome. Garber et al reported an adult case of Henoch–Schönlein purpura associated with anti-Ro (SSA) and antiphospholipid syndrome with recurrent deep vein thrombosis. Sokol et al reported a case of a 15 year old girl who had features of Henoch–Schönlein purpura and stroke, with transient IgA antiphosphatidylethanolamine antibody in her serum and CSF.

Henoch–Schönlein purpura and antiphospholipid syndrome are multisystem diseases which may affect the skin, joints, gastrointestinal tract, kidneys, brain, testes, myocardium, and lungs. The pathogenetic mechanisms underlying Henoch–Schönlein purpura are still poorly understood. However, IgA abnormalities suggest an immunological basis. Antiphospholipid syndrome also has an immunological basis related to antiphospholipid antibodies and/or phospholipid binding proteins. The question remains whether it is a coincidental association or the same disease with different manifestations.

REFERENCES