CASE REPORT

Primary antiphospholipid syndrome presenting as complicated Henoch–Schönlein purpura

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A child showing signs of Henoch–Schönlein purpura developed a right tibiofibular vascular thrombosis. Antiphospholipid antibody tests were positive for both lupus anticoagulant and anticardiolipin antibodies. This suggests that an antiphospholipid syndrome should be considered in cases of Henoch–Schönlein purpura and antiphospholipid antibodies should be measured to determine whether prophylactic antithrombotic measures are needed to prevent thrombotic manifestations.

The antiphospholipid syndrome is a thrombotic disorder characterised by recurrent arterial or venous thrombosis or recurrent pregnancy loss and the presence of circulating antibodies directed against phospholipids.[1] The features of antiphospholipid syndrome are well recognised in children, including venous or arterial occlusion syndromes, non-stroke neurological events such chorea, Perthes' disease, and thrombocytopenia.[1] The syndrome may be secondary to systemic lupus erythematosus or other rheumatic processes; in the absence of such diseases, antiphospholipid syndrome is considered as a primary syndrome.

Henoch–Schönlein purpura is the most common vasculitic disease affecting children.[2] The diagnosis is straightforward when the clinical features include non-thrombocytopenic purpura, arthralgias, abdominal pain, and glomerulonephritis. We present a case of a lower limb arterial thrombosis associated with antiphospholipid syndrome in a 6 year old boy who presented with Henoch–Schönlein purpura.

CASE REPORT

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 extensive 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. Oropharyngeal, 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 ileofemoral 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). Laboratory tests showed anaemia (haemoglobin 73 g/l), leucocytosis (white blood cell count 35 × 10^9/l with 85% neutrophil polynuclear cells), platelet count 410 × 10^9/l, and erythrocyte sedimentation rate 64 mm/h; prothrombin time, complement proteins C3 and C4, anti-DNA, antinuclear, and anti-Sm antibodies were normal, as were serological tests for syphilis (VDRL) and HIV. Antithrombin III, protein C, and protein S were also normal. A lupus anticoagulant was detected on haematological testing; anticardiolipin antibodies were: IgG 21 UGPL/ml and IgM 54 UMPL/ml (normal values in our laboratory are: IgG 10 UGPL/ml and IgM 10 UMPL/ml).

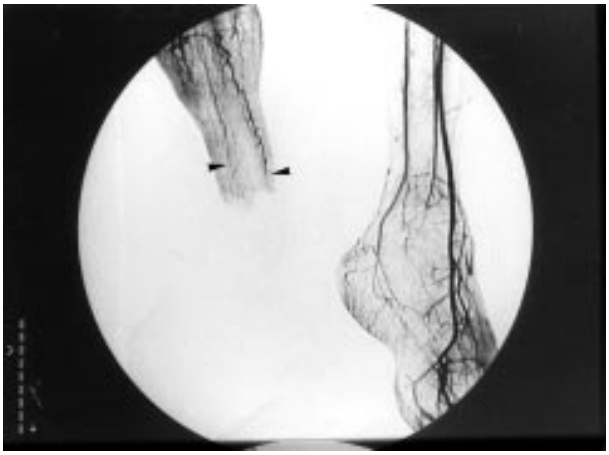

Figure 1 Lower limb arteriogram image showing arterial thrombosis of anterior tibial, and posterior tibial peroneal arteries (arrowheads).

He was treated with heparin (500–600 U/kg/day) and antibiotics (ceftazidime, fosfomycin, amikacin). Dry gangrene of the right foot and the lower leg extremity developed, resulting in amputation. His wound healed without further complications. The patient was discharged taking aspirin (5 mg/kg/day). He had no other thrombotic events or Henoch–Schönlein purpura manifestations during four years of follow up.

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.[1] 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.[3] Subsequent arterial and venous occlusions proved to be caused by a primary antiphospholipid syndrome with multisystemic manifestations[1 4]: purpura, vaso-occlusive manifestations with high anticardiolipin antibodies and no systemic lupus erythematosus features; all symptoms resolved after anticoagulant therapy.

Antiphospholipid antibodies have been associated with vasculitic disease such as Henoch–Schönlein purpura but

without any pathogenic events of antiphospholipid syndrome.[5 6] Garber *et al* reported an adult case of Henoch–Schönlein purpura associated with anti-Ro (SSA) and antiphospholipid syndrome with recurrent deep vein thrombosis.[7] 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.[8]

Henoch–Schönlein purpura and antiphospholipid syndrome are multisytem diseases which may affect the skin, joints, gastrointestinal tract, kidneys, brain, testes, myocardium, and lungs.[1 2]

The pathogenetic mechanisms underlying Henoch–Schönlein purpura are still poorly understood. However, IgA abnormalities[2] 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.

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