Thrombocytopenia.

Neurological events such as chorea, Perthes’ disease, and including venous or arterial occlusion syndromes, non-stroke antiphospholipid syndrome are well recognised in children, who presented with Henoch–Schönlein purpura.

We present a case of a lower limb arterial thrombosis. Purpura, arthralgias, abdominal pain, and glomerulonephritis. When the clinical features include non-thrombocytopenic 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).

He was treated with heparin (500–600 U/kg/day) and antibiotics (cefazidime, 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. 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, vasooclusive 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. 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.

Authors’ affiliations
K Monastiri, H Selmi, B Tabarki, M Yacoub, A S Essoussi, Department of Pediatrics, Farhat Hached Hospital, Sousse, Tunisia
T Mahjoub, Laboratory of Hematology, Farhat Hached Hospital

REFERENCES
7 Garber ME, Mohr BW, Calabrese LH. Henoch-Schönlein purpura associated with anti-Ro(SSA) and antiphospholipid antibody syndrome. J Rheumatol 1993; 20:1964–6.
Primary antiphospholipid syndrome presenting as complicated Henoch–Schoénlein purpura

K Monastiri, H Selmi, B Tabarki, M Yacoub, T Mahjoub and A S Essoussi

Arch Dis Child 2002 86: 132-133
doi: 10.1136/adc.86.2.132

Updated information and services can be found at:
http://adc.bmj.com/content/86/2/132

These include:

References
This article cites 8 articles, 2 of which you can access for free at:
http://adc.bmj.com/content/86/2/132#BIBL

Email alerting service
Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Topic Collections
Articles on similar topics can be found in the following collections

Immunology (including allergy) (2018)
Drugs: cardiovascular system (514)
Stroke (227)

Notes

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/