

Infantile Acute Haemorrhagic Oedema in a Child with von Willebrand's Disease

Sir,

Infantile acute haemorrhagic oedema (IAHE) is an acute form of cutaneous leukocytoclastic vasculitis, which mainly involves capillaries and postcapillary venules of the dermis. It occurs in children under the age of 2 years and is clinically characterized by moderate fever and by the sudden appearance of an inflammatory oedema of the limbs, mainly the extremities, and of the head, associated with erythematous-oedematous purpuric plaques. These lesions rapidly enlarge and coalesce, forming polycyclic figures and/or ecchymotic cockade plaques with a characteristic target-like appearance. Visceral involvement is generally absent. Laboratory findings show normal platelet count without any coagulation abnormalities. Microscopic haematuria, mild proteinuria and increased blood urea nitrogen levels, when present, are usually transient (1, 2). A complete spontaneous recovery occurs in 1 to 3 weeks (mean 12 days) without any significant long-term complications or recurrences. The aetiology of IAHE still remains unknown. The frequent association with a preceding infection, immunization or drug intake suggests a cell-mediated response to an infective agent or an allergic reaction to a drug. Histopathologic findings show a leukocytoclastic vasculitis, with or without fibrinoid necrosis, and a lymphohistiocytic perivascular infiltrate with extravasation of erythrocytes (1, 2).

We here describe the occurrence of IAHE in a child affected by von Willebrand's disease (vWD).

CASE REPORT

Ten days after the start of an upper respiratory tract infection, a 9-month-old boy developed round purpuric, oedematous patches with a cockade pattern. These lesions were symmetrically distributed on the legs (particularly the feet) and on the ears. Inflammatory symmetric oedema of the feet was also present.

The child was otherwise in good health, without any accompanying symptoms.

Laboratory values revealed a haemoglobin level of 11 g/dl, a leukocyte count of 13,930/mm³ with a differential count of 48% neutrophils, 38% lymphocytes, 7% monocytes, 3% eosinophils, and 1% basophils. The platelet count was 314,000/mm³ and erythrocyte sedimentation rate was slightly increased (25 mm/hour; normal value < 15 mm/hour). Prothrombin time (PT) was 92% (normal value 70–110%), partial thromboplastin time (PTT) ratio: 1.46 (normal value 0.85–1.25), fibrinogen 350 mg/dl (normal value 150–600 mg/dl). Other laboratory tests, i.e. liver and renal function, complement, protein C, serum immunoglobulins, Streptozyme test, aspartate aminotransferase and circulating immune complex all showed normal values. Antibody titres against adenovirus and *Mycoplasma pneumoniae* were negative. Urinalysis showed a few microscopic red cells without protein. Throat and urine cultures were negative; pharyngeal swabs for respiratory syncytial virus, adenovirus, influenza A and B virus and parainfluenza virus 1,2,3 were negative.

A biopsy was not performed because the parents refused.

All skin lesions regressed after a week without any treatment. After 10 days we repeated the urinalysis, which did not show microscopic red cells. The previous abnormal coagulation tests were all within the normal limits: PT=89%, PTT ratio=1.27, antithrombin III=108% (normal values >80%), Factor VIII c=93%, Factor VIII ag=92%, while von Willebrand factor was 49% (normal values 70–150%).

A subsequent familial study revealed that the mother showed the same mild von Willebrand factor defect without any clinical signs.

DISCUSSION

At the time of observation our patient had tender symmetric oedematous and target-like purpura. In addition the lack of any visceral involvement, the patient's age and the rapid, benign course of the disease completely fit the diagnostic criteria of IAHE. In fact, Henoch-Schönlein purpura arises in older children and is generally associated with joint pain and/or abdominal pain, renal involvement, and gastrointestinal bleeding. Although there are some clinical similarities between IAHE and Henoch-Schönlein purpura, recent reports indicate that they may be considered two separate entities (3–8). In IAHE cutaneous lesions are characterized by medallion-like or cockade purpura with no extracutaneous involvement and the patient's age is approximately 2 years or less.

All cases of IAHE reported in the literature show normal haematologic and coagulation values, except for one patient who developed a severe gastrointestinal involvement and intravascular disseminated coagulation with fatal outcome (9) and another case with mild reduction of PT value (60%) without any clinical relevance (10).

vWD encompasses a heterogeneous group of disorders, involving the von Willebrand portion of the factor VIII complex. The incidence of vWD is not available, but according to some authors at least 1%–3% of the general population may show coagulation abnormalities consistent with vWD (11, 12).

Patients with vWD show a mild to moderate bleeding tendency in muco-cutaneous surfaces: nosebleeds, menorrhagia, prolonged oozing from cuts, and increased bleeding after trauma or surgery. The bleeding time is usually prolonged and the PTT may also be prolonged.

However, some patients with coagulation abnormalities consistent with vWD report no bleeding symptoms; as von Willebrand factor behaves as an acute phase reactant protein (11) PT and PTT are sometimes in the normal range, so that no coagulation abnormalities can be observed if only these two tests are performed. We observed a similar situation at the second test in our case, but the laboratory values were consistent with a diagnosis of vWD.

The association between vWD and IAHE here described is, to our knowledge, the first reported in the literature. It may be casual but if other observations of IAHE associated with vWD are reported in the future this association may be considered not fortuitous.

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BOOK RECEIVED

An Atlas of Vulval Disease, 2nd ed. by M. Leibowitch, R. Staughton, S. Neill, S. Barton and R. Marwood, 1997. 196 pages. ISBN 1-85317-431-9. Hardcover. Price £49.95. Martin Dunitz Publ. London.

This new edition has been updated with new excellent colour illustrations and expanded with sections such as treatment, plastic repair, malignant disease and patient information. The clear and didactic writing makes this book very valuable in clinical practice where patients with vulvar problems may appear.

The Editor

Strategies for Immunointerventions in Dermatology edited by Günter Burg and Reinhard G. Dummer, 1997. 418 pages, 35

Figures. ISBN 3-540-62680-8. Hardcover. Price US\$ 129. Springer Verlag, Berlin.

Fifty-two experts have contributed to this volume containing chapters on basic aspects of immunointerventions in dermatology, various immunosuppressive and antiinflammatory compounds, immunostimulatory substances, and vaccination therapies. There are also clinical aspects on various dermatoses including also the common disorders as contact dermatitis, atopic dermatitis, psoriasis, bullous diseases, vasculitis and viral infections as well as rheumatic diseases, neoplasms and malignant melanoma. After each chapter there are many references up to 1996. The book is of great value to doctors taking care of patients with these disorders when immunotherapy can be needed.

The Editor