et al[5] and could have accounted for the lung lesion seen in our patient. Nevertheless, the peculiar evolution of this case, in which the vascular lesions suggesting periarteritis nodosa preceded the other clinical signs typical of relapsing polychondritis, tends to confirm the suggestion by MacAdam *et al*[3] and Neild *et al*[5] that at least some patients with relapsing polychondritis fall into the range of systemic vasculitis.

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Brugmann Hospital and Free University, Brussels, Belgium
G SOMERS, MD, clinical assistant, department of medicine
P POTVLIEGE, MD, senior assistant, department of pathology

Thrombocytopathy associated with autoimmune haemolytic anaemia

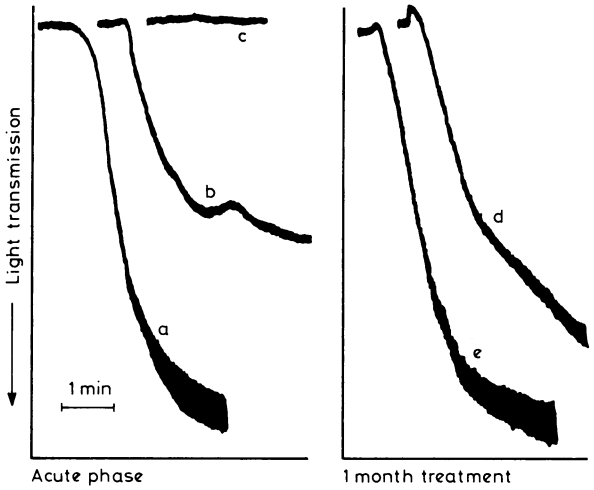
Thrombocytopathy associated with acquired deficiency of the platelet storage pool of adenine nucleotides (acquired storage pool disease) is being increasingly recognised as a complication of several conditions, including disseminated intravascular coagulation[1] and systemic lupus erythematosus (SLE).[2] We report a case of a patient who presented with an autoimmune haemolytic anaemia and later developed arterial insufficiency of the left hand. The platelet defect first became apparent during plasmapheresis, when an accumulation of platelet aggregates in the machine was observed.

Case report

A 30-year-old woman presented with malaise, painful swelling, and paraesthesiae of the forearm and fingers of the left hand. There was no history suggestive of Raynaud's phenomenon, and she was not taking the contraceptive pill. She had blotchy erythema of the left forearm, cyanosis of the fingers of the left hand, and splinter haemorrhages in the nail beds. All peripheral pulses were present. Investigations showed an autoimmune haemolytic anaemia: haemoglobin concentration 9·1 g/dl, reticulocytes 16%, white cell count $7{\cdot}8 \times 10^9$/l, platelets 230×10^9/l, ESR 170 mm in 1st h (Westergren). The direct Coombs test was positive and serum contained anti Wr^a. Cryoglobulins were not present. The results of investigations for SLE including LE cells, DNA-binding antibodies, and antinuclear antibodies were negative. Plasma viscosity was normal. There was no laboratory evidence of disseminated intravascular coagulation (PI 12·5 s, kaolin cephalin clotting time 48·5s, fibrinogen concentration 130 mg/100 ml, fibrinogen degradation products $<10\ \mu$g/ml). The patient had a bruising tendency but a bleeding time was not estimated. An arch arteriogram showed severe tapering of the distal and digital arteries of the left forearm and hand.

She was treated with intravenous Rheomacrodex (dextran 40 injection) and heparin and later prednisolone (60 mg/day) was added. The condition of the left hand worsened, however, with the development of gangrene of the tips of the fingers. A two-litre plasmapheresis was performed and a mass of platelet aggregates was observed in the machine at the end of the procedure. Platelet aggregation studies performed at this stage (figure) showed a diminished response to a standard suspension of acid-insoluble collagen (Diamed Diagnostic) and an abnormal second-phase aggregation to 4 μM ADP (Sigma) but aggregation to 1·0 mM sodium arachidonate (Sigma) was normal. Measurement of total adenine nucleotides on perchloric acid extracts[3] showed diminished total nucleotides and reduced nucleotide release in response to 1 unit/ml human thrombin (WHO 1st International Standard 70/157). Total adenine nucleotides and released nucleotides were 99·1 nmol/10^9 platelets and 32·3 nmol/10^9 platelets, respectively (control values $167{\cdot}1 \pm 28{\cdot}2$ nmol/10^9 platelets and $78{\cdot}2 \pm 18{\cdot}6$ nmol/10^9 platelets, respectively, $n=12$). A further plasmapheresis was performed two days later and again a mass of platelet aggregates was observed. The results of tests to demonstrate platelet antibodies by platelet factor-3 availability[4] were negative.

The response to plasmapheresis was good with improvement of the circulation to the fingers and demarcation of the areas of gangrene. At this stage azathioprine (100 mg/day) was added to her treatment. One month later her haemoglobin concentration had risen to 13·1 g/dl, the reticulocytes were 1·2%, and the ESR had fallen to 14 mm/h. Platelet aggregation studies now showed normal aggregation to collagen and ADP (see figure). The total platelet count and released adenine nucleotides had returned to normal (162·1 nmol/10^9 platelets and 85·9 nmol/10^9 platelets respectively).



Photometric recordings of aggregation in platelet-rich plasma showing abnormal responses to ADP (b) and collagen (c) during acute phase of illness. Aggregation to sodium arachidonate (a) was normal. After one month's treatment platelet aggregation showed completely normal aggregation to ADP (d) and collagen (e) identical to controls.

Comment

It is being increasingly recognised that an in vivo-platelet release reaction due to circulating aggregating agents may lead to the formation of platelet aggregates and to thrombosis.[1] In this patient the tendency for platelet aggregation to occur was shown during plasmapheresis. Because of rheological factors not all platelets that undergo a release reaction are concerned in forming aggregates and this leads to the circulation of a population of haemostatically incompetent platelets, which may lead to a bleeding tendency. In this patient the abnormal platelet aggregation study results and the diminished total and released adenine nucleotide concentrations were typical of acquired storage pool disease. Normal aggregation in response to sodium arachidonate excluded an aspirin-like thrombocytopathy.

The mechanism for the acquisition of the platelet defect in this patient is uncertain, since we failed to demonstrate the presence of platelet antibodies; nevertheless, the methods available for detecting platelet antibodies are not totally reliable. The presence of red cell antibodies, the clinical response of the thrombotic episode to plasmapheresis and immunosuppressive treatment, and the return to normal of platelet function on this treatment all indicate an immunological basis for the platelet defect—for example, circulating immune complexes or undetected platelet antibodies. Platelet injury due to immunological mechanism may be more common than realised as a contributing cause in unexplained thrombotic episodes, and early diagnosis might be important in the management of such patients.

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University of Liverpool and Liverpool Royal Infirmary, Pembroke Place, Liverpool 3
N H RUSSELL, MB, MRCP, research fellow in haematology
J P KEENAN, FIMLS, senior chief medical laboratory scientific officer
M A FRAIS, MB, MRCP, medical registrar

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