nephrotic syndrome in that most patients with associated renal vein thrombosis have membranous glomerulonephritis.[5]

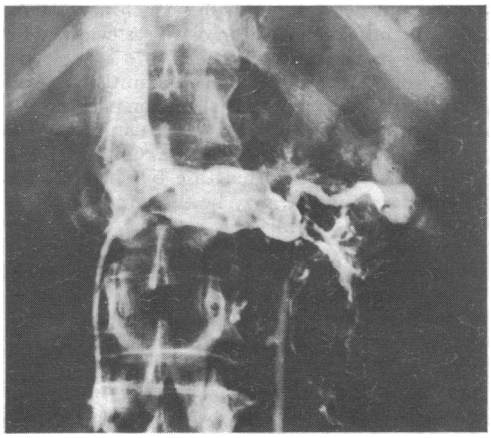
We describe here a patient who presented with lupus erythematosus, nephrotic syndrome, and unilateral renal vein thrombosis and was found to have another type of histological lesion: focal proliferative glomerulonephritis.

Case report

A 45-year-old woman admitted to our unit in February 1976 was found to have malar erythema, bilateral pleural effusion, and ankle oedema. Her blood pressure was 120/90 mm Hg. When she was 26 this woman had had proteinuria and oedema during the first trimester of her second pregnancy but had had no evidence of renal failure or hypertension. Delivery had been normal. Two years after this pregnancy she had begun to have pain in her joints, and five years before admission she had developed malar erythema and occasional fever.

Laboratory investigation showed: packed cell volume of 40%, white cell count $5{\cdot}1 \times 10^9$/l (5100/mm^3); erythrocyte sedimentation rate 48 mm in 1 h; platelet count $196 \cdot 10^9$/l (196 000/mm^3); urea 10 mmol/l (60 mg/100 ml); serum creatinine 150 μmol/l (1·7 mg/100 ml); creatinine clearance 44 ml/min/1·73 m^2; total protein 44 g/l; albumin 17 g/l; cholesterol 6·4 mmol/l (248 mg/100 ml); and triglycerides 3·0 mmol/l (268 mg/100 ml). Urine analysis showed proteinuria of 14 g/day with 15-20 red cells, 2 or 3 leucocytes, and 3 or 4 granular casts per high-power field. LE cells were found, and she was positive for antinuclear antibodies at a titre of 1/640. C3(B_1A) was 0·470 g/l (normal: 0·52-1·15 g/l) and plasma fibrinogen was 3·54 g/l, with no serum fibrinogen degradation products.

Chest radiographs showed bilateral pleural effusion. Bilateral renal phlebography showed complete occlusion of the left intrarenal venous system, with a large thrombus in the principal venous trunk and collateral drainage circulation throughout the ovarian vein (see figure). Biopsy of the left kidney showed that out of 25 glomeruli three were completely sclerosed, 13 were normal on light microscopy, and nine had proliferative segmental lesions with focal thickening of the basement membranes, areas of necrosis, and adhesions. There were mild tubulointerstitial lesions with lymphocyte accumulation, slight fibrosis, oedema, and tubular atrophy. Immunofluorescent deposits of IgG, IgM, C1q, and C3 were shown in the mesangium and basement membranes of almost all glomeruli. There were no deposits of IgA, IgE, C4, or fibrinogen.



Left renal phlebography showing complete occlusion of intrarenal venous system, thrombus in principal trunk, and collateral drainage circulation throughout ovarian vein.

The patient was treated with prednisone (40 mg/day) and azathioprine (100 mg/day). During the next two months renal function worsened, but after three-day pulse treatment with methylprednisolone 1 g/day it improved and she was discharged on oral prednisone (30 mg/day) and azathioprine (100 mg/day). During the next 11 months kidney function was sustained without hypertension and with a proteinuria of 4-6 g/day.

Comment

In all previous cases of lupus erythematosus associated with the nephrotic syndrome and renal vein thrombosis the histological appearances, when documented,[4 5] have shown membranous glomerulonephritis. Ours is the first case to show focal proliferative glomerulonephritis.

The pathogenesis of the triad remains obscure. Although the idiopathic nephrotic syndrome seems to be associated with a hypercoagulable state characterised by increased concentrations of factors V, VII, VIII, and X, fibrinogen, and platelets, the coagulation abnormalities in lupus erythematosus are complex. Thrombocytopenia is often found and almost half the patients have qualitative defects in the platelets, with a decrease in platelet aggregation induced by collagen, adenosine diphosphatase, and adrenaline. Circulating anticoagulants against clotting factors may be found, which seems to reduce the tendency toward thrombosis. On the other hand, lupus vasculitis favours peripheral vascular occlusions as well as superficial and deep vein thrombophlebitis, and an increase of fibrinolitic split products has been reported, which suggests that slow intravascular coagulation may occur.

To our knowledge there have been no controlled trials to determine whether renal vein thrombosis worsens the prognosis of patients with the lupus nephrotic syndrome. Prospective studies in which renal phlebography and biopsy are performed in patients with the lupus nephrotic syndrome are needed to establish whether renal vein thrombosis is associated more often with membranous nephropathy than with focal proliferative lesions and whether this has any prognostic significance.

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Aid for the disabled

The Vac-Pac system was originally conceived to maintain the patient in the correct position during surgery. It was later thought that this principle could easily be used in the conscious patient who through injury or disease was unable to maintain a comfortable resting position: in particular this might prove useful for patients with deformity or muscular weakness due to muscular dystrophy. Such patients often have great difficulty in sleeping owing to an inability to maintain a comfortable position.

Method

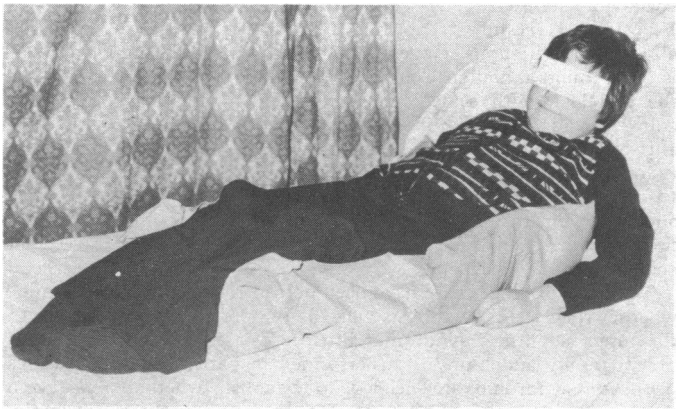
Vac-Pac consists of thousands of polystyrene beads within a plastic envelope which can be moulded to shape. When a vacuum is created within the envelope the beads clump together, becoming solid and maintaining their shape. The device reverts to its flat shape on releasing the vacuum. The vacuum required can easily be created by a small hand-operated vacuum pump such as the Ambu Minipump. The total cost is about £75.

Usually the child is brought into hospital for a few days so that both parents and child can familiarise themselves with the technique. With the patient in the desired position the Vac-Pac is moulded around the body contour and then suction applied (see figure). The Vac-Pac may be used lined or unlined.

Discussion

Most paediatric hospitals see the unfortunate sufferers of muscular dystrophy in either neurological or orthopaedic clinics. Vac-Pac has been found ideal for these patients, its particular advantage lying in its facility for altering shape as the patient grows or his deformities deteriorate. Thus the Vac-Pac can be used as an aid to sleep and also to maintain a comfortable sitting position during play.

The results of using this device have been extremely encouraging. To date, four patients aged between 9 and 16 years all with Duchenne type of muscular dystrophy have used this method. All reported



Vac-Pac comfortably moulded to the body contour.

improvement in both general comfort and sleep. The sphere of application for Vac-Pac is probably far wider than just muscular dystrophy.

I would like to thank Mr A Glass for allowing me access to his patients and for helpful advice. The photograph is by the Department of Medical Illustration, North Manchester General Hospital. Vac-Pac is marketed by Howmedica (UK) Limited. Ambu Pumps are available from Oxylitre Limited of Manchester.

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Autoimmune haemolytic anaemia with anti-A autoantibody

In many cases of autoimmune haemolytic anaemia the target antigen corresponds to an antigenic entity defined by isoantibodies that have blood group specificity. Usually the Rh-complex is implicated,[1 2] here we report a case in which the autoantibody showed specificity for A_1 and A_2 erythrocytes.

Case report

A 2-year-old girl was referred because of persistent listlessness after an upper respiratory tract infection. Examination showed pallor, jaundice, and hepatosplenomegaly. Results of investigations were: haemoglobin 4·1 g/dl; red cell count 1720×10^9/l (1 720 000/mm^3); white cell count $5·2 \times 10^9$/l (normal differential); platelet count 120×10^9/l; erythrocyte sedimentation rate 150 mm in first hour; reticulocytes 2·7%; 6 nucleated red cells/100 white cells; pronounced rouleaux formation; normoblastic hyperplasia in bone marrow; haemosiderinuria and methaemalbuminaemia detected; serum bilirubin concentration 58 μmol/l (3·4 mg/100 ml); no evidence of intracorpuscular erythrocytic defects on screening; viral screen normal; IgG 96 (normal 43-142) U/ml, IgM 400 (47-220) U/ml, IgA 50 (18-115) U/ml, no M component.

Initial progress with steroid treatment was satisfactory, and a mild relapse after one month responded to a small increase in prednisolone.

On referral a direct antiglobulin test (DAGT) result was positive (titre 1/128) due to complement binding. Incubation produced lysis of the patient's and donor A_1 erythrocytes due to antibodies in the patient's serum. Tests with control serum gave negative results. The antibodies in the patient's serum showed specificity for A_1 and A_2 cells (see table). The patient was group A_1 and remained so after clinical remission. The specificity of the antibody for the A antigen was confirmed when absorption of the patient's serum with A_1 secretor saliva removed the autoanti-A antibodies and left the anti-B antibodies. Furthermore, the patient's erythrocyte eluate contained anti-A antibodies but no antibodies against B or O cells. Two factors suggested that the autoantibody was IgM—namely, incubation of the serum with dithiothreitol removed both the anti-B and autoanti-A antibodies, and Sephadex filtration showed maximal antibody activity in the 19S fraction.

The autoantibodies disappeared for a short time with treatment, but were present later when the DAGT result was positive. By then the autoantibodies had changed in reactivity, binding to group O and fetal cells in addition to group A erythrocytes. An erythrocyte eluate, however, showed activity against A_1 and A_2 cells but not against group O cells.

Comment

There has been one report[3] of autoanti-A antibodies causing intravascular haemolysis, but their specificity has been questioned.[4] The autoantibodies in our patient did not show any cross-reactivity with either group O or cord erythrocytes at the time of referral. The antiglobulin test result, though positive with specific anticomplement sera, was unexpectedly negative with anti-IgM sera. This may be explained either by a weak antiserum having been used or by the fact that IgM on the erythrocyte was masked by C3.

That the antibody was IgM would be consistent with the type of blood group specificity shown, since naturally occurring anti-A and anti-B antibodies are of the IgM subclass. The lytic nature of the antibody shown in vitro also supports this conclusion and may explain the presence of the intravascular haemolysis, as evidenced by urinary haemosiderin and methaemalbuminaemia.

At follow-up the autoantibody that was initially specific for A_1 and A_2 appeared to be broadening in its reaction pattern. Whether this change was due to treatment or part of the natural evolution of the antibody type in this patient is unknown. Our findings, however, support those of the other reported case by showing that a major blood group antigen, group A, may be the target antigen in human autoimmune haemolytic anaemia.

We thank Miss E Manson for her technical expertise; Dr K James and Miss J Wallace for fractionating the serum; Dr J W Farquhar for allowing us to report this case; and Miss K Denholm for typing the manuscript.

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Results of standard antibody screening of patient's serum and erythrocyte eluate on referral. Specificity for group A cells shown

	Presence in patient's serum of antibodies against:					Presence in erythrocyte eluate of antibodies against:				
	O erythrocytes (six separate donors)	Fetal cells	A_1	A_2	B	O erythrocytes (two separate donors)	Fetal cells	A_1	A_2	B
Saline (at 4°C)	−	−	+	+	+ + +	−	−	+ +	+ +	−
Saline (at room temperature)	−	−	±	±	+	−	−	+ +	+ +	−
Saline (at 37°C)	−	−	±	±	±	−	−	+	+	−
Enzyme (at room temperature)	−	−	−	−	±	−	−	+ +	+ +	−
Enzyme (at 37°C)	−	−	−	−	±	−	−	+ +	+ +	−
IAGT result	−	−	+	+	+ +	−	−	±	±	−

IAGT = Indirect antiglobulin test.