Three months after the first episode of purpura she presented again with diffuse purpura. The platelet count was 6×10^9/l (6000/mm^3), and the results of examination of the bone marrow was consistent with the diagnosis of idiopathic thrombocytopenic purpura. She had clearly had a relapse. The results of tests for LE cells, antinuclear factor, and red-cell antibodies were negative. Cellular and humoral immunity function tests were normal. The thrombocytopenia and purpura persisted despite treatment with steroids and azathioprine, so a splenectomy was performed when she was 6 years old. After this she had a complete clinical and haematological recovery.

Comment

All but 5% of patients with thrombocytopenic purpura are considered to have the idiopathic form, in which no preceding infection can be shown. One-half of this group have the true idiopathic form and the other half the postviral idiopathic form. Possibly, therefore, a patient who is thought to have the true idiopathic form might move to the postviral group when an infectious illness can be shown to have preceded the onset of purpura. This may have been the case with our patient. She fulfils the criteria proposed by Feldman[3] for the diagnosis of toxoplasmosis. A titre of 1/4096 in the dye test and a rising titre in two serum specimens are compatible with the diagnosis of a recently acquired infection.

Toxoplasma infection is much more common in people with compromised immunological states. This raises the possibility that both the idiopathic thrombocytopenic purpura and the toxoplasmosis in our patient resulted from an altered immune mechanism that was not detected by standard laboratory procedures. Cell-mediated immunity may play a part in the pathogenesis of idiopathic thrombocytopenic purpura. Recently, blast transformation of lymphocytes in the presence of autologous platelets was shown.[4] Is the production of antiplatelet antibodies mediated through the inhibition of suppressor T cells? Or are there two parallel mechanisms: one of cell-mediated immunity and the other of humoral immunity? Is toxoplasma infection the result of an altered immune state, or did the parasite cause such a change in cell-mediated immunity that idiopathic thrombocytopenic purpura appeared? These and other questions await an answer.

Interestingly, we could not detect autoantibodies in the sera of our patient after splenectomy. In our laboratory we found a significant increase in the incidence of autoantibodies in patients who have had splenectomies. In view of our patient's condition, we suggest that the term "postviral idiopathic thrombocytopenic purpura" should be extended to "postinfectious idiopathic thrombocytopenic purpura" to include the possible infectious agents that, although they are not viruses, may play a part in causing thrombocytopenic purpura.

[1] Doan CA, Bouroncle BA, Weiseman BK. Idiopathic and secondary thrombocytopenic purpura. *Ann Intern Med* 1960;**53**:861-9.
[2] McClure PD. Idiopathic thrombocytopenic purpura in children: diagnosis and management. *Pediatrics* 1974;**55**:68-74.
[3] Feldman HA. Toxoplasmosis. *N Engl J Med* 1968;**279**:1370-5.
[4] Wybran J, Fudenberg HH. Cellular immunity to platelets in idiopathic thrombocytopenic purpura. *Blood* 1972;**40**:856-61.

(*Accepted 4 March 1980*)

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Primary thrombocythaemia in monozygotic twins

The aetiology of myeloproliferative disorders is unknown but there is some evidence that genetic factors may play a part. Although there are several reports[1 2] of familial polycythaemia there are very few of primary thrombocythaemia. This report is of primary thrombocythaemia in monozygotic twins.

Case report

A 35-year-old man (A) had lost 6 kg in weight when he presented with a 10-week history of partial blindness in one eye. He had a right subhyloid haemorrhage and his spleen was enlarged 7 cm below the costal margin. He was anaemic and had a platelet count of 954×10^9/l (954 000/mm^3). His asymptomatic twin brother (B) had a radiologically enlarged spleen and a platelet count of 1330×10^9/l (1 330 000/mm^3) but he was not anaemic. The full blood counts are given in the table. B's blood film was morphologically normal but that of A showed poikilocytes, teardrop cells, and target cells. Their bone marrows yielded only cells of the normal male karyotype. The morphology of marrow smears and bone trephines from both men were similar. The smears contained no stainable iron, erythropoiesis was normoblastic, and there were no dyserythropoietic changes. There were sheets of platelets and numerous megakaryocytes. There was no increase of reticulin and no fibrosis in the bone trephines. All other investigations, including measurement of the leucocyte alkaline phosphatase concentration, were normal except that A had hyperuricaemia, with uric acid concentration 0·59 mmol/l (10 mg/100 ml) (normal 0·15-0·42 mmol/l (2·5-7·0 mg/100 ml)).

Blood counts in monozygotic twins with primary thrombocythaemia

	Age when tested (years)	Haemoglobin (g/dl)	Mean cell vol (fl)	White cells (10^9/l)	Platelets (10^9/l)	Red cells (10^{12}/l)
A	35	11·8	87	12·4	954	3·84
B	35	13·5	88	14·9	1330	4·39
Father	67	11·4	93	6·7	328	3·59
Mother	65	15·7	90	8·5	250	4·92
Sister	37	12·3	88	6·3	235	3·93
Daughter of A	15	12·7	90	7·8	390	4·28
Son of A	11	12·3	84	8·6	390	4·48
Son of B	7	11·8	81	7·4	350	4·41

Conversion: SI to traditional units—Mean cell volume: 1 fl = 1 μm^3.

The red cell genotype of the twins and their close relations together with tissue typing studies suggested that the twins were monozygotic. None of their relations had thrombocytosis. The twins were both treated for primary thrombocythaemia. After initial treatment with radioactive phosphorus the platelet count of both men fell below 400 10^9/l (400 000/mm^3) within four months. By then A had gained 7 kg and his haemoglobin concentration had risen to 13·6 g/dl. During the subsequent six years neither has developed polycythaemia but both have had several courses of busulphan when their respective platelet counts rose above 400×10^9/l.

Comment

Dedichen *et al*[3] in a paper describing nine patients with haemorrhagic thrombocythaemia included a 40-year-old woman with five similarly affected siblings. The platelet count of four of the siblings was not available but the fifth, a man, had splenomegaly associated with thrombocythaemia. Beretta Anguissola and Prato[4] described thrombocythaemia associated with gastrointestinal haemorrhage in three male members of three generations of an Italian family. Flickers and Speck[5] described a 47-year-old man with primary thrombocythaemia. He had been taking thiotepa for two years when cytogenic studies showed hypoploidy in 8 of 70 karyotypes. His sister, who had hepatosplenomegaly and thrombocythaemia, refused to have a marrow aspirate, but probably her brother's chromosomal abnormalities were the result of cytotoxic therapy.

The present report is unique in that it describes primary thrombocythaemia in monozygotic twins. There is no evidence that any of their close relations has a similar disorder, but the presentation of the same myeloproliferative disease in the twins is evidence that a genetic factor may play some part in its pathogenesis.

I thank Professor S Peart for referring the patient, Dr S Lawler for the cytogenetic studies, and Dr R Sanger for red cell genotyping.

[1] Modan B. Polycythaemia. A review of epidemiological and clinical aspects. *J Chronic Dis* 1971;**18**:605-45.
[2] Levin WC, Houston EW, Ritzmann, SE. Polycythaemia vera with Ph' chromosomes in two brothers. *Blood* 1967;**30**:503-12.
[3] Dedichen J, Refven O, Ytreuus K. Essential thrombocythaemia. *Nord Med* 1959;**62**:1265-70.
[4] Beretta Anguissola A, Prato V. Trombocitosi ereditaria, prima osservazione. *Minerva Med* 1961;**52**:4545-8.
[5] Flickers M, Speck B. Thrombocythaemia. Familial occurrence and transition into blastic crisis. *Acta Haematol* (*Basel*) 1974;**51**:257-65.

(*Accepted 4 March 1980*)

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