Autoimmune Haemolytic Anaemia Associated with Rheumatoid Arthritis and Paroxysmal Nocturnal Haemoglobinuria

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Forman et al. [1984] have described 2 patients with paroxysmal nocturnal haemoglobinuria (PNH) and autoimmune haemolytic anaemia. More recently, Conti et al. [1985] described a patient with PNH with autoimmune haemolytic anaemia and autoimmune thrombocytopenia. I would like to report a patient with PNH and autoimmune haemolytic anaemia associated with rheumatoid arthritis.

An 82-year-old lady originally presented in June 1976 with aplastic anaemia confirmed by bone marrow biopsy. She was treated with regular red cell transfusions and oxymetholone 150 mg/day, and a partial remission was achieved after 6 months. Oxymetholone had to be stopped because of the unacceptable side-effect of hirsutism, but in spite of this, partial remission was maintained. In May 1980, the lady developed seropositive rheumatoid arthritis affecting mainly the hands and knees. She subsequently required regular red cell transfusions for an unexplained iron deficiency anaemia.

In July 1985, she developed haemoglobinuria and was admitted for further investigation. Examination revealed pallor, mild jaundice and typical rheumatoid joint deformities of her hands and knees. The spleen was moderately enlarged.

Investigations: Hb 6.5 g/dl, WBC $3.8 \times 10^9/l$, platelets $100 \times 10^9/l$, reticulocyte count 14%. Peripheral blood film showed spherocytosis and polychromasia. Bone marrow biopsy showed reduced cellularity with absent iron stores. Biochemistry: total bilirubin $50 \mu\text{mol/l}$, direct bilirubin $9 \mu\text{mol/l}$, AST 34 U/l, ALT 16 U/l, LDH 2,160 U/l. Immunoglobulins were normal. Plasma haemoglobin was elevated at 99 mg/l with absent serum haptoglobin, and urinary haemosiderin was positive. Rheumatoid and antinuclear factors were positive.

Immunohaematological studies revealed strongly positive direct Coombs' test with broad-spectrum antiglobulin serum. Further studies with monospecific antiglobulin serum revealed an IgG- and C3d-coating on the patient's cells. The eluate prepared from the patient's erythrocytes had anti-c rhesus specificity. Ham's test, sucrose lysis test and insulin test were all positive.

The patient has refused treatment with steroids because of previous side-effects, she is therefore receiving regular red cell transfusions of washed cells. In addition, she is taking oral iron and folic acid. Evidence of continuing mild haemolysis persists.

This is another case of a rare association between PNH and autoimmune haemolytic anaemia of which only 3 cases have so far been described. Further studies are required to determine the incidence of autoimmune disorders in PNH patients.

References

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