Heterozygous Beta-Thalassaemia and Cold Agglutinin Disease in a Surinamic Man

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As far as we can determine this is the first report of this nature.

Case Report

A 48-year-old Surinamic man arrived in the Netherlands 2 months before hospitalisation. The descriptive diagnosis, haemolytic anaemia, had been made 3 years previously in Suriname, for which he received 30 mg prednisone daily. He had complaints of palpitation, dyspnoea, tingling in the hands, increasing fatigue and cramps in the extremities without Raynaud's phenomenon. On physical examination of this ill looking man we noted a Cushingoid face, yellow sclerae, no lymph-adenopathy or hepatosplenomegaly.

Laboratory Findings

ESR 110 mm, Hb 6.1 g/dl, Ht 0.19, reticulo-cytes 4%, WBC $6.0 \times 10^9/l$, the peripheral blood picture under normal circumstances presents aggregation of the erythrocytes and a normal picture of the erythrocytes after warming the slide. Plasma iron $36 \mu\text{mol/l}$, total plasma iron binding capacity $37 \mu\text{mol/l}$, total bilirubin $50 \mu\text{mol/l}$, indirect fraction $47 \mu\text{mol/l}$, LDH 443 U/l, haptoglobin 0 mg/dl. In the bone marrow aspirate the erythro-poiesis was three times as active as the myelo-poiesis. Red cell survival ($^{51}\text{Cr T}_{50}$) was 18.2 days during prednisone and 15.5 days without prednisone (normal 26–32 days), the heart-spleen-liver index was normal. It was concluded that the patient was indeed suffering from a haemolytic anaemia. To determine the nature of this, the following investigations were performed: Hb electro-phoretic pattern, HbAa 4.4% (n 1.9–3.0%), HbF

1.3% (n 0.5–1.5%). Direct antiglobulin test: negative with anti-IgA, C, M, D, E, positive with anti-complement antibodies. Identification of the antibodies: cold agglutinins and haemolysins of specificity anti-I.

The titre of the cold agglutinin was more than 1/500. The concentration of complement component Cs was decreased. Immunoelectrophoretic pattern: IgA 100 IU/ml (n 40–225), IgM 150 IU/ml (n 45–335), IgG 60 IU/ml (n 65–200); IgM consisted of a linear band, as seen with a paraprotein. Immunofluorescence pattern of the bone marrow: IgA 22%, IgM 24%, IgG 33%, kappa 70%, lambda 30% (normal). Reaction of Wassermann, VDRL and Paul Bunnell were negative. Erythrocyte enzyme patterns normal. Vitamin B12 concentration 315 pg/ml (normal). On the basis of these findings the diagnosis of heterozygous β -thalassaemia combined with cold agglutinin disease was established. We slowly diminished and finally stopped prednisone

without precipitating any acceleration of the anaemia. The patient returned to Suriname because he felt the cold Dutch weather was the cause of his troubles.

Comment

This patient is suffering from α -heterozygous thalassaemia and cold agglutinin disease. The two types of anaemia appear to be unrelated.