Letters to the Editor

Systemic lupus erythematosus in a patient with beta-thalassemia major

Sirs,

Systemic lupus erythematosus (SLE) is an autoimmune disease while beta-thalassemias are common hereditary disorders which result in reduced or absent beta-globin chain synthesis. Although the coexistence of sickle cell disease and SLE has been described (1), the occurrence of beta-thalassemia with SLE is rare (2, 3). We describe the case of a man with beta-thalassemia major and SLE affecting mainly the joints and kidneys.

A 37-year-old Greek man presented at the outpatient Rheumatology Clinic of the University Hospital of Ioannina on October 2002 with fatigue, arthritis, fever (38°C), anemia and acute renal failure. In 1974 beta-thalassemia major had been diagnosed based on hemoglobin (Hb) electrophoresis and genotype characterization and the patient was being followed up in the Hematology Clinic. Blood transfusions have been required since 1989 and desferoxamine was administered regularly.

Physical examination revealed moderate palomagnathy, arthritis of the knees and wrists, and icteric conjunctiva. Laboratory results showed Hb 7.4 g/dl, erythrocyte sedimentation rate: 131 mm/h, creatinine 2.2 mg/dl, urea 99 mg/dl, total bilirubin 3.1 mg/dl (direct 0.74 mg/dl), and direct Coombs positive. Twenty-four hour urine protein was 648 mg. The immunological evaluation showed positive antinuclear antibodies (ANA) (titer: 1/1280, homogenous pattern), positive anti-dsDNA antibodies: 79 IU/ml (negative < 10 IU/ml), C3: 75 mg/dl (range 16-181 mg/dl), and C4: 5 mg/dl (range 16-40 mg/dl). The case was negative for antiphospholipid antibodies.

The patient was treated with methylprednisone (60 mg/day) followed by tapering and hydration were administered. Renal biopsy showed mesangial glomerulonephritis. Therefore, the immune status of the patient as well as the effect of multiple allo- and autologous blood transfusion can induce antibody formation. The perpetuation of antibody formation may result from the continuous stimulation of the immune system. SLE patients with beta-thalassemia trait may be at a higher risk of development of SLE. Our patient had a low C4:CD8 ratio of 0.71 and the HLA typing was A2, A30, B14, B18, C2, C8, DR1, DR16, DQ5 and DQ51. In November 2002 the diagnosis of SLE was confirmed by positive antinuclear antibodies: 79 IU/ml (titer: 1/1280, homogenous pattern), C3: 75 mg/dl, and C4: 5 mg/dl (titer: 1/1024, linear pattern). In addition genotyping and genotype characterization and the patient was being followed with HLA typing: B18, DR16 that have been associated with lupus. Furthermore, multiple transfusions may alter the immunological response of thalassemic patients. Experimental transfection of plasma containing alloantibodies has been shown to lead to autoantibody formation. Therefore, the immune status of the patient and the effect of multiple allo- and autologous blood transfusion can induce antibody formation. The perpetuation of antibody formation may result from the continuous stimulation of the immune system. Autoantibody production has been described in thalassemia patients (7) with variable clinical significance. They may exhibit the characteristics of natural autoantibodies or, under unclear circumstances, they may become the pathogenic autoantibodies that are found in SLE. It is known that T cell subsets play a pivotal role in the pathogenesis of SLE. On the other hand, T lymphocytes from thalassemic patients are activated in vivo and they present several abnormalities (8). Thus, the lymphocyte background in thalassemic patients may under the influence of a triggering factor contribute to the development of SLE. Our patient had a low C4:CD8 ratio, which has been described in lupus nephritis patients (8). In addition desferoxamine, which modifies T-cell-mediated immune responses, may also play a role in SLE expression (9, 10).

In conclusion, we described a patient with beta-thalassemia major and SLE. Further studies will clarify whether the association of these two diseases is real or if it constitutes an occasional coexistence. 

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References


Seronegative spondyloarthropathy associated with Takayasu’s arteritis in a child

Sirs,

Takayasu’s arteritis (TA), a relatively uncommon disease in childhood, is a chronic aortopathogenic vasculitis that involves the aorta and its main branches (1). Ankylosing spondylitis is characterized by progressive inflammatory arthritis of the spinal and sacroiliac joints (2). Both of the diseases are infrequent and their association is even more rare.

A 14-year-old girl with a history of leg and low back pain for 4 years, who had previously been followed at various centers with the diagnosis of acute rheumatic fever and juvenile rheumatoid arthritis was admitted to our hospital. She had been operated on for a right ureteropelvic stricture one year ago. Two months after the operation the patient was admitted to her local hospital with signs of severe hypertension unresponsive to medical treatment. Radiological examinations that included DMX angiography

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