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ΑΡΧΕΙΑ ΕΛΛΗΝΙΚΗΣ ΙΑΤΡΙΚΗΣ 2007, 24(5):509

Clinical Immunology Quiz - Case 2

M. Speletas,[1]
F. Kalala,[1]
Z. Daniil[2]

[1]*Department of Immunology and Histocompatibility*
[2]*Department of Respiratory Medicine, Medical School, University of Thessaly, Larissa, Greece*

A 45-year-old male was admitted to the hospital complaining of increasing breathlessness and productive cough. His medical problems commenced in early childhood with recurrent episodes of pneumonia and he reported frequent episodes of sinusitis and respiratory infections as an adult. Upon physical examination, diffuse rhonchi and inspiratory crackles were heard at both the middle and lower respiratory areas, while no hepatosplenomegaly or adenopathy was observed. Laboratory studies revealed a white blood count of 5.9×10^9/L (normal values: $4\text{-}10\times10^9$/L), hemoglobin 13.8 g/dL (normal values: 14–16 g/dL), platelet count 197×10^9/L (normal values: $140–400\times10^9$/L). The white count differential was as follows: neutrophils 77.7%, lymphocytes 19.3%, monocytes 3%, without the presence of blasts. The erythrocyte sedimentation rate was 65 mm for the first hour (normal <20 mm) and C-reactive protein levels were 6.1 mg/dL (normal <0.8 mg/dL). Renal and liver function, electrolytes, and uric acid were all normal. High-resolution computed tomography of the chest revealed multiple, extensive, variably sized cystic bronchiectasis in both lungs. Peripheral blood of the patient was referred to the Immunology Lab for immunophenotyping that revealed reduced number of B-cells (fig. 1, Tab. 1).

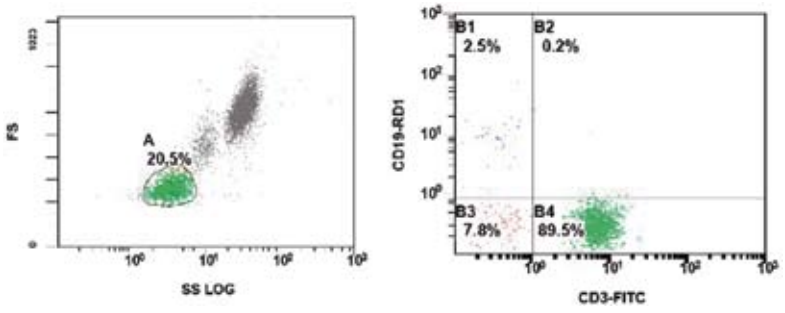

Figure 1. Flow cytometry analysis showing reduced number of B-cells.

Which is the next laboratory step?

Serum protein electrophoresis showed hypogammaglobulinemia and the quantitative evaluation of serum immunoglobulins revealed a marked decrease of IgG and IgA levels (tab. 1). The above findings (B-cell reduction, hypogammaglobulinemia and a medical history of recurrent respiratory infections from early childhood) were consistent with the diagnosis of common variable immunodeficiency (CVID).

Table 1. Lymphocyte subsets and quantitative serum immunoglobulins of the patient.

Blood lymphocyte subpopulations ($\times10^9$/L)		
Total lymphocyte count	1.215	[1.0–3.5]
T lymphocytes		
CD3	1.167	[0.9–2.5]
CD4	0.554	[0.5–1.5]
CD8	0.583	[0.25–1.0]
B lymphocytes		
CD19	0.031	[0.1–0.5]
Quantitative serum immunoglobulins (mg/dL)		
IgG	331	[874–1690]
IgA	28	[99–300]
IgM	78	[64–249]

Comment

CVID is the most prevalent primary immunodeficiency presenting in childhood or adult life. Peaks of onset occur in children aged 1–5 years and in persons aged 16–20 years. Interestingly, more than two-thirds of patients are aged 21 years or older when CVID is diagnosed. Most patients have reduced serum levels of IgG and IgA, normal or slightly reduced levels of IgM and normal or low numbers of B-cells. About one-third of the patients have some degree of abnormality of cell-mediated immunity. Affected individuals experience recurrent respiratory bacterial infections and a delayed diagnosis (as in our patient) can lead to permanent damage to the bronchi, resulting in bronchiectasis and pulmonary arterial hypertension. Autoimmune phenomena and a higher prevalence of malignancies (especially of lymphomas) are other common complications of the disease. The mainstay of treatment for CVID is Ig replacement therapy (400–600 mg/kg every 3–4 weeks) that can stop the cycle of recurrent infections.

Diagnosis: *Common variable immunodeficiency (CVID)*

Corresponding author:

M. Speletas, Department of Immunology and Histocompatibility, Medical School, Univesrity of Thessaly, University Hospital of Larissa, GR-41110 Larissa, Greece. Tel +302410681862, Fax +302410682580, e-mail: maspel@med.uth.gr