

CONTINUING MEDICAL EDUCATION ΣΥΝΕΧΙΖΟΜΕΝΗ ΙΑΤΡΙΚΗ ΕΚΠΑΙΔΕΥΣΗ

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ΑΡΧΕΙΑ ΕΛΛΗΝΙΚΗΣ ΙΑΤΡΙΚΗΣ 2008, 25(2):260

Clinical Immunology Quiz – Case 4

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Peripheral blood from a 34-year old female with lymphocytopenia, found during routine exams, was referred to the Immunology Lab for immunophenotyping. CD4+ lymphocytopenia (absolute number 224 per μL ; normal range 500–1500) (fig. 1A) was revealed. Further evaluation did not detect anti-HIV-1 and -2 antibodies and the patient did not fulfil the diagnostic criteria of non-Hodgkin lymphoma (NHL). Thus, the diagnosis of idiopathic CD4+ T-lymphocytopenia (ICL) was taken into account.

Which is the next laboratory test?

ICL is a common finding in patients with HIV infection and NHL, diseases that were excluded in our patient. However, a more detailed medical history revealed that the patient displayed dryness of the mouse (xerostomia) and eyes during the last two years that deteriorated during the last 2 months. Thus, further immunologic exams were performed, displaying positivity for antinuclear antibodies (ANA, title 1:1280), anti-ENA (>100 IU/mL, normal <20 IU/mL), anti-SSA (>100 IU/mL, normal <20 IU/mL) and anti-SSB (43.677 IU/mL, normal <20 IU/mL) antibodies (fig. 1B). Furthermore, a labial salivary gland biopsy exhibited the presence of infiltration by inflammatory cells, confirming the Sjögren's syndrome (SS) diagnosis.

Comment

ICL is detected in about 5% of patients with primary SS and is correlated mainly with the presence of anti-SSA antibodies. Since immunophenotyping is not included in the initial diagnostic approach of patients with suspected SS, it is possible that ICL in SS is underestimated. Bearing in mind that ICL has been implicated as a risk factor for the development of NHL, a condition with increased frequency in SS patients, the search for ICL in patients susceptible or diagnosed with SS should be encouraged.

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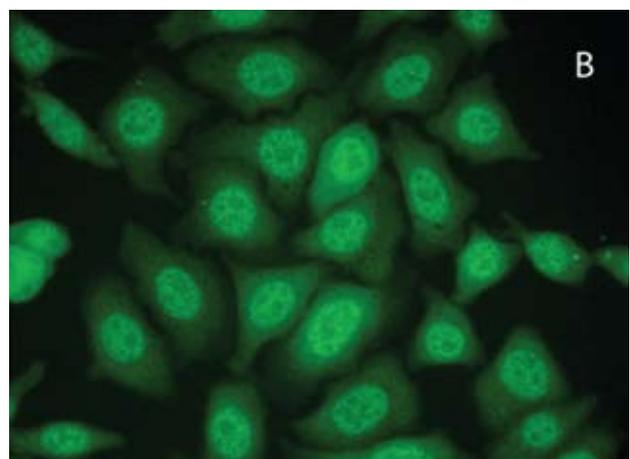
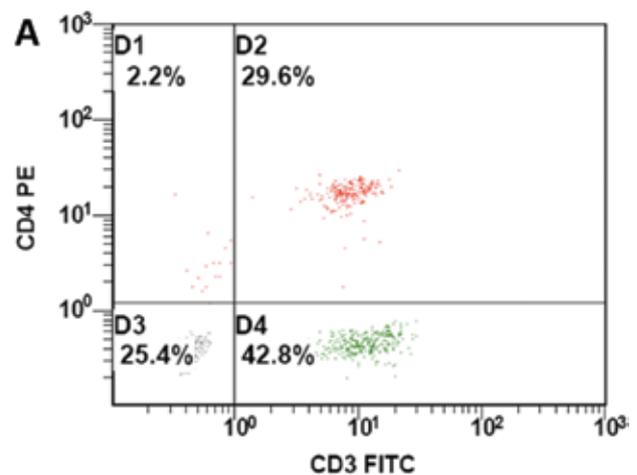


Figure 1. A. CD3 and CD4 expression on lymphocytes. B. ANA immunofluorescence on HEp-2 cells: Speckled pattern of fluorescence attributed to a high titer of anti-SSA antibodies.

Diagnosis: Idiopathic CD4+ lymphocytopenia in a patient with Sjögren's syndrome