Case report

Simultaneous appearance of mycosis fungoides and chronic lymphocytic leukemia in the same patient

K. KONSTANTOPOULOS¹,*, V. KAPSIMALIS², G. VAIPOULOS¹, CH. KOKKINIS², TH. PAPADAKI², K. PSARRA² and J. EKONOMIDOU²

¹ First Department of Internal Medicine, Athens University Medical School at Laikon Hospital, Athens, Greece
² Department of Immunology, Evangelismos Hospital, Athens, Greece

Abstract—A patient is presented having simultaneously chronic lymphocytic leukemia with a monoclonal B-lymphocyte population and mycosis fungoides with atypical T-cell population in the skin lesion and in the enlarged lymphoid nodes confirmed by detailed phenotyping.

Key words: Chronic lymphocytic leukemia; mycosis fungoides.

CASE REPORT

The patient, an 85-year-old Greek housewife presented with a pruritus rash on the thighs and axilla. The plaques had appeared 2 years before her present admission. The skin biopsy from the involved areas revealed histology typical for mycosis fungoides; at that time, her hematology showed a mild leukocytosis (WBC count 11 × 10⁹/l); no further investigation was conducted as the patient did not accept any further work-up and was lost to follow-up.

When, 2 years later she presented for deterioration of the skin lesions and persistence of the pruritus, on clinical examination she was found to be anemic; enlargement of axillary, inguinal and mediastinal lymph nodes was found but liver or spleen were not palpable. Hematology revealed anemia (Hb 100 g/l) and lymphocytosis (WBC 11 × 10⁹/l, lymphocytes being 70%). Peripheral blood lymphocyte marker analysis was performed according to standard protocol using the appropriate panel of monoclonal antibodies and the Epics XL MLL Coulter Electronics flow

*To whom correspondence should be addressed. K. Konstantopoulos MD, University of Athens School of Medicine, First Department of Medicine at Laikon Hospital, Athens, GR-11527, Greece.
cytometer; a monoclonal B-lymphocyte population was demonstrated (sIg-lambda, CD19, CD23, CD5 positive). Serum HTLV-1 antibodies were negative.

The bone marrow aspirate at that time indicated a small lymphocyte infiltration. Skin biopsy showed an epidermal atrophy and an intense infiltration by atypical lymphocytes and large cells with convoluted nuclei; the T-cells dominated by immunophenotype (LCA, CD3, CD4, CD5 positive; CD8, CD7, CD27, CD19, CD22, CD30 negative). Lymph node biopsy revealed a complete infiltration of the whole node architecture by small and large lymphocytes; immunophenotyping was similar to that of the skin infiltrates.

Molecular investigation of T and B cell monoclonality was conducted on material from both peripheral blood and lymph nodes by PCR amplification of the immunoglobulin heavy chain gene (IgH) and T-cell receptor $\gamma$-chain (Tcr-$\gamma$) using consensus primers directed to the Framework-3 (FR3) region and joining (J) segment of the genes. A monoclonal rearrangement of IgH and Tcr-$\gamma$ genes was detected. A similar study was not conducted on skin material for technical reasons.

DISCUSSION

Chronic lymphocytic leukemia (CLL) a B-cell malignancy (in 95% of the cases) involves the skin in some 8% of all cases [1, 2]. Mycosis fungoides is a low-grade cutaneous T-cell lymphoma (C-TCL); the co-existence of CLL with MF is rare [3, 4]. Benign conditions histologically similar to C-TCL, named MF simulants or Pseudo-T-cell lymphomas, can occur as reactive (polyclonal) T-cell infiltrates not in conjunction with CLL but mainly related to other factors as drugs, chemicals, dermatitis, arthropods or infections [5]. It should be noted that as a detailed immunological-molecular study is not always available, it is highly believed that many ‘C-TCL’ cases co-occurring with B-lymphocyte malignancies, in fact may be MF simulants or Pseudo-CTCLs [5]. A correlation of the concurrent T/B lymphoproliferative diseases with HTLV-1 infection is also reported [6] but it was not confirmed in our case.

All findings in our case indicate the concomitant existence of both a T-cell (Mycosis fungoides) and a B-cell malignancy (CLL) in the same patient, practically evolving over the same period of time, but retaining a rather clear ‘departmentalisation’ the B-clone dominating in peripheral blood whereas the T-clone is mainly restricted to the skin and lymph nodes. These compartments in the malignant clones, as indicated by morphology and immunophenotyping studies seem clear; in this context, we would like to mention that the molecular detection in the lymph node material of IgH clonal gene rearrangement by a very sensitive technic as PCR, given the considerable existence of blood circulating across it, cannot be interpreted as proving the origin, nor the homing of the B-cell clone in it.

Admittedly, the lack of a molecular study in skin material weakens the validity of our observation, although the immunophenotyping data are rather clear as far as the skin infiltration by T-cell population. A Southern blotting based molecular