

## T-CELL RECEPTOR GENE REARRANGEMENT STUDIES IN SÉZARY SYNDROME.

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### ABSTRACT

Sézary syndrome (SS) is a cutaneous T-cell lymphoma characterized by pruritic erythroderma, generalized adenopathy and circulating Sézary cells in peripheral blood. The Sézary cells' percentage and morphology necessary for the diagnosis are still unknown; therefore, recently T-lymphocyte receptors gene rearrangement has been employed thus allowing to study the monoclonality of lymphoproliferative disorders. In order to verify the specificity and the diagnostic relevance of the above mentioned method, the DNA obtained from the peripheral blood of 17 subjects (10 affected by SS and 7 by inflammatory erythroderma) was analyzed by the Southern Blot Analysis (SBA). A monoclonal gene rearrangement of the TcR  $\beta$ -chain was shown in 6 subjects affected by SS. In 2 patients the analysis was repeated during the therapy with recombinant interferon  $\alpha$ -2a (rIFN $\alpha$ -2a), in one patient the clonal population had disappeared. In 4 patients with CSCs and germline pattern in the peripheral blood ("false negatives") gene rearrangement bands were obtained in skin and lymphnodes except for in one patient. Seven controls with inflammatory erythroderma presented a germline pattern.

These results suggest that the TcR gene rearrangement, although it is a useful diagnostic tool, must always be associated with clinical, histopathological and immunophenotypical data; Furthermore, this study highlights the need to make consecutive sampling, possibly with material from various organs (peripheral blood, lymphnodes, skin).

### KEY WORDS

*T-cell receptor, gene rearrangement, Southern blot analysis, cutaneous T-cell lymphoma*

### INTRODUCTION

Sézary syndrome (SS) is a cutaneous T cell lymphoma (CTCL) characterized by pruritic erythroderma, generalized adenopathy and circulating atypical lymphoid cells in peripheral blood (circulating Sézary cells, CSC) (1). There are different opinions regarding

the percentage of CSC (2,3,4,5) and the morphological criteria (5,6) necessary to confirm the diagnosis: Cases of SS with moderately atypical nuclei of the circulating lymphocytes (7) and, on the contrary, reactive dermatitis or benign lymphoproliferative

