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CONTINUING MEDICAL EDUCATION
ΣΥΝΕΧΙΖΟΜΕΝΗ ΙΑΤΡΙΚΗ ΕΚΠΑΙΔΕΥΣΗ

Hematology Quiz – Case 51

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ΑΡΧΕΙΑ ΕΛΛΗΝΙΚΗΣ ΙΑΤΡΙΚΗΣ 2017, 34(1):136–138

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A 61-year-old woman presented to the Outpatient Department because of generalized skin rash, alopecia, nail dystrophy and intense pruritus. Two years ago the patient developed eczematous skin rash; there were skin patches of varying size, involving mainly the unexposed areas of the body, and were accompanied by itching. A tentative diagnosis of chronic eczematous dermatitis had been made, but the lesions had not responded to standard topical therapy. A skin biopsy had been performed which had shown lymphoid infiltration, mainly perivascular, without microabscesses and large atypical lymphocytes. Immunophenotyping had revealed predominance of CD4+ helper cells, with absent CD7 and Leu-8 expression. The diagnosis of "large plaque para-

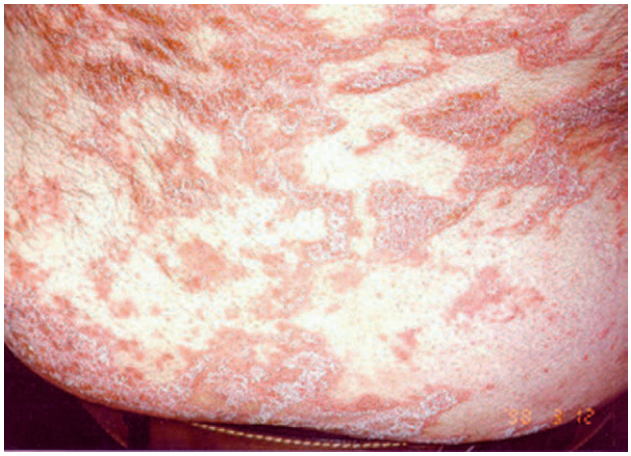

Figure 1

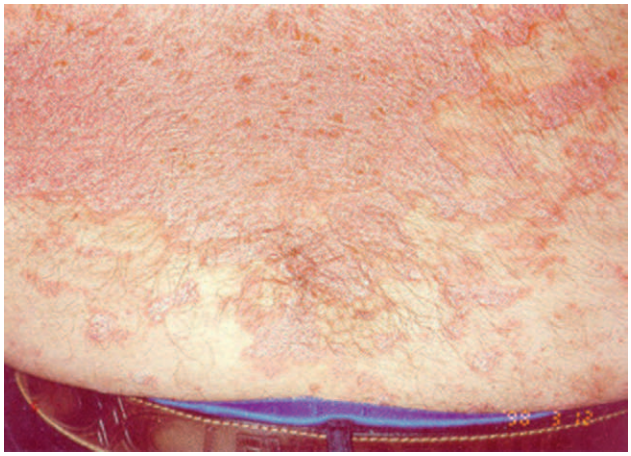

Figure 2

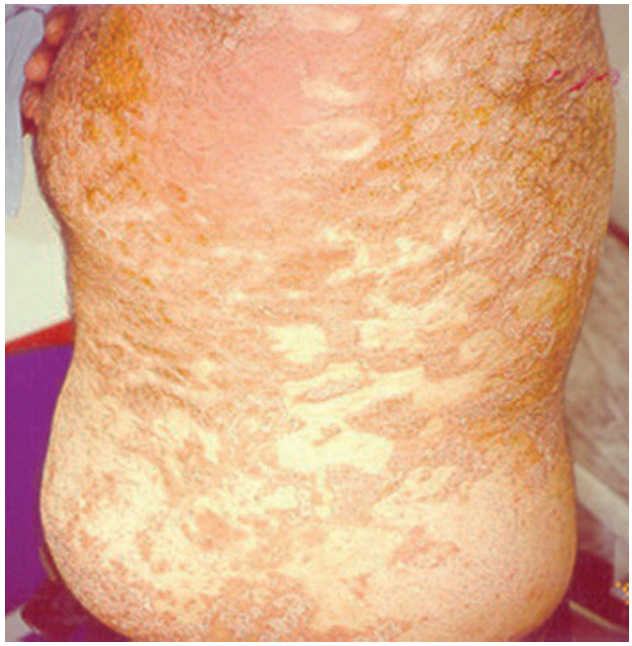

Figure 3

psoriasis" was made and the appropriate treatment resulted in the improvement of skin lesions and of itching. One and a half years later the skin lesions and the pruritus recurred, and the patient was admitted to our Department. On physical examination there were scaly macules, and erythematous brown patches and plaques with serpiginous borders in the whole body (figures 1 to 3). There were also superficial atrophic scars, alopecia and nail dystrophy. Cervical and supraclavicular microlymphadenopathy (smaller than 0.5 cm in diameter, painless and mobile), and mild, non-tender splenomegaly (2 cm below costal margin) were also present. The hematology profile was as follows: Hb 14.2 g/dL,

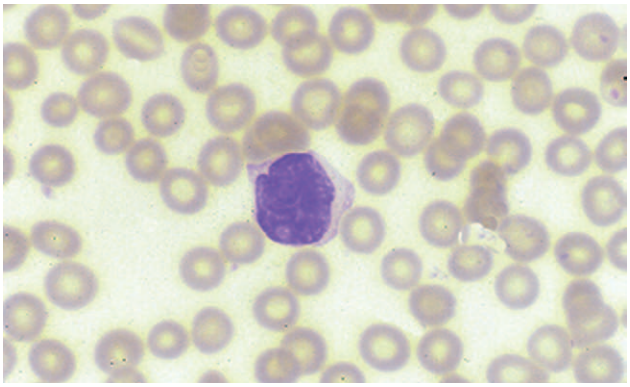

Figure 7

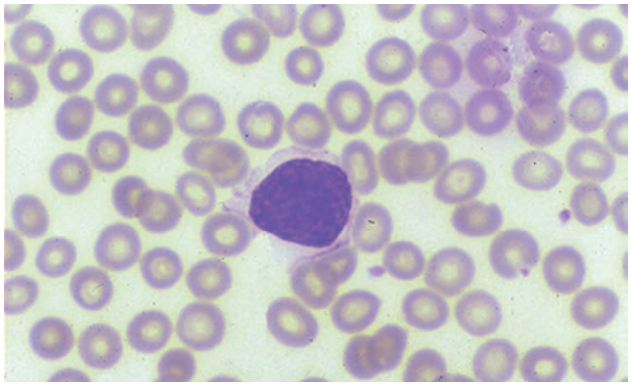

Figure 4

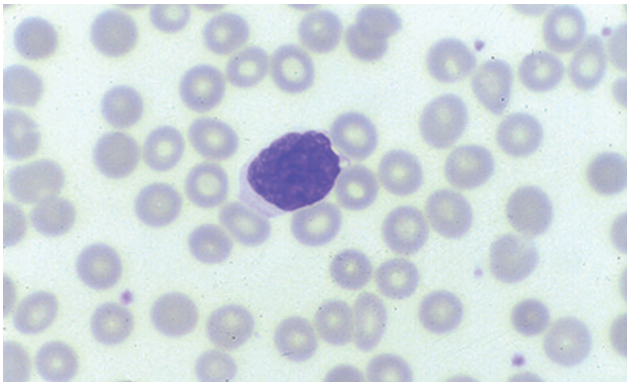

Figure 8

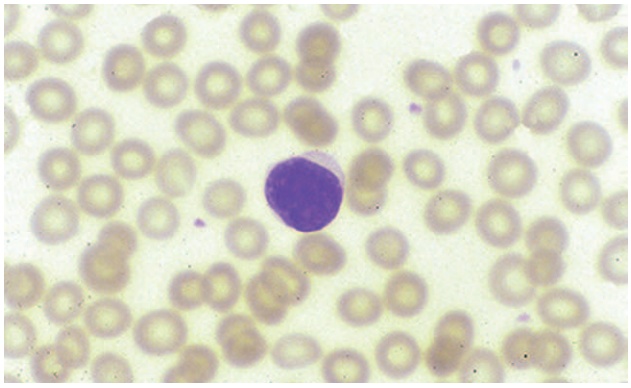

Figure 5

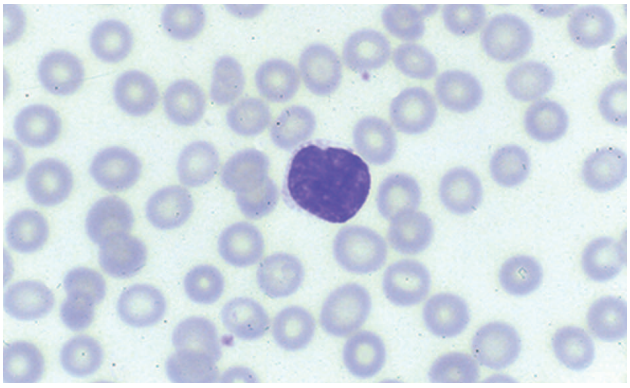

Figure 9

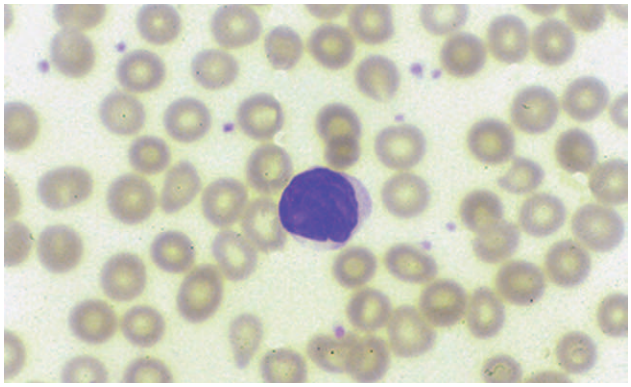

Figure 6

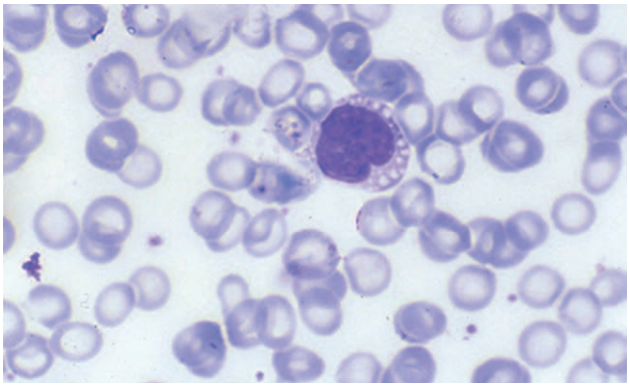

Figure 10

Ht 43%, WBC 6.7×10^9/L (differential count: neutrophils 51%, lymphocytes 43%, monocytes 3%, eosinophils 1%, and atypical lymphocytes 2%) and platelets 198×10^9/L. Peripheral blood smear showed atypical lymphocytes (figures 4 to 10). Serum biochemical tests revealed only a mild hypergammaglobulinemia without a monoclonal component. Computed tomography scans of the chest, abdomen and pelvis were normal. Bone marrow biopsy was also negative for lymphocytic infiltration. Lymph node histology revealed reactive changes. Skin biopsy revealed a moderately dense, band-like, infiltrate of numerous, small to intermediate-size lymphocytes in the superficial papillary dermis with epidermal infiltration and formation of microabscesses. Immunophenotyping confirmed the origin of these cells: CD2+, CD3+, CD4+, CD5+, CD1-, CD8-, CD25-. Total skin electron beam therapy and interferon administration resulted in a significant improvement of skin lesions and of patient's quality of life.

Comment

It is a characteristic generalized exfoliative erythroderma with extended epidermal infiltration by atypical lymphoid cells. Mycosis fungoides is a chronic T-cell cutaneous lymphoma that reduces initially in the skin and lymph nodes, but later it expands and infiltrates the spleen and the bone marrow. Many investigators believe that in association with Sézary syndrome it represents the manifestation of the same entity (Sézary syndrome when there are circulating abnormal lymphocytes).

Sézary syndrome represents a peripheral T-cell lymphoma with a leukemic picture. It consists of the leukemic equivalence of mycosis fungoides. The abnormal circulating cells in these cases are known as Sézary's cells which are differentiated in two types according to their size: (a) the type with large cells, and (b) the type of small cells.

The large Sézary's cells are almost tetraploid and are rarely seen. They are of large size than the neutrophils and sometimes the monocytes, they also have a large size round or ovaloid nucleus with deep reflexions and a coarse chromatin pattern and a high N/C ratio (4/5 of cell area) often presenting a cerebriform appearance of small size or not well visible nucleoli. The cytoplasm is basophilic without azurophilic granulation. Sézary's cell variant with small cells (Lutzner's cells) is the most frequent type. These cells have the size of a small lymphocyte but a nucleus containing corrugations, with cerebriform chromatin large Sézary's cells and the scanty cytoplasm may contain numerous perinuclear vacuolation (PAS positive staining). These small cells often have a higher N/C ratio than the small lymphocytes.

With a-napthyl acetate esterase (ANAE) there is a typical granular staining as in the normal T-helper cells, as well as a granular β-glucuronidase staining.

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