

CONTINUING MEDICAL EDUCATION ΣΥΝΕΧΙΖΟΜΕΝΗ ΙΑΤΡΙΚΗ ΕΚΠΑΙΔΕΥΣΗ

Hematology Quiz – Case 9

A 61-year-old man presented with easy fatigue, shortness of breath and headache, particularly with exercise, leg cramps and an erythematous papular rash, which had appeared on the upper extremities three months before his admission and increased gradually in number. He had a family history of secondary acute leukaemia (his mother died because of a secondary leukaemia 19 years after having chemotherapy for Hodgkin's lymphoma). His medical history included hypertension, and benign prostatic hypertrophy. On admission, his temperature was 36.6 °C and his blood pressure was 146/90 mmHg under amiloride and hydrochlorothiazide treatment in combination with metoprolol tartrate administration. The physical examination revealed skin lesions, 2–7 mm in diameter that were not painful, tender or pruritic, and were spread over the head (fig. 1) and the upper extremities (fig. 2). Other physical and neurological examinations showed no abnormalities. The respiratory and heart sounds were clear.

Laboratory tests indicated anaemia, with a haemoglobin level of 9.8 g/dL, leukocyte count of $10.8 \times 10^9/L$ (differential count: neutrophils 42%, lymphocytes 37%, monocytes 11%, eosinophils 1% and blasts of monocytic morphology 9%; fig. 3), and thrombocytopenia ($62 \times 10^9/L$). Lactate dehydrogenase (628 IU/L) and uric acid (8.4 mg/dL) were increased. C-reactive protein was negative. An electrocardiogram revealed premature ventricular contractions, and bradycardia with complete right bundle branch block.



Figure 1



Figure 2

ARCHIVES OF HELLENIC MEDICINE 2008, 25(6):842–843
ΑΡΧΕΙΑ ΕΛΛΗΝΙΚΗΣ ΙΑΤΡΙΚΗΣ 2008, 25(6):842–843

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The bone marrow aspiration showed an infiltration by blasts (62%) that were peroxidase positive. The marrow blasts displayed positive immunophenotypes for CD33, CD13, CD34, CD64, CD68, CD117, and HLA-DR. Chromosome analysis revealed normal

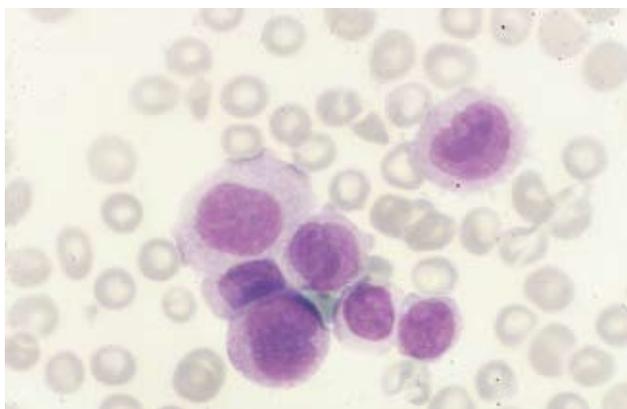


Figure 2

karyotypes of these cells. The skin biopsy specimen revealed the infiltration of blast cells in the dermis and subcutaneous fat tissue. These cells were identified with the chloracetate esterase stain, and were positive for leukocyte common antigen (CD45), CD33, CD117, and HLA-DR.

Based on the above laboratory and immunophenotypic characteristics the diagnosis was established and the patients started the appropriate therapy. The skin lesions disappeared and became pigmented after two courses of this therapy, and the patient achieved CR. He remained well, with no evidence of relapse nine months after his diagnosis.

Comment

Patients with leukaemia often show skin complications. The skin lesions can be categorized into two groups: nonspecific infiltration or "leukemids", such as vasculitis, purpura, exfoliative erythroderma and erythema nodosum; specific infiltration "leukaemia cutis". Leukaemia cutis is a condition characterized by the appearance of specific cutaneous infiltrates by leukemic cells. In addition to this condition, aleukemic leukaemia cutis is characterized by the absence of other signs of leukaemia, including pathological bone marrow findings. Although leukaemia cutis is reported in 2% of AML mainly in the M4 and M5 subtypes, it is occasionally also reported in patients with the M1, M2, and M3 subtypes. In most cases, such lesions are not painful, tender or pruritic. The immunophenotypic analysis of monocytoid blasts in the skin includes positive reactions for CD11b, CD13, CD14, CD33, CD45 (LCA), CD68, HLA-DR and, occasionally, CD15.

The differential diagnosis of the skin lesions in leukaemia includes metastatic carcinoma, fungal infections, drug eruptions, leukocytoclastic vasculitis and Sweet's syndrome. In general, the prognosis for patients with leukaemia cutis is poor and almost half of them died within six months of the diagnosis. Intensive chemotherapy for the underlying leukaemia was selected in most cases that have been reported in the literature and is considered to-date the standard care for these patients.

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