

CONTINUING MEDICAL EDUCATION ΣΥΝΕΧΙΖΟΜΕΝΗ ΙΑΤΡΙΚΗ ΕΚΠΑΙΔΕΥΣΗ

Hematology Quiz – Case 18

A 69-year-old woman presented with fatigue, mainly in exercise and an erythematous papular rash, which had appeared on her trunk one month before her admission and increased gradually. Her medical history included hypertension, hyperlipidemia and coronary disease.

On admission, her temperature was 36.8 °C and her blood pressure was 150/90 mmHg under amiloride and hydrochlorothiazide treatment in combination with metoprolol tartrate administration.

The physical examination revealed pallor and skin lesions, 5–25 mm in diameter that were not painful, tender or pruritic, and were spread over the trunk (fig. 1). Other physical and neurological examinations showed no abnormalities. The respiratory and heart sounds were clear.

Laboratory tests indicated anemia, with a hemoglobin level of 9.5 g/dL, leukocyte count of $12.6 \times 10^9/L$ (differential count: neutrophils 36%, lymphocytes 40%, monocytes 12%, eosinophils 1% and blasts of monocytic morphology 11%), and thrombocytopenia ($88 \times 10^9/L$). Lactate dehydrogenase (715 IU/L) and uric acid (8.1 mg/dL) were increased, while there were no other abnormalities in the biochemical profile of the patients.

The bone marrow aspiration showed an infiltration by blasts (55%) that were peroxidase positive. The marrow blasts displayed positive immunophenotyping for CD33, CD13, CD15, CD41a, CD56, CD64, CD68, CD117 and HLA-DR. A skin biopsy was performed which also revealed infiltration in the dermis and subcutaneous fat tissue by leukemic cells that were positive for CD33, CD15, CD45, CD56, CD68 and HLA-DR. Chromosome analysis was normal.

Based on the above laboratory and immunophenotypic characteristics the diagnosis was established and the patient started the appropriate therapy. The skin lesions disappeared and became pigmented after two courses of this therapy, and the patient achieved CR. She remained well, with no evidence of relapse nine months after her diagnosis. However, four months post therapy initiation, fine, transverse depressed lines that were parallel and evenly spaced developed on all of her fingernails (fig. 2).

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Comment

Acute myeloblastic leukemia (AML) may be associated with extramedullary infiltrates of leukemic blasts at diagnosis. Clinically, the extramedullary infiltrates include tumor nodules (myeloid or granulocytic sarcoma), skin infiltrates (leukemia cutis), meningeal infiltrates, gingival infiltrates (gum hypertrophy), or hepatosplenomegaly. Extramedullary infiltrates have been reported more commonly in myelo-monoblastic and monoblastic subtypes of AML. Our case had a myelo-monoblastic acute leukemia subtype. The frequency of extramedullary infiltrates in AML is reported to be between 20–40%. Chromosomal abnormalities, including t(8;21) and 11q23 abnormalities have been associated with extramedullary infiltrates manifesting as lymphadenopathy and gingival hypertrophy. 11q23 abnormalities are found more frequently in FAB-M4 and M5 AML; however, our patient had not such an anomaly in conventional cytogenetics. Several studies have established a correlation of extramedullary infiltrates with a low complete response rate and a shorter overall survival in these patients.

Differences in the cell surface phenotype are a potential biological explanation for the extramedullary infiltrates of some leukemic



Figure 1



Figure 2

cells. CD56, first described as a marker of natural-killer cells, has been shown to be identical to the previously recognized neural-cell adhesion molecule and to modulate homotypic neuronal growth. An association of CD56 with extramedullary infiltrates of AML has been reported. It is of great interest that our patient's blasts were CD56 positive.

Chemotherapy has been associated with changes in the nails that are related to interruption of mitotic activity of the proximal nail matrix and include mainly Beau's lines and onychomadesis. Beau's lines appear as transverse depressions in the nail plates. The depth of the depression represents the extent of damage. The duration of the insult may be estimated by the longitudinal width of the damage; normal nail growth is about 1 mm every six to ten days. Beau's lines may be caused by infection, surgery, trauma, Raynaud disease, pemphigus or chemotherapy (i.e. cyclophosphamide, vincristine, doxorubicin and taxoids). Our patients had received doxorubicin-based chemotherapy.

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