

Acute Myeloid Leukemia: Significance of the Skin Test

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Abstract Granulocytic sarcoma is generally associated to acute myeloblastic leukemia, either as its first clinical manifestation, tissue expression of the disease, or initial symptom of its recurrence. Less than 1 percent of patients will present with prominent extramedullary disease (i.e., myeloid sarcoma, also called granulocytic sarcoma, myeloblastoma, or chloroma). We report the case of a young patient suffering from granulocytic sarcoma of her right ankle which is symptom of its recurrence.

Keywords: granulocytic sarcoma, dermatological symptoms, ankle, knee, acute myeloid leukemia

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1. Introduction

Leukemia is a malignant neoplasm affecting the hematopoietic system. Following a generalization phase in the bone marrow and subsequent appearance of leukemic cells in the peripheral blood, extramedullary manifestation can occur in various organs of the body, including the skin. The classification of various types of leukemia is based on the biological behavior of disease as well as the morphological, immunophenotypical, and cytogenetic characteristics of neoplastic cells in acute and chronic, lymphocytic or myeloid forms of disease [1,2]. Acute leukemia has been divided into sub-types by the French-American-British (FAB) Cooperative Group: acute myeloid leukemia (AML) into types M0–M7 [3,4]. The diagnosis of AML was defined using the World Health Organization (WHO)'s definition of >20% blasts in the bone marrow or peripheral blood [3].

Dermatological symptoms seen in leukemia are divided by clinical and histopathological criteria into unspecific and specific skin changes. Unspecific skin changes are mostly dermatological diseases which have been associated with leukemia and which develop on the basis of abnormal hematopoiesis or as an expression of a cutaneous paraneoplastic disorder. [4,5]. Less than 1 percent of patients will present with prominent extramedullary disease (i.e., myeloid sarcoma, also called granulocytic sarcoma, myeloblastoma, or chloroma) [6]. Granulocytic sarcoma is generally associated to acute myeloblastic leukaemia, either as its first clinical manifestation, tissue expression of this disease, or initial symptom of its recurrence [7,8].

We report the case of a young patient suffering from granulocytic sarcoma (GS) of her right ankle which is symptom of its recurrence.

2. Case Report

A 46-year old female was diagnosed with AML-M5 in October 2010 due to peripheral smear (%35 myeloblasts), flow cytometric immunophenotype and result of bone marrow biopsy. She suffered weight loss, general situation defect, expectoration, cough and fever. Her body temperature was 38.3°C, pulse rate was 102 bpm and respiratory rate was 15/min. Laboratory values were as follows (Table 1). There was no family history for any disease. No abnormally enlarged lymph nodes were palpable in any part of her body. Her abdomen was not distended, and her spleen and liver were not palpable. We reviewed the extremity and performed further immunohistochemical analysis to determine whether or not the tumoral cells were myeloid. The patient was started induction chemotherapy with cytosine arabinoside and idarubicin. Remission was determined by marrow aspiration with no blast cells at day 27th after first chemotherapy cycle. A third cycle high dose cytarabine 2x3 gr/m²/day was given. During the third chemotherapy cycle, we observed a purple skin lesion which was fluffy, rough, measuring about 2.5x1.5 cm, mobile, demarcated, ulcerated and painless. It was on right ankle (Picture 1). New peripheral smear and bone marrow aspiration revealed recurrence. Excisional biopsy from right ankle was performed. The pathological diagnosis was granulocytic sarcoma (GS). Bone marrow biopsy revealed recurrence of acute myeloblastic leukaemia, which had identical characteristics compared to the initial sample.

Table 1. Laboratuvar parameters of patient

Parameter	Value	Normal Range
Leukocyte count	5,100/ μ L	4.000-10.000/ μ L
Haemoglobin	10,6 g/dL	13-17 g/dL
Haematocrit	33,5%	39,5-50,3%
Mean corpuscular volume	85,5 fL	80,7-90,5 fL
Mean corpuscular haemoglobin (MCH)	29,6 pg	27,2-33,5 pg
Mean platelet volume (MPV)	14,1 fL	6,6-10,6 fL
Platelet count	68.000/ μ L (in EDTA tubes)	150.000-450.000 / μ L
Platelet count	72.000/ μ L (in citrate tubes)	150.000-450.000 / μ L
Prothrombin time	12 sec	9-13,5 sec

**Picture 1.**

3. Discussion

Cutaneous paraneoplastic disease at AML includes pyoderma gangrenosum, Sweet syndrome, and insect-sting-like skin changes [4,5].

Less than 1 percent of patients will present with prominent extramedullary disease (i.e., myeloid sarcoma, also called granulocytic sarcoma, myeloblastoma, or chloroma) [6]. Extramedullary disease may present simultaneously with or precede bone marrow disease, and may be seen in relapse. Sites of isolated myeloid sarcoma include bone, periosteum, soft tissues, and lymph nodes, and less commonly the orbit, intestine, mediastinum, epidural region, uterus, and ovary [9,10,11].

GS is a rare entity that may precede the clinical manifestations of AML (it is often misdiagnosed like this case) or may occur as a relapse of a nonleukemic AML [8]. The patient's diagnosis of AML with isolated(GS) was, however, more rapid than typically observed despite the initial lack of circulating myeloblasts on peripheral blood

smear. So physical examination should be made carefully, GS may be first symptom of recurrence.

4. Conclusion

Any skin lesion at AML should be carefully evaluated and the physician should always be alert about GS.

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