

In the name of God

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Leukemia Cutis

(Answer to the Clinical Quiz of the Previous Issue)

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Abstract:

Skin infiltration in leukemia occurs rarely. Leukemia cutis has a wide range of cutaneous manifestations from maculopapular to tumorous stage. Patients with Leukemia cutis usually have concomitant systemic leukemia, but occasionally skin involvement precedes the involvement of the bone marrow or peripheral blood. Complete remission could be achieved with chemotherapy, however, the long term prognosis is poor.

Keywords: Leukemia cutis, Myelomonocytic leukemia

Case presentation and complete clinical course of the patient:

The patient is a 40 year old man, a case of acute myelomonocytic leukemia AML M4 (CD13+, CD33+, CD14+), who received induction of remission by 7;3 (cytarabine 100mg/m² and Idarubicin 12mg/kg) regimen, he also received 5;2 (cytarabine 100mg/m² and Idarubicin 12mg/kg) as consolidation treatment. He was candidate for allogenic stem cell transplantation from full HLA matched sibling, but a few days after consolidation chemotherapy he developed generalized erythematous raised skin papules that became confluent in some areas. Skin biopsy was taken and histology showed a diffuse infiltration of the dermis consisting of large cells with round to oval nuclei with strong positivity for myeloperoxidase (MPO) consistent with leukemic involvement. Bone marrow aspiration showed 70% blast.

He received FLAG (fludarabine 25mg/m², cytarabine 2gr/m² and GCSF 300µg daily) as salvage regimen and all of the skin lesion disappeared. Allogenic stem cell transplant was done from full HLA matched sibling. After transplant he had grade IV graft versus host disease (GVHD) with skin, gastrointestinal and liver involvement.

He died due to severity of GVHD and multiple organ failure.

Leukemia cutis:

Skin infiltration in leukemia occurs rarely, In some cases skin infiltration was evident at presentation and in others it developed while on treatment. The lesions

varied from maculopapular to tumorous stage. Complete remission could be achieved with chemotherapy, however, the long term prognosis is poor.⁽¹⁾

Leukemia cutis (LC) is a nonspecific term used for cutaneous manifestations of any type of leukemia. LC has a wide range of cutaneous manifestations, which can make it difficult to clinically distinguish from other skin lesions. Patients with LC usually have concomitant systemic leukemia, but occasionally skin involvement precedes the involvement of the bone marrow or peripheral blood.⁽²⁾

Leukemia cutis is a specific lesion of leukemia in which malignant hematopoietic cells invade the skin. It is a rare manifestation with unknown incidence. The lesions usually appear at the time of diagnosis of systemic disease or thereafter, but occasionally can occur before peripheral blood or bone marrow involvement (aleukemic leukemia cutis).⁽³⁾ Nonspecific cutaneous lesions occur more often and should be differentiated from specific lesions.⁽³⁾

The incidence of leukemia cutis varies depending on the type of leukemia. It is most frequently associated with acute myeloblastic leukemia, with higher incidences in the monocytic (AML-M5) and myelomonocytic (AML-M4) subtypes.^(4, 5)

The clinical appearance is variable with erythematous to violaceous papules or nodules being the most frequent lesions, followed by infiltrated plaques, to a generalized cutaneous eruption and erythroderma. They are frequently asymptomatic. Solitary lesions of AML may

present as an ulcerating greenish tumor termed chloroma.⁽⁶⁾

Aleukemic leukemia cutis is a rare condition characterized by the invasion of leukemic blasts into the skin before their appearance in the peripheral blood. Leukemia cutis usually occurs in patients with myeloid leukemia, especially the myelomonocytic and monocytic types of acute myeloblastic leukemia⁽⁷⁾

Retrospective studies show that cutaneous involvement of leukemia is associated with poor long-term survival. Despite aggressive therapy, most patients die within a few months.⁽⁸⁾ Leukemia cutis appears to be associated with a higher percentage of extramedullary leukemic involvement at other sites. Central nervous localization of acute monocytic leukemia occurs more frequently in patients with leukemia cutis (17 % versus 5-7 %)⁽⁹⁾

Other hematologic malignancies that can involve skin are:

1. Precursor B lymphoblastic leukemia⁽¹⁰⁾

2. Adult T Cell Leukemia Lymphoma⁽¹¹⁾

3. T-cell prolymphocytic leukemia⁽¹²⁾

4. Acute myelomonocytic and acute monocytic leukemia^(4,7)

5. Myelodysplastic syndrome (RAEB)⁽¹³⁾

6. Hodgkin's and nonhodgkin's lymphoma^(14,15)

Cutaneous involvement in myelodysplastic syndrome (MDS) was seen and is a poor prognostic factor, particularly is associated with progression to acute leukemia. However, the pathology of patient's lesions revealed a more sparse sprinkling of atypical mononuclear cells indicative of an inflammatory recruitment of leukemic cells to the dermis. Nonetheless, the guarded prognosis of this high-risk subtype of MDS mandates continued monitoring for development of LC and progression to leukemia.⁽¹³⁾



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