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Chronic Myeloid Leukemia Cutis: A case report

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Abstract
Leukemia cutis [LC] is the infiltration of neoplastic leukocytes or their precursors into the epidermis, the dermis, or the sub cutis, resulting in clinically identifiable cutaneous lesions. LC may follow, precede, or occur concomitantly with the diagnosis of systemic leukemia.
Here we present a case report of Chronic Myeloid Leukemia [CML] cutis in a 32-year-old female who was a known case of CML and presented with multiple erythematous nodules all over her body. Cytological study from the aspirates of these nodules revealed MPO [Myeloperoxidase] positive immature cells from myeloid series.

Keywords: Chronic, Myeloid, Leukemia, Cutis

Introduction
LC is defined as cutaneous infiltration by neoplastic leukocytes [myeloid or lymphoid] resulting in clinically identifiable cutaneous lesions. When composed of neoplastic granulocytic precursors, LC has been designated as myeloid sarcoma, granulocytic sarcoma, primary extra medullary leukemia or chloroma. When composed of neoplastic monocytic precursors [monoblasts and promonocytes], LC also has been designated as monoblastic sarcoma. LC has been described in patients with acute myeloid leukemia, chronic myeloproliferative disease, including CML, myelodysplastic syndromes, and myelodysplastic or Lymph proliferative diseases. In patients with chronic diseases, skin involvement is associated with transformation into a plastic phase and suggests disease progression [1].
The diagnosis of LC alone is nonspecific, and immunophenotyping is very important for generating a specific, clinically useful diagnosis. MPO and lysozyme are helpful in discriminating between myeloid and non-myeloid cells. MPO is strongly positive in most neoplasms of granulocytic lineage, and it can be weakly positive in a subset of monocytic neoplasms. In general, the development of LC portends a poor prognosis. Several studies indicate that, in the presence of LC in patients with AML or CML, the disease will follow an aggressive course and the survival is short [1].

Case report
A 32-year-old female, known case of CML (chronic phase) presented with [Fig.1] multiple erythematous nodules over the extremities and trunk.
On examination, she had a severe pallor, cervical lymphadenopathy, and hepatosplenomegaly. Her hemoglobin was 5.5gm/dl, total leucocyte count was 3.77 lakh/µl and differential nucleated cell count [Fig. 2] was as follows: blasts: 3%; promyelocytes: 4%; myelocytes: 8%; metamyelocytes: 20%; band forms: 8%, neutrophils: 25%, lymphocyte: 8%, eosinophil: 8%, basophil: 5%; monocyte: 1% and nRBC's: 10%. Her platelets count was 1.9 lakhs/cu.mm.
Aspiration cytology smears from the cutaneous nodule [Fig. 3] revealed mixture of mature and immature cells of myeloid series which were positive for myeloperoxidase [Fig. 4].

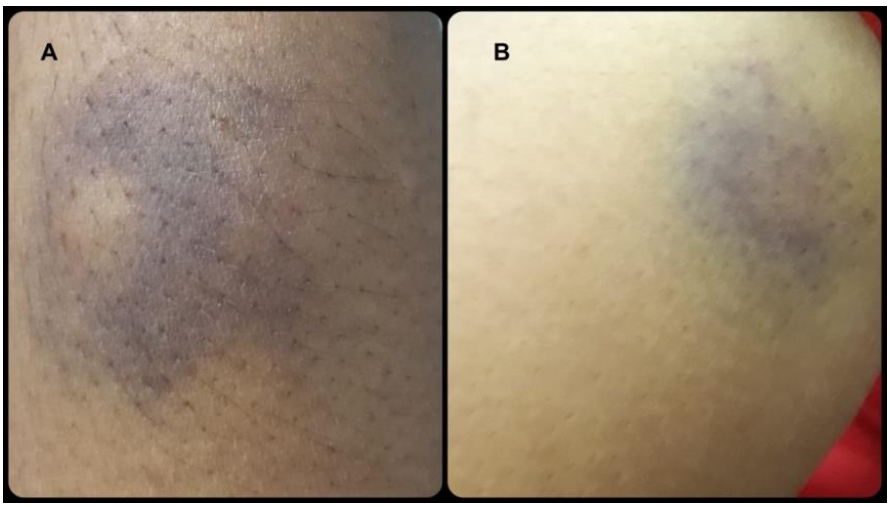

Fig 1: Gross examination of the patient showing multiple erythematous nodules over her both arms

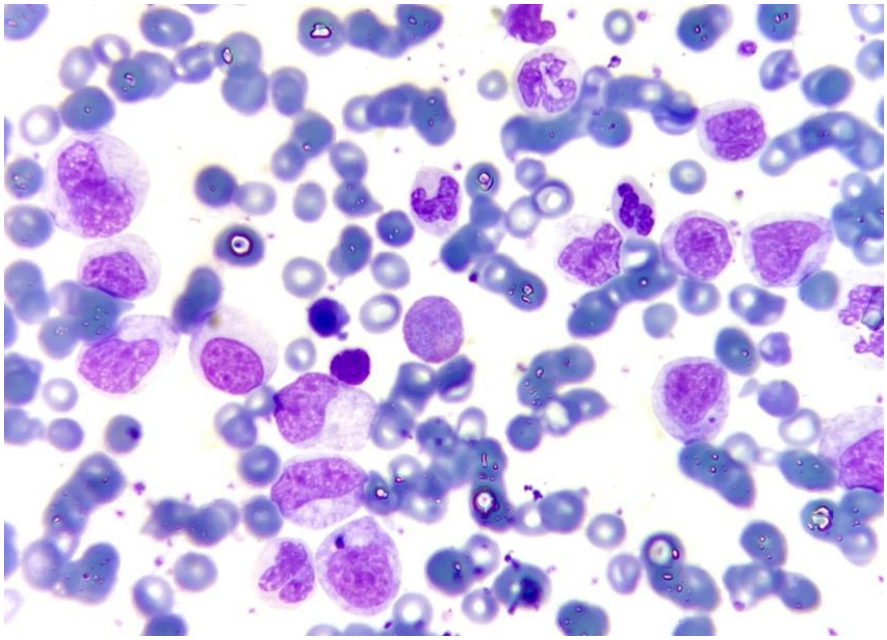

Fig 2: Peripheral smear shows immature cells from myeloid series, [Leishman's stain: 100 X]

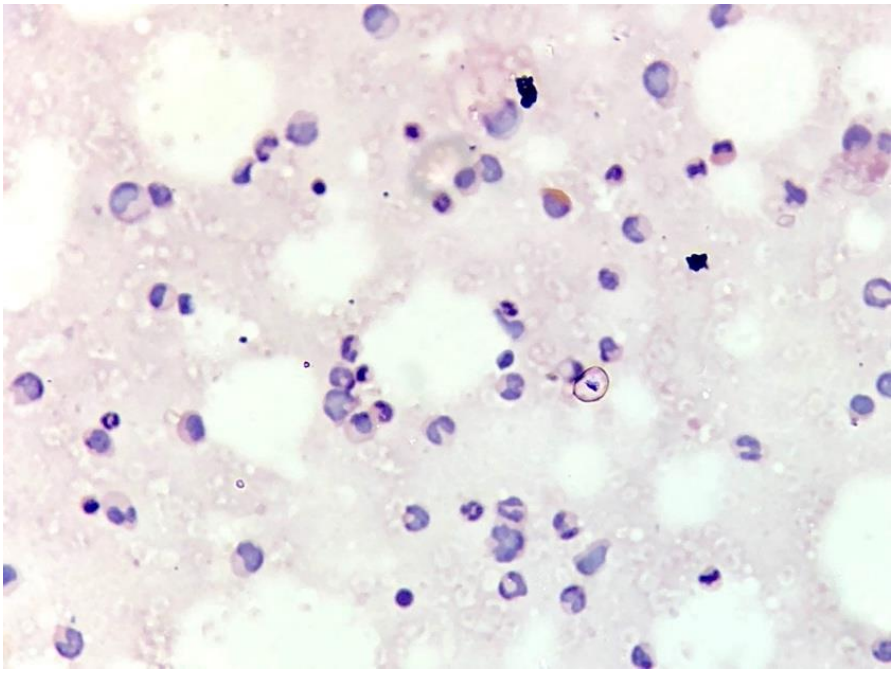

Fig 3: FNAC of the erythematous skin nodules: revealed immature cells from myeloid series. [PAP: 10X]

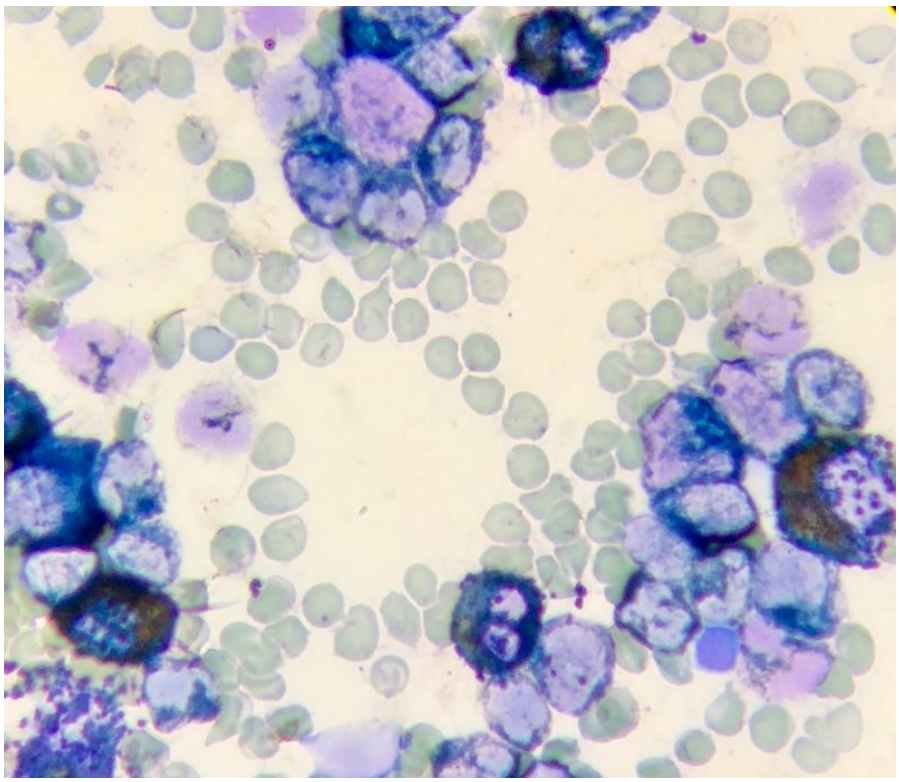

Fig 4: Shows MPO positive immature cells from myeloid series [MPO: 40X]

Discussion

LC is a nonspecific term used for cutaneous manifestations of any type of leukemia. Patients with LC usually have concomitant systemic leukemia, but occasionally skin involvement precedes the involvement of the bone marrow or peripheral blood. Thus, a skin biopsy can be the first indication of the presence of leukemia in a subset of patients. The immunophenotyping of routinely processed skin biopsy specimens is very useful in establishing the diagnosis of LC [1].

LC is the infiltration of neoplastic leucocytes or their precursors into the skin resulting in extensive clinical manifestations. Skin lesions may also represent the first clinical manifestation of this disease accompanied by normal peripheral smear which is termed as a leukemic leukemia. The commonest clinical presentation could range from erythematous or lilaceous macules, papules, plaques, or nodules. LC is described mostly in AML and acute myelocytes monocytic leukemia, it is rare in CML and is seen mostly during the blast crises. Presence of LC indicates a poor prognosis with an average survival of 9.4 months [2].

LC needs to be distinguished from sweet syndrome which is a cutaneous par neoplastic manifestation in patients of hematological malignancies which is characterized by the presence of mature neutrophils in the dermis [2]. We ruled out sweet syndrome as our case showed extensive infiltration of the dermis with immature granulocytes.

Patients with LC may have single or multiple skin lesions. The lesions are usually described as lilaceous, red-brown, or hemorrhagic papules, nodules, and plaques of varying sizes. Erythematous papules and nodules are reported as the most common clinical presentation. Legs are involved most, followed by arms, back, chest, scalp, and face. Leukemic infiltration tends to preferentially occur at sites of previous or concomitant inflammation. A particular type of leukemia can produce different skin lesions during the disease, even in the same patient. Most cases of LC occur after a diagnosis of systemic leukemia has been established. Concomitant involvement of skin and systemic leukemia has been observed in up to one third of the cases, and occasionally [<10% of cases], skin infiltration can occur before bone marrow or peripheral blood involvement and in the absence of systemic symptoms. A leukemia cutis lesions are usually widespread and papulonodular [1].

Cutaneous manifestations of leukemia vary in a broad spectrum. Li L *et al.* found cutaneous nodules in 40.0%, papules in 33.3%, Macules in 10% and masses in 40.0% of the patients in his study [3].

The development of LC imparts a poor prognosis, with a mortality of about 88 % within one year of the diagnosis [4].

Conclusion

LC occurs in 10-15% of patients of AML and even less frequently in CML [1]. The lesions, most commonly occur as erythematous nodules or papules and involves the extremities followed by trunk, face, and scalp. Most of these cases are seen after the diagnosis of leukemia has been established [4]. The nodules are typically firm or rubbery in consistency and can become pruritic if the patient is thrombocytopenic [5]. Presence of leukemia cutis indicates a poor prognosis with an average survival of 9.4 months [6].

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