Subacute dermal monocytic sarcoma: report of a case of monocytic leukemia with initial dermal presentation

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Monocytic sarcoma is a neoplasm of immature monocytes involving any extramedullary site such as skin. The definite diagnosis is important for adequate therapy, which is often delayed because of misdiagnosis. We report a 51-Year-old diabetic woman who presented with several erythematous itchy papules and plaques on her scalp, trunk and upper extremities during a five-month period. Clinical differential diagnoses were scleredema diabeticorum, scleromyxoma, mucinosis, eruptive histiocytosis, leprosy and sarcoidosis. Skin biopsy showed features of monocytic sarcoma. We present this case to alert dermatologists and dermatopathologists of erythematous skin lesions clinically resembling bland-looking dermatoses such as scleredema diabeticorum, scleromyxoma, mucinosis and eruptive histiocytosis, which, in rare instances, may be a presenting feature of a monoblastic leukemia with initial dermal presentation.

Keywords: leukemia cutis, monocytic leukemia, monocytic sarcoma

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INTRODUCTION

Generally, monocytic sarcoma is a rare cutaneous manifestation of a neoplastic change in myeloid cells line with dermal infiltration of monoblasts preceding or concurrently bone marrow or peripheral blood involvement. Monocytic sarcoma may presented with bacterial ecthyma, bullous lesions, disseminated erythematous maculaes, papules and nodules or an erythematous maculopapular rash that may be clinically resembled viral exanthema [1-3]. When such presentations preceding the signs and symptoms of systemic malignant involvement, proper diagnosis is very difficult for dermatologists.

Extramedullary accumulation of immature myeloid cells identify myeloid sarcoma in the World Health Organization (WHO) classification that can develop in any part of the body, but commonly involved sites include subperiosteal bone structures of the skull, paranasal sinuses, sternum, ribs, vertebrae, and pelvis. Lymph nodes and skin are also commonly involved. They may precede or occur concurrently with acute myeloid leukemia, or may be a sign of the blastic transformation of chronic myeloproliferative disorders or myelodysplastic syndromes. They may also be a sign of relapse in the treated patients [4-6]. The rate of occurrence is approximately 1.4% to 9% of patients with acute myeloid leukemia (AML). They are signified by a diffuse infiltrate composed of medium to large size cells. Myeloid sarcoma is frequently mistaken for non-Hodgkin lymphoma (NHL), small round cell tumor (neuroblastoma, rhabdomyosarcoma, Ewing's sarcoma (primitive neuroectodermal tumor (PNET), and medulloblastoma), and undifferentiated carcinoma. About 50% of cases are misdiagnosed when immunohistochemistry (IHC) methods are not used. The most common suggested diagnosis was that of a NHL. Four variants can be distinguished with IHC: granulocytic, monoblastic, myelomonoblastic and megakaryoblastic. Monocytic sarcoma is a neoplasm of immature monocytes involving any extramedullary site such as skin. The definite diagnosis is important for adequate

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therapy, which is often delayed because of misdiagnosis [5].

CASE REPORT

We report a 51-year-old diabetic woman who presented with several erythematous itchy papules and plaques on her scalp, trunk and upper extremities since 5 months before admission at the dermatology clinic of Farshchian Hospital in Hamadan, Iran (Figure 1). Clinical differential diagnosis were scleredema diabeticorum, scleromyxoma, mucinosis, eruptive histiocytosis, lepra and sarcoidosis. She had a drug history of metformin, glibenclamide, gemfibrozil and dexamethasone. She had no family history of malignancy. Physical examination revealed axillary lymphadenopathy but no evidence of hepatosplenomegaly.

Pathologic differential diagnosis of the previous skin biopsy in two months later were dermatofibrosarcoma protuberans and histiocytosis. Microscopic evaluation of the skin biopsy show neoplastic proliferation of atypical cells with kidney shape lobulated nuclei and prominent eosinophilic cytoplasm that arranged with diffuse pattern in dermis. Mitotic activity and apoptosis are obvious. Epidermal and adnexal invasion were absent (Figures 2,3). The diagnosis is consistent with monocytic sarcoma. The peripheral blood smear revealed:10700 white blood cell (WBC) count with a differentiation of 37% monocytes; 18% monoblasts, 35% lymphocytes, 2% neutrophils, and 1% eosinophils (Figure 4). Bone marrow biopsy and aspiration were suggestive for AML-M5 (Figure 5).

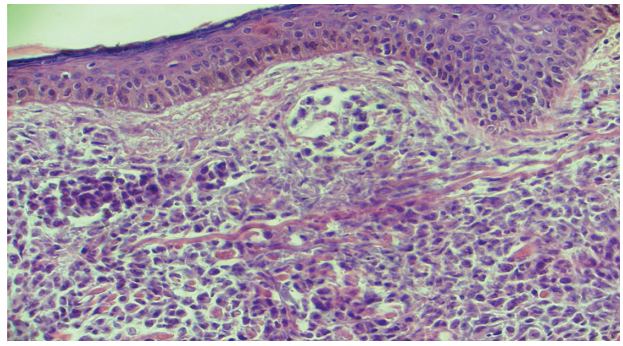

Figure 2. Skin biopsy show neoplastic proliferation of atypical cells with diffuse pattern in dermis (H&E ×10)

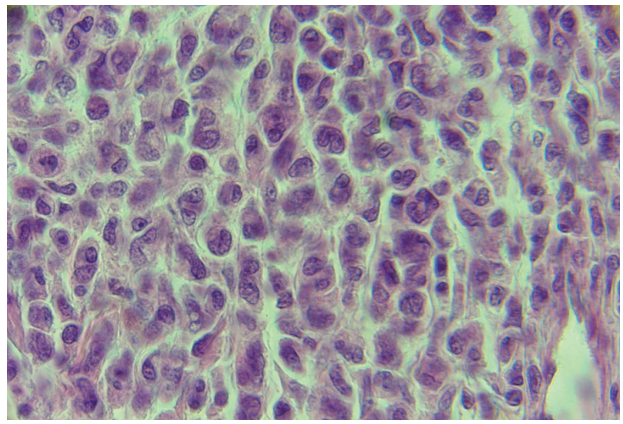

Figure 3. Skin biopsy show neoplastic proliferation of atypical cells with kidney shape lobulated nuclei (H&E ×40)

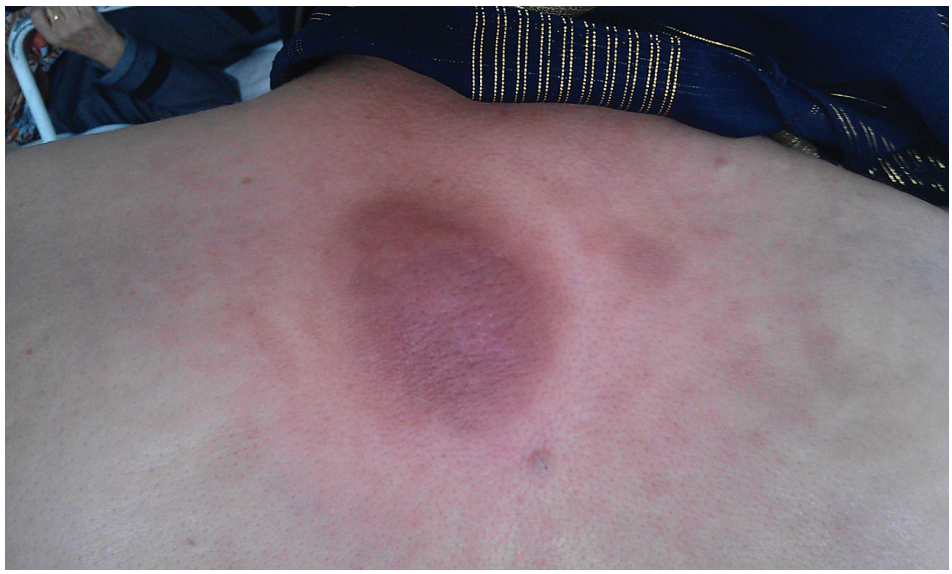

Figure 1. Pruritic erythematous papules and plaque on the upper trunk.

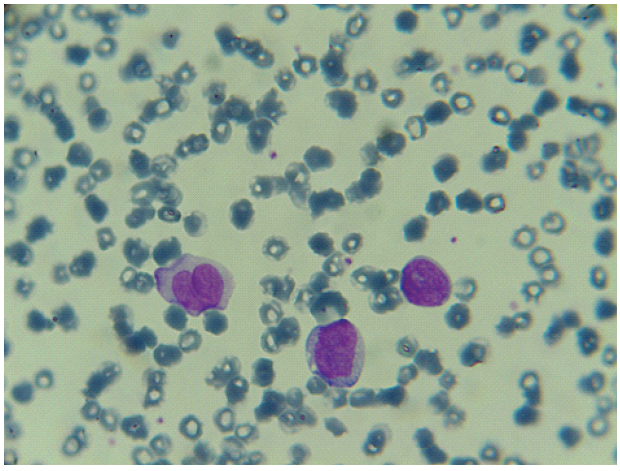

Figure 4. Peripheral blood smear revealing monoblasts (Giemsa ×100)

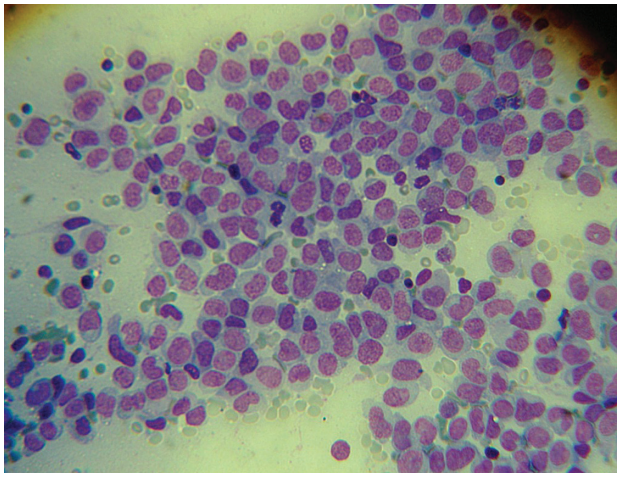

Figure 5. Bone marrow biopsy and aspiration was suggestive for AML-M5 (Giemsa ×100)

The patient died during chemotherapy secondary to disseminated intravascular coagulation (DIC).

DISCUSSION

We present this case to alert dermatologists and dermatopathologists of erythematous skin lesions clinically resembling bland-looking dermatoses such as scleredema diabeticorum, scleromyxoma, mucinosis and eruptive histiocytosis, which, in rare instances, may be a presenting feature of a monoblastic leukemia with initial dermal presentation. It is difficult for dermatopathologists to distinguish this entity (monoblastic leukemia cutis) even using IHC methods since monoblasts are negative for hemopoietic precursor cell antigens like CD34, terminal deoxynucleotidy1 transferase (TdT) and CD117 [7]. The CD34 is expressed by the blasts of most forms of leukemia, but not by the cells of most malignant lymphomas. CD68, normally expressed by macrophages, is found on the cells of myeloid and myelomonocytic leukemias. An important caveat is that myeloblasts react with a variety of reagents that also identify lymphoid cells, including CD43, CD45RO, and CD20. IHC is sometimes positive for CD 7, CD 79a, and CD 56 [7]. The expression of CD 99 and CD 117, which can now be done on paraffin sections may be helpful to detect blasts of granulocytic origin [5].

Since there is immunophenotypic differences between skin and bone marrow blasts using the algorithm for immunohistochemical approach to diagnose leukemia cutis in skin specimen is necessary and CD3/CD20, CD43, MPO, CD68, CD56, CD117 should be assessed, respectively [6,8]. NHL and myeloid sarcoma (MS) may have some similar morphological features and both express some leukocyte antigens, such as CD43 and CD45. B-cell and T-cell lymphomas were excluded by negative stains for CD20, CD79a and CD3, respectively. Similarly blastic NK cell lymphomas express CD43, CD4, CD56, and, unlike MS, are negative for MPO, CD33, CD117, lysozyme, and CD68 [9].

Nevertheless, a practical way to confirm the diagnosis of MS, in routine practice, is the demonstration of alpha-1 -antitrypsin (A1AT) and/or lysozyme by the immunoperoxidase technique, on fixed paraffin-embedded tissues [10]. The diagnosis is not true in about 50% of cases when IHC is not applied. Myeloid sarcomas should be treated in the same way as acute myeloblastic leukemia. Disease progression and prognosis are identical for the both conditions [5].

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