

Different faces of leukemia cutis: Presenting as purpura fulminans and lupus like butterfly rash

Sir,

Leukemia cutis poses diagnostic difficulty due to its varied clinical manifestations. Appearance of lesions of leukemia cutis indicates a poor prognosis. We report two cases of leukemia cutis with varied presentations.

Our first patient, a 27-year-old male, a known case of myelodysplastic syndrome (MDS) for 2 years, presented with fever, hemorrhagic bullae, erythematous and purpuric plaques on the face and upper extremities for 10 days [Figures 1 and 2]. He had taken oral Norfloxacin for fever. Bone marrow biopsy done 1 year ago showed hypocellular marrow with dysplastic changes in myeloid series with megaloblastic anemia. For his MDS, the patient was on oral hematinics and had received blood transfusions in the past. He had no history of bleeding from any other site. On general examination, the patient was febrile (38.4°C), had tachycardia and was normotensive. His respiratory rate was increased (20/min). He had pallor. A clinical differential diagnosis of Sweet's syndrome, bullous fixed drug eruption and purpura fulminans was considered.

His hemoglobin was 9.1 g/dl. Total and differential counts were normal. Peripheral smear detected macrocytosis, polychromasia, and a few abnormal cells (six myelocytes, a few schistocytes). Platelets were decreased (70,000/mm³). Liver and renal function parameters were deranged [Serum glutamic oxaloacetic transaminase (SGOT): 66 IU (up to 40); Serum glutamic pyruvic transaminase (SGPT): 143 IU (up to 40); Blood urea nitrogen (BUN): 20 mg% (10–15)]. His urine showed 40–50 RBCs/hpf, with no growth on blood culture. Hematological tests showed disseminated intravascular coagulation (DIC) [fibrinogen degradation product: 80–160 µg/ml (0–5); fibrinogen: 643 mg/dl (200–400); prothrombin time: 10.8 seconds (7.4–10.5); partial thromboplastin time:

37.6 seconds [22.3–33.4]). Skin biopsy of the purpuric plaque from the face showed normal epidermis, superficial and deep nodular, interstitial as well as perivascular infiltrates of myeloid blast cells extending to subcutaneous fat and erector pilorum muscle with extravasation of RBCs [Figure 3]. Bone marrow biopsy could not be done at this stage to rule out leukemia due to the fulminant course of the disease. The patient died within a day.

The second patient, a 48-year-old female, diagnosed of acute myeloid leukemia, for 1.5 months, was referred for multiple asymptomatic edematous, erythematous and purpuric plaques on the face, simulating a butterfly rash [Figure 4]. There were purpuric plaques and papules on the upper extremities with a few lesions showing central blistering for 10 days, along with intermittent fever. There was no other significant history. On general examination, she had pallor and submandibular lymphadenopathy. Systemic examination was normal.

A clinical differential diagnosis of Sweet's syndrome, leukemia cutis was considered.

The patient was anemic (Hb: 7.8 g%). Total counts were raised (56,800/mm³). Peripheral smear showed 70% myeloblasts, P-19, L-10. Platelets were decreased (39,000/mm³). Bone marrow aspiration was hypercellular with decrease in all three cell lines and 90% blast cells. The patient was in DIC [fibrinogen degradation products: 654 mg/dl (200–400); di-dimer products: positive]. Skin biopsy showed grenz zone, superficial and deep perivascular as well as interstitial infiltrates of blast cells involving the subcutaneous fat and erector pilori muscle. Immunohistochemistry of skin biopsy was CD43+ and myeloperoxidase positive, suggestive of myeloid leukemia. The patient was started on injection cytarabine 150 mg s.c. BD; however, she succumbed to death within 7 days.

There are a few reports of leukemia cutis in our



Figure 1: Purpuric plaques on the face



Figure 2: Hemorrhagic bullae simulating purpura fulminans

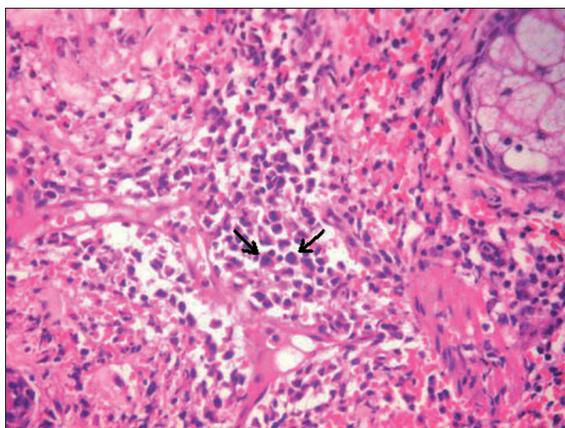


Figure 3: Interstitial myeloid blast cells (marked with arrows) (H and E, x200)



Figure 4: Purpuric plaque on the face resembling butterfly like rash

country. The reported prevalence by Angis *et al.* is 2.9–3.7%.^[1] Seven percent of the cases present as aleukemic leukemia cutis. Leukemia cutis manifests in different types of leukemia with highest involvement in adult T cell leukemia, followed by acute myelogenous leukemia as seen in our second case.^[2] It can also occur in patients with MDS and may be an early manifestation of leukemic transformation in these patients as seen in our first case.^[3] Approximately, 10–40% of MDS cases terminate in acute leukemia.^[4] This may be important in identifying high-risk patients.

Cutaneous presentations of leukemia cutis are described in Table 1. Hemorrhagic bulla and butterfly like rash, as seen in our cases, have been reported in a few cases in literature. Hemorrhagic lesions, as seen in our first case, are commonly found in the subtypes of leukemia that are associated with coagulopathies. The etiology for bullae is less well understood. No association has been found with the type of leukemia cutis and butterfly rash. It only highlights one of the unusual modes of presentation. In our patient with MDS, cutaneous purpura was observed when the platelet count was 72,000/mm³, whereas in the second patient the platelet count was 39,000/mm³. Baer *et al.* have not attributed the development of purpura in lesions of leukemia cutis to thrombocytopenia or hyperleukocytosis.^[2] It may be due to capillary fragility or leukemic infiltration of the capillaries.

The survival rate is 30% at 2 years in patients with acute myeloid leukopenia (AML) without skin lesions as compared to 6% in patients with skin lesions indicating grave prognosis of leukemia cutis. Both our patients had a fatal outcome. These cases have been reported due to their uncommon manifestations and to emphasize that a dermatologist is quite instrumental in the diagnosis of leukemia cutis.

Table 1: Morphologic types of leukemia cutis^[5,6]

Erythematous or skin colored or violaceous macules, papules, plaques, nodules – 60%

Exanthematous eruptions

Purpura*

Bullae*

Ulcers

Butterfly like rash*

Leonine facies

Others: Stasis dermatitis, erythema nodosum, erythema annulare centrifugum, pyoderma gangrenosum, urticaria, urticaria pigmentosum, guttate psoriasis, chronic paronychia, subungual leukemia cutis and macular erythema

*Seen in our patients

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