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Aleukemic leukemia cutis

Sami S. Yones, Dip Derm, MSc, FCD, MD; Mohammed Ghazavi Khorasgani , MD; Taher Iqbal, Dip Derm, MSc; Nazirin Arrifin, MRCP; Rohaj Mehta, MRCP; Gosial Skibinska, MD; Asmaa Karim, MD; Charlot Grech, MRCP; Gleb Ivanov, MD, MRCP, FRCPath*; Mohson Khorshid, MD, MRCP

e-mail: yones5@yahoo.com

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Dermatology Department
Basildon and Thurrock University Hospitals

* Hematology Department
Southend University Hospital

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Abstract:

It is a rare condition characterised by the infiltration of skin by leukemic cells before their appearance in the peripheral blood or bone marrow. We report a case of aleukemic leukemia cutis in a fit gentleman, who rapidly progressed to acute myeloid leukomia. However, the outcome was fatal within 3 months of diagnosis of skin lesions.

Case Report:

An 82 year old retired policeman was referred to dermatology department with several weeks' history of a symptomatic generalised erythematous papulo-nodular rash on face, upper limbs, trunk, and lower limbs following a flu jab.

He had a past medical history of asthma and hypertension and he was on ibesartan and salbutamol inhaler. There was no improvement following oral and topical steroid and also changing his antihypertensive medication Ibesartan to doxazocin by his GP.

On examination he looked very well with no lymphadenopathy or organomegally. The rash **(Fig 1a, 1b, 1c, 1d)** was reddish-purple infiltrative, firm, papulo-nodular lesions about 0.5 to 1.5 cm in diameter with a smooth surface on the torso and proximal parts of the extremities. All blood tests including full blood counts,

erythrocyte sedimentation rate, liver and renal functions tests, blood film, autoantibody screen and X-ray chest were normal.

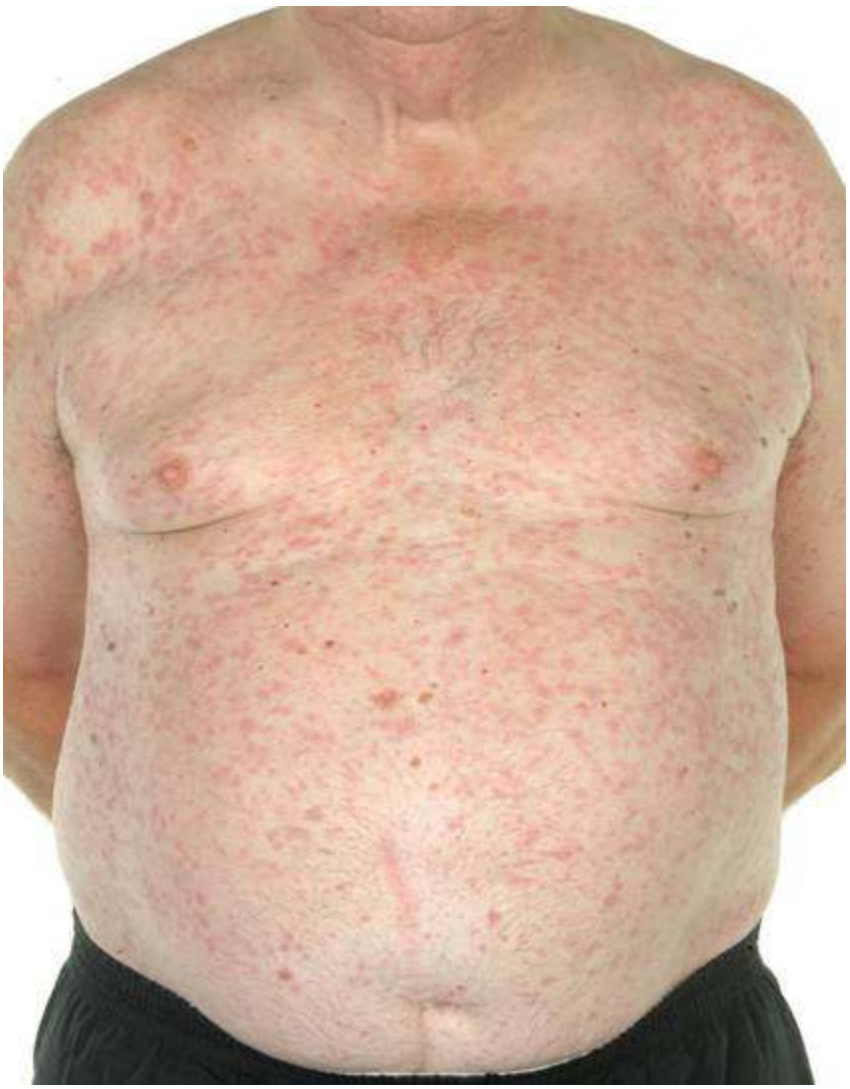

Fig 1a: Leukaemia cutis affecting the trunk

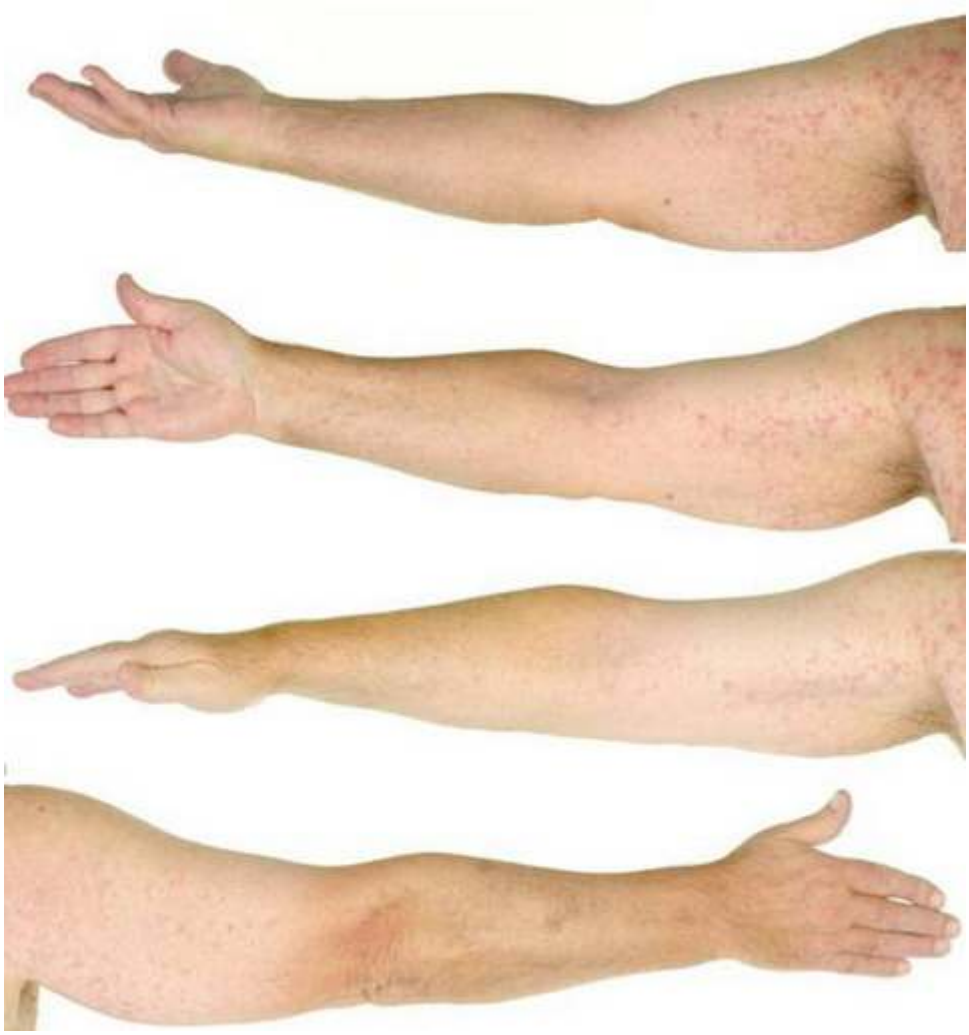

Fig 1b: Leukaemia cutis affecting the arms

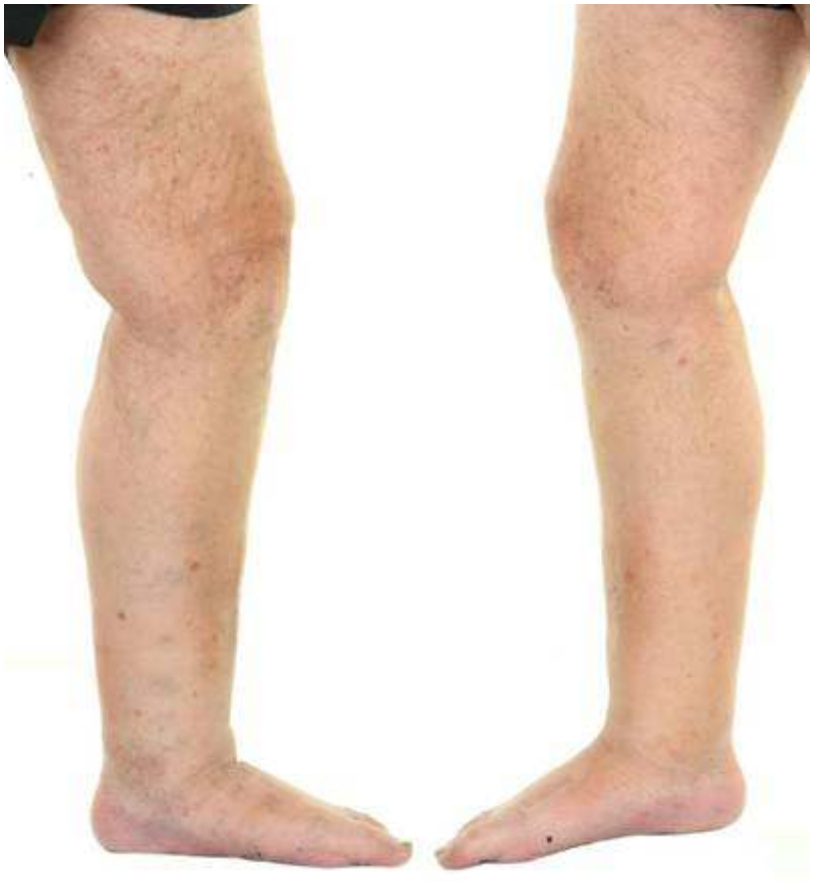

Fig 1c: Leukaemia cutis affecting the legs.

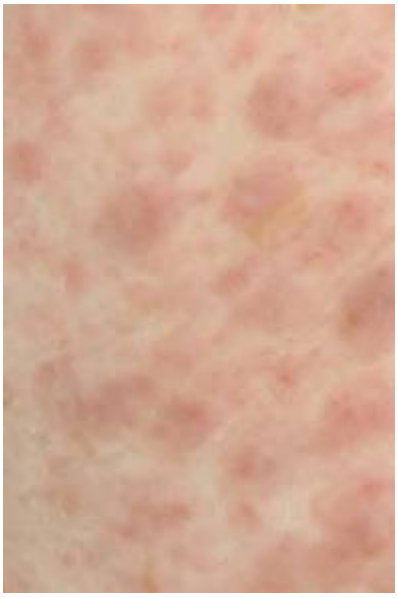

Fig 1d: Close view of the leukaemia cutis lesions.

A skin biopsy was taken from the chest and he was referred to the haematologist urgently with the suspicious clinical diagnosis of leukaemia cutis. By the time he was seen by the haematologist, the rash started to coalesce into geographic patches covering the lower trunk, abdomen, and proximal extremities.

Second blood film 3 months following the appearance of the skin lesions showed 20 % circulating myeloid blasts **(Fig. 2)**, positive to CD33, CD117, and FLT3 phenotype. Skin biopsy showed infiltrative sheets of atypical blastoid cells in the dermis mainly perivascular and periadnexal **(Fig. 3a, 3b)** with positive myeloperxidase staining **(Fig. 4)**, compatible with the diagnosis of acute myeloid leukaemia (AML) which was confirmed by the bone marrow biopsy.

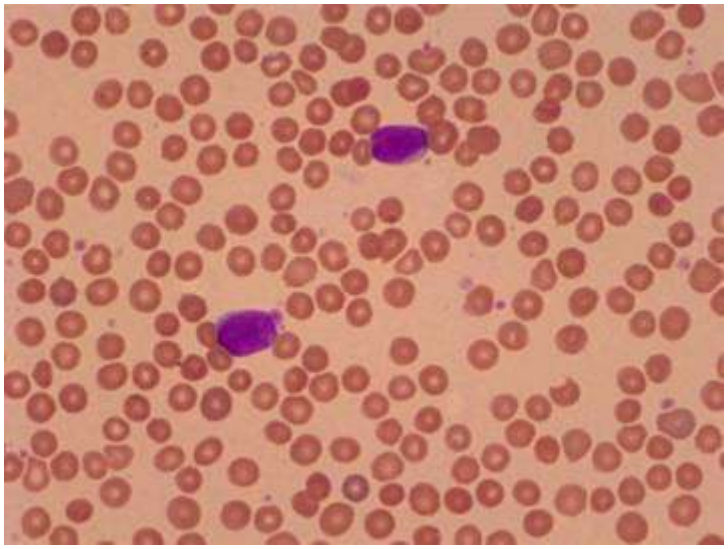

Fig 2: A peripheral blood smear containing blast cells having a little cytoplasm.

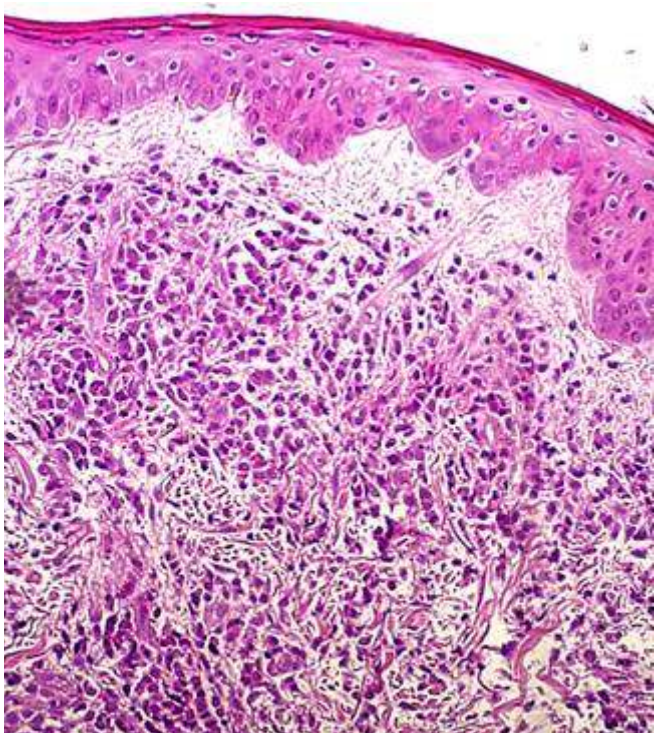

Fig 3a: Atrophic epidermis, grenz zone and diffuse infiltrate of blast cells in the dermis (H & E, X 40).

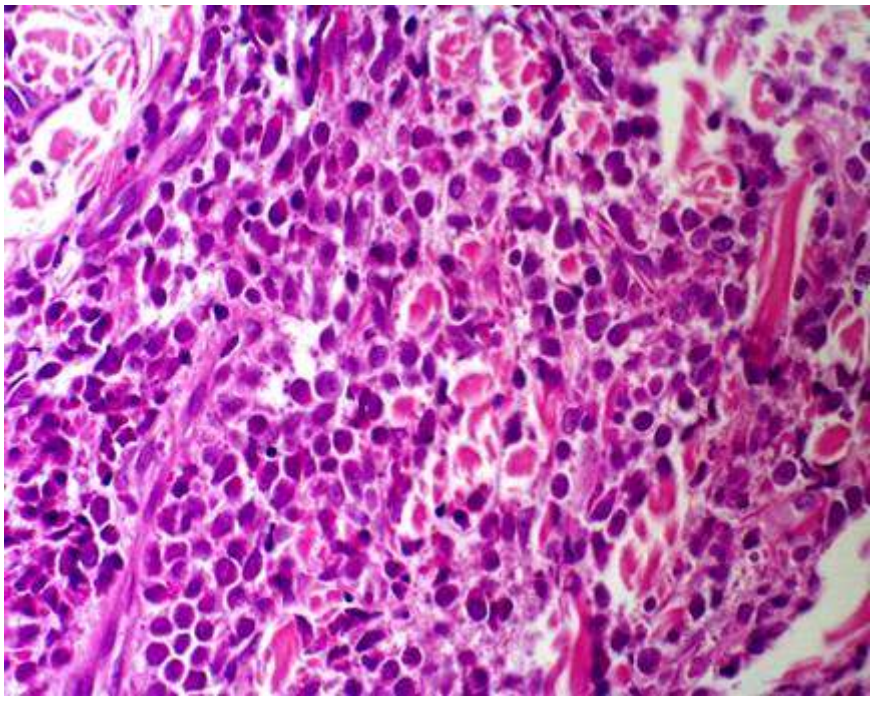

Fig 3b: (H & E, X 100), demonstrates densely blast cells in the dermis.

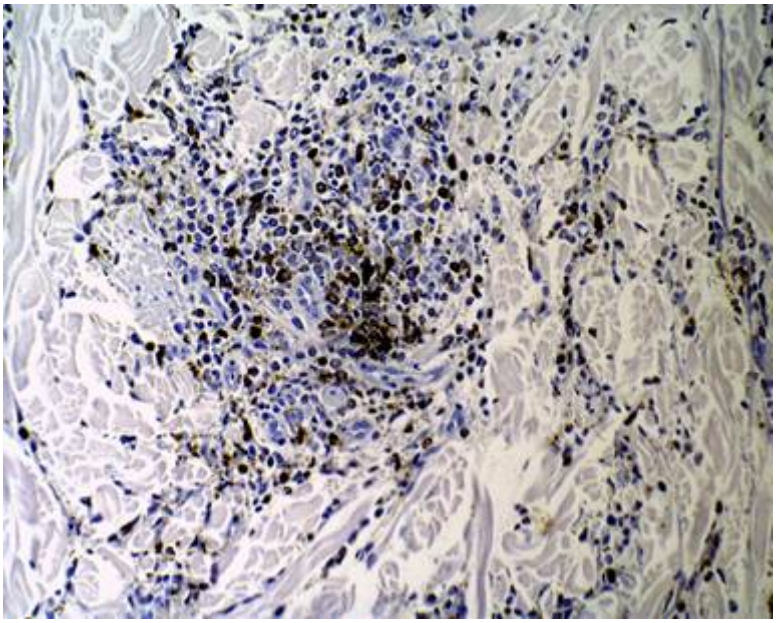

Fig 4: Meloperxidase(X100) staining shows positive blast cells.

Discussion:

A leukaemic leukaemia cutis (ALC) is a rare condition with a poor prognosis that has been described as skin involvement by leukaemic cells before their appearance in the peripheral blood or bone marrow [1]. In most cases of leukaemia cutis, systemic disease precedes the development of skin lesions. However, in as many as less than 10% of patients with leukaemia cutis [2], localized skin lesions occur prior to bone marrow infiltration and systemic symptoms (aleukemic leukaemia cutis or primary extramedullary leukaemia).

A literature review showed that aleukaemic leukaemia cutis confined to the skin is extremely rare and commonly misdiagnosed [3,4]. Its recognition is important because early diagnosis can lead to a better prognosis.

As it was mentioned earlier in our case he was under treatment with his GP for several weeks before he was seen by a dermatologist; he was generally well with even normal investigation. There is no clear evidence between the effects of skin infiltration on leukaemia prognosis. However ALC is usually associated with a very poor prognosis in leukaemic patients [3,5].

Wilkins and Janes reported a case of ALC in a 57 years old lady who has had a fatal outcome within 11 months of the diagnosis of skin lesions [3]. Recently Dekio et al described a case of ALC in a middle aged lady who had passed away within 4 months of the skin rashes [6]. More recently Vishalakshi et al presented a case of ALC in a young man who had died within 3 months of the diagnosis of the skin eruption [7].

Aleukaemic leukaemia cutis is easily to be misdiagnosis, and has always been a dermatological curiosity. This case has been reported to emphasize the important of the clinical diagnosis with the combined role of dermatologist, dermatopathologist and haematologist to achieve early correct diagnosis.

Acknowledgment

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