Leukaemia cutis

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Introduction

Leukaemia cutis (LC) is defined as cutaneous infiltration by neoplastic leucocytes (myeloid or lymphoid), resulting in clinically identifiable cutaneous lesions[1]. LC has been described in patients with acute myeloid leukaemia (AML), chronic myeloproliferative disease and myelodysplastic syndromes[2]. We report a case of LC with skin lesions without systemic features.

Case report

A 14 year old, previously healthy girl presented with multiple erythematous, hyperpigmented, plaques and nodules over the face for 4 weeks (Figure 1).

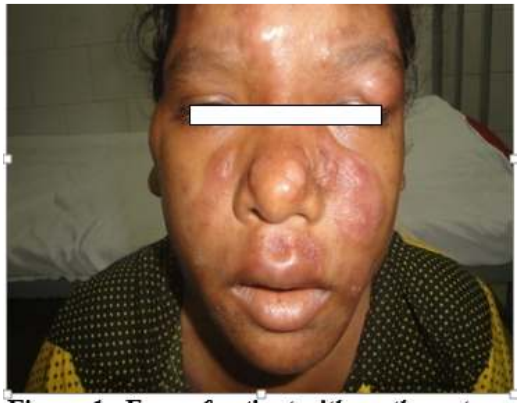

Figure 1: ***Face of patient with erythematous, hyperpigmented, plaques and nodules***

**Permission given by parents to publish photograph*

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She had been treated with topical steroids and antihistamines for above complaints. There was cervical lymphadenopathy but no pallor or hepatosplenomegaly. She also had low grade fever for 10 days. A complete blood count and peripheral blood smear did not show any significant findings. Based on physical examination cutaneous leukaemia was suspected. Biopsy was performed from a suspicious skin lesion on face and the histopathology report confirmed LC. The section showed the leukaemic cells forming nodular aggregate with a sheet like pattern separated from epidermis by a clear zone of upper dermis. The epidermis was focally thinned out but did not show any evidence of tumour infiltration. The tumour cells were monotonous with round hyperchromatic nucleus, inconspicuous nucleolus and scanty cytoplasm. Occasional mitotic figures were also seen (Figure 2).

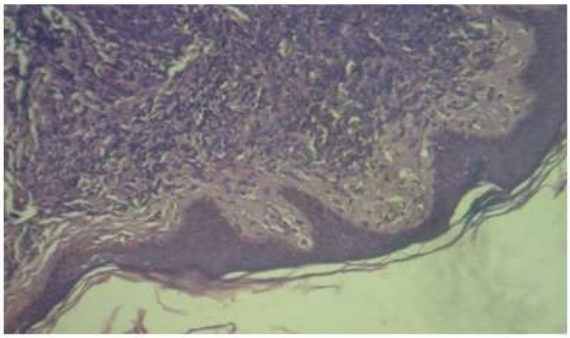

Figure 2: ***Haematoxylin and eosin stain (200X): microphotograph shows leukaemic cells in a nodular aggregate in lower dermis with clear zone of separation from epidermis***

A bone marrow aspirate confirmed precursor B-cell acute lymphoblastic leukaemia (ALL) with 25% blast cells. On immunophenotypic analysis, the neoplastic cells were positive for CD45, CD10, CD19, CD22, HLA-DR. Cytogenetic studies showed normal 46XX karyotype and ALL chromosomal translocations were negative. Chest x-ray did not show enlarged thymus. Cerebrospinal fluid (CSF) study showed no blast cells. Induction therapy was started immediately using protocol ALL ICBFM 2009. On the 20th day of induction chemotherapy, the soft tissue and skin lesions had almost completely regressed.

Discussion

LC occurs in 10-15% of patients with AML and less frequently in chronic myeloproliferative disease[3]. Of infants with congenital leukaemia 25-30% develop skin involvement[4]. Congenital acute leukaemia, most frequently associated with LC is AML[5]. Lesions of LC are usually single or multiple, violaceous, red-brown, or haemorrhagic papules, nodules and plaques of varying sizes. Legs are involved most commonly, followed by arms, back, chest, scalp, and face. Leukaemic infiltration tends to preferentially occur at sites of previous or concomitant inflammation[6].

Most cases of LC occur after a diagnosis of systemic leukaemia has been established. In one third cases, concomitant involvement of skin and systemic leukaemia has been observed, and in less than 10% of cases, skin infiltration can occur before bone marrow or peripheral blood involvement in the absence of systemic symptoms. This is known as "aleukaemic LC" or "primary extra-medullary leukaemia" and occurs predominantly with AML and very rarely in ALL. Aleukaemic leukaemia cutis lesions are usually widespread and papulonodular[7,8]. Up to 30% of children with congenital leukaemia have skin infiltration, and this is one cause of the "blue berry muffin" appearance[9]. Most paediatric patients with LC have high leukaemic tumour load and hepatosplenomegaly[10].

Diagnosis of LC is based on the morphologic pattern of skin infiltration, cytologic features and the immunophenotypic characteristic of tumour cells. Correlation with clinical data, bone marrow and peripheral blood findings is often helpful to confirm diagnosis[11]. LC is a local manifestation of an underlying systemic disease and treatment is aimed at eradicating systemic disease by using systemic chemotherapy as well as local therapy[12]. In general, development of LC has a poor prognosis. Patients with congenital leukaemia seem to be an exception because LC does not confer a worse prognosis in this clinical setting. Our patient represents a type of leukaemia cutis of acute lymphoblastic leukaemia.

References

1. Weedon D. Cutaneous infiltrates: lymphomatous and leukaemic. Skin Pathology. 2nd ed. New York: Churchill Livingstone; 2002:1118-20.

2. Brunning RD, Matutes E, Flandria F, et al. In: Jaffe ES, Harris NL, Stein H, et al, editors. World Health Organization Classification of Tumours: Pathology and Genetics of Tumours of Haematopoietic and Lymphoid Tissues. Lyon, France: IARC Press; 2001:104-5.

3. Agis H, Weltermann A, Fonatsch C, et al. A comparative study on demographic, hematological, and cytogenetic findings and prognosis in acute myeloid leukemia with and without leukemia cutis. *Annals of Hematology* 2002; **81**:90-5. http://dx.doi.org/10.1007/s00277-001-0412-9
PMid: 11907789

4. Zhang IH, Zane LT, Braun BS, et al. Congenital leukaemia cutis with subsequent development of leukaemia. *Journal of the American Academy of Dermatology* 2006; **54**(2 suppl.):S22-S27. http://dx.doi.org/10.1016/j.jaad.2005.04.038
PMid: 16427986

5. Resnik KS, Brod BB. Leukaemia cutis in congenital leukaemia: analysis and review of the world literature with report of an additional case. *Archives of Dermatology* 1993; **129**:1301-6. http://dx.doi.org/10.1001/archderm.1993.01680310071012

6. Cho-Vega JH, Medeiros LJ, Prieto VG, Vega F. Leukaemia cutis. *American Journal of Clinical Pathology* 2008; **129** (1):130-42. http://dx.doi.org/10.1309/WYACYWF6NGM3WBRT
PMid: 18089498

7. Appelbaum FR, Rowe JM, Radich J, et al. Acute myeloid leukaemia. *Hematology Am Soc Hematol Educ Program* 2001:62-86. http://dx.doi.org/10.1182/asheducation-2001.1.62

8. Greer JP, Baer MR, Kinney MC. Acute myeloid leukaemia in adults. In: Greer JP, Foerster J, Lukens JN, et al, eds. Wintrobe's Clinical Hematology. Philadelphia, PA: Lippincott Williams & Wilkins; 2003:2097-142.

9. Resnik KS, Brod BB. Leukaemia cutis in congenital leukaemia analysis and review of the world literature with report of an additional case. *Archives of Dermatology* 1993; **129**: 1301-6. http://dx.doi.org/10.1001/archderm.1993.01680310071012

10. Magro CM, Morrison CD, Heerema N, et al. T-cell prolymphocytic leukaemia: an aggressive T cell malignancy with frequent cutaneous tropism. *Journal of the American Academy of Dermatology* 2006; **55**:467-77. http://dx.doi.org/10.1016/j.jaad.2006.04.060 PMid: 16908353

11. Boccara O, Laloum-Grynberg E, Jeudy G, Aubriot-Lorton MH, Vabres P, de Prost Y, Pacquement H, Brousse N, Fraitag S, Bodemer C. Cutaneous B-cell lymphoblastic lymphoma in children: a rare diagnosis. *Journal of the American Academy of Dermatology* 2012; **66**:51- 7. http://dx.doi.org/10.1016/j.jaad.2010.10.040 PMid: 21745698

12. Kahwash SB, Qualman SJ. Cutaneous lymphoblastic lymphoma in children: report of six cases with precursor B-cell lineage. *Pediatr Dev Pathol* 2002; **5**:45-53. http://dx.doi.org/10.1007/s10024-001-0104-4