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RARE PRESENTATION OF ACUTE MYELOID LEUKAEMIA IN A SUDANESE CHILD

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Abstract:
Classically, children with acute leukaemia present with symptoms and signs related to infiltration of the bone marrow by leukaemic cells, such as fever, pallor and fatigue, cutaneous and mucosal bleeding and bone pain. Also anorexia and weight loss. This is a case of acute myeloid leukaemia (AML.) which presented with tongue bleeding and then part of the tongue separated completely from the rest of the tongue and it fell from his mouth, few days later it regenerated to a normal tongue. Few days after regeneration of the right side of the tongue, the same previous scenario occurred in the left side, and then regenerated to normal size. This indicates an unusual presentation of AML.

Introduction

Acute myeloid leukaemia (AML) is a clonal malignant disease of the bone marrow in which haematopoietic progenitor cells are arrested at an early stage of development due to acquired genetic alterations that lead to failure of differentiation and to over proliferation [(1)]. The etiology for most cases of AML is unclear [(2)], Classically, children with acute leukaemia present with symptoms and signs related to infiltration of the bone marrow by leukaemic cells [(3)], such as fever, pallor and fatigue, cutaneous and mucosal bleeding. Also bone pain, anorexia and weight loss. Massive lymphadenopathy and hepatosplenomegaly are less common, except in infants with AML [(4)]. Gingival hyperplasia is occasionally observed, while chloromas, solid tumours consisting of blasts, often occur in the cranium. Central nervous system is involved at diagnosis in approximately 15% of cases, while testicular infiltration is rare [(5, 6)]. Disseminated intravascular coagulation (DIC) can be observed in all AML subtypes but is much more common in acute promyelocytic leukaemia (APL) [(7)]. Acute myeloid leukaemia (AML) accounts for nearly 15% of all leukaemias in children [(8)], and (AML) is characterized by a block in differentiation and an unregulated proliferation of myeloid progenitor cells. While the cause of AML in children is unknown, risk factors that have been identified include exposure to toxins such as ethanol and pesticides [(9)]. The carcinogenic effects of radiation on the young have been reported after intrauterine exposures and after exposures during childhood [(10)]. This article describes a rare presentation of acute myeloid leukaemia in a Sudanese patient.

Case Report

A ten year old Sudanese boy was referred to paediatrics unit with change in colour of his tongue for three days. The condition started when his mother noticed that the right half of her child tongue changed to whitish colour, after one day it became bluish in colour then on the third day changed to black. He started to bleed from his tongue, few days later the affected half of the tongue became rounded mass like a ball, it separated completely from the rest of the tongue and it fell from his mouth (Plate 1). Few days later it regenerated to a normal tongue (Plate 2). Examination revealed ill emaciated, pale, febrile and tachypneic

child with small size submandibular enlarged lymph nodes. Chest examination indicated evidence of chest infection. Spleen and liver were just palpable. Local examination of the tongue indicated rounded mass 5 cm in diameter black in colour stained with blood, soft, freely mobile with a pedicle originated from the tongue. Few days after regeneration of the right side of the tongue, the same previous scenario occurred in the left side, and then regenerated to normal size. Complete blood count revealed: TWBS 100 000 cells/ mm^3, Hb. 6g/dl, Platelets Count 50 000 cells/ mm^3 , ESR 151 mm/r. Bone marrow revealed hypercellularity, erythropoiesis very much reduced with deserythromaturation, granulopoiesis replaced with blast cells, which consisted 80% of maturated cells, megakaryocytes reduced, features consistent with acute myeloid leukaemia. Cytology of the ulcerative mass of the tongue showed many epithelial cells with scattered inflammatory cells (neutrophils). Chest X- ray showed wide mediastinum (Plate 3). He was diagnosed as acute myeloid leukaemia, transfused twice, started initial treatment and referred for chemotherapy. But unfortunately the patient died so there was no time for performing cytochemistry which was not available in the patient area to determine the sub type of acute leukaemia.

Discussion

AML requires a minimum of 30% leukemic cells in the bone marrow for the diagnosis of AML.[(1)] Infections in acute leukaemia are often accompanied by fatal septicemia and pneumonia [(11)] or other traditionally sign and symptoms just like fever, pallor and fatigue, cutaneous and mucosal bleeding, bone pain, anorexia and weight loss. In this case the lesion observed is a chloroma of the tongue made up of leukaemic infiltration. The frequency of the head and neck region to be affected by leukemic infiltration varies, according to the literature, between 12% and 48%.In this region, the most frequently affected sites are the soft palate, the rhino-pharynx, orbit, salivary glands, scalp and face. Uncommon sites are: the jaw, facial nerve, lips, nasal cavity, maxilla and temporal bone [(12)] .There was a rare case reported but in the maxillo- ethmoid area [(12)]. Also there was another one intra-oral and it was not in the tongue. It was in the alveolar socket of extracted tooth as ulcerated mass [(13)]. Infections and anemia are the major causes of death in leukemic patients [(14)]. Untreated, acute leukemia has an aggressive course, with death occurring within six months or less [(15)] According to our knowledge AML does not presented with tongue affected and then the tongue regenerated. So this case indicated an unusual presentation of acute myeloid leukaemia.

References

1. Krstovski N, Janic D, Dokmanovic L. Childhood acute myeloid leukaemia. Journal of Pediatric Sciences 2010; 2: e23

2. Deschler B, Lübbert M. Acute myeloid leukemia: epidemiology and etiology. Cancer. 2006; 107: 2099-107.

3. T Sharma T; Grewal J; Gupta S; and Murray P I. Ophthalmic manifestations of acute leukaemias: the ophthalmologist's role. *Eye* 2004; 18: 663–672.

4. Choi, S I and Simone, J V. Acute nonlymphocytic leukemia in 171 children. Paediatrics Blood & Cancer 2006; (2) 2: 119–146.

5. Rubnitz JE, Gibson B, Smith FO. Acute myeloid leukemia. *Pediatr Clin North Am.*2008; 55: 21-51.

6. Hutspardol, S H. Common Problems in Pediatric Oncology.
Available from: http://elearning.medicine.swu.ac.th/pedia/wp-content/uploads/2009/06/oncology17may2010.pdf

7. Kingsley EC, Durie BGM, Garewal HS: Acute promyelocytic leukemia. *West J Med* 1987; 146: 322-

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327.

8. Stein-Wexler R, Wootton-Gorges SL, West DC. Orbital granulocytic sarcoma: an unusual presentation of acute myeloid leukaemia. *Pediatr Radiol* 2003; 33:136-9.

9. Peter B. Langmuir, Richard Aplenc, Beverly J. Lange. Acute myeloid leukaemia in children. *Clinical Haematology* 2001; 14: 77-93.

10. Miller RW. Special susceptibility of the child to certain radiation-induced cancers. Environ Health Perspect. 1995; 103: 41-4.

11. O.M. KoriechK, F.Z. Al-Dash. Skin and bone necrosis following ecthyma gangrenosum in acute leukaemia: report of three cases. *Clinical and Experimental Dermatology* 2006;13: 78 – 81.

12. Ferri E, Minotto E C, Ianniello F, Cavaleri S, Armato E, and Capuzzo P. Maxillo-ethmoidal chloroma in acute myeloid leukaemia: Case report. *Acta Otorhinolaryngol Ital.* 2005; 25(3): 195–199.

13. Papamanthos M K, Kolokotronis A E, Skulakis H E, Angela-Monika A. Fericean A A, Zorba M T and Matiakis A T. Acute Myeloid Leukaemia Diagnosed by Intra-Oral Myeloid Sarcoma. A Case Report. *Head Neck Pathol.* 2010; 4(2): 132–135.

14. Greenburg M and Glick M. Burket's oral medicine diagnosis and treatment. 10th ed. BC Decker Inc.; 2003. p.443-8.

15. McKenna SJ. Leukemia. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod.* 2000; 89: 137-9.

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