Case Report

Maxilla Unilateral Swelling as the First Diagnostic Symptom of Acute Lymphoblastic Leukemia Relapse: A Case Report

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Abstract:

Acute Lymphoblastic Leukemia (ALL) is the most prevalent hematological malignant tumor during childhood. Unilateral infiltration into the gums is less prevalent and more often observed in the AML type.

A 12-year-old girl with symptoms of pain and swelling in the buccal vestibule and also at the posterior part of the right palate of the maxilla was referred to a private dental office. The patient had been inflicted by ALL and had undergone complete chemotherapy. A week prior to her admittance into the hospital, the workup of the patient's blood revealed her recovery. The clinical and radiographic evidence did not show any dental problems. The histological examinations on the patient's jaw revealed the correct diagnosis of ALL and the patient underwent chemotherapy for the second time.

This case has been reported to point out that intraoral unilateral swelling of the upper jaw may be propounded as the primary diagnostic symptom of ALL.

Key Words: Precursor Cell Lymphoblastic Leukemia-Lymphoma; Hematologic Neoplasms; Child

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INTRODUCTION

Acute Lymphoblastic Leukemia (ALL) is the most common hematological malignant tumor in childhood and it is responsible for a great number of different oral problems. The leukemic cells are the result of colonial proliferation of abnormal stem cells or progenitor cells which replace the normal hematological cells in the bone marrow, thus causing its failure [1]. These replacement cells, which lack the ability of differentiation and maturation, enter the blood circulatory system and infiltrate into the other organs [2]. The oral cavity, especially the gingival tissue, is one of the sites that may be involved by leukemic cell infiltration or inflammatory reactive hyperplasia, resulting in gingival enlargement [1]. The primary symptoms of ALL generally include anemia, bleeding tendency, pain, splenomegaly and lymphadenopathy. In rare cases, the primary symptoms may be the oral and intra oral findings, such as toothache, which are not pathognomonic [3]. The ALL oral problems, which are classified into three groups, develop during the diagnosis period or treatment. The first group of oral manifestations is caused by the infiltration of leukemic cells into the oral structures. The second occurs due to the

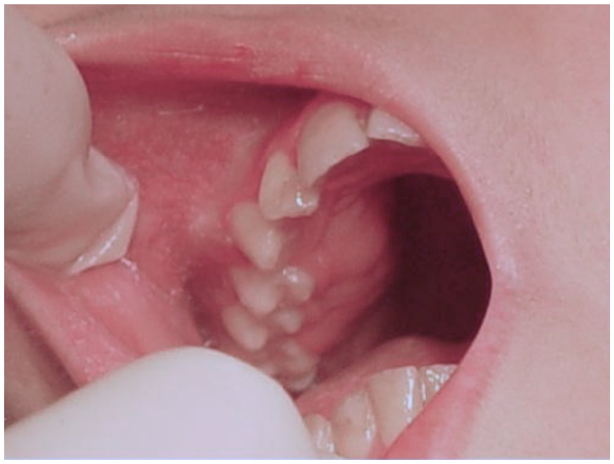

Fig 1. Swelling of right vestibule and palatal of maxilla.

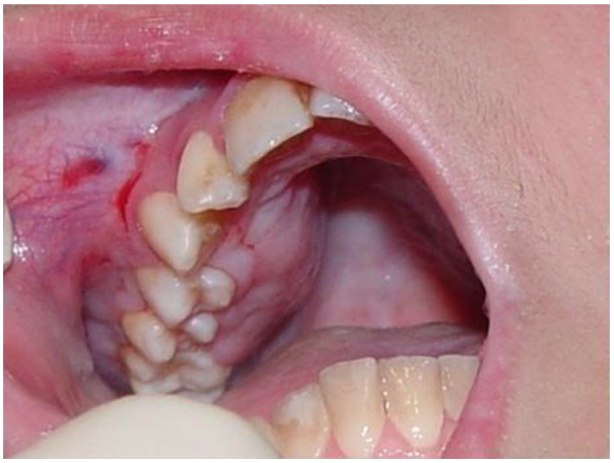

Fig 2. The Biopsy of buccal vestibule.

characteristic of myelophthisic sickness which includes the symptoms of anemia, bleeding tendency and predisposition to infection. The third is caused by anti-leukemic treatment such as cytotoxic effects [4].

CASE REPORT

A 12-year-old girl was referred to a private dental office due to pain and swelling of the right vestibule and palate of maxilla (Fig 1). She received right kidney transplantation five years ago and has been using immunosuppressive drugs. The patient was afflicted by ALL six months ago and underwent chemotherapy including rituximab, cyclophosphamide, vincristine and prednisolone. The chemotherapy lasted six months. The result of complete blood work-up did not show any sign of blood cell alteration.

A biopsy was performed (Fig 2) on the mucosal tissue of the buccal vestibule. The tissue structure was beige to white, completely soft like fish meat, with differential diagnosis including leukemic infiltration and secondary tumor.

The pathological testing procedure of immunohistochemistry and C-Kit (CD 117) confirmed the diagnosis of ALL and the patient underwent chemotherapy again with stronger drugs. The patient died due to severe side effects of chemotherapy after 3 months.

DISCUSSION

This study is one of the rare case reports of ALL relapse during consolidation therapy manifesting as unilateral pain, swelling and teeth mobility because of leukemic cell infiltration which were the only intra-oral symptoms of ALL. According to Boggs et al study [5], infiltration of the leukemic cells is seen in ALL. In White's study [6], infiltrations of leukemic cells were found in the gingival tissue. The existence of leukemic cutis in ALL within the oral mucous is also due to the infiltration of leukemic cells [7]. Sometimes this infiltration in the structure of PDL results in an intensive periodontal destruction, periodontal disease and loosening of the teeth. This periodontal destruction accompanied by the intensive destruction of alveolar bone is possible as the initial manifestation of ALL [6]. Moreover, pericoronitis is one of the primary symptoms of ALL due to infiltration of leukemic cells in the gingival tissue [8]. Karimi and Eshghi [9] have also reported a case containing massive swelling of the maxilla, mandible and palate in addition to severe hypertrophy of the gingiva caused by infiltration of leukemic cells inside the bone and the gingiva accompanied by irregular breathing and swallowing problems due to ALL. Occasionally, the initial symptoms of ALL manifest as severe hypertrophy of the gingiva, jaws and also the bone [10].

Hiraki et al [3] reported a case of ALL, in which the primary symptoms included generalized tooth pain and Numb Chin Syndrome with intraoral symptoms including teeth loosening, gingival abscess and partial bone destruction in some areas.

The biopsy showed leukemic cell infiltration into the gingiva. Study on 56 ALL adult patients showed 23% extra-medullary infiltration in non-lymphopoietic tissues such as the testis, CNS, skin, gingiva and pleural cavities [11]. The secondary malignant neoplasm (SMN) is common in children who have been treated for ALL. The prevalent neoplasms as SMN include BCC, adenocarcinoma, SCC, meningioma, malignant histiocytoma, glioblastoma, osteosarcoma, Ewing sarcoma, fibroblastic sarcoma, lymphoma and thyroid carcinoma. The risk for SMN 15 years after treatment of ALL is estimated as 1.26% [12]. The incidence of T-cell ALL following the treatment of AML has been reported after 3 years of remission [13]. Acute leukemia occurrence as secondary malignancy is extremely rare and secondary AML is more common, which in certain cases is propounded as the secondary primary tumor [14]. The occurrence of secondary ALL (SALL) after the treatment of ALL is highly rare; although it is possible the SALL rate after the primary treatment of ALL is underestimated with misdiagnosis of the relapse [15]. The prognosis of ALL relapse is very poor, especially in the patients of early bone marrow relapse. The highest rate of relapse is also observed in patients who have completed the second phase of remission [16].

CONCLUSION

Although the systemic signs such as fatigue, fever and bleeding tendency are common as the primary symptoms of ALL, special attention should be paid to the oral signs including pain, loosening of teeth and gingival abscess without any specific reasons such as decay or periodontal disease. These oral signs may be the initial symptoms of malignant disease such as ALL. In addition, although gingival biopsy by itself not sufficient, it may help in diagnosis.

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REFERENCES

1-Abdullah BH, Yahya HI, Kummoona RK, Hilmi FA, Mirza KB. Gingival fine needle aspiration cytology in acute leukemia. J Oral Pathol Med 2002 Jan;31(1):55-8.

2-Wadhwa N, Vohra R, Shrey D, Iyer VK, Garg S. Unilateral hypopyon in a child as a first and sole presentation in relapsing acute lymphoblastic leukemia. Indian J Ophthalmol 2007 May-Jun;55(3): 223-4.

3-Hiraki A, Nakamura S, Abe K, Takenoshita Y, Shinohara M, Shirasuna K. Numb chin syndrome as the initial symptom of acute lymphoblastic leukemia, report of three cases. Oral Surg Oral Pathol Oral Radiol Endod 1997 May; 83(5):555-61.

4-Vinckier F, Declerck D. Oral manifestations in leukemic children and their diagnostic value. Acta Stomatol Belg 1989 Oct;86(3):219-26.

5-Boggs DR, Wintrobe MM, Cartwright GE. The acute leukemias analysis of 322 cases and review of the literature. Medicine 1962;41:163-225.

6-White GE. Oral manifestations of leukemia in children. Oral Surg Oral Med Oral Pathol 1970 Mar;29(3):420-7.

7-Michaud M, Baehner RL, Bixler D, Kafrawy AH. Oral manifestations of acute leukemia in children. J Am Dent Assoc. 1977 Dec;95(6):1145-50.

8-Aronovich S, Connolly TW. Pericoronitis as an initial manifestation of acute lymphoblastic leukemia: a case report. J Oral maxilofac Surg 2008 Apr;66(4):804-8.

9-Karimi M, Eshghi P. Unusual lymphoblastic leukemia/lymphoma in Eastern Iran Indian J Pediatr 2006 Jul;73(7):619-22.

10-Premalata CS, Madhumathi DS, Lakshmidevi V, Pradeep R, Appaji L, Mukherjee G. Gum hyper-

trophy–an unusual presenting feature in a case of precursor T-cell lymphoblastic leukemia. Turk J Hematol 2008;25:201-4.

11-Jensen IM, Jensen PD, Ellegard J, Bastrup-Madsen P, Hokland P. Extramedulary manifestation among adult patient with acute lymphoblstic leukemia (ALL). Ugeskr Laeger 1991 Apr 15; 153(16):1125-9.

12-Borgmann A, Zinn C, Hartmann R, Herold R, Kaatsch P, Escherich G et al. Secondary malignant neoplasms after intensive treatment of relapsed acute lymphoblastic leukaemia in childhood. Eur J Cancer 2008 Jan;44(2):257-68. Epub 2007 Nov 5.

13-Tsuboi K, Komatsu H, Miwa H, Iida S, Banno S, Wakita A et al. T-cell acute lymphoblastic leukemia as a secondary leukemia after a 3-year remission of acute myelocytic leukemia. Int J Hematol 2003 Jun;77(5):518-21.

14-Hijiya N, Ness KK, Ribeiro RC, Hudson MM. Acute leukemia as a secondary malignancy in children and adolescents: current findings and issues. Cancer 2009 Jan 1;115(1):23-35.

15-Zuna J, Cave H, Eckert C, Szczepanski T, Meyer C, Mejstrikova E, et al. Childhood secondary ALL after ALL treatment. Leukemia. 2007 Jul;21(7):1431-5.

16-Hamed TM, Gaynon P. Relapsed acute lymphoblastic leukemia: current status and future opportunities. Curr Oncol Rep 2008 Nov;10(6):453-8.