Case Report

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T-cell Lymphoblastic Lymphoma in the Maxilla and Mandible of a Child: A Rare Case Report

Dentistry Section

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ABSTRACT

T-cell lymphoblastic lymphomas (T-LBL), defined as neoplasms of immature T cells, are the most common paediatric T-cell lymphoma. These account for approximately 90% of all lymphoblastic lymphomas. The primary manifestation of T-LBL rarely occurs in the oral cavity. In this case report, we describe a case of primary T-LBL affecting the maxilla and mandible of a 10-year-old male patient. This is the first case of T-LBL reported in this region.

We emphasize that early diagnosis of aggressive lesions in the maxilla or mandible is one of the responsibilities of oral physicians, who can help patients to overcome the many challenges of malignant diseases.

Keywords: Aggressive lesions, Neoplasm, Oral cavity, Paediatric T-cell lymphoma

CASE REPORT

A 10-year-old male child visited the Department of Oral Medicine, School of Dentistry, Mashhad, Iran, on July 2012 with the chief complaint of swelling over both sides of the face which had appeared since two months ago. Swellings were progressively increasing up to the present size. Medical and family history was non-contributory. He reported no weight loss, wheezing, nausea, vomiting and abdominal pain. Previous diagnosis of abscess by a general dentist at the local dental clinic was made two months ago for which he received amoxicilin and metronidazole for seven days, but the swelling did not subside.

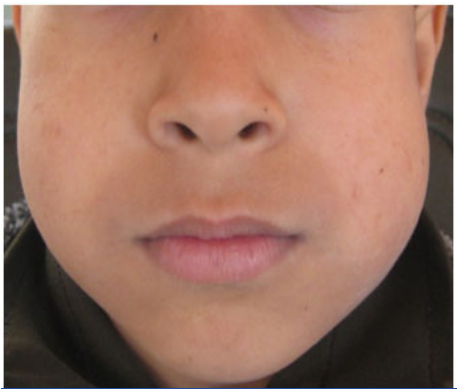
On extra oral examination, bilateral diffuse swellings were observed in both sides of the face, left side being bigger, 5X4X4 cm, extending superiorly from the zygomatic arch and inferiorly to the border of the mandible [Table/Fig-1]. Skin over the swellings on both sides of the face had a smooth surface and was normal in colour. The consistency of involved areas was firm, without any pain, tenderness or local rise in temperature. Regional lymph nodes were not palpable. Intraorally, swellings were observed in the mandibular and maxillary buccal vestibules bilaterally as well as in the right palatal region. In the mandibular buccal vestibules, the extension was from the lateral incisor to the retromolar pad in the left and from the second deciduous molar to the retromolar pad in the right. Palatal swelling from right deciduous canine to tuberosity and bilateral buccal vestibular obliteration was seen in the maxilla [Table/Fig-2].

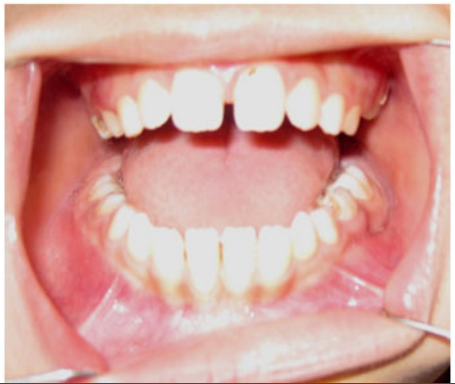
The consistency of swellings on palpation was firm in the buccal vestibules and bony hard in the palate. Mobility was not detected in the associated teeth, the occlusion was unaltered [Table/Fig-3]. Overlying surface had normal mucosa and tenderness was not observed. Vitality test suggested that all the teeth were vital and there were no symptoms of numbness, tenderness and pain.

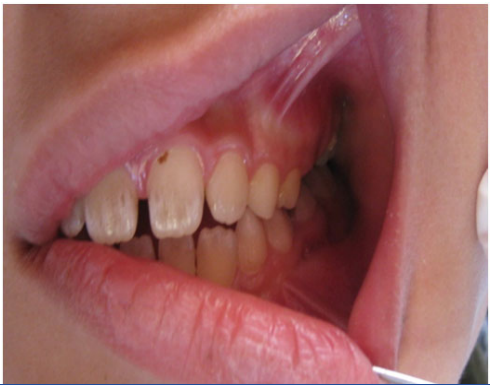
Based on the history and clinical examination (increased volume of the face, presence of an intra-oral mass, involvement of all posterior quadrants of maxilla and mandible), a provisional diagnosis of burkitt's lymphoma was given. Notably, Burkitt's lymphoma is a rare and rapidly progressive tumour, especially in patients at the first decade of life. The following differential diagnoses were considered: other non-Hodgkin's lymphomas, leukemia and undifferentiated carcinomas and sarcomas.

The patient was subjected to radiographic examination including panoramic and occlusal radiographs [Table/Fig-4]. The panoramic radiograph showed a unilocular ill-defined radiolucency in the unerupted second premolar region of the left side of the mandible.

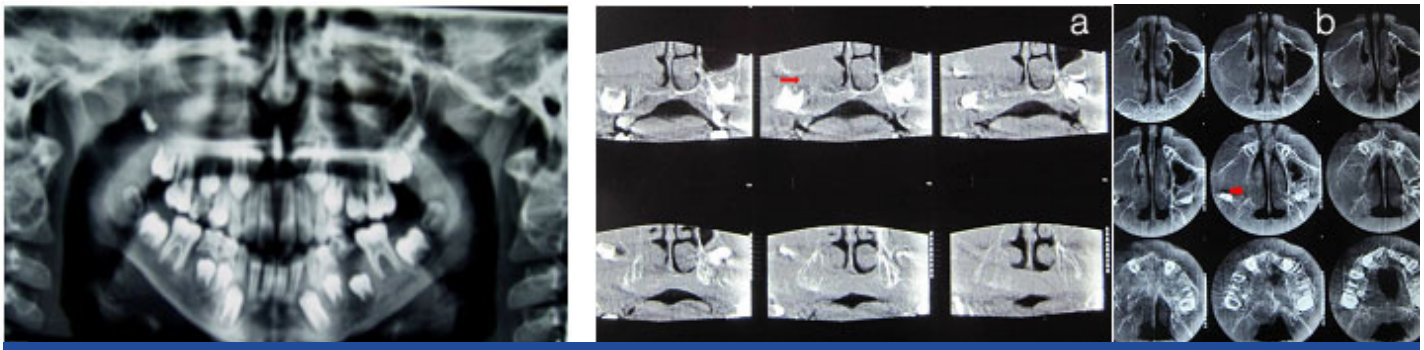
To assess the exact boundaries and invasion into the adjacent areas, cone beam computed tomography (CBCT) was performed. CBCT revealed a large soft tissue density lesion in the right maxillary sinus, causing erosion of medial wall of the maxillary sinus with extending medially into the ethmoid sinus. The maxillary teeth were not involved. It was also evident that the inferior rectus muscle and the buccal soft tissue were invaded [Table/Fig-5a&b]. Furthermore, routine blood investigations were carried out. Haemoglobin level



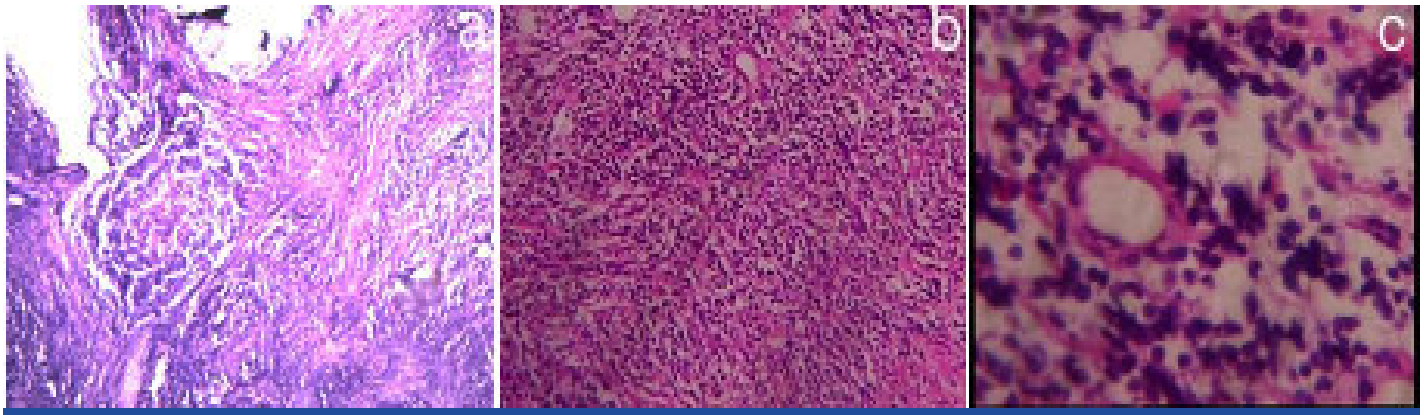




[Table/Fig-1]: An extraoral photograph showing bilateral diffuse swellings in both sides of the face and the facial asymmetry **[Table/Fig-2]:** Intraoral appearance of the lesion, with swelling of the mandibular and maxillary buccal vestibules bilaterally **[Table/Fig-3]:** Photograph showing patient occlusion



[Table/Fig-4]: Panoramic radiograph showing a unilocular ill-defined radiolucency in the unerupted second premolar region **[Table/Fig-5a,b]:** Computed tomography examination showing a large soft tissue density lesion in the right maxillary sinus, causing erosion of medial wall of the maxillary sinus with extending medially into the ethmoid sinus (a: coronal view; b: axial view)



[Table/Fig-6a-c]: Photomicrograph showing diffuse undifferentiated proliferation of small cells with a little amount of cytoplasm and hyperchrome nuclei (H & E staining, a: 40X, b: 100X, c: 400X)

was 11.1 g/dl (normal 13-17 g/dl); white blood cell count and platelet count were within the normal range.

An incisional biopsy of intraoral mass was performed under local anaesthesia. Histopathological examination of haematoxylin and eosin stained section showed a diffuse undifferentiated proliferation of small cells with a little amount of cytoplasm and hyper-chrome nuclei in desmoplastic connective tissue stroma. It was finally diagnosed as malignant small round cell tumour [Table/Fig-6a-c].

On immunohistochemical analysis, tumour cells were positive for LCA, TDT, CD-3, CD-10 and CD-99 and negative for CK and vimentin. Thus the final diagnosis of T cell lymphoblastic lymphoma was made. The patient was referred to the Department of Paediatric Oncology. Bone marrow aspiration, cerebrospinal fluid (CSF) cytology and chest X-Ray were carried out. No lymphoblastic cell was detected on the cerebrospinal fluid (CSF) cytology. Bone marrow biopsy revealed normal cellular marrow as well as normal chest X-Ray.

The patient received six cycles of chemotherapy at Doctor Sheikh Children Hospital in Mashhad, Khorasan, Iran. During follow-up the patient has been in complete remission for approximately two years from the initial diagnosis and treatment.

DISCUSSION

Misdiagnosis of a maxillary and mandibular swelling as abscesses, even when there are no signs and symptoms of infection, is very alarming. These delays in prompt diagnosis of aggressive lesions or malignant neoplasia may worsen the prognosis of these patients. Lymphoblastic lymphoma (LBL) accounts for about one third of childhood non-Hodgkin's lymphomas (NHL) [1]. Approximately 90% of all LBLs are in the form of T-cell lymphoblastic lymphoma (T-LBL), developing from immature T cell precursors, which is the most common paediatric T-cell lymphoma [2]. The cytological and clinical features of T-LBL are similar to those of acute lymphoblastic leukemia (ALL) – both represent the same spectrum of disease and are arbitrarily separated by the degree of bone marrow involvement. Patients with more than 25% lymphoblasts in their marrow are considered as having T-ALL, whereas those with a lower degree of marrow involvement are diagnosed as having T-LBL [1,3].

The primary manifestation of both T-cell lymphoblastic lymphoma (T-LBL) and B-cell lymphoblastic lymphoma (B-LBL) rarely occur in the oral cavity. Cox et al., reported the first case of B-LBL in the oral cavity of 46-year-old white woman. The patient presented with a mass in the right mandible which was initially treated as an inflammation of odontogenic origin, delaying the correct diagnosis [4]. Cavalcante et al., also reported a case of B-LBL in the maxilla of a 6-year-old child. The patient presented with swelling of the right maxilla which was initially treated based on the hypothesis of dentoalveolar abscess by dentist [5]. To the best of our knowledge, the present case is the first case of T-LBL manifested in the oral cavity while, B-LBL has been previously reported [4,5].

Clinically, T-LBL is more common in males and usually presents with a large mediastinal tumour and often with symptoms of respiratory distress. In addition, skin, bone, gonadal or central nervous system (CNS) may sometimes be involved. However, in the current case, the patient had no mediastinal involvement [3,6].

Clinical staging evaluations of T-LBL include computed tomography (CT) of the chest and abdominopelvis, bone marrow evaluation and lumbar puncture. There is complication in the diagnosis of small round cell tumours of bone marrow and other neoplasms including Ewing's sarcoma, small cell osteosarcoma, rhabdomyosarcoma, lymphoma, neuroblastoma, and neuroendocrine carcinoma due to the overlapping histology and similar clinical and radiographic characteristics. Lymphomas can be characterized by haematoxylin-eosin staining of tumour sections; however, immunophenotyping is often employed as a main diagnostic method.

The tumour cells of T-LBL are typically positive for terminal deoxynucleotide transferase (TdT), a unique DNA polymerase which is present in T-cell and B-cell precursors and is used to differentiate between LBL and Burkitt's lymphoma. Furthermore, these cells exhibit the expression of CD1a, CD2, CD3, CD4, CD5, CD7 and CD8.

Among these markers, CD7 and cytoplasmatic CD3 (cCD3) are positive in most cases. The only reliable lineage-specific is the surface CD3 (sCD3). Co-expression of CD4 and CD8 is frequently observed and CD10 is variably expressed. In addition to TdT expression, CD99, CD34 and CD1a are the most specific markers. The myeloid antigens CD13 and CD33 are positive in 19–31% of cases and their presence does not rule out the T-LBL/ALL diagnosis [7]. Consistent with previous observations, the tumour cells of the present case were positive for TDT, CD-3, CD-10 and CD-99 markers.

T-LBL in children is commonly classified based on the stage system of Murphy [1,3,8]. Currently, standard therapy for LBL involves CNS prophylaxis and intensive multidrug leukemia chemotherapy protocols, which are based on regimens with 7-10 drugs including methotrexate, prednisone, cyclophosphamide, vincristine, cytarabine, Elspar, etoposide, thioguanina, nitrosoureas and anthracyclines, on a type C basis [7,9]. In the present case, a 4-drugs regimen containing a combination of vincristine, Elspar, danomycin and methotrexate was used for chemotherapy.

The application of recent intensive therapeutic strategies for treatment of immature T-cell neoplasms has significantly improved the outcome in children with T-LBL. Consequently, long-term overall survivals in the range of 80–95% can be achieved with current protocols [10].

CONCLUSION

Early diagnosis of lesions in the maxilla or mandible is one of the responsibilities of dental surgeons, who could along with other health professionals; help patients overcome the many challenges of malignant diseases.

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