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CASE REPORT

Burkitt's Lymphoma Presenting as Maxillary Swelling: Case Report

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Abstract
In the head and neck region, lymphomas are the most frequently seen malignant lesions after squamous cell carcinoma. Burkitt's lymphoma is a malignant, highly aggressive non-Hodgkin's lymphoma. It is a B-cell type that generally presents in the oral region as a rapidly growing mass, which is usually misdiagnosed as odontogenic infection. In this case report, we present the diagnosis of Burkitt's lymphoma in a 3-year-old boy patient who was admitted with odontogenic abscess and facial swelling complaint.

Key words: Burkitt's lymphoma, odontogenic abscess, chemotherapy.

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Introduction

Burkitt's lymphoma (BL) is a rare monoclonal proliferation of B-lymphocytes and is classified as a poorly differentiated lymphocytic lymphoma (Ziegler, 1977). The tumor was first described in 1958 as a malignancy that occurs among African children (Burkitt, 1958). This tumor, which predominantly affects children, seems to be the fastest growing tumor in humans with exuberant proliferation (Ziegler, 1977). BL in three main variants: endemic, sporadic and immunodeficiency-associated types. The endemic form frequently involves the jaw bones and the abdomen of equatorial African children, whereas the sporadic form usually presents as an abdominal mass in adult patients from North America and Europe. The immunodeficiency-associated variant has a similar clinical presentation as that of sporadic subtype, with rare orofacial involvement (Bieging et al., 2010).

The clinical presentation of BL in the maxillofacial area is variable. It is characterized by

the rapid progression of symptoms with frequent multifocal extranodular involvement, including central nervous system involvement.
Within the oral cavity, this tumor can progress rapidly and appears as facial swelling or an exophytic mass involving the jaws (Kikuchi et al., 2012). In this case, we report the diagnosis and treatment of Burkitt's lymphoma presenting as facial swelling revealed by intraoral and extraoral swelling.

Case Report

A three-year-old boy presented with facial swelling and pain. Upon clinical examination, a slightly tender, sessile, firm, non-fluctuant mass 2 cm in diameter was found in the buccal sulcus in the right maxilla. The upper right deciduous first and second molars were not carious but were slightly mobile. No periodontal pockets were found. Upon extraoral examination, swelling similar to maxillary abscess was seen (Fig. 1-A). No organomegalia and lymphadenopathy were found. Moreover, no associated systemic symptoms were observed. No tumor masses or lymph nodes were clinically apparent in the head and neck region.

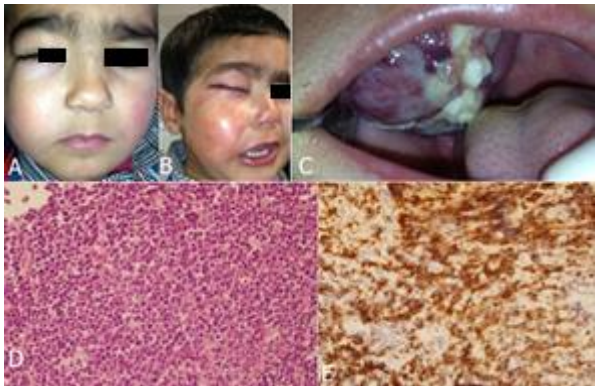

Figure 1. *A*: An extraoral appearance on day of admission. *B:* An extraoral appearance at one week following the biopsy. *C:* An intraoral appearance at one week following the biopsy. Oral examination showed gingival erythema, ulceration, suppuration and swelling extending buccally. *D:* Upon histologic examination, monotonous cells with round to oval nuclei, multiple nucleoli, and dark blue vacuolated cytoplasm with numerous mitotic figures were identified. Tingible body macrophages made a starry-sky pattern (X400, H&E). *E:* Immunohistochemically cells were positive for CD20 and negative for CD34 and Tdt. Ki67 was positive in almost 100% of the cells (X400, Ki67).

The consent form was obtained from the patient's parents and, incisional biopsy of the lesion was made under local anesthesia. Seven days after his first presentation, the patient was taken to the hospital again because of the rapid deterioration of his condition (Figs. 1 B-C). Pathological and immunohistochemical findings were consistent with BL (Figs. 1 D-E).

To evaluate the stage of BL, computerized tomoghraphies (CT) were taken. CT of the thorax was normal, but CT of the abdomen revealed ileoilial invagination, which is a sign of abdominal involvement. The right maxillary sinus was completely invaded. As the anterior, lateral, and superior walls were destroyed, the lesion extended to the subcutaneous region, right nasal cavity, and right orbital cavity. The right orbita was displaced superiorly and anteriorly by the compression of the lesion (Fig. 2 A, B). The tumor did not invade the brain, and the rest of the paranasal sinuses were normal.

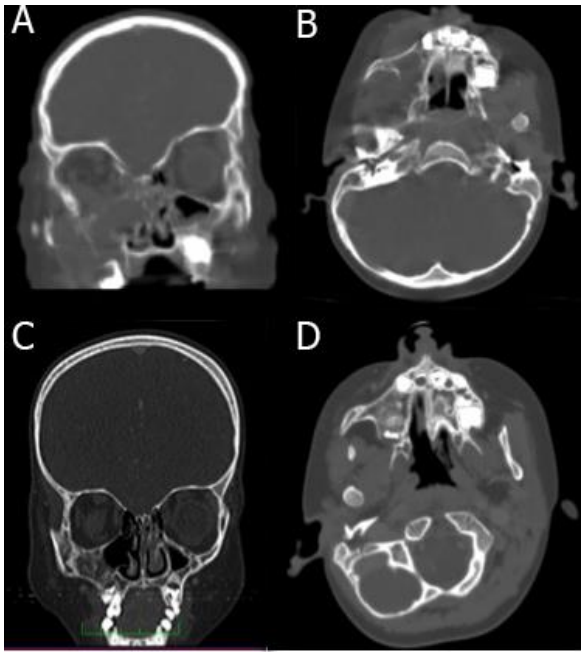

Figure 2: *A:* Coronal section shows maxillary sinus, nasal and orbital involvement. *B:* Axial section. *C and D*: Computed tomography axial and coronal image showing disappearance of previously observed lesions six months after chemotherapy.

Chemotherapy was started immediately and the remission of the lesions was corrected by CT images (Fig. 2 C, D). A remarkable resolution of

the intraoral disease, represented by stabilized teeth and resolved alveolar mucosa swelling, was observed within three weeks after chemotherapy (Fig. 3-A, B) and a 12-month follow-up (Fig. 3-C, D). The patient is being followed up closely.

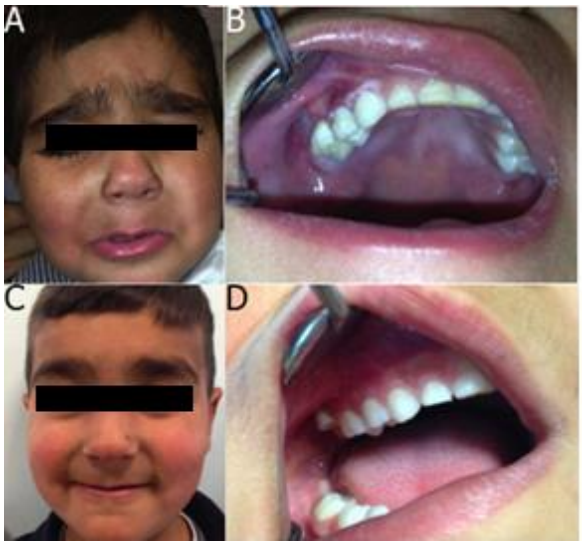

Figure 3. *A and B:* Post treatment photos showing resolution of facial and intraoral swelling after 3 weeks of chemotheraphy. *C and D*: After 12-month follow-up

Discussion

Lymphomas are malignant neoplasms of the cells of lymphoid tissues and it has two subgroups: Hodgkin's lymphoma and Non-Hodgkin lenfoma (NHL) (Molyneux et al., 2012). In the middle of the 20th century, Dr. Denis Burkitt described a malignancy that he had often seen in young African children. This lesion predominantly affected the jaws and the abdomen (Burkitt, 1958). Later, Michael Anthony Epstein, Yvonne Barr, and Bert Achong showed a herpes virus in a biopsy specimen taken from BL. This virus is known as the Epstein–Barr Virus (EBV), which is considered a potential etiologic agent (Epstein et al., 1964).

BL is a high-grade B-cell type NHL. It has three subtypes: endemic, sporadic, and immunodeficiency-related type. The endemic form of the disease is mostly seen among African children, who are nine years old on average, and is strongly linked to the EBV. It mainly affects the jaw (60%–80%) and other facial bones; it is less commonly seen in the abdomen and in the bone marrow (Banthia et al., 2003). Sporadic cases have emerged outside Africa. The mean age of presentation of the sporadic form is higher than that of the endemic form, i.e., 11–15 years of age (Kikuchi et al., 2012). Despite having the same histological features as the endemic form, the sporadic form is rarely associated with EBV infection, usually involves the abdomen (60% – 80%), and is rarely seen in the head and neck region (Banthia et al., 2003, Mbulaiteye et al., 2009). The immunodeficiency-related type is mainly seen in AIDS patients, and less than 40% of cases are associated with the EBV (Molyneux et al., 2012). In our case, the EBV titers were negative, which is a sign of the sporadic form of the disease. However, the jaw involvement and relatively younger age of the patient (three years old) are far from the common characteristics of the sporadic form.

BL is the fastest growing human tumor; the cells of the BL cycle have a 24 h–48 h period. Histopathologic examination of a biopsy specimen reveals monomorphic medium-sized cells with a high doubling rate. Macrophages have a "starry-sky" appearance as they contain apoptotic tumor cells. Gingival swelling or rapidly growing tumor masses in the oral cavity are regarded as the most common initial symptom. Therefore, making a diagnosis based only on clinical examinations is difficult. BL has no specific clinical symptoms, and it is often misdiagnosed as odontogenic infections (Balasubramaniam et al., 2009; Sasaki et al., 2011). Clinical symptoms may vary depending on the affected site. In the head and neck region, BL may cause facial asymmetry within a short time, and it can present with similar features of odontogenic infections. Nasal obstruction, rhinorrhea, facial swelling, unilateral tonsillar enlargement, cervical lympadenopathy, numb chin syndrome, loosening of teeth, ulceration, and rapidly growing mass with or without pain can also be noted (Balasubramaniam et al., 2009; Nikgoo et al., 2009; Sasaki et al., 2011). In these circumstances, patients tend to visit oral and maxillofacial surgery clinics. Therefore, clinicians should further investigate suspicious cases for possible malignancies. Intensive chemotherapy is the preferred treatment modality for BL. The rapid doubling rate of the cells makes them sensitive to cytotoxic agents, and except in the advanced stages of BL, the outcome of combination chemotherapy is excellent in children, with a cure rate of approximately 90% (Banthia et al., 2003,

Molyneux et al., 2012). Despite the fact that the patient lives outside Africa, where the prevalence of the endemic form of the disease is high, some characteristics, such as younger age and the jaw involvement, are compatible with the endemic form. However, the patient was EBV negative, which is a sign of the sporadic form.

Conclusion

Clinicians should be concerned when faced with a child patient who presents with unexplained hypermobility of teeth and swelling that is not associated with caries and apical periodontitis. In such cases, a biopsy and a radiological image should be taken immediately. The cause of the swelling must be determined, such as in our case. Orofacial swelling should be examined carefully in terms of differential diagnosis.

Informed Consent: Necessary information using the patient information form and consent form was taken.
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