Int J Clin Exp Med 2021;14(11):2556-2561
www.ijcem.com /ISSN:1940-5901/IJCEM0131622

Case Report

Oral and maxillofacial swelling may be a sign of other systemic diseases: two cases report

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Received February 7, 2021; Accepted August 16, 2021; Epub November 15, 2021; Published November 30, 2021

Abstract: Factors that contribute to children's oral and maxillofacial swelling are complex, and they can be originated from injections, trauma, and tumors, etc. In the Pediatric Dental Clinic, we often receive patients with odontogenic maxillofacial swellings. However, not all facial swellings are caused by dental infections. Therefore, it's necessary for pediatric dentists to precisely and quickly distinguish non-odontogenic swelling from a clinical aspect. It is of great significance to have a good command of these clinical reserves. The objective of the present article is to show two cases of non-odontogenic maxillofacial swelling in children; one is a 5-year-old boy with Burkitt's lymphoma who showed a fast-growing gingival mass associated with bilateral mandibular molar displacement, and the other is an 8-year-old boy with a mandibular cyst showing no apparent mass region on the gingiva of the left side.

Keywords: Oral and maxillofacial swelling, children, non-infectious disease

Introduction

Burkitt's lymphoma (BL) is an aggressive B-cell lymphoma that grows rapidly and usually happens in children. BL was first described by Dennis Burkitt [1], yet recently, BL was defined as small non-cleaved cell lymphoma [2]. The clinical presentation of Burkitt's lymphoma predominantly presents as jaw swelling and may be caused by various factors, according to epidemiological data [3-5]. The diagnosis of BL relies on morphologic findings, immunopheno typing results, and cytogenetic features [4], and recently Dave SS's team pointed out that molecular diagnosis-Gene-expression profiling was an accurate and quantitative method to distinguish BL from large-B-cell lymphoma [6]. Although BL can be sensitive to chemotherapy, overall mortality of endemic Burkitt's lymphoma remains high due to limited access to health care, late presentation and high relapse rate with advanced-stage disease [7]. For example, a 5-year-old boy, died in this case, unfortunately. Therefore, it is essential to get access to diagnosis and effective treatment in time to improve the survival rate. As pediatric dentists, we need to have related knowledge of Burkitt's lymphoma and refer these patients to professional doctors.

Jaw cysts in children are also an uncommon disease that pediatric dentists encounter at times. In this article, we report about a benign cyst which happened on the jawbone, and jaw cysts present similarities with Odontogenic radicular cyst in clinical and radiographic aspects. The final diagnosis relies on the examination of a biopsy or surgical excision specimens. Though most jaw lesions tend to be benign, sometimes they can be locally aggressive and cause cosmetic and functional damage [8], like the second case which is reported hereafter. Consequently, it is essential to refer children with jaw cysts in a timely manner to specialized clinics preventing injury to the development of jaw bone.

Therefore, this paper highlights the clinical, radiographic and histopathological features of Burkitt's lymphoma and a jaw cyst to help dental practitioners recognize such cases readily and facilitate prompt and potentially life-saving referrals.

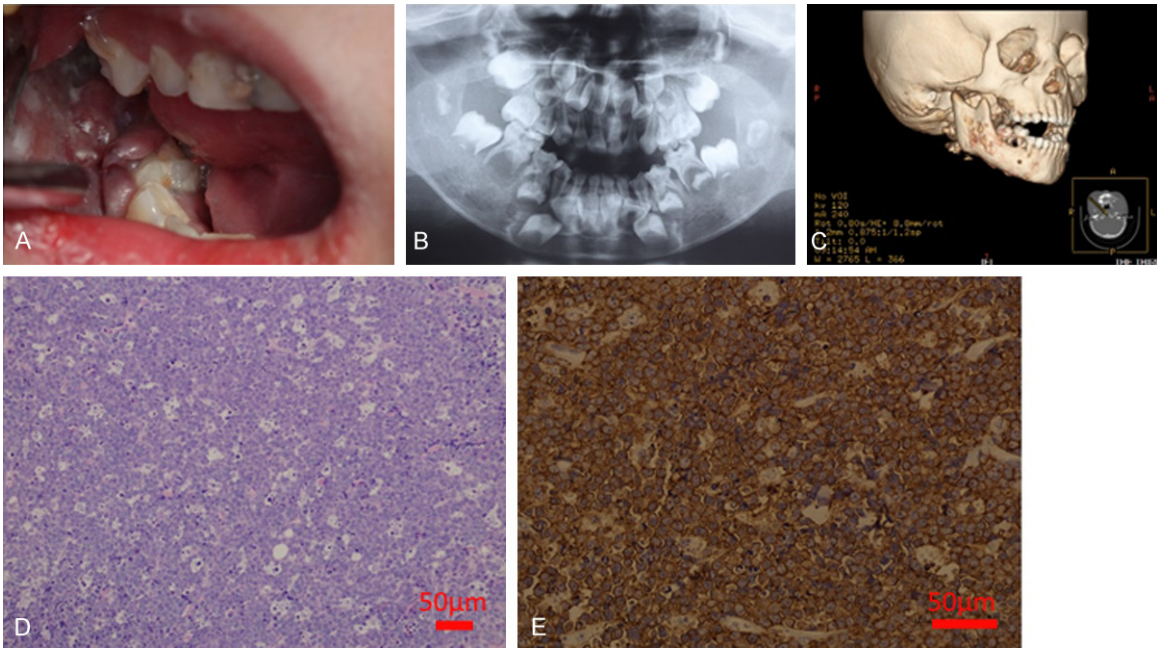

Figure 1. The clinical presentation of a 5-year-old boy. A. Intraoral examination revealed bilateral mandibular molars displacement and gingival mass. B and C. Mandibular molar disappearance and dissolution-like osteolysis. D and E. Pathological results. D. The H&E showed lymphoid-liked tumor cells. E. Ki-67 immunohistochemical results revealed that the proliferation of tumor cells was approaching 100%, which was one of the diagnosis bases of the BL. Scale bar = 50 μm.

Case presentation

Case 1

A 5-year-old boy was referred to the pediatric dentistry department of the stemmatological hospital to evaluate a facial swelling that was present for almost 2 months. The boy was diagnosed with a dental infection and treated with antibiotics for over one month in the district dental clinic. However, the swelling didn't decrease but continued growing.

Extraoral examination showed a firm, non-tender swelling at the angular region of the right mandible and a severe limitation of opening of the mouth. Intraoral examination revealed a fast-growing gingival mass associated with bilateral mandibular molar displacement (**Figure 1A**). Panoramic radiography confirmed the expulsion of mandibular molars with root resorption (**Figure 1B**). Dissolution-like osteolysis was shown in the mandibular body with CT three-dimensional scan (**Figure 1C**).

Laboratory tests showed the patient was negative of HIV and EB virus. Incisional biopsy of the intraoral mass was performed and the result was reported as Burkitt's lymphoma (**Figure 1D**, **1E**). As shown in **Figure 1**, the tumor cells are lymphoid-like cells whose proliferation rate was nearly 100% (**Figure 1E**), forming a classic starry-sky appearance. Based on the pathology results, the patient was then referred to the department of pediatric oncology and hospitalized immediately. Then through thoroughly detailed examination, the patient was finally diagnosed with Grade IV Burkitt's lymphoma with kidney involved. The patient received three chemotherapy sessions. However, he passed away six months later due to multiple organ injury.

Characters: BL is a high-grade Non-Hodgkin's lymphomas (NHL) that mainly affects the immune system and predominately B-cells [1, 2]. BL has a high tendency to occur in Africa and Papua New Guinea [3, 9]. According to studies, people that have limited access to health care, often have diagnosis delay and high relapse rates with late-stage disease findings that contribute to the high mortality rate of BL [7, 9-11]. Therefore, doctors should have knowledge of BL intraoral presentation in cases of delayed treatment, even though the inci-

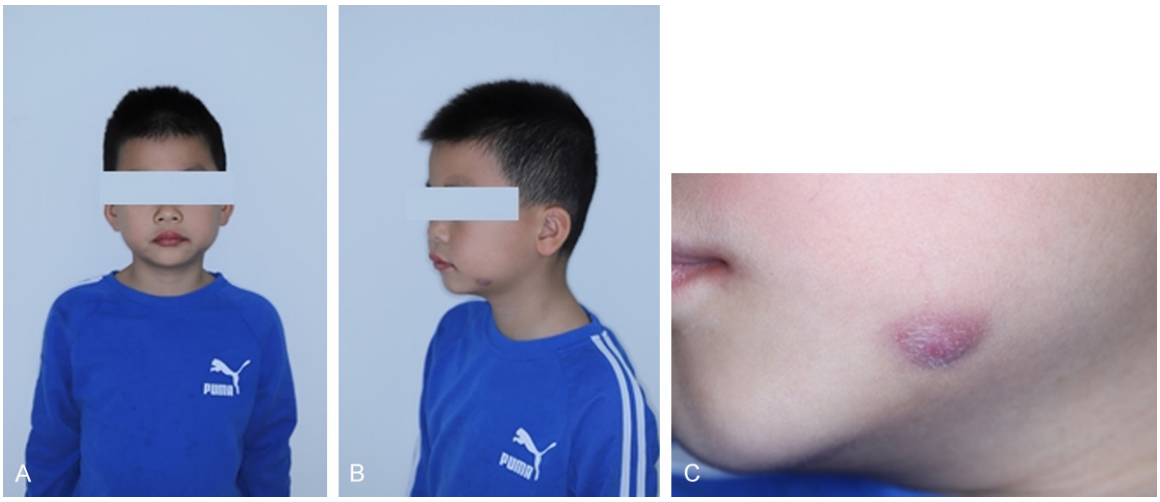

Figure 2. Extraoral examination of the boy. A, B. Craniofacial asymmetry could be seen with a mass on the left cheek. C. The mass was adherent to the skin and toughly touched with a red surface.

dence is much lower in China. Previous data has indicated that the characters of BL are various. Still, the most common presentation is in oral and maxillofacial and symptoms, including fast-growing gingival mass, toothache or dental displacement and jaw swelling.

Additionally, the premolar or molar region is the most frequently involved [4]. In our case, the chief sign of disease was gingival mass associated with jaw swelling. In making a diagnosis, there still exist challenges to distinguish BL from other aggressive B-cell NHL. Therefore, a bone marrow aspirate and histopathologic biopsy examination are important to make an accurate diagnose. Given multicenter studies, intensive combination chemotherapy is the first choice for children with BL.

Case 2

A 7-year-old boy was referred to our institution for repeated swelling of the left cheek without toothache for 40 days. The swelling was slightly relieved after anti-inflammatory treatment (the drug and dose were not specified). However, the left cheek swelling occurred again 20 days later.

Extraoral examination showed a craniofacial asymmetry with a 2.0 cm mass on the left cheek (**Figure 2A**, **2B**). As shown in **Figure 2C**, the mass surface was red, and adherent to the skin. Additionally, there was a little bit of tenderness.

Intraoral examination showed no apparent mass on the gingiva of the left side (**Figure 3Aa-Ac**). Besides, no caries on the crown of the canine and molars and the supporting structures including the periodontal ligament gingival tissue, appeared normal. However, the CT result revealed a well-circumscribed and unilocular radiolucent area at the root apex of the first permanent molar 36 (**Figure 3Ba-Bd**). Detailed CT analysis (from sagittal, axial and coronal section) represented that this radiolucent area measured approximately 1.0×2.0 cm in diameter, closely surrounded by tooth 36.

Surgical curettage of the region was performed and histopathological examination (HE) revealed a cyst of the jaw (**Figure 3C**, **3D**).

Characters: Jaw cysts are uncommon in clinical practice and especially non-radicular cysts in pediatric patients. The estimated incidence in the pediatric population is 7-15%, depending on the age range from 0-18 years old [8, 12, 13]. Generally, most cases tend to be benign, and there is no clinical manifestation and they are usually not aware of them except for infections [12, 14], such as the example in our case. These lesions usually represent slow and expansive growth, chronic swelling but sometimes are associated with bone destruction and recurrence [15]. Unilocular jaw cyst is predominantly seen in the mandible, usually involving the posterior and they show a well-demarcated unilocular shadow in radiological investigations [14]. CT or cone-beam CT is an ideal

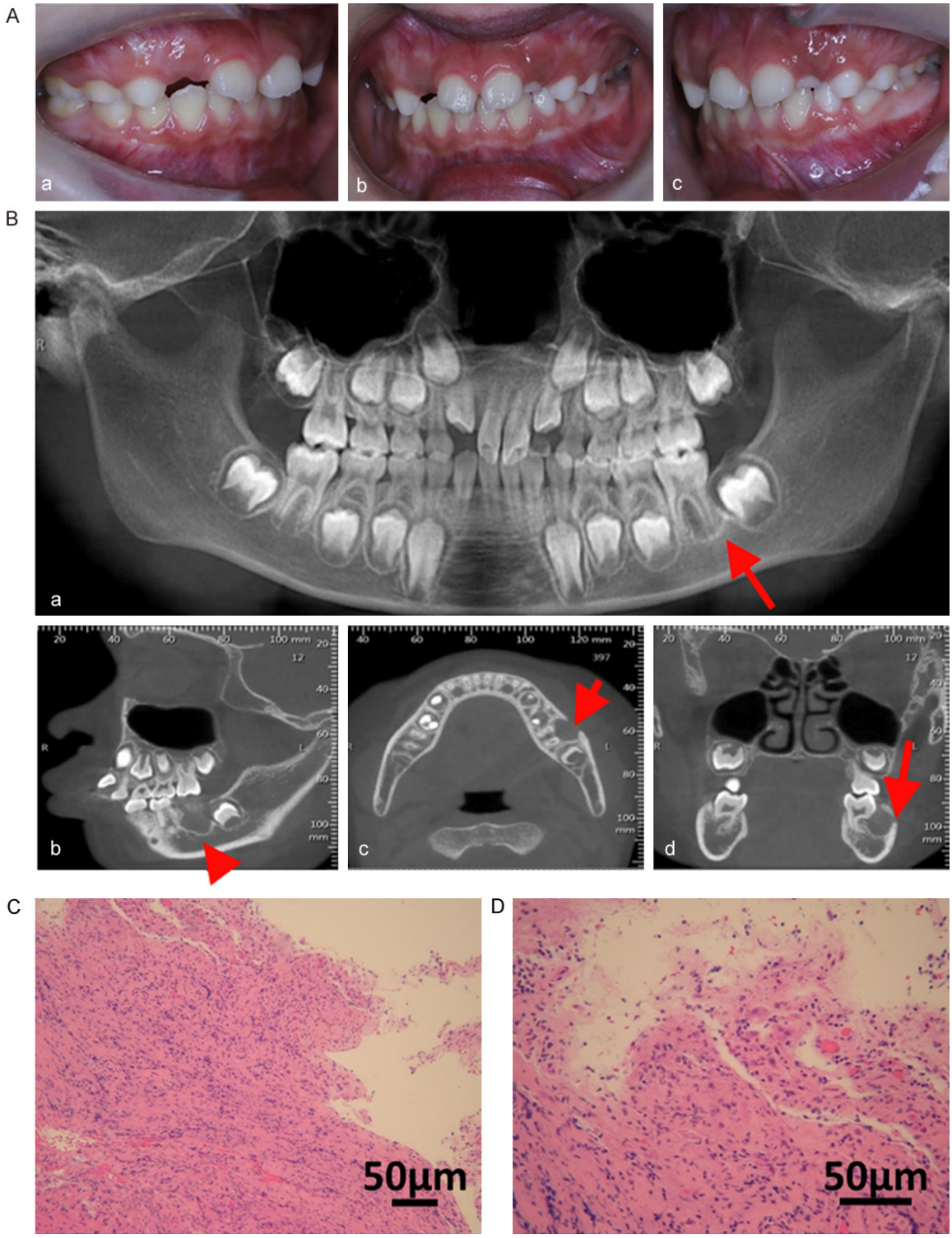

Figure 3. Intraoral examination, CT and pathology results of the boy. (Aa-Ac) Intraoral pictures of left, front and right bites. There was no obvious mass in the pictures. (Ba-Bd) Cone-beam CT represented a well-demarcated unilocular radiolucency at the apex of the root of tooth 36 from the sagittal, axial and coronal section of computed tomographic scan (b-d), respectively. (C and D) A cyst of the jaw, and fibroblast proliferation could be seen in the cyst wall. Scale bar = 50 μm.

investigation to discover intraosseous lesions and provides the most detailed information about the cortical thickness and structural integrity for operative planning [16, 17].

Occasionally these cysts have tenderness and to the touch and they feel like a ping-pong ball [8, 12, 14]. Therefore, knowledge of maxillofacial cysts in children and teenagers is essential to diagnose early and provide treatment quickly. Early lesions can be successfully treated with irradiation or cautery and curettage.

Discussion

Factors that contribute to children's oral and maxillofacial swelling are complex, and they can be originated from injections, trauma and tumors, etc. In the Pediatric Dental Clinic, we often receive patients with an odontogenic maxillofacial bump. However, not all facial swellings are caused by dental infections or trauma. Pediatric oral maxillofacial non-odontogenic cysts are unusual, and data on them is sparse, despite many studies on jaw cysts. These lesions may be either symptomatic or asymptomatic and could be identified via careful clinical and radiographic examinations. Since these non-odontogenic swellings are challenging to diagnose straight forwardly, knowledge of maxillofacial cysts in children is necessary for dentistry practitioners. These situations often require a certain level of clinical exposure and expert skill in examining or managing and radiological investigations. Hence, general dentists need to have updated knowledge on odontogenic and non-odontogenic cysts or swellings to get allow a quick diagnosis, early treatment and favorable prognosis.

Acknowledgements

This work was financially supported by grants from General program of Jiangsu Natural Science Foundation (BK20191121). Six Talent Peaks Project in Jiangsu Province (2019-wsw-128) and Nanjing funding project for students studying abroad (2019-B-43).

Disclosure of conflict of interest

None.

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