Short communication

Widely disseminated sporadic Burkitt lymphoma initially presented as oral manifestations in a 6-year-old boy

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ABSTRACT

Burkitt lymphoma, a subtype of non-Hodgkin's lymphoma, is an aggressive neoplasm with three variants that are endemic, sporadic, and immunodeficiency associated. We present an unusual case of sporadic Burkitt lymphoma in a 6-year-old boy who initially presented with hypermobile teeth and no other specific signs or symptoms. On dental radiography, the patient was found to have alveolar bone resorption adjacent to the maxillary first molars, with the appearance of floating teeth. In addition, magnetic resonance imaging (MRI) showed extensive soft tissue masses involving four quadrants of the jaws. A definitive diagnosis of Burkitt lymphoma was made based on tissue and bone marrow biopsy. Subsequent images, including abdominal computed tomography (CT) and bone scan, revealed wide dissemination of the lymphoma into the abdominal cavity, pancreas, and numerous bones. This case suggests the possibility of dental complaints as an initial clinical manifestation of sporadic Burkitt lymphoma and emphasizes the role of dentists in early detection of the disease to improve prognosis.

1. Introduction

Burkitt lymphoma should be diagnosed as quickly as possible and requires prompt intervention because it is a rapidly growing high-grade malignant neoplasm.^{1,2} However, the clinical features of oral lymphoma, especially of lesions involving alveolar bones are not specific, dentists often misdiagnose them as tooth-related diseases or any other problem and attempt to solve the problem with dental treatment. For example, dentists usually evaluate hypermobile teeth with a history of jaw pain or floating teeth with resorption of the adjacent alveolar bone as dental abscess or Langerhans cell histiocytosis, respectively. This might delay diagnosis and treatment, thereby worsening the prognosis. We observed a case of stage IV sporadic Burkitt lymphoma in a 6-year-old boy that initially presented as hypermobile teeth and jaw expansion, with no other specific signs or symptoms. This case highlights the dentist's role in the early detection of Burkitt lymphoma involving the jaw because the prognosis of the patient can be improved if the diagnosis is made as quickly as possible.

2. Case report

A 6-year-old boy visited for an evaluation of hypermobility of the

maxillary right first molar. The boy's past medical history was non-specific, and he had no history of progressive systemic symptoms such as fatigue, weakness, or fever.

Intraoral examination revealed painless gingival and mucosal swelling in both posterior maxillas and mandibles. Furthermore, the right mandibular first molar was tilted lingually. A periapical radiograph (Fig. 1A) showed severe alveolar bone resorption around the maxillary right first molar, with uncompleted root development. A panoramic (Fig. 1B) view showed loss of the lamina dura around all four first molars. Enlarged follicular spaces were also observed in both mandibular second premolars. Cone-beam computed tomography (CT) images (Fig. 1C and D) showed large tumorous lesions occupying both maxillary sinuses, causing partial resorption of the sinus wall.

Histopathologic findings of the buccal mucosa of the upper right first molar showed sheets of monomorphic lymphoid cells and intervening phagocytic cells, imparting a "starry-sky appearance". Immunohistochemical staining was positive for CD10, CD20, and MUM1, which are specific to B-cell lymphocytes and late stages of B-cell differentiation. Ki-67 staining was positive in more than 95% of the cells, revealing high proliferative activity. Overall, histopathology results supported the diagnosis of Burkitt lymphoma. The patient was immediately referred to the Hematology-Oncology Department for

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Fig. 1. (A) A periapical view showing no bone support around the maxillary right first molar. (B) A panoramic radiograph showing loss of the lamina dura around all four first molars and the bone crypts of all second molars. (C & D) Cone-beam CT images showing diffuse bone resorption and tumorous lesions occupying both maxillary sinuses.

definitive care.

Subsequent abdominal CT, bone scan, and positron emission tomography (PET)-CT (Fig. 2) revealed multiple lesions involving the right abdominal cavity, pancreas, multiple retroperitoneal lymph nodes, and left gastric, splenic and portocaval areas as well as multiple bone lesions. Bone marrow involvement was also observed. The patient was diagnosed with stage IV Burkitt lymphoma. The patient was treated with several courses of intensive chemotherapy, and has been in

remission for 20 months.

Due to the retrospective nature of this study, it was granted an exemption in writing by the institutional review board (IRB) of Pusan National University Dental Hospital (IRB No. PNUDH-2017-13).

3. Discussion

General dentists are inexperienced in diagnosing Burkitt lymphoma

Fig. 2. PET-CT images showing multiple lesions involving the jaw, abdominal cavity, vertebrae and four limbs.

of the jaw in non-endemic areas due to the rarity of this disease. Burkitt lymphoma, a non-Hodgkin's lymphoma (NHL), is a malignant B-cell tumor classified into endemic, sporadic, and immunodeficiency associated subtypes.^{1,2} Although histopathologic findings are similar among the three forms, they have different clinical features with respect to age distribution and area of prevalence. The endemic form that is frequently found in Africa is widely reported to occur in early childhood at a peak age of 6 years; in contrast, the sporadic form has no geographic predilection and occurs mainly in children and adolescents. The endemic form commonly involves the jaw bones and abdomen and typically presents as tooth mobility, jaw expansion, and an abdominal mass.^{1,2} Conversely, jaw involvement is not common in the sporadic form.^{3,4}

In our case, floating teeth was also the only problem detected before other symptoms appeared, even though most organs were already involved and the disease was clinically advanced. Thus, the presence of floating teeth on plain radiography reflecting that destructive process in the maxilla and/or mandible might suggest sporadic Burkitt lymphoma as well as infectious, hematologic or metabolic causes including Langerhans cell histiocytosis. Recently, a similar case of sporadic Burkitt lymphoma with dental manifestations was reported in a 12-year-old boy. The boy was admitted with a one-week history of urinary retention and dysuria due to a prostate mass.⁵ Preceding these symptoms, he had complaints of jaw pain for 4 weeks that had initially been treated as a dental abscess. Based on these cases, dentists should acknowledge that although jaw involvement is rare, it is a potential site for Burkitt lymphoma.

In diagnosing Burkitt lymphoma involving the jaw, dental radiography can play an important role; indeed, radiographic changes in the jaw are usually present prior to the emergence of clinical signs of the disease. Radiologically, the earliest signs are loss of the lamina dura and enlargement of developing tooth crypts,⁶ which can be firstly recognized by general dentists. The radiographic features of Burkitt lymphoma involving the jaw are osteolytic lesions in the jaw around the permanent tooth buds, attenuation of the lamina dura, displacement of developing teeth, widening of follicular spaces, and a floating appearance of teeth.^{6–9} Thus, the unexplained hypermobility of teeth and the displacement of developing teeth with cortical attenuation in a child

were ominous signs that created suspicion regarding malignant lesions.

Although Burkitt lymphoma is an aggressive neoplasm that has a poor prognosis, the cure rate of the disease depends on its stage. A study on oral Burkitt lymphoma reported that even stage III patients had a 97% survival rate. In contrast, the prognosis of stage IV Burkitt lymphoma is still dismal: the survival rate of stage IV patients was substantially lower at 27%, suggesting that early diagnosis of Burkitt lymphoma is crucial.¹⁰ Thus, dentists should be aware that dental problems involving floating teeth and/or alveolar bone resorption might be the first clinical manifestation of Burkitt lymphoma. After our prompt biopsy, the extent of the disease was identified, and the patient underwent chemotherapy immediately. Accordingly, the patient had a good prognosis, and he has been in remission for 20 months.

Conflict of interest

None.

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