African Type Burkitt’s Lymphoma: A Case Report

Burkitt’s lymphoma (BL) is an extranodal malignancy with distinct epidemiological, clinical, pathological, immunological, and molecular cytogenetic characteristics. The prognosis for Burkitt’s lymphoma in the past was poor, with a median survival time of only 11 months. More recent trials with more intensive, multiagent chemotherapeutic protocols show a 68% remission rate after 38 months follow-up.

In this case report we present of a 10-year-old male patient with Burkitt’s lenfoma diagnosed clinically and histopathologically, treated by chemotherapy and followed up eighteen months post chemotherapy.

KEYWORDS
Burkitt’s Lymphoma, Gingival enlargement
INTRODUCTION

Burkitt’s lymphoma (BL) is an extranodal malignancy with distinct epidemiological, clinical, pathological, immunological, and molecular cytogenetic characteristics. Burkitt’s lymphoma is a malignancy of B-lymphocyte origin that represents an undifferentiated lymphoma. During the 1950s, Dennis Burkitt described rapidly growing jaw and abdominal lymphoid tumors in East African children. The Tumor, Burkitt’s lymphoma, is the human cancer most closely linked with a virus. Ebstein-Barr virus is associated with 90% of African patients with BL, but this percentage is considerably lower for BL seen in other parts of the world. The reason for the association between BL and Ebstein-Barr virus remains unknown. The peak age of the African BL is between 5 and 7 years. The male to female ratio ranges between 2:1 and 6,5:1 and is much higher in patients under 13 with an incidence of 0,1 to 0,3/100,000.

The clinical presentation of BL is characterized by rapid progression of symptoms with frequent multifocal extranodal involvement, including central nervous system involvement. With in the oral cavity, this tumour can progress very fast and appears as a facial swelling or exophytic mass involving the jaws. Involvement of facial bones and oral cavity occurs in less than 30% of cases in most series. The African form of BL most frequently manifests itself as rapidly growing, extranodal jaw tumors in young children, but it also may be first detected as an abdominal involving the kidneys or ovaries. The growth of the tumor mass may produced facial swelling and propitosis. Pain, tenderness, and paresthesia are usually minimal, although marked tooth mobility may be present because of aggressive destruction of the alveolar bone.

The radiographic features are consistent with a malignant process and include a radiolucent destruction of the bone with ragged, ill-defined margins. This process may begin as several smaller sites, which eventually enlarge and coalesce. Patchy loss of the lamina dura has been mentioned as an early sign of BL.

BL histopathologically represents an undifferentiated, small, noncleaved B-cell lymphoma. The lesional tissue invades as broad sheets of tumor cells and exhibits round nuclei with several prominent nucleoli and numerous mitoses. A classic starry-sky pattern is associated with lesional tissue, a phenomenon that is caused by the presence of macrophages within the tumor tissue. Tumors with a similar histomorphology, commonly referred to as American Burkitt’s lymphoma, have been observed in other countries where the neoplasms usually first detected as an abdominal mass.

BL lesions have a dramatic response to chemotherapy, particularly cyclophosphamide. The tumor also has been shown to be sensitive to methotrexate, vincristine, and cytarabine. Combinations of drugs have achieved remissions in more than 90% of patients. Unfortunately, most experience recurrences and ultimately die of their disease. The prognosis for Burkitt’s lymphoma in the past was poor, with a median survival time of only 11 months. More recent trials with more intensive, multiagent chemotherapeutic protocols show a 68% remission rate after 38 months follow-up.

In this case report we present of a 10-year-old male patient with Burkitt’s lymphoma from Turkey diagnosed by gingival overgrowth, alveolar bone loss, tooth mobility and histopathologically, treated by chemotherapy and followed up eighteen months post chemotherapy.

CASE REPORT

A ten year old boy presented with a six week history of painful and progressive swelling of the jaw bilaterally. It was considered as dental abscess at the local hospital and antibiotic therapy was considered for two weeks. The lesions of the gingiva and the swelling of the jaw did not resolve. He had also headache, fever and weight loss. A gingival biopsy was performed at the local hospi-
tal and histopathological examination was reported as “chronic inflammatory process”.

The patient admitted to our hospital with the same complaints in November 2002. Oral clinical examination revealed gingival overgrowth in both maxilla and mandible and marked tooth mobility. The marginal and attached gingivae in almost all teeth, on both buccal-labial palatal-lingual aspect, were severely inflamed, bright red in appearance, and hyperplastic (Figs 1 and 2).

Initial physical examination showed a marked swelling of the mandibula bilaterally, facial edema and also swelling of the cheeks extending to the zygomatic arcus on the left. Two lymph nodes are palpable, each smaller than 0.5 cm in the right anterior cervical region. The rest of the physical examination was unremarkable.

The orthopantomographic examination revealed alveolar bone resorption (Fig 3). A new biopsy was performed from the right lower gingiva. Both the new biopsy and the previous one were reported as “Burkitt’s lymphoma” (Fig 4).

The initial laboratory examinations of the patient were as follows: Hb:9.1 g/dl, WBC: 7800/mm$^3$, platelets: 315,000/mm$^3$ with a normal differential; serum biochemistry was normal except a LDH of 1737 IU. Bone marrow examination revealed diffuse involvement by lymphoblasts; computerized tomography of the head showed lytic lesions on the right frontoparietal region with an epidural soft tissue component and skin involvement; also a left sphenoidal lytic lesion with a soft tissue mass extending to the anterior of the temporal lobe and the orbit extraaxially.
was detected (Fig 5). A thoracic and abdominal computerized tomography showed bilateral kidney lesions. The other bones except the facial ones were normal on plain X-rays. The cytological examination of cerebrospinal fluid was negative.

With the diagnosis of stage IV Burkitt’s type non-Hodgkin’s lymphoma; we treated the patient with LMB-B chemotherapy regimen which included vincristine, cyclophosphamide, prednisolone, adriamycine, high dose methotrexate with leucovorine rescue. The patient had a complete response to chemotherapy. In April 2003, chemotherapy was ended when the patient was in complete remission. He is still under regular follow-up with no evidence of disease eighteen months after cessation of treatment. Fig 6 and 7 shows the post chemotherapy clinical and radiographic picture of normal tooth development.

**DISCUSSION**

Diagnosis of Burkitt’s lymphoma, especially when the sole presentation is in the maxillofacial region, is very difficult. The clinical presentation of the disease may mimic a wide variety of disorders more commonly found within the jaws. Because primary presentation of the disease is often in the mouth and jaws, a high index of suspicion is required from the medical staff in order to assure early diagnosis and a better prognosis of the disease. Clinical differential diagnosis of Burkitt’s lymphoma should include: acute dentoalveolar abscess, osteomyelitis, rhabdomyosarcoma, periapical lesions, ameloblastoma, eosinophilic granuloma, multiple myeloma, leukemia, and other fibro-osseous lesions. The signs and symptoms of oral Burkitt’s lymphoma include mobile teeth, toothache, oral masses, gingival enlargement, pain, and jaw expansion. Additional to all these findings Ugboko et al. reported a case with a lower lip paresthesia. BL is one of the most rapidly growing tumors, doubling in size every 24 hours. Because of its extremely rapid growth rate, prompt diagnosis before the initiation of specific treatment is imperative for a favorable prognosis.
Alveolar bone resorption and lamina dura loss has been seen in radiological examination. In the literature, radiographic findings of the involved jaw have been well described by Burkitt, Adatia, Hupp et al., Wood et al., Anavi et al., and Hanazawa et al. Our findings were similar. According to our experience, dentists and maxillofacial surgeons should evaluate oral and radiologic findings as facial swelling and gingival enlargement, alveolar bone loss with a suspect of BL considering the rapid growth rate and poor diagnosis.

REFERENCES


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