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## Burkitt's lymphoma: A child's case presenting in the maxilla. Clinical and radiological aspects

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### Abstract

Burkitt's lymphoma (BL) is a neoplasm which, despite its very aggressive behaviour is potentially curable. It typically affects the paediatric population. BL belongs to the non-Hodgkin lymphomas group, and is the first human tumour undoubtedly related to a viral origin (Epstein-Barr virus). Two main clinical subtypes are recognized: endemic or African type, and sporadic type; HIV associated BL constitutes a third type. Although common in endemic BL, maxillary involvement is rare in sporadic cases. This, together with the clinical lack of specificity associated to this location, makes diagnosis difficult. New chemotherapeutic protocols achieve a high survival rate. Most important prognostic factors are location and tumour stage. We report a paediatric case of BL presenting in the maxilla, with a review and a description of the characteristics of the disease.

**Key words:** Burkitt's lymphoma, non hodgkin lymphomas, maxillary swelling, chemotherapy.

### Introduction

BL is an aggressive tumour typically, but not exclusively, affecting children. It belongs to the undifferentiated non-Hodgkin lymphomas and it is an extra nodal tumour composed of a monoclonal proliferation of undifferentiated B cells (1). BL is related to Epstein-Barr virus (EBV) (1-3). Three clinical subtypes are admitted: African or endemic, American or sporadic, and HIV associated subtypes (4). The Irish surgeon Denis Burkitt

first reported in 1958 its high prevalence in the maxillae of Central-African children (5-7); its presence in the rest of the world was later observed. BL represents 3-5% of all lymphomas. It usually affects children, representing up to 40% of all children's head and neck lymphomas (3). Endemic BL affects African communities in the central and sub-Saharan areas, where its incidence is 5-15 cases per 100000 children. The incidence of sporadic subtype, in the rest of the world is 1-3 cases per

100000 children (2). HIV associated subtype represents 35-40% of non-Hodgkin lymphomas appearing in positive HIV population. Although silent in early stages, BL is a rapidly growing and expanding tumour. Signs and symptoms depend on primary location of the tumour and its degree of spread (3). Endemic subtype mainly affects maxillary bones (5,6), but secondarily other facial bones and abdominal organs may become affected. Sporadic subtype usually presents as an abdominal or pelvic mass, and Waldeyer's ring is commonly affected (3).

From 1970 on, the development of a combined protocol of chemotherapeutic agents and the use of new antibiotics have determined an improvement in the prognosis of BL affected patients. Anatomical sites and early treatment are today considered main prognostic factors.

### Case Report

A male white 5 year-old child from Córdoba province (Spain) was admitted to the Oral and Maxillofacial Surgery Unit in July 1993 because of a one month painful hard swelling in his right upper bucal sulcus accompanied by a non adherent right submandibular adenopathy. No diplopia, anosmia or teeth mobility were detected. Among his past history, rickets could be elicited. Orthopantomogram showed upper right maxillary bone destructuration (Fig. 1). Waters view revealed destruction of the right zygomaticomaxillary buttress and maxillary sinus opacification. Computed tomography (CT) showed a 5x5 cm soft density mass affecting the right maxillary sinus, orbital floor, and lateral nasal wall, that laterally displaced the zygomatic arch (Fig. 2). No tumour was detected at any other location. A biopsy was taken and the result was Burkitt's lymphoma. Microscopic description reported a diffuse "starry sky" pattern. Phenotype was positive for CD20 and negative for CD2. The tumour showed a high proliferative index (MIB1) and a marked expression of p53 and Rb genes. Neither in situ hybridation nor immunohistochemical techniques showed evidence of EBV. Once haematology unit had staged the disease, chemotherapy composed of cyclophosphamide, vincristine, metotrexate, and prednisone (COMP), with intrathecal metotrexate was instituted. Later on, loco regional radiotherapy with a total dose of 26 Gy with Co-60 photons on the right maxillary sinus, orbital floor, nasal cavity, hard palate and first lymph node echelon was applied.

Twelve years after diagnosis, clinical and radiographic follow up confirms a complete remission of the disease. In this moment, the patient is under orthodontic treatment and shows right maxillomandibular hypoplasia with moderate external facial asymmetry. Orthopantomogram shows lack of development of right maxillary teeth, probably related to chemo and radiotherapeutic treatment received at the age of five years (Fig. 3).



Fig. 1. Orthopantomogram at presentation.



Fig. 2. CT showing the tumour in right maxillary sinus.



Fig. 3. Orthopantomogram at ten years of age, showing a retained upper right canine and some other dental alterations caused by radiotherapy and chemotherapy received at the age of 5 years.

**Discussion**

Paediatric non-Hodgkin lymphomas evolve more rapidly than those in adulthood (3). There exist clinico-pathological differences among the different subtypes of BL (Table 1). Mean age of presentation in the endemic subtype is 7 years of age, while that in sporadic subtypes is 14 years of age (5). Maxillary involvement in the endemic subtype is age-dependant; 75% of affected children under 5 years of age develop maxillary tumour whereas only 25% of those over 14 years of age do. This fact may suggest a stimulating effect induced by growth factors involved in child's dental and osseous development (2). Upper jaw is twice affected as lower jaw and several quadrants may be simultaneously affected (2). BL represents 45% of all maxillary tumours among children under 14 years of age in Sub-Saharan Africa (1). Males are affected three times more often than girls (8). In sporadic cases of BL bone maxillary tumours appear only in 12-20% , and there appears to be no geographic, climate or age relationship. HIV related BL mainly affects lymph nodes, and bone marrow is involved in only 30% of cases (9).

EBV appears in 97% of endemic LB, but only in 20-30% of non endemic cases (4,8). In the case here reported, there was no evidence of EBV.

Temporary or definitive teeth loss or displacement, gingival expansion, numb chin, and tooth ache secondary to pulpal infiltration with tumour cells (what is characteristic in this tumour) are early signs of BL (1,9-11). Radiologically, the tumour appears as a radiolucent lesion with early loss of the lamina dura around developing teeth (1,8,12). Symptoms due to the general affection of the patient, or to compression of adjacent anatomical structures by the tumour, such as pharynx, trachea, superior cava vein or central nervous system (CNS) may appear (3,4). Bone scintigraphy shows hypercaptation in early stages. Bone marrow biopsy, cytogenetic studies, and immunophenotypic analysis support definitive

diagnosis of the disease. The classical histological pattern is conformed by a thick and diffuse monoclonal proliferation of small dark cytoplasm noncleaved neoplastic cells with some interspersed clear macrophagic cells (1-4). The high mitotic index observed in the slices indicates BL's high proliferative activity whose tumoral duplication time is approximately as high as 26 hours (4,8). In both endemic and sporadic clinical subtypes, t(8,14) translocation appears (1-3, 8-10) with differences at the molecular level with regard to the sites of chromosomal rupture.

Intensive chemotherapy has allowed a survival rate to five years for early stages up to 90-100% and a larger disease free period, with less long term toxicity. Local complications include caries, mucositis, gingivitis, herpetic stomatitis and candidiasis (13,14). New chemotherapeutic agents such as daunorubicine, arabinoside-C, tioguanide and hidroxiurea have been added to the classical treatment scheme COMP. When involvement of CNS is suspected prophylaxis by means of intrathecal instillation with metotrexate or citarabine is recommended. Bone marrow transplantation is an alternative in cases of persistence in spite of intensive treatment. Monoclonal antibodies constitute a future therapeutic strategy in the treatment of these tumours (3). Due to its rapid spread, surgical resection its limited to those cases with obstructive symptomatology compromising survival (2). Cranio-spinal irradiation may be effective as prophylaxis of spread to the CNS and as a treatment alternative to achieve immediate relief of symptoms derived from compression to CNS structures. The role of radiotherapy as a coadjuvant treatment for the disease affecting the jaws is still to be determined. According to literature, a dose of 7-30 Gy is recommended (3). In the case here reported, the total dose of radiation was 26 Gy.

As in the case here shown (Fig. 3), children which have received radiation may develop noticeable impairment

**Table 1.** Differential characteristics of BL subtypes. BL: Burkitt's lymphoma; t: translocation; HIV: human immunodeficiency virus.

BL clinical subtype	Endemic	Sporadic	HIV related
Epidemiology	Central Africa and New Guinea	Rest of the world: USA, Europe	Rest of the world: USA, Europe
Incidence	10 cases/10 <sup>5</sup>	1-3 cases/10 <sup>5</sup>	6 /10 <sup>3</sup> AIDS cases
Genetics	t(8,14) breaking point over c-myc	t(8,14) breaking point in c-myc	t(8,14) breaking point in c-myc
Location	Jaws and facial bones in children	Abdominal mass in older children and adults	Lymphatic organ in adults
Survival with treatment	Good (90%)	Good (90%)	Lower (40%)
Serology for EBV	97%	20-30%	20-30%

in the development of dental and maxillofacial structures, such as blunt roots, incomplete calcification, facial asymmetric growth and occlusal changes (15,16). Specific periodontal problems may also appear showing a decrease of the osteoblastic and osteoclastic activity in the alveolar bone, almost complete lack of osteocytes, and degeneration of fat marrow. Odontoblastic activity stops in developing teeth, with the result of agenesis and microdontias (15,17).

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