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Bilateral Burkitt's Lymphoma of Maxillary Sinuses: A Case Report

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Abstract **Case Report**

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. Imaging and histology play a crucial role in the diagnosis. More than 100 craniomaxillofacial cases have been reported in published studies and have had a variety of stemming locations, manifestations, complications, and other characteristics. The present case report introduces a unique case of BL originating in the maxillary sinuses bilaterally.

Keywords: Burkitt's lymphoma, maxillary, pediatric, CT scan.

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BACKGROUND

Lymphomas are malignant neoplasms of lymphocyte cell lines that are more common in the head and neck than squamous cell carcinoma. They are classified into two types: Hodgkin's (HL) and Non-(NH). Hodgkin's (NHL). HL often manifests as a nodal illness, with a preference for the head and mediastinal nodes. NHL is a diverse collection of neoplasms that typically arise in lymph nodes, although up to 24%-40% of cases occur in non-lymph nodal areas such as the gastrointestinal tract, skin, bone, and Waldeyer's ring [1]. Burkitt's lymphoma (BL) is an aggressive type of non-B-cell Hodgkin's lymphoma that is widespread in Africa and sporadic elsewhere. It is commonly diagnosed in children and young adults, although it is only rarely detected in middle-aged individuals. The mandible, maxilla, and abdomen are commonly affected in the endemic type.

Orofacial BL is commonly found in the jaw bones and is connected with tooth movement, dental discomfort, and jaw extension. Life-threatening complications such as airway and gastrointestinal blockage, as well as acute renal failure, have been recorded with BL [2]. Since this tumor has an aggressive behavior, immediate histopathologic and cytogenetic evaluation and early treatment are essential. In this case, we describe the diagnosis and treatment of Burkitt's lymphoma, which manifested as face enlargement with intraoral and extraoral swelling.

CASE REPORT

A 3 year old patient, without any particular pathological history, presented to the emergency room with a right jugal swelling, dyspnea and a progressive alteration of the general state. Clinical examination revealed a large, firm, painful right jugal mass with no inflammatory signs associated with homolateral exophthalmos. The examination also revealed abdominal distension. No organomegalia nor lymphadenopathy were found. The rest of the examination was normal. The biological workup was unremarkable. CT scans were performed urgently, showing two tumor processes centered on the maxillary sinuses, the larger one being right and responsible for significant bone lysis with endo-orbital and endo-nasal invasion but without endo-cranial extension (Figure 1).

CT of the thorax was normal, but CT of the abdomen revealed bilateral nodular nephromegaly in relation to lymphomatous infiltration (Figure 2). The diagnosis of Burkitt's lymphoma was strongly suspected and chemotherapy was immediately started. After 8 days of treatment, a follow-up CT scan was performed, showing a clear regression in size of the two maxillary sinus lesions and the renal lymphomatous infiltration indicating a good response to the chemotherapy (Figure 3 and 4). The patient is currently still under control and seems to have a good evolution.

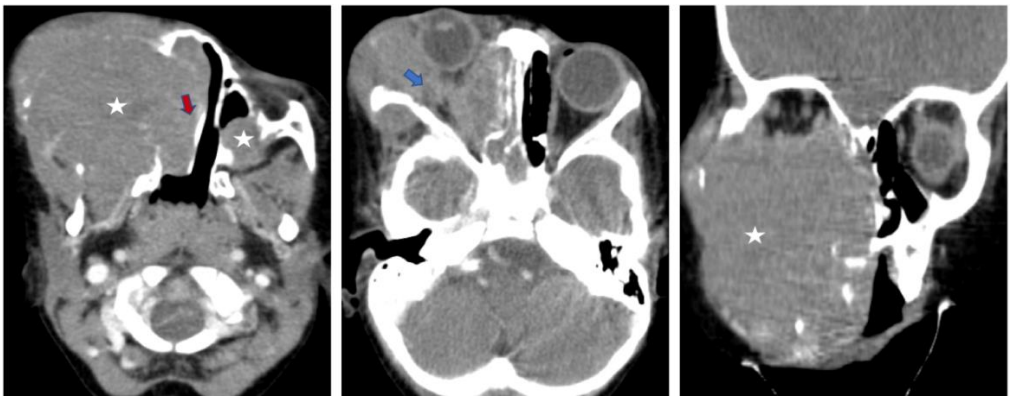

Figure1: Facial CT scan showing two tumor processes centered on the maxillary sinuses (white star), the larger one being on the right and responsible for significant bone lysis with endo-orbital (red arrow) and endo-nasal (blue arrow) invasion but without endo-cranial extension

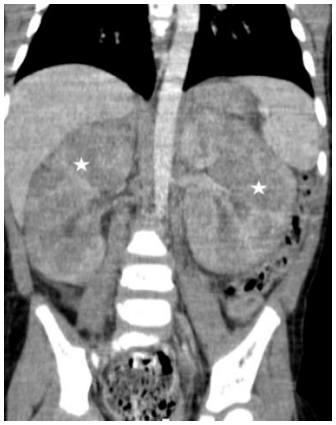

Figure 2: Abdomen CT scan showing bilateral nodular nephromegaly

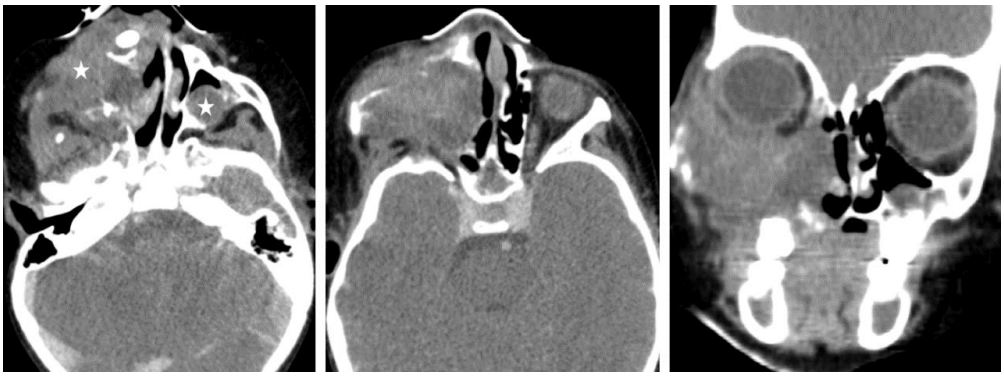

Figure 3: Post chemotherapy facial CT showing a clear regression of the sinusal masses

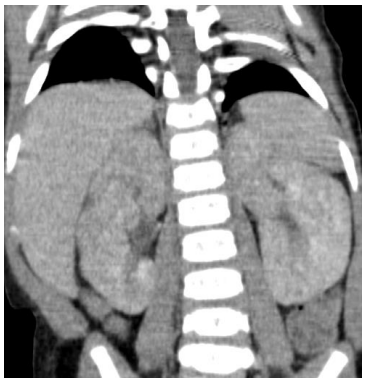

Figure 4: Post chemotherapy abdominal CT showing a clear regressions of the nodular nephromegaly

DISCUSSION

Primary NHL seldom affects the nasal cavities and paranasal sinuses. Lymphomas with main extra-nodal locations include the liver, soft tissue, dura, bone, stomach, intestine, bone marrow, and others [3]. There are no defined clinical signs of BL, and they might vary according on the afflicted region. Head and neck BL most commonly manifests as lymphadenopathy with main involvement of the nasal cavity and paranasal sinuses. The maxillary sinus is usually implicated in paranasal BL, while the sphenoethmoidal sinuses are less commonly affected. Nasal obstruction, epistaxis, headache, and unilateral face, cheek, and nasal edema are the most typical presenting symptoms of sinonasal lymphomas. [4-6].

These symptoms are typically associated with chronic rhinosinusitis. The clinical manifestations of BL in the head and neck area are highly dependent on the anatomic location of involvement. As a result, BLs in this location may be misdiagnosed as other diseases in the early stages. Sinonasal lymphoma might show as a polypoid mass-like lesion on endoscopy. As a result, it's easy to mistake it with paranasal sinusitis, polypoid illness, or other tumorous disorders [6]. A high index of suspicion is required to guarantee prompt referral to a specialist center. Because the illness has an extraordinarily quick doubling time (24 to 48 h), delays in treatment may raise the risk of cancer spreading to other regions of the body [6].

Initially, BL can appear on imaging as many ill-defined radiolucencies that eventually merge to form growing bigger lucencies. The cortical margins of the lesions dissolve due to their fast growth. The crypt of the growing teeth and the erupted teeth are significantly shifted. The lamina dura is damaged, as are the cortical boundaries of surrounding structures such the maxillary sinus, nasal floor, orbital walls, and mandibular inferior border. In certain situations, the cortical bone sheath of the inferior alveolar canal is lost. Periosteal involvement can cause a "sun-ray" look, which is uncommon [7].

Squamous cell carcinoma, ossifying fibroma, osteosarcoma, Langerhans cell disease, salivary gland cancers, particularly mucoepidermoid carcinoma, and fibrous dysplasia are also prevalent causes of BL [8-9]. CT and magnetic resonance imaging can assist assess the degree of the primary disease, and a whole body disease workup should include positron emission tomography and bone marrow and cerebrospinal fluid examination. To establish the diagnosis, the least intrusive method should be used as soon as possible [10]. In rare situations, abdominal and chest CT scans to assess for distant illness dissemination are needed. Whenever feasible, the main lesion should be biopsied directly, ideally with an endoscope.

On CT, it is sometimes difficult to distinguish malignancies from other inflammatory processes. Any questionable discoveries should be biopsied and frozen section examined. BL is very vulnerable to cytotoxic medicines since it is one of the quickest growing tumors. As a result, complicated chemotherapy, which includes cyclophosphamide, vincristine, methotrexate, and prednisone, is the major treatment method [4]. Long-term survival rates for BL regimens confined to the head and neck area have reached 90%. Individuals with advanced stages illness, a bulky mass, a high lactate dehydrogenase level, and involvement of the central nervous system (CNS) or bone marrow have a bad prognosis [6]. Our patient with stage I BL responded well to treatment, as seen by our follow-up outcomes.

CONCLUSION

Burkitt's lymphoma in the paranasal sinuses is rare but demands prompt diagnosis. In the case of a child with facial swelling with suspected tumour lesions, urgent imaging and endoscopic biopsy are needed. The importance of early cancer diagnosis of BL needs for increase awareness and education. Late and advanced presentation of cancers impacts on treatment, prognosis and ultimately survival.

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