International Journal of Contemporary Pediatrics
Larissa MT et al. Int J Contemp Pediatr. 2025 Feb;12(2):318-320
http://www.ijpediatrics.com
pISSN 2349-3283 | eISSN 2349-3291

DOI: https://dx.doi.org/10.18203/2349-3291.ijcp20250016

Case Report

Burkitt lymphoma: an unusual location in children, report of a case

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Received: 23 December 2024
Accepted: 06 January 2025

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ABSTRACT

Burkitt lymphoma (BL) is the most common malignant non-Hodgkin lymphoma in African children. It is a very aggressive lymphoma characterized by a high degree of proliferation. If left untreated, the outcome is quickly fatal. The aim of this work is to present an unusual location of BL. We report the case of an 11-year-old patient with bilateral ocular swelling that has been developing for 2 months associated with swelling spread over the entire body in the form of papules and nodules associated with a deterioration in general condition. A paraclinical assessment, namely orbital magnetic resonance imaging (MRI), revealed an enhanced tumor process in the bilateral upper-lateral eyelid orbital regions and the right supra-nasal paramedian facial soft tissues, diffuse filling of the sinuses, diffuse meningeal enhancement of the peri-brain dura; and histology of a nodule confirmed BL. We reported a case of BL with an unusual ocular and cutaneous presentation, which caused a delay in diagnosis and treatment, thus endangering the vital prognosis of our patient.

Keywords: BL, Unusual location, Child, Libreville

INTRODUCTION

Burkitt lymphoma (BL) is the most common non-Hodgkin's malignant lymphoma in African children. A distinction is classically made between BL called endemic in Africa, so-called sporadic BL in other regions of the globe, and BL linked to HIV.[1] It is the most common malignant lymphoma and the first childhood cancer in Sub-Saharan Africa and represents 40 to 70% of malignant tumors.[2] BL can invade several tissues, but the clinical presentation is often variable.[3] Abdominal location is the most common according to the literature (around 90%).[3-5] The other location is ENT (oropharynx, nasal involvement, tonsils). Bone marrow infiltration is described in almost 30% of cases, the central nervous system can be affected in 20% of cases. Cytopuncture is a simple, rapid, inexpensive and less invasive means of diagnosis with good sensitivity.[6] Well-codified care allows recovery; however, relapses can occur and complications or even death in the event of a delay in diagnosis and treatment.

CASE REPORT

We reported the case of an 11-year-old patient with left eyelid swelling (Figure 1) that has been developing for 2 months associated with swelling spread over the entire body consisting of areas of solid and raised skin (papules) and other larger swellings(nodules) (Figure 2), with alteration of the general condition. The patient consults our department 2 months after the onset of symptoms. The examination on arrival noted a right eyelid swelling occupying the entire eye and nodules or non-painful indurated papules scattered throughout the body

associated with tinnitus and hypoacusis, we also noted swollen breasts and indurated. An assessment of the extension carried out using orbital MRI revealed an enhanced tumor process in the bilateral upper-lateral eyelid orbital regions and the right supra-nasal para-median facial soft parts, a diffuse filling of the ethmoidal, maxillary and sphenoid sinuses, diffuse meningeal enhancement of the peri-encephalic dura mater (Figure 3) and histology of a nodule made it possible to confirm Burkitt's lymphoma by the presence of a cluster of atypical lymphocytes of medium size, with a little basophilic cytoplasmabundant and vacuolar, containing a round or oval hyperchromatic nucleus, a proliferation of atypical lymphocytes of hyperchromatic medium size, on a necrotic/apoptotic background with the seat of the proliferation being dermo-hypodermal of medium hyperchromatic size, on a background necrotic/apoptotic with the seat of proliferation being dermo-hypodermal (Figure 4). The evolution is marked by the death of the patient before the start of chemotherapy.

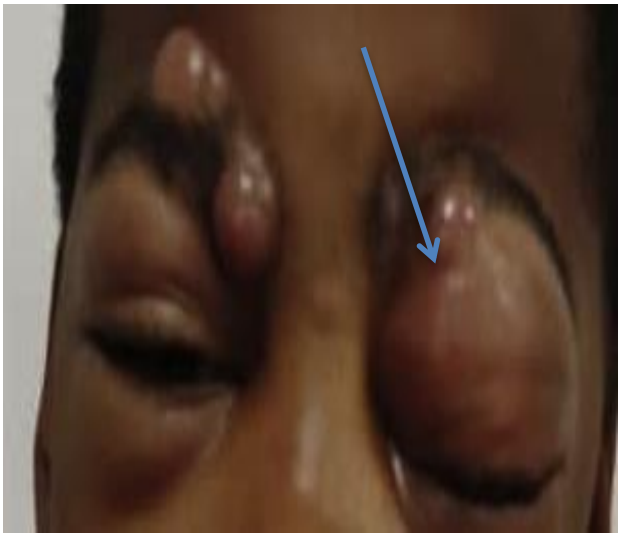

Figure 1: Left eyelid swelling and nodules.

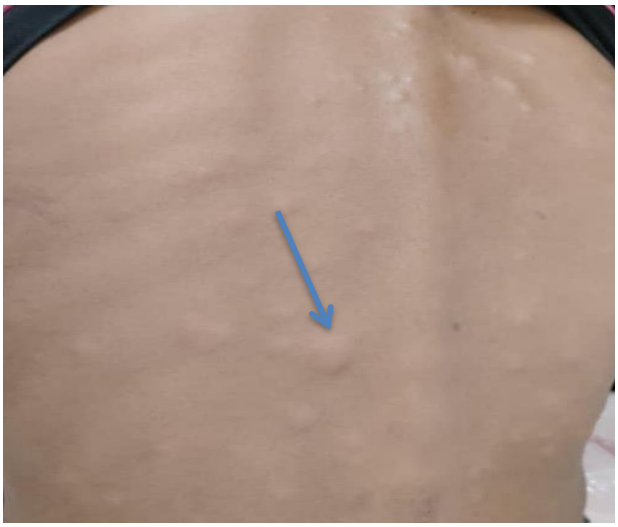

Figure 2: Disseminated skin papules and nodules.

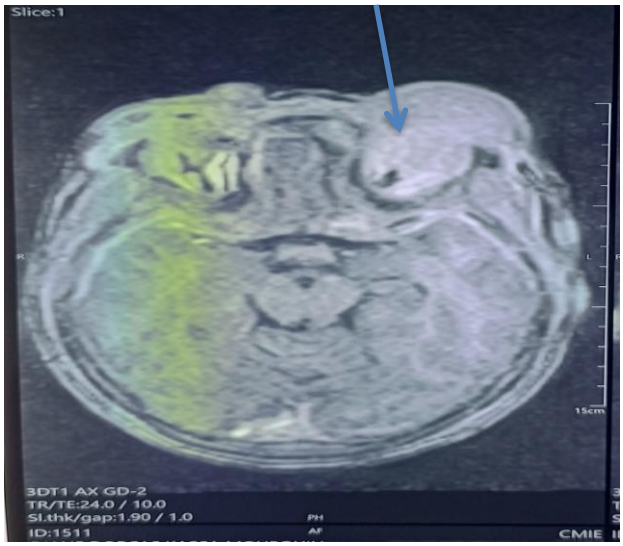

Figure 3: Orbital MRI which shows a tumor process enhanced in the eyelid orbital regions.

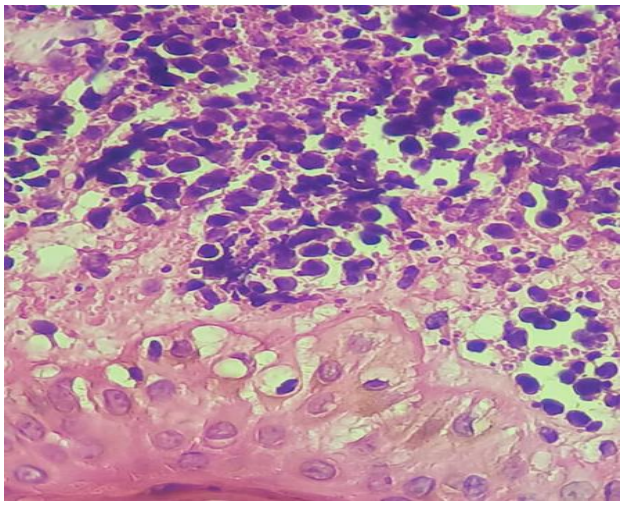

Figure 4: Clusters of atypically sized lymphocyte with little basophilic cytoplasm abundant and vacuolar containing a nucleus containing a round hyperchromatic core or oval.

DISCUSSION

BL is the most common malignant non-Hodgkin lymphoma of children. It is common between 4 and 15 years of age with a peak frequency at 10 years of age.[2] The facial tumor was considered the main clinical presentation of the endemic form before the popularization of abdominal ultrasound. The sporadic form usually found in high-income countries is abdominal.[7] BL can invade several tissues, but the clinical presentation is often variable [3] Abdominal location is the most common according to the literature (around 90%).[3-5] The other location is ENT (oropharynx, nasal involvement, tonsils). Bone marrow infiltration is described in almost 30% of cases, the central nervous system can be affected in 20% of cases. In our case it was

an unusual localization which affects several organs because we noted an ocular, cutaneous and sinus localization and breast involvement with both breasts which were increased in volume and stony consistencies thus classifying our patient at the stage III according to Murphy's classification. Like many authors from Sub-Saharan Africa, the majority (85.3%) of their patients were in Murphy stages III and IV, and the proportions reported varied from 69 to 83%.[8,9] Other unusual localizations have been noted by other authors, Blanc et al showed a rectal localization of the Burkitt in a child who presented a picture of intractable constipation associated with rectal bleeding, Zidani et al found a cerebral localization manifested by drowsiness and intracranial hypertension syndrome and cerebellar syndrome, El Fahssi presented a case of childhood Burkitt lymphoma revealed by acute intestinal intussusception.[5,10,11] The confirmatory diagnosis of BL is anatomopathological and cytogenetic. Cytology is a simple and rapid technique that allows an initial diagnosis to be obtained in just a few minutes. The biopsy sample of the tumor mass is the test which allows diagnostic confirmation.[12] Burkitt lymphoma is the fastest growing human tumor whose pathology reveals a high mitotic rate, monoclonal proliferation of B lymphocytes, and a "starry sky" pattern of benign macrophages that have engulfed apoptotic malignant lymphocytes.[12] In this case we do not have this starry sky appearance but we have the presence of a cluster of atypical lymphocytes of medium size, with a scanty and vacuolar basophilic cytoplasm, containing a hyper chromatic round or oval nucleus, a proliferation of atypical lymphocytes of hyperchromatic medium size, on a necrotic/apoptotic background with the site of dermo-hypodermal proliferation hyperchromatic medium size. The extension was rapid in the patient with invasion with diffuse filling of the ethmoid, maxillary and sphenoid sinuses, diffuse meningeal enhancement of the peri-encephalic dura mater, we also noted rapidly progressive indurated bilateral breast masses judging from rapid evolution as found in the literature.

CONCLUSION

Burkitt's lymphoma in African children usually manifests itself with abdominal and maxillofacial involvement, however it must be considered in the face of ocular and skin involvement, these atypical forms can cause the diagnosis to be erroneous as in our clinical case. Delayed diagnosis and delay in treatment are life-threatening.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Larissa MT, Eurudys MMA, Edmee MMN, Claire LF, Raissa KM, Mariela LMA, et al. Burkitt lymphoma: an unusual location in children, report of a case. Int J Contemp Pediatr 2025;12:318-20.