

Burkitt's Lymphoma Involving the Nasosinusal Area: Report of a Case and Review of the Literature

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Abstract

Case Report

Burkitt's lymphoma is a high-grade malignant non-Hodgkin's lymphoma, it is an undifferentiated B-cell lymphoma characterized by a distinctive histological pattern commonly referred to as the "starry sky" appearance that manifests in two clinical forms: the African or endemic form, which primarily affects children in Central Africa, and the American or sporadic form, initially described in North America. His naso-sinusal presentation is relatively rare, the diagnosis relies mainly on biopsy with histological examination. Treatment involves polychemotherapy and adjunctive therapies. In this report, a rare case of 16 years old boy of Burkitt lymphoma that originated in the sphenoid sinus extended to the nasal cavities, we will discuss the diagnosis of Burkitt's lymphoma of the naso-sinusal area in relation to the literature on this subject.

Keywords: Burkitt's lymphoma, starry sky, polychemotherapy and adjunctive therapies, naso-sinusal presentation.

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INTRODUCTION

Burkitt's lymphoma was initially identified in 1958, in an African child presenting with a malignant growth in the mandible. Recognized as a highly aggressive B-cell non-Hodgkin's lymphoma. Around 10% to 34% of all cases of non-Hodgkin's lymphomas originate from sites outside of the lymph nodes [1, 2]. Among these cases, nasal or paranasal lymphomas represent fewer than 3% of all malignant lymphomas originating from extra nodal sites [3]. Sinus-related primary diseases are rare, with occurrences in the sphenoid sinus being even rarer.

CASE REPORT

We present a case of a 16-year-old patient who presented with frequent left-sided headaches. He had epistaxis and progressive nasal obstruction. During examination, it was found that a bulging mass completely obstructed the right nostril. The mass exhibited a necrotic surface and appeared pinkish in color. CT scan demonstrated opacity of the posterior ethmoid and sphenoid sinuses filling the Rosenmuller's fossae extended to the right nasal cavity, with evidence of erosion of the anterior wall of the sphenoid (Figure 1).; Magnetic Resonance Imaging (MRI) demonstrated a homogenous soft-tissue lesion occupying the sphenoid

sinus and invading the right cavernous sinus (Figure 2). The patient underwent trans sphenoidal biopsy and the lesion was histologically diagnosed as non-Hodgkin's lymphoma, diffuse large B-cell type.

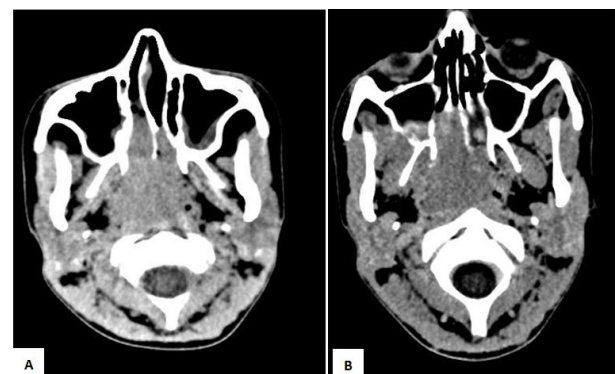


Fig 1: CT parenchymal window without injection and (A) parenchymal window with injection; (B) showed homogenous mass of soft tissue intensity enhanced after injection in the right nasal cavity, involving the posterior ethmoid and sphenoid sinuses filling the Rosenmuller's fossae extended to the right nasal cavity, with evidence of erosion of the anterior wall of the sphenoid

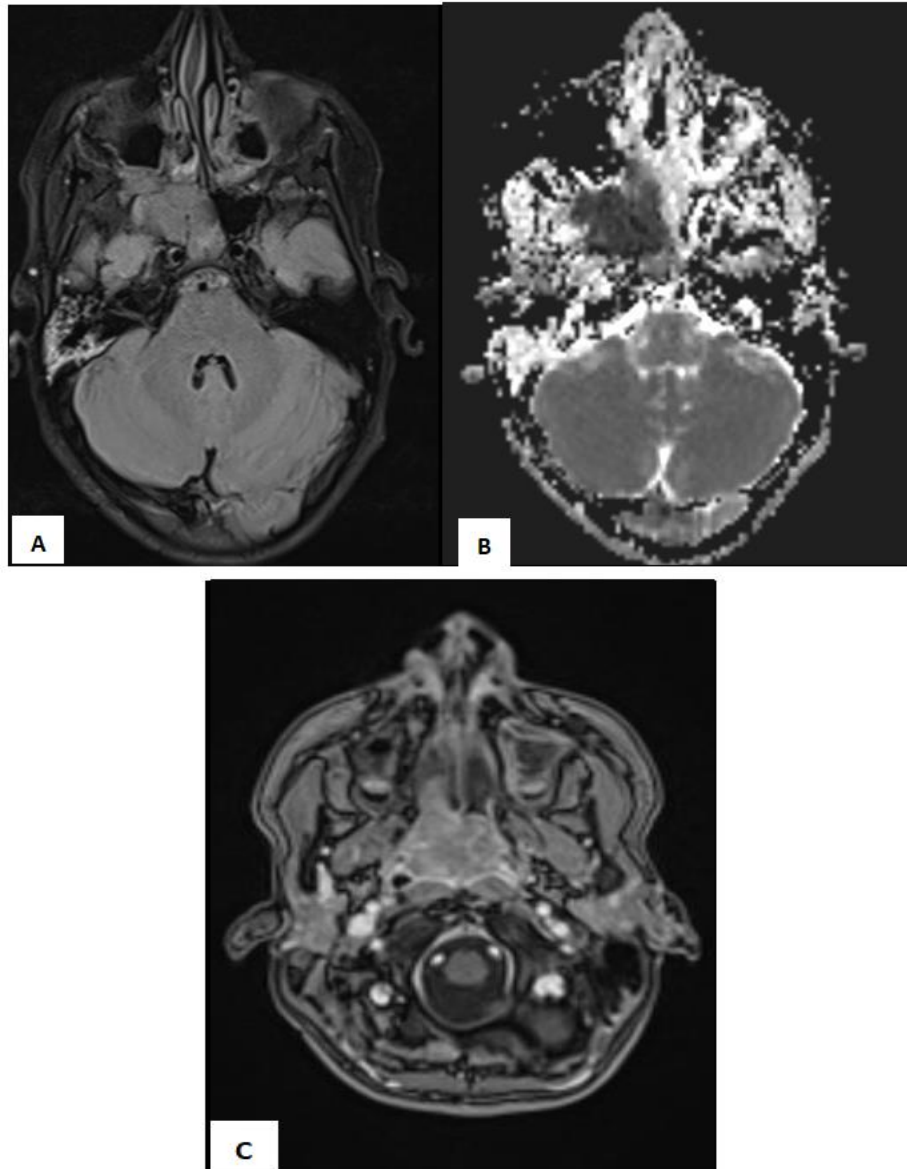


Figure 2: (A) Axial Flair; (B) Axial Diffusion ADC; (C) Axial Gadolinium-enhanced MRI demonstrated a homogenous soft-tissue lesion occupying the sphenoid sinus and invading the right cavernous sinus

DISCUSSION

Lymphoma is a malignant neoplasm of the lymphoreticular system arising from B cells or T cells; Burkitt's lymphoma accounts for roughly 40% of all cases of childhood non-Hodgkin's lymphoma and approximately 3% to 4% of all childhood malignancies diagnosed annually [4].

The International Working Formulation classified non-Hodgkin's lymphoma into low, intermediate, or high grades, which are indicative of prognosis. While low and intermediate grades of NHL are more common in adults, over 90% of children diagnosed with non-Hodgkin's lymphoma have high-grade tumors [6].

Based on this finding, Burkitt's lymphoma was categorized as a subtype of non-Hodgkin's lymphoma,

with further classification into either the endemic (African) form or the non-endemic (American) form

Burkitt's lymphoma is associated with characteristic chromosomal alterations. The translocation of the "c-myc" gene is often induced by the Epstein-Barr viral oncogene. The most common location of this translocation in the genome is from the q24 region of chromosome 8 to the q32 region of chromosome 14 {t(8;14) (q24;q32)}. Other variants have been described, such as from chromosome 8 to chromosome 2 {t(8;2) (q24;p12)} or to chromosome 22 {t(8;22)(q24;q11)} [10, 11].

Often, Burkitt's lymphoma manifests with a painless mass as its sole symptom. When located in the abdomen, it can result in nausea, vomiting, and pain, typically due to intestinal obstruction. In cases where the

tumor is in the head and neck area, patients may experience symptoms such as nasal congestion, rhinorrhée, facial swelling, enlarged tonsils, and swelling of lymph nodes in the neck. This issue can be mistakenly identified as nasal polyps or chronic sinusitis. Consequently, in the initial phases, Burkitt's lymphoma might be inaccurately managed as an upper airway infection or another inflammatory condition in the study conducted by Cuadra-Garcia *et al.*, at the Massachusetts General Hospital, 58 cases of sinonasal lymphoma were analyzed. Nasal obstruction emerged as the most prevalent symptom, despite only one instance of Burkitt's lymphoma being reported in the study.

While clinical manifestations of Burkitt's lymphoma may resemble those of allergic rhinitis and acute rhinitis, the tumor exhibits rapid growth and spreads through bloodborne dissemination.

Radiological imaging plays a crucial role in various aspects, such as evaluating tumor spread, identifying bone damage, detecting mucosal thickening, and determining the optimal biopsy site and approach. While computed tomography (CT) excels in displaying intricate bone structures, magnetic resonance imaging (MRI) is effective in assessing most areas of bone destruction and offers several additional benefits. MRI is particularly adept at distinguishing between tumors, mucosal thickening, or trapped sinus secretions [5].

If feasible, the primary lesion should be directly biopsied, possibly utilizing an endoscope. Additionally, lumbar puncture, bone marrow aspiration, chromosomal studies, and a comprehensive laboratory profile should be conducted, including assessing LDH levels and EBV titers.

Other studies, including our case, have been verified as diffuse large B-cell lymphomas. The primary site of the lymphoma, along with its histologic grade, T/B phenotype, and clinical stage, could serve as significant prognostic factors in primary non-Hodgkin's lymphoma of the sinonasal cavities

Reviewing multiple reports indicates that the most favorable treatment outcomes are observed with the CHOP (cyclophosphamide, adriamycin, vincristine (oncovin), and prednisone) regimen, administered at three-week intervals. Rituximab, a therapeutic antibody targeting the CD20 surface antigen commonly found in lymphoma cells, is often combined with CHOP to enhance treatment response in lymphomas expressing the CD20 antigen, as observed in our case. Following chemotherapy, loco-regional radiotherapy is frequently administered at a dosage of 30 to 40 Gy [8, 9].

Non-Hodgkin's lymphomas are frequently treated with, and respond to, a combination of chemotherapy and radiotherapy

Non-Hodgkin's lymphomas affecting the sphenoid sinus are seldom contemplated in the differential diagnosis of a mass in the sphenoid sinus without accompanying systemic symptoms like weight loss, night sweats, or fever. This is likely attributed to the extremely infrequent occurrence of this disease [7].

CONCLUSION

Naso-sinusal tumors are very rarely indicative of Burkitt's lymphoma. Nevertheless, it is crucial for the otolaryngologist to consider Burkitt's lymphoma and its differential diagnoses. It primarily affects younger children. Prompt initiation of chemotherapy following early diagnosis, prior to dissemination, is crucial for a favorable prognosis. Hence, maintaining awareness of this rare disease and conducting early endoscopic biopsies are even more imperative when managing pediatric patients

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