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Case Report

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Primary Burkitt Lymphoma of the Maxillary Sinus in an Immunocompetent Adult: A Case Report

Konstantinos Chaidas[1], Artemis Zarkadi[1], Nikolaos Tepelenis[2], Sotirios Papouliakos[3]

[1]Democritus University of Thrace School of Medicine, University Hospital of Alexandroupolis, Department of Ear, Nose, and Throat, Alexandroupolis, Greece
[2]General Hospital of Athens "G. Gennimatas", Department of Pathology, Athens, Greece
[3]General Hospital of Athens "G. Gennimatas", Department of Ear, Nose, and Throat, Athens, Greece

Abstract

Primary Burkitt lymphoma (BL) of the maxillary sinus is rare, particularly in immunocompetent adults, and only a few cases have been reported in the literature. Its clinical presentation may mimic benign conditions, such as chronic rhinosinusitis or neuropathic facial pain, potentially resulting in diagnostic delay. We present the case of a 38-year-old male with a five-month history of isolated left-sided facial pain. Comprehensive clinical, radiological, and histopathological evaluation established the diagnosis of BL, and the patient subsequently received systemic chemotherapy. This case highlights the importance of considering BL in the differential diagnosis of persistent or atypical sinonasal symptoms, particularly those unresponsive to empirical treatment. Prompt biopsy and histopathological and immunohistochemical evaluation are essential for establishing the diagnosis and initiating appropriate treatment without delay.

Keywords: Burkitt lymphoma, paranasal sinus neoplasms, maxillary sinus, immunocompetence, rhinology, case report

Introduction

Burkitt lymphoma (BL) is a non-cleaved B-cell non-Hodgkin lymphoma that accounts for less than 5% of adult lymphoma cases (1,2). Sinonasal lymphomas are generally uncommon, representing less than 1% of all malignancies in the head and neck region. Although BL predominantly affects children and immunocompromised individuals, its occurrence in immunocompetent adults is uncommon, and involvement of the sinonasal region is exceedingly rare. The association between viral infections and lymphoid malignancies is well established, with Epstein-Barr virus (EBV) and human immunodeficiency virus (HIV) recognized as key etiological factors in BL. Characterized by an extraordinary proliferation rate, BL typically exhibits rapid local expansion and early dissemination, often leading to swift clinical deterioration and facial deformities in cases involving the head and neck region (3). Given its rarity confined to the paranasal sinuses, particularly in non-immunocompromised adults, we present an extremely rare case of BL originating from and limited to the left maxillary sinus in an immunocompetent adult, highlighting the diagnostic challenges and the need for heightened clinical suspicion.

ORCID IDs of the authors:
K.C. 0000-0002-4428-7248
A.Z. 0009-0008-9884-9016
N.T.
S.P. 0000-0001-6164-7279

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Corresponding Author:
Konstantinos Chaidas, MD;
konchaidas@gmail.com

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Case Presentation

A 38-year-old Caucasian male presented to the otorhinolaryngology outpatient clinic with a five-month history of progressively worsening left-sided facial pain associated with numbness over the left cheek. The patient reported no other symptoms. There was no relevant medical or surgical history. Clinical examination did not reveal any abnormality such as nasal obstruction or cervical lymphadenopathy. There was normal function of all cranial nerves.

A computed tomography (CT) scan of the paranasal sinuses revealed a soft-tissue mass within the left maxillary sinus with erosion of the posterior wall and possible compression of the infraorbital nerve (Figure 1). A subsequent magnetic resonance imaging (MRI) demonstrated similar findings, with a soft-tissue mass partially occupying the left maxillary sinus and extending posteriorly into the adjacent pterygopalatine fossa.

Considering the above findings, the patient underwent left-sided endoscopic middle meatal antrostomy and diagnostic tumor biopsy. Intraoperatively, a white, round, and friable tumor within the maxillary sinus was seen (Figure 2).

Histopathological evaluation demonstrated the presence of a monomorphic infiltrative population of medium-to-large-sized lymphoid cells exhibiting a classic "starry-sky" pattern, characterized by numerous tangible-body macrophages scattered amongst the highly proliferative neoplastic B-lymphocytes (Figure 3). Immunohistochemical staining yielded the following profile: the neoplastic cells were positive for Pan-B markers (L26/CD20, CD10, CD79a, and Bcl-6) and negative for CD3, CD5, CD23, CD30, and BCL-2. A notably high Ki-67 proliferative index of 98% was documented, confirming the diagnosis of BL. Serological testing for EBV, HIV, and hepatitis B and C was negative. Further imaging to determine disease staging, including contrast-enhanced CT of the chest, abdomen, and pelvis, as well as a bone scan, revealed no evidence of metastatic disease (Ann Arbor Stage IE).

The patient was promptly referred to the Haematology department and initiated on an intensive, short-course chemotherapy regimen following the R-CODOX-M/IVAC protocol, comprising rituximab in combination with cyclophosphamide, doxorubicin, vincristine, methotrexate/ifosfamide, etoposide, and cytarabine. The patient completed four cycles of therapy, without major complications. A follow-up CT scan performed two months after treatment demonstrating complete remission. A positron-emission tomography (PET)/CT scan at one-year follow-up was also unremarkable. At the two-year follow-up, the patient remains clinically and radiologically free of disease recurrence.

Informed consent was obtained from the patient.

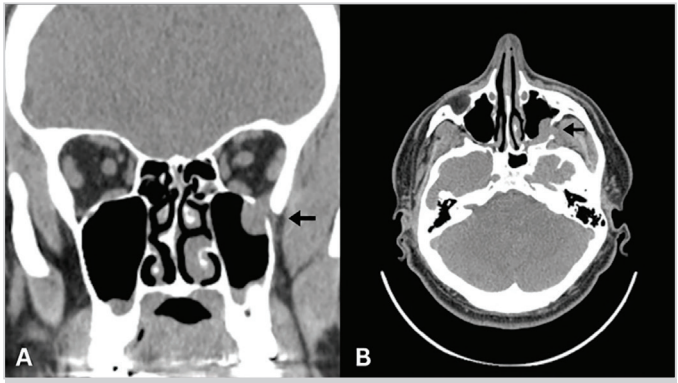

Figure 1. Coronal (A) and axial (B) view of a computed tomography showing partial opacification of the left maxillary sinus by a soft-tissue mass originating from the superior and lateral walls. The mass exhibited locally aggressive characteristics causing erosion of the posterior sinus wall and possible compression of the infraorbital nerve

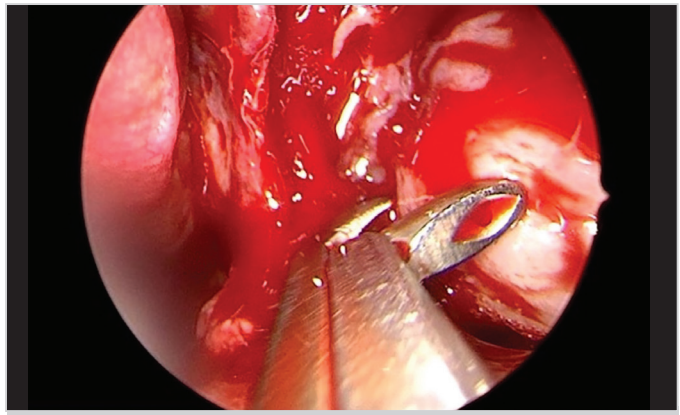

Figure 2. Intraoperative imaging of the left maxillary sinus showing the presence of a white, friable lesion

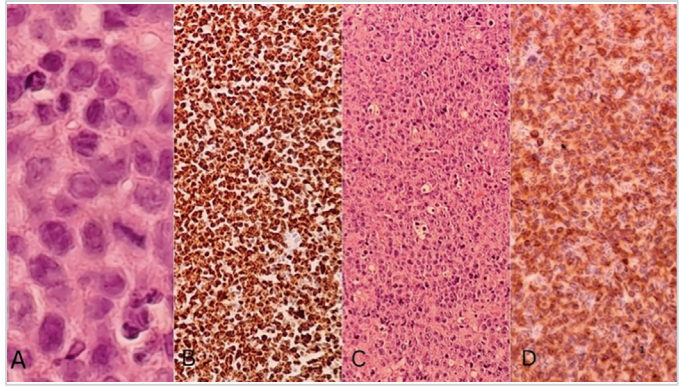

Figure 3. A. There is a lymphoid population of Burkitt-like cells featuring an atypical mitosis; B. BCL2 immunostain with dim expression; C. The proliferation rate of the lymphoma is almost 100% with the Ki67 immunostain; D. There exists an evident Starry sky pattern among the lymphoid cells

Discussion

BL was first described in Central Africa by Denis Burkitt in 1958 and is recognized as one of the most aggressive B-cell non-Hodgkin lymphomas. Although BL was historically classified into three clinical subtypes (endemic, sporadic, and immunodeficiency-associated), the recent 5th edition of the World Health Organization classification employed a molecular classification based on EBV status, distinguishing EBV-positive from EBV-negative BL (4).

In the head and neck region, BL most commonly presents as cervical lymphadenopathy (3); however, primary extranodal involvement of the paranasal sinuses is extremely rare, especially in the absence of immunosuppression. To the best of our knowledge, only a few cases of BL primarily affecting the maxillary sinus have been reported in the literature until now (3,5).

The diagnostic challenge of BL in otorhinolaryngology resides in its non-specific clinical presentation. Symptoms such as nasal obstruction, facial swelling, or headache frequently mimic benign conditions such as chronic rhinosinusitis or nasal polyposis, often resulting in diagnostic and therapeutic delay-an issue of critical importance given the tumor's exceptionally rapid doubling time of 24-48 hours (5,6). The present case illustrates a particularly atypical presentation, wherein neuropathic facial pain caused by infraorbital nerve encasement was the sole symptom. Such a subtle manifestation, particularly in the absence of typical sinonasal complaints, highlights the importance of considering BL in the assessment of rapidly progressive or atypical sinonasal lesions.

Therefore, diagnostic evaluation must be thorough and expeditious (7). Cross-sectional imaging utilizing contrast-enhanced CT and MRI is vital for delineating local disease extent, while PET-CT is recommended for systemic staging. Laboratory investigations should include a complete blood count, serum lactate dehydrogenase (LDH), renal and hepatic function tests, tumor lysis parameters, and viral serology for HIV, EBV, and hepatitis B and C. A surgical biopsy remains the preferred diagnostic method, providing tissue for histopathological and immunohistochemical analysis. Lumbar puncture is essential to exclude central nervous system involvement, with bone marrow assessment reserved for cases demonstrating clinical or laboratory indications of leukemic transformation.

Management of BL requires intensive, short-course, multi-agent chemotherapy, frequently including rituximab, an anti-CD20 monoclonal antibody. The selection of treatment protocols depends on tumor burden, serum LDH levels, performance status, and disease stage (1). The currently used chemotherapy regimens in patients with disease limited to the head and neck have yielded 90% long-term survival (3).

Conclusion

This case highlights the significance of sustaining a heightened level of suspicion for BL in patients presenting with persistent, unilateral sinonasal symptoms that do not respond to conventional therapy. Early imaging, prompt biopsy, and multidisciplinary management are crucial for facilitating a timely diagnosis and curative intervention, even among immunocompetent individuals who are HIV- and EBV-negative, considering the tumor's highly aggressive characteristics.

Ethics

Informed Consent: Informed consent was obtained from the patient.

Footnotes

Authorship Contributions

Surgical and Medical Practices: K.C., N.T., S.P., Concept: K.C., S.P., Design: K.C., A.Z., S.P., Data Collection and/or Processing: K.C., A.Z., N.T., S.P., Analysis or Interpretation: K.C., A.Z., N.T., S.P., Literature Search: K.C., A.Z., Writing: K.C., A.Z., N.T., S.P.

Conflict of Interest: The authors declare that they have no conflict of interest.

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Main Points

- Primary Burkitt lymphoma of the maxillary sinus is a quite rare entity in immunocompetent adults, yet it should be included in the differential diagnosis of rapidly progressive or atypical sinonasal lesions.
- Early recognition supported by advanced imaging and definitive histopathological and immunohistochemical confirmation is crucial for avoiding diagnostic delay.
- Short-course, multi-agent chemotherapy regimens have demonstrated favorable outcomes and represent the cornerstone of effective management, offering a promising prognosis when initiated early.

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