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Case Report

Carotid space Burkitt lymphoma presenting with headache: A case report ☆,☆☆

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ABSTRACT

Burkitt's lymphoma presents with diverse clinical manifestations, including rare extranodal occurrences. This report documents a unique case of primary Burkitt's lymphoma located in the carotid space, an infrequent presentation with significant diagnostic implications. A 25-year-old male presented with the sudden onset severe headache and nausea. Imaging revealed an infiltrative lesion encompassing the right carotid sheath, extending into the superior retropharynx and nasopharynx. Diffusion-weighted imaging (DWI) and apparent diffusion coefficient (ADC) mapping demonstrated significant restriction. A core needle biopsy confirmed Burkitt's lymphoma diagnosis. Following the initiation of chemotherapy, follow-up imaging showed significant tumor regression. This case highlights the diagnostic challenges associated with carotid space tumors and underscores the pivotal role of imaging modalities such as computed tomography (CT) and magnetic resonance imaging (MRI) in their comprehensive evaluation. Multidisciplinary management, including aggressive chemotherapy, is imperative for achieving favorable outcomes in rare extranodal lymphomas like Burkitt's. Further research is warranted to refine diagnostic and therapeutic approaches for managing such complex cases.

Introduction

Burkitt's lymphoma is characterized by its highly aggressive nature, presenting in three histologically and phenotypically similar yet clinically diverse forms: endemic, nonendemic (sporadic), and immunodeficiency-associated [1,2]. The endemic variant is primarily associated with the Epstein-Barr virus (EBV) detected in nearly all cases [3]. This subtype

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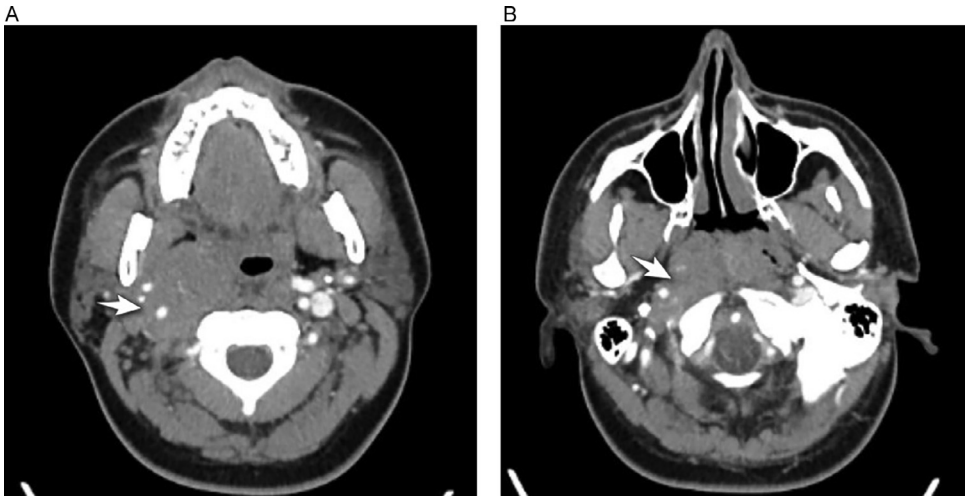

Fig. 1 – Contrast enhanced CT scan. (A) A slightly enhancing mass is observed in the right carotid space, causing splaying of the internal and external carotid arteries with parapharyngeal space obliteration, (B) The mass extends into the nasopharynx and superior retropharyngeal space.

predominantly affects children and shows a distinct male predilection.

The mandible, facial bones, kidneys, gastrointestinal tract, ovaries, breast, and other extranodal sites are commonly involved. Although the neck is less frequently affected compared to other regions, Burkitt's lymphoma has been documented in the sinonasal and orbital regions, as well as in less typical sites such as the nasopharynx, postnasal space, parapharyngeal space, mastoid, and oral cavity [4–7].

Extranodal Burkitt's lymphoma involving the carotid space has been reported in only a limited number of individual case studies. Here, we present a rare case of primary Burkitt's lymphoma occurring in the carotid space of a young male.

Patient information

A 25-year-old man presented with a sudden, severe headache radiating to the right ear, accompanied by nausea. He denied any history of trauma, violent sneezing, cervical massage, or abnormal movements. The patient had no significant medical or surgical history, nor did he report any relevant family history. His vital signs were within normal limits. On physical examination, the right tonsil was observed to be have shifted to the left side. An initial diagnosis of a peritonsillar abscess prompted a referral to the radiology department for a CT scan.

Neck CT and MRI revealed the presence of an infiltrative lesion within the right carotid sheath, extending superiorly into the retropharynx and nasopharynx, with visible extension to the jugular foramen (Figs. 1 and 2). The abnormal tissue in the right carotid space exhibited hypoattenuation on CT, intermediate signal intensity on T1-weighted MRI, and high signal intensity on T2-weighted MRI, with slight homogeneous contrast enhancement. Significant restriction (with 458 mm2/s ADC value) was noted on diffusion-weighted images. No cervical lymphadenopathy was identified. The initial differential diagnosis included idiopathic inflammation, such as IgG4-related disease, and lymphoma.

A subsequent core needle biopsy of the right parapharyngeal mass confirmed Burkitt lymphoma, with positive immunohistochemical staining for CD20, CD10, BCL-6, Ki-67, and PAX5. A brain MRI without gadolinium was unremarkable. Laboratory investigations revealed no abnormalities.

Chemotherapy, including the R-CODOX-M/IVAC (Magrath) regimen, was initiated. Follow-up MRI showed a partial response, with significant tumor regression (Fig. 3).

Discussion

Understanding the intricate anatomy of the carotid space and mastering the nuances of differentiating between various pathologies is crucial for making precise diagnoses [8]. Anatomopathological studies have identified fibrofatty tissue and scattered neutrophils as predominant components of the carotid sheath. Nonetheless, in some instances, lymphoid aggregates primarily composed of B-cells have been observed [9]. This suggests the possibility of lymphomas, including Burkitt's lymphoma, originating from or affecting the carotid arteries.

Burkitt's lymphoma presenting as a tumor in the carotid space is exceedingly rare and often poses diagnostic challenges due to its atypical location and nonspecific clinical manifestations [10]. In the differential diagnosis of a carotid space mass, key considerations include idiopathic inflammation, such as IgG4-related disease, and lymphoma, including Burkitt's lymphoma. IgG4-related disease typically presents with a chronic, painless swelling. It can involve multiple organs, often demonstrating diffuse enhancement and homogeneity on imaging, along with characteristic storiform fibrosis and a dense lymphoplasmacytic infiltrate on histopathology. In contrast, Burkitt's lymphoma is more aggressive, often

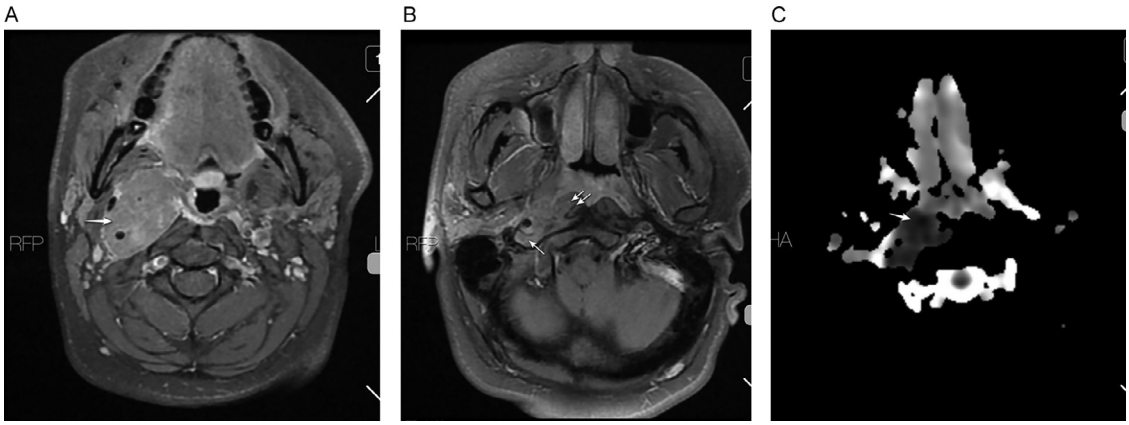

Fig. 2 – MRI findings. (A) An infiltrative mass is identified within the right carotid sheath, infiltrating into the superior retropharynx and nasopharynx, (B) Superior extension is visible to the level of the jugular foramen. Involvement of the longus capitis muscle at same level depicted (C) Significantly restricted diffusion is evident in axial ADC map.

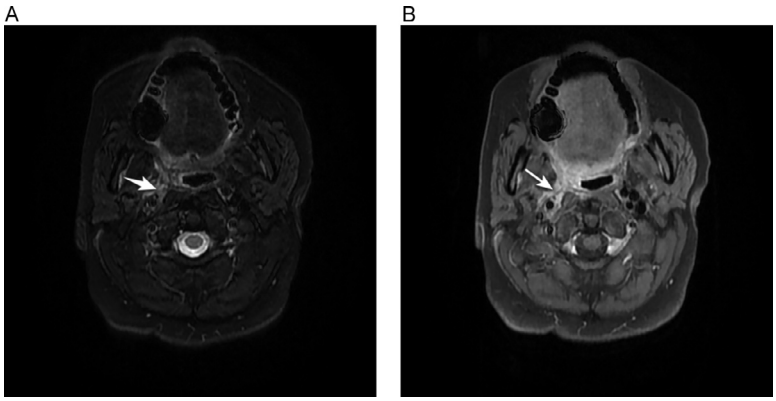

Fig. 3 – Follow-up MRI. (A) Axial T2 sequences shows slight residue of tumor. (B) Axial post contrast images shows significant regression of tumor.

presenting with rapidly growing masses and systemic symptoms. Imaging features of Burkitt's lymphoma typically include an infiltrative mass with significantly restricted diffusion on DWI/ADC images, indicative of high cellularity. Unlike IgG4-related disease, which rarely causes significant vascular involvement, Burkitt's lymphoma may extend into adjacent structures but generally does not cause luminal narrowing of arterial vessels. Additionally, histopathological examination is crucial, as Burkitt's lymphoma is characterized by a high proliferation index (Ki-67 nearly 100%) and specific immunohistochemical markers, such as CD20, CD10, and BCL-6, which are absent in IgG4-related disease. These distinguishing features are critical for accurate diagnosis and appropriate treatment planning [11–14].

In our case report, imaging modalities, including neck CT and MRI, provided valuable insights into the tumor's vascular involvement and characteristics. These imaging techniques are essential for the comprehensive evaluation and differential diagnosis of carotid space lesions.

Lymphomatous involvement on CT scans can exhibit diverse enhancement levels and may extend extensively. Lymph node conglomerations may infiltrate deep neck spaces, surrounding vital structures. Notably, lymphoma typically does not lead to luminal narrowing of arterial blood vessels, which aids disease differentiation [6]. The complementary information obtained from CT and MRI helped delineate the anatomy and guided treatment planning. Contrast-enhanced CT and CTA provide detailed visualization of vascular structures, enabling a thorough assessment of tumor vascularity and potential carotid artery encasement. In our case, a contrast-enhanced CT scan revealed a slightly hyper-enhancing mass with splaying of the internal and external carotid arteries, along with extension into the nasopharynx and superior retropharyngeal space.

MRI with gadolinium provides high-resolution imaging of the carotid arteries and surrounding structures, aiding in evaluating tumor extent and vascular involvement. In our case, MRI demonstrated an infiltrative mass within the right carotid sheath, extending into the superior retropharynx and nasopharynx, providing additional support for diagnosing a carotid space tumor. Significant restricted diffusion was observed in DWI/ADC images. This finding, combined with clinical and histopathological correlation, led to the diagnosis of Burkitt's lymphoma.

The treatment of carotid space tumors often requires a multidisciplinary approach involving surgery, chemotherapy, and radiotherapy tailored to tumor characteristics and patient factors. In Burkitt's lymphoma, aggressive chemotherapy regimens like R-CODOX-M/IVAC have demonstrated favorable outcomes in specific cases, emphasizing the significance of precise diagnosis and timely treatment initiation.

In conclusion, our case report highlights the importance of multimodal imaging, including CT and MRI, in characterizing carotid space tumors, especially in rare entities such as Burkitt's lymphoma. Further research and collaboration are warranted to refine diagnostic algorithms and therapeutic strategies for effectively managing these complex cases.

Patient consent

We wish to confirm that the patient information in the manuscript titled "Carotid Space Burkitt Lymphoma Presenting with Headache: A case report" has been included with the patient's informed consent. The patient has authorized the use of their case details in the printed, online, and licensed editions of a medical journal. Additionally, the patient has been given the chance to review the general description of the manuscript's content and all the photographs from the investigations that are being submitted for publication.

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