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Case Report

Nasopharyngeal Burkitt lymphoma masquerading as adenoid hypertrophy in an adult: a diagnostic challenge

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ABSTRACT

This case report presents the diagnostic challenges encountered in identifying nasopharyngeal Burkitt lymphoma (BL) in an 18- year-old male initially misdiagnosed with adenoid hypertrophy. BL, a rare and aggressive B-cell non-Hodgkin's lymphoma (NHL), typically manifests in cervical lymphnodes or facial bones, making nasopharyngeal involvement unusual. The patient's symptoms progressed from nasal blockage to oropharyngeal dysphagia and neck swelling, prompting further investigation. Radiological assessments, including a CT scan, were inconclusive, leading to a diagnosis through excisional biopsy and immunohistochemistry, confirming BL with CD20, CD10, and Bcl-6 expression. Successful treatment involved six cycles of chemotherapy, resulting in complete remission. This case underscores the importance of considering rare malignancies in atypical presentations and highlights the role of comprehensive diagnostic approaches in achieving accurate and timely management.

Keywords: Nasopharyngeal BL, Adenoid hypertrophy, Misdiagnosis, Diagnostic challenge, Immunohistochemistry, CT scan, Complete remission

INTRODUCTION

Adenoid hypertrophy is rare in adults, and many cases are misdiagnosed due to insufficient examination of the nasopharynx by indirect posterior rhinoscopy.[1] Chronic inflammation is linked to the presence of lymphoid hyperplasia in the adult nasopharynx, including the persistence of childhood adenoids. Adenoid tissue that has regressed may begin to proliferate again in response to irritants and infections.[1] A very affects the cervical lymph nodes or facial bones in the head and neck.[2,3] BL is classified as a malignant tumor of mature, low-frequency B-cell lymphocytes and is a subtype of NHL. It constitutes 1-3% of NHL cases and is distinguished by an aggressive tumor. [4] Since the otolaryngologic symptoms of BL are frequently non-specific, there is a risk that a diagnosis will be delayed. In the present report, we present a case of an 18-year-old who had BL of the nasopharynx misdiagnosed as adenoid hypertrophy.

CASE REPORT

An 18-year-old male patient reported to the ENT OPD with complaints of headache and nasal blockage for 4 months. On examination, the paranasal sinuses (PNS), neck and throat were within normal limits. Initial nasal endoscopy revealed a pinkish mass in the nasopharynx, which was determined to be adenoid hypertrophy (Figure 1). The patient reported to the OPD a week later, when the patient's nasal blockage worsened with oropharyngeal

dysphagia and the rapid onset of bilateral swelling in the neck. On clinical inspection, the oropharynx showed swelling posterior to the soft palate. Lymph node examination showed that level 5 lymph nodes were enlarged bilaterally. Diagnostic nasal endoscopy showed a massive mass completely obstructing choana (Figure 2). In suspicion of a nasopharyngeal malignancy, the patient was advised to undergo a CECT of neck with PNS, although inconclusive, which led to diagnosis of lymphoma/nasopharyngeal malignancy (Figure 3). Patient underwent excisional biopsy of lymph node and nasopharyngeal mass under general anaesthesia. HPE-suggestive of poorly differentiated malignant neoplasms (Figure 4). Immunohistochemistry validated CD20, CD10 and Bcl-6 expression in lymphoid neoplasia. Ki-67 proliferation rate was highest (100%), supporting diagnosis of BL (Figure 5). IHC was not positive for CK, CD3, bcl-2, MUM-1, C-myc, or Tdt (Figure 6). A PET scan was done before starting patient on chemotherapy (Figure 7). After being referred to department of medical oncology, patient received six rounds of intravenous rituximab, etoposide, vincristine, adriamycin, and endoxan. Following chemotherapy, patient recovered well. Patient had no mass lesion on endoscopy, or on the second PET scan four months after therapy.

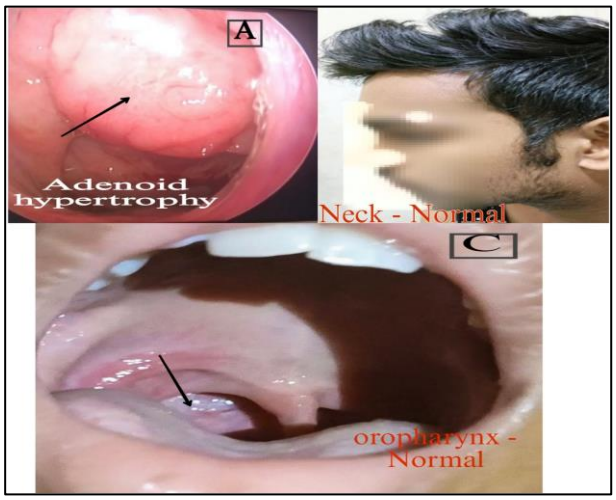

Figure 1 (A-C): Diagnostic nasal endoscopy-showing adenoid hypertrophy.

Figure 2 (A-C): Diagnostic nasal endoscopy-mass completely obstructing choana, oropharyngeal swelling posterior to soft palate and bilateral neck swelling.

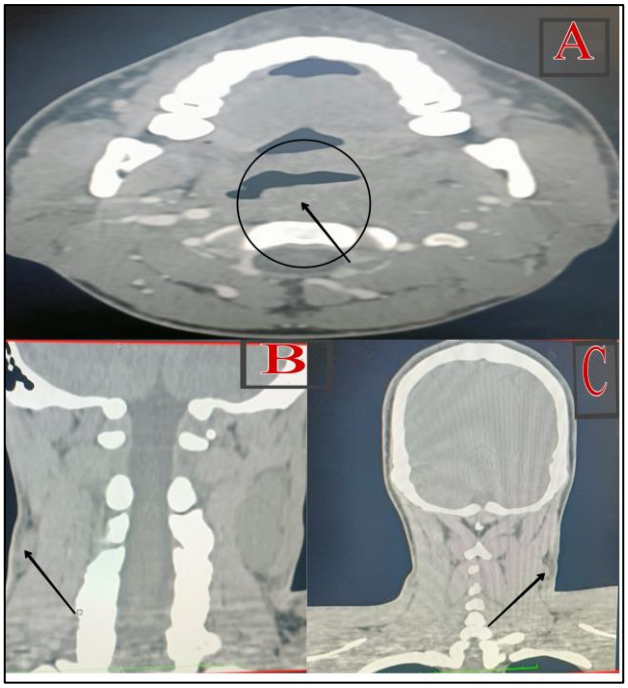

Figure 3 (A-C): Contrast enhanced CT of neck with PNS of homogeneous mass lesion epi-centered in pharyngeal mucosal space with retropharyngeal and bilateral cervical lymphadenopathy.

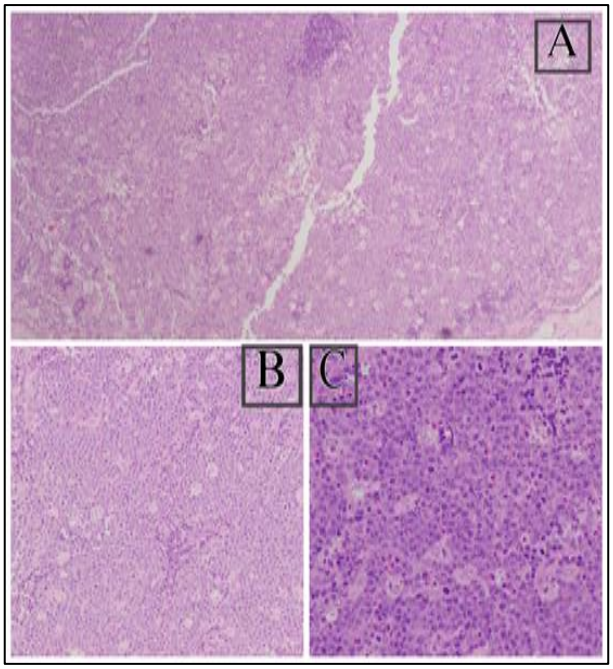

Figure 4 (A-C): Histopathological examination-Right posterior triangle lymph node showing effaced architecture and infiltration by monotonous cells with thin capsule. Starry sky pattern because of tingible body macrophages and biopsy from Nasopharyngeal mass- uniform, monotonous sheets of medium sized lymphoid cells, round nuclei of individual cells, increased N:C ratio, prominent nucleoli with scanty cytoplasmic rim.

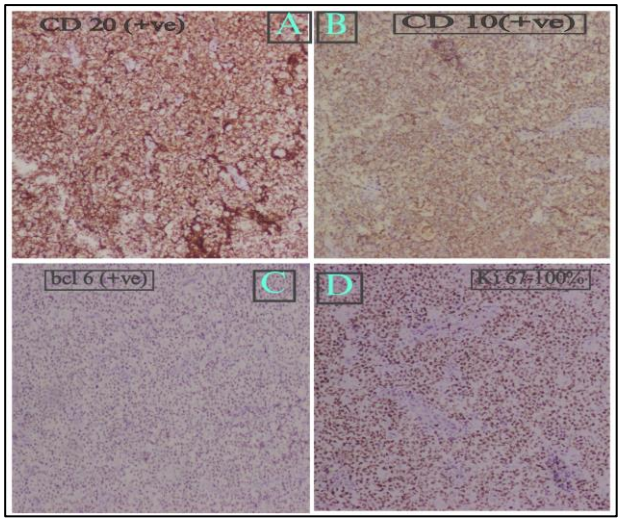

Figure 5 (A-D): Immunohistochemistry-neoplastic lymphoid cells express CD20, CD10, bcl6, Ki67 proliferation index-100%.

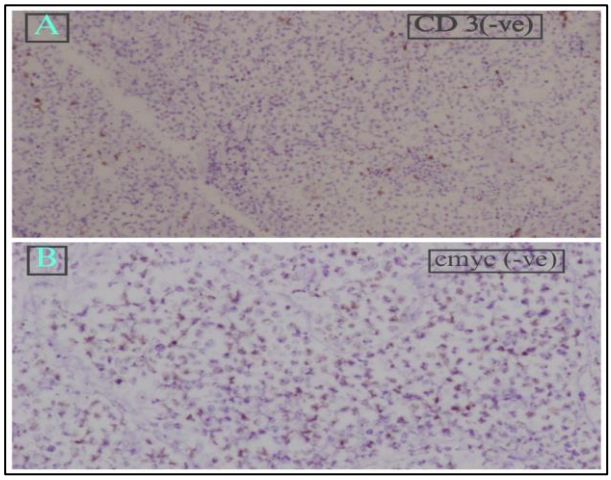

Figure 6 (A and B): Immunohistochemistry-negative to CD3 and C-myc.

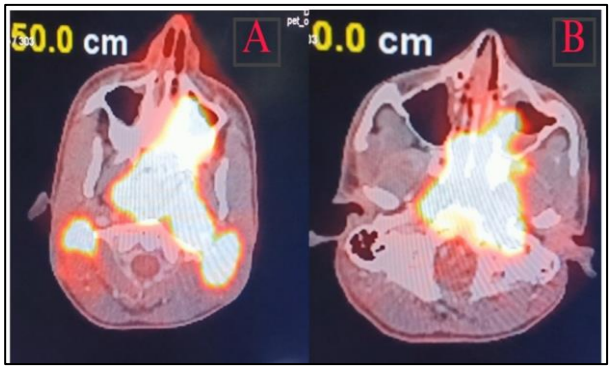

Figure 7 (A and B): PET CT-large ill-defined hypermetabolic space occupying soft tissue lesion involving nasopharynx, extending to posterior nasopharynx, left lateral nasopharyngeal wall and oropharynx with hypermetabolic cervical lymphadenopathy.

DISCUSSION

Hodgkin's lymphoma/NHL, is one of the 2 primary groups of lymphomas. NHL is further subdivided into natural killer cell lymphoma, T-cell lymphoma, and B-cell lymphoma.[5] The most prevalent type of lymphoma in the head and neck area is NHL, which is often a B-cell lymphoma. In pediatric age range, NHL is frequently of a high grade. About 30% of NHL cases are BL, which is typically aggressive in nature.[6] Lymph nodes, especially those in abdomen/head and neck region, are common sites for BL. Skin, bones, or central nervous system are possible additional sites.[6] In present case, it was located in the nasopharyngeal region. It was previously believed that BL could only be found in tropical Africa, not at higher elevations/in areas with very moderate temperatures. More BL happens when there is more rainfall.[6] Correlation between geographical and climatic factors points to a connection between BL and falciparum malaria.[6] There are 3 different types of BL: endemic (African) form, which is the most prevalent worldwide; the sporadic (American) form, which is more frequently observed in North America; and immunodeficiency-linked variety.[7] Immunodeficiency type and sporadic type are less common in the head and neck region and more common in the abdomen and bone marrow.[8] Nonetheless, Epstein-Barr virus is linked to African form of BL, which frequently affects nasofacial bones and surrounding soft tissues.[9] Approximately 58% of BL cases in the head and neck are located in maxilla/mandible.[10] The mandibular involvement site is linked to the clinical features of this aggressive neoplasm, which include proptosis, facial swelling, loosening of teeth, and jaw swelling.[10] Though very few cases have been documented in the nasal cavity and nasopharynx, it is less common in the nose and nasopharynx. Owing to rarity of BL in the nasopharynx, in present case report, authors emphasize misdiagnosis of adenoid hypertrophy for BL. In our case, the patient had nasal blockage and a headache and showed worsening symptoms such as oropharyngeal dysphagia and the rapid onset of bilateral swelling in the neck.

A CT scan is a crucial radiological test that's used to assess tumor staging and the extent and severity of the disease. The most effective medical test to identify the lesion's location is CT. Currently, functional and anatomical evaluations are conducted using positron emission tomography and CT scans for tumor staging and post-treatment monitoring.[11] In this case, CECT showed a large, poorly defined lesion involving the nasopharynx. Excisional biopsy and in situ hybridization are other tests helpful to get correct diagnosis. Histopathological reports showed poorly differentiated malignant neoplasm.

However, immunohistochemistry is a useful method for identifying particular tumor cells. For example, anti-CD20 positive and anti-CD3 negative immune staining tests can be used to confirm that tumor originated from B lymphocytes and to rule out the possibility that the tumor originated from T lymphocytes.[12] Immunohistochemical analyses in this instance revealed expression of CD20, CD10, and bcl-6 in lymphoid neoplasia. The highest

possible Ki-67 proliferation rate (100%) supported the BL diagnosis. CK, CD3, bcl-2, MUM-1, C-myc, and Tdt were not detected by IHC. Leukemia, blastoid mantle-cell lymphoma, high-grade B-cell lymphoma, and other variants are included in differential diagnosis.[13] The differential diagnosis in our case was lymphoma, nasopharyngeal malignancy, and extraosseous chordoma.

The definitive treatment of BL is often based on the patient's age and the location of tumor. There are different treatment options for BL, such as surgery, radiotherapy, chemotherapy, and radioimmunotherapy. The role of surgery in BL is still controversial. Surgical treatment is often required in cases of organic obstruction of airway, in conditions like optic nerve decompression, if the orbit is affected, or in case of a diagnostic biopsy. BL dramatically responds to chemotherapies that induce regressions of tumor and often lead to long-term remission.[13] In this case, 6 cycles of intravenous rituximab, etoposide, vincristine, adriamycin, and endoxan were given. Following chemotherapy, the patient recovered well. Patient had no mass lesion on endoscopy or, on the second PET scan four months after therapy. This could be attributed to an understanding of disease biology and the advancement of chemotherapeutic drugs.

CONCLUSION

The diagnosis of BL of the nasopharynx is confirmed by histopathological analyses and immunohistochemistry. BL presents with non-specific, gradual symptoms ranging from nasal blockage to mouth breathing, rarely detected in the nose and nasopharynx. Medical professionals should exercise a high degree of clinical suspicion in order to identify the pathology in its polymorphism due to the variety of clinical presentations and the nature of the disease. Since BL is an aggressive neoplasm and is frequently regarded as a serious medical illness, it is always necessary to treat the disease as soon as possible.

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