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Case Report

Primary diffuse large B-cell lymphoma of the nasal cavity: a rare aggressive entity

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ABSTRACT

Lymphomas are heterogeneous malignancies that arise from the clonal proliferation of lymphoid cells at different stages of maturation. Non-Hodgkin's lymphoma (NHL) often involves extranodal sites, such as the gastrointestinal tract, skin, bone, and brain. The most common subtype of NHL is diffuse large B-cell lymphoma (DLBCL), which has varied clinical and pathological findings depending on the primary site. DLBCL arising primarily from the nasal cavity is a rare and distinct entity, as it constitutes only 0.2-2% of all cases of NHL. It has a wide range of presentations from mild early-stage symptoms to severe forms in later stages. The symptoms are primarily due to the obstruction by the tumor mass. Caution should be given for early diagnosis and initiation of treatment, as it is an aggressive tumor and can extend rapidly to adjacent structures. Here, in this report, an unsuspected case of primary nasal cavity DLBCL in a 70-year-old male is described, demonstrating the unusual location of DLBCL and the significance of its accurate diagnosis and treatment.

Keywords: Nasal cavity, Lymphoma, Immunohistochemistry

INTRODUCTION

Lymphomas constitute 14% of all head and neck malignancies.[1] They are classified as Hodgkin's and non-Hodgkin's lymphomas (NHL), of which NHL has a higher frequency of involving the extranodal sites. The most common extranodal sites involved are the gastrointestinal tract, skin, bone, and brain. Extranodal NHL arising in the head and neck region is more commonly of B-cell lineage, predominantly diffuse large B-cell lymphoma (DLBCL).[2] This aggressive tumor has varied clinical and pathological findings depending on the primary site.[3]

Primary nasal cavity DLBCL is a rare and distinct entity. Here we describe one such rare case of primary nasal cavity DLBCL in a 70-year-old man so as to create awareness among clinicians dealing with this enigmatic entity.

CASE REPORT

A 70-year-old male patient presented in the outpatient department with a chief complaint of right nasal obstruction for 6 months, not relieved by nasal decongestants. It was insidious in onset and gradually progressive. He also experienced intermittent episodes of rhinorrhoea and pain over the right maxillary area for 2 months. It was not associated with epistaxis, sneezing, itching, headache, fever, or anosmia. There was no history of weight loss and night sweats. The patient had no history of trauma or tuberculosis contact. His medical history for any major disease or prior surgeries, as well as family history of any cancer, was non-contributory. On local examination, a diffuse swelling was present over the right side of the nose extending to the right maxillary area. On anterior rhinoscopy, an irregular mass measuring 1.5×1.5 cm was present within the right nasal cavity, which was soft to firm in consistency and did not bleed on touch. The

left nasal cavity appeared normal. There were no palpable lymph nodes or organomegaly, and the patient's vitals were within normal limits. No other abnormality was detected on his systemic examination. All the laboratory investigations were within normal limits. The patient was advised for contrast enhancement computed tomography (CECT) head and neck, followed by fine needle aspiration cytology (FNAC). CECT head and neck revealed a poorly defined infiltrative homogenously enhancing mass lesion involving the right-side nasal cavity, bilateral ethmoid sinuses, and right maxillary sinus. Associated hypodense retained secretions were also noted in the right maxillary sinus antrum and the right sphenoid sinus due to a block of sinus drainage pathways. The lesion was extending to the extraconal space of the right orbit. The coronal bone window image revealed an associated bony erosion of the medial and inferior walls of the right orbit, right nasal turbinates, right-sided uncinate process, septae of bilateral ethmoid air cells, and roof of bilateral ethmoid sinuses (floor of anterior cranial fossa). No obvious intracranial extension was seen (Figure 1a). Axial CT image through the neck showed a few homogenously enhancing enlarged and bulky bilateral cervical lymph nodes at levels Ib and II (Figure 1b). Imaging findings were consistent with a malignant/ aggressive sinonasal mass lesion.

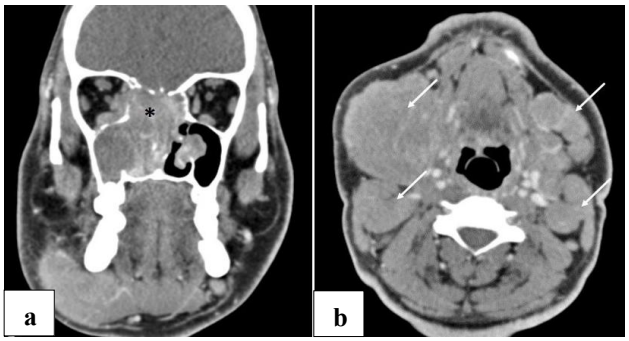

Figure 1: (a) Coronal CECT image showing a homogenously enhancing infiltrating mass lesion (black asterisk) involving the right side nasal cavity, and (b) axial CT neck revealing homogenously enhancing bilateral cervical lymph nodes (white arrows).

The patient, however, decided against the FNAC procedure. He was then advised to undergo a nasal mass biopsy. On histopathological examination of the nasal mass biopsy, hematoxylin and eosin-stained sections showed sheets of monomorphic population of medium to large-sized atypical lymphoid cells with pleomorphic nuclei, vesicular chromatin, and prominent nucleoli. Mitotic activity was 3-4/10 high power field. Areas of necrosis were present (Figure 2). A primary diagnosis of lymphoma was made. On immunohistochemistry (IHC), the tumor cells were positive for leukocyte common antigen (LCA), CD20, B-cell lymphoma 2 (BCL2), multiple myeloma oncogene-1 (MUM1), BCL6, and CD99. The Ki-67 proliferation index was 70% (Figure 3). Tumor cells were negative for cytokeratin (CK), CD10, CD5, synaptophysin, chromogranin, cyclin D1, CD3, CD30, TdT, CD56, S100, HMB45, vimentin, α-smooth muscle actin, and desmin. Based on these morphological and immunohistochemical findings, a final diagnosis of the non-germinal center B-cell subtype of DLBCL, right nasal cavity was made.

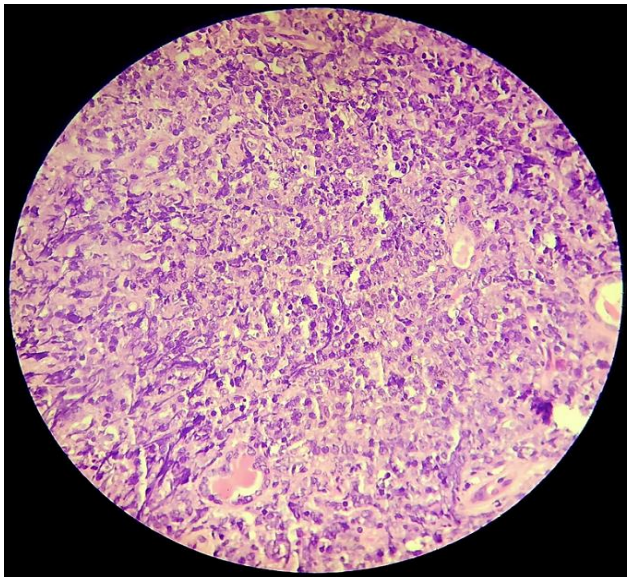

Figure 2: Photomicrograph showing nodal parenchyma effacement by tumor cells (H and E, x20).

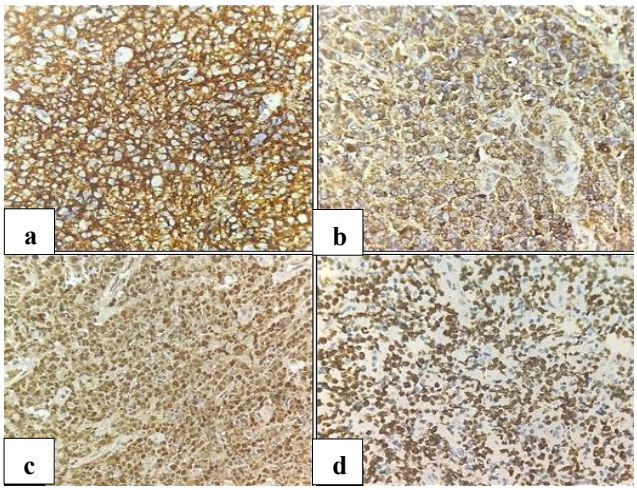

Figure 3: Tumor cells exhibiting (a) LCA immunopositivity (IHC, x20), (b) positive CD20 immunoexpression (IHC, x20), (c) MUM1 immunopositivity (IHC, x20), and (d) high Ki-67 proliferation index (IHC, x20).

Upon further follow-up, the patient was advised to undergo a fluorodeoxyglucose (FDG)-positron emission tomography (PET) scan to confirm the primary origin of the tumor. The PET scan revealed no other significant hypermetabolic abnormalities, except for activity in the anterior nasal cavity on the right side and bilateral cervical lymph nodes. He was started on chemotherapy, which was

a multidrug regimen consisting of rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisolone (R-CHOP) therapy. Unfortunately, he was lost to follow-up.

DISCUSSION

DLBCL is the most common adult NHL, 40% of which are extra-nodal in origin. Its incidence is higher in India, which ranges from 34% to 60%.[4] It predominantly affects males (55%), with the median age of presentation being 64 years.[5] Primary DLBCL arising in the nasal cavity is a rare presentation, as it constitutes only 0.2-2% of all cases of NHL.[6] Nasal cavity lymphomas are more commonly T-cell and NK cell subtypes of NHL, with extra-nodal involvement of the tonsils, throat, and paranasal sinuses being predominantly B-cell lymphomas.[7]

Nasal cavity DLBCL can present at the initial stages, but with very non-specific and relatively mild symptoms.[8] The common presenting symptoms are nasal obstruction, epistaxis, headache, and unilateral facial, cheek, or nasal swelling with or without B symptoms (fever, weight loss, and night sweats). The nasal soft tissue mass can be friable, ulcerative, necrotic, or polypoidal, which can rapidly extend to the adjacent structures.[2,8] In our case patient had nasal obstructive symptoms and did not have any B symptoms. Nevertheless, most of these symptoms can mimic other head and neck diseases (granulomatous as well as neoplastic), therefore, a high index of suspicion is required for early diagnosis of this rare entity.[7,9-11] A collaboration of various investigations is required to clinch its diagnosis. Once it is clinically suspected, imaging techniques such as CT and magnetic resonance imaging (MRI), although not specific, can help ascertain the extent and pattern of the disease. Additionally, CT offers the advantage of assessing for bony remodelling, or bone destruction, compared to MRI.[7]

FNAC has become a routine procedure for the pathological diagnosis of head and neck lesions.[12-17] A variety of non-neoplastic and neoplastic conditions can involve the sinonasal region. However, FNAC is seldom performed in this region. Nevertheless, authors have documented that FNAC is a reliable diagnostic procedure in the majority of sinonasal pathology cases, particularly when combined with clinical and radiological data.[9,18] However, the definitive diagnosis is made by biopsy and histopathological examination supported by ancillary studies, including IHC, fluorescence in situ hybridization, and flow cytometry.[8] The morphological subtypes of DLBCL include centroblastic, immunoblastic, and anaplastic variants. On IHC, they show positivity for B-cell markers like CD20, PAX5, CD79a, CD19, and CD22 with a high Ki-67 proliferation index. Molecular subtypes broadly correlate with different stages of B-cell differentiation and include subtypes: germinal center B-cell (GCB) and activated B-cell type (ABC)/non-GCB. This molecular classification has been the basis of prognostication in DLBCL for the last two decades.[19] On the other hand, gene expression profiling is the gold standard for identifying the cell of origin, but its applicability is limited due to high cost. Various IHC algorithms are utilized, with the Hans algorithm being widely accepted; it employs three markers: CD10, BCL6, and MUM1.[3,19] In our case, tumor cells were positive for MUM1, a marker of non-GCB DLBCL.

Early detection with precise categorization is very crucial in DLBCL cases. Researchers have observed that non-GCB DLBCL has worse outcomes and increased death rates.[4,20] Also among patients with the non-GCB subtype, inferior event-free and overall survival were reported in CD99-positive patients.[21]

The treatment of nasal DLBCL is similar to other sites, which is R-CHOP regimen-based chemotherapy with or without radiation.[2] In the study by Kwak et al, the final local control rate following radiotherapy was reported to be 94%.[22] However, another study indicated that there is no significant difference in survival rates between chemotherapy with or without rituximab, or chemotherapy plus radiotherapy compared to chemotherapy alone.[6,23]

The prognosis of the disease depends on the stage and extent of it. Nevertheless, primary nasal cavity and paranasal sinus lymphoma demonstrate aggressive behavior and often relapse.[6] In a nationwide cohort study on sinonasal B-cell lymphomas, an overall survival rate of 56% was found in primary sinonasal DLBCL, and 5-year progression-free survival was 50%. Also, 22% of the patients had recurrence.[3] Therefore, close monitoring and long-term follow-up are required in cases of primary nasal DLBCL. Unfortunately, our case was lost to follow-up, so the assessment of the treatment effect was not possible.

CONCLUSION

Primary DLBCL of the nasal cavity, although rare, should be considered in the differential diagnosis of nasal masses. Due to its varied presentations, it can be easily misdiagnosed. Therefore, recognizing its typical morphology is essential to avoid diagnostic errors. It is essential to conduct a thorough clinical history assessment and a detailed workup, including histopathological examination of the nasal mass in patients with this aggressive tumor. This approach is crucial for timely intervention and appropriate treatment. Additionally, further insight is needed to gain a better understanding of the behaviour of DLBCL that occurs at such unusual locations.

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