

Primary Non-Hodgkin's Lymphoma of The Nasal Cavity: Unusual Site and Unusual Presentation

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Abstract

Non-Hodgkin's lymphoma (NHL) is a heterogeneous subgroup of lymphomas with characteristic morphological and molecular features. Although lymph nodes are the most common organ of involvement, such as liver, stomach, intestine and bone marrow may also be affected on occasions. Primary non-Hodgkin's lymphoma (NHL) of the nasal cavity is an extra nodal form of NHL characterized by association with Epstein-Barr virus (EBV) infection, predominance of T-cell phenotype and a worse prognosis compared to nodal lymphoma. Although relatively common in China, nasal NHL is considered a rare entity in other countries of South East Asia. We are presenting a case of nasal NHL which was initially misdiagnosed as inverted papilloma and treated with surgical resection.

Keywords: Non-Hodgkin's Lymphoma; Paranasal lymphoma; Nasal lymphoma

Introduction

Lymphomas are malignant neoplasm of lympho-reticular system [1]. Non Hodgkin's lymphoma (NHL) is a heterogeneous subgroup of lymphomas with characteristic morphological and molecular features. Although lymph nodes are the most common organ of involvement, such as liver, stomach, intestine and bone marrow may also be affected on occasions [2]. Involvement of the nasal cavities by primary NHL is rare. These are classically associated with Epstein-Barr virus (EBV) infection and hence are common in those regions where EBV infection is endemic [3]. Although relatively common in China, nasal NHL is considered a rare entity in other countries of South East Asia [4,5]. There is no consensus on the management of these rare malignant neoplasm [6]. We are presenting a case of nasal NHL which was initially misdiagnosed as inverted papilloma and treated with surgical resection.

Case Report

A 50 year old gentleman presented with history of epistaxis and nasal blockade of 4 month duration. On examination, he had a polypoidal mass completely filling the left nasal cavity. Contrast enhanced computed tomography (CECT) of the nasal cavity revealed a well-defined, heterogeneously enhancing soft tissue mass occupying the entire nasal cavity with extension into the left maxillary sinus causing mass effect with bulging of nasal septum to the opposite side which was suggestive of inverted papilloma (Figure 1a). Biopsy of the mass was not conclusive of any specific malignancy even after 2 attempts. In view of CECT findings and clinical presentation of the patient, tentative clinical diagnosis was inverted papilloma and hence surgery was planned. Medial maxillectomy was performed. Intra and post-operative course were uneventful and patient was discharged on 3rd post operative day. Histopathological examination revealed monomorphic, large cells with eccentric nucleus and moderate to abundant cytoplasm with plasmablastic differentiation which was suggestive of Non Hodgkin's lymphoma of plasmablastic differentiation (Figure 1b and 1c). CECT thorax and abdomen were done as a part of staging. There was no evidence of disease at any other site and hence patient was staged as IE limited disease according to Ann Arbor staging system. He was treated with CHOP chemotherapy comprising of Cyclophosphamide 750 mg/m², Doxorubicin 50 mg/m², Vincristine 1.4 mg/m², Prednisolone 100 mg/m² for 5 days for a total of 6 cycles. At last follow up, patient was alive and disease free.

Discussion

Primary non-Hodgkin's lymphoma (NHL) of the nasal cavity is an extra nodal form of NHL characterized by association with Epstein-Barr virus (EBV) infection, predominance of T-cell phenotype and a worse prognosis compared to nodal lymphoma. In the west, paranasal sinus NHL is more common. In contrast, in the east, nasal NHL is more common than the NHL of the paranasal sinus [7].

Xiong et al. reported the largest series of primary non-Hodgkin's lymphoma (NHL) of the nasal cavity [8]. Median age of presentation was 44 years (Range: 11-79 years) and males were affected more often than females. Maxillary sinus was the most frequent site of involvement followed by the ethmoid sinus and the nasopharynx. The most common presenting symptoms include nasal obstruction, epistaxis and headache. Diplopia and blurred vision are rare infrequent symptoms associated with nasal NHL. The present patient is a middle aged male who presented with nasal obstruction and extension of the tumour into the maxillary sinus.

Macroscopically, these tumours are sub mucosal in contrast to squamous cell carcinomas which are ulcerative [9]. Angio invasion and necrosis are the two characteristic histological features of sino-nasal NHL [10]. NHL of the nasal cavity are predominantly T-cell lymphomas, whereas NHL of the paranasal sinuses are B-cell lymphomas.

Staging of the patients is according to Ann Arbor system [11]. In addition to CECT of the para nasal sinuses, CECT of the abdomen and thorax are required for adequate staging. Management is according to the stage of the disease. Stage IE is treated with chemotherapy or radiotherapy alone whereas stage IIE/IIIE and IVE require varying

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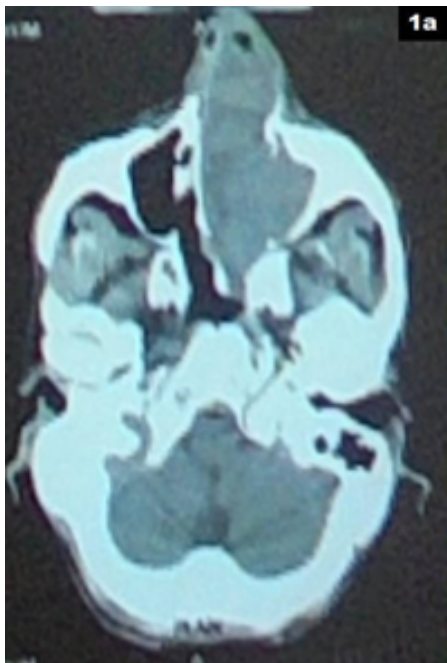


Figure 1A: Contrast enhanced computed tomography (CECT) of the nasal cavity.

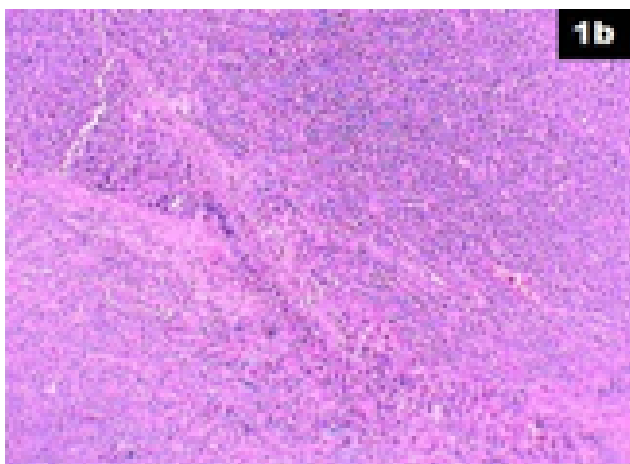


Figure 1B: Histopathological examination revealed monomorphic, large cells with eccentric nucleus.

combination of chemotherapy and radiotherapy [12-15]. The present patient had limited stage IE disease and hence was treated with chemotherapy alone.

In general, patients with nasal NHL have worse prognosis compared to other extra nodal sites of NHL. Stage is the most important factor determining prognosis. Patients with Stage IE disease confined to the nasal cavity have improved survival with estimated five-year overall survival (OS) and disease free survival (DFS) of 90% and 89%, respectively. In contrast, patients with Stage IIE, IIIE, and IVE have a worse prognosis with uniformly fatal outcome irrespective of the use of chemotherapy.

Conclusion

The purpose of presenting this case is to highlight the rarity of nasal

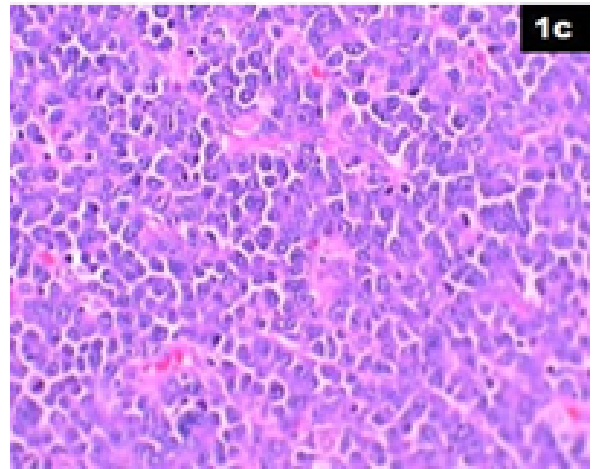


Figure 1C: Histopathological examination of cytoplasm with plasmablastic differentiation.

NHL in Indian subcontinent and stress the need for better awareness among treating oncologists, pathologist and radiologists about this disease entity.

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