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 polypoidal mass completely filling the left nasal cavity. Contrast enhancement of the nasal cavity revealed a well-defined, heterogeneous 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.

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. 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.

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/IIE and IVE require varying

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

References
