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CASE REPORT

Primary mantle cell lymphoma of the nasopharynx: a rare clinical entity☆

Linfoma primário de célula do manto da nasofaringe: uma entidade clínica rara

Ji-Hun Kang[a], Young-Dae Park[a], Chang-Hoon Lee[b], Kyu-Sup Cho[a,*]

[a] *Department of Otorhinolaryngology and Biomedical Research Institute, Pusan National University Hospital, Busan, Republic of Korea*
[b] *Department of Pathology, Pusan National University School of Medicine, Pusan National University Hospital, Busan, Republic of Korea*

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Introduction

Most non-Hodgkin's lymphomas (NHL) in the head and neck region develop in the extranodal lymphatic system of the Waldeyer ring.[1] Within the Waldeyer ring, the nasopharynx is the second most common site of disease after the tonsil. Primary nasopharyngeal lymphoma is much less common, occurring in only 8% of all NHL of the head and neck,[2] and diffuse large B-cell lymphoma (DLBCL) is the most common histologic type.[3] Mantle cell lymphoma (MCL) is a distinct subtype of B-cell lymphoma and comprises approximately 5–10% of all lymphomas.[4] MCL is characterized by an aggressive clinical course, and there is a pattern of frequent relapse after conventional chemotherapy.[4] MCLs involving the nasopharynx and oropharynx are extremely rare, and have not been reported in the literature, to the best of the authors' knowledge. This case report describes a rare clinical presentation of primary MCL arising in the nasopharynx and extending to the oropharynx. This study was approved by the institutional review board of Pusan National University Hospital.

Case report

A 66-year-old male with both nasal obstruction and ear fullness visited the authors' clinic. He denied fever, chills, and weight loss. The endoscopic examination revealed obstruction of both posterior choanae by a huge nasopharyngeal mass, accompanied by necrotic material. No cervical lymphadenopathies were felt. Paranasal sinus computed tomography (CT) showed a homogenous solid mass with mild enhancement involving both nasopharyngeal walls and extending to the upper oropharynx (Fig. 1). On magnetic resonance (MR) images, the homogenous mass demonstrated low signal intensity on T1-weighted images (T1WIs), intermediate signal intensity on T2WIs, and moderate enhancement on gadolinium-T1WIs (Fig. 2). A transnasal endoscopic biopsy of nasopharyngeal mass was performed. Histopathologic examination showed diffuse infiltration of

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* Corresponding author.
E-mails: choks@pusan.ac.kr, ckssmj@hanmail.net (K.S. Cho).

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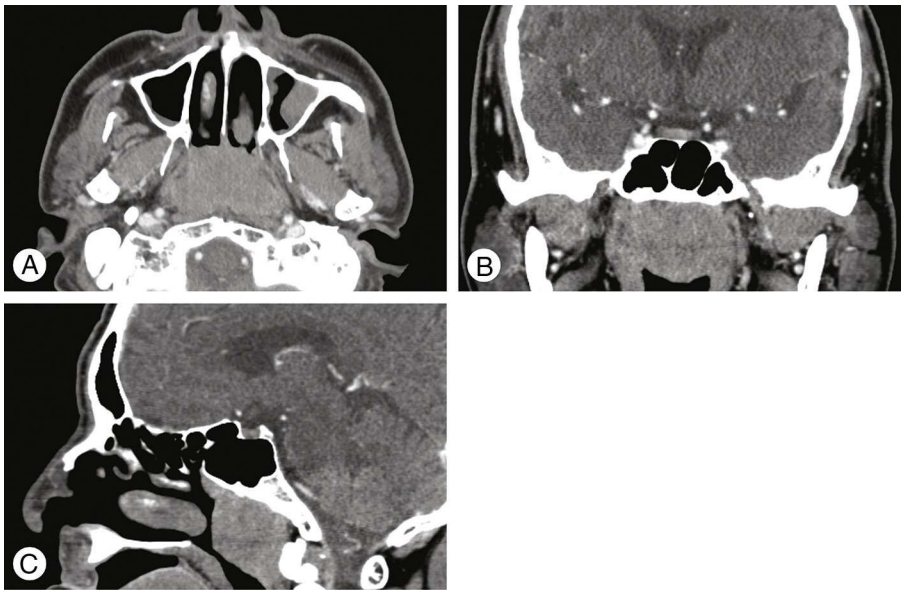

Figure 1 Computed tomography (CT) of paranasal sinus. Contrast-enhanced CT images show a bilateral, homogenous, mildly-enhanced solid mass from nasopharynx extending to upper oropharynx on axial (A), coronal (B), and sagittal (C) view.

small lymphocytic cells with mild nuclear atypia (Fig. 3A). Immunochemical staining revealed that the neoplastic lymphocytes were strongly positive for CD 20, CD5, and cyclin D1 (Fig. 3B–D). These findings were consistent with MCL. An upper gastrointestinal endoscopy, bone scan, and CT scan of the chest, abdomen, and pelvis were all reported as normal. A bone marrow biopsy showed no abnormalities. The patient was staged IE according to the Ann Arbor staging system. The patient received four cycles of R-CHOP chemotherapy and radiotherapy (total dose, 40 Gy). After 24 months of post-therapy follow-up, the patient exhibited no evidence of residual or recurrent disease.

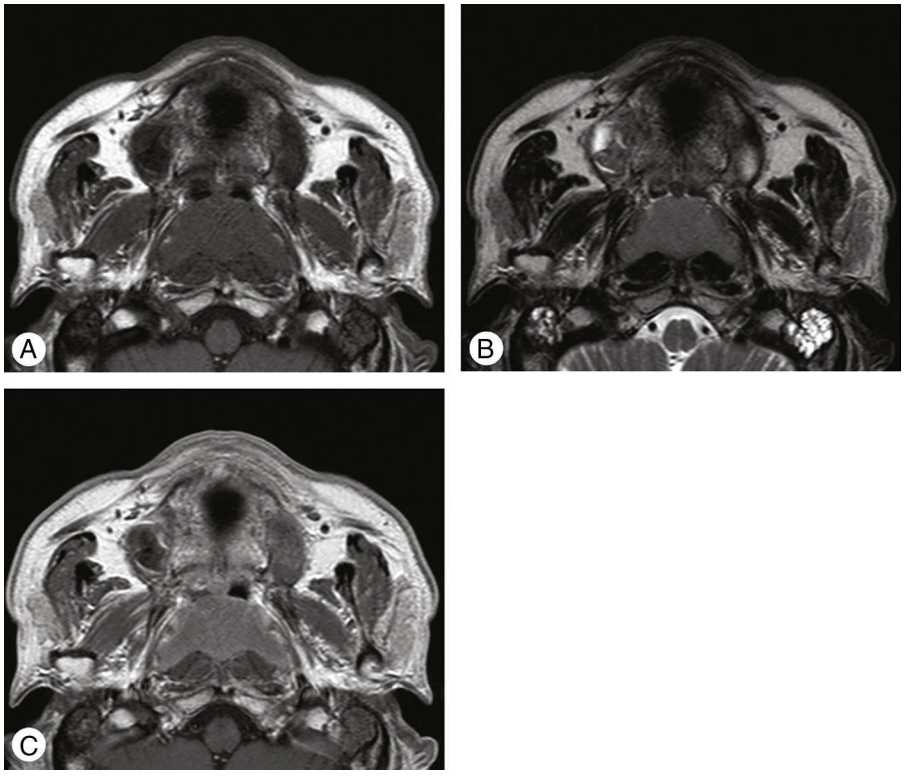

Figure 2 Magnetic resonance (MR) images of nasopharyngeal mass. Bilateral nasopharyngeal mass reveals low signal intensity on T1-weighted image (T1WI) (A) and intermediate signal intensity on T2WI (B). Postcontrast image (C) demonstrates moderate enhancement.

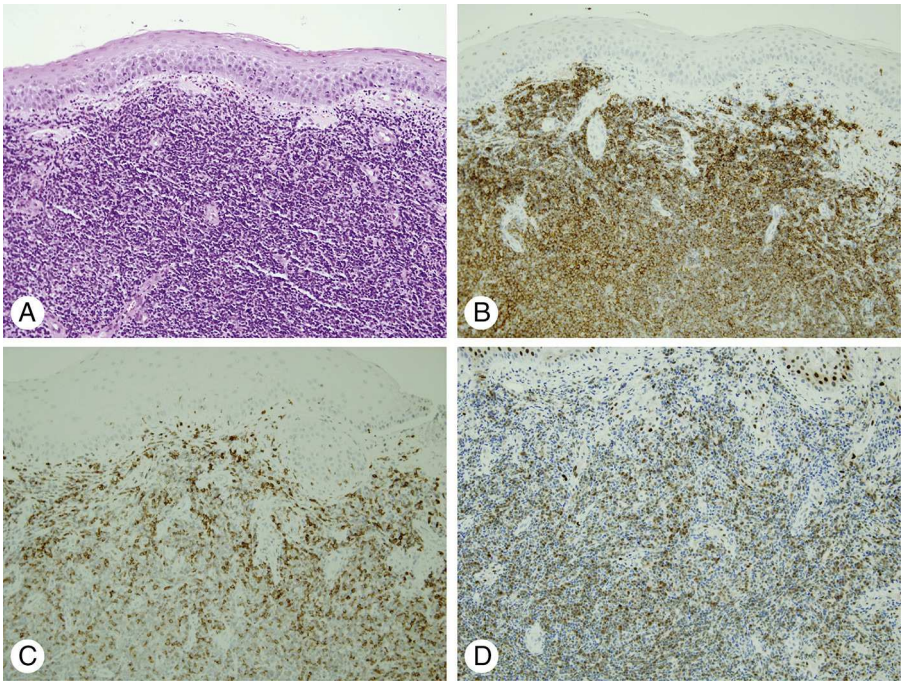

Figure 3 Histopathologic findings of nasopharyngeal mass. (A) Microscopic finding shows diffuse infiltration of small lymphocytic cells with mild nuclear atypia (H&E, ×200). Immunohistochemical staining shows strong positivity to anti-CD20 (B), anti-CD5 (C), and anti-cyclin D1 (D) antibody (×200).

Discussion

MCL is a subtype of B-cell lymphoma, derived from CD5-positive antigen-naïve pregerminal center B-cells within the mantle zone that surrounds normal germinal center follicles.[5] MCL cells generally over-express cyclin D1 due to a t (11:14) chromosomal translocation in the deoxyribonucleic acid.[5,6] The cause is unknown and no inherited predisposition has been identified. It accounts for about 5% of adult NHL in the United States, and moreover, the incidence of MCL has been increasing over the last decade, especially among elderly patients.[7] The population most commonly affected consists of men with a median age of 60 years.[7]

Clinically, MCL usually presents with stage III or IV disease and extensive lymphadenopathy, hepatosplenomegaly, and bone marrow involvement. One-quarter of patients are found to also have peripheral blood involvement.[5] Extranodal disease occurs less frequently, but when present, it typically may be found in the gastrointestinal tract and Waldeyer's ring.[6] In the extremely rare cases when MCL involves the nasopharynx, it presents with a nasopharyngeal mass. Described herein is the first case report of MCL arising in the nasopahrynx.

MCL is diagnosed by examination of affected tissue, obtained from a biopsy of a lymph node, tissue, bone marrow, or blood phenotype, which shows the typical morphology of monomorphic small- to medium-sized lymphoid cells with irregular nuclear contours.[8] Immunophenotyping is commonly used with MCL cells that are CD20+, CD5+, and positive for cyclin D1, whereas negative for CD10 and Bcl-6.[8] In most of patients with MCL, t (11:14) and other genetic changes cause excess production of cyclin D1, which is an early event in MCL.[9] MCL is presently staged by using a modified Ann Arbor system. This patient had stage I disease at presentation, with lesions of the nasopharynx and oropharynx, which was successfully treated by immunochemotherapy with radiotherapy.

Most MCL patients receive treatment following diagnosis and staging. A number of chemotherapy and rituximab combinations, such as R-CHOP, are used to treat patients with MCL. Although the addition of rituximab, a monoclonal antibody, has significantly improved the overall outcome, the five-year overall survival is as low as 40% in MCL patients.[5,10]

Conclusion

The first case of primary MCL arising in the nasopharynx and extending to the oropharynx has been described, which was successfully treated by immunochemotherapy with radiotherapy. This entity should be recognized and adequately diagnosed because it may have a more aggressive clinical course than other types of NHL in the head and neck. A detailed morphologic evaluation with thorough immunophenotyping is essential for an accurate diagnosis.

Conflicts of interest

The authors declare no conflicts of interest.

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