Case Report

Intraocular Involvement of a Nasal Natural Killer T-Cell Lymphoma: A Case Report

Jae Ho Yoo, Soo Young Kim, Kyu Bong Jung, Jung Joo Lee, Sang Joon Lee
Department of Ophthalmology, Kosin University College of Medicine, Busan, Korea

Malignant lymphomas that develop in the orbit and ocular adnexa account for 8% of all extranodal lymphomas [1, 2]. Most ocular and orbital lymphomas are non-Hodgkin’s B-cell lymphoma [1]. At these sites, with the exception of B-cell lymphoma, lymphoblastic diseases are rare and occur in only 1% to 3% of cases [2]. Ocular and orbital adnexa T-cell lymphomas are rare. There are a few cases of nasal natural killer T-cell lymphoma (NKTL) reported in the medical literature [1-4]. Additionally, pathologically confirmed intraocular involvement of nasal NKTL, such as the case reported here, is very rare [3].

Case Report

A 57-year-old woman presented with a three-day history of photophobia and diplopia in the left eye. One-month previously, she was diagnosed with nasal NKTL of the right nasal cavity. Ophthalmic examination revealed conjunctival injection and ptosis. The left pupil was fully dilated and non-reactive to light. Ocular motion was restricted on left-upper gaze. Five days later, anterior uveitis developed and persisted despite topical steroid treatment. An orbital magnetic resonance imaging was without specific findings, however, ophthalmoplegia, vitreous opacity, and an iris mass were observed. A diagnostic anterior chamber aspiration was performed. Aqueous humor aspiration revealed 35% morphologically atypical lymphocytes. After an intravitreal triamcinolone injection, radiotherapy and chemotherapy were administered; this resolved the uveitis and iris mass. When refractory uveitis or orbital pseudotumor occurs in patients with nasal NKTL, ocular and orbital involvement of the NKTL should be considered.

Key Words: Extranodal NK-T-cell lymphoma
munohistochemical staining was positive for CD56 (Fig. 2C) and CD3 (Fig. 2D). The patient was diagnosed with nasal NKTL with ocular involvement. After receiving radiotherapy at 900 cGy, her visual acuity improved and her
anterior uveitis, vitreous opacity, and iris mass resolved (Fig. 3A and 3B). However, the patient’s condition continued to deteriorate despite radiotherapy and chemotherapy, and her ptosis and ophthalmoplegia gradually progressed. She died of sepsis three months after diagnosis.

Discussion

Extranodal NKTL, including nasal NKTL, previously known as lethal midline granuloma, is a definitive diagnostic entity according to the World Health Organization lymphoma classification [1,4,5]. The nasal cavity is the most common site of involvement. However, histopathologically identical tumors may be identified at other extranodal sites, including the skin, muscle, gastrointestinal tract, liver, kidney, and retroperitoneal space [6,7]. Orbital and adnexal involvement has been infrequently reported in patients with this disorder [6]. There are few reported cases of nasal NKTL involving the orbit and/or ocular adnexa [2,6]. Coupland et al. [2] and Woog et al. [6] reported the formation of chronic uveitis and vitritis as intraocular manifestations of NKTL. Cimino et al. [3] did report a case of histologically confirmed nasal NKTL with intraocular involvement, however, in most cases intraocular involvement has not been confirmed histologically. The presence of intraocular involvement raises the possibility of leptomeningeal or central nervous system dissemination, and prompts consideration for external-beam radiotherapy to the eye and orbit in addition to systemic chemotherapy [1,6,8]. NKTL involving the ocular adnexa is generally a rapidly progressing disease, with a short survival from time of diagnosis, despite standard therapy [6,9,10]. This was true in the current case as well; the patient died three months after diagnosis. Since NKTL with ocular and orbital involvement is very rare and is characterized by rapid disease progression and a poor prognosis, NKTL should be considered in the differential diagnosis of uveitis or orbital pseudotumor refractory to therapy.

Conflict of Interest

No potential conflict of interest relevant to this article was reported.

References

1. Choi KH, Lee SJ, Suh YL, Kim YD. Nasal-type natural killer/T-cell lymphoma of the orbit. J Korean Ophthalmol Soc 2004;45:2145-50.
2. Coupland SE, Krause L, Delecluse HJ, et al. Lymphoproliferative lesions of the ocular adnexa. Analysis of 112 cases. Ophthalmology 1998;105:1430-41.
3. Cimino L, Chan CC, Shen D, et al. Ocular involvement in nasal natural killer T-cell lymphoma. Int Ophthalmol 2009;29:275-9.
4. Hon C, Kwok AK, Shek TW, et al. Vision-threatening complications of nasal T/NK lymphoma. Am J Ophthalmol 2002;134:406-10.
5. Al-Hakeem DA, Fedele S, Carlos R, Porter S. Extranodal NK/T-cell lymphoma, nasal type. Oral Oncol 2007;43:4-14.
6. Woog JJ, Kim YD, Yeatts RP, et al. Natural killer/T-cell lymphoma with ocular and adnexal involvement. Ophthalmology 2006;113:140-7.
7. Nakamura S, Suchi T, Koshikawa T, et al. Clinicopatho-
logic study of CD56 (NCAM)-positive angiocentric lymphoma occurring in sites other than the upper and lower respiratory tract. *Am J Surg Pathol* 1995;19:284-96.

8. Cheung MM, Chan JK, Lau WH, et al. Primary non-Hodgkin’s lymphoma of the nose and nasopharynx: clinical features, tumor immunophenotype, and treatment outcome in 113 patients. *J Clin Oncol* 1998;16:70-7.

9. Chan JK, Sin VC, Wong KF, et al. Nonnasal lymphoma expressing the natural killer cell marker CD56: a clinicopathologic study of 49 cases of an uncommon aggressive neoplasm. *Blood* 1997;89:4501-13.

10. Chan JK. Natural killer cell neoplasms. *Anat Pathol* 1998;3:77-145.