Case Report

Nasal extranodal NK/T-cell lymphoma mimicking inflammatory polyp: a case with indolent clinical behavior

Qing-Fu Zhang¹, Yu-Nan Han², Li-Mei Sun¹, Ying Tian³, Xue-Shan Qiu¹

¹Department of Pathology, The First Affiliated Hospital and College of Basic Medical Sciences of China Medical University, Shenyang 110001, People’s Republic of China; ²Department of Breast Surgery, The First Affiliated Hospital of China Medical University, Shenyang 110001, People’s Republic of China; ³Department of Otorhinolaryngology, The First Affiliated Hospital of China Medical University, Shenyang 110001, People’s Republic of China

Received October 9, 2015; Accepted November 20, 2015; Epub January 1, 2016; Published January 15, 2016

Abstract: Extranodal natural killer (NK)/T-cell lymphoma, nasal type (ENKTCL-NT) is an uncommon subtype of non-Hodgkin lymphoma with an aggressive course and poor prognosis, which is mainly seen in Asia and Latin America. ENKTCL is associated with the Epstein-Barr virus (EBV) and is characterized by vascular destruction and prominent necrosis. We reported a 53-year-old Chinese man who was diagnosed with ENKTCL-NT without tissue necrosis and angioinvasion. In this case, the lesion has a polypoid growth pattern and the clinical course was indolent, it mimicked nasal inflammatory polyps and was difficult to diagnose.

Keywords: Extranodal natural killer (NK)/T-cell lymphoma, inflammatory polyp, immunohistochemistry

Introduction

Extranodal natural killer (NK)/T-cell lymphoma (ENKTCL) is a category of NK cell-derived neoplasms recognized as a distinct entity by the WHO classification of lymphoid tumors in 2008. Extranodal natural killer (NK)/T-cell lymphoma, nasal type (ENKTCL-NT) is uncommon and is distinctive in geographical distribution, accounting for 7% to 10% of all non-Hodgkin lymphomas in Asia and Latin America and rarely prevalent in Western countries [1]. ENKTCL-NT mainly affects the nasal cavity and paranasal sinuses, and most patients primarily present lesions in the nasal cavity or lesions above the laryngopharynx. It is highly aggressive, with poor prognosis and a short survival period [2].

In this study, we reported a case of polypoid ENKTCL-NT without its typical features, such as vascular destruction and prominent necrosis, but instead having indolent biological behavior. Our case suggests that ENKTCL-NT with a polypoid growth pattern may be confused in diagnosis with nasal inflammatory polyp.

Case description

A 53-year-old Chinese man had a history of bilateral nasal polypsis resection 20 years ago. The patient presented to the hospital with a progressive right nasal obstruction that lasted for 50 days. The right nasal cavity had purulent secretions without bleeding discharge. In the recent two weeks, the patient noticed that the right side of the face was swollen without obvious tenderness, and both eyelids and bulbar conjunctivas were red and swollen. In physical examination, both inferior turbinates were hypertrophic, with no obvious deviation of the nasal septum and no abnormal secretions in the left nasal cavity. Nasal endoscopic examination revealed numerous polyp-like tissues almost completely filling the right nasal cavity. The mucous membrane of surface of the right nasal cavity was unsmooth with a grey appearance. The purulent secretions contained no evident hemorrhage and necrosis. Computed tomography (CT) showed that multiple high-density masses occupied the right
maxillary sinus, ethmoid sinus, and frontal sinus, and no obvious bone destruction was observed (Figure 1). After ineffective anti-inflammatory treatment, functional endoscopic sinus surgery (FESS) was performed to remove the nasal polyp-like lesions of the right nasal cavity and to clean out and enlarge sinus openings. The patient also received chemotherapy and radiotherapy after the surgery.

Materials and methods

The biopsy specimen was fixed in 10% buffered formalin solution and embedded in paraffin, sectioned at 4 µm, and then stained with hematoxylin and eosin (H&E). Immunohistochemical staining was performed using the streptavidin-peroxidase system (Ultrasensitive, MaiXin Inc, Fuzhou, China) according to the instructions of manufacturer. Heat-induced epitope retrieval was performed. The following antibodies (MaiXin Inc, China, prediluted) were used: Cytokeratin (pan), CD3ε, CD20, Pax-5, granzyme B, TIA-1, CD56, CD4, CD8, S-100, Desmin, Chromogranin A, Synaptophysin, and Ki-67. Epstein-Barr virus encoded RNA (EBER) was detected by in situ hybridization (ISH). Positive and negative controls were evaluated appropriately for each procedure.

Pathological findings

Histopathological analysis showed that the tumor was in polypoid-growth configuration, covered with pseudostratified ciliated columnar epithelium, and epithelial cell shedding was observed in a small part of the tumor surface. Normal glands of the mucosa were decreased; cells of the residual glands showed acidophilic degeneration in the cytoplasm, and numerous lymphocyte infiltrations were seen in the subepithelium (Figure 2A). Tumor cells varied in density and distribution; local tumor cells invaded glands and formed lymphatic epithelial lesions. Neoplastic cells showed similar small to intermediate size and shape, irregular nuclear contours, clear cytoplasm, and dispersed karyorrhexis, but lacked cell necrosis and apoptosis (Figure 2B). Intercellular substance was loose and edematous, absent in lymphoid follicles, infiltrated with dense plasma cells and small lymphocytes, and some small blood vessels were dilated and congested (Figure 2C). Focally, the tumor cells exhibited an angiocentric growth pattern, but vascular wall damage and fibrinoid necrosis were not identified in these sections (Figure 2D). Immunohistochemical staining showed that the tumor cells were positive for cytoplasmic CD3ε (Figure 3A), CD56 (Figure 3B), and the cytotoxic molecules (granzyme B, TIA-1), but negative for cytokeratin (pan), B cell markers (CD20 and Pax-5), S-100, Desmin, Chromogranin A, and Synaptophysin. CD4 and CD8 shows that a subset of cells were positive. EBER in situ hybridization (ISH) staining (Figure 3C) showed that nearly all neoplastic cells were positive; the Ki-67 (Figure 3D) labeling index was about 80%. Based on the histopathological and immunohistochemical findings, we confirmed the pathological diagnosis of ENKTCL-NT.

Discussion

ENKTCL-NTis a distinct group of aggressive lymphoid neoplasm. It is now clear that the majority of these tumors express the NK-cell phenotype, although a few express the T-cell phenotype. Nearly all cases have evidence of EBV infection and have positive reactivity for EBV by ISH [3]. This case was an indolent ENKTCL-NT and was absent of characteristic histologic findings, such as angiocentric growth patterns and obvious tissue necrosis. This patient underwent surgical resection of the lesions in the form of nasal polyp removal. The morphological findings were subtle and similar to nasal inflammatory polyps. Also, the patient’s clinical history and examinations may cause misdiagnosis, and ENKTCL-NT could have been easily overlooked.
Previous reported lymphomas, which had the growth pattern of multiple mucosal polyps, usually occurred in the gastrointestinal tract, including the esophagus, stomach, duodenum, and intestines. Multiple lymphomatous polyposis (MLP) is characterized by the formation of multiple mucosal polyps [4]. Histological findings of MLP tend to be classified as mantle cell lymphoma [5], follicular lymphoma [6], small lymphocytic lymphoma [7], diffuse large cell lymphoma [8], or mucosa-associated lymphoid tissue lymphoma [9].

The polypoid structures in ENKTCL-NT may suggest that the tumor is benign, thus causing it to easily be misdiagnosed as nasal inflammatory polyps. The ENKTCL-NT showed a broad cytomorphological spectrum, and polypoid-like morphological findings caused the possibility of overlooking the diagnosis of ENKTCL-NT [10]. Here, we presented this case to indicate that multiple polypoid nasal lesions could also occur in ENKTCL-NT, and pathologists should be aware of this rare histological morphology in order to avoid misdiagnosis.

NK/T-cell lymphomas are characterized by an aggressive clinical course and poor response to treatment, so patients with ENKTCL-NT have poor survival outcome, with the five-year overall survival rate being 60.3% [11]. In the newly proposed model for ENKTCL (Korean prognostic model), which included B-symptoms, stages, LDH levels, and regional lymphadenopathy, four prognostic groups were identified as important adverse factors for survival [12]. However, the relationship between the histological morphology and cytological grading of ENKTCL-NT and the prognosis does not have specified research.

In conclusion, this case presented polypoid-like structures under microscopic examination and lacked the typical vascular destruction and necrosis of ENKTCL-NT. The patient has a 20-year nasal polyp disease history, he was
treated with complete remission in combination with radiotherapy and chemotherapy, and there was no recurrence after a 4-month follow-up. Thus, this was an indolent case of ENKTCL-NT. We speculated nasal ENKTCL may also have a differentiation lineage, with different histological morphology and different prognostic implications, but more case studies are needed to explain this phenomenon.

Acknowledgements

This work was supported by the Program for Liaoning Innovative Research Team in University (No. LC2015029), the Natural Science Foundation of Liaoning Province of China (No. L2015598), and the Program of Science and Technology Department of Liaoning Province (No. 2013225021).

Disclosure of conflict of interest

None.

Address correspondence to: Xue-Shan Qiu, Department of Pathology, The First Affiliated Hospital and College of Basic Medical Sciences of China Medical University, Shenyang 110001, People’s Republic of China. E-mail: xsqiu@mail.cmu.edu.cn

References


Nasal extranodal NK/T-cell lymphoma


