Primary Natural Killer/T-Cell Lymphoma of the Ribs

Kostaların Birincil Doğal Öldürücü/T-Hücreli Lenfoma

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To the Editor,

Extranodal natural killer/T-cell lymphoma (ENKTCL) is a highly aggressive disease that is strongly associated with Epstein-Barr virus (EBV). ENKTCL can involve the nasal cavity, brain, skin, gastrointestinal tract, and upper respiratory tract [1]. Primary bone lymphoma is not common in ENKTCL, with only a few cases reported [1,2].

An 81-year-old male patient presented to the cardiovascular thoracic surgery department for evaluation of an incidentally detected mass of the right third rib. He had undergone surgical resection and chemotherapy for rectal cancer 3 years prior. Laboratory findings revealed hemoglobin of 114 g/L, red blood cell count of 3.91×10^{12}/L, white blood cell count of 5.01×10^{9}/L, and platelet count of 226×10^{9}/L. His β2-microglobulin level was 2.9 mg/L. Magnetic resonance imaging revealed a large mass of the right third rib. The mass was 9.6 cm in size with high signal intensity on T2-weighted imaging. Imaging with fluorodeoxyglucose-positron emission tomography/computed tomography (FDG-PET/CT) showed an FDG-avid osteolytic lesion in the third rib (Figures 1A and 1B). The clinical impression suggested bone metastasis of rectal cancer or osteosarcoma.

A needle-biopsy specimen revealed diffuse infiltration of lymphoid cells. Immunohistochemical staining showed positivity for CD3, granzyme B, and CD56. In situ hybridization was positive for EBV-encoded RNA (EBER) (Figures 1C-1F). The tumor appeared within the rib with no other site involvement, and the patient had no previous ENKTCL history. Based on these findings, the pathological diagnosis was primary bone NK/T-cell lymphoma. After diagnosis, the patient underwent treatment with the SMILE regimen (dexamethasone, methotrexate, ifosfamide, L-asparaginase, and etoposide). However, his condition deteriorated and he refused further therapy including other chemotherapy regimens or radiotherapy. At the last follow-up in October 2022, the patient remained alive.

Primary bone NK/T-cell lymphoma was first reported as a spinal tumor in 2008 [3]. To our knowledge, only four cases were reported between 2008 and 2021, including the present case [2,3,4]. Of these four patients, three were from China and one was from South Korea. The fact that all patients were Asian shows consistency with ENKTCL being more prevalent in Asia. The range of these patients' ages was 49-81 years and all were male. Radiological findings of primary bone NK/T-cell lymphoma have been nonspecific. In all cases, the primary bone NK/T-cell lymphoma was described as a nodular lesion or mass [2,3,4]. Thus, differential diagnosis is challenging before biopsy.

Histopathological characteristics reveal diffuse infiltration of atypical lymphoid cells. Tumor cell size is variable, ranging from small to large. In the present case, immunohistochemical findings revealed tumor cell positivity for CD3, CD56, and granzyme B. Both NK cells and T-cells were found to be positive for EBER via in situ hybridization.

The prognosis of ENKTCL is very poor. The overall survival time was less than 3 months in all previously reported cases [2,3,4]. Specific treatment guidelines have not been established yet, but the SMILE regimen may be an effective treatment approach for ENKTCL. Recently, the effectiveness of stem cell transplantation has also been evaluated [4]. Since primary bone NK/T-cell lymphoma is an extremely rare entity, more case reports would help elucidate the clinicopathological characteristics of this entity and establish optimal treatment guidelines.

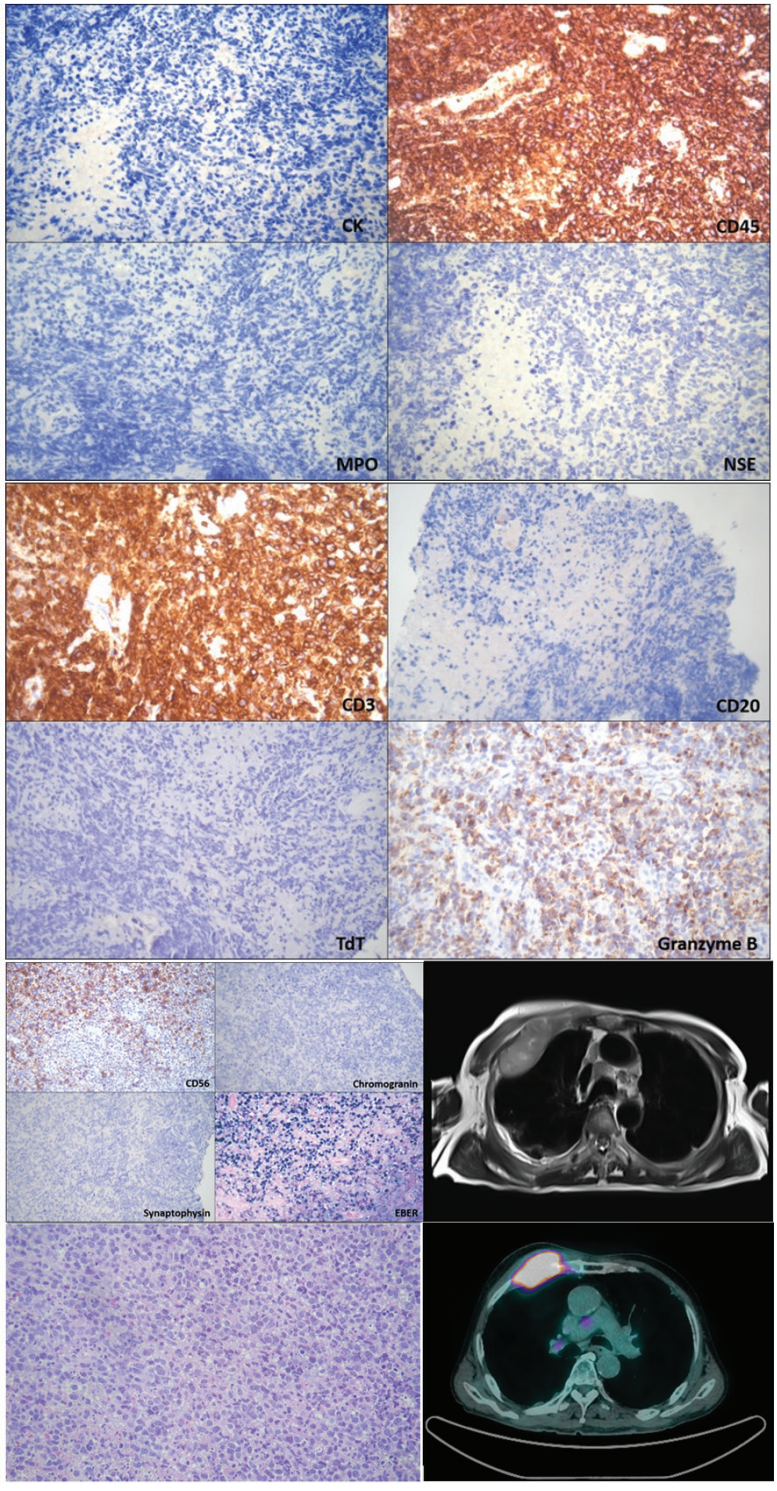

Figure 1. Radiological and histopathological findings of primary bone natural killer/T-cell lymphoma: (A) The tumor presented as a 9.6-cm mass with high signal intensity on T2-weighted imaging. (B) The mass showed a fluorodeoxyglucose-avid osteolytic lesion in the third rib on fluorodeoxyglucose-positron emission tomography/computed tomography. (C) The mass was composed of variably sized lymphoid cells with a diffuse pattern. Apoptotic bodies were observed (hematoxylin and eosin, 400^{x}). (D) Immunohistochemical staining showed positivity for CD45. Tumor cells were negative for cytokeratin (CK), myeloperoxidase (MPO), and neuron-specific enolase (NSE). These findings revealed the tumor to be a lymphoid neoplasm (original magnification, 400^{x}). (E) Immunohistochemical staining showed positivity for CD3 and granzyme B and negativity for CD20 and TdT. These findings suggested that the tumor was natural killer/T-cell lymphoma (original magnification, 400^{x}). (F) Immunohistochemical staining showed positivity for CD56 and negativity for chromogranin and synaptophysin. These findings excluded the possibility of a neuroendocrine tumor. In addition, in situ hybridization was positive for EBV-encoded RNA (EBER) (original magnification, 400^{x}).

Keywords: Lymphomas, T-cell neoplasms, Lymphoid cells neoplasms, NK cell neoplasms

Anahtar Sözcükler: Lenfomalar, T-hücreli neoplazmalar, Lenfoid hücreli neoplazmalar, NK hücreli neoplazmalar

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Ethics

Ethics Committee Approval: This case report was approved by the Jeonbuk National University Hospital Institutional Review Board (approval no. IRB 2021-11-002).

Informed Consent: Patient consent was obtained for publication.

Authorship Contributions

Concept: K.M.K., K.Y.J., J-Y.K., A.R.A.; Design: K.M.K., K.Y.J., J-Y.K., A.R.A.; Data Collection or Processing: K.M.K., K.Y.J., J-Y.K., A.R.A.; Analysis or Interpretation: K.M.K., K.Y.J., J-Y.K., A.R.A.; Literature Search: K.K., K.J., A.A.; Writing: K.M.K., K.Y.J., J-Y.K., A.R.A.

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