Case Report

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대한영상의학회지

MR Imaging Characteristics of Primary T-Cell Lymphoma of the Cauda Equina: A Case Report and Literature Review

말총의 원발성 T세포 림프종에서 MR 영상 소견: 증례 보고와 문헌 고찰

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Primary central nervous system lymphoma is a rare form of extranodal non-Hodgkin lymphoma, and primary T-cell lymphoma of the cauda equina is extremely rare. We describe a case involving a 56-year-old female who presented with low back pain and radiating leg pain for 4 months. MRI of the lumbar spine revealed an elongated, multinodular intradural lesion of approximately 10 cm from the L4 body to the S2 body level with iso-signal intensity on T1-weighted imaging, heterogeneous iso- and high-signal intensity on T2-weighted imaging, and a heterogeneous intense enhancement on gadolinium contrast-enhanced T1-weighted imaging. A peripheral T-cell lymphoma of the cauda equina was diagnosed on the basis of immunohistochemical and T-cell receptor gamma gene rearrangement analysis after intradural biopsy of the mass.

Index terms Lymphoma, T-Cell, Peripheral; Magnetic Resonance Imaging; Lumbosacral Region; Cauda Equina

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INTRODUCTION

Spinal lymphoma accounts for only 3.3% of all cases of lymphoma of the central nervous system (CNS), which in turn accounts for 1% of all lymphoma (1). Based on our review of the English literature published between 1996 and 2019, only 2 cases of primary T-cell lymphoma of the cauda equina (2, 3) have been reported. Because most of the primary CNS lymphoma (PCNSL) is B-cell lymphoma (4), the imaging characteristics of T-cell lymphoma remain unclear. We report a case of primary T-cell lymphoma at the

cauda equina and describe the relevant MRI characteristics.

CASE REPORT

A 56-year-old female presented with a 3-year history of low back pain. Her medical history was insignificant, with no history of surgery. Her symptoms were insidious and relieved with epidural steroid injections. However, the injection therapy ceased to be effective for 8 months before presentation. She experienced severe radiating pain from the gluteal area to the bilateral posterior thighs and lateral side of the calf, and the first toe, with bilateral leg weakness and fecal incontinence since 4 months before presentation. Although an epidural steroid injection was administered for the radiating pain 1 month before presentation, the pain persisted.

MRI of the lumbar spine was performed to evaluate the refractory back pain. MRI revealed a relatively well-defined, elongated, multinodular intradural lesion of from the L4 body to the S2 body level of approximately 10 cm. The mass had iso-signal intensity to the spinal cord on T1-weigted imaging. Most of the mass revealed iso-signal intensity on T2-weighted imaging; however, few peripheral portions demonstrated high signal intensity similar to the cerebrospinal fluid (CSF). After gadolinium contrast enhancement, the mass showed intense heterogeneous enhancement primarily in the portions of high signal intensity on T2-weighted imaging and along the outer dural wall. The lesion occupied the central spinal canal from the L4 to S2 level completely (Fig. 1A).

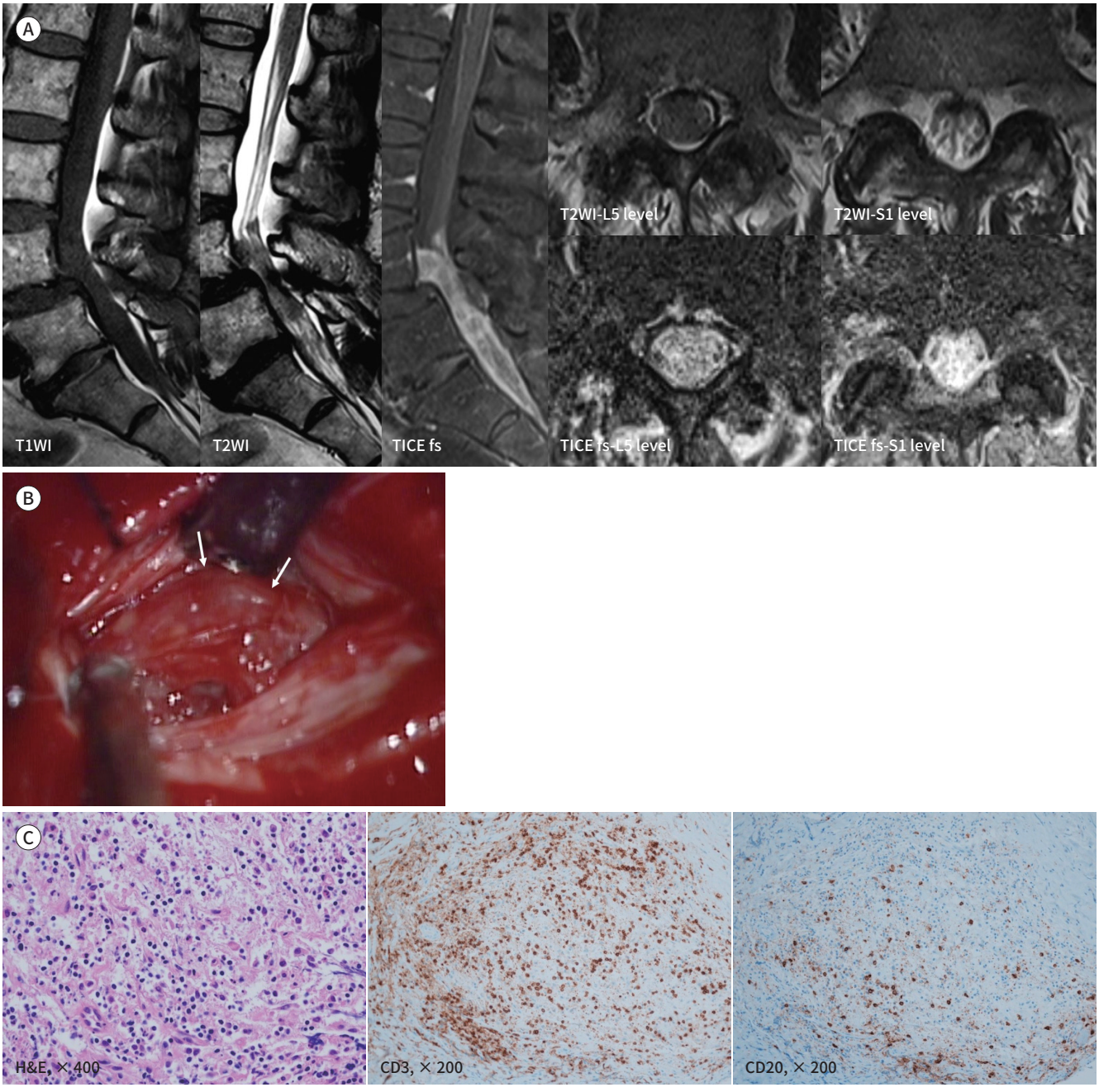
We planned a surgical spinal biopsy. Routine hematology and biochemistry evaluations at admission were normal. The white blood cell count was 11/mm^3, and the total protein level was high (105.7 mg/dL; normal range, 8–43 mg/dL) in the CSF study. Fluorine-18 fluorodeoxyglucose (^{18}F-FDG) PET/CT suggested only a focal, intense hypermetabolic lesion at the spinal canal at the L4-S2 level (maximal standardized uptake value = 6.4). Chest and abdominal contrast-enhanced CT examinations revealed no other abnormality.

Decompressive bilateral laminectomy at L4/5 revealed bulging intradural masses; intradural biopsies were performed subsequently (Fig. 1B). Hematoxylin-eosin staining revealed dense lymphocytic and histiocytic infiltration with granulomatous inflammation along the dural and neural tissues. The repeated epidural injections were considered the cause of granulomatous inflammation. Immunohistochemistry of the tissue was positive for CD3, a marker for T-cell lymphoma in infiltrative lymphocytes, and CD20, a marker for B-cell lymphoma in some cells (Fig. 1C) (5). Multiplex polymerase chain reaction fragment analysis revealed monoclonal T-cell receptor gamma gene rearrangement, indicating a T-cell origin of the lymphoma. Although immunohistochemistry revealed CD20 positivity in some cells, CD20 expression has been reported in peripheral T-cell lymphoma cases (5). The final pathologic diagnosis was primary peripheral T-cell lymphoma basis other examinations including ^{18}F-FDG PET/CT.

Symptoms including radiating leg pain, leg weakness, numbness, or fecal incontinence resolved gradually after L4/5 decompression. She was treated with intravenous chemotherapy with high-dose methotrexate and high-dose cytarabine for PCNSL every 3 weeks, and the fourth chemotherapy cycle is ongoing currently without major adverse events.

The study protocol conformed to the ethical guidelines of the 1975 Declaration of Helsinki.

Fig. 1. Primary T-cell lymphoma of the cauda equina in a 56-year-old female.
A. An approximately 10-cm-long multinodular mass is located in the cauda equina from the L4 to the S2 body level. On the T1-weighted sagittal image, the lesion is slightly heterogeneous and isointense with the spinal cord. On the T2WI, the lesion is heterogeneously isointense to hyperintense with the spinal cord. On the enhanced fat-suppressed T1WI, the lesion is heterogeneous and shows strong enhancement, particularly along the dural wall. On the T2WI and enhanced fat-suppressed T1-weighted axial images at the L5 vertebral body level, the nodular lesion shows isointensity with heterogeneous enhancement. On the T2WI and enhanced fat-suppressed T1WI at the S1 vertebral body level, the entire spinal canal is tightly packed with the thickened nerve roots, and the peripheral portions of the nerve roots show hyperintensity with bright enhancement.
B. After total laminectomy at the L4/5 level, an abnormal whitish red intradural bulging mass is seen (arrows), which was biopsied.
C. Histopathologic and immunohistochemical examination of the mass on the cauda equina after a biopsy. H&E staining (× 400) shows that the lymphoma cells had infiltrated into the neural tissue with a high nucleus-to-cytoplasm ratio. Immunohistochemistry indicates that most of the atypical large lymphoma cells are positive for CD3 and some are positive for CD20 (× 200).
fs = fat saturated, H&E = hematoxylin and eosin, T1CE = contrast enhanced T1WI, T1WI = T1 weighted image, T2WI = T2 weighted image



DISCUSSION

PCNSL accounts for only 1%–2% of non-Hodgkin lymphoma. In addition, spinal cord PCNSL is extremely rare (6). Spinal lymphoma accounts for 3.3% of all CNS lymphoma, which constitutes only 1% of all lymphoma (1). To our knowledge, the incidence of primary lymphoma in the cauda equina remains unreported.

Most cases of PCNSL in the cauda equina present with cauda equina syndrome including low back pain, weakness and numbness of the lower limbs, and urinary dysfunction (7).

Primary spinal intramedullary lymphoma usually shows a diffusely enlarged spinal cord on MRI. Most PCNSLs are of B-cell origin, and PCNSLs of T-cell origin are extremely rare (4). The typical MRI features of B-cell lymphoma at cauda equina include isointense or hyperintense enlargement of the cauda equina relative to the spinal cord's intensity on T1-weighted imaging and hyperintense on T2-weighted imaging. Furthermore, it shows marked enhancement after gadolinium administration (3, 8).

The lesion in our patient was a solid mass lesion with diffuse nerve root thickening. T1-weighted MRI showed isointensity with the spinal cord; T2-weignted imaging showed heterogeneous isointensities and hyperintensities and avid heterogeneous enhancement particularly along the peripheral dural wall.

According to our review of the English literature, only 2 cases of T-cell lymphoma among the primary lymphoma of the cauda equina have been reported, and the reported imaging findings vary among those studies (2, 3). Ooi et al. (2) reported ill-defined intrathecal masses isointense with the CSF on T1-weighted imaging that show significant enhancement on contrast-enhanced T1-weighted imaging; however, they did not report signal intensity on T2-weighted imaging. Morita et al. (3) reported a mass lesion isointense with the CSF on T1-weighted imaging and hypointense on T2-weighted imaging, with homogeneous enhancement after contrast administration. Basis the findings of the 2 previous reports and our case, spinal cord T-cell lymphoma is likely isointense with the CSF on T1-weighted and of variable intensity on T2-weighted imaging with intense contrast enhancement regardless of the homogeneous or inhomogeneous pattern.

The main differential diagnoses of primary tumors arising at the cauda equina include ependymoma, schwannoma, and so forth (9). Myxopapillary ependymomas are the most common tumor in the cauda equina. Myxopapillary ependymoma is known to originate from the glial cells of the filum terminale and accounts for 13% of all spinal cord ependymoma (10). Typically, myxopapillary ependymoma appears as a sausage-shaped mass on the cauda equina with T1 hypointensity, T2 hyperintensity, and intense contrast enhancement (10).

Schwannomas are benign nerve sheath tumors. Schwannoma has traditionally been cited as the most common primary intradural extramedullary spinal tumor; it typically arises from the dorsal spinal nerve root and foraminal extension. Spinal cord schwannoma is rare. On MRI, schwannoma presents as a well-circumscribed T1 hypointense and T2 hyperintense mass with intense contrast enhancement. Large schwannoma lesions may show heterogeneous enhancement (10).

Paragangliomas are also spinal tumors. These are neuroendocrine tumors known to involve the adrenal gland (pheochromocytoma), carotid body (carotid body tumor), jugulotym-

panic region (glomus jugulare and glomus tympanicum), and vagus nerve (vagal paraganglioma). Spinal paraganglioma is rare and accounts for 3%–4% of cauda equina tumors. Imaging features of paraganglioma include isointensity on T1-weighted imaging, heterogeneous isointensity to hyperintensity on T2-weighted imaging, and intense contrast enhancement. Owing to high vascularity, intratumoral hemorrhage can lead to a salt-and-pepper appearance. The cap sign refers to T2 hypointensity seen along the boundary of the mass and is believed to be hemosiderin deposition (10).

In conclusion, primary T-cell lymphoma of the cauda equina is an extremely rare tumor among non-Hodgkin lymphomas. This case report presents MRI findings of primary T-cell lymphoma of the cauda equina. Differentiating between B-cell lymphoma and T-cell lymphoma in the cauda equina basis MRI findings alone is difficult. Therefore, other studies such as CSF cytology, FDG PET/CT, and ultimately tissue biopsy of the cauda equina are needed for definitive diagnosis.

Author Contributions

Conceptualization, L.G.Y., S.K., K.H.S.; data curation, K.Y., S.K., K.H.S.; formal analysis, K.H.S., K.S., L.G.Y.; investigation, K.Y., K.S.; methodology, K.Y.; project administration, L.G.Y.; resources, S.K., K.H.S.; software, K.Y.; supervision, L.G.Y.; validation, K.Y.; visualization, K.Y.; writing—original draft, K.Y., L.G.Y.; and writing—review & editing, L.G.Y.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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말총의 원발성 T세포 림프종에서 MR 영상 소견: 증례 보고와 문헌 고찰

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중추신경계의 원발성 림프종은 비호지킨림프종의 드문 형태이다. 그중에서도 말총의 원발성 T세포 림프종은 극히 드물다. 이 증례 보고는 4개월 전부터 시작된 허리통증과 하지방사통을 주소로 내원한 56세 여성의 증례에 대한 것이다. 요추 MRI에서 10 cm 크기의 길다란 다결절의 경막 내 병변이 4번 요추에서부터 2번 미추까지 있었으며 T1 강조영상에서는 등신호강도이고 T2 강조영상에서는 비균질적인 등신호강도와 고신호강도, 가돌리늄 조영증강 T1 강조영상에서는 비균질적인 강한 조영증강을 보였다. 말총 종괴에 대한 수술적 경막 내 생검을 시행하였고 면역조직화학염색과 T-cell receptor gamma 유전자 재배열 분석을 통한 진단은 말총의 말초 T세포 림프종이었다.

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