

Diffuse Large B-cell Lymphoma of the Sacral Nerve Root ; Presenting as a Polyradiculoneuropathy

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Primary spinal cord lymphomas are rare, and are either extra-/intradural masses with leptomeningeal infiltration or intramedullary in nature. The authors present a patient with a diffuse large B-cell lymphoma involving the sacral nerve root, extension to extradural space, and the cranial nerve.

KEY WORDS : Diffuse large B-cell lymphoma · Nerve root · Polyradiculoneuropathy.

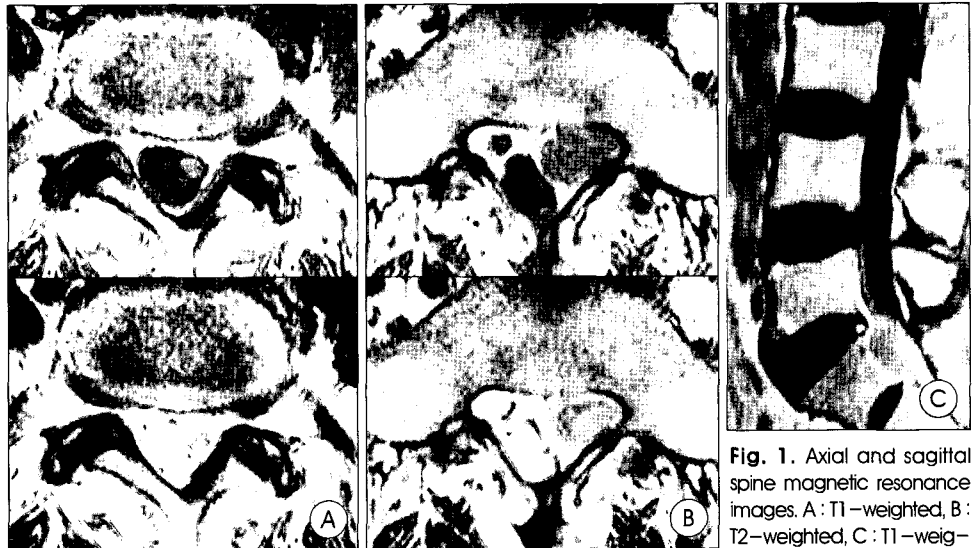
Introduction

Primary spinal cord lymphomas are rare^{4,8,9}, and either extra-/intradural masses with leptomeningeal infiltration or intramedullary in nature^{9,10}. These two patterns of growth do not generally overlap. The authors present a patient with a diffuse large B-cell lymphoma involving the sacral nerve root, extension to extradural space, and the cranial nerve. The literature regarding isolated nerve root lymphoma and lymphomatous leptomeningitis was reviewed with an emphasis on the clinical, pathological and radiological entities of nerve sheath tumor or thickened nerve root, and the general management of these diseases.

Case Report

A 60-year-old man presented with a 3-month history of intractable left leg pain and plantar flexion weakness of

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hted after gadolinium administration. Note, S1 nerve root lesion.

Fig. 1. Axial and sagittal spine magnetic resonance images. A : T1-weighted, B : T2-weighted, C : T1-weighted after gadolinium administration. Note, S1 nerve root lesion.

foot for one month. He had taken analgesics for his leg pain at a private clinic, but there was no response to them.

History and examination

On physical examination, the patient did not show organomegaly, peripheral lymphadenopathy or any cutaneous lesions. The cranial nerve examination was remarkable for a dysfunction of right third cranial nerve. He demonstrated a left positive straight leg-raising test at 45 degrees, motor weakness of plantar-flexion, diminished sensation to the pinprick test on the S1 dermatome and diminished ankle jerk on the left side. Routine laboratory tests and chest radiograph were normal. Computerized tomography after myelogram and magnetic resonance images showed a S1 nerve root



Fig. 2. Axial and coronal brain T1-weighted magnetic resonance images after administration of gadolinium. Note right oculomotor enhancement at cisternal portion.



Fig. 3. Intraoperative photograph showing the S1 nerve root and extradural component. Note check-marker delineating the S1 nerve root from extradural component.

at the cisternal portion on T1-weighted gadolinium images (Fig. 2).

Operation

An L5-S2 laminectomy was performed. The lesion was found to involve the left S1 nerve root. The bony neural foramen was somewhat widened, but grossly intact.

Foraminotomy and facetectomy delineated the nerve root lesion that was intradurally and extradurally located (Fig. 3).

On opening the dura, the transdural extension of the lesion became obvious and the nerve root was concentrically enlarged. Accordingly, the mass lesion including the extradural space was removed.

Histopathologic examination

The mass was tan colored and measured $1.2 \times 1.6 \times 4.5$ cm. A histologic examination revealed a diffuse infiltrate of monomorphic large neoplastic lymphoid cells. The nuclei were mainly round, with vesicular chromatin and membrane-bound nucleoli. Numerous mitotic figures and abundant karyorrhectic debris were also noted (Fig. 4). Immunohistochemical staining of paraffin-embedded tissue revealed that the neoplastic lymphoid cells were positive for the pan-B cell markers, CD20 and CD79a (Fig. 5A), but negative for the T-

lesion, which was hypointense on T1-weighted images and mild enhancement after gadolinium administration (Fig. 1). The brain magnetic resonance images showed an enhanced right oculomotor nerve

cell markers, UC-HL1, CD3 and CD5 (Fig. 5B).

In addition, the neoplastic cells were negative for CD30.

Taken together, these histologic findings and the immunohistochemical results were compatible with diffuse large B-cell lymphoma.

Postoperative course

Postoperatively the patient was promptly relieved of intractable left leg pain. His plantar flexion weakness still remained, but there was no further developed motor weakness. The patient was not immunocompromised, and a test for human immunodeficiency virus proved negative. The MR images of brain were interpreted to be consistent with lymphoma. So, we started up systemic combination chemotherapy (CHOP : Cyclophosphamide, Doxorubicin, Vincristine, Prednisone) and craniospinal irradiation (6000cGy for whole brain & lumbosacral area, each other). He was well after treatment, but died of sepsis after 8 months.

Discussion

In this case, the clinical features were those of polyradiculoneuropathy, while the computed tomography and MR images of the spine favored a nerve sheath tumor. However, the MR images of the spine and brain suggested that the lesions might represent a metastasis. The operative findings demonstrated a partly intradural and extradural tumor that appeared to have arisen from the S1 root, and which had the typical gross appearance of a neurofibroma or a form of lymphomatous leptomeningitis. However the histologic

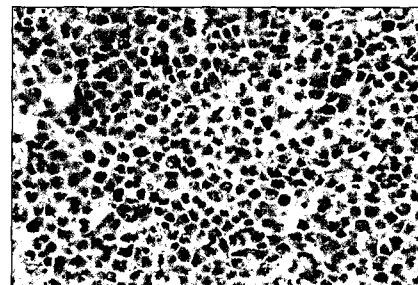


Fig. 4. Photomicrograph. Diffuse proliferation of large lymphoid cells with monomorphic round to oval nuclei (H & E, original magnification X 400).

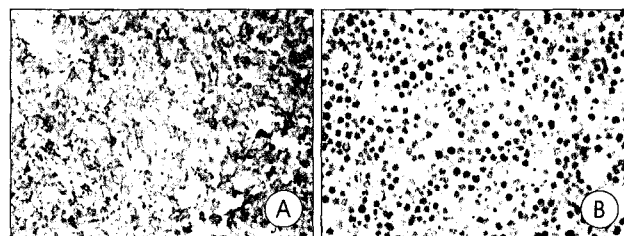


Fig. 5. Photomicrographs. A : Large lymphoid cells marked by CD20. (X 400), B : A few CD3 positive T-lymphocytes are shown whereas the large tumor cells are completely negative for CD30 (X 400).

Lymphoma of Sacral Root

diagnosis was diffuse large B-cell lymphoma. The intra- and extradural extension of a lesion confined to the nerve root, observed at surgery, is our finding for a lymphoma involving the spine. Recently, Viswanathan et al¹¹⁾ reported a similar case affecting a lumbar nerve root with vertebral body.

Although primary extradural lymphoma and extradural extension of retroperitoneal lymphoma through the neural foramina are not uncommon, intradural involvement is more characteristically observed in lymphomatous leptomeningitis, which affects multiple spinal and cranial nerve roots^{2,3,5)}. The tumor described here had some characteristic features, as follows; the involvement of the spinal root with extradural extension, the lack of involvement of the surrounding soft tissue or of the bone of the neural canal, and cranial nerve deposit, resembling aspects of lymphomatous leptomeningitis^{2,3,5)}. The prognosis for primary CNS lymphoma without treatment is poor, and median survival is 4.6 months⁵⁾. The initial treatment of primary CNS lymphoma for all patients, including the elderly, should be chemotherapy using a CHOP(Cyclophosphamide, Doxorubicin, Vincristine and Prednisone) or high-dose methotrexate-based regimen. Although cranial irradiation has often been combined with methotrexate, the unacceptably high incidence of late neurotoxicity, i.e. white matter lesions associated with severe cognitive dysfunction, particularly in older patients, has caused many to eliminate radiotherapy, especially in those older than 60 years^{1,7)}. Radiation therapy can salvage for whom relapse after chemotherapy¹⁾. Intrathecal methotrexate is frequently used in combination with high-dose methotrexate, but its benefits are not established except in case of positive-cerebrospinal fluid^{5,6)}. Operative intervention in such cases may be avoided if a definitive diagnosis is made by biopsy of relevant tissues, based on radiological clues. Initially, we might have chosen systemic chemotherapy after obtaining a histologic diagnosis through biopsy. However, we adopted operation for following reason; the tumor mass resembled a form of nerve sheath tumor based on spinal MR images, causing intractable leg pain and motor weakness.

Although his brain MR images could be interpreted to be consistent with lymphoma and suggests metastasis, we did not confirm the exact primary origin of the lymphoma in this patient. We report a case of sacral nerve root lymphoma extending to the extradural space, and presented as a polyradiculoneuropathy.

Conclusion

In conclusion, involvement of a spinal nerve root by lymphoma, although rare, should be suspected in all cases of nerve sheath tumor.

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