Solitary intradural extramedullary lymphoma of the cervical spine

Case report

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The authors report on the case of a 64-year-old man with solitary intradural extramedullary non-Hodgkin lymphoma of the cervical spine. The lesion mimicked the appearance of meningioma on MR imaging. Positron emission tomography showed increased accumulation of fluorine-18–labeled fluorodeoxyglucose only in the cervical lesion. Serum levels of C-reactive protein and soluble interleukin-2 receptor were mildly elevated. At surgery, the intradural tumor in the subarachnoid space was totally extirpated. Based on histopathological findings, diffuse, large B-cell type non-Hodgkin lymphoma was diagnosed. Postoperatively, the patient was treated with 2 courses of chemotherapy by intrathecal injection of methotrexate, cytarabine, and prednisolone and 4 courses of intravenous rituximab, an antibody binding to CD20 on the surface of B cells. All preoperative symptoms completely resolved after surgery. Two years postoperatively, the patient was faring well with no evidence of local recurrence or new lesions at any other site. To the best of the authors’ knowledge, this case is the first reported instance of solitary intradural extramedullary non-Hodgkin lymphoma of the cervical spine. (DOI: 10.3171/2009.11.SPINE08735)

Key Words • intradural tumor • cervical spine • lymphoma • non-Hodgkin disease

Non-Hodgkin lymphomas involving the spine and/or spinal cord are not rare. In previous reports, however, almost all lesions have been epidural or intramedullary, presenting as part of systemic lymphoma or direct invasion from a paravertebral lesion.3,5 To the best of our knowledge, no previous reports have documented cases with solitary intradural extramedullary non-Hodgkin lymphoma of the cervical spine. In the present report we describe the clinical and radiographic features of an extremely rare case of solitary intradural extramedullary non-Hodgkin lymphoma of the cervical spine.

Case Report

History and Examination. This previously healthy 64-year-old man presented with a 5-month history of intolerable right neck and shoulder angle pain, progressive motor weakness of the right upper and lower extremities, and clumsiness of bilateral hands. Physical examination revealed right dominant hypesthesia of the upper extremities. Right upper-extremity muscles displayed Grade 2/5 power. Grip strength of the right hand was decreased to 3 kg. All deep tendon reflexes were exaggerated, and a positive Hoffmann reflex was identified bilaterally. Gait was severely spastic. The patient reported mild dysuria and constipation. Neither eruptions nor lymphadenopathy were present.

Preoperative radiographs of the cervical spine showed neither osteolytic changes nor scalloping. Magnetic resonance imaging demonstrated a homogeneous mass encroaching on the cord at the C3–4 level: T1-weighted images showed an intradural tumor, homogeneous and isointense compared with muscle (Fig. 1A), and T2-weighted images demonstrated a homogeneous tumor hyperintense to muscle (Fig. 1B). The tumor displayed homogeneous enhancement on contrast-enhanced MR images (Fig. 1C) and appeared to be widely based in the dura mater (Fig. 1D). These MR imaging findings indicated a lesion mimicking the appearance of a meningioma. Positron emission tomography showed increased accumulation of FDG only in the cervical lesion. The SUV max of the cervical lesion was 3.04 (Fig. 2). A blood workup demonstrated a slightly increased leukocyte count (9260/μl) and an elevated C-reactive protein serum level (5.0 mg/dl). The level of sIL-2R was mildly elevated to 813 U/ml. Given these results, the preopera-

Abbreviations used in this paper: FDG = fluorine-18–labeled fluorodeoxyglucose; sIL-2R = soluble interleukin-2 receptor; SUV max = maximum standardized uptake value.
The differential diagnosis was meningioma, lymphoma, or inflammatory process.

**Operation.** Total extirpation of the tumor was performed through an open-door laminoplasty from C-3 to C-4. An intradural tumor in the subarachnoid space seemed to adhere to one of the right C-4 dorsal nerve rootlets. Gross findings revealed a firm, yellowish-white, avascular tumor that was encapsulated. The lesion involved the ventral dura mater but had not expanded to the extradural space (Fig. 3). Intraoperative examination of a frozen section showed malignancy comprising small round cells.

**Pathological Examination.** Histopathological examination showed a diffuse population of large lymphoid cells with atypical nuclei on staining with H & E (Fig. 4 upper). Immunohistochemical studies demonstrated a diffuse population of large cells with strong positivity for the B-cell marker CD20 (Fig. 4 lower) and negativity for the T-cell marker CD3. Based on these histopathological findings, diffuse, large B-cell type non-Hodgkin lymphoma was diagnosed.

**Postoperative Course.** Computed tomography studies of the chest, abdomen, and pelvis and a bone marrow biopsy procedure were performed postoperatively for staging evaluation. The CT scans showed no apparent lymphadenopathy. No heterogenic cells were found in the bone marrow biopsy specimen. Postoperative examination of CSF revealed no heterogenic cells. The cervical lesion was thus considered to represent the solitary and primary expression of non-Hodgkin lymphoma in this case. The patient was treated with 2 courses of chemotherapy consisting of intrathecal injection of methotrexate, cytarabine, and prednisolone and 4 courses of intravenous rituximab, an antibody binding to CD20 on the surface of B cells.

After surgery, all preoperative symptoms completely resolved. At 2 years postoperatively, the patient was faring well with no evidence of local recurrence or new lesions at any other site.

**Discussion**

Non-Hodgkin lymphoma involving the CNS is not rare, with a reported incidence of 5–11% of cases. Spinal involvement occurs as a metastatic lesion in the advanced stages in 0.1–5.9% of patients with non-Hodgkin lymphoma, although primary intramedullary spinal cord non-Hodgkin lymphoma has been described in a few case reports. Conversely, intradural extramedullary lymphoma has rarely been reported. To the best of our knowledge, only 2 reports have documented an intradural extramedullary lesion. However, in 1 of these 2 cases, whether the tumor was solitary or part of systemic lesions was not clarified. Although Heran et al. reported the first case of primary intradural extramedullary Hodgkin lymphoma of the cervical spine, mediastinal and parapharyngeal lymphadenopathies were seen in that case. Thus, the present case is the first instance of presumed solitary intradural extramedullary lymphoma of the cervical spine.

In the present case, an intradural tumor in the subarachnoid space seemed to adhere to one of the right C-4 dorsal nerve rootlets and showed involvement of the ventral dura mater with continuity; however, the lesion had not extended to the extradural space, as confirmed intraoperatively and on CT and MR imaging. Intraoperative findings suggested that the tumor might arise from the dura mater. The tumor had an encapsulated form, although a lymphoma is usually not encapsulated. Nevertheless, a study of primary lymphoma of the lacrimal gland has shown that excised lymphomas appeared en-
capsulated in all 13 patients. Therefore, some lymphomas can have an encapsulated form.

Regarding MR imaging, quite similar findings were shown in the present case and the case described by Heran et al. both tumors were homogeneously enhanced lesions widely based in the dura mater, and the borderline between tumor and dura mater showed nearly right angles, not sharp angles. Based on these findings, the lesion mimicked the appearance of a meningioma. Conversely, a study of MR imaging features in primary lymphoma of the CNS showed that 100% of lesions were iso- or hypointense on T1-weighted images, 47% were hyperintense on T2-weighted images, and 74% had a homogeneous enhancement pattern. The signal pattern in the present case was considered typical of lymphoma. Therefore, the possibility of lymphoma should be considered when an intradural tumor mimics the appearance of a meningioma on MR imaging, although the number of patients with such findings has thus far been limited to only 2.

For preoperative differential diagnosis, several investigators have reported high sensitivity of FDG-PET for lymphoma. These studies have shown sensitivities of 62–100%. The mean SUV_{max} for non-Hodgkin lymphoma is reportedly 3.2–43.0. The SUV_{max} is reportedly associated with tumor grade, indolent or aggressive nature, and histology. High-grade lymphomas display higher FDG uptake (SUV_{max} 17.2 ± 9.7) than low-grade tumors (SUV_{max} 6.7 ± 2.9). Regarding diffuse large-cell lymphoma classified as high-grade tumor, the SUV_{max} of these lesions has been reported as 3.2–35.0. In the present case of diffuse large B-cell lymphoma, the SUV_{max} of the lesion (3.04) did not contradict these earlier findings, although this score was within the lower reported range.

As a useful marker for the diagnosis of lymphoma, serum sIL-2R levels are reportedly elevated in patients with lymphoma and are influenced by the stage and cell type of the tumor. In a previous report, the median serum sIL-2R level was 617 U/ml in Stages 1 and 2, compared with 2188 U/ml in advanced Stage 3 or 4. The mean serum sIL-2R level of T-cell lymphoma is reportedly higher (3260 U/ml) than that of B-cell lymphoma (1473 U/ml).
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In the present case, the serum sIL-2R level (813 U/ml) was considered typical of Stage 1 B-cell lymphoma. Altogether, the possibility of lymphoma must be considered when an intradural tumor mimics the appearance of meningioma on MR imaging, and FDG-PET and an evaluation of the serum sIL-2R level should be performed to facilitate the differential diagnosis before surgery.

**Disclosure**

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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