A CASE OF PRIMARY SPINAL INTRAMEDULLARY LYMPHOMA

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A 41-year-old male was admitted to our clinic with a 1-month history of left hemiparesis. He had marked left arm weakness. The diagnostic work-up revealed an intramedullary mass at spinal level C2-4. Laminectomies were performed at C2-3-4 and the tumor was subtotally resected. Histological examination identified the mass as a non-Hodgkin’s diffuse B-cell lymphoma. The patient was treated with corticosteroids, chemotherapy, and adjuvant radiotherapy. The residual tumor tissue had completely disappeared by 6 months of follow-up; however, the patient presented with intraventricular metastasis at 11 months postsurgery. © 2001 by Elsevier Science Inc.

KEY WORDS
Primary spinal lymphoma, non-Hodgkin’s lymphoma, intramedullary, chemotherapy, radiotherapy, recurrence.

Non-Hodgkin’s lymphomas are rare tumors that account for only 0.3% to 1.5% of all central nervous system neoplasms (CNS) [1,4,6,11]. To date, only 11 cases of primary intramedullary spinal cord lymphoma have been reported in the literature [4,7–9,12,14–17,18]. Here we report an unusual case of primary spinal intramedullary lymphoma.

CASE REPORT

A 41-year-old male was admitted to our clinic with left hemiparesis of 1 month’s duration and marked left arm weakness. The patient was able to walk, his deep tendon reflexes were hyperactive, and there was a positive Babinski’s sign on the left. Spinal magnetic resonance imaging (MRI) results showed enlargement of the spinal cord and an intramedullary mass between C2 and C4 (Figure 1A). We performed bilateral laminectomies at vertebras C2, C3, and C4, and exposed a yellowish-white intramedullary tumor. Only subtotal resection was possible because of the invasive nature of the mass. Small tumor nodules that were observed on the exposed spinal cord and some of the visible spinal nerve roots were noted as metastatic seeding.

The pathology report identified the tumor as a non-Hodgkin’s diffuse B-cell lymphoma (Figure 1B). The patient exhibited no additional neurological deficits in the early postoperative period, and was prescribed 32 mg/day dexamethasone to be taken in four equal doses. Contrast-enhanced (Gd-DTPA) MRI conducted immediately after surgery showed residual tumor tissue at C3 and C4. There were no abnormal findings on contrast-enhanced computed tomography (CT) and MRI studies of the thoracic and lumbosacral spinal cord, cranium, abdominopelvic area, and chest. Histopathological examination of a bone marrow aspirate and biopsy were also normal. Biochemical testing of the patient’s cerebrospinal fluid revealed slightly elevated glucose (151 mg/dL) and protein (440 mg/dL) levels. Hematological investigations including complete blood count, erythrocyte sedimentation rate, blood chemistry, and protein electrophoresis were normal, and the patient tested negative for human immunodeficiency virus. The diagnosis was primary intramedullary lymphoma of the cervical spinal cord.

Based on the diagnosis and the fact that there was residual tumor tissue, the patient was started on a chemotherapy protocol of vincristine (1.4 mg/m² i.v., first day of the cycle only), doxorubicin (50 mg/m² i.v., first day only), cyclophosphamide (750 mg/m² i.v., first day only), and prednisolone (100 mg per os, days 1–5). The chemotherapy protocol was repeated every 21 days for 6 cycles. After the fourth cycle, adjuvant radiotherapy was administered in the cervical region, with a total radiation dose of 4500 cGy. The remaining two chemotherapy cycles were completed after radiotherapy.
### Summary of Previously Reported Primary Intramedullary Lymphoma Cases

<table>
<thead>
<tr>
<th>Author</th>
<th>Age</th>
<th>Sex</th>
<th>Location of Tumor</th>
<th>Cell Types</th>
<th>Radiological Examination</th>
<th>In Vivo Diagnosis</th>
<th>Extramedullary Involvement</th>
<th>Extracerebral Involvement</th>
<th>Treatment</th>
<th>Post Mortem Examination</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bruni et al (1977)</td>
<td>53</td>
<td>M</td>
<td>C1-T1</td>
<td>Lymphocytes, mononuclear cell, some plasma cells, histiocytes</td>
<td>Myelography</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>+</td>
</tr>
<tr>
<td>Herbst et al (1976)</td>
<td>51</td>
<td>M</td>
<td>T12-L1</td>
<td>Mixed histiocytic lymphoma, lymphoblastic lymphoma</td>
<td>Myelography</td>
<td>Biopsy</td>
<td>Cerebellum, brainstem</td>
<td>—</td>
<td>Irradiation; Chemotherapy</td>
<td>−</td>
</tr>
<tr>
<td>Fisher (1979)</td>
<td>51</td>
<td>F</td>
<td>T1-T2</td>
<td>Lymphoblastic lymphoma</td>
<td>Myelography</td>
<td>Biopsy</td>
<td>Pons, midbrain</td>
<td>Supravacular node 3 yrs before</td>
<td>Irradiation; Chemotherapy</td>
<td>−</td>
</tr>
<tr>
<td>Slager et al (1982)</td>
<td>45</td>
<td>F</td>
<td>C1-C5</td>
<td>Mononuclear cell, some plasmocytoid cells</td>
<td>Myelography</td>
<td>—</td>
<td>Midline cerebellum</td>
<td>—</td>
<td>—</td>
<td>+</td>
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<tr>
<td>Mitsumoto et al (1980)</td>
<td>60</td>
<td>F</td>
<td>L1-L5</td>
<td>Diffuse histiocytic lymphoma</td>
<td>Myelography</td>
<td>Biopsy</td>
<td>Parietooccipital lobe</td>
<td>—</td>
<td>Irradiation; Chemotherapy</td>
<td>+</td>
</tr>
<tr>
<td>Hautzer et al (1983)</td>
<td>26</td>
<td>F</td>
<td>T8-L2</td>
<td>Lymphocytes, mononuclear cell, plasmocytes, histiocytes</td>
<td>Myelography</td>
<td>—</td>
<td>Optic nerve, thalamus</td>
<td>—</td>
<td>—</td>
<td>+</td>
</tr>
<tr>
<td>Slowik et al (1990)</td>
<td>49</td>
<td>F</td>
<td>C3-C6</td>
<td>Non-Hodgkin’s lymphoma (no definite type)</td>
<td>CT myelography</td>
<td>Biopsy</td>
<td>—</td>
<td>—</td>
<td>Irradiation; Chemotherapy</td>
<td>−</td>
</tr>
<tr>
<td>Wong Chung et al (1991)</td>
<td>56</td>
<td>M</td>
<td>Conus</td>
<td>Non-Hodgkin’s lymphoma</td>
<td>CT myelography</td>
<td>—</td>
<td>Lymph node 6 months before</td>
<td>—</td>
<td>Irradiation; Chemotherapy</td>
<td>−</td>
</tr>
<tr>
<td>Bekar et al (Present report)</td>
<td>41</td>
<td>M</td>
<td>C2-C4</td>
<td>B-cell, non-Hodgkin’s lymphoma</td>
<td>CT MRI</td>
<td>Biopsy</td>
<td>—</td>
<td>—</td>
<td>Subtotal resection, Irradiation, Chemotherapy</td>
<td>−</td>
</tr>
</tbody>
</table>
At 6 months after surgery, contrast-enhanced cervical MRI showed no residual intramedullary lesion (Figure 1C). By the ninth month, the patient’s hemiparesis had improved and he was showing no additional neurological deficits. However, at 11 months postsurgery he was admitted to the emergency department with sudden-onset motor aphasia and tetraparesis. Contrast-enhanced cranial MRI revealed small tumor nodules in the third and fourth ventricles (Figure 1D). Spinal MRI at this stage showed no lesion.

**Discussion**

Less than 1% of all non-Hodgkin’s lymphomas are intramedullary tumors [1,6,13]. Individuals at particularly high risk for developing CNS lymphoma include transplant recipients and patients with acquired immunodeficiency syndrome, congenital immune deficiency, or Epstein–Barr virus infection [2,10,11]. Most intramedullary non-Hodgkin’s lymphoma lesions are found in the upper thoracic or lower cervical regions of the spinal cord [1,13]. Contrast-enhanced CT or MRI should be done at the first sign of neurologic symptoms. Even in an immunologically normal patient, when unexpected remission is observed on CT or MRI after steroid treatment, the diagnosis of primary central nervous system lymphoma (PCL) must be considered [3,11]. Management of PCL is based on histologic diagnosis [1].

The conventional treatment methods of irradiation and corticosteroid therapy often achieve complete remission in cases of primary CNS non-Hodgkin’s lymphoma, but the average patient survival time is only 10 to 18 months. Corticosteroid treatment (24 mg/day dexamethasone divided in three doses) has been shown to reduce tumor size and decrease morbidity [1,5]. Chemotherapy, another of the conventional methods, can lead to remission but does not cure [9,11]. One report also described success in treating subarachnoid tumor deposits with intrathecal administration of methotrexate [1]. The apparent tumor seeding we observed during surgery made us suspicious that the detected mass might represent metastasis from another site in the patient’s spinal cord; however, we found no other tumor. On this basis, we diagnosed and treated the detected mass as primary lymphoma of the spine. Even though the patient completed six cycles of chemotherapy and underwent follow-up radiation treatment, he went on to develop intraventricular metastasis.

Surgical debulking was carried out in the case presented in this report, but we believe that the most important steps in the approach to spinal cord lymphoma are biopsy and histological diagnosis, followed by chemotherapy, adjuvant radiotherapy, and corticosteroid administration. The physician should always maintain a high index of suspicion for recurrence in any case of CNS non-Hodgkin’s lymphoma.

**REFERENCES**

COMMENTARY

This is an interesting case report on a primary spinal intramedullary lymphoma, which is a rare lesion in the spinal cord. Again, we can appreciate the need for histological diagnosis of any intraspinal cord tumor, because MRI cannot determine it with certainty.

I am puzzled by Figure 1C, in which the authors combined a T2-weighted image with intravenous Gd-DTPA. This is very unusual—we have no experience with this MRI sequence, and neither does our neuroradiologist.

Lymphomas are infiltrative tumors. The strategy should be to obtain a biopsy without any attempt at removal, followed by adjunctive chemotherapy, as is conducted for brain lymphoma. This would have been our policy in such a case. I assume that the authors gave radiation therapy after four cycles of intravenous chemotherapy because they did not observe complete remission on MRI. We would question the use of intrathecal chemotherapy. In brain lymphomas, intraventricular chemotherapy is given through a permanent catheter via an Ommaya reservoir; this method should also be used in intramedullary lymphomas, through either a ventricular or a lumbar route, or both.

Finally, I suspect that this patient died shortly after the discovery of the intracranial metastases. A postmortem examination would have been extremely interesting in this case of a rare intramedullary lesion.

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