Primary intradural extraarachnoid Hodgkin lymphoma of the cervical spine

Case report

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This 42-year-old man presented with an intradural extramedullary mass in his lower cervical spine. On imaging studies the lesion mimicked the appearance of a meningioma. At surgery, the mass was found to be an intradural extraarachnoid tumor. An intraoperative pathological examination determined the lesion to be a Hodgkin lymphoma. The procedure was limited to biopsy sampling and the patient was treated further with adriamycin, bleomycin, vincristine, and dacarbazine chemotherapy, after which remission was demonstrated both clinically and on images. Analysis of a frozen section obtained during the procedure aided in the diagnosis of the tumor, thus preventing further resection and the potential neurological complications associated with more radical resection. To the authors’ knowledge this is the first reported case of a presumed primary intradural extraarachnoid Hodgkin lymphoma.

KEY WORDS • spinal cord compression • intradural tumor • cervical spine • lymphoma • Hodgkin disease

Abbreviation used in this paper: CT = computed tomography.
The plantar responses were downgoing. Sensory testing revealed reduced pain and light-touch perception in the C-5 and C-6 dermatomes on the left side and the C-8 and T-1 dermatomes on the right. The remainder of the neurological examination revealed normal findings.

Routine blood workup demonstrated no abnormality. The peripheral blood counts were within the normal range and had a normal differential. Oblique cervical radiographs as well as the initial chest x-ray study showed no abnormality. Magnetic resonance imaging of the spinal cord revealed an asymmetrical intradural, extraaxial homogeneously enhancing lesion extending from C-5 to T-1. There was no evidence of bone involvement or extraradicular disease. Involvement of the dura mater was present with appreciable circumferential enhancement around the cord. The lesion appeared to encase the anterior and left side of the spinal cord, and appeared to be based in the dura mater (Fig. 1). The preoperative differential diagnosis was that of an unusual dura-based lesion, possibly a meningioma, metastasis, lymphoma, or inflammatory process.

Operation. A C-6 laminectomy and biopsy sampling of the lesion were performed on March 19, 2001. At surgery, a firm, yellowish-gray, relatively avascular, friable tumor in the intradural extraarachnoid space was removed easily from the adjacent tissues. The surrounding arachnoid and dura mater appeared normal as did the cord and cerebrospinal fluid. There was significant mass effect on the spinal cord, but no obvious changes in the bone to suggest bone involvement or a long-standing process. Examination of a frozen section determined the diagnosis to be a Hodgkin lymphoma (Fig. 2). The procedure was then terminated and the wound closed.

Pathological Examination. Permanent histological preparation and examination by a neuropathologist and hematopathologist confirmed a lymphoproliferative neoplasm characterized by a prominent sclerosing background and heterogenic population of lymphoid cells. We observed large cells with prominent nucleoli, several classic Reed–Sternberg cells (Fig. 3A), smaller cells with morphological characteristics of reactive lymphocytes, and eosinophils scattered throughout the tumor and stroma. Immunohistochemical studies confirmed the large cells to be strongly immunoreactive for CD30 (Fig. 3B), and many of the reactive-appearing lymphocytes were immunopositive for T-cell markers (MT1 and CD3) (Fig. 3C). A small number of lymphoid cells were immunoreactive for L26. Very few tumor cells were immunoreactive for muramidase and cells occasionally stained positive for myeloperoxidase. The cells displaying eosinophil-like characteristics were immunopositive for chloracetate esterase. These findings confirmed the diagnosis of a Hodgkin lymphoma.

Treatment. Postoperatively, the patient commenced treatment with high-dose dexamethasone for his neurological symptoms. A staging assessment was performed, which included a chest x-ray study; CT scans of the chest, abdomen, and pelvis; a bone scan; a bone marrow biopsy procedure; and a hematological consultation and workup. The CT scan demonstrated a mediastinal lymph node on the left side measuring 10 mm in size in the aorticopulmonary window as well as several small left paratracheal nodes all smaller than 10 mm in size. Evaluation of bone marrow aspirates of the iliac crests revealed the absence of Hodgkin disease. On the basis of this assessment, the Hodgkin lymphoma was classified as Stage IVA, and the spinal lesion was believed to be the primary tumor given its nature as the predominant focus of disease. The patient

![Fig. 1. Magnetic resonance images.](image-url)
Primary intradural Hodgkin lymphoma

then began a regimen of adriamycin, bleomycin, vincristine, and dacarbazine chemotherapy.

Postoperative Course. The patient’s neurological symptoms rapidly improved and eventually resolved. Following the completion of the planned six courses of chemotherapy, clinical evaluation and repeated staging studies, including serial MR imaging of the cervical spine, performed at 6-monthly intervals demonstrated complete clinical and imaging remission at the 4-year follow-up examination.

Discussion

Spinal cord compression due to lymphoproliferative neoplasms is much more frequently caused by non-Hodgkin lymphoma than Hodgkin disease. Primary intramedullary spinal cord lymphoma has been described in a number of case reports, and all have been of the non-Hodgkin variety. A review of the literature revealed only a few cases of spinal cord compression due to primary Hodgkin lymphoma, and all lesions so far have been extradural in origin. The reported incidence of spinal cord compression caused by epidural disease ranges from 1.15 to 7.6%. The incidence of epidural spinal cord compression as the initial manifestation of Hodgkin disease and the only site of clinical involvement is approximately 0.2%. Secondary intradural spread, either intramedullary or leptomeningeal, is rare in Hodgkin disease, usually presenting late after systemic therapy, and is associated with a poor prognosis. There have been no reports of primary Hodgkin lymphoma located in the intradural compartment. It is difficult to comment on the cell of origin in this location because no aggregates of lymphoid tissue are present within the intradural compartment. The mechanism may not be dissimilar from the development of primary central nervous system lymphoma, which in itself is debatable.

In one large retrospective review of 426 patients with Hodgkin lymphoma, compression of either the spinal cord or nerve roots was demonstrated in 25 but in only two of them was the spine problem the presenting complaint. None of the patients in that study had intradural lesions. The presenting feature in patients with spinal or radicular involvement was localized back pain before the onset of long tract disturbances. Our patient experienced a similar clinical course. In most cases of spinal cord compression due to advanced Hodgkin disease, spinal involvement was noted approximately 4 years after the initial diagnosis, lending further support to our presumption that the spinal disease was the primary focus.

The present report represents the first case of a primary intradural extradural Hodgkin lymphoma. The patient presented with radicular symptoms and subsequent spinal cord compression. An intradural extramedullary lesion with an apparent dura-based extension was visible on imaging. Because the preoperative diagnosis was not certain, we proceeded to undertake only a limited exposure with a plan to attain a biopsy sample and perform analysis of a frozen section. The intraoperative and subsequent permanent sections were diagnostic of a Hodgkin lymphoma with the presence of typical Reed–Sternberg cells. Based on the intraoperative pathological findings, the surgical procedure was terminated without proceeding and further radical resection was not undertaken, thereby avoiding the potential for morbidity associated with nerve root or cord injury.

Within the spinal cord, Hodgkin lymphoma appears to spread directly among structures such as paravertebral masses, along the neural foramina, or intradural metastases. This differs from the hematopoietic

![Fig. 2. Photomicrograph of a frozen section obtained during tumor biopsy. A heterogenic population of lymphoid cells with prominent Reed–Sternberg cells is seen (arrows). Original magnification × 40.](image)

![Fig. 3. Photomicrographs of H & E–stained permanent sections obtained during the tumor biopsy procedure. A: A prominent sclerosing background and heterogenic population of lymphoid cells are demonstrated. Several classic Reed–Sternberg cells are identified (arrows). Smaller cells have the morphological characteristics of reactive lymphocytes. B: The larger cells are strongly immunoreactive for CD30. C: Staining for the T-cell marker CD3 demonstrates immunopositivity among the smaller reactive-appearing lymphocytes. Original magnification × 40.](image)
spread to the vertebrae that is characteristic of other tumors.\textsuperscript{1,2,7,9,11,16,20} In our case the initial onset of radicular pain may have been associated with the tumor arising within the root sleeve and extending farther intradurally. There was no evidence of paravertebral or foraminal disease in this case, or a primary tumor elsewhere. Given the mechanism of spread, plain radiographs and CT scans may not reveal findings of Hodgkin lymphoma manifesting in patients who present with spinal cord compression because bone invasion is infrequent.\textsuperscript{5,8,13} Magnetic resonance and CT myelography are the most sensitive imaging modalities for demonstrating tumor impinging on neural elements.\textsuperscript{8,13}

Although an intradural extramedullary location for the initial presentation of Hodgkin disease is rare, the management of the disease is similar to that of Hodgkin lymphoma found at other sites. In the latter cases, there is no role for resection aside from tumor biopsy procedure to establish a diagnosis. In cases of Hodgkin disease in which there is extradural extension, good response to radiation therapy and chemotherapy is reported and is associated with a high rate of good functional recovery (86%), complete response (61%), and long-term survival.\textsuperscript{9} For the unusual case in which the patient presents with spinal cord compression as the initial symptom of Hodgkin disease, the therapeutic options include chemotherapy alone, radiation therapy alone, or combined-modality therapy once the diagnosis is known and the appropriate staging evaluation has been completed.\textsuperscript{9} Because of the leptomeningeal blood supply to the tumor, it is not clear whether the blood–brain barrier may pose a hindrance to the penetration of chemotherapeutic agents. Consideration of alternative treatment regimens may thus be necessary if initial agents fail.\textsuperscript{3}

Conclusions

Hodgkin disease may rarely present as an intradural extramedullary mass and may resemble an intradural meningocele, which may present with spinal cord compression due to extradural content.\textsuperscript{10} Although the primary involvement of the spinal cord is rare, the potential for significant neurological deterioration is high. In Hodgkin disease, a preoperative pathological examination is advocated and results from Hodgkin’s disease with chemotherapy. A report of two cases and a review of the literature. Am J Med 84: 555–558, 1988


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References


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