Hodgkin’s Lymphoma Presenting as Spinal Cord Compression: A Case Report

Arianna R. Gregg BS, MPH1,3, Abhijay Kumar BS2,3, Francis Joseph Lauzier BS2,3, Mallory Brosious MD3

1University of Nevada, Reno, NV, USA
2Wayne State University School of Medicine, Detroit, Michigan
3Department of Internal Medicine, Henry Ford Hospital, Detroit, MI, USA

*Corresponding Author: Arianna R. Gregg BS, MPH, Department of Internal Medicine, Henry Ford Hospital, Detroit, MI, USA. Email: ariannagregg@med.unr.edu

Abstract

Spinal cord compression is an extremely rare yet worrisome complication that may arise in individuals with Hodgkin lymphoma (HL). Spinal cord compression is found in roughly 0.2% of HL cases during the initial diagnosis. Considering HL as a rare but important differential diagnosis in cases of spinal cord compression is crucial due to the potential for prolonged time-to-diagnosis, subsequent suboptimal management and poor patient outcomes. We present the case of a rare yet severe initial presentation of spinal cord compression in a patient with advanced nodular sclerosis HL. We aim to increase awareness, diagnostic work up, and optimal management of advanced HL resulting in severe spinal cord compression.

Keywords: Hodgkin’s lymphoma, spinal cord compression

1. INTRODUCTION

HL is a rare monoclonal lymphoid neoplasm derived from B cells which commonly originates in the cervical lymph nodes.1 The occurrence of HL exhibits a bimodal pattern in terms of age distribution, with an initial peak among young adults aged 20-24 years and a second larger peak in incidence during the eighth decade of life.2 Classical HL is the predominant pathologic type affecting children, adolescents, and young adults. The nodular sclerosis subtype of classical HL most commonly occurs in females between the ages of 15 and 34 years.2 Over the past four decades, significant advancements in treatment have resulted in a remarkable improvement in survival rates, with HL in children and young adults attaining a 5-year survival rate greater than 95%.2

The classic presentation of HL is painless lymphadenopathy, with associated low-grade fever, night sweats, and weight loss. Approximately 80% of patients diagnosed with classical HL exhibit painless lymphadenopathy, predominantly affecting the supraclavicular and cervical lymph nodes. The clinical presentation of spinal cord compression occurs in approximately 0.2% of cases at the time of initial HL diagnosis.5

Since HL rarely presents as spinal cord compression, it is often not considered in the differential diagnosis of patient’s presenting with complaints of back pain and/or symptoms of neurological deficits. Therefore, it is crucial to increase awareness of cases with extra-nodal involvement to reduce the time it takes to diagnose the condition and minimize the occurrence of late complications. This case report highlights a rare but severe initial manifestation of spinal cord compression in a patient with advanced nodular sclerosis HL. Our objective is to raise awareness about the diagnostic workup and optimal management of advanced HL leading to severe spinal cord compression.

2. CASE DESCRIPTION

A 27-year-old-male with no significant past medical history presented to the emergency department with complaints of progressively worsening lower back pain followed by 3-4 days of acute bilateral lower extremity weakness and decreased sensation. He also endorsed fevers,
night sweats and a 50-pound weight loss over the few months prior. He denied urinary and bowel incontinence. Physical exam was significant for decreased sensation at and below the level of T4 as well as spinal point tenderness from the mid-thoracic level down to the sacrum. Cervical, bilateral supraclavicular, axillary and inguinal lymphadenopathy was also noted. Labs were significant for anemia of chronic disease, reactive thrombocytosis, elevated lactate dehydrogenase and elevated uric acid.

MRI of the entire spine demonstrated an extensive bone marrow replacing process throughout the visualized osseous structures as well as innumerable enlarged lymph nodes concerning for lymphoma. In addition, there was extensive extrasosseous soft tissue spread of disease involving the epidural, foraminal, and paraspinal soft tissues with multifocal areas of spinal cord compression and displacement, greatest at the T4 and T5 levels where there was moderate to severe cord compression and abnormal T2 hyperintense cord edema extending caudally to the stenosis, seen greatest at the T3-T4 level. There was also moderate to severe spinal canal narrowing at T10-T11 without cord signal abnormality. The constellation of findings was most suspicious for lymphoma or leukemia with extrasosseous extension.

Figure 1. Sagittal MR images. (A) T2 weighted images showing extensive extrasosseous soft tissue spread of disease involving the epidural, foraminal, and paraspinal soft tissues of the lower cervical, thoracic, and lumbar spine with multifocal areas of spinal cord compression and displacement, greatest at the T4 and T5 levels where there was moderate to severe cord compression and abnormal T2 hyperintense cord edema (A) extending caudally to the stenosis, seen greatest at the T3-T4 level. There was also moderate to severe spinal canal narrowing at T10-T11 without cord signal abnormality. (B) T1 weighted pre-contrast (C) T1 weighted post-contrast images show that the lesions are enhancing.

Figure 2. Axial MR images. (D) T1-weighted with gadolinium showing epidural lesion at T4 resulting in spinal cord compression. The lesion also extends into the foramen and paraspinal soft tissues.

Given the severe spinal cord compression seen on imaging, the patient was started on intravenous dexamethasone. Neurosurgery stated the patient was not a surgical candidate given the extent of disease. The patient underwent right axillary excisional lymph node biopsy which confirmed Classical HL, nodular sclerosing subtype; CD15 and CD30 positive.
Hodgkin’s Lymphoma Presenting as Spinal Cord Compression: A Case Report

Imaging established stage IV-B. Our initial plan was to urgently start chemotherapy inpatient, in turn reserving radiotherapy for potential use later. Unfortunately, prior to initiation of systemic chemotherapy, the patient developed acute worsening of his bilateral lower extremity weakness and subsequently underwent a short course of palliative radiotherapy (20 Gy in 5Fx) to the large, thoracic spinal-cord adjacent lymph nodes in an effort to shrink the lymphomatous disease and restore neurological function. Following radiation, an outpatient 6-cycle chemotherapy treatment of BV-AVD (Brentuximab-Vedotin conjugate, Adriamycin (Doxorubicin), Vinblastine, and Dacarbazine) was initiated (administration every 2 weeks over 6 months, 12 treatments total) with supportive tbo-filgrastim after each chemotherapy treatment. He was discharged on an oral prednisone taper. Approximately 1.5 months after discharge from the hospital, and after 2 cycles of chemotherapy, the patient had regained full strength in his bilateral lower extremities.

3. DISCUSSION

HL is a rare monoclonal lymphoid neoplasm derived from B cells which commonly originates in the cervical lymph nodes and is characterized by the presence of scattered large mononuclear Hodgkin cells and multinucleated Reed-Sternberg cells within a background of non-neoplastic inflammatory cells. It is categorized into two main pathological types: classical and nodular lymphocyte predominant. HL accounts for approximately 0.5 to 1% of adult cancers.

The majority of patients diagnosed with classical HL typically present with lymphadenopathy. The commonly affected nodal sites in classical HL include the cervical, mediastinal, supracavicular, and axillary regions. Constitutional symptoms, commonly referred to as "B" symptoms, including fever, weight loss, and drenching night sweats, are observed in approximately 40% of cases.

Spinal cord compression is observed in only 0.2% of cases at the initial diagnosis of HL and in around 6% of cases during progression of the disease. Symptoms of spinal cord compression include chronic back pain as well as sensory, motor, and autonomic neurological disorders. According to one study, spinal cord compression was found to be the primary indication of an undetected malignancy in 75% of cases. Motor deficit was the prevailing symptom among all patients, while pain was reported by 60% of them, and sphincter dysfunction was experienced by 43%.

When spinal epidural infiltration occurs in patients with HL, the thoracic segment is the most commonly affected site. Studies have suggested that the spread of disease in classical HL follows the natural direction of lymphatic flow. Extramodal involvement in HL can occur through hematogenous dissemination or more commonly develops through direct spread from retroperitoneal or thoracic lymph nodes.

In patients with HL and spinal cord compression, the priority is to restore neurologic function. Aside from emergent administration of dexamethasone, there is currently no clear consensus on the optimal treatment approach for HL presenting with spinal cord compression. The choice of treatment approach may depend on individual patient factors, extent of the disease and neurological deficits, the severity of compression and speed of neurologic deterioration.

HL with epidural involvement has shown favorable response rates to specific chemotherapy and radiotherapy treatments. Complete clinical response has been reported in 61% of cases, while functional recovery has been observed in 86% of cases. Some argue that given the positive response to chemotherapy and radiotherapy, emergency decompressive laminectomy should be reserved for cases of advanced disease or relapse, particularly when severe and progressive neurological symptoms are present. However, retrospective studies have reported that patients who underwent laminectomy experienced better neurological recovery compared to those who received exclusive chemotherapy or radiotherapy treatments.

In contrast, a study by Correale et al. found that the prognosis did not differ when comparing patients who underwent laminectomy combined with chemotherapy and radiotherapy to those who underwent only chemotherapy and radiotherapy.

4. CONCLUSION

In conclusion, our case highlights the importance of prompt diagnosis and treatment in rare cases of advanced HL presenting with severe spinal cord compression. Given the lack of consensus on optimal treatment of spinal cord
compression in those with a new diagnosis of HL, our case supports the use of radiotherapy followed by chemotherapy in cases presenting with rapidly deteriorating neurologic function.

REFERENCES


doi:10.1080/10790268.2018.1427519


