Extranodal hairy cell leukemia presenting in the lumbar spine

Case report

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The authors report on a 54-year-old man who presented with a lumbar vertebral body lesion and an adjacent epidural lesion that was found to be hairy cell leukemia (HCL). The patient presented with gradual onset of back pain and intermittent lower-extremity radicular symptoms. He did not have splenomegaly or peripheral blood count abnormalities. Admission MR imaging revealed an L-5 vertebral body lesion and a lumbar epidural lesion extending from L-3 to S-2. An [¹⁸F]fluorodeoxyglucose–PET study showed numerous sites of osseous involvement. The patient underwent minimally invasive surgical biopsy sampling of the epidural lesion. Histopathological examination revealed extranodal HCL. After treatment with a 5-day course of cladribine, the patient’s symptoms resolved, and at the 16-week follow-up visit there was no radiographic or metabolic evidence of disease. Hairy cell leukemia rarely involves neurological structures, but this patient responded well to standard treatment. This case demonstrates the value of tissue biopsy procedures instead of aggressive resection and the use of minimally invasive techniques to treat an HCL spinal lesion. (DOI: 10.3171/SPI.2008.9.10.374)

**KEY WORDS**  • epidural lesion  • extranodal disease  • hairy cell leukemia  • lumbar spine

Hairy cell leukemia is an uncommon, indolent, B-cell, lymphoproliferative disorder infrequently associated with neurological symptoms.⁴,⁶ The disease typically presents with peripheral blood cytopenias, splenomegaly, and circulating hairy cells.⁴ We report an unusual presentation of HCL with a spinal lesion in a 54-year-old man who presented with back pain and intermittent lower-extremity radicular symptoms without splenomegaly or abnormal peripheral blood counts. An MR image revealed an L-5 VB lesion and an adjacent epidural lesion extending from L-3 to S-2. After minimally invasive surgical biopsy sampling of the epidural lesion, histopathological examination revealed extranodal HCL. Although osteolytic lesions of the spinal column have been previously reported in patients with HCL,³ extranodal spinal epidural HCL causing neural compression is rare. In this report we discuss the neurological manifestations of HCL and demonstrate a favorable response to standard treatment.

Abbreviations used in this paper: HCL = hairy cell leukemia; VB = vertebral body.

**Case Report**

**History and Examination.** This 54-year-old man with a history of obesity and anxiety developed progressively severe back pain and intermittent numbness and weakness of his lower extremities over a 3-month period. He underwent lumbar spine MR imaging that revealed a heterogeneously enhancing epidural lesion extending from L-3 to S-2, with involvement of the L-5 VB (Fig. 1). Subsequent full-body CT scans did not reveal additional abnormalities; specifically, he did not have hepatosplenomegaly or adenopathy. A skeletal survey did not reveal any osteolytic lesions. As part of the staging evaluation, an [¹⁸F]fluorodeoxyglucose–PET scan was performed that showed numerous osseous lesions in the proximal femurs, right anterior iliac wing, and the L-3 and L-5 VBs. No soft-tissue lesions were identified. No abnormalities were found on laboratory evaluations, which included a complete blood count, serum chemistry panels, quantitative immunoglobulin studies, β₂-microglobulin detection, and serum and urine immunoelectrophoresis. Before presenting to our institution, the patient underwent a fine-needle aspiration of the epidural lesion that implicated...
The patient was discharged home on the same day as his surgery. His neurological status was unchanged after surgery. Histopathological examination of the tissue sample revealed a homogeneous population of intermediate-sized cells with distinct borders and a “fried-egg” morphological appearance implicating involvement by HCL. Immunohistochemical analysis showed the cells to be CD20-positive B cells that expressed tartrate-resistant acid phosphatase and CD25, consistent with a morphological diagnosis of HCL (Fig. 2). Subsequent bone marrow biopsy sampling demonstrated focal and interstitial involvement of the bone marrow by HCL. The immunophenotype of these cells, as determined by flow cytometry, was CD19-positive B cells that coexpressed CD11c, CD103, and CD25—a combination that is diagnostic of HCL.

Five weeks after surgery the patient received a 5-day course of cladribine. This treatment was complicated by neutropenic fever and Guillain–Barré syndrome, but the patient fully recovered from these complications. All neurological symptoms and back pain resolved within 1 week of therapy. Follow-up MR imaging performed 5 weeks after chemotherapy showed no evidence of lumbar disease. The PET scan findings also normalized, with no evidence of residual avid enhancement indicating the disease. A bone marrow biopsy performed 4 months post-treatment showed no definitive evidence of HCL. At 8 months of follow-up the patient remains clinically stable with normal hematological parameters and no neurological symptoms.

Operation. Because the patient had no acute or consistent neurological deficits, he underwent a minimally invasive L4–5 laminotomy for biopsy sampling of the epidural mass. The CT scan did not demonstrate evidence of L-5 VB destruction, and therefore, spinal stabilization was not required. Using fluoroscopic guidance, a 2.5-cm incision was made 2 cm off of the midline above L4–5. Serial tubular retractors were inserted over a K-wire to expose the L4–5 interlaminar space. An L4–5 laminotomy was performed and the underlying ligament was removed. Firm, pearly-white tissue was identified in the epidural space, and a biopsy specimen was obtained and sent to the pathology laboratory.

Postoperative Course. The patient was discharged home on the same day as his surgery. His neurological status was unchanged after surgery. Histopathological examination of the tissue sample revealed a homogeneous population of intermediate-sized cells with distinct borders and a “fried-egg” morphological appearance implicating involvement by HCL. Immunohistochemical analysis showed the cells to be CD20-positive B cells that expressed tartrate-resistant acid phosphatase and CD25, consistent with a morphological diagnosis of HCL (Fig. 2). Subsequent bone marrow biopsy sampling demonstrated focal and interstitial involvement of the bone marrow by HCL. The immunophenotype of these cells, as determined by flow cytometry, was CD19-positive B cells that coexpressed CD11c, CD103, and CD25—a combination that is diagnostic of HCL.

Spinal epidural neural compression is a rare occurrence in HCL. In a review of 108 patients with HCL, ~ 5% developed neurological complications from the disease. The most common cause of neurological symptoms was infection, but a single patient had VB invasion and radiculopathy from epidural spread. In another series of 116 patients with HCL, a single patient developed spinal cord compression and paralysis caused by an HCL lesion. Lytic osseous lesions of the spinal column have been reported in patients with HCL. However, these reports do not include MR imaging evaluation, and therefore, the frequency of epidural lesions associated with osseous lesions of the spinal column is unknown. The MR imaging characteristics of the epidural lesion included high-intensity signal on T1- and T2-weighted images and a heterogeneous enhancement pattern. At surgery, the lesion appeared pearly white. This appearance is different from the green color of granulocytic sarcomas (chloroma).
that occur in myeloid leukemias. The ability to detect and monitor the disease in our patient by using $^{18}$F-fluorodeoxyglucose–PET is also unique, because the avid enhancement of HCL on PET scans is not well established.

Surgical management of HCL of the spine should be tailored to the individual patient. In the present case, the diagnosis of HCL was not known at presentation, and therefore, a definitive biopsy finding was needed. As demonstrated in our case, fine-needle aspirations are often insufficient to confirm the subtype of lymphoproliferative disorder. Aggressive Surgical decompresion may be necessary for patients with acute neurological deficits, and in patients with significant VB destruction, spinal stabilization may be required. However, medical treatment of HCL is highly effective, and therefore, surgical biopsy sampling without aggressive decompression may be a reasonable option for patients without a neurological deficit at presentation.

The treatment of HCL consists of chemotherapy with purine analogs, most commonly with cladribine. In patients with a classic presentation of HCL, a single 5-day course of cladribine results in a nearly 100% overall response rate, with most patients achieving complete remission. At long-term follow-up of 10 years, approximately one-third to one-half of patients will have experienced relapse. Predictive factors for relapse include lower pretreatment hemoglobin and higher total white blood cell count. The impact of extranodal involvement on relapse risk, as in our case, is unknown due to the rarity of this presentation.

Conclusions

Hairy cell leukemia can have atypical presentations including back pain and symptoms from neural compression. An adequate biopsy specimen is critical for distinguishing HCL from other more common lymphoproliferative and plasma cell disorders. Although HCL lesions of the spine are rare, spine surgeons should be aware of this disease.

**Disclaimer**

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

**References**