Primary Malignant Lymphoma in a Spinal Cord Presenting as an Epidural Mass with Myelopathy: A Case Report

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We report the case of a 47-year-old man who presented with progressive paraparesis and sphincter changes over 2 weeks. Magnetic resonance imaging revealed a spinal epidural mass from T9 to L2. We performed a decompressive laminectomy and mass removal. The histopathology was consistent with a small lymphocytic lymphoma. No metastatic lesion was noted in the chest and abdomen-pelvic computerized tomography (CT) and positron emission tomography computerized tomography (PET-CT) scan. The final diagnosis was primary spinal lymphoma, so we performed chemotherapy combined with radiotherapy. At one year follow-up, he had no neurological deficit and no recurrence on neurologic and radiologic exams. Primary spinal cord lymphomas should be considered in the differential diagnosis of spinal cord tumors. Early surgical management is mandatory to achieve a recovery of neurologic function, especially if the patient has a neurological deficit.

Key Words: Primary Spinal Lymphoma • Spinal Epidural Mass • Decompressive laminectomy • Chemo-radiotherapy

INTRODUCTION

Malignant lymphoma (ML) can involve the central nervous system either primarily or by secondary spread, which tends to occur late in the disease as part of widespread dissemination. Lymphoma presenting as primary tumors of the spinal cord are extremely uncommon. The rate of epidural mass formation in all cases of MLs is 0.8-2.8. Owing to this rate of occurrence, a small number of research studies and case reports dominate the literature. We report a rare case of primary spinal epidural lymphoma that presented with progressive myelopathy.

CASE REPORT

A 47-year-old man presented with progressive lower extremity weakness and numbness associated with fecal and urinary incontinence over 2 weeks. The patient did not complain of fevers, night sweats, or weight loss. A physical examination revealed hypoesthesia on the medial aspect of both thighs with bilateral motor weakness (Gr 4-4+). Anal tone decreased and urinary incontinence was observed. The vibration senses were reduced below the T10 level. There was no abnormality of laboratory findings except the erythrocyte sedimentation rate was elevated (ESR-32 mm/h; reference value - less than 20 mm/h). Preoperative magnetic resonance imaging (MRI) of the thoracicolumbar region showed an isointense spinal medulla on T1-weighted images and a slightly hyperintense T2-weighted images of the spinal cord extending from T9-L2. The mass was of the homogeneous signal intensity and exhibited diffuse enhancement with gadolinium (Fig. 1A-E). The patient underwent a decompressive partial hemilaminectomy of T9-L2 and tumor removal. Intraoperatively, a grayish epidural mass was identified and resected to decompress the cord and cauda equina. Histopathologic analysis of the tumor specimen showed infiltration of a small lymphocytic lymphoma. Proliferation of small blastic cells, with a lymphoblastic aspect and starry sky pattern, was seen. Further immunohistochemical characterization showed that results from tumor testing were positive for CD20, CD3, CD5, Bcl-2, Cyclin D1 and negative for CD23, CD10 and T-cell markers (Fig. 2). We performed the further evaluations because of indicated lymphoma characteristics. The MRI of the brain and cervical spine were normal. Further work-up, including computed tomography (CT) scans of the chest and the abdomen, iliac crest bone marrow biopsy, and ultrasound of the lymph nodes showed no extramedullary lymphoma manifestation. The patient had no history of immune disorder, and
human immunodeficiency virus (HIV) testing was negative. The treatment regimen consisted of radiotherapy, 40 Gy administered in 20 fractions, from T9-L2, followed by combination chemotherapy with cyclophosphamide, adriamycin, oncovin, and prednisone (CHOP). A follow-up MRI one year after the surgery showed that the lesions of lymphoma had disappeared (Fig. 1F). After surgery, all preoperative symptoms completely resolved. At one year postoperatively, the patient was faring well with no evidence of local recurrence or new lesions at any other site.

**DISCUSSION**

Primary central nervous system lymphoma is a rare form of extranodal lymphoma, particularly isolated primary spinal lymphoma. Compression of the spinal cord as the first manifestation of the primary spinal lymphoma is particularly rare, with an incidence of <5% [15]. In our report, the patient initially presented with compression of the spinal cord manifesting as hypoesthesia and motor weakness of the lower extremities. Although the mechanism of epidural mass formation without systemic lymphoma has not been clarified, the origin of the mass is thought to be bone marrow [10]. A small amount of lymphocytic cells can migrate to the spinal epidural space directly through the harversian canals of the vertebral bone or hematogenously via the epidural venous plexus, followed by a mass formation even before systemic lymphoma.

Findings from the CT and MRI provided only indirect diag-
Primary Malignant Lymphoma in Spinal Epidural Mass

Fig. 2. (A) Photomicrograph of a monomorphous specimen and comprised of small lymphocytes with a blend cytological appearance. A heterogenous mixture of cells including prolymphocytes, lymphoplasmacytoid cells or features of disease progression with an increased number of larger cells (paraimmunoblasts) and mitoses or an aberrant phenotype should be considered in the small lymphocytic lymphoma (Haematoxylin and eosin, ×400.) (B) Immunohistochemical staining showing tumor cells positive for CD20 (EnVision, original magnification ×200).

Therefore, concurrent chemoradiotherapy treatment was provide (CHOP; cyclophosphamide, adriamycin, oncovin, and prednisone+radiotherapy; 40 Gy administered in 20 fractions) after pathologic confirmation. Monnard et al observed local control of 88% and a 5-year overall survival of 69% with combined modality treatment. In a multivariate analysis, they found that the combined modality treatment was statistically superior to RT alone. Also, they reported a post-treatment complete neurologic response in 25% of patients who had initial motor deficits. The remaining 75% had partial recovery. In their multivariate analysis, they have found that complete neurologic response was the most significant favorable prognostic finding with regard to overall survival (p=0.001). In the present case, because of the manifestations of compression, timely surgical decompression was required to allow for a recovery of nerve function even before the diagnosis had been made. After surgery, the neurologic function in our patient was completely recovered. Therefore, every effort should be made to recognize and treat this disease appropriately.

CONCLUSION

Primary small lymphocytic lymphomas are rare, especially with an initial presentation of paraparesis. A primary small lymphocytic lymphoma of the spine should be considered in the differential diagnosis of primary spine tumors. Surgical decompression and tumor removal are mandatory to recover neurologic function, if the patient had a neurologic deficit. The combined modality treatment after surgical decompression resulted in improvement of the neurologic symptoms and remission of the lymphoma.
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