Primary Epidural non-Hodgkin’s Lymphoma in Clinical Stage IEA Presenting with Paraplegia and Showing Complete Recovery after Combination Therapy

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A 70-year-old man was admitted to our hospital because of paraplegia. A spinal epidural tumor (Th VII-XI) was diagnosed by magnetic resonance imaging and resected. Histological examination of the tumor showed non-Hodgkin’s lymphoma of the diffuse large cell type (Lymphoma Study Group classification), with a B cell phenotype. The clinical stage was IEA by the Ann Arbor classification. Radiation therapy of the involved field was performed (total dose: 40 Gy), followed by six courses of modified cyclophosphamide, adriamycine, vincristine, prednisolone (CHOP) therapy. He was discharged walking unaided 10 months later and has shown no evidence of relapse in the 26 months since discharge.

Key words: spinal-epidural tumor, malignant lymphoma, spinal cord compression

Introduction

Extranodal non-Hodgkin’s lymphoma accounts for 24–48% of all non-Hodgkin’s lymphoma (1–3). The common primary sites of such extranodal disease include Waldeyer’s ring and the stomach. Reports of primary epidural non-Hodgkin’s lymphoma are very rare. Here, we report a patient with primary epidural non-Hodgkin’s lymphoma, who presented with paraplegia due to spinal cord compression and showed complete recovery after combination therapy.

Case Report

A 70-year-old man developed progressive weakness in both legs from February 20, 1989 and paraplegia occurred two weeks after the onset. He was admitted to our hospital and was diagnosed as having an epidural tumor lying between the Th7 and Th11 levels by magnetic resonance imaging (Fig. 1A). Examination of cerebrospinal fluid by lumbar puncture before operation showed an increase of protein (102 mg/dl, normal value 10–40 mg/dl). Vertebral laminectomy and subtotal resection of the tumor were performed at the Department of orthopedics on March 20, 1989. Histological examination of the tumor showed non-Hodgkin’s lymphoma (diffuse large cell lymphoma according to the lymphoma Study Group classification) (Fig. 2). Immunohistological examination of tumor cells disclosed them to be MB1 (+), MxPanB (+), MT1 (−) and UCHL1 (−) in paraffin section. The clinical stage was determined to be Ann Arbor IEA after further work-up. Radiotherapy of the involved field was performed to a total dose of 40 Gy, followed by six courses of modified CHOP therapy (Fig. 3). No residual mass was shown by magnetic resonance imaging after radiotherapy and chemotherapy (Fig. 1B). Examination of cerebrospinal fluid by lumbar puncture also showed no abnormalities. Voluntary movements of both legs gradually improved during treatment. Following rehabilitation, he was discharged from our hospital walking unaided on December 4, 1989. He has been well since discharge and has shown no evidence of relapse at the time of writing this paper (June 1991).
Discussion

Among patients with non-Hodgkin's lymphoma, the incidence of extranodal lymphoma is 20–48%, but extranodal lymphoma affecting the epidural space as the primary site is very rare (1–3). Only ten cases diagnosed as primary epidural lymphoma have been reported in Japan, including this case (4). At our department, of 110 patients with primary extranodal non-Hodgkin's lymphoma (0.9%) (data not shown), the present patient was the only case of primary epidural non-Hodgkin's lymphoma found. According to previous reports, the frequency of spinal compression in non-Hodgkin's lymphoma ranges from about 0.1 to 5.8%, but these frequencies also include secondary epidural non-Hodgkin's lymphoma (5–8). Therefore, the frequency of primary epidural non-Hodgkin's lymphoma is thought to be much lower than suggested by these figures. Primary epidural non-Hodgkin's lymphoma has been reported to affect older individuals and to predominantly have a B cell phenotype. The most prevalent primary site is the thoracic spine (6, 9). The present patient had all of these characteristics.

The proper management of patients with primary epidural non-Hodgkin's lymphoma is not yet certain and the value of laminectomy is unclear because non-Hodgkin's lymphoma is a very radio- and chemo-sensitive
tumor (9). In this patient, laminectomy was performed to treat rapidly progressive paraplegia and to obtain a diagnosis, because of the lack of other sites of involvement. Epelbaum et al. have stated that when it is possible to obtain a tissue specimen from a site other than the spine and there is no rapidly progressive severe neurologic impairment, treatment based mainly upon intensive chemotherapy and irradiation may be sufficient (9). Recently, it was also suggested that chemotherapy alone may be a sufficient treatment for this disease (10).

The prognosis for functional recovery in patients with spinal cord compression due to epidural non-Hodgkin’s lymphoma is relatively better than that of patients with metastatic carcinoma, but it is very rare for a patient to be discharged walking unaided after being paraplegia as happened in this case (11).

**References**


