Spinal Burkitt’s Lymphoma Mimicking Dumbbell Shape Neurogenic Tumor: A Case Report and Review of the Literature

You-Sub Kim, Jung-Kil Lee, Ki-Young Choi, Jae-Won Jang

Department of Neurosurgery, Chonnam National University Medical School & Research Institute of Medical Sciences, Gwangju, Korea

Non-Hodgkin's lymphoma (NHL), a disease which may involve the spine, is frequently associated with advanced disease. Radiculopathy caused by spinal root compression as the initial presentation in patients with NHL is very rare and thought to occur in less than 5% of cases. A 69-year-old woman complained of a history of low back pain with right sciatica for 1 month prior to admission. Computed tomography and magnetic resonance imaging of the lumbar spine showed a dumbbell-shape epidural mass lesion extending from L2 to L3, which was suggestive of a neurogenic tumor. After paraspinal approach and L2 lower half partial hemilaminectomy, total excision of the tumor was achieved, followed by rapid improvement of back pain and radiating pain. The lesion was confirmed to be Burkitt’s lymphoma by histopathological examination. We then checked whole-body PET-CT, which showed multifocal malignant lesions in the intestine, liver, bone and left supraclavicular lymph node. Although a rare situation, Burkitt’s lymphoma should be considered in the differential diagnosis for patients presenting with back and lumbar radicular pain without a prior history of malignancy. Burkitt’s lymphoma could be the cause of dumbbell-shape spinal tumor.

Key Words: Burkitt’s lymphoma ・ Dumbbell-shape tumor ・ Epidural ・ Radiculopathy

INTRODUCTION

The spinal epidural space is an uncommon presenting site in non-Hodgkin’s lymphoma (NHL), and accounts for 9% of spinal epidural tumors and 0.1-3.3% of all lymphomas\(^8,10\). Burkitt’s lymphomas are small noncleaved B-cell lymphomas with highly aggressive clinical features. It is characterized by rapid progression, early hematogenous dissemination, and a propensity to spread to the bone marrow and the central nervous system (CNS)\(^1\). Three clinical variants of Burkitt’s lymphoma are described in the World Health Organization classification: endemic, sporadic, and immunodeficiency-associated types. Endemic Burkitt’s lymphoma refers to those cases occurring in African children, usually in the age between 4 and 7 years. Sporadic Burkitt’s lymphoma occurs worldwide, accounting for 1-2% of lymphomas in adults and up to 40% of lymphomas in children in the USA and Western Europe. Immunodeficiency-associated Burkitt’s lymphoma occurs mainly in patients infected with human-immunodeficiency virus\(^7\). CNS disease, found in less than 15% of sporadic cases at diagnosis, can include involvement of the meninges, infiltration of cranial nerves, intraparenchymal brain disease, or a paraspinal mass\(^8,13\). When Burkitt’s lymphoma develops in the epidural space and presents with neurologic deficits from compression of the spinal cord or spine nerve root, it is frequently associated with advanced disease\(^3,20\). Thoracic segments are predominantly affected regions, but any spinal region can be affected. In patients with NHL, spinal cord compression as the primary presentation is rare and thought to occur in less than 5% of cases\(^4,6,12,16\). We describe a patient who initially presented with spine nerve root compression, which was later revealed to be Stage 4E of Burkitt’s lymphoma.

CASE REPORT

A 69-year-old woman admitted with a 1-month history of low back pain radiating down to the right leg. The patient had been healthy before admission and her medical history was unremarkable except for several year history of chronic renal disease, which had been treated at a local clinic. Neuro-
logical examination showed hypoesthesia on the right L2-3 sensory dermatome and diminished motor power of right hip flexion and limiting gait. The right straight leg raising test was positive at 30 degrees of elevation. She showed normal rectal tone and no bladder-urinary dysfunction. In initial blood investigations, complete blood cell counts, protein, electrolytes were within normal ranges. There was no other evidence of hematologic disorder in laboratory examination. Plain X-ray films showed no abnormality. Magnetic resonance imaging (MRI) of the lumbar spine showed a well-demarcated, posterolateral extradural mass lesion between L2 and L3, with extension through the spinal foramen (Fig. 1). This mass lesion was isointense relative to the spinal cord on T1- and T2-weighted images. And heterogeneous enhancement was appreciated after administration of gadolinium. Computed tomography (CT) showed a poorly defined, slightly enhancing lesion involving the central canal and the right paraspinal area at the body level of L2-L3. Intervertebral foramina widening or bony destructive/sclerotic lesion was not demonstrated. Due to the dumbbell shape, we initially assumed the tumor was a benign neurogenic tumor affecting the L2 and L3 roots on the right. And we chose paraspinal approach because of the large extraforaminal portion of the tumor. Under prone position, midline hockey stick incision and dissection of paraspinal muscle were performed. After the exposure of L1-2-3-4 intertransverse space, L2-3 intertransverse ligament was removed. Also L2 and L3 transverse process was cut for visualization of the tumor. Careful dissection and removal of tumor were performed without injury of L2 and L3 root. Subsequently, L2-L3 half hemilaminectomy was needed for removal of intraspinal portion of the tumor. The epidural mass occupied the right L3 root with extension to the foramen. It was a white-brown in color, soft and fragile in consistency and uncapssulated mass. So it was hard to dissect due to fragility and uncapssulation but gross total removal was possible. The post-operative course was uneventful and the patient’s symptoms were improved soon.

Histopathological examinations disclosed a lymphoid lesion, which was diagnosed as Burkitt’s lymphoma. The tumor consisted of a single population of medium sized cells with abundant basophilic cytoplasm and multiple small nucleoli producing a starry-sky pattern with frequent mitotic figures (Fig. 2). Immunohistochemical studies demonstrated that the tumor cells were positive for CD20, CD79a, BCL-6, CD10, and EBV, but not for BCL-2 (Fig. 3). The proliferation marker Ki-67 was expressed in almost all the tumor cells, thus confirming Burkitt’s lymphoma.

Clinical postoperative reevaluation with whole-body positron emission tomography (PET) discovered multifocal malignant lesions in the intestine, liver, bone and left supraclavicular lymph node (LN) (Fig. 4). Finally, the staging of the disease
revealed Stage 4E of Burkitt’s lymphoma. The patient was transferred to the oncology department and recommended for receiving chemotherapy. But she refused chemotherapy for her underlying diseases such as chronic kidney disease, asthma and cardiac problem.

**DISCUSSION**

Burkitt’s lymphoma is a rare and aggressive B cell tumor, involves typically extranodal sites.

Adult patients with Burkitt’s lymphoma present with abdominal masses, B symptoms, tumor lysis, bone marrow involvement (70%) and leptomeningeal involvement (up to 40%). Diagnostic workup in the acute phase should include an MRI and/or CT scan of the spine and tissue sampling during surgery. MRI is the initial procedure of choice for evaluation of acute spinal cord or root compression. It provides good anatomical detail, offers more information about bone and soft tissue involvement, and potentially characterizes the tissue for the tumor mass itself. Following decompressive surgery, chemotherapy would be the initial treatment of choice in most patients with intermediate and high grade NHL, followed by radiotherapy in localized presentations. Complete remission of Burkitt’s lymphoma was sometimes reported after treating with dose-intensive, multi-agent chemotherapy regimens that incorporates CNS prophylaxis. There are two highly effective regimens, which are CODOX M (cyclophosphamide, vincristine, doxorubicin, high-dose methotrexate) and IVAC (ifosfamide, etoposide and high dose cytarabine). For CNS prophylaxis, intrathecal chemotherapy using cytarabine or methotrexate can be added. In the results obtained with 4 cycles of CODOX MI/IVAC protocol in pediatric patients, the 1-year event-free survival (EFS) rate was reported to 85%. However, the study about the overall survival rate of older patients is insufficient.

Patients with radiculopathy or myelopathy caused by spine root compression due to an unknown lesion require surgical decompression for the diagnosis and treatment. Although spinal chemotherapy for secondary spinal epidural Burkitt’s lymphoma seems to be an effective treatment protocol, the aggressive nature of the tumor necessitates immediate intervention to minimize neurologic dysfunction. Surgery provides the most rapid decompression of nerve tissue compared to chemotherapy and radiotherapy, both of which may take several days for decompression to occur. The role of surgery in this case was to achieve immediate neural decompression and to obtain an adequate specimen for a definitive pathological diagnosis.

Spinal tumors that extend into the vertebral canal and paraspinal spaces through the intervertebral foramen are so-called dumbbell-shaped spinal tumors. Basically, any mass occurring in the vertebral canal space, intervertebral foramen, or paraspinal space can be dumbbell shaped. The most common causes of spinal dumbbell lesions are benign neurogenic tumors, such as schwannomas or neurofibromas. The percentage of neurogenic tumors among dumbbell-shaped spinal tumors has been reported to be in the range from 68.8 to 81.3%. Dumbbell-shaped spinal tumors are usually thought to be of neurogenic origin, but this is not always the case. Various neoplastic and non-neoplastic causes, originating at an intradural and/or extradural compartment, may also lead to dumbbell-shape spinal tumors with/without intervertebral foraminal widening. There are many kinds of dumbbell-shaped spinal tumors other than neurogenic ones, and several cases of dumbbell-shaped spinal lymphoma have been reported. Most benign tumors have a tendency to grow slowly and extend into the intervertebral foramen, so they usually cause intervertebral foraminal widening. On the other hand, malignant tumors grow rapidly and are easily extended into the vertebral canal. In the present case, a benign neurogenic tumor was suspected due to the typical dumbbell-shape appearance. However, the possibility of other tumors was also raised on preoperative imaging findings. Many neurogenic tumors display some typical features such as regular margins, enlarged intervertebral foramen, and cystic/hemorrhagic changes. Lack of bony involvement on plain films or CT scan provides an important clue to the diagnosis. Extradural compression of the cord in the presence of normal radiographs may suggest a lymphoma. Finally, dumbbell-shaped Burkitt’s lymphoma was diagnosed in healthy immunocompetent women.
because of radicular symptom due to spine root compression as her initial manifestation.

CONCLUSION

Spinal cord or root compression as the initial presenting feature of lymphoma is rare. Although rare, Burkitt’s lymphoma should be considered in the differential diagnosis when a patient without a prior history of malignancy presents with back pain followed by spinal cord or root compression by a tumor. Burkitt’s lymphoma could be the cause of dumbbellshape spinal tumor too.

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