Primary Spinal Epidural Lymphoma
Mimicking Epidural Abscess in a Diabetic
- A Case Report -

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= Abstract =

당뇨 환자에서 경막외 농양과의 감별을 요한 원발성 척추 경막외 임파종
- 중례보고-

Primary spinal epidural lymphoma (SEL), i.e. occurring in the absence of any detectable extraspinal lymphoproliferative disorder, is an unusual cause of spinal cord compression. The authors report a 48-year-old, diabetic woman presented with back pain followed by acute paraparesis and voiding difficulty. She had been treated with acupuncture on her back before admission, and complete blood count showed leukocytosis with neutrophilia and increased erythrocyte sedimentation rate (ESR). Thoracic spine magnetic resonance imaging (MRI) revealed an epidural mass extending from T5 to T8 with compression of the spinal cord. Emergency decompressive laminectomy was performed with a tentative diagnosis of spinal epidural abscess, but a B-cell lymphoma was final pathologic diagnosis. Further staging showed no other sites of lymphoma, and the spinal lesion was treated by chemotherapy and radiotherapy.

The authors stress that primary SEL can mimic spinal epidural abscess (SEA) in the diabetic patient and should be a diagnostic consideration in patients with a syndrome of acute spinal cord compression manifested by a prodrome of back pain and neuroimaging consistent with an epidural compressive lesion, especially in a diabetic.

KEY WORDS Primary spinal epidural lymphoma • Epidural abscess • Spinal cord compression • Diabetic.

Introduction

Since the first description by Welch and Ginsburg\(^9\), lymphoma occupying the extradural space has been known to be an important cause of spinal cord compression. Extranodal presentations account for 15 to 30% of all lymphoma cases\(^9\). Of these, the spinal extradural involvement represents only 0.8 to 2.8%\(^9\). Cord compression, one of the most common neurological syndromes seen in spinal lymphoma, may occur in up to 7% of all lymphomas\(^9\).

The authors found no previous report of primary spinal epidural lymphoma (SEL) mimicking spinal epidural abscess (SEA) in a diabetic patient and believe that SEL should be a diagnostic consideration in patients with a syndrome of acute spinal cord compression manifested by a prodrome of back pain and neuroimaging consistent with an epidural compressive lesion, especially in diabetics without prior history of malignancy.
Case Report

A 48-year-old female presented with acute paraparesis and voiding difficulty. She was diabetic and used to have acupuncture treatment for chronic low back pain. She had multiple acupuncture treatments on the back area 1 month before presentation. She had a prodromal period of aggravated back pain for 15 days prior to the onset of myelopathy, followed by urinary retention.

On admission, she had mild tenderness on paravertebral muscle in the thoracolumbar region. Neurologic examination revealed Grade II weakness of motion of both hip and knee joints with complete paralysis of both ankle joints. Hypesthesia and sensory dissociation were noted below T8 dermatome level. Deep tendon reflexes were not seen in the lower limbs. Babinski sign was equivocal on both sides and ankle clonus was not present. Anal sphincter tone was decreased. Her body temperature was 38°C. A complete blood count showed leukocytosis, 17800/µL of white blood cell (WBC) count with neutrophilia of 88.2%. Erythrocyte sedimentation rate (ESR) was increased to 48 mm/hour. Random blood sugar level was 329mg/dL and hemoglobin A1C level was 8.2% of total hemoglobin (reference 4-6% of total hemoglobin). Liver function tests were normal. She was not immunocompromised or infected with the human immunodeficiency virus.

Plain radiographs of the thoracic spine showed absence of bony involvement. Emergency thoracic spine magnetic resonance imaging (MRI) revealed evidence of an enhancing epidural mass extending from T5 to T8, with antero-laterally displaced spinal cord (Fig. 1).

With a tentative diagnosis of spinal epidural abscess, the patient underwent emergency laminectomy for decompression of the spinal cord and tissue diagnosis. After laminectomy, a whitish glistening fibrogelatinous material

![Preoperative thoracic spine MRI. A & B] Sagittal T2-weighted image (T2WI) & T1-weighted image (T1WI) demonstrating a well-defined epidural mass, extending from T5 to T8 with compression of the spinal cord. The lesion shows homogeneous signal intensity, slightly lower than that of the spinal cord. C] Gadolinium-enhanced T1WI shows marked enhancement of the epidural mass. D & E] Pre- & Gadolinium-enhanced T1WI showing the epidural mass and antero-laterally displaced spinal cord.](image-url)
was noted in the epidural space. The mass compressing the dural sac was removed thoroughly.

Pathologic diagnosis of the epidural mass was a non-Hodgkin’s lymphoma (NHL). Histological subtyping according to the International Working Formula and immunohistochemical study revealed diffuse large cell lymphoma of B-cell origin (Fig. 2). Systemic investigations for occult disease, including hematology and biochemistry, chest radiograph, computed tomography (CT) of the abdomen and pelvis, iliac crest bone marrow biopsy and peripheral blood examination, did not reveal anything other than the mass lesion strictly limited to the extradural space (Stage 1E).

Additional chemotherapy and radiotherapy were given by the consulting oncology team. One month after surgery, she recovered from the paraparesis and achieved bladder control.

**Discussion**

Spinal extradural lymphoma can be an unusual cause of spinal cord compression and it represents only 0.8 to 2.8% of all extranodal presentations, which account for 15 to 30% of all lymphoma cases. Cord compression, one of the most common neurological syndromes seen in spinal lymphoma, may occur in up to 7% of all lymphomas.

Spinal cord compression occurs during the course of non-Hodgkin’s lymphoma (NHL) in from 0.1% to 10.2% of patients. Such involvement tends to develop late in the course of established disease, when dissemination has occurred, thus cord compression as the initial presenting feature of lymphoma is relatively uncommon. Primary spinal epidural lymphoma (SEL), i.e. occurring in the absence of any detectable extraspinal lymphoproliferative disorder, is an extremely rare cause of spinal cord compression.

Epidural extension from adjacent vertebral and paravertebral lesions is a common growth pattern in spinal lymphoma. However, hematogeneous involvement of the epidural space is possible. Less commonly, NHL may arise subdurally or within the spinal cord, and the disease may take on the behaviour of primary cerebral lymphoma.

Extradural presentations of lymphoma have been descri-
bed in all age groups, including childhood, but tend to cluster in the fifth to sixth decades. The symptoms are those to be expected with any epidural tumor, and as with other causes of malignant extradural compression, back pain tends to precede the onset of myelopathy by a variable period of time. Few studies have specifically evaluated the clinical features where the spine is the primary site of presentation. One found the most common presenting feature to be lower limb weakness. Most authors have shown the clinical picture consisted of two phases. The first, a prodrome of local back pain, sometimes accompanied by radicular pain, aggravated by coughing or sneezing. Haddad, et al. found the median duration of this pain before diagnosis to be 6 months. The second phase of spinal cord compression followed within 2-8 weeks.

The tumor has a tendency to spread over several vertebrae and has a definite predilection for the thoracic spine followed by the lumbar, cervical and lumbosacral levels. The location of lymphoma in our case was also T5 to T8 level. Mullins et al. suggested that patients with lesions involving segments T5-T8 have a less-favourable outcome because there are fewer radicular arteries to the spinal cord and because this region may be more susceptible to ischemic injury secondary to epidural compression.

Plain radiographs of the spine are not helpful in most cases compared to those of patients with metastatic carcinoma causing extradural cord compression, but may show pedicular erosion or vertebral body destruction. Thus, the lack of bony involvement on plain films or computed tomography (CT) scan often provides an important clue to diagnosis. Botterell, et al. emphasized that extradural compression of the spinal cord in the presence of normal radiographs should suggest a process such as lymphoma.

In diabetics, especially when the patient reveals infection signs such as leukocytosis and increased ESR as in our case, epidural abscess must be considered in differential diagnosis and they are usually fusiform, centered on and contiguous with the diseased disc and adjacent vertebral bodies, and often show lower signal than normal disc in MRI.

Most lymphomas give lower signal than fat and slightly higher signal than that of muscle on T1-weighted image (T1WI), with a homogeneous appearance, and are isointense with or give lower signal than fat and higher signal than muscle on T2-weighted image (T2WI). Contrast-enhanced T1WI affords good detection and delineation of epidural lymphoma, paravertebral extension, vertebral involvement and thereby of the full extent of the lesion, and are helpful for demonstrating the sites of spinal cord compression.

Spinal epidural hematoma or spinal subdural hematoma must be also considered in differential diagnosis when the epidural mass gives high signal on both T1WI and T2WI. Lymphoid malignancies are isointense with marrow on T2WI. As most metastatic carcinomas and sarcomas give higher signal than fat on T2WI it may be possible to distinguish these pathologies.

Lyons, et al. stressed that primary spinal epidural NHL should be a diagnostic consideration in the older patient with a syndrome of spinal cord compression manifested by a prodrome of back pain, followed by a rapid neurological deterioration, no prior history of cancer, normal plain spine radiographs, and neuroimaging consistent with an extradural compressive lesion.

The treatment of SEL is based on prognostic indicators such as histology, age and the extent of disease. Because of the known radiation sensitivity of lymphoma, extradural lymphoma has traditionally been treated by radiotherapy, either alone or followed by chemotherapy after initial decompressive surgery. But, recent studies have suggested an improved survival and tumor control with surgery followed by combined chemotherapy and radiotherapy.

Unlike other extradural malignancies, in which up to 26% of patients will be worse after surgery, evidence suggests that patients with lymphoma will improve or remain stable. The lack of bony involvement may contribute to this improved outcome, but this remains speculative. Primary spinal extradural non-Hodgkin’s lymphoma, as our case, is known to have a potentially favourable outcome if diagnosed and treated early, the lesion being markedly sensitive to radiation and chemotherapy.

The overall mean survival of patients with all types of extradural tumor is known to be 8 to 9 months, with less than 10% of patients surviving more than 1 year. The prognosis for patients with primary spinal epidural lymphoma are reportedly more favourable, with about 50% of patients surviving more than 3 years. Its occurrence in otherwise healthy patients, sensitivity to treatment, and favorable prognosis as compared with other malignant extradural tumors underscores the need to define this condition better.
Conclusions

The authors report a diabetic patient with a clinical picture of acute spinal cord compression, which is an uncommon initial presentation of non-Hodgkin’s lymphoma. Primary spinal epidural lymphoma can mimic spinal epidural abscess in the diabetic patients and should be a diagnostic consideration in those patients with a syndrome of acute spinal cord compression manifested by a prodrome of local back pain and neuroimaging consistent with an epidural compressive lesion, especially in diabetics without prior history of malignancy.

References


당뇨 환자에서 경막외 농양과의 감별을 위한 원발성 척추 경막외 임파종
- 중재보고-

김세훈·임동준·조태형·정용구·이동갑·이기찬·서중근

= 국문 요약 =

당뇨 환자에서 경막외 농양과의 감별을 위한 원발성 척추 경막외 임파종의 임상적 특징에 대한 보고이다. 임상적으로 건드림, 통증, 운동한상, 감각저하, 소변반사 등 증상이 관찰되었고, 진단은 자기공명영상 검사, 유리신경신경활성검사, 유리신경조직검사 등이 시행되었다. 임상적 특징은 통증 7%, 건드림 48%의 높은 반도를 보였으며, 신경 발달과의 감별은 주로 통증의 유무로 이루어졌다. 임상적 특징을 감별할 수 있는 기준으로 제시하였다.

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