MRI of Primary Spinal Epidural Lymphoma: case report

DA-CHUNG LIOU†  JUNG-MAO CHOU†

Department of Radiology†, Department of Pathology†, Zuoying Armed Forces General Hospital

We present a case with primary thoracic spinal epidural non-Hodgkin’s lymphoma which involved neuroforamen in the absence of spinal cord compression or paraspinal muscle extension. Magnetic resonance imaging (MRI) showed a lesion with homogeneous enhancement following the administration of gadolinium.

The prognosis of primary spinal epidural lymphoma with minimal symptoms during the early stage is relatively good as compared to those with classical signs of cord compression with more obvious abnormalities on images.

Primary epidural spinal lymphoma accounts for 0.9-6.5% of non-Hodgkin’s lymphoma (NHL). Primary spinal epidural NHL (SENHL) is less common as compared to metastatic lesions and represents 10-30% of all epidural malignancies [1]. Primary SENHL has a potentially favorable outcome if diagnosed and treated early. We present a patient with SENHL, which was diagnosed early by MRI and treated immediately.

MRI has replaced myelography and computed tomography (CT) in the examination of epidural neoplasms. It is therefore important to determine the MRI characteristics of these lesions for early diagnosis and treatment [1].

CASE REPORT

A 52 year-old man complained of lower back pain for a month and progressive weakness of the lower right limb sought consultation at our neurosurgical OPD. The patient had diabetes mellitus for 3 years and hypertensive cardiovascular disease for 2 years. However, the laboratory tests showed within normal limits. MRI of the T-L spine was performed under the impression of HIVD.

The MRI showed a fusiform iso-intense lesion in the posterior epidural space from T8-12 level on sagittal T1WI image (Fig. 1a) and high signal intensity on sagittal T2WI image (Fig. 1b). Homogeneous enhancement of the lesion was found following the administration of gadolinium (Fig. 1c). On the axial MR images, the lesion occupied the posterior epidural space and extended into the right neuroforamen (Fig. 2). According to the series of MRI findings, a spinal epidural malignancy was highly suspected, thus he was admitted immediately and operated upon. The pathologic diagnosis was diffuse large B cells epidural non-Hodgkin’s lymphoma, T8-12 (Fig. 3a, 3b). The patient was discharged one month later but continued to receive radiotherapy and chemotherapy.

Reprint requests to: Dr. Da-Chung Liou
Department of Radiology, Zuoying Armed Forces General Hospital.
No. 553, Chun Hsiao Road, Kaohsiung 813, Taiwan, R.O.C.
DISCUSSION

The growth pattern of spinal lymphoma may be divided into three types according to its main location: paraspinal, vertebral, and epidural. However, the epidural location is less common than the paraspinal and vertebral locations [2].

Epidural lymphomas account for 9% of epidural spinal tumors and 0.1-3.3% of all lymphomas. Non-Hodgkin’s lymphomas have a spinal location in 0.1-6.5% of cases [3]. The origin of SENHL remains unknown. Rubinstein believes that it stems from the lymphatic tissue present in the epidural space, while other authors are in support of the hypothesis that SENHL originates from the paravertebral lymph nodes or from the vertebral body, which later extends to the epidural space. Recent immunocytochemical and electron microscopic studies have identified that the tumor most frequently originates from the B lymphocytes [3].

SENHL usually presents in the fourth to fifth decade of life and tends to occur in males (66-76%) [3]. The tumor has a tendency to spread over several vertebrae and has a definite predilection for the thoracic spine followed by lumbar, cervical, and sacral level. Some authors consider this to be a result of both the greater length of the thoracic portion in relation to others and to the particular lymphatic drainage of the vertebral column [3]. The clinical findings of SENHL include backache, limb weakness, paraplegia, motor impairment, and bladder control impairment [4]. Cord compression is one of
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the more common neurological syndromes seen in spinal epidural lymphoma, which occurs in up to 7% of cases and more frequently in NHL than in Hodgkin’s disease [1, 4, 5].

According to the report by Macvicar et al., there are three points that should be emphasized. First, the prognosis for cord compression due to epidural lymphoma is far better than cord compression in metastatic carcinoma. Second, the presence of epidural lymphoma does not adversely affect the overall prognosis of the disease. Third, early diagnosis and treatment of epidural lymphoma carries an excellent prognosis [4]. It is therefore important to detect epidural disease as early as possible. Plain radiographs of the spine are of minute aid and have no practical value as the only sign visible from plain films is bone destruction. Myelography reveals thecal indentation, encasement, or blockage, but it is still not adequate in evaluating extradural lesion. CT has clear advantages in the demonstration of epidural tumors when compared with myelography and plain radiography. It is possible to determine the relationship of an epidural mass to the underlying bone or adjacent retroperitoneal or mediastinal soft tissue masses [4], but it is difficult to survey the whole cervical, thoracic, or lumbar spine if the lesion extends over more than one segment of the spine. MRI is a non-invasive procedure in evaluation of the location and the extent of a paraspinal soft tissue mass. MRI is also helpful in the demonstration of bone marrow infiltration which is helpful in the staging of the disease [2, 6].

An epidural infiltration of spinal lymphoma is relatively homogeneous on MRI. Most lymphomas showed lower signal intensity than fat and slightly higher signal intensity than muscle on T1WI, and are isointense or have a lower signal intensity than fat and a higher signal intensity than muscle on T2WI. Homogeneous enhancement was found following the administration of gadolinium and is infiltrative extending along the epidural space [1, 2]. These observations suggest that lymphoma has characteristic appearance on MRI, and therefore can be differentiated from other epidural tumors.

We present this rather exceptional case of SENHL because it primarily emerged from the epidural space, rather than the epidural extension from adjacent vertebral or paraspinous lesion. Additionally, our case was found in an early stage in the absence of cord compression, and the early diagnosis and treatment of non-compressive epidural lymphoma improves the prognosis.

SENHL can be confused with epidural hematoma, epidural abscess, neurofibromatosis, herniated disk, metastasis and sarcoma [1, 7]. Spinal epidural hematomata revealed high signal intensity on both T1WI and T2WI; epidural abscesses are fusiform in shape, center in location, contiguous with the diseased disc and adjacent vertebral bodies, and often shows lower signal intensity than a normal disk. Neurofibromatosis is usually located in intradural, extramedullary space; it is a well-defined mass with a dumbbell configuration and widening of intervertebral foramen.

Figure 3. a. Histopathology (200x): Microscopically, the section showed a picture of malignant lymphoma with large round, oval, polygonal cells and focal pleomorphic nuclei (arrow). b. Tumor cells showed positive for CD20 in immunohistochemical study (arrow), which was indicative of diffuse large B-cell lymphoma.
Neurofibromatosis shows homogeneous isointense to cord on T1WI MRI and hyperintense on T2WI MRI as compared to surrounding fat. A herniated disk shows low signal intensity on both T1WI and T2WI MRI without gadolinium enhancement, associated with displacement of the posterior longitudinal ligament and epidural fat of relative high signal intensity on T1WI MRI. Most metastatic carcinomas and sarcomas show higher signal intensity than fat on T2WI MRI and heterogeneous enhancement after gadolinium administration. However, the metastatic carcinoma often involves the spinal and paraspinal regions. Thus, it may be possible to distinguish these pathologies from SENHL [1, 7, 8].

The treatment of epidural lymphoma is a subject of debate. Radiotherapy has been the mainstay of treatment in most cases, in combination with surgery and/or chemotherapy [3, 4].

Spinal epidural lymphoma always involves several segments of the spine, which may have a paraspinal extension and are accompanied by diffuse vertebral marrow signal changes. In some rare cases of early stage spinal epidural lymphoma, it may be confined to the epidural space.

REFERENCE
原發性脊椎硬膜上淋巴癌之磁振造影影像：病例報告

劉大忠¹  柯榮茂²

國軍左營總醫院  放射診斷科¹  病理科²

我們報告一個有關原發性胸椎硬膜上 non-Hodgkin 氏淋巴癌，它因為早期發現，只侵犯右側神經孔，沒有造成脊髓的壓迫或脊椎外的侵犯。磁振造影在 T1-weighted 影像上顯示和脊髓相同訊號的病灶，在 T2-weighted 影像上顯示高訊號的病灶，並且在注射對比劑後顯示均勻顯影的影像。

原發性脊椎硬膜上的淋巴癌，如果早期發現只有輕微的症狀或局部的病灶，它的預後比那些典型的脊髓壓迫症狀或廣泛性的侵犯好很多。