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## CASE REPORT

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# Primary Epidural Spinal Lymphoma: a Rare Disease Entity

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### ABSTRACT

*Primary spinal epidural lymphoma is a rare entity and one of the differential diagnoses of an epidural spinal mass. This report describes two patients who presented with symptoms of cord compression subsequently proven to be due to primary spinal epidural lymphoma. The imaging features of this condition are discussed.*

*Key Words: Lymphoma, non-Hodgkin; Spinal cord neoplasms; Tomography, X-ray computed*

## 中文摘要

### 罕見的原發性脊柱硬膜外淋巴瘤

李嘉樂、張智欣、袁學遠、AT Ahuja

原發性脊柱硬膜外淋巴瘤十分罕見，為脊髓硬膜外腫塊鑒別診斷之一。本文描述兩名患者病發時有脊髓壓迫症狀，後被確診為原發性脊柱硬膜外淋巴瘤。本文同時討論此症的影像學。

### INTRODUCTION

Primary spinal epidural lymphoma (PSEL) is a rare disease entity and is defined as lymphoma primarily occurring in the epidural space in the absence of other systemic localisation, brief clinical history, and no or only secondary erosion of the vertebral body.<sup>1,2</sup> It must be distinguished from secondary spread to the epidural space from elsewhere, as the prognosis of primary epidural lymphoma is more favourable if diagnosed or treated early; 50% of patients survive more than three years.<sup>3</sup> Commonly epidural lymphoma presents with features of cord compression, preceded by pain.<sup>3</sup>

### CASE REPORT

#### Case 1

A 34-year-old woman presented with low back pain for

a few weeks in November 2009. She then developed subacute onset of bilateral lower limb weakness. On clinical examination, a sensory level was elicited at T10.

Computed tomography (CT) showed a hypodense homogeneously enhancing epidural lesion extending from T7 to T12/L1 levels, with neuroforaminal extension and indicative features of spinal cord compression at these levels. Associated pre- and paravertebral soft-tissue thickening at T12 level (Figures 1 and 2) was also noted. The differential diagnosis included infection, leptomeningeal tumour spread, or a primary spinal canal tumour.

A CT-guided biopsy of the paravertebral soft-tissue thickening at T12 was performed. Histology showed

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**Figure 1.** Contrast-enhanced computed tomography shows an enhancing epidural soft tissue lesion (white arrow) at the right posterolateral aspect of the spinal canal. Abnormal similarly enhancing soft tissue is also present in the right paravertebral region (black arrow).

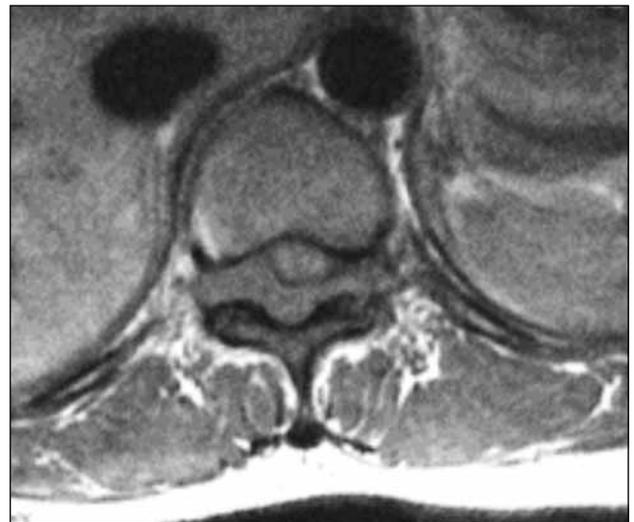


**Figure 2.** Contrast-enhanced computed tomography reveals the extension of the soft tissue lesion into the right neural foramen (white arrow), likely due to spread into the paravertebral region as shown in Figure 1 (black arrow). Biopsy confirmed the lesion to be a primary spinal lymphoma.

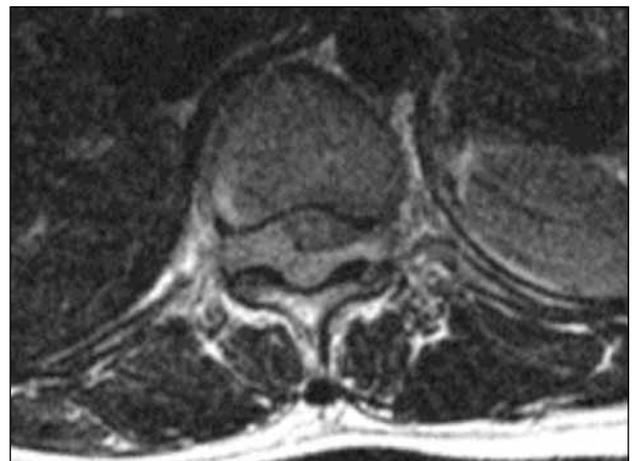
it to be malignant low-grade non-Hodgkin's B-cell lymphoma. In view of the progressively deteriorating lower limb power, debulking surgery for spinal cord decompression was performed. Postoperative whole-body CT revealed residual tumour at the operative site and no other supradiaphragmatic abnormal foci (including the brain). The bone marrow examination was negative. The diagnosis was PSEL. Chemotherapy was started and a follow-up CT showed significant decrease in size of the residual tumour.

## Case 2

A 35-year-old woman presented with bilateral thigh and low back pain for one month in October 2008. She then presented with subacutely evolving bilateral lower limb weakness. Physical examination revealed lax anal tone and decreased sensation in the peri-anal region. Magnetic resonance imaging (MRI) showed an extradural homogeneous lesion in the posterolateral aspect of the spinal canal from T11 to L1 levels. The lesion was mainly T1 hypointense and T2 hyperintense and enhanced homogeneously following gadolinium injection (Figures 3 to 6).



**Figure 3.** An axial T1-weighted magnetic resonance image shows the slightly T1-hypointense lesion in the epidural space at the right posterolateral aspect of the spinal canal.



**Figure 4.** An axial T2-weighted magnetic resonance image shows the lesion is mainly T2-hyperintense and displaces / compresses the spinal cord.



**Figure 5.** A gadolinium-enhanced axial T1-weighted magnetic resonance image shows homogeneous enhancement of the lesion and right neural foraminal spread (white arrow). Biopsy confirmed it to be primary spinal lymphoma.



**Figure 6.** A gadolinium-enhanced sagittal T1-weighted magnetic resonance image better delineates the extent of the tumour from T10 to L1 spinal levels.

In view of deteriorating lower limb neurological symptoms and signs, spinal canal decompressive surgery was performed. Histology revealed a diffuse large B-cell lymphoma. Re-staging whole body CT showed no other area of involvement and bone marrow examination was negative, confirming the diagnosis was PSEL. Chemotherapy was started and the patient responded well.

## DISCUSSION

Non-Hodgkin's lymphoma of the spinal cord was first described by Welch (as cited in Routh<sup>4</sup>). While PSEL accounts for about 9 to 10% of spinal tumours, 0.1 to 6.5% of non-Hodgkin's B-cell lymphomas have epidural spinal involvement.<sup>1-4</sup> The origin of PSEL is still unknown. Some believe that it is derived from lymphatic tissue in the epidural space,<sup>5</sup> while others suggest it originates from the paravertebral lymphoid rests or from the vertebral body, which later extends to the epidural space.<sup>3</sup>

There is a propensity for non-Hodgkin's lymphoma to involve the extradural space, which is in contrast to Hodgkin's disease.<sup>3</sup> Histologically, PSEL is mostly of B-cell origin; intermediate-, low-, or high-grade T-cell variants are rarely encountered.<sup>6</sup>

The disease tends to manifest in the fifth to sixth decade, although childhood presentation also occurs.<sup>2,3,7,8</sup> Some authors have suggested a male predominance while others state there is no gender predilection.<sup>1,3,7</sup> The above two cases are atypical in terms of age (being in their third decade), and causing a diagnostic dilemma prior to available histology.

As in the two cases presented here, the thoracic region is the most commonly involved, followed by the cervical, lumbar, and lumbosacral regions.<sup>3,9</sup> This may be related to the relatively longer length of the thoracic spine and the lymphatic drainage of the vertebral column.<sup>1</sup> Thoracic involvement usually has poor prognosis.<sup>7</sup> Within the epidural space, the posterolateral position is a common site, as occurred in these two cases.<sup>3,7</sup>

Imaging plays an important role in the diagnosis of PSEL. Plain radiographs are usually normal with occasional vertebral scalloping by the mass (evident in 30-42% of cases).<sup>3</sup> Cross-sectional imaging by MRI and CT have replaced myelography, as they provide more useful information regarding the site and cause of cord compression, and assessment of possible

systemic involvement. On CT, its density is variable (from hypodense, isodense to hyperdense) and shows significant uniform contrast enhancement as in case 1. On MRI, lymphoma has a characteristic appearance, the signal being lower than fat and slightly higher than muscle on T1-weighted images, and having lower than fat and higher than muscle on T2-weighted images.<sup>3</sup> The tumours are isointense to spinal cord on T1- and T2-weighted sequences. Moreover, CT has an advantage over the MRI in assessing the intrinsic bony abnormalities. In addition, the long acquisition time of MRI may be degraded by patient movement. However, MRI provides better soft tissue information and thus greater anatomic detail.<sup>3</sup>

Importantly, PSEL should be differentiated from the other lesions, such as haematoma, abscess, and metastasis.<sup>7,10</sup> Spinal epidural haematomas show variable density on CT and signal intensity on MRI, depending on the age of the blood, but it shows no contrast enhancement. Epidural abscesses tend to be central and contiguous with diseased disc and adjacent vertebral bodies,<sup>7,10</sup> and are usually fusiform. On CT, they reveal rim-enhancing lesions, and MRIs usually show lower signal than normal disc and rim enhancement after gadolinium. While metastatic carcinomas may yield similar imaging features to PSEL with CT and MRI, they have higher signal intensity than fat on T2-weighted images and heterogeneous contrast enhancement.

For cord decompression, the mainstay of treatment is surgery. The tumour is markedly sensitive to radiation and chemotherapy as demonstrated in the above two cases.<sup>1,2,9</sup>

To conclude, PSEL should be suspected if: (1) there is spinal cord compression with initial pain followed by acute neurological deficit; (2) there is absence of any previous history of cancer; (3) there is normal bony

structure in plain radiographs or CTs; and (4) cross-sectional imaging (CT or MRI) reveals an extradural contiguous level compressive soft tissue lesion.<sup>7,11,12</sup> Absence of systemic involvement should be elicited by a brief clinical history, which is the hallmark of diagnosis.<sup>7,12</sup>

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