



## Spinal Epidural Diffuse Large B-Cell Lymphoma Presenting as Cervical Myelopathy: A Case Report

Chung-Che Lu<sup>1</sup>, Chao-Hung Yeh<sup>1\*</sup>, Shih-Sung Chuang<sup>2</sup>

<sup>1</sup>Department of Neurosurgery, Chi-Mei Medical Center, Tainan, Taiwan

<sup>2</sup>Department of Pathology, Chi-Mei Medical Center, Tainan, Taiwan

\*Corresponding author: Chao-Hung Yeh, Department of Neurosurgery, Chi-Mei Medical Center, Tainan, Taiwan

**Citation:** Lu CC, Yeh CH, Chuang SS. (2021) Spinal Epidural Diffuse Large B-Cell Lymphoma Presenting as Cervical Myelopathy: A Case Report. Ann Case Report 6: 675. DOI: 10.29011/2574-7754.100675

**Received Date:** 03 July, 2021; **Accepted Date:** 09 July, 2021; **Published Date:** 14 July, 2021

### Abstract

Diffuse large B-cell lymphoma involving spinal epidural space is relatively rare and there is often a delay in its diagnosis. The case describes a 56-year-old male presented with 2-month history of neck pain radiating to the right upper limb. He was initially diagnosed with cervical degenerative disease and has undergone rehabilitation. However, he experienced progressive symptoms of myelopathy including weakness and numbness of all four limbs and dysuria all within a span of one week. The magnetic resonance imaging (MRI) revealed a C6-T2 epidural tumor with extrathecal compression and bilateral neuroforamen extension. Emergent C6-T2 decompressive laminectomy and tumor removal were performed thereafter. Histopathological examination of this tumor revealed diffuse large B-cell lymphoma. The neurological deficits gradually improved after surgery followed by six cycles of chemotherapy with rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisolone (R-CHOP).

**Keywords:** Epidural; Diffuse large B-cell lymphoma; Spine; Surgery

### Introduction

Diffuse large B cell lymphoma (DLBCL) is the most common lymphoma and accounts for approximately 25 percent of all non-Hodgkin lymphomas (NHL). Diffuse large B-cell lymphoma located in the spinal epidural space accounts for only 1.8% of cases [1,2]. The symptoms and signs of the disease depend on the location and size of the tumor. Prodromal symptoms can include localized pain accompanied by radicular pain that can persist for several months to a year. Rapid neurological deterioration then occurs as a result of spinal cord compression, therefore it is often missed and may be misdiagnosed, missing the opportunity of timely intervention to prevent tumor progression [3].

### Case report

We would like to report a case of a 56-year-old male who presented with 2-month history of neck pain radiating to the right upper limb. He was initially treated conservatively, but the symptoms progressed within a week, of which includes weakness

and numbness of all four limbs and dysuria, hindering his ability to walk. He also reported one month history of fever and night sweats. The past medical history includes type 2 diabetes and benign prostatic hyperplasia.

On neurological examination, weakness of the right distal upper limbs (2/5), left distal upper limbs (3/5) and lower limbs (4/5) was observed. Hypoesthesia below C6 dermatome and lower limbs hyperactive reflexes were also noted. Anal sphincter tone was intact. Physical examination demonstrated positive Lhermitte's sign and bilateral Spurling's test with negative Hoffman's sign and Babinski sign.

Initially, the X-rays of the cervical spine were obtained, which showed no obvious abnormality except for degenerative changes. Laboratory evaluation on admission demonstrated anemia (Hb: 6.8g/dL). No abnormality of other peripheral blood, electrolytes and tumor marker was found. The MRI showed enhancing mass lesions at C5-T4 posterior paraspinous regions, C6-T2 epidural space with bilateral neuroforamen extension and extrathecal compression, enlarged lymph nodes up to 3cm at bilateral IB/II/V regions and a right tongue base enhancing nodule (2.2cm) (Figure 1).

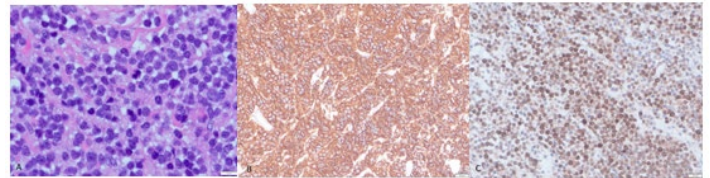


**Figure 1:** (A) Lateral cervical X-ray showing no obvious abnormality except for degenerative changes. (B) Sagittal T2-weighted pre-operative MRI showing C5-T4 posterior paraspinal regions and epidural mass. (C) Sagittal T1 weighted with gadolinium showing the lesion enhances with contrast. (D) Axial T2 weighted MRI showing the mass extended to bilateral neuroforamen and posterior paraspinal muscle with extrathecal compression.

Emergent C6-T2 decompressive laminectomy and tumor removal was performed using the posterior midline approach. A greyish soft tumor between paraspinal muscle and interspinous ligament was found during the procedure. The spinous process and lamina were intact without tumor infiltration. After laminectomy, a greyish epidural tumor was identified dorsally, extending from C6-T2 and bilateral neuroforamen, causing extrathecal and nerve root compression. This tumor was grossly removed for decompression. There was no dural defect or intradural invasion observed.

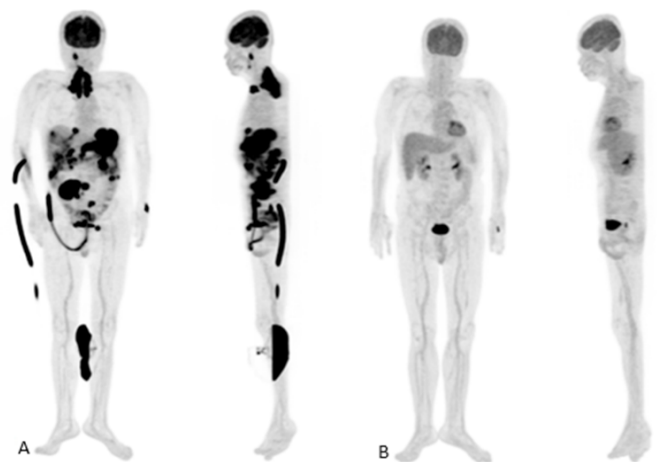
The tumor removed was sent for a pathology exam, and it showed that the tumor was of diffuse large B-cell lymphoma origin. Immunohistochemistry report showed CD10 (+), CD20 (+),

BCL-6 (+), CD3 (-), CD30 (-), BCL-2 (-), MUM1 (-), MYC (-) of neoplastic cells. Positivity of Ki-67 in 70% of tumor cells showed high proliferation index. The diagnosis was diffuse large B-cell lymphoma of germinal center phenotype (Figure 2).



**Figure 2:** (A) The atypical lymphocytes are medium to large-sized with a mitosis in the central-upper part (HE x1000) (B) These atypical lymphocytes are diffusely positive for CD20, indicating a B-cell lymphoma (CD20 x400) (C) Around 70% of these atypical lymphocytes are positive for Ki67, consistent with a high-grade tumor, i.e., Diffuse large B-cell lymphoma (Ki67 x400).

Further imaging including abdominal, chest CT and PET/CT demonstrated a high probability of lymphoma in right tongue base, right cardiophrenic angle, liver surface, stomach, mesentery, retroperitoneum and bilateral inguinal regions (Figure 3). Bone marrow biopsy indicated mature B-cell lymphoma with marrow involvement and he was consequently diagnosed with stage IV diffuse large B-cell lymphoma. The postoperative period was uneventful with successful recovery in neurological symptoms. The patient was referred to our oncology department on day 21 postoperatively for six cycles of R-CHOP chemotherapy. Follow-up PET/CT after chemotherapy showed good response (Figure 3).



**Figure 3:** (A) PET/CT before chemotherapy showing high probability of lymphoma in posterior neck, right tongue base, right cardiophrenic angle, liver surface, stomach, mesentery, retroperitoneum and bilateral inguinal regions. (B) PET/CT after six cycles of chemotherapy showing no evidence of FDG-avid lymphoma on whole body.

## Discussion

Lymphoma is a heterogeneous family of lymphoid malignancies, which typically develop in lymph nodes but may occur in almost any tissue. It can be classified into Hodgkin's lymphoma or non-Hodgkin's lymphoma, with the latter being more common. DLBCL is one of the most common lymphoid neoplasms in adults, accounting for 30% of non-Hodgkin lymphomas and 37% of B-cell tumors. Like most other NHLs, there is a male predominance with approximately 55% of cases [4]. The incidence increases with age and the median age at presentation is 64 years old for all patients [5]. DLBCL develops in single or multiple nodal sites, however, 40% of cases may arise from extra-nodal sites, such as the gastrointestinal tract, skin, bone marrow, salivary glands, liver and CNS [6]. Lymphomas located in the spinal epidural space are rare and account for only 0.1 to 3.3% of cases in previous reports [7]. At the time of diagnosis, approximately 60% of the spinal epidural involved DLBCL cases were advanced disease (stages III–IV) [1].

Spinal epidural lymphoma is thought to involve the paraspinal soft tissues initially, such as the paravertebral ganglion or epidural lymphoid tissue, and subsequently invade around the cord via the vertebral foramen without destroying bony structures [6,8]. In this particular case, the tumor involved both the paraspinal muscle and epidural space but the bony structures were intact. The symptoms and signs of spinal epidural lymphomas depend on the location and size of the lesions. Localized back pain or neck pain, sometimes accompanied by radicular pain to the legs or hands, can be the early symptoms that persist for several months to a year. In addition, sensory deficit and paresthesia may sometimes be observed. Within two to eight weeks, rapid neurological deterioration occurs due to spinal cord compression which causes symptoms like limb weakness and impairment of bladder and bowel function [9]. For this patient, the initial features included neck pain radiating to right upper limbs which lasted for 2 months, which was treated as cervical degenerative disc disease. However, it progressed and developed symptoms of cervical myelopathy including weakness and paresthesia of all four limbs and bladder dysfunction. This required immediate surgical decompression.

Early surgical decompression is suggested in the case of rapid and severe neurologic impairment, especially in cases of acute paresis or loss of bowel and bladder control. Moreover, surgical histological diagnosis is an accurate method of obtaining a definitive histopathological tissue sample, hence surgical biopsy is indicated when the diagnosis is not yet established [3,6].

Different combinations of chemotherapy have been used in the treatment of DLBCL. Six cycles of the R-CHOP regimen is considered the gold standard. In young patients with aggressive B-cell non-Hodgkin lymphoma and favorable prognosis, we suggest four cycles of R-CHOP rather than six or more cycles of

R-CHOP due to the former having the same efficacy but reduced toxicity [10]. As for limited stage DLBCL, combined modality therapy and chemotherapy alone can achieve comparable outcomes but are also linked with adverse effects [11]. Treatment with radiation therapy alone is not recommended for patients with limited stage DLBCL [12]. In conclusion, our case of a otherwise healthy 56-year-old man, was diagnosed early and underwent surgery, resulting in significant neurological improvement, good response to chemotherapy and a complete neurological recovery.

## References

1. Wada N, Kohara M, Ikeda J. (2010) Diffuse large B-cell lymphoma in the spinal epidural space: A study of the Osaka Lymphoma Study Group. *Pathol Res Pract*. 206: 439-444.
2. Swerdlow S, Campo E, Harris NL. (2008) World Health Organization Classification of Tumours of Haematopoietic and Lymphoid Tissues. Lyon, France; IARC.
3. Pokhrel NB, Prasad R, Paudel S. (2020) Primary spinal epidural non-Hodgkin's diffuse large B-cell lymphoma: A case report. *Clin Case Rep*. 8: 2276-2280.
4. Morton LM, Wang SS, Devesa SS. (2006) Lymphoma incidence patterns by WHO subtype in the United States, 1992-2001. *Blood*. 107: 265-276.
5. Shenoy PJ, Malik N, Nooka A. (2011) Racial differences in the presentation and outcomes of diffuse large B-cell lymphoma in the United States. *Cancer*. 117: 2530-2540.
6. Pinheiro JP, Rato J, Rebelo O, Costa G. (2020) Primary spinal epidural lymphoma: a rare entity with an ambiguous management. *BMJ Case Rep*. 13: e233442.
7. Monnard V, Sun A, Epelbaum R. (2006) Primary spinal epidural lymphoma: patients' profile, outcome, and prognostic factors: a multicenter Rare Cancer Network study. *Int J Radiat Oncol Biol Phys*. 65: 817-823.
8. Seo JY, Ha KY, Kim MU. (2014) Spinal cord compression by B-cell lymphoma, unclassifiable, with features intermediate between diffuse large B-cell lymphoma and Burkitt lymphoma in a patient seropositive for human immunodeficiency virus: a case report. *J Med Case Rep*. 8: 324.
9. Epelbaum R, Haim N, Ben-Shahar M. (1986) Non-Hodgkin's lymphoma presenting with spinal epidural involvement. *Cancer*. 58: 2120-2124.
10. Poeschel V, Held G, Ziepert M. (2019) Four versus six cycles of CHOP chemotherapy in combination with six applications of rituximab in patients with aggressive B-cell lymphoma with favourable prognosis (FLYER): a randomised, phase 3, non-inferiority trial. *Lancet*. 394: 2271-2281.
11. Kwak YK, Choi BO. (2017) Treatment outcome of diffuse large B-cell lymphoma involving the head and neck. *Medicine (Baltimore)*. 96: e7268.
12. Spicer J, Smith P, MacLennan K, Hoskin P. (2004) Long-term follow-up of patients treated with radiotherapy alone for early-stage histologically aggressive non-Hodgkin's lymphoma. *Br J Cancer*. 90: 1151-1155.