

Primary Diffuse Large B-Cell Non-Hodgkin's Lymphoma of the Thoracic Spine Presented Initially as an Epigastric Pain

Abstract

Non-Hodgkin's lymphoma (NHL) compromises the vast majority of lymphomas and predominantly takes on the form of B-cell lymphomas. More specifically, 30% of all newly diagnosed cases of NHL in the United States (US) are of diffuse large B-cell lymphoma (DLBCL) type, making it the most prevalent form of NHL in the US. Arising from either nodal or extra-nodal lymphatic tissue origin, DLBCL is an aggressive tumor which is fatal if left untreated. Primary central nervous system lymphoma is rare; however, when diagnosed, it presents as a DLBCL in 90% of patients. Herein, we present an elderly male complaining initially of acute epigastric pain but soon afterward developed acute spinal cord compressive symptoms; subsequently, it was found to be caused by a primary DLBCL diagnosed in the thoracic spinal cord. This case report presents a rare condition with unexpected initial presentation, and we attempt to illustrate the importance of early detection and treatment of DLBCL in attaining more favorable prognostic and survival rates among patients. Written consent was obtained from the patient after reading a written summary of the case report. This consent was checked and approved from the Scientific Board of the University of Aleppo.

Keywords: Diffuse large B-cell lymphoma, epidural mass lesion, epigastric pain, non-Hodgkin's lymphoma, spinal tumor

Introduction

Non-Hodgkin's lymphoma (NHL) compromises the vast majority of lymphomas. Thirty percent of all newly diagnosed cases of NHL in the United States are of the Diffuse Large B-cell Lymphoma (DLBCL) type, making it the most prevalent form of NHL in the US. DLBCL is an aggressive tumour which is fatal if left untreated.^[1]

Primary Central nervous system (CNS) lymphoma is rare, but when diagnosed it presents as a DLBCL in 90% of patients.^[2]

We present a case of an elderly male with unexpected initial presentation and rapid neurological deterioration. However, he was treated in time and fortunately showed an excellent recovery.

Case Report

A 60-year-old Middle Eastern male, with no significant medical history, presented to the emergency room complaining of acute nonradiating epigastric pain started

about 6–7 h earlier. There was no vomiting, nausea, or anorexia. There was no jaundice, pallor, or history of loss of weight. The temperature was 37.5°C. Abdominal examination was negative for hepatomegaly, splenomegaly, abdominal tenderness, or rebounding pain. There were no signs of lymphadenopathy. A clinical diagnosis of acute gastritis was made, and the patient was given proton pump inhibitor and discharged.

Two days later, the pain started to radiate toward the back, and the patient started to suffer from severe thoracic back pain. Four days later, the pain started to radiate toward both lower limbs with the subtle beginning of the weakness of the lower limbs and progressed within a few hours later to inability to walk, with intact neurological function of the upper extremities.

He denied any history of trauma, recent history of viral illness, or illicit drug use. On neurological examination, the patient was found to have bilateral numbness and motor impairment in the lower limbs with muscle strength of 2/5. Hyperactive patellar and Achilles reflexes were observed. Pain

**Fakhr Fakhouri,
Nihad Shoumal¹,
Batoul Obeid²,
Ayman Alkhoder³**

Department of Neurosurgery, Krankenhaus der Barmherzigen Brüder, Trier, Germany, Departments of ¹Neurosurgery and ²Pathology, Aleppo University Hospital, Aleppo, Syria, ³Department of Medicine, Emory University School of Medicine, Atlanta, GA, USA

Address for correspondence:

*Dr. Fakhr Fakhouri,
Department of Neurosurgery,
Krankenhaus der Barmherzigen
Brüder, Nordallee 1, 54292
Trier, Germany.
E-mail: fakhrf@msn.com*

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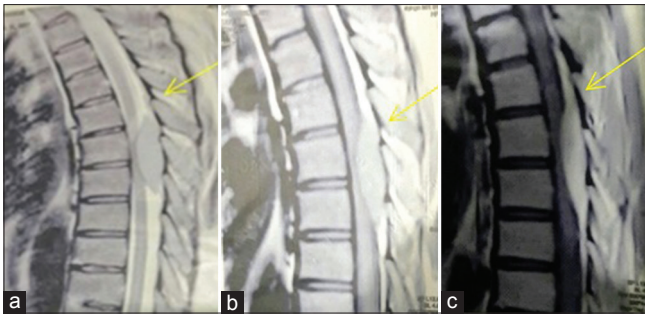


Figure 1: (a) Magnetic resonance imaging T2. (b) Magnetic resonance imaging T1. (c) Magnetic resonance imaging T1 with contrast

and light touch sensation were objectively normal; there was no bowel or urinary incontinence. The straight leg raise test was negative bilaterally.

Magnetic resonance imaging revealed a space-occupying lesion in the spinal canal at the T6/T7 level without detection of any lesions elsewhere [Figure 1]. Because of suspected metastasized tumor, further investigations were undertaken. Computed tomography (CT) scan of the neck, thoracic, abdomen, and pelvis were performed and did not reveal any masses, enlarged visceral organs, or lymph nodes. A bone marrow biopsy presented no marrow involvement.

In view of the rapid progression of the neurological deficit, the patient emergently underwent a neurosurgical decompression through partial laminectomy with total resection of the extradural mass.

The pathohistological examination of the biopsy revealed diffuse malignant infiltration of large atypical lymphoid cells, large vesicular nuclei, prominent nucleoli, and coarse chromatin.

Numerous mitotic cells were also present [Figure 2a]. Immune stains were positive for CD20 and leukocyte common antigen [Figure 2b] and negative for CD3 [Figure 2c] and CD30 [Figure 2d]. These findings are consistent with diffuse large B-cell lymphoma. After the diagnosis was made, the patient underwent chemotherapy.

Day after day, the patient gradually regained the strength in all muscle groups and was able to walk 1 month after surgery. Furthermore, the patient has been followed up for over 2 years without any signs of recurrence both clinically and radiographically [Figure 3].

Discussion

Non-Hodgkin's lymphoma (NHL) usually presents with a fast-growing and symptomatic mass, usually located in the neck, chest, or abdomen. However, it may present as a mass lesion anywhere in the body, including the spinal canal.^[3] Lymphoma in the epidural space of the spine is very rare and clinically presents as back pain of the thoracic spine, followed by neurologic deterioration. In addition, NHL presenting for the first time as spinal cord compression is rare and occurs in between 1% and 5% of cases.^[2]

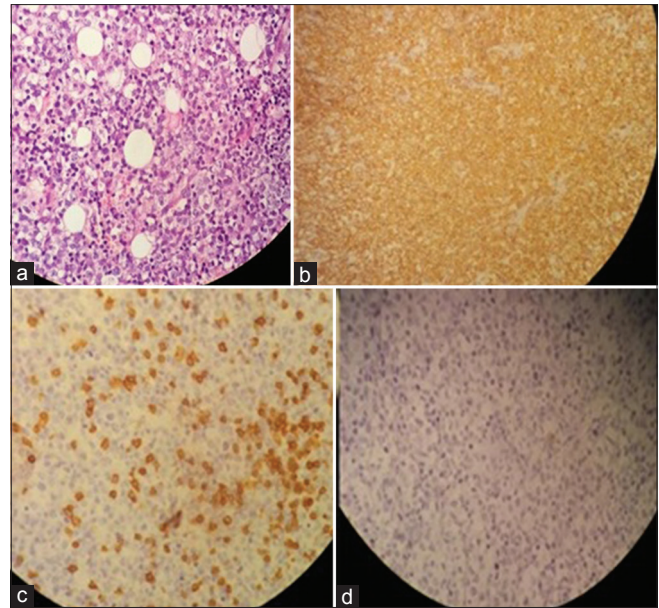


Figure 2: (a) Pathohistological examination of the biopsy revealed diffuse malignant infiltration of large atypical lymphoid cells, large vesicular nuclei, prominent nucleoli, and coarse chromatin. Numerous mitotic cells were also present. (b) Immune stains were positive for CD20 and leukocyte common antigen. (c) Immune stains were negative for CD3. (d) Immune stains were negative for CD30

The most prominent notification in our case is how the patient's initial presentation of epigastric pain for 2 days; then, the pain started to radiate toward the back, and finally, the symptoms and signs of the spinal cord appeared. Neurological deterioration usually occurs over several days or weeks. In our case, it progressed very rapidly within very few hours. Therefore, the surgical intervention has been done as an emergency. Interestingly, abdominal pain could be more frequent as thought an initial presentation of spinal epidural mass lesions such as a spontaneous spinal epidural hematoma^[3] or a spinal epidural abscess.^[4]

It indeed does not initially come one's mind to consider NHL in the differential diagnosis of a patient with acute epigastric pain that converts later to a thoracic back pain; however, in the context of an appropriate constellation of sudden onset and localized thoracic back pain in an elderly patient, which significantly increases despite the treatment with analgesics, mass lesion warrants careful consideration as an etiologic entity and further investigation is needed to confirm or rule out any suspected tumor mass. If a mass is present, especially in the epidural space, CT-guided biopsy is needed to determine the underlying etiology.

The treatment for spinal cord compression associated with NHL lacks definitive evidence but includes chemotherapy, radiotherapy, corticosteroid, and surgery. One approach proposed immediate surgery for patients presenting with profound neurology cord compression symptoms. Chemo- and radiotherapy should be started after surgery unless the patients were not eligible for such additional therapy modality due to poor condition. For those with mild-to-moderate cord

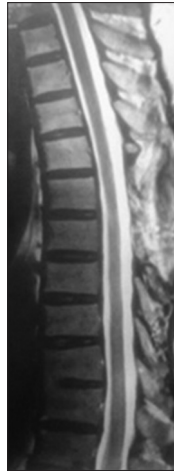


Figure 3: Magnetic resonance imaging showing no tumor or any signs of recurrence after 2 years

compression symptoms, an initial conservative trial of chemoradiotherapy may be initiated. If symptoms deteriorated or failed to improve within 2 weeks of initiating conservative treatment, surgery is definitely indicated.^[5]

A study conducted by Chang *et al.* reported complete recovery of neurologic symptoms in all patients who underwent decompressive surgery compared with 20% of those in the nonsurgical group. Furthermore, they noted a trend for improved median survival in the surgical group.^[6]

Conclusion

In this case, the patient presented complaining of epigastric pain. This pain radiated later to the back, and after a few days, the patient started to have bilateral weakness of the lower extremities. Therefore, the decompressive intervention must have been done as an emergency. According to the symptoms in our case report, maybe, we should keep in mind that the epigastric pain associates sometimes with the epidural spinal masses at the thoracic spine. Physicians should include NHL in the differential diagnosis of a newly diagnosed epidural spinal tumor to expedite diagnosis before the development of neurological

deterioration. An early diagnosis, in the time when chemoradiation could be sufficient, could save the surgical intervention.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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