Marginal Zone Lymphoma, A Rare Variant Of Non-Hodgkin Lymphoma: A Case Report

Non-Hodgkin Lenfomanın Nadir Bir Varyantı Olan Marjinal Bölge Lenfoması: Bir Olgu Sunumu

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Geliş Tarihi/Received: 06.11.2024 Kabul Tarihi/Accepted:11.12.2024

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Hippocrates Medical Journal / Hippocrates Med J, 2024, 4(3)135-137 DOI: 10.58961/hmj.1596949

Abstract

Marginal zone lymphomas are low-grade non-Hodgkin lymphomas originating from post-germinal center B cells. The disease is subdivided into extra-nodal, nodal, and splenic marginal zone lymphomas. The features, clinical course, and treatment of the disease vary considerably depending on the location of involvement. Here, we present a case of a patient who underwent surgery for a mass causing compression in the spinal cord and was diagnosed with marginal zone lymphoma. A patient presented to the neurosurgery clinic with complaints of back pain and a mass was detected on lumbar magnetic resonance imaging at the lumbar level that significantly narrowed the spinal cord. Excisional pathology of the mass was consistent with lymphoid follicle structures with prominent germinal centers and expanding marginal zones, and immunophenotyping was performed. The present case is a case of advanced-stage marginal zone lymphoma with extra-nodular involvement. Marginal zone lymphomas usually tend to involve areas with high antigenic stimulation such as the skin, ocular adnexa, and salivary glands, but in our case, the site of involvement was the spinal cord and has been rarely reported in the literature.

Keywords: Non-Hodgkin Lymphoma, Marginal Zone Lymphoma, Spinal Cord

Özet

Marjinal bölge lenfomaları, post-germinal merkez B hücrelerinden kaynaklanan düşük dereceli non-Hodgkin lenfomalardır. Hastalık, ekstra-nodal, nodal ve splenik marjinal bölge lenfomaları olarak alt bölümlere ayrılır. Hastalığın özellikleri, klinik seyri ve tedavisi, tutulumun yerine bağlı olarak önemli ölçüde değişir. Burada, omurilikte basıya neden olan bir kitle nedeniyle ameliyat edilen ve marjinal bölge lenfoma tanısı konulan bir olguyu sunuyoruz. Sırt ağrısı şikayetiyle nöroşirürji polikliniğine başvuran ve lomber manyetik rezonans görüntülemesinde, lomber seviyede omuriliği önemli ölçüde daraltan bir kitle tespit edildi. Kitlenin eksizyonel patolojisi belirgin germinal merkezler ve genişleyen marjinal zonlara sahip lenfoid folikül yapıları ile uyumlu bulunmuş ve immünfenotipleme yapılmıştır. Mevcut vaka, ekstra-nodüler tutulumu olan ileri evre marjinal zon lenfoma vakasıdır. Marjinal zon lenfomaları genellikle cilt, oküler adneksler, tükürük bezleri gibi yüksek antijenik uyarım olan yerleri tutma eğilimindedir ancak vakamızda tutulum yeri omurilikti ve literatürde nadir olarak bildirilmiştir.

Anahtar kelimeler: Non-Hodgkin Lenfoma, Marjinal Bölge Lenfoması, Omurilik

INTRODUCTION

Marginal zone lymphomas are low grade non-Hodgkin lymphomas that originate from post-germinal center B cells. The disease is subdivided into extra-nodal, nodal, and splenic marginal zone lymphomas. Disease characteristics, clinical course and treatment vary significantly depending on the location of involvement (1). Herein, we present a case operated upon due to a mass causing compression in the spinal cord and diagnosed as marginal zone lymphoma.

CASE REPORT

Fifty-one-year-old male patient without any chronic disease and no history of regular medication applied to the neurosurgery outpatient clinic with the complaint of back pain for the last six months and that has been irresistible in recent days. The pain was extending to the left knee. In the lumbar magnetic resonance imaging of the patient, whose neurological examination did not reveal any pathology, a mass constricting the spinal cord significantly at the lumbar level and exerting pressure on the filum terminale and cauda equina anterior fibers was detected. Excisional pathology of the mass was consistent with lymphoid follicle structures with prominent germinal centers and expanding marginal zones. In the immunophenotype, expression of CD20 was seen without CD5 or CD10 expression. The Ki-67 proliferation index was 5-10% suggestive of marginal zone lymphoma. The patient was referred to our clinic. He had no B symptoms but had a complaint of urinary incontinence for the last three days. His family history was unremarkable for any hematological disease. There was no palpable lymphadenopathy or hepatosplenomegaly in physical examination. Complete blood count and liver and renal biochemistries were normal. LDH was found to be 124 U/L (below the upper limit of normal). Repeat neurological examination was normal and the changes in repeat lumbar magnetic resonance imaging performed upon urinary incontinence complaints, were secondary changes associated with the previous surgery. For staging, positron emission tomography was performed that demonstrated 18- flouro-deoxyglucose uptake of lymph nodes above and below the diaphragm. SUVmax values were ranging between 2.93 and 3.60. Lymphoma involvement was not detected in the bone marrow biopsy. Cytogenetic and fluorescence in-situ hybridization test results were not gathered yet.

Rituximab combined with bendamustine regimen was started and planned to be given for six cycles.

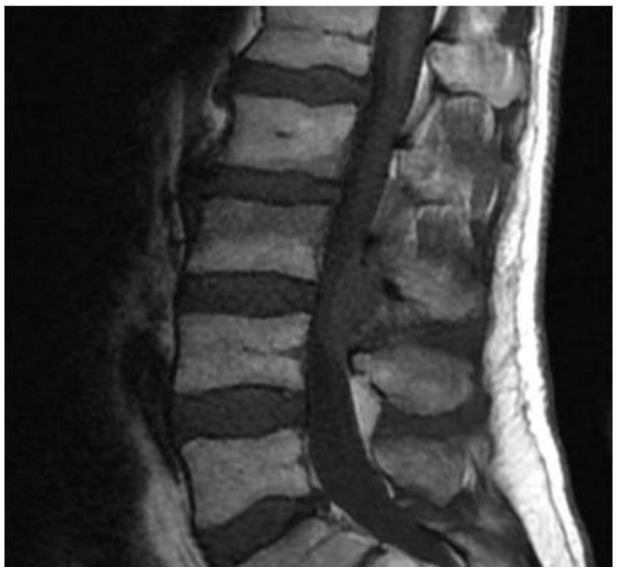

Figure 1. Spinal mass at L3-L4

DISCUSSION

The current case is an advanced stage marginal zone lymphoma case with extra-nodular involvement. Although the marginal zone lymphoma tends to involve the locations with high antigenic stimulation like skin, ocular adnexa, salivary glands; herein the involvement site was the spinal cord. There are primary marginal zone lymphomas reported in the literature involving the spinal cord or relapsing in the spinal cord (2, 3). The indication for systemic treatment was the threatened end-organ function. Rummel et al showed better progression free-survival and better tolerability with bendamustine-rituximab compared to CHOP-rituximab regimen. Thereby, we preferred bendamustine-rituximab in the current case.

Ethical Declarations:

Not required. Consent was obtained from the participant for this case report.

Conflict of Interest:

None declared.

Financial Disclosure:

None declared.

Author Contribution:

None declared by the author

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