Interesting Image

Mol Imaging Radionucl Ther 2025;34:156-158 *DOI:10.4274/mirt.galenos.2024.26879*

Neurolymphomatosis: The Sinister Face of Lymphoma

Nörolenfomatozis: Lenfomanın Kötü Yüzü

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Abstract

Neurolymphomatosis (NL) is a rare clinical condition characterized by the infiltration of malignant lymphocytes into the cranial or peripheral nerves, nerve roots, or plexus. Diagnosis can be clinically challenging due to its variable presentation. It usually occurs in B cell lymphoma; however, a few cases of extranodal killer/T cell lymphoma. Most cases present at a secondary site in patients with primary site in remission. ^{18}Fluorine fluorodeoxyglucose positron emission tomography/computed tomography plays an important role in the early detection of NL, resulting in timely treatment. We present a case of a 24-year-old male with nasal natural killer T cell lymphoma who initially responded to treatment but relapsed with NL based on clinical and radiological findings.

Keywords: Neurolymphomatosis, natural killer T cell lymphoma, ^{18}Fluorine fluorodeoxyglucose positron emission tomography/computed tomography, lymphom

Öz

Nörolenfomatozis (NL), nadir görülen bir klinik durumdur ve kranial veya periferik sinirlerin, sinir köklerinin veya pleksusların malign lenfositler tarafından enfiltrasyonudur. Değişken klinik sunumu nedeniyle tanısı zor olabilir. Genellikle B hücreli lenfoma ile birlikte görülür; ancak, ekstra nodal doğal öldürücü/T hücreli lenfomalı birkaç olguda bildirilmiştir. Çoğu zaman, birincil bölge remisyonda olan hastalarda ikincil bir bölgede ortaya çıkar. ^{18}Flor florodeoksiglukoz pozitron emisyon tomografisi/bilgisayarlı tomografi, NL'nin erken tespitinde önemli bir rol oynar ve zamanında tedavi edilmesini sağlar. Klinik ve radyolojik bulgulara dayanarak, başlangıçta tedaviye yanıt veren ancak daha sonra NL ile birlikte tekrarlayan 24 yaşında bir erkek nazal doğal öldürücü T hücreli lenfomalı olguyu sunuyoruz.

Anahtar kelimeler: Nörolenfomatozis, doğal öldürücü T hücreli lenfoma, ^{18}Flor florodeoksiglukoz pozitron emisyon tomografisi/bilgisayarlı tomografi, lenfoma

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Received: 20.08.2024 **Accepted:** 23.12.2024 **Epub:** 23.05.02025 **Publication Date:** 03.05.2025

Cite this article as: Gill SM, Hassan A, Qureshi PAAA, Bashir H. Neurolymphomatosis: the sinister face of lymphoma. Mol Imaging Radionucl Ther. 2025;34:156-158.

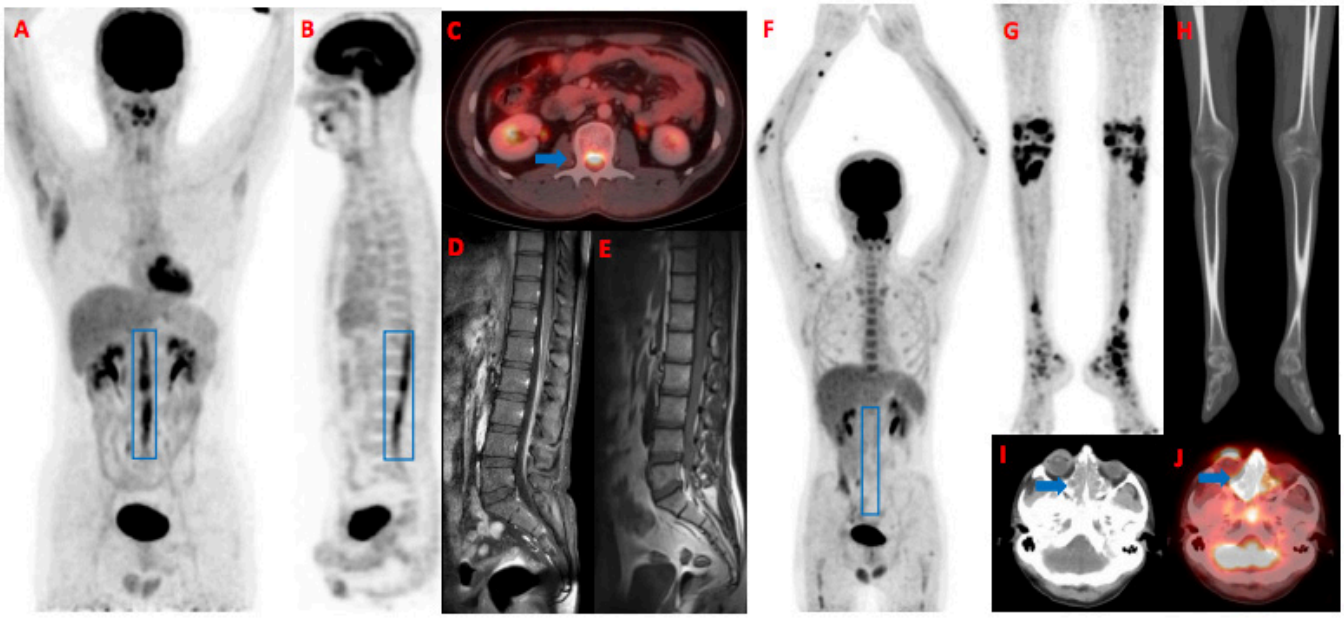

Figure 1. A 24-year-old male patient diagnosed with natural killer T cell lymphoma (NKTL) presented to our department for ^{18}Fluorine fluorodeoxyglucose 2-deoxy-glucose positron emission tomography/computed tomography (^{18}F-FDG-PET/CT). There was clinical suspicion of disease relapse due to complaints of backache, heavy headedness, and diplopia for 1 week. On examination, his glascow coma scale score was 15/15; power in upper limbs was unremarkable; however, power in lower limbs was reduced to 1/5 along with decreased tone and reflexes. His fundoscopy findings were unremarkable. Contrast-enhanced brain magnetic resonance imaging (MRI) of the patient was unremarkable. His ^{18}F-FDG PET/CT showed hypermetabolic mild thickening of the conus medullaris and cauda equina [standardized uptake value (SUV) 6.6, liver SUV 2.9] highly concerning for neurolymphomatosis (NL) [A,B maximum intensity projection (MIP) and sagittal PET blue boxes and C fused PET/CT blue arrow]. The primary tumor site (i.e., nasal cavity) showed minimal thickening along the nasal turbinate without significant FDG uptake. Contrast-enhanced spine MRI also revealed diffuse thickening, edema and abnormal enhancement of the conus medullaris and cauda equina, suggestive of lymphomatous infiltration [C and D, Sagittal pre- and post-contrast T1-weighted imaging (T1WI) MRI]. Cerebrospinal fluid analysis revealed cluster of differentiation 56+ malignant cells, and flow cytometry confirmed central nervous system relapse. He was then treated with high-dose chemotherapy and radiotherapy. Response evaluation ^{18}F-FDG PET/CT showed interval resolution of FDG uptake in the conus medullaris (F, MIP image blue box); however, there was progression at multiple sites, including bone marrow (G and H, MIP and coronal CT), preseptal region, and nasopharynx (I and J, axial CT and fused images, blue arrows). At this point, the patient was referred for palliative treatment; and after a period of about 2 months on palliative treatment, the patient succumbed to the complications of disease.

NL is a rare condition. Diagnosis can be clinically challenging due to its variable presentation. Literature review shows that, most commonly, it is associated with non-hodgkin lymphoma (1,2). Approximately 80% of the cases are associated with B cell lymphoma compared with T cell lymphoma. Among T cell lymphomas, NKTL is a rare entity that occurs at unusual sites, such as the central nervous system, skin, and nasopharynx. Despite aggressive treatment, it usually follows a rapidly progressive course (3). It is important to differentiate NL from non-cancerous conditions, such as the Miller Fischer variant of Guillain-Barre Syndrome, inflammatory radiculopathy, neuropathies, and chemoradiotherapy-induced damage to the nerves. That history proves to be of paramount importance in making definitive diagnosis. Although histopathology remains the gold standard for diagnosing NL, due to its limitations, MRI and PET-CT play a major role in the diagnosis of NL, with sensitivity of 87.5% and 100%, respectively (4,5,6). Biopsy is associated with an increased risk of permanent nerve damage, and it is usually reserved for cases that are not diagnosed on history and imaging. Within our institution, over a span of the last 15 years, there are 9 reported cases of NL on ^{18}F-FDG PET/CT (7,8), of which only our patient was of NKTL. Therefore, clinicians reporting PET scans should be cognizant of the fact that NL is likely to represent the aggressive extra-nodal lymphoma spectrum.

Ethics

Informed Consent: An informed consent was obtained from the patient.

Footnotes

Authorship Contributions:

Concept: S.M.G., P.A.A.A.Q., Design: S.M.G., A.H., P.A.A.A.Q., H.B., Data Collection or Processing: S.M.G., P.A.A.A.Q., Analysis or Interpretation: S.M.G., A.H., P.A.A.A.Q., H.B., Literature Search: S.M.G., Writing: S.M.G., A.H., P.A.A.A.Q.

Conflict of Interest: No conflicts of interest were declared by the authors.

Financial Disclosure: The authors declare that this study has received no financial support.

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