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Case report

An Extremely Rare Intersection: Neurolymphomatosis in a Patient with Burkitt Lymphoma Detected by 18F-Fluorodeoxyglucose Positron Emission Tomography/Computed Tomography

Ali Ozan Oner, Kursat Okuyucu[1], Engin Alagoz[1], Bilal Battal[2], Nuri Arslan[1]

Department of Nuclear Medicine, Afyon Kocatepe University, Afyon, Departments of [1]Nuclear Medicine and [2]Radiology, Gulhane Military Medical Academy, Ankara, Turkey

Abstract

Neurolymphomatosis (NL) is a rarely seen neurologic involvement of the systematic lymphoma. Its diagnosis is challenging, and requires biopsy. In cases where biopsy is not appropriate, 18F-fluorodeoxyglucose positron emission tomography/computed tomography (FDG-PET/CT) may aid in diagnosis. Here, we present a 54-year old male patient diagnosed with Burkitt lymphoma who underwent FDG-PET/CT in order to evaluate the treatment response after chemotherapy and radiotherapy. On viewing PET/CT images of the patient who complained of pain and weakness in his upper extremities after therapy, linear FDG uptake was observed in bilateral cervical 5 (C5), left cervical 6 (C6), bilateral cervical 7 (C7), and right lumbar 4 (L4) nerve roots. Magnetic resonance imaging (MRI) revealed dilation and thickening of nerve roots consisted with FDG uptake observed on PET/CT images. Since biopsy was not performed, histopathological diagnosis could not be established. However, overlapping of clinical, PET/CT, and MRI findings strongly suggested the presence of NL. As is the case of this patient, in cases with non-Hodgkin lymphoma, a combined evaluation of FDG-PET/CT and MRI modalities aid in the establishment of the diagnosis of NL.

Keywords: 18F-fluorodeoxyglucose, Burkitt lymphoma, neurolymphomatosis, positron emission tomography/computed tomography

Introduction

Neurolymphomatosis (NL) is an uncommon clinical entity caused by the infiltration of malignant lymphocytes into cranial nerves, peripheral nerves, nerve roots, or plexuses.[1] The peculiar presentations of NL are progressive sensorimotor peripheral neuropathy, cranial neuropathy, or plexopathy.[1] The diagnosis of NL is very difficult since these symptoms are also manifested by other diseases such as polyradiculopathy, mononeuropathy, Guillain-Barre syndrome, *cauda equina* syndrome, and chronic inflammatory demyelinating polyneuroradiculopathy.[2] Nerve biopsy is considered to be the gold standard for diagnosing NL.[3] In cases not appropriate for biopsy, recent imaging techniques, which have improved resolutions to the extent that affected neural structures can easily be detected such as 18F-fluorodeoxyglucose positron emission tomography/computed tomography (FDG-PET/CT) and magnetic resonance imaging (MRI) may aid in the diagnosis of NL.[4] FDG-PET/CT is being increasingly used for the diagnosis, staging and evaluation of treatment response in non-Hodgkin lymphoma (NHL) and it also appears to be a highly sensitive diagnostic method for the identification of NL.[5] In the literature, there are a lot of case reports about NL detected by FDG-PET/CT and many of these are infiltration of diffuse large B-cell lymphoma. But here, we present a patient with Burkitt lymphoma and neurologic complaints in the upper extremities on whose FDG-PET/CT were found linear hypermetabolic lesions along the peripheral nerve roots, suggestive of NL.

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Address for correspondence:
Dr. Ali Ozan Oner, Department of Nuclear Medicine, School of Medicine, Afyon Kocatepe University, Afyon, Turkey. E-mail: draliozanoner@gmail.com

Case Report

A 54-year old male patient who had swelling of the left testicle was diagnosed with Burkitt lymphoma by biopsy. On primary staging FDG-PET/CT, there were FDG accumulations in the lungs, bones, stomach, head of pancreas, right kidney, liver, spleen, thyroid gland, cervical, mediastinal, and intraabdominal lymph nodes with very high maximum standardized uptake value (SUVmax) (between 17 and 42) consistent with widespread metastasis. He was treated with Larsson+R-DHAP protocol. Pain and weakness developed in his upper extremities after therapy. Peripheral neuropathy secondary to aggressive therapy was suspected by the clinician at first glance. He underwent control FDG-PET/CT in order to evaluate treatment response 3 months after chemotherapy. Although all metastatic foci disappeared, linear FDG uptake was observed in bilateral C5, left C6, bilateral C7, and right L4 nerve roots with SUVmax values between 3.6 and 7.1 [Figure 1]. MRI revealed dilation and thickening of nerve roots where FDG uptake was observed on PET/CT images [Figure 2]. Since biopsy was not performed, histopathological diagnosis could not be established. However, overlapping of clinical, FDG-PET/CT, and MRI findings strongly suggested the presence of NL.

Discussion

As NL is extremely rare, it is a clearly defined entity.[5] Malignant lymphocytes seize the peripheral nervous system. NL appears in neurologic manifestation as the least common form of lymphoma. Baehring *et al*. declared that NL comprises 10% of all primary lymphomas of the nervous system and 0.2% of all NHLs.[6] Furthermore, metastasis rate of NHL to the nervous system is 8.5–29% and 10% of these have a predilection for the peripheral nervous system.[6]

There is a diagnostic dilemma in the detection of NL. A specific, optimal diagnostic tool is not present yet and its clinical symptoms are quite similar to those of other reasons of nonneoplastic neuropathies. The gold standard method to establish the diagnosis of NL is a nerve biopsy. But nerve biopsy is not possible in all cases because biopsy of the affected nerve might be difficult or the procedure could cause neurological sequelae. Recent studies have shown that state-of-the-art imaging modalities presented promising results for the detection of NL. Grisariu *et al*. found that FDG-PET/CT and MRI were diagnostic in 77% and 84% of the patients, respectively, in their retrospective study of 50 patients with NL while cerebrospinal fluid cytology and biopsy of nerve were positive in 40% and 88% of the patients, respectively.[7] MRI shows dilation, diffuse thickening, and contrast enhancement in the affected region of NL.[7] FDG-PET/CT is being widely used in primary staging, restaging, evaluation of treatment response in Hodgkin lymphoma (HL) and NHL with high specificity, sensitivity, and accuracy.[5] Positive findings on FDG-PET/CT are highly suggestive of a diagnosis of NL, especially in the presence of neurologic symptoms and with a history of known malignancy.[8]

In our case, it was difficult to understand the neurologic complaints of the patient before FDG-PET/CT. The clinician initially thought of peripheral neuropathy secondary to aggressive therapy. When linear moderate FDG uptake was observed in cervical nerve roots responsible for the neurologic symptoms of the upper extremity, we suspected of NL. A nerve biopsy was not possible for the patient. Whereupon MRI was requested to support FDG-PET/CT and it reinforced the diagnosis of NL, revealing the typical findings of dilation and thickening of nerve roots where FDG uptake was observed on PET/CT images. Although

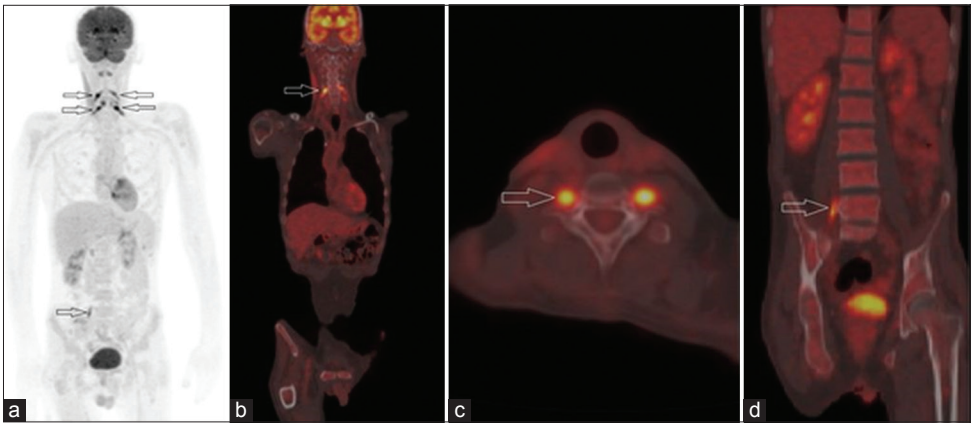

Figure 1: Positron emission tomography/computed tomography (PET/CT) maximum intensity projection (MIP) image (a) shows involvement of cervical and lumbar spinal nerve roots (arrows). Arrows indicate 18F-fluorodeoxyglucose (FDG) uptake by cervical nerve root on coronal fusion image (b), by the nerve root of C7 on axial fusion image (c), by the nerve root of L4 on coronal fusion image (d)

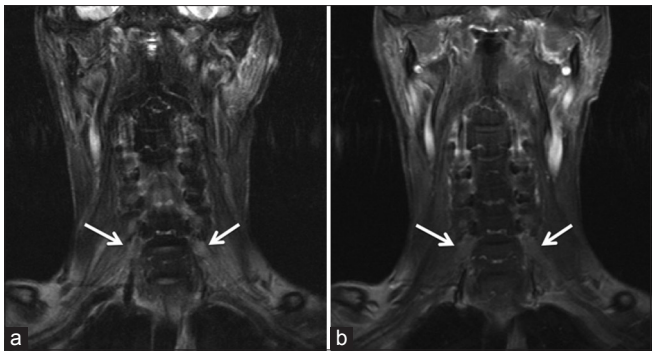

Figure 2: Coronal T2-weighted (a) and contrast-enhanced T1-weighted (b) magnetic resonance images show thickening and moderate contrast enhancement in bilateral C7 spinal nerve roots (arrows)

FDG-PET/CT and MRI may complement each other for the diagnosis of NL, if there is not an occasion for performing a nerve biopsy, FDG-PET/CT should be the first-line imaging modality because it has some unique advantages over MRI. It is able to show functional alterations that precede the anatomical changes. On the other hand, integration of CT to FDG-PET combines anatomical detail with functional information and yields excellent anatomofunctional information, increasing accuracy and detection capability. Apart from this, it can visualize the disease extent with its whole-body scanning capability. FDG-PET/CT can show subclinical lesions and early infiltration into peripheral nerves not detectable by MRI.[5] FDG-PET/CT detected L4 root involvement additionally without any complaint associated with it in our case. The combined evaluation of FDG-PET/CT and MRI contributed to the establishment of diagnosis of NL in our patient and FDG-PET/CT helped us in understanding our patient's neurologic symptoms and neglected an invasive approach.

Conclusion

When nerve biopsy and histopathological diagnosis are not possible, positive FDG-PET/CT findings strongly suggest the diagnosis of NL, especially in the presence of known NHL and with a history of neurologic symptoms. The positive findings on FDG-PET/CT are multiple, linear, and usually moderate uptake conforming to the nerve roots, plexuses, and peripheral or central nerves.

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