Neurolymphomatosis of brachial plexus: A rare clinical presentation and the role of ^{18}F-FDG PET/CT

Case Report

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ABSTRACT

Neurolymphomatosis (NLY) is a type of rare disease with poor prognosis, characterized by infiltration of nerves, nerve roots and plexus. We report a case of 54-years old female patient non-diabetic and normotensive diagnosed with chronic lymphocytic leukemia presented with pain in the neck and arm and numbness in the ipsilateral hand. The patient underwent ^{18}F fluorodeoxyglucose PET/CT scan, which showed increased tracer accumulation in the right neck and supraclavicular region in the brachial plexus. ^{18}F-FDG PETCT is highly sensitive technique for early localization of NLY than MRI or CT alone.

Key words: Neurolymphomatosis; Brachial plexus; Rare clinical presentation; ^{18}F-fluorodeoxyglucose PET/CT

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INTRODUCTION

Neurolymphomatosis (NLY) is an uncommon disease and rare neurologic disorder characterized by extranodal lymphoma, where the tumor cells invade the cranial nerves, nerve plexus, nerve root, spinal nerve roots, trunk nerves or peripheral nerves [1]. Mostly NLY occurs in aggressive subtype of leukemia or non-Hodgkin's diffuse large B-cell lymphoma (NHL) [2-4]. Predominantly, presentation is in diffuse large B-cells lymphoma, although rare cases were observed bearing follicular lymphoma, mantle cell lymphoma and peripheral T-cell lymphomas literature [5, 6]. Differential diagnosis includes nerve damage from radiation neuritis, leptomeningeal lymphomatosis, nerve root compression, herpes zoster, paraneoplastic syndromes and lymphoma-associated vasculitis [7, 8]. Here we present a rare case of NLY in a patient with chronic lymphocytic leukemia.

CASE PRESENTATION

A 54 years old female patient non-diabetic and normotensive diagnosed with chronic lymphocytic leukemia m (CLL) in February 2016. The patient was on follow up when she developed pain in her right arm and neck with progressive numbness in the right hand. The patient underwent ^{18}F- fluorodeoxyglucose PET/CT scan, which showed increased tracer accumulation in the right neck and right supraclavicular region which proven to be neurolymphomatosis on nerve biopsy (Figure 1).

DISCUSSION

NLY is a rare disorder and its incidence in currently not well known but it has been reported that NLY is more related to non-Hodgkin lymphoma (90%) and less likely to acute leukaemia (10%) [1]. In our case the patient was suffering with CLL when she developed neuropathic symptoms later on proved to be tumour involvement of brachial plexus. (Figure 1). Given the low incidence of the NLY, it should be considered when patient complains of neurological symptoms especially in the case of haematological malignancy. The symptoms may be site specific or patient may present with non-specific symptoms usually pain at a specific area, may or may not be associated with any feeing of swelling. There are different presentations of the disease where Baehring et al. described four patterns including pain, cranial neuropathy with or without pain, painless involvement of the peripheral nerves, and painless or painful involvement of single nerve [5]. In case of brachial plexus involvement as in our case, the patient mainly presents with shoulder or neck pain and a rapid onset of weakness, numbness, and paraesthesia [9]. 76% of cases with NLY present with painful symptoms hence a careful clinical examination can salvage the patient from further complications [1].

The diagnosis of NLY depend on careful patient observation, patient history, clinical /neurological examination followed by MRI or PET/CT of the suspected region. However, it is critical to distinguish these signs from other non-malignant conditions such as inflammatory radiculopathy, neurofibromatosis, and peripheral nerve sheath malignancy, [1]. Sensitivity of MRI is reported to be 40% [1] whereas, literature shows that PET-CT, sensitivity for identifying malignant peripheral nerve lesions is 87.5-100%, therefore it is especially useful in patients who have already been diagnosed with hematologic malignancy [10].

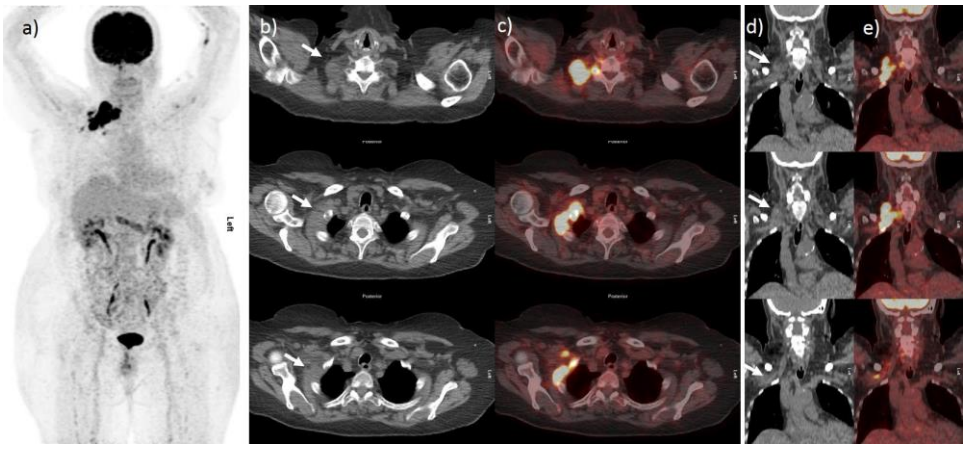

Fig 1. ^{18}F-FDG PET/CT scan of a 54 years old female diagnosed with chronic lymphocytic leukemia in February 2016. Where in a) represents MIP, b-c) CT and fused PET/CT slices, d-e) CT and fused PET/CT slices. The PET scan is showing intense uptake in the corresponding CT lesion (white arrows) in the region of the brachial plexus.

PET-CT shows abnormal FDG uptake. PET/CT is still reliable source of managing the patients despite of good sensitivity of biopsy 88% because it is sometime difficult to perform biopsy of the relevant parts and possibility of permanent nerve damage [11]. Cerebrospinal fluid cytological evaluation can be helpful in some patients, although its sensitivity is reported as 21%, which is quite low [5]. A nerve conduction test is useful for localization of the affected lesion [10]. There are no standard guidelines or treatment regimens for NLY. Chemotherapy, radiotherapy and targeted therapy has been tried in combination with intrathecal therapy, however survival rely mostly on the early diagnosis of the NLY, limiting the damage to the nerves involved.

In conclusion, Neurolymphomatosis is a rare disease with poor prognosis, therefore it should be considered wherever patient comes across neurological deficits. A prompt PET/CT is advantageous over MRI, CT and CSF cytology in detecting and localizing early disease hence limiting the possible complications by early diagnosis.

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