Case Report

Positron emission tomography/computerized tomography imaging of multiple focus of neurolymphomatosis

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ABSTRACT

Neurolymphomatosis is defined as infiltration of the peripheral nervous system by malignant lymphocytes in the presence of lymphoma. In this case, we described multiple neurol involvement and findings of ^{18}F-fluorodeoxyglucose positron emission tomography/computerized tomography in a 35-year-old female diagnosed with B-cell lymphoma.

Keywords: Lymphoma, neurolymphomatosis, positron emission tomography/computerized tomography

INTRODUCTION

Neurolymphomatosis (NL) is defined as infiltration of the cranial nerves, peripheral nerves, nerve roots, or plexus by malignant lymphocytes in the presence of lymphoma and remains a rare cause of peripheral neuropathy. It can also involve the cerebro-spinal fluid, brain and spinal cord.[1] Histopathologic involvement of NL is characterized by tumor cell infiltration of the endoneurium and perineurium. The B-cell type of non-Hodgkin lymphoma tends to be the predominant type involving the peripheral nerves. NL tends to occur with aggressive subtypes such as diffuse large B-cell lymphoma and has a variable response to current therapeutic treatments, carrying a poor prognosis. It is most common in patients who relapse, secondarily presenting solely with neurolympomatosis despite ongoing complete remission at original or other sites outside the nervous system. It presents clinically with varying degrees of peripheral neuropathy. On imaging, it is most often demonstrated by diffuse or nodular nerve root or nerve sheath thickening, and sometimes, though very rare, a mass like appearance.[2,3]

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Quick Response Code:

Website: www.ijnm.in

DOI: 10.4103/0972-3919.142632

CASE REPORT

In this case, we described multiple neural involvement and findings of fluorodeoxyglucose positron emission tomography/computerized tomography (FDG PET/CT) in a 35-year-old female diagnosed with diffuse stage IV large B-cell lymphoma, originally diagnosed in April 2013, involving the stomach, duodenum, pancreas, kidneys, and epicardium of the heart. The patient received two cycles of rituximab, cyclophosphamide, doxorubicin, vincristine, prednisolone chemotherapy, and intrathecal methotrexate. She presented 3 months later with neurological changes including blurry vision, cranial nerve VI palsy with diplopia, photophobia, left lower facial droop, right upper extremity weakness, including right wrist and hand weakness, and right leg pain. She reported having mild imbalance during walking with intermittent fevers and night sweats.

Positron emission tomography/CT demonstrated multiple areas of increased metabolic activity along several diffusely enlarged nerves, most significant along the left trigeminal nerve distribution, including V1 through V3 and near the left mandible and left tongue base [Figure 1]. Involvement within the right C7 nerve root and its distal right musculocutaneous nerve distribution was also identified. There was involvement of the bilateral sciatic nerves with extension distally along the left distal left femoral nerve near the popliteal fossa [Figure 2]. These findings were consistent with NL. In addition, there was questionable involvement of the cranial nerve XII on the right.

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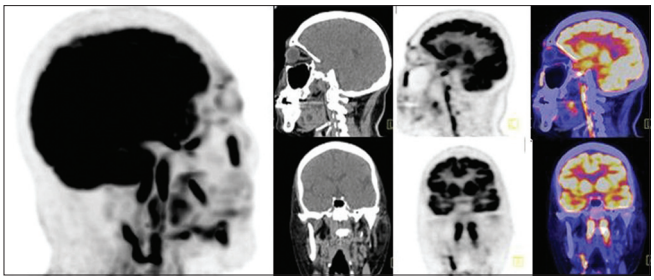

Figure 1: Cranial maximal intensity projection images of fluorodeoxglucose positron emission tomography/computerized tomography is demonstrating multiple hipermetabolic neurol involvement most significant along the left trigeminal nerve distribution, including V1 through V3 and near the left mandible and left tongue base as well as involvement within the right C7 nerve root and its distal right musculocutaneous nerve distribution

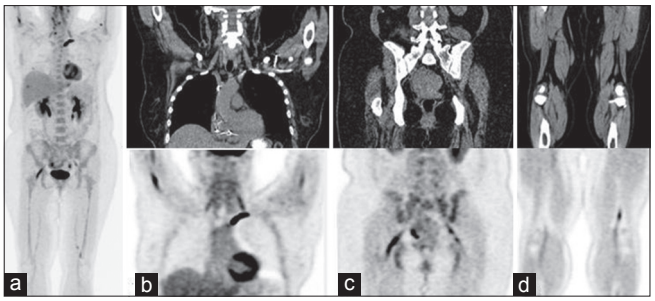

Figure 2: Whole body maximal intensity projection (a) and coronal images (b-d) of fluorodeoxyglucose positron emission tomography/computerized tomography. There is involvement of bilateral Sciatic nerve involvement most common neuropathy in neurolymphomatosis

DISCUSSION AND CONCLUSION

Neurolymphomatosis as defined by lymphomatous involvement of peripheral and cranial nerves remains rare. This usually develops in patients with nonHodgkin's lymphoma, often presenting after remission, with its sole re-presentation within the neurologic system. It may present clinically as painful polyneuropathy or polyradiculopathy, followed by cranial neuropathy, painless polyneuropathy and peripheral mononeuropathy.[1,2,4] Of note, most of the lymphomatous involvement of peripheral nerves described in the literature involves the sciatic nerve.[5] It is described during or following chemotherapy with good response to the original presenting systemic disease. Most case reports demonstrate diffuse or solitary thickening of the nerves/nerve sheath and nerve roots, and rarely, as a mass.[6-8] The differential considerations are Gillian Barré syndrome, herpes zoster, demyelinating polyradiculoneuropathy, vinca alkaloid toxicity, mechanical compression of nerve roots, and systemic amyloidosis.[2] In diagnosis of NL the sensitivity of nerve biopsies was 80%, of MRI was 40% and cerebrospinal fluid examination was 21%.[2]

During the progressive disease or therapy following aggressive lymphoma, peripheral neuropathy may be difficult to distinguish and the differential, particularly given the patient's immune compromised state, remains broad. NL fortunately remains rare as it carries a poor prognosis. However, PET/CT is useful diagnostic tool in the evaluation and follow-up of the patient's, often sufficient for diagnosis in the appropriate clinical setting, with characteristic neuroimaging findings; primarily demonstrated by diffuse or nodular nerve root or nerve sheath thickening, and rarely, a mass like appearance.

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How to cite this article: Peterson J, Caliskan B, Bonyadlou S. Positron emission tomography/computerized tomography imaging of multiple focus of neurolymphomatosis. Indian J Nucl Med 2014;29:252-3.
Source of Support: Nil. **Conflict of Interest:** None declared.