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The Diagnostic Dilemma of Neurolymphomatosis

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Neurolymphomatosis (NL) defined as infiltration of the central nervous system or the peripheral nervous system (PNS) by malignant lymphoma cells is a rare clinical entity. However, the increasing use of fluorodeoxyglucose positron-emission tomography (FDG-PET) and magnetic resonance imaging in evaluating PNS disorders is resulting in; this condition being recognized more frequently. Here; we report five NL patients and review the current literature. We report five patients with non-Hodgkin's lymphoma (NHL) and NL, all of whom were men aged 47–69 years. The clinical presentation varied from symmetrical peripheral neuropathy to mononeuropathy. Peripheral neuropathy was the presenting manifestation of a systemic lymphoma in two patients (40%). Neuroimaging as well as whole-body FDG-PET helped in determining the correct diagnosis in all of the patients. NL is an unusual presentation of NHL resulting from infiltration of the PNS by malignant lymphomatous cells. While evaluating peripheral neuropathy, a high degree of suspicion of NL is required since the presenting symptoms vary, conventional radiology has only modest sensitivity, and a pathological diagnosis is often difficult. FDG-PET helps in the early diagnosis and treatment of this condition.

Key Words neurolymphomatosis, diffuse large-B-cell lymphoma, mononeuropathy, fluorodeoxyglucose positron-emission tomography.

INTRODUCTION

Neurolymphomatosis (NL), defined as invasion of the peripheral nervous system (PNS) (cranial nerves, peripheral nerves, roots, and plexus) by lymphoma cells, is a rare clinical entity;[1] being the least common way in which lymphoma can involve the PNS. It is important to differentiate NL, the common mode of PNS involvement in lymphoma, from other causes of PNS involvement in lymphoma (e.g., paraneoplastic PNS involvement, and radiation-and chemotherapy-induced damage to the PNS), since their treatment modalities differ.[2] The increasing use of positron emission tomography (PET) and magnetic resonance imaging (MRI) in evaluating PNS disorders is resulting in increasing numbers of NL cases being recognized.

Here we report five NL patients and review the current literature regarding the diagnosis and management of this rare condition.

PATIENT 1

Mr. A, a 69-year-old male, presented to us with a two month history of shooting pains in the bilateral gluteal regions and the back of the thigh, followed 1 month later by pin prick paresthesias in both upper limbs and the right half of the face, along with asymmetrical quadriparesis affecting the legs more than the arms, low-grade fever, dry cough, orthop-

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