Bilateral Vagus Nerve Neurolymphomatosis Diagnosed Using PET/CT and Diffusion-Weighted MRI

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Abstract: In neurolymphomatosis, malignant lymphocytes infiltrate the peripheral nervous system in the presence of a known or unknown hematological malignancy. This report describes the findings of diffusion-weighted MRI and 18F-FDG PET/CT in a 65-year-old man with hoarseness. Results revealed a mass with restricted diffusion on diffusion-weighted imaging in the right visceral vascular space, increased uptake of 18F-FDG, and other masses at distant peripheral nerves. Restaging PET/CT showed involvement of the right brachial plexus and right sciatic nerve. Biopsy and immunohistochemistry of the right vagus nerve and cervical lymphadenopathy revealed a diffuse large B-cell non-Hodgkin lymphoma.

Key Words: neurolymphomatosis, MRI, diffusion, PET/CT

REFERENCES
FIGURE 1. A 65-year-old man was investigated for hoarseness. Laryngoscopy revealed paralysis of his right vocal cord, and clinical examination showed a palpable right cervical lymph node. T1- (A) and T2-weighted (B) MRI of the skull base in the axial plane and contrast-enhanced T1-weighted fat-saturated images in the axial (C, D) and coronal (E) planes show an ill-defined, elongated mass in the right vascular space between the internal carotid artery and the jugular vein. There was intermediate signal intensity in both T1- and T2-weighted sequences and homogeneous enhancement extending through the cisternal segment of the vagus nerve. These features are nonspecific, and diagnosis of a malignant peripheral nerve sheath tumor was considered. F to H, Lymph node specimen. F, Diffuse large B-cell lymphoma, not otherwise specified; hematoxylin-eosin staining; magnification, ×100. G, Diffuse membrane labeling for CD20; magnification, ×400. H, Few reactive lymphocytes labeled for CD3; magnification, ×400. Neurolymphomatosis (NL) is a rare neurologic presentation of non-Hodgkin lymphoma (NHL) and leukemia and involves neural infiltration by malignant lymphocytes. Neurolymphomatosis usually precedes the systemic disease and diagnosis of the condition requires (1) histological demonstration of malignant lymphocytes within the peripheral or cranial nerve, root or plexus; or (2) radiologic or intraoperative evidence of nerve enlargement beyond the dural sleeve and/or enhancement in the presence of a primary central nervous system lymphoma or systemic NHL. There are 4 major clinical presentations: painful polyneuropathy or polyradiculopathy, cranial neuropathy, painless peripheral polyneuropathy, and peripheral mononeuropathy. In 58% of cases, more than 1 anatomical structure is affected. Neurolymphomatosis is distinct from subarachnoid seeding or perineural infiltration of an epidural lymphoma, inflammatory neuropathies, and neuropathic complications of treatment.
FIGURE 2. Restaging MRI performed 2 months later. Axial T1- (A), T2- (B), diffusion-weighted imaging (DWI) (C), apparent diffusion coefficient (D), contrast-enhanced T1-weighted imaging in the axial (E), and coronal planes with fat suppression (F) showing an increase in the size of the lesion in the right vagus nerve and a new lesion in the left vagus nerve. Intermediate signal in T1- and T2-weighted images, restricted diffusion, and homogeneous enhancement were observed. MRI findings of enlarged neural structures with contrast enhancement constitute the result most indicative of this diagnosis, with sensitivity of 40% to 70% when associated with a known history of NHL. MRI sensitivity is greater than that achieved with cerebrospinal fluid analysis, which is reported to range from 21% to 40%; however, sensitivity with both these techniques is very low compared with the sensitivity of 80% achieved with nerve biopsy.\(^1,^2\) Restricted diffusion on DWI may serve to differentiate lymphoma from other tumors because of its high cellularity.\(^3\) Concomitant findings at PET/CT and MRI in NL have already been published,\(^4\) but to the best of our knowledge, this is the first case of NL showing restricted diffusion on DWI and its association with PET/CT, which may be useful in defining diagnosis.
FIGURE 3. PET/CT (A) and MRI fused with PET/CT (B) show increased uptake bilaterally in the vagus nerves. PET/CT and 3D volume rendering technique (C, D) show increased uptake at the visceral vascular spaces (long thin arrows), in left cervical lymphadenopathy (short thin arrow), at the projection of a paravertebral nerve root at the level of the lower thoracic spine and at the left lumbosacral plexus (short thick arrows). The lesions shown in vascular spaces are in agreement with the findings of the DW images (Figs. 2C, D). A restaging PET-CT and 3D volume rendering technique (E, F) performed 7 months later show new sites of uptake involving the right brachial plexus (long thin arrow) and right lumbosacral plexus (short thin arrow) and improvement at the other sites, reflecting a mixed metabolic response and progression of the disease, indicative of significant intrapatient heterogeneity of gene expression profiles. If clinical presentation, cerebrospinal fluid analyses, and imaging studies are inconclusive, biopsy of neural tissue is required.\(^1,2\) The PET/CT sensitivity of 87% is helpful in diagnosing NL in the presence of a known hematological malignancy, even in the case of lesions in the subclinical phase and principally when the set of tests used for evaluation yield inconclusive results.\(^2,5,7\) The present case also demonstrates the usefulness of FDG-PET/CT in NL for the evaluation of therapeutic response, as previously reported by Talanow and Shrikanthan.\(^8\)