Neurolymphomatosis as a manifestation of relapsed primary cardiac lymphoma

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A 68-year-old Japanese woman was admitted to our hospital because of congestive heart failure and sinus node dysfunction. ^{18}F -Fluorodeoxyglucose-positron emission tomography/computed tomography (FDG-PET/CT) revealed a cardiac tumor involving right atrium of the heart (Fig. 1a, b), and the tumor was diagnosed as primary cardiac diffuse large B cell lymphoma by open chest biopsy. The immunophenotype of the lymphoma cells was $\text{CD}3^-$, $\text{CD}5^-$, $\text{CD}10^-$, $\text{CD}19^-$, $\text{CD}20^+$, $\text{CD}22^+$, smIgM^+ , and $\text{smIg}\kappa^+$. The

Fig. 1 Images of FDG-PET/CT. **a** A maximum intensity projection (MIP) image of the whole body and **b** a fusion image (horizontal section) at the onset of primary cardiac lymphoma. These images show the lymphoma mainly involved right atrium (**a** black arrow, **b** white arrow). **c** A MIP image of the whole body, and **d** a coronal section of the abdomen and **e** a horizontal section of the lumbar region of the fusion images at the onset of neurolymphomatosis. These images show that the linear lesions emerged from intervertebral foramens (**d, e** white arrowheads) and are located along brachial plexus, lumbosacral plexus, brachial nerves, and sacral nerves (**c** black arrowheads). These also show the small recurrence of the lymphoma in the heart (**c, d** white arrows). **f** A MIP image of the whole body 3 weeks after the start of re-administration of high-dose methotrexate. This shows that the abnormal FDG-uptake disappeared. In these figures, physiological FDG uptake is observed in the brain, gastro-intestinal tract, urinary tracts, a urinary catheter, and the operation scar of the open chest biopsy on the sternum

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staining for Epstein–Barr virus-encoded small RNA was negative. After administration of 3 courses of CHOP-like regimen with rituximab, the cardiac tumor was dramatically

shrunk; however, she developed nausea. Cerebrospinal fluid (CSF) analysis revealed leptomeningeal involvement of the lymphoma. After 3 courses of high-dose-methotrexate-containing regimen, CSF analysis and FDG-PET/CT revealed complete remission of the disease. Two months later, however, she experienced rapid onset of neurogenic bladder dysfunction, paralysis of legs, and numbness of extremities. Magnetic resonance imaging for brain and vertebra and CT scan of the whole body revealed no abnormal findings. CSF analysis revealed absence of abnormal cells and normal glucose level, but slight increased level of total protein (69.1 mg/dl). FDG-PET/CT, however, showed high accumulations along brachial plexus and nerves, lumbosacral plexus, and sacral nerves (Fig. 1c–e), suggesting neurolymphomatosis. After re-administration of high-dose methotrexate, her neurological symptoms subsided immediately, and the FDG-PET/CT after 3 weeks showed no abnormal uptake (Fig. 1f).

Primary cardiac lymphoma is a rare malignant lymphoma accounting for 0.5% of all the extranodal malignant lymphomas, and neurolymphomatosis is also a rare manifestation of lymphoma. To our knowledge, this is the first report of neurolymphomatosis as a manifestation of relapsed primary cardiac lymphoma. Although it has been difficult to make a successful diagnosis with primary cardiac lymphoma, newly developed FDG-PET/CT enables to visualize the lymphomatous lesions in the heart clearly. Neurolymphomatosis is often misdiagnosed mainly because CT scan hardly detects the lesions. Our case suggests that FDG-PET/CT is a powerful tool for detecting the lesions of neurolymphomatosis. In conclusion, we encountered a rare case with primary cardiac lymphoma evolving into neurolymphomatosis, suggesting that FDG-PET/CT is very useful in diagnosing these conditions.