Neurolymphomatosis: A Case Study of Diffuse Large B-Cell Lymphoma

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Neurologic changes can be seen during treatment for hematologic malignancies such as lymphoma and leukemia. Differential diagnoses for neurologic changes remain vast, and etiologies may result from malignant, nonmalignant, and paraneoplastic syndromes. This article describes a case study of a man with neurolymphomatosis, a rare neurologic manifestation seen in B-cell and non-Hodgkin lymphoma and acute leukemias.

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xperiencing dyspnea and profound weakness, B.L., a 64-year-old man, presented to the emergency room in February 2011. He had a white blood cell count of 4,300 mcl, hemoglobin level of 5.3 g/dl, and platelet count of 16,000 mcl. The initial peripheral blood smear appeared to show acute lymphoblastic leukemia. However, the final pathology revealed high-risk, CD20-positive, stage IVB diffuse large B-cell lymphoma. B.L. also presented with high-risk features including bone marrow involvement, a palpable spleen, elevated lactate dehydrogenase, and poor performance status.

B.L. was treated with standard R-CHOP (rituxan, cyclophosphamide, vincristine, doxorubicin, and prednisone) chemotherapy for six cycles. Although the initial staging lumbar puncture was negative, with his aggressive leukemic phase presentation and bone marrow involvement, B.L.'s central nervous system was prophylaxed during treatment with intrathecal methotrexate biweekly. Post-treatment positron-emission tomography/computed tomography (PET/CT) scan was consistent with complete remission, and cerebrospinal fluid analysis remained negative throughout treatment. B.L. then was seen at a local academic

medical center for consideration of peripheral stem cell transplantation.

During pretransplantation evaluation, B.L. was noted to have evolving progressive new lower extremity weakness over three to four weeks, necessitating the use of a wheelchair. That new weakness delayed the transplantation as the next four months involved multiple hospitalizations for continued neurologic deterioration. His neurologic changes included development of left third cranial nerve palsy and severe bilateral motor neuropathy with a component of sensory neuropathy. Repeat PET/CT scan of the brain and spine and cerebrospinal fluid analysis remained negative. A magnetic resonance imaging (MRI) scan was not performed because B.L. had a permanent pacemaker.

Neurology was consulted and B.L. was diagnosed with an atypical Guillian-Barre syndrome and treated with high-dose steroids and IV gammaglobulin. B.L. had a minor initial response with neurologic improvement as shown by some ability to move his legs. When B.L. began to decline neurologically again, the clinical course was reevaluated. The diagnosis was changed to chronic inflammatory demyelinating polyneuropathy, and B.L. was rechallenged with high-dose steroids and

IV gammaglobulin, despite no evidence of recurrent disease. B.L.'s condition continued to decline, and seven months after his initial diagnosis (three months after the onset of neurologic decline), he transitioned to palliative care, where he died. Given the unusual neurologic manifestations, a postmortem examination was performed of the spinal cord and nerves, which showed diffuse infiltrates of his original diffuse large B-cell lymphoma within the peripheral nerves of the cauda equina, leading to paralysis and a final diagnosis of neurolymphomatosis (NL).

Neurolymphomatosis

NL is a rare neurologic manifestation most often associated with B-cell non-Hodgkin lymphoma but also seen with leukemia where malignant cells infiltrate the nerve root, resulting in neurologic deterioration (Grisariu et al., 2010). The incidence of NL is unknown because a majority of the information available is based on case reports, with Grisariu et al. (2010) citing only 72 cases of NL occurring since 1980. The main characteristics associated with NL include painless or painful involvement of singular or multiple peripheral nerves, nerve roots, or cranial nerves (Baehring, Damek, Martin, Betensky, & Hochberg, 2003; Cheung et al., 2012; Grisariu et al., 2010).

Diagnostic Evaluation

Diagnosing NL often is challenging because the symptoms can fit a variety of diagnoses. Patients often present with diffuse disease, but single-site peripheral nerve involvement with no evidence of disease elsewhere, including the cerebrospinal fluid, has been reported (Baehring et al., 2003; Gan, Azad, Cher,