Teaching NeuroImages: Neurolymphomatosis

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A 64-year-old woman with a history of diffuse large B-cell lymphoma presented with 3 months of progressive pain and weakness. Examination was notable for asymmetric face, arm, and leg weakness with absent reflexes. CSF was normal twice, including cytopathology and flow cytometry. PET showed widespread avidity (figure 1), including the bilateral brachial and lumbosacral plexi. Nerve conduction study/EMG showed reduced motor amplitudes but normal sensory responses, indicating that Wallerian degeneration had not yet progressed distal to the dorsal root ganglia—underscoring the urgency of timely diagnosis and treatment. Sciatic nerve biopsy confirmed neurolymphomatosis (figure 2). She received 2 cycles of rituximab plus ifosfamide, carboplatin, and etoposide chemotherapy with resolution of PET avidity, followed by autologous bone marrow transplant. Neurolymphomatosis is characterized by malignant invasion of nerves, often presenting with severe, asymmetric pain.¹ CSF studies have low sensitivity; thus, diagnosis often depends on PET and biopsy.² Symptoms and imaging abnormalities often resolve with systemic chemotherapy; however, relapse is common—with 1 large case series showing a median survival of 10 months and 24% survival at 36 months.²

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Figure 1 PET showed widespread fluorodeoxyglucose avidity (A), including right facial nerve (B), sciatic and femoral nerves (C), bilateral brachial and lumbosacral plexi (D), and C3/C4 roots (E), with resolution of PET avidity after chemotherapy (F)

Author contributions

S.R. DeBoer and S. Lesche: drafting/revising the manuscript, data acquisition, study concept or design, analysis or interpretation of data, accepts responsibility for conduct of research, and will give final approval. F. J. Rodriguez: data acquisition, analysis or interpretation of data, accepts

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Figure 2 Sciatic nerve fascicle biopsy (400×)

H&E (A) shows infiltration and disruption by malignant cells. Anti-CD20 immunostain (B) demonstrating large neoplastic B cells, in a background of reactive CD3⁺ T cells (not shown).

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