

## Tumefactive Appearance of Hematologic Malignancies Involving Peripheral Nerves: A New Imaging Pattern Stepan Capek MD; Marie-Noelle Hebert-Blouin MD; Ross Puffer MD; Carlo Martinoli MD; Matthew A. Frick MD; Kimberly K. Amrami MD; Robert J. Spinner MD

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### Introduction

The diagnosis of neurolymphomatosis (NL) can be enigmatic, especially in primary disease. MRI plays a crucial role: it helps not only better localize the disease (and improve the yield of targeted biopsy), but can provide clues to identify the etiology. The nerves are typically described to be enlarged, hyperintense on T2WI sequences and enhancing on gadolinium scans. This description is highly non-specific and can be seen is other conditions as chronic inflammatory demyelinating polyneuropathy. We recently became aware of a "tumefactive" pattern of NL, neuroleukemiosis (NLK) and neuroplasmacytoma (NPLC), which we believe is exclusive to hematologic diseases affecting peripheral nerves.

### **Material & Methods**

We defined the "tumefactive" appearance as "complex, fusiform, circumferential tumor masses encasing the involved peripheral nerves. Both structures show varying level of homogenous enhancement on gadolinium scans and hyperintense signal on T2WI. The nerves appear to be infiltrated by the tumor causing separation of individual fascicles". We retrospectively reviewed our institutional series of all cases of NL (n=52) diagnosed between 1998 and 2014 in search for this pattern; 2 extra cases of NL, 3 cases of NLK and 1 case of NPLC were added to the series based on their known appearance.

# Results

We identified 20 "tumefactive" lesions in 18 patients. Mean age was 64.4 years. Nine patients were females and 9 males. Patients presented as recurrence (n=7), NL as the first symptom of a systemic disease (n=5), neurological involvement during the course of the disease (n=3), and as primary NL or NLK (n=3). The patients presented with weakness (n=15), numbness (n=10), pain (n=10) or paresthesias (n=6). Two patients had no symptoms.

Fig 1 - An example of high-grade NL

An axial (A) and sagittal (B) T2 fat saturated (FS) images demonstrate an enlarged sciatic nerve with preserved fascicular architecture (arrowheads) surrounded by a soft-tissue mass (arrows). Similar situation is depicted on the gadolinium enhanced spoiled gradient recall (SPGR) images (C,D; sciatic nerve – arrowhead, tumor mass - arrows). Results

The brachial plexus ( =7) was mostly affected, followed by the sciatic nerve (n=6) and lumbosacral plexus (n=3). 4 patients had involvement of other nerves. Biopsy was performed and was positive in all cases (n=18). The final diagnosis was diffuse large B-cell lymphoma (DLBCL)(n=12)(Fig. 1), acute myeloid leukemia (n=2), marginal zone lymphoma (n=1)(Fig. 2), low grade non-Hodgkin lymphoma (NHL) not otherwise specified (n=1), low grade NHL with plasmacytic differentiation (n=1) and plasmacytoma (n=1)(Fig. 3).



An axial (A) and sagittal (B) T2 FS MR images demonstrate the infiltrated sciatic nerve (arrowheads) surrounded by a hyperintense lymphomatous mass (arrows). The mass (arrows) and the nerve (arrowheads) show similar level of enhancement on gadolinium SPGR sequences (C,D).

### Fig 3 - An example of NPLC



An axial T2 FS image (A) demonstrates the enlarged and infiltrated sciatic nerve (arrowhead) surrounded by a tumorous mass (arrows), which together with the nerve (arrowheads) avidly enhances on gadolinium enhanced sagittal (B) and axial (C) SPGR images. A coronal combined Fdeoxy-glucose PET/CT image demonstrates an area of increased uptake (D – arrow), which correlates with the tumor seen on MRI.

#### Conclusions

We present a new imaging pattern of tumefactive neurolymphomatosis, neuroleukemiosis or neuroplasmacytoma in a series of 18 cases. We believe this pattern is associated with hematologic diseases directly involving the peripheral nerves. Knowledge of this association can provide a clue to clinicians in establishing the correct diagnosis.