Multifocal acquired demyelinating sensory and motor neuropathy

Figure MRI and fascicular biopsy of the right brachial plexus

(A) MRI of the right C7 and C8 nerve roots (arrowheads) and brachial plexus (arrow) demonstrated marked swelling and hyperintensity on T2-weighted sequences. (B) Fascicular biopsy of the right brachial plexus revealed numerous large "onion-bulbs" (reflecting chronic demyelination/remyelination), surrounding small, thinly myelinated axons on Toluidine blue staining (arrows).

A 54-year-old man presented with a 10-year history of progressive weakness, tingling, and pain involving the left lower and right upper extremities. Examination revealed moderate weakness of right radial, median, ulnar, and left tibial-innervated muscles with sensory loss in right hand digits 4/5 and the left sole. Electrophysiologic testing disclosed a left sciatic neuropathy (tibial division) and right sensorimotor brachial plexopathy with axon loss physiology. A diagnosis of multifocal acquired demyelinating sensory and motor neuropathy (MADSAM), a multifocal variant of chronic inflammatory demyelinating polyneuropathy, was made following an MRI and fascicular biopsy of the right brachial plexus¹ (figure). Muscle strength, paresthesias, and pain improved following IV immunoglobulin therapy.²

Araya Puwanant, MD, David N. Herrmann, MBBCh, Rochester, NY

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Correspondence & reprint requests to Dr. Puwanant: araya_puwanant@urmc.rochester.edu

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