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[The Importance of Rare Subtypes in Diagnosis and Treatment of Peripheral Neuropathy: A Review.](#)

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Abstract

IMPORTANCE: Peripheral neuropathy is a prevalent condition that usually warrants a thorough history and examination but has limited diagnostic evaluation. However, rare localizations of peripheral neuropathy often require more extensive diagnostic testing and different treatments. **OBJECTIVE:** To describe rare localizations of peripheral neuropathy, including the appropriate diagnostic evaluation and available treatments. **EVIDENCE REVIEW:** References were identified from PubMed searches conducted on May 29, 2015, with an emphasis on systematic reviews and randomized clinical trials. Articles were also identified through the use of the authors' own files. Search terms included common rare neuropathy localizations and their causes, as well as epidemiology, pathophysiology, diagnosis, and treatment. **FINDINGS:** Diffuse, nonlength-dependent neuropathies, multiple mononeuropathies, polyradiculopathies, plexopathies, and radiculoplexus neuropathies are rare peripheral neuropathy localizations that often require extensive diagnostic testing. Atypical neuropathy features, such as acute/subacute onset, asymmetry, and/or motor predominant signs, are frequently present. The most common diffuse, nonlength-dependent neuropathies are Guillain-Barré syndrome, chronic inflammatory demyelinating polyneuropathy, multifocal motor neuropathy, and amyotrophic lateral sclerosis. Effective disease-modifying therapies exist for many diffuse, nonlength-dependent neuropathies including Guillain-Barré syndrome, chronic inflammatory demyelinating polyneuropathy, multifocal motor neuropathy, and some paraprotein-associated demyelinating neuropathies. Vasculitic neuropathy (multiple mononeuropathy) also has efficacious treatment options, but definitive evidence of a treatment effect for IgM anti-MAG neuropathy and diabetic amyotrophy (radiculoplexus neuropathy) is lacking.

CONCLUSIONS AND RELEVANCE: Recognition of rare localizations of peripheral neuropathy is essential given the implications for diagnostic testing and treatment. Electrodiagnostic studies are an important early step in the diagnostic evaluation and provide information on the localization and pathophysiology of nerve injury.

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