

Peripheral nerve: Myelin, compact, uncompact, widely spaced and anti-Myelin Associated Glycoprotein neuropathy.

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Abstract

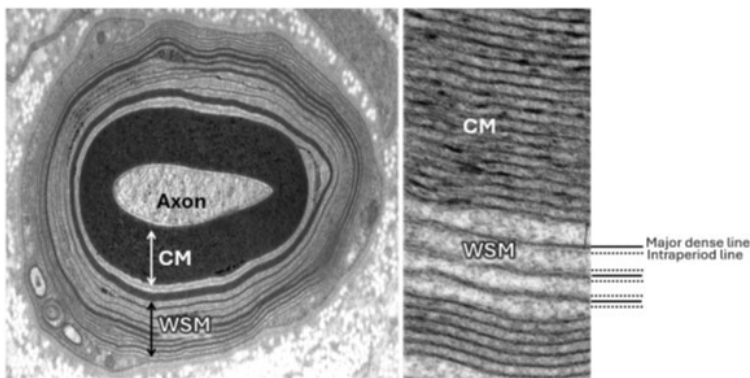
Learning Objectives:

1. Understand normal peripheral nerve anatomy, histology and ultrastructure
2. Identify the ultrastructure of normal myelin and artefacts in myelin.
3. Understand the pathogenesis of anti-Myelin Associated Glycoprotein (MAG) neuropathy
4. Identify the ultrastructure of widely spaced myelin and uncompact myelin.

Case Report: A 70-year-old male with a history of B-cell lymphoma presented with chronic progressive peripheral neuropathy. Differential diagnoses included amyloidosis, intraneural lymphoma, and anti-MAG neuropathy. Sural nerve biopsy plastic sections showed reduced numbers of large myelinated fibres with no evidence of amyloid or lymphoma. Ultrastructurally, these large myelinated fibres showed widely spaced myelin, characteristic of anti-MAG neuropathy, Figure1.

Discussion: Anti-MAG neuropathy results from IgM autoantibodies targeting the myelin-associated glycoprotein (MAG). These antibodies disrupt myelin compaction, leading to widely spaced lamellae. I will discuss the origin of anti-MAG antibodies, the location of MAG in myelin, and the mechanism of myelin disruption. I will differentiate widely spaced myelin from uncompact myelin.

Key Takeaway: Widely spaced myelin is the diagnostic ultrastructural feature of anti-MAG neuropathy.



Left panel, a sural nerve fibre shows a widely spaced myelin (WSM) of the outer layers, compared to normal compact myelin (CM). Right panel, higher magnification showing myelin lamellae separation between the intraperiod line (.....), with intact major dense line (—), diagnostic of anti MAG neuropathy.

References

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