American Association of Neuropathologists

Peripheral Nerve Biopsies

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Case-Based Questions (please see page 3 for answers)

1.	Different types of vasculitis can affect peripheral nerves. Two main subgroups are patients with secondary vasculitic neuropath in the context of systemic vasculitis and those with nonsystemic vasculitic neuropathy (NSVN). The following statement is true about NSVN:	
	a.	NSVN typically affects the larger vessels in a nerve biopsy.
	b.	NSVN is typically more rapidly progressive than cases of systemic vasculitis.
	c.	NSVN can be difficult to diagnose because of the lack of systemic manifestations.

2.	One possible finding in peripheral nerve biopsies are amyloid deposits. What statement is true about patients with this finding:	
	a.	The typing of the amyloid found in a biopsy has important implications for treatment.
	b.	Amyloid is always deposited around epineurial vessels.
	c.	The amyloid found in peripheral nerve biopsies is usually beta amyloid.
	d.	The most common pattern of nerve damage in amyloid neuropathy is one of demyelination.

3.	Leprosy is a common cause of infectious peripheral neuropathy in some parts of the world. What statement about leprous neuropathy is true:	
	a.	Leprous neuropathy is always associated with a pattern of granulomatous inflammation.
	b.	Identifiable organisms may be absent in tuberculoid leprosy but are typically found in the lepromatous form.
	c.	Bacterial cultures are a reliable test to detect Mycobacterium leprae.

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Correct Answers and Rationales

Question 1 Correct Answer and Rationale**: C. NSVN can be difficult to diagnose because of the lack of systemic manifestations.**

Rationale: NSVN typically presents with a more slowly progressive course than patients with nerve involvement by systemic vasculitis. The lack of systemic manifestations is often one of the reasons for a delay in diagnosis. NSVN typically affects small vessels in a nerve biopsy.

Question 2 Correct Answer and Rationale: **A. The typing of the amyloid found in a biopsy has important implications for treatment**.

Rationale: Amyloid neuropathy can be the result of primary amyloidosis (i.e. derived from immunoglobulin light chains) or secondary amyloidosis (e.g. transthyretin). Beta amyloid is not found in peripheral nerve biopsies. Determining the type of amyloid clinically or through laboratory testing is important to guide treatment decisions, especially with the advent of new drugs for some forms such as transthyretin amyloidosis (PMID: 38923548). Amyloid deposits can be found in different distributions in a nerve biopsy but are most often endoneurial. The predominant pattern of nerve damage is one of axonal degeneration.

Question 3 Correct Answer and Rationale: **B. Identifiable organisms may be absent in tuberculoid leprosy but are typically found in the lepromatous form.**

Rationale: Leprosy is one of the infectious etiologies of peripheral neuropathy and can have a long incubation period of multiple years. The different tissue manifestations represent a spectrum ranging from the tuberculoid and the lepromatous form. The former is characterized by extensive, destructive inflammation with granulomatous features but often no identifiable organisms. The latter is characterized by numerous organisms within foamy macrophages and Schwann cells. M. leprae is an obligate intracellular parasite and therefore difficult to grow in culture. Molecular testing can confirm the diagnosis.