

Amyloid Stain (AS)

Neuropathy can be an early manifestation of systemic amyloidosis. Such patients often present with a painful distal and symmetric neuropathy and autonomic symptoms. It can begin as a small fiber neuropathy and progress to involve the large sensory and motor axons (Yang et al, 2010). Involvement of other organs can also cause myopathy, congestive heart failure, or renal failure.

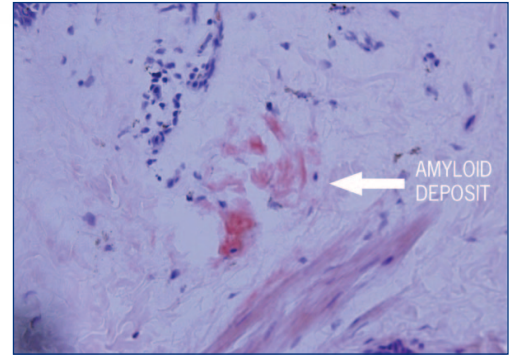
Biopsy studies can show deposits of amyloid fibrils in the nerves and other tissues, including the skin (Nagasaka et al, 2009). The amyloid fibrils appear as reddish deposits using Congo red staining, and as apple-green birefringence using polarized light microscopy (bottom image).

Amyloid deposits differ in composition, based on the underlying cause. Primary amyloidosis is associated with monoclonal gammopathy and the amyloid fibrils are made up of fragments of the monoclonal immunoglobulin light chains (Kissel and Mendell, 2001). In secondary, or hereditary amyloidosis, the fibrils are composed of mutated proteins, most often transthyretin, and less frequently apoprotein A-1 or gelsolin (Mendell, 2001). Amyloid deposits, once detected, can be typed using immunohistochemistry.

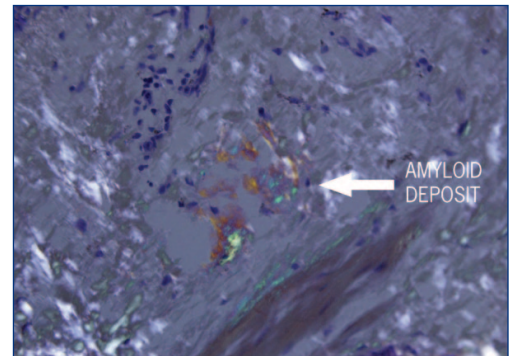
Amyloid deposits can occur in a spotty distribution, so that the absence of amyloid deposits in a skin biopsy does not rule out the diagnosis of amyloid neuropathy. Biopsy of other tissues including abdominal fat, bone marrow, rectal mucosa, or another affected organ, including nerve, may be required to make the diagnosis in individual patients.

**More information on the amyloid stain, including a complete list of references can be found on our website.*

Positive Amyloid Stains



Amyloidosis in skin of patient with amyloid neuropathy, as detected by Congo Red stain. The arrow points to the amyloid deposits.



Amyloidosis observed with polarizing light microscopy. The arrow points to the apple-green birefringence of the amyloid deposit, previously detected by Congo Red staining, confirming the diagnosis.