# Amyloidosis

Amyloidosis may be a systemic or localized process. It may occur spontaneously or in response to chronic disease processes.

## Classification

Systemic amyloidosis can be hereditary or acquired. The autosomal dominant hereditary transthyretin amyloidosis and the acquired light-chain amyloidosis, the result of a plasma cell dyscrasia, are multisystem disorders with cardiovascular, autonomic and peripheral nerve involvement. Primary amyloidosis is characterized by no detectable plasma cell dyscrasia or abnormal serum proteins; secondary amyloidosis involves systemic disease, in which the breakdown of cells is protracted.

Primary localized amyloid deposits in bone are unusual and any bone may be involved. Skeletal amyloid deposits often have an associated soft-tissue mass that may contain variable amounts of calcification.

The lesions grow slowly and can produce significant local destruction of bone and soft tissue. It is this tumor-like appearance and behavior that make the diagnosis difficult to establish on imaging studies.

see Spinal amyloidoma.

see Transthyretin amyloidosis.

## Diagnosis

There are numerous investigational modalities available to diagnose systemic amyloidosis and to assess the extent of organ involvement, but it is frequently misdiagnosed due to its heterogeneous clinical presentations and misleading investigation findings. An accurate and timely diagnosis of amyloid neuropathy can greatly impact on the outcomes for patients, especially as there will soon be new gene-silencing treatments for hereditary transthyretin amyloidosis [1].

1)
Kapoor M, Rossor AM, Jaunmuktane Z, Lunn MPT, Reilly MM. Diagnosis of amyloid neuropathy. Pract Neurol. 2018 Dec 30. pii: practneurol-2018-002098. doi: 10.1136/practneurol-2018-002098. [Epub ahead of print] PubMed PMID: 30598431.