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Amyloidosis

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Amyloidosis describes a group of rare diseases characterised by the deposition of abnormal insoluble proteins in the interstitium between cells, causing organ damage. Clinical hallmarks can be renal insufficiency, congestive heart failure, gastrointestinal symptoms, neuropathy, and skin changes.

A common cause of primary amyloidosis is the production of monoclonal immunoglobulin light-chains by a lymphoproliferative disease, e.g. multiple myeloma. This form is named AL-Amyloidosis.

Familial amyloidosis, also termed familial amyloid polyneuropathy or TTR-associated amyloidosis (ATTR), is characterised by a mutation in the TTR-gene (Transthyretin, transporter of thyroxine and retinol).

In AA-amyloidosis fibrils that are composed of fragments of serum amyloid-A protein are deposited between cells. AA-amyloidosis is the most common form of amyloidosis worldwide and can develop during the course of chronic inflammatory diseases as Mediterranean fever or rheumatoid arthritis. The organs that are mainly affected by AA-amyloidosis are liver, spleen and the kidneys. Neuropathy and congestive heart failure are uncommon.

In this issue of MEMO, Dr. Dörler and Dr. Pölzl review the diagnosis and management of cardiac involvement [1]. Dr. Heininger highlights the renal aspects of amyloidosis [2], Dr. Graziadei focuses on the gastrointestinal complications of these diseases [3].

One of the first publications on amyloidosis came from Heidelberg in 1859, showing that amyloid is consisted of proteins instead of carbohydrates [4]. Dr. Kimmich and colleagues from the amyloidosis center in Heidelberg

summarise the treatment options for patients with AL-amyloidosis [5].

The availability of sensitive tests, e.g. free light chain assays and the implementation of novel drugs into the treatment of patients with AL-amyloidosis, have considerably changed our management. Diagnosis is now often made early in the course of the disease and with a modern management, which is in most cases an interdisciplinary undertaking; more patients can be offered treatment. These achievements in diagnosis and therapy have changed AL-amyloidosis from a uniformly fatal disease to one that can be effectively treated in many patients so that haematological remissions and more and more organ responses can be achieved. Although there is much room for improvement.

As a consequence of this progress in the diagnosis and management of AL-amyloidosis, physicians must be aware of the clinical symptoms of this rare disease.

Conflict of interest

The author declares that there is no conflict of interest.

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