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Severe dysautonomy revealing AL amyloidosis with multiple organ involvement

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Background

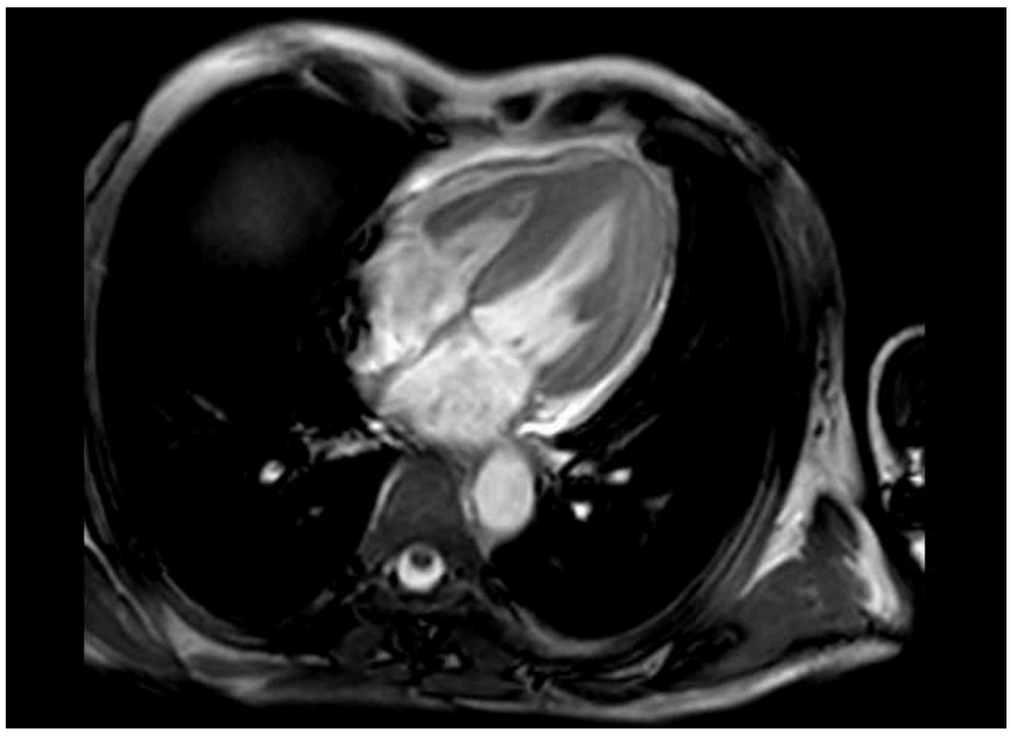
Amyloidosis constitutes a large spectrum of diseases characterized by the extracellular deposition of fibrils composed of several proteins. It can virtually affect any organ, and may lead to cell damage and organ function impairment, with potential life-threatening complications. We reported a case of severe autonomic neuropathy as the initial presentation of AL lambda amyloidosis associated to multiple myeloma (MM).

Case presentation

A 48-year old man was admitted after multiple episodes of syncope without prodromes. For several months, he complained of alternating diarrhea and constipation, pollakiuria and impotence, and then developed slight paresthesia of the upper and lower extremities, in addition to a severe weight loss related to anorexia. An initial work-up including electromyography, brain CT, lumbar puncture, PET-CT and coronarography failed to identify any abnormality, but the presence of a serum lambda M-protein raised the suspicion of amyloidosis. In addition to bowel and bladder dysfunction, he presented a dramatic orthostatic hypotension that kept him unable to stand or sit. Rectal biopsy confirmed the diagnosis of **lambda AL amyloidosis**. Further investigations revealed a salivary gland, kidney, gut and heart involvement and a SD IIA, ISS 2 MM. Therapy with bortezomib-cyclophosphamide-dexamethasone was interrupted after 2 cycles for grade 3 sensitive polyneuropathy, in the absence of hematological response, and replaced by a daratumumab-lenalidomide-dexmethasone combination. A partial hematological response was achieved after four cycles without any clinical benefit. Despite the use of high-dose midodrine, physiotherapy and stockings, the patient remained bedridden, and finally died of nosocomial infections.

Conclusion

We report a case of AL lambda amyloidosis with multiple organ involvement, including a severe autonomic neuropathy that was lately diagnosed and led to life-threatening complications, despite the use of new drugs. Amyloidosis remains largely underdiagnosed, early diagnosis is the cornerstone of late organ damage and better outcome.



Cardiac MRI showing left hypertrophic cardiomyopathy

AL amyloidosis :
Target organs
Heart 82%
Kidney 68%
Liver 14%
Soft tissues 17%
PNS 12%
ANS 10%

ANS, autonomous nervous system; PNS, peripheral nervous system – adapted from ASH Educ Program 2017