Lumbar spinal canal stenosis: An early sign of amyloid transthyretin related amyloidosis

Alessandro Graziani¹, Patrizia Cenni², Matteo Lisi³, Marco Domenicali⁴, Ludovico Graziani²

¹Department of Internal Medicine, Santa Maria Delle Croci, Ravenna, Italy
²Department of Radiology, Santa Maria Delle Croci, Ravenna, Italy
³Department of Cardiology, Santa Maria Delle Croci, Ravenna, Italy
⁴Department of Internal Medicine, University of Bologna, Bologna, Italy
⁵Department of Genetic Medicine, Bambino Gesù Children’s Hospital, Roma, Italy

Amyloid transthyretin-related amyloidosis (ATTR) onsets due to the extracellular multi-organ deposition of misfolded transthyretin, a serum protein that synthesizes mainly in the liver. Two different forms of the disorder have been identified to date, namely wild type ATTR (wtATTR), previously referred to as “senile” since it was mainly diagnosed in the elderly; and an inherited ATTR (hATTR), caused by mutant transthyretin.

ATTR amyloidosis is often overlooked or misdiagnosed owing to its non-specific presentation. Amyloid deposits can determine musculoskeletal manifestations, such as carpal tunnel syndrome (CTS), lumbar spinal canal stenosis (LSCS), or distal biceps tendon rupture (DBTR) several years before any cardiac manifestations, particularly in patients with wtATTR.

Cardiac manifestations of wtATTR (wtATTR-CA) include aortic stenosis, hypertrophic cardiomyopathy, heart failure with preserved ejection fraction, and hypertensive cardiomyopathy, although the cardiac signs and symptoms resemble those of other cardiovascular conditions of different etiology during the course of the disease [1].

We present radiological images of an 80-year-old man who had wtATTR-CA and LSCS. At the age of 65, he had had bilateral CTS. Ten years later, he began to report pain and loss of strength in the lower limbs mainly localized in the buttocks and quadriceps. Computed tomography (CT) of the spine showed a LSCS due to ligamentum flavum hypertrophy (LFH) considered to result from fibrous degeneration (Figure 1A). wtATTR-CA was diagnosed (Figure 1C) four years later. Genetic investigations yielded a negative result for hATTR. Upon further investigation, spinal magnetic resonance imaging (Figures 1D, E) and a second CT scan of the spine (Figure 1B) showed significant LFH with narrowing of the spinal canal.

The LF covers the rear surface of the spinal dural sac and has a protective effect on the spinal cord, controlling the extension of the intervertebral movement. In the elderly, LFH is one of the most frequent causes of LSCS, and its onset has been related to a degenerative fibrotic condition. Patients usually complain of intermittent claudication, low back pain and leg numbness, and pain [2]. Several authors recently reported amyloid deposits in elderly patients with LFH and LSCS. In a recent study, where 250 patients underwent surgery for LSCS due to LFH, Eldhagen et al. identified an amyloid presence in 88.4% of the histological samples, and ATTR was found in 37% of the cases [3]. In a large group of 324 patients who underwent surgery for LSCS, Godara et al. found wtATTR in 13% of the cases. wtATTR-CA was diagnosed in two of those cases. LF amyloid deposits could thus be considered an early manifestation of systemic ATTR disease [4]. Compared with CTS, LFH is a less known musculoskeletal wtATTR manifestation and is generally attributed to changes in fibrotic ligaments. Unfortunately, ⁹⁹mTc-diphosphonate scintigraphy, which is a useful diagnostic tool to detect wtATTR-CA, is not able to demonstrate LF amyloid deposits [5].

A correct interpretation of these manifestations, together with the cardiological
signs of the disease, allows for early diagnosis and faster access to therapy. For this reason, improved inter-specialty communication is required for managing patients with ATTR amyloidosis.

**Article information**

**Conflict of interest:** None declared.

**Open access:** This article is available in open access under Creative Common Attribution-Non-Commercial-No Derivatives 4.0 International (CC BY-NC-ND 4.0) license, allowing to download articles and share them with others as long as they credit the authors and the publisher, but without permission to change them in any way or use them commercially. For commercial use, please contact the journal office at kardiologiapolska@ptkardio.pl.

**REFERENCES**


