Multidisciplinary approach for the early detection of amyloid in patients who undergo carpal tunnel syndrome or lumbar stenosis surgery

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Background/Introduction: Symptomatic carpal tunnel syndrome (CTS) and lumbar stenosis (LS) seem to precede the cardiac manifestations of cardiac amyloidosis (CA) due to transthyretin amyloidosis (ATTR), so they could represent early markers of CA. CTS is more commonly present in CA due to wild-type ATTR (ATTRwt), although it can also be present in its hereditary variant (ATTRv) and in primary amyloidosis caused by immunoglobulin light chain deposition (AL).

Purpose: Evaluate the prevalence of amyloidosis in patients undergoing CTS or LS surgery, in an endemic area of the TTR mutation Val50Met (ATTRv).

Methods: 237 operated patients have been included (183 CTS, 54 LS) in whom an intraoperative biopsy was obtained (synovial tissue or flexor retinaculum in CTS; ligamentum flavum in LS) for histopathological analysis using Congo Red staining for amyloid detection and immunohistochemistry (IHC) or mass spectrometry (MS) for subtyping in amyloid A (AA), kappa, lambda and ATTR.

Also, a blood and urine test to rule out a monoclonal component and a cardiac scintigraphy (CS) with 99mTc-DPD to detect myocardial uptake were performed, which analysis was visual by Perugini scale and semiquantitative by heart-to-contralateral-lung ratio (H/CL). A cardiac SPECT/CT was performed in cases with a positive planar imaging, according to the ASNC/EANM Cardiac Amyloidosis Practice Points.

Results: Total of 204 biopsies were obtained (154 CTS, 50 LS), 16 of them were amyloid positive (8 CTS, 8 LS). All IHC were negative for AA, kappa and lambda. IHC for ATTR and MS analysis are still pending. 33 operated patients without biopsy (29 CTS, 4 LS).

203 laboratory tests and CS have been performed: 5/203 CS were positive (grade 2-3) in 4 patients who underwent CTS surgery (2 with positive biopsies for amyloid and 2 without biopsy) and 1 LS with also positive biopsy. In 15/203 laboratory tests, a monoclonal component was detected.

149/178 completed cases with negative biopsy, laboratory test and CS have completed the study (all tests negative).

Positivity for amyloid has been obtained in 18/237 cases (7.6%): 16 positive biopsies (2 with positive blood/urine test and negative CS, 3 with positive CS and negative blood/urine test and 11 with negative blood/urine test and CS) and 2 positive CS without biopsy and negative blood/urine test.

In the follow-up (2.41 years) of patients with positivity, 27.8% (5/18) manifests cardiomyopathy, 4 of them with positive CS, and 88.9% (16/18) has previously other orthopedic manifestations.

Conclusion: The estimated prevalence of amyloidosis in our series of surgically treated patients with CTS or LS is 7.6%