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Different Types of Amyloidosis Revealed with Positive Cardiac Scintigraphy with Tc-99M DPD-SPECT

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Abstract : Introduction: Transthyretin amyloidosis (ATTR) is a rare but serious infiltrative disease. Myocardial scintigraphy with DPD has emerged as the most effective, non-invasive, highly sensitive, and highly specific diagnostic method for cardiac ATTR amyloidosis. However, there are cases in which additional laboratory investigations reveal AL amyloidosis or other diseases despite a positive DPD scintigraphy. We describe the experience from the Onassis Cardiac Surgery Center and the monitoring center for infiltrative myocardial diseases of the cardiology clinic at AHEPA. Materials and Methods: All patients with clinical suspicion of cardiac or extracardiac amyloidosis undergo a myocardial scintigraphy scan with Tc-99m DPD. In this way, over 500 patients have been examined. Further diagnostic approach based on clinical and imaging findings includes laboratory investigation and invasive techniques (e.g., biopsy). Results: Out of 76 patients in total with positive myocardial scintigraphy Grade 2 or 3 according to the Perugini scale, 8 were proven to suffer from AL Amyloidosis during the investigation of paraproteinemia. Among these patients, 3 showed Grade 3 uptake, while the rest were graded as Grade 2, or 2 to 3. Additionally, one patient presented diffuse and unusual radiopharmaceutical uptake in soft tissues throughout the body without cardiac involvement. These findings raised suspicions, leading to the analysis of κ and λ light chains in the serum, as well as immunostaining of proteins in the serum and urine of these specific patients. The final diagnosis was AL amyloidosis. Conclusion: The value of DPD scintigraphy in the diagnosis of cardiac amyloidosis from transthyretin is undisputed. However, positive myocardial scintigraphy with DPD should not automatically lead to the diagnosis of ATTR amyloidosis. Laboratory differentiation between ATTR and AL amyloidosis is crucial, as both prognosis and therapeutic strategy are dramatically altered. Laboratory exclusion of paraproteinemia is a necessary and essential step in the diagnostic algorithm of ATTR amyloidosis for all positive myocardial scintigraphy with diphosphonate tracers since >20% of patients with Grade 3 and 2 uptake may conceal AL amyloidosis.

Keywords : AL amyloidosis, amyloidosis, ATTR, myocardial scintigraphy, Tc-99m DPD
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