Cardiac amyloidosis in patients undergoing aortic valve replacement for aortic stenosis

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Background: Transthyretin cardiac amyloidosis (ATTR) has been increasingly recognized in patients with degenerative aortic stenosis (AS). In some reports, the uptake of Tc-99m labeled bone radiotracers in cardiac amyloidosis has been documented. Tc-99m pyrophosphate (PYP) scintigraphy in the absence of evidence of a monoclonal gammopathy was diagnostic for transthyretin cardiac amyloidosis, providing a cost-effective and non-invasive technique with a specificity and positive predictive value of nearly 100%. We sought to determine the prevalence of ATTR as detected by the bone scan tracer among the patients with severe AS requiring surgical valve replacement.

Methods: We retrospectively analyzed clinical and echocardiographical data for 44 patients with severe AS requiring surgical valve replacement between Jan. 2009 and Dec. 2016. All eligible patients were offered Tc-99m PYP scintigraphy. Retention of Tc-99m PYP in the heart was assessed using both a semiquantitative visual score (range, 0 [no uptake] to 3 [uptake greater than bone]). Positive uptake was defined score 2 and 3.

Results: Myocardial deposition of Tc-99m PYP (Score 2–3) was identified in 4 of 44 patients (9%), all $>$70 years and 75% male. Patients with myocardial deposition of the tracer were older (78±8 years vs. 70±12 years), and had more mean interventricular septum thickness (18±3 mm vs. 14±5 mm). Both groups had at least ejection fraction and abnormal global longitudinal strain with no significant difference between groups. Pre-operative serum median NT-pro BNP level was similar between two groups, but post-operative improvement of NT-pro BNP was larger in non-deposition of the tracer group. During the post operative follow-up, survival was significantly worse if patients had amyloid deposition compared with no deposition subjects (25% vs. 7.5%).

Conclusion: Incidental transthyretin cardiac amyroidosis had a prevelance of 9% among patients undergoing surgical aortic valve replacement and was associated with a poor outcome.