A COMPARISON OF AORTIC DISSECTION IN THE PATIENTS WITH OR WITHOUT MARFAN SYNDROME AMONG KOREAN POPULATION

ACC Poster Contributions
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Background: Of cardiovascular manifestations in Marfan syndrome (MFS), aortic dissection (AD) is a substantial contributor to premature mortality among MFS patients. We hypothesized that patients with AD and MFS show different behaviors and prognosis from those with AD and non-MFS. We compared the clinical presentation, dissection characteristics and outcomes of patients with or without MFS among Korean population.

Methods: We reviewed patients with AD who were diagnosed from December 1994 to March 2009 at our institute. Clinical observation was conducted for 34 ± 35 months and all patients underwent multi-detector computed tomography during follow-up period. The following parameters to evaluate clinical outcomes were analyzed: significant progression of dissection, aneurysm formation, re-operation for recurrent dissection/aneurismal dilatation, and death.

Results: Of the 464 patients with AD, 53 patients (11.4%) had MFS. Compared with non-Marfan patients, patients with MFS more likely developed AD at younger age (33 ± 10 vs 57 ± 13 years, p < 0.0001), had prior cardiac surgery (13.2% vs 2.9%, p = 0.003) and less tended to be hypertensive (9.4% vs 74%, p < 0.0001). Aortic root dilatation and accompanying aortic valve regurgitation were more prominent in patients with MFS (p < 0.0001). Despite a similar incidence of type A AD between patient with and without MFS (66.0% vs 56.9%, p = 0.238), surgical repairs were more likely undergone in those with MFS (81.1% vs 65.1%, p = 0.020). During follow-up periods of our study, patients with MFS were more likely to develop of aortic dilatation and expansion of dissection (43.8% vs 21.8%, p = 0.003) and showed higher rate of re-operation (32% vs 8.7%, p < 0.0001). Overall outcomes in patients with MFS were far worse than those without MFS (45.8% vs 24.5%, p = 0.0062).

Conclusions: Our study demonstrated different characteristics and poorer outcome of aortic dissection in patients with Marfan syndrome compared with non-Marfan patients. Based on these finding, we emphasize the importance of accurate diagnosis and surveillance of aortic dissection as well as timely management including prophylaxis in patients with Marfan syndrome.