Coronary Artery Involvement by Chronic Graft-Versus-Host Disease Presenting as Sudden Cardiac Arrest

Ko J.K., You J.J., Im H.J.
Department of Pediatrics, Ulsan University College of Medicine, Seoul, Korea

Introduction: We present a case with chronic graft-versus-host disease (GVHD) who experienced sudden cardiac arrest 21/2 years after allogeneic peripheral blood stem cell transplantation (allo-PBSCT). Sudden cardiac arrest was resulted from myocardial ischemia caused by coronary obstruction.

Case: A 44 month old boy was admitted to the intensive care unit due to sudden collapse at the emergency department (ED) visiting with fever. He was diagnosed at 6 months with chronic granulomatous disease by CYBB gene mutation. Since 3 month of age, he has suffered from multiple abscesses at body and internal organ such as liver and kidney. Allo-PBSCT using HLA matched unrelated donor was performed at 13 months. His post-transplantation course was complicated by CMV infection and acute gut and cutaneous GVHD. His chronic GVHD manifested as elevated hepatic enzymes and sclerosis of skin with multiple joint contractures. For a week before visiting ER, he had shown upper respiratory infection-like symptoms with poor oral intake. At ED, he suddenly collapsed and was successfully resuscitated from ventricular fibrillation. On admission, echocardiography showed no abnormality, however, ECG showed abnormal q-wave and ST changes in the inferior leads, suggesting coronary event. Cardiac magnetic resonance imaging revealed myocardial thinning and hypokinesia of left ventricle with a weak contrast uptake consistent with subendocardial infarct. Coronary artery imaging with multidetector computed tomography showed obliterative narrowing of left anterior descending artery (LAD) and luminal irregularity at the right coronary artery (RCA) and left circumflex artery (LCX). Coronary angiography revealed complete obstruction of proximal LAD, subtotal obstruction of mid LCX and mild narrowing at the RCA. Distal LAD was filled by the small collateral arteries from RCA and LCX. His poor general condition didn’t permit further intervention and after 6 months of follow-up, he died suddenly at home.

Conclusions: The coronary artery disease is only rarely occurring cardiac event in children and cardiac complications associated with GVHD are uncommon. Coronary artery involvement, albeit rare, should be recognized as one of the important manifestation of chronic GVHD in children.