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Cardiovasc Pharm Open Access 2018, Volume 7
DOI: 10.4172/2329-6607-C1-006

15th International Conference on

Pediatrics and Pediatric Cardiology

February 19-20, 2018 | Paris, France

Long-term outcome of Kawasaki disease complicated by a large coronary aneurysm

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Kawasaki disease (KD) is an acute febrile disease of unknown etiology. It causes systemic inflammation of vessels through the whole body and especially affects coronary arteries in children younger than 5 years of age. The disease leads to coronary artery aneurysms in ≈25% of untreated cases. Treatment within 10 days of illness onset with high dose intravenous immunoglobulin (IVIG) and aspirin has proven effective in dramatically reducing the risk of cardiac complications. However, approximately 20% of patients who require a second dose of IVIG for KD do not adequately respond to this treatment and require additional medications such as corticosteroid or infliximab. KD was first identified in Japan by Tomisaku Kawasaki in 1967, and has now been described worldwide. In Korea, the annual incidence of KD was 194.7 per 100,000 children in 2014, and the coronary aneurysm rate was 1.7%. The natural history of coronary arterial inflammation in KD was demonstrated to occur through 3 mechanisms as reported previous studies: (1) No coronary artery changes, (2) Mild, transient dilatation that resolves within 4-6 weeks, (3) Necrotizing arteritis that forms an aneurysm. Coronary arterial aneurysm can progress to subacute/chronic vasculitis, luminal myofibroblastic proliferation, and laminar non-occlusive thrombosis. Its phenotypes are myocardial infarction, possible progression to a normal luminal dimension, or further progression to complex stenosis with calcification. In 1% of cases with such damage, diffuse dilatation of the coronary arteries can progress to a giant aneurysm. The progression to giant aneurysm increases the risks of acute thrombosis and long-term myocardial ischemia. The severity of coronary artery luminal lesions determines the risk range for long-term management. Such risk assessment depends on the patient's maximal Z score in any branch by echocardiography or coronary angiography/angio-computed tomography. According to recent published guidelines, large/giant aneurysms are defined as Z score ≥10 or absolute inner diameter ≥8 mm, and medium aneurysms are those with a Z score ≥5 to <10 or an absolute inner diameter <8 mm. The treadmill exercise test has been known as the least sensitive method to detect myocardial ischemia in patients with KD. Stress myocardial scintigraphy has been shown to be useful for risk stratification, but we should consider its radiologic hazard. Korean nationwide data collected between 1990 and 2011 reported, and showed 239 patients with coronary aneurysms of diameter >6 mm. Severe stenosis or occlusion of the coronary artery were analyzed, and percutaneous transluminal coronary balloon angioplasty was performed in 10 patients, stent placement in 9 patients, and percutaneous transluminal coronary rotational ablation in 3 patients. Thirteen patients presented with suggestive myocardial infarction, 14 patients underwent coronary artery bypass graft surgery, and 5 patients died during the follow-up period. For KD patients with giant coronary aneurysm, careful planning of follow-up is mandatory for long-term manage, and aggressive treatments with transcatheter or surgical intervention are needed.

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