Editorial

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Long-Term Outcome of Kawasaki Disease Complicated by a Large Coronary Aneurysm

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Kawasaki disease was first identified in Japan by Tomisaku Kawasaki in 1967,[1] and has now been described worldwide. In Korea, the annual incidence of KD was 194.7 per 100000 children in 2014, and the coronary aneurysm rate was 1.7%.[2]

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.[3] 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.[4][5]

The natural history of coronary arterial inflammation in KD was demonstrated to occur through 3 mechanisms (Fig. 1):[3][6] 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.[3][6] 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.[7]

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. 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.[3][5] The treadmill exercise test is the least sensitive method to detect myocardial ischemia in patients with KD.[8] Stress myocardial scintigraphy has been shown to be useful for risk stratification.[8]

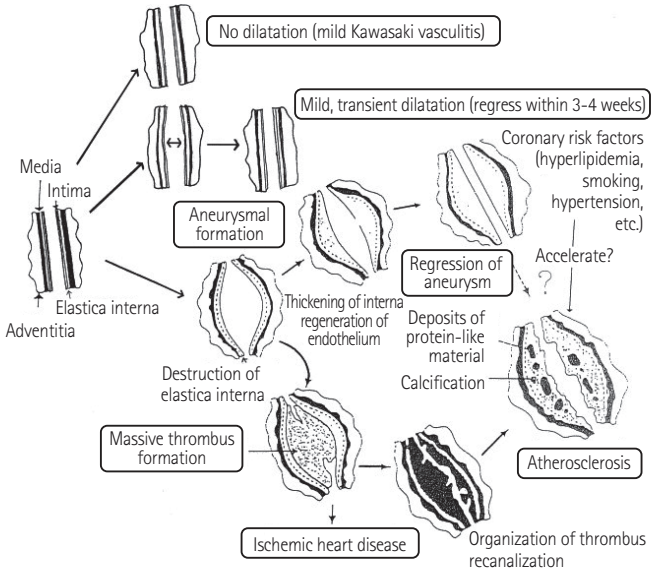

Fig. 1. The natural history of coronary arterial inflammation in Kawasaki disease.[6] Printed with permission from Elsevier.

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In data of 76 Japanese patients with giant aneurysm of diameter ≥8.0 mm from 1972 to 2011,[8] 46 (61%) underwent catheter and surgical coronary interventions, and 12 showed acute myocardial infarction. Of all patients, 7 died and 1 underwent a heart transplantation, resulting in 95%, 88%, and 88% survival rates at 10, 20, and 30 years after the onset of KD, respectively. The giant aneurysm was located in the right coronary artery only in 17 patients, left coronary artery only in 13 patients, and both coronary arteries in 46 patients. Although one patient with chest discomfort showed 99% stenosis of the left anterior descending artery in cardiac catheterization, his treadmill exercise stress test did not show ischemic changes on electrocardiogram.

Korean nationwide data collected between 1990 and 2011 was reviewed by Jang et al.[9] 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.

A Korean study of long-term prognosis for 71 patients with KD complicated by a coronary aneurysm from 1986 to 2013 was compared between coronary aneurysms sized 6-8 mm and those ≥8 mm. Myocardial infarction only occurred in 7 patients with coronary aneurysm ≥8 mm in diameter, and the median elapsed time from KD onset to myocardial infarction was 4.0 months (range: 0.5-31.2 months). Despite the loss of one patient, there was no statistical difference in the cumulative coronary intervention rate for the two groups of patients with aneurysm.[10]

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.

References

1. Kawasaki T. Acute febrile mucocutaneous syndrome with lymphoid involvement with specific desquamation of the fingers and toes in children. *Arerugi* 1967;16:178–222.
2. Kim GB, Park SH, Eun LY, et al. Epidemiology and clinical features of Kawasaki disease in South Korea, 2012–2014. *Pediatr Infect Dis J* 2017;36:482–5.
3. McCrindle BW, Rowley AH, Newburger JW, et al. Diagnosis, treatment, and long-term management of Kawasaki disease: a scientific statement for health professionals from the American Heart Association. *Circulation* 2017;135:e927-99.
4. Abrams JY, Belay ED, Uehara R, Maddox RA, Schonberger LB, Nakamura Y. Cardiac complications, earlier treatment, and initial disease severity in Kawasaki disease. *J Pediatr* 2017;pii: S0022-3476(17):30655-8. [Epub ahead of print] doi: 10.1016/j.jpeds.2017.05.034
5. Lim YJ, Jung JW. Clinical outcomes of initial dexamethasone treatment combined with a single high dose of intravenous immunoglobulin for primary treatment of Kawasaki disease. *Yonsei Med J* 2014;55:1260-6.
6. Kato H. Cardiovascular complications in Kawasaki disease: coronary artery lumen and long-term consequences. *Prog Pediatr Cardiol.* 2004;19:137-45.
7. Golshevsky, D, Cheung, M, Burgner, D. Kawasaki disease: the importance of prompt recognition and early referral. *Aust Fam Physician* 2013;42:473-6.
8. Suda K, Iemura M, Nishiono H, et al. Long-term prognosis of patients with Kawasaki disease complicated by giant coronary aneurysms: a single-institution experience. *Circulation* 2011;123:1836-42.
9. Jang GY, Kang IS, Choi JY, et al. Nationwide survey of coronary aneurysms with diameter >6 mm in Kawasaki disease in Korea. *Pediatrics Int* 2015;57:367-72.
10. Bang JS, Kim GB, Kwon BS, et al. Long-term prognosis for patients with Kawasaki disease complicated by a large coronary aneurysm (diameter ≥6 mm). *Korean Circ J* 2017;47:516-22.

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