Case Report

Segmented Coronary Artery Aneurysms and Kawasaki Disease

Mohammad Yoosef Aarabi Moghadam, MD, Hojat Mortazaeian, MD, Mehdi Ghaderian, MD*, Hamid Reza Ghaemi, MD

Shaheed Rajaei Cardiovascular, Medical and Research Center, Tehran University of Medical Sciences, Tehran, Iran

Received 15 July 2010; Accepted 23 February 2011

Abstract

Kawasaki disease (KD) is an acute vasculitis syndrome of unknown etiology. It occurs in infants and young children, affecting mainly small and medium-sized arteries, particularly the coronary arteries. Generalized microvasculitis occurs in the first 10 days, and the inflammation persists in the walls of medium and small arteries, especially the coronary arteries, and changes to coronary artery aneurysms.

We report the case of a 10-month-old girl referred to our center three months after the onset of disease due to the aneurysms of the coronary arteries. During the acute phase of her illness, she received 2 gr/kg intravenous gamma globulin; and after her referral to us, the patient was treated by antiaggregant doses of acetylsalicylic acid (ASA) (5 mg/kg) and Warfarin (1 mg/daily). At three months' follow-up, the aneurysms still persisted in the echocardiogram.

J Teh Univ Heart Ctr 2011;6(2):89-91

This paper should be cited as: Aarabi Moghadam MY, Mortazaeian H, Ghaderian M, Ghaemi HR. Segmented Coronary Artery Aneurysms and Kawasaki Disease. J Teh Univ Heart Ctr 2011;6(2):89-91.

Keywords: *Mucocutaneous lymph node syndrome • Coronary aneurysm • Child*

Introduction

Kawasaki disease (KD), first described in Japan in 1967 by Kawasaki, is an acute vasculitis syndrome of unknown etiology occurring in infants and young children and affecting mainly small and medium-sized arteries, particularly the coronary arteries. Eighty percent of the patients are younger than 5 years of age and boys are more susceptible than girls with a ratio of approximately 1.5:1.[1] The disease occurs most frequently in winter and spring. Young infants have the highest rate of coronary artery aneurysm formation and often exhibit incomplete clinical presentations.[1] Giant coronary artery aneurysms (internal diameters > 8mm) are seen in 0.5% to 1% of adequately treated patients.[2, 3] Incomplete cases are most frequent in infants, who unfortunately also have the highest likelihood of developing coronary disease.[4]

Case Report

A 10-month-old girl referred to our center three months after the onset of acute disease (her parents described an acute phase with a fever of 10 days' duration). During the acute phase of her illness, she had a polymorphous exanthema

*Corresponding Author: ***Mehdi Ghaderian,*** *Assistant Professor of Pediatric Cardiology, Department of Pediatric Cardiology, Jundishapor University of Medical Sciences, Golestan Medical, Educational and Research center, Ahwaz, Iran. 6135733118. Tel: +98 61 13743063. Fax: +98 61 13743063. E-mail: ghader_45@yahoo.com.*

with perianal desquamation, a bilateral bulbar conjunctival injection, and changes in lips and oral cavity mucosa. She received 2 gr/kg intravenous gamma globulin (IVIG); and once more, the IVIG infusion caused a persistent fever. In follow-up by echocardiography, she had aneurysms of the coronary arteries and was therefore referred to our center. Echocardiographic examination demonstrated multiple aneurysms and diffuse dilatation of the coronary arteries (Figure 1).

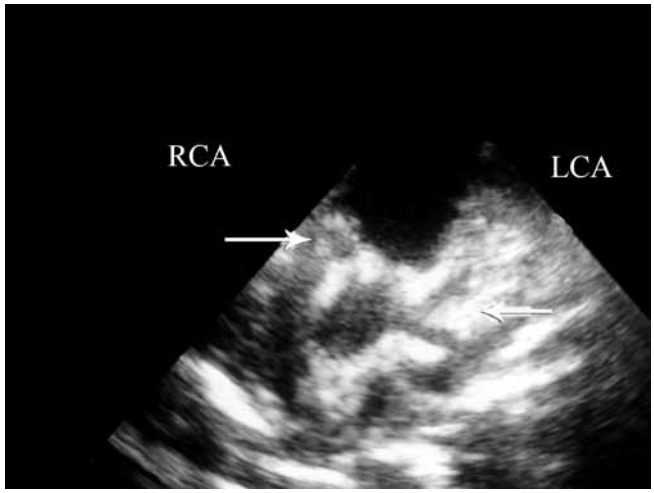

Figure1. Short-axis parasternal echocardiography demonstrating diffuse dilatation of the coronary arteries (arrows)
LCA, Left coronary artery; RCA, Right coronary artery

No impairment of the left ventricular function was found. The standard twelve-lead electrocardiogram showed no ischemia or dysrhythmias. The coronary angiograms showed diffuse segmented aneurysms (resembling beads on a string) of the right coronary artery and left anterior descending artery from the ostium to the distal segment (Figures 2 & 3).

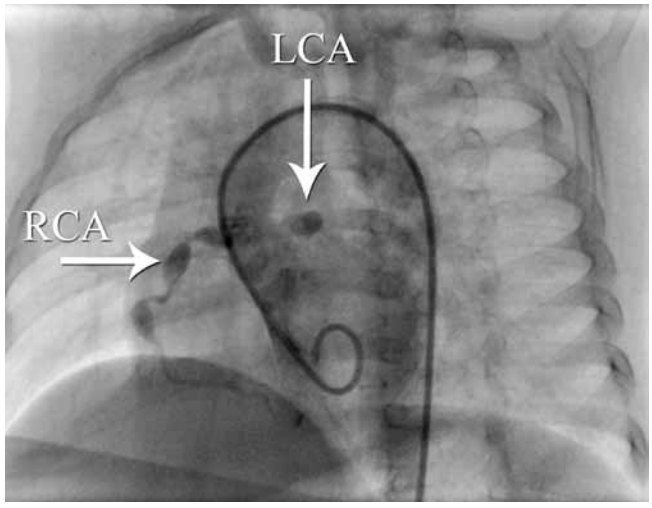

Figure 2. Segmented aneurysms of the left coronary artery (LCA) and right coronary artery (RCA) in angiogram (arrows)

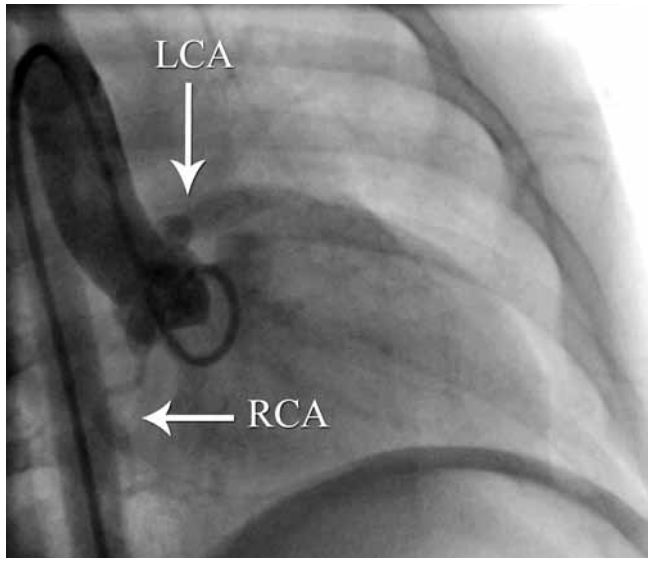

Figure 3. Segmented aneurysms in the right coronary artrey (RCA) and dilatation in the left coronary artery (LCA) in angiogram (arrows)

The patient was treated by antiaggregant doses of ASA (5 mg/kg) and Warfarin (1 mg/daily). At this point, she was followed up via echocardiography. At three months' follow-up, the aneurysms still persisted in the echocardiogram without any new complications such as thrombosis, ventricular dysfunction, and myocardial infarction.

Discussion

KD is an acute febrile vasculitis of unknown etiology, although an infectious or toxic triggering agent is suspected by some authors. Its incidence presents regional variations that oscillate between 108 in 100,000 children younger than 5 years of age in Japan and 10 in 100,000 children younger than 5 years old in the United States.[5] The acute phase of KD usually starts with upper respiratory or gastrointestinal prodromal symptoms. It is followed by an abrupt onset of high fever, accompanied by skin rash, conjunctival injection, reddening and fissuring of lips, erythema of the buccal mucosa, strawberry tongue, nonsuppurative cervical lymphadenitis, and erythema and edema of the hands and feet.[1] ASA and IVIG should be started within 10 days from fever onset in order to prevent coronary involvement.

Overall, 2%-6% of children with KD treated with IVIG develop coronary artery disease; it tends to increase to 20%-30% in patients who do not receive IVIG. Giant coronary artery aneurysms are seen in 0.5% to 1% of adequately treated patients. Unfortunately, if the diagnosis is not established and treatment is not instituted, some patients may suffer sudden death secondary to myocardial infarction or coronary aneurysm rupture, or may develop serious asymptomatic coronary disease that is unrecognized until

the symptoms of myocardial ischemia develop later in life.[4] The prevalence of coronary aneurysms is higher in infants less than 12 months old. Shulman et al.[6] reported that about 50% of coronary abnormalities regress within 5 years. At three months' follow-up via echocardiography, our patient's aneurysms were still persistent in size.

References

1. Takahashi M, Newburger JW. Kawasaki disease (mucocutaneous lymph node syndrome). In: Allen HD, Driscoll DJ, Shaddy RE, Feltes TF, eds. Moss and Adam's Heart Disease in Infants, Children and Adolescents: Including the Fetus and Young Adults. 7th ed. New York: Lippincott Williams & Wilkins; 2008. p. 1243-1256.
2. Moura C, Costa P, Silva JC. Giant coronary aneurysms in Kawasaki disease. Pediatr Cardiol 2008;29:230-231.
3. Aarabi Moghadam MY, Mearji M, Sayadpoor K. Study of cardiac involvement prevalence in 61 pediatric case of Kawasaki disease. IJMS 2004;11-extra.1:361-366.
4. Moreno N, Méndez-Echevarría A, de Inocencio J, Del Castillo F, Baquero-Artigao F, García-Miguel MJ, de José MI, Aracil J. Coronary involvement in infants with Kawasaki disease treated with intravenous gamma-globulin. Pediatr Cardiol 2008;29:31-35.
5. Shulman ST, McAuley JB, Pachman LM, Miller ML, Ruschhaupt DG. Risk of coronary abnormalities due to Kawasaki disease in urban area with small Asian population. Am J Dis Child 1987;141:420-425.