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Case Report

A 14.5-Year-Old Boy with Coronary Obstruction after Kawasaki Disease: A Case Report

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Abstract

Kawasaki disease (KD) is a febrile vasculitis and is considered a leading cause of acquired coronary artery disease in children. A clinically critical complication is the coronary artery aneurysm, which may progress and lead to coronary stenosis or even obstruction. Herein, we describe a 14.5-year-old boy with a history of KD at 6 months old, who developed multiple aneurysms along all the coronary branches. During the follow-up at the age of 14 years, the left coronary artery aneurysms regressed, while the aneurysm of the right coronary artery persisted and was complicated by obstruction at its proximal part, according to computed tomography angiography. However, the patient at the last follow-up was asymptomatic and well.

The serious nature of KD coronary complications warrants follow-up visits. Since echocardiography alone may fail to reveal stenosis or obstruction, other adjunct follow-up imaging modalities such as conventional, computed tomography, and magnetic resonance angiography should be performed in patients with coronary aneurysms.

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Keywords: *Mucocutaneous lymph node syndrome; Coronary aneurysm; Coronary stenosis*

Introduction

Kawasaki disease (KD), a febrile vasculitis, was first described in 1967 by a Japanese pediatrician.[1,2] KD is a leading cause of acquired coronary artery disease in children.[3] Typical forms occur in children under 5 years old.[4] The most important complication of KD is coronary involvement, the most serious of which is the coronary artery aneurysm (CAA), with a prevalence rate of nearly 25%.[1,4] Coronary involvement may change with time: it may regress or progress toward stenosis or obstruction. Stenosis usually occurs more gradually than regression.[1] Medications like intravenous immunoglobulin can decrease the risk of CAAs.[5]

Echocardiography can assess the proximal parts of the coronaries. More evaluation can be done by angiography, which is the gold standard of diagnosis.[6] Computed tomography (CT) angiography is a suitable substitute for angiography because not only is it noninvasive and associated with fewer complications but also it confers accurate evaluation.[6]

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Case Report

A 14.5-year-old boy was admitted to the hospital at 6 months old due to prolonged fever, skin rash, lymphadenopathy, and a KD diagnosis. On physical examination, he had maculopapular rashes, normal heart sounds without significant murmurs, and distal edema. Echocardiography revealed multiple coronary aneurysms (the right coronary artery [RCA] =10 mm {giant}, the left anterior descending artery [LAD] =6 mm {large}, and the left circumflex artery [LCX] =5.6 mm {large}). Warfarin and aspirin were prescribed because of the giant coronary aneurysm, and single-dose immunoglobulin (2 g/kg) was infused intravenously. After the resolution of his symptoms, the patient was discharged and followed up routinely through serial visits and echocardiography, performed every 3 months initially for the first year and then every 6 months for 10 years and yearly afterward until 14.5 years old. Twelve months after the diagnosis, echocardiography showed the complete regression of the LAD and LCX aneurysms, whereas the aneurysm in the RCA had decreased in size. First-time angiography was done about 2 years after the diagnosis, and it showed multiple RCA aneurysms (the biggest one 5×10 mm in size) and 20% narrowing between 2 aneurysms. During the follow-up, the patient was symptom-free and had a normal electrocardiogram (ECG). After about 9 years of follow-up, given the decreased size of the RCA aneurysms, warfarin was discontinued, and dipyridamole was added to ASA.

Ten years after the diagnosis, angiography revealed a small, single RCA aneurysm with some narrowing. Four years later, CT angiography showed intimal thickening at the origin and the proximal part of the RCA, diffuse significant calcified plaques (the coronary calcium score =115 [near occlusion]) at the proximal third of the RCA, adequate collaterals from the left-sided coronary artery branches (Figure 1) without spotty calcification or the napkin-ring sign, and a normal left ventricular ejection fraction.

Since then, the patient has been on regular follow-ups. He was asymptomatic at his last follow-up visit with normal ECG (due to coronary collaterals) (Figure 2) and ventricular function, and he was on Plavix, ASA, and propranolol.

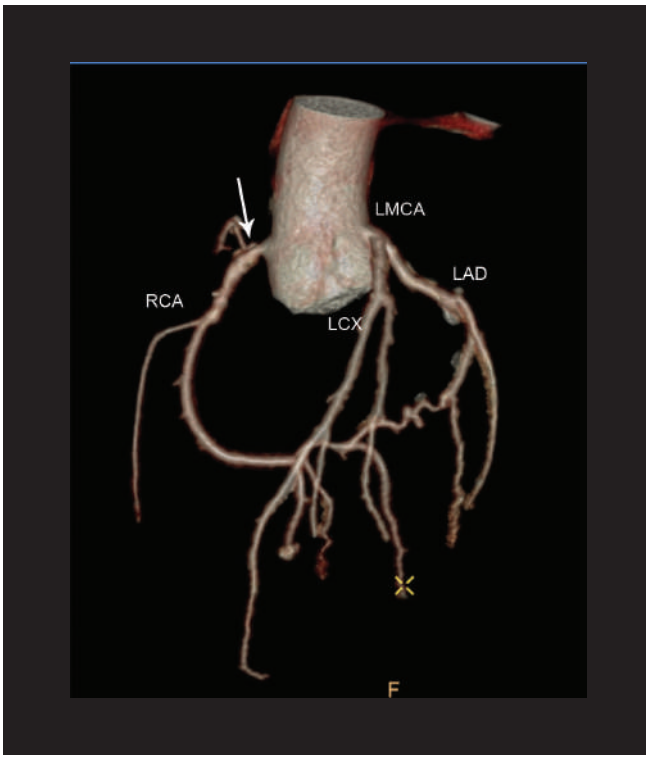
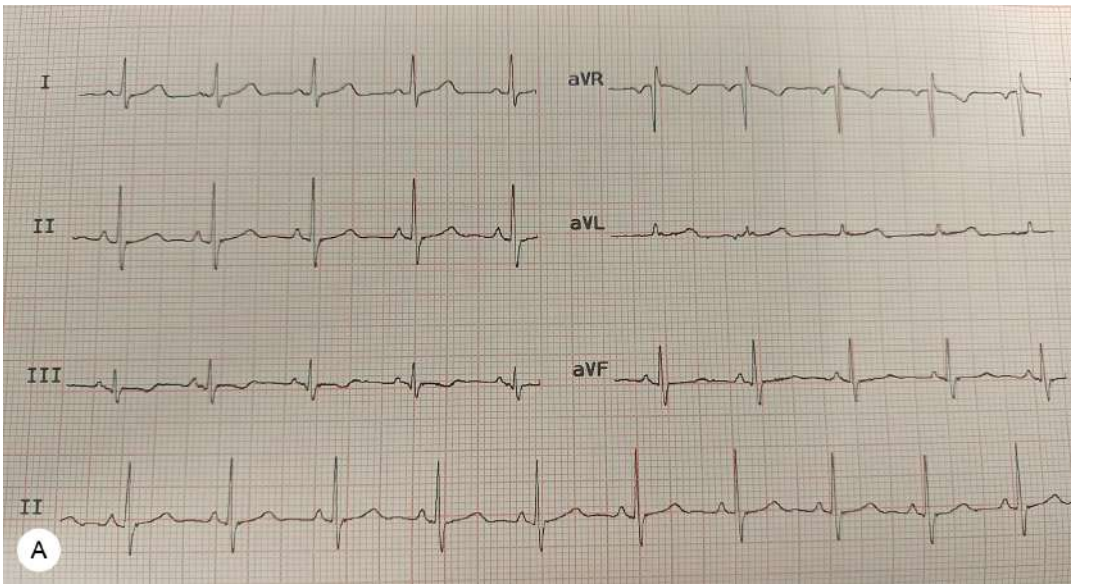


Figure 1. Coronary computed tomography angiography shows intimal thickening at the origin and the proximal part and significant calcified plaques (near occlusion) at the proximal third of the RCA (arrow).
RCA, Right coronary artery; LMCA, Left main coronary artery; LAD, Left anterior descending artery; LCX, Left circumflex artery



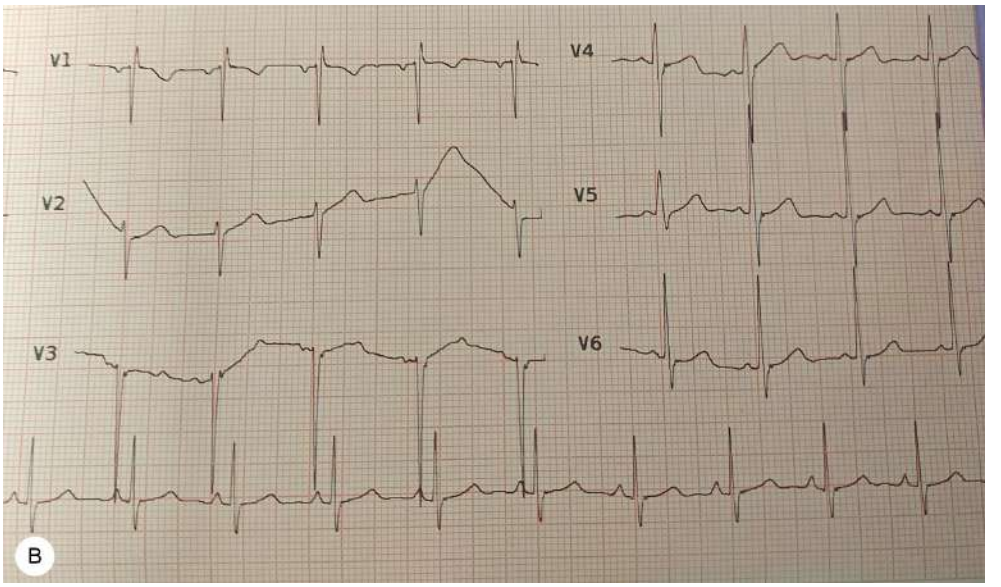


Figure 2. The last electrocardiogram of the patient shows a normal sinus rhythm and a normal axis without ischemic changes.

Discussion

The case we present herein focuses on the complications of KD, the most significant of which are coronary complications. Among these complications, CAA and stenosis (as a late one) are the causes of mortality and morbidity in KD. First described in 1967 in Japan,[1, 2] KD happens in young children (<5 y/o) with prolonged fever and mucocutaneous changes.[3] It is one of the notable causes of acquired coronary artery disease in children, with a male-to-female ratio of about 1.5/1. One of the most serious complications of KD is the CAA, which is more prevalent in young infants. Coronary artery dilatation and perivascular brightness appear between 30% and 50% of patients with KD in the acute phase. The late consequences of KD manifest themselves in patients with coronary involvement at presentation.[4] Medications like intravenous immunoglobulin can decrease the risk of CAAs from 25% to between 3% and 5% in treated patients.[5]

Patients with a history of coronary involvement and aneurysms may be at risk of myocardial infarction, sudden death, or heart failure due to stenosis or thrombosis.[3] Calcification or thrombosis in aneurysms can be misleading, resulting in the underestimation of the aneurysm by echocardiography.[7] In their investigation, van Stijn et al[8] concluded that the accuracy of CT angiography in detecting CAAs was about twice that of echocardiography and reported that aneurysms that might be missed by echocardiography were usually located in the LAD, the LCX, and the distal segment of the RCA.

Kato et al[3] evaluated 594 Japanese patients for KD complications 10 to 12 years after the onset of the disease and found that coronary artery stenosis, heart failure, and infarction constituted the late complications. Some other studies have also shown the occurrence of stenosis, occlusion, or calcification at the site of previous aneurysms whether or not the patient is symptomatic.[3]

A study conducted in 2005 concluded that patients with moderate-to-large aneurysms (>6 mm) had a greater risk of not only irreversible changes in the coronary arterial wall but also stenosis.[9]

As was discussed above, the risk of coronary stenosis, as a late complication, is higher at the site of previous aneurysms, and CT angiography is more accurate for evaluating such a complication. We reached a similar conclusion in our case.

Large or giant aneurysms warrant more evaluation by angiography about 6 to 12 months after the diagnosis, with further follow-up determined according to the results of the evaluation and clinical presentations of the patient.[3] Some studies have recommended more evaluation by CT angiography about 2 to 3 months after the diagnosis.[8] Kim JW et al[10] claimed that the accuracy of CT angiography in the assessment of the coronary arteries was more than that of magnetic resonance (MR) angiography in patients with KD. Furthermore, imaging modalities such as CT and MR angiography should be employed in the follow-up of KD patients with CAAs, and different medications or interventions might be needed based on the evaluations. In our case report, we followed up the patient every 6 months on medication.

Conclusion

According to our case and others, a long follow-up in patients with Kawasaki disease is strongly recommended, and echocardiography is not the ideal diagnostic tool for evaluating stenosis or obstruction in the coronary arteries. It is advisable to use different modalities like ECG, angiography, CT angiography, and MR angiography to evaluate patients with Kawasaki disease, especially those with cardiac symptoms,

ischemic changes in ECG, abnormal ejection fractions, or new-onset mitral regurgitation. These modalities can also be used to discontinue anticoagulants or antiplatelet treatment in patients whose coronary artery aneurysms show regression at follow-ups.

References

1. Senzaki H. Long-term outcome of Kawasaki disease. Circulation 2008;118:2763-2772.
2. Gordon JB, Kahn AM, Burns JC. When children with Kawasaki disease grow up: myocardial and vascular complications in adulthood. J Am Coll Cardiol 2009;54:1911-1920.
3. De Ferranti SC. Burns JC. Newburger JW. Kawasaki disease (Mucocutaneous Lymph Node Syndrome). In: Allen HD, Shaddy RE, Penny DJ, Feltes TF, Cetta F, eds. Moss and Adams' Heart Disease in Infants, Children, and Adolescents: Including the Fetus and Young Adult. 9th ed. Philadelphia/Baltimore/New York/London/Buenos Aires/Hong Kong/Sydney/Tokyo: Wolters Kluwer; 2016. p. 1355-1371.
4. Gordon JB, Burns JC. Management of sequelae of Kawasaki disease in adults. Glob Cardiol Sci Pract 2017;2017:e201731.
5. Yan F, Zhang H, Xiong R, Cheng X, Chen Y, Zhang F. Effect of Early Intravenous Immunoglobulin Therapy in Kawasaki Disease: A Systematic Review and Meta-Analysis. Front Pediatr 2020;8:593435.
6. Ghareep AN, Alkuwari M, Willington F, Szmigielski W. Kawasaki disease: diagnosis and follow-up by CT coronary angiography with the use of 128-slice dual source dual energy scanner. A case report. Pol J Radiol 2015;80:526-28.
7. Tsuda E, Kamiya T, Ono Y, Kimura K, Kurosaki K, Echigo S. Incidence of stenotic lesions predicted by acute phase changes in coronary arterial diameter during Kawasaki disease. Pediatr Cardiol 2005;26:73-79.
8. van Stijn D, Planken RN, Groenink M, Streekstra GJ, Kuijpers TW, Kuipers IM. Coronary artery assessment in Kawasaki disease with dual-source CT angiography to uncover vascular pathology. Eur Radiol 2020;30:432-441.
9. Mandal S, Pande A, Mandal D, Sarkar A, Kahali D, Panja M. Various coronary artery complications of Kawasaki disease: series of 5 cases and review of literature. J Cardiovasc Dis Res 2012;3:231-235.
10. Kim JW, Goo HW. Coronary artery abnormalities in Kawasaki disease: comparison between CT and MR coronary angiography. Acta Radiol 2013;54:156-163.