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Case Report

Late onset Kawasaki disease with multiple coronary arterial aneurysms: A case report ☆

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ABSTRACT

Kawasaki disease is an acute, self-limiting, systemic vasculitis of small and medium-sized arteries. It predominantly occurs in children under 4 years of age, though rarely older children can also be affected. This disease is the leading cause of acquired heart disease in children, with coronary aneurysms being a hallmark finding. The risk of coronary complications necessitates regular monitoring and possible preventative treatment with thromboprophylaxis. Here we discuss a rare case of a 10-year-old boy who exhibited typical symptoms of Kawasaki disease and was found to have multiple coronary artery aneurysms through diagnostic imaging.

Introduction

Kawasaki disease (KD), also referred to as mucocutaneous lymph node syndrome, is a form of childhood vasculitis that primarily affects small and medium-sized muscular arteries, including the coronary arteries. It is a self-limiting febrile condition of unknown etiology, though genetic and environmental factors may play a role. KD has incidence ranging between 5 and 22 cases per 100,000 children less than 5 years old [1]. KD predominantly affects young children; about 80% of cases are seen in children under 4 years old, with the highest frequency around 9 to 11 months [2]. Instances of KD are rare in children younger than 3 months or older than 8 years [3]. KD is regarded as the leading cause of acquired heart disease in children, with coronary artery aneurysms being the hallmark finding. If untreated, the disease can lead to coronary complications in up to 25% of cases, resulting in a mortality rate of approximately 2% [3].

Classic (typical) KD is diagnosed based on the presence of a fever lasting 5 or more days, accompanied by 4 out of the following findings: bilateral conjunctival injection, oral changes like cracked and erythematous lips and strawberry tongue, cervical lymphadenopathy, extremity changes such as erythema or desquamation of palm and sole, and a polymorphous rash [4]. Incomplete (atypical) KD is diagnosed when

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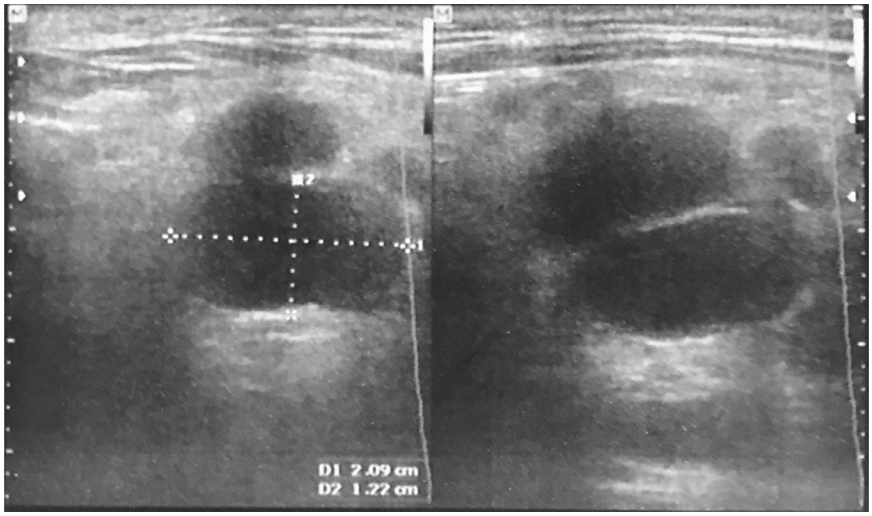

Fig. 1 – Ultrasonogram of neck revealed few enlarged left cervical lymph nodes.

a patient has fever lasting 5 or more days and 2 or 3 of these clinical signs [4].

Here we discuss a rare case of a 10-year-old boy who exhibited clinical manifestation of classic KD and was found to have coronary artery aneurysms, involving both right and left coronary arteries, through diagnostic imaging.

Case presentation

A 10-year-old boy presented with a high-grade fever that reached up to 103.6 degrees Fahrenheit, accompanied by a polymorphous rash and swelling of both hands and feet, persisting for several days. He exhibited daily spikes in temperature but did not report shortness of breath, cough, chest pain, diarrhea, or vomiting. Upon clinical examination, he displayed a macular and papular rash primarily on his trunk and proximal extremities, pustules on his palms and soles, swelling in the hands and feet, a strawberry tongue, swollen and cracked lips, and nonexudative conjunctival injection. Additionally, few palpable enlarged lymph nodes were noted on the left side of his neck.

Laboratory investigations revealed an increased erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP), as well as leukocytosis with a predominance of neutrophils. In light of the clinical findings, a diagnosis of KD was made. Ultrasound of the neck revealed few enlarged lymphnodes in the left cervical chain (Fig. 1). Transthoracic Echocardiography (TTE) revealed a dilated mid right coronary artery (RCA) of 4.1 mm (z-score +3.4) and mid left anterior descending artery of 3.9 mm (z-score +3.2), suggestive of coronary aneurysms.

The patient started receiving treatment with a single infusion of 2 g/kg of intravenous immunoglobulin (IVIG) and oral aspirin at a dose of 60 mg/kg/day until the fever resolved. There was noticeable improvement in the patient's overall condition along with remission of fever. Echocardiograms were performed daily for the first week and then on a weekly basis.

A computed tomography (CT) coronary angiogram (Figs. 2A-C) revealed medium and small-sized aneurysms of the mid-right coronary artery, with a maximum diameter of 5.6 mm, and mid-left anterior descending artery, with a maximum diameter of 4.3 mm. He was discharged from the hospital on dual antiplatelet therapy and kept under regular follow-up.

Discussion

KD is diagnosed based on clinical criteria [5]. There is no specific test for it, but certain lab results—such as elevated ESR, elevated CRP, hyponatremia, hypoalbuminemia, and and TTE findings of coronary aneurysms—can support the diagnosis and help distinguish it from other illnesses. The most common conditions confused with KD are scarlet fever and measles [6]. These can be differentiated by the characteristics and distribution of the rash: KD features a polymorphous rash primarily on the hands and feet, whereas measles progresses from head to extremities and scarlet fever mainly affects the axillary and inguinal folds [6]. Additionally, KD often involves swelling of the hands and feet. Scarlet fever, in contrast to KD and measles, does not present with conjunctivitis. Measles can also be identified by Koplik's spots [6].

Coronary anomalies like aneurysms can appear in KD within the first week of the illness [7]. TTE is preferred for detecting these aneurysms due to its lack of radiation, though it may not clearly show the distal coronary arteries [8]. Angiography offers the most accurate vascular assessment. Aneurysms are categorized by size as small (<5 mm internal diameter), medium (5–8 mm internal diameter), or giant (>8 mm internal diameter) [9]. CT coronary angiography, a noninvasive approach, is crucial for assessing aneurysms, including their number, distribution, size, potential intraluminal thrombi, and related complications like myocardial infarction. It also plays a vital role in the ongoing monitoring of coronary aneurysms. KD may also present with other cardiac is-

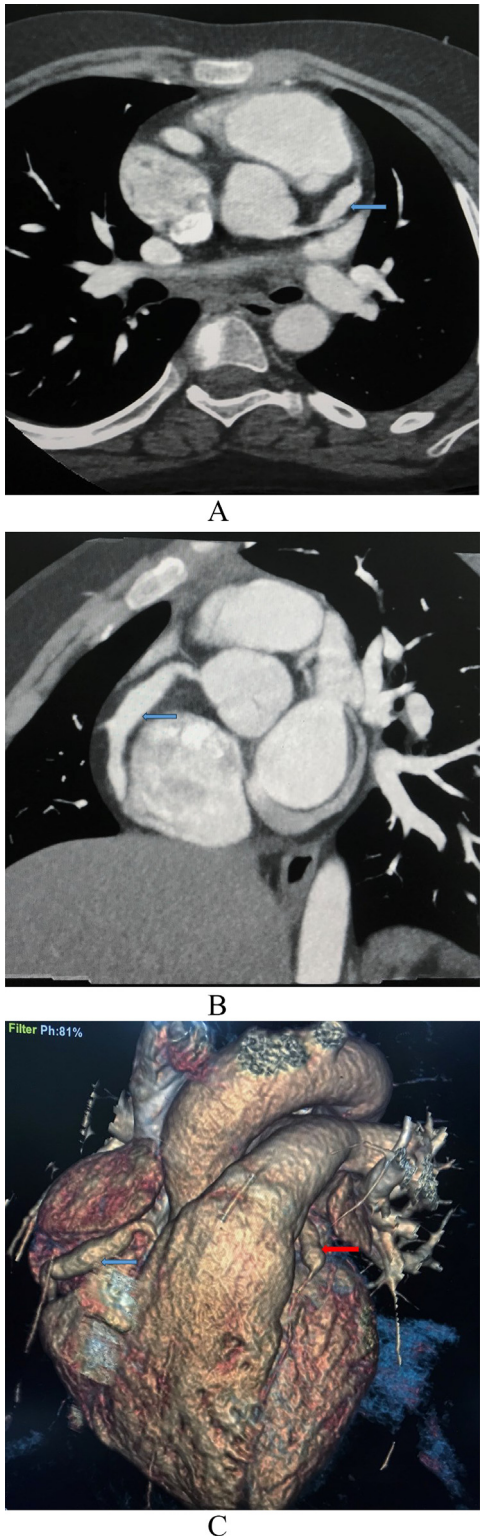

Fig. 2 – (A) Oblique axial view of CT Coronary angiogram showing fusiform aneurysm of left anterior descending artery (blue arrow). (B) Oblique coronal view of CT Coronary angiogram depicting right coronary artery aneurysm (blue arrow). (C) Reformatted CT coronary angiography image showing aneurysms in left anterior descending artery (red arrow) and mid right coronary artery (blue arrow).

sues such as pericardial effusion, myocarditis, valvular regurgitation, premature atherosclerosis, embolic phenomena, and fistula formation [10]. Coronary stenosis typically affects the left coronary artery more, while thromboses are more common in the right [11]. Magnetic resonance (MR) angiography can also assess aneurysms and myocardial perfusion but are less favored due to availability issues, long scan times, and potential for motion artifacts.

Preventing aneurysms is a key goal in treating KD [3]. The standard treatment includes intravenous immunoglobulin (IVIG) and aspirin [12]. If there's no initial response, a second dose of IVIG may be administered with or without corticosteroids or other treatments [12]. The severity and presence of coronary aneurysms at the time of diagnosis guide the treatment plan and the need for intense, long-term cardiovascular monitoring and thromboprophylaxis [12]. Once large aneurysms form, the risk of severe cardiac complications significantly increases. Timely diagnosis and treatment with IVIG can drastically reduce the risk of coronary artery complications from 25% to 5% [3]. Coronary aneurysms may remain stable, regress, or enlarge over time, but about half regress spontaneously within 2 years of disease onset [13].

Conclusion

KD predominantly affects younger children, usually those under 4 years old, but it can also occur in older children, where a high level of clinical suspicion is necessary for detection. While KD is typically a self-limiting illness, involvement of the coronary arteries can lead to serious health consequences. Prompt and accurate diagnosis and intervention are crucial to reduce or prevent severe coronary complications. This case emphasizes the importance of considering KD when diagnosing febrile children with rash and cervical lymphadenopathy, and highlights the need for early use of echocardiography and CT coronary angiography to check for potential coronary issues.

Patient consent

Written informed consent was obtained from the patient's parents for publication of this case report and any accompanying images.

REFERENCES

[1] Elakabawi K, Lin J, Jiao F, Guo N, Yuan Z. Kawasaki disease: global burden and genetic background. Cardiol Res 2020;11(1):9–14.
[2] Rowley AH, et al. Kawasaki syndrome. In: Gershon AA, Hotes PJ, Katz SL, et al., editors. Krugman's infectious diseases of children. Philadelphia, PA: Mosby Inc; 2004. p. 323–35.
[3] Yeom JS, Woo HO, Park JS, Park ES, Seo JH, Youn HS. Kawasaki disease in infants. Korean J Pediatr 2013;56(9):377–82.

[4] Kobayashi T, Ayusawa M, Suzuki H, Abe J, Ito S, Kato T, et al. Revision of diagnostic guidelines for Kawasaki disease (6th revised edition). Pediatr Int 2020;62(10):1135–8.
[5] Newburger JW, Takahashi M, Gerber MA, Gewitz MH, Tani LY, Burns JC, et al. Diagnosis, treatment, and long-term management of kawasaki disease: a statement for health professionals from the committee on rheumatic fever, endocarditis and kawasaki disease, council on cardiovascular disease in the young, American Heart Association. Circulation. 2004;110(17):2747–71.
[6] Lim KYY, Chua MC, Tan NWH, Chandran S. Reactivation of BCG inoculation site in a child with febrile exanthema of 3 days duration: an early indicator of incomplete Kawasaki disease. BMJ Case Rep 2020;13(12):e239648.
[7] Dominguez SR, Anderson MS, El-Adawy M, Glodé MP. Preventing coronary artery abnormalities: a need for earlier diagnosis and treatment of Kawasaki disease. Pediatr Infect Dis J 2012;31(12):1217–20.
[8] Margossian R, Lu M, LuAnn Minich L, Bradley TJ, Cohen MS, Li JS, et al. Predictors of coronary artery visualization in Kawasaki disease. J Am Soc Echocardiogr 2011;24(1):53–9.
[9] Dajani AS, Taubert KA, Takahashi M, Bierman FZ, Freed MD, Ferrieri P, et al. Guidelines for long-term management of patients with Kawasaki disease. Report from the Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease, Council on Cardiovascular Disease in the Young, American Heart Association. Circulation. 1994;89(2):916–22.
[10] Tsujioka Y, Handa A, Nishimura G, Miura M, Yokoyama K, Sato K, et al. Multisystem imaging manifestations of Kawasaki disease. Radiographics 2022;42(1):268–88.
[11] Broncano J, Vargas D, Bhalla S, Cummings KW, Raptis CA, Luna A. CT and MR imaging of cardiothoracic vasculitis. Radiographics 2018;38(4):997–1021.
[12] McCrindle BW, Rowley AH, Newburger JW, Burns JC, Bolger AF, Gewitz M, et al. Diagnosis, treatment, and long-term management of kawasaki disease: a scientific statement for health professionals from the American Heart Association. Circulation [Internet] 2017;135(17):e927–99. [Accessed March 9, 2024]Available from: https://www.ahajournals.org/doi/10.1161/CIR.0000000000000484 .
[13] Sasaguri Y, Kato H. Regression of aneurysms in Kawasaki disease: a pathological study. J Pediatr 1982;100(2):225–31.