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Case Report: Late Identification of Children with Complete Kawasaki Disease with Left Coronary Artery Aneurysm in Remote Area: Kutai Barat

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Abstract: This case report discusses a 6-year-old child with Kawasaki disease and findings of an aortic aneurysm. The patient's complaints and clinical presentation raised suspicion of Kawasaki disease. Although delayed, an echocardiogram was performed approximately one week after the onset of symptoms to assess potential complications that could worsen if left undetected. This case report emphasizes the significance of diagnostic imaging in detecting cardiac complications in Kawasaki disease cases, as well as the importance of follow-up examination and treatment planning for the patient. Despite limitations in facilities and access, this case report emphasizes that monitoring echocardiographic findings supports the prevention of worsening aneurysms.

Keyword: Kawasaki disease, aortic aneurysm, early identification, echocardiography

INTRODUCTION

Kawasaki disease was initially identified in Japan and predominantly affects children under the age of five. Over time, cases have been increasingly reported in developing countries. In Indonesia, fewer than 200 cases are documented annually, with the majority occurring in children below five years of age. However, the oldest reported case involved a 16-year-old child.[1]

This disease is characterised by acute systemic vasculitis, presenting with symptoms such as conjunctival injection, oral mucosal changes, extremity alterations, and cervical lymphadenopathy. One of the most frequent complications is the development of aortic aneurysms, which may lead to myocardial ischemia if effective treatment is not administered.[2]

Aortic aneurysms can regress over time, but also have the potential to enlarge, eventually forming giant aneurysms. Additionally, stenotic changes and calcification may occur, progressing long after the initial onset of the disease. [3]

The detection of aortic aneurysms is primarily achieved through echocardiography, a preferred method in pediatric patients due to its non-invasive nature and absence of radiation exposure. [4]

Treatment with intravenous immunoglobulin (IVIG) and aspirin has been shown to reduce the risk of aortic aneurysm formation during the acute phase of the illness, as well as alleviate clinical symptoms. In cases with persistent aneurysms, consideration may be given to dual antiplatelet therapy and beta-blockers. [5]

METHOD

Case Presentation

This case involves a 6-year-old child who initially presented with a fever lasting more than seven days that did not improve despite taking antipyretic. Additional symptoms included abdominal pain, cracked lips, and peeling skin. The child also complained of significant redness affecting the eyes, mouth, and palms of the hands. These symptoms were unprecedented for the patient. Subsequently, the child was taken to a paediatrician's office and was suspected of having Kawasaki disease within the same week. Aspirin was administered at the paediatrician's clinic, and the physician recommended clinical follow-up at the hospital the following week.

By the second week, the redness in the eyes and mouth had diminished, and the fever had subsided. No other complaints were reported. The child's weight was recorded at 15.9 kg, and height at 116 cm. Laboratory tests revealed a hemoglobin level of 12.6 g/dL, decreased mean corpuscular volume (MCV) of 73.4 fL, reduced mean corpuscular hemoglobin (MCH) of 23.8 pg, lower mean corpuscular hemoglobin concentration (MCHC) of 32.4 g/dL, elevated leukocyte count of 13,500/µL, increased platelet count of 514,000/µL, and an erythrocyte sedimentation rate (ESR) of 73 mm/hour. Urinalysis showed no abnormalities.

Figure 1. Clinical photograph of a patient with Kawasaki disease

Echocardiographic examination revealed dilation of the left coronary artery measuring 0.4 cm and the right coronary artery measuring 0.2 cm. An electrocardiogram (ECG) was not performed for this patient. The patient's family was informed that the standard management for children with this condition involves administration of intravenous immunoglobulin (IVIG) and that the patient would be referred to Samarinda for treatment. However, due to a shortage of IVIG stock, the patient was unable to receive this therapy at that time. Instead, the patient was treated with aspirin at a dose of 100 mg four times daily (30 mg/kg/day) for two weeks.

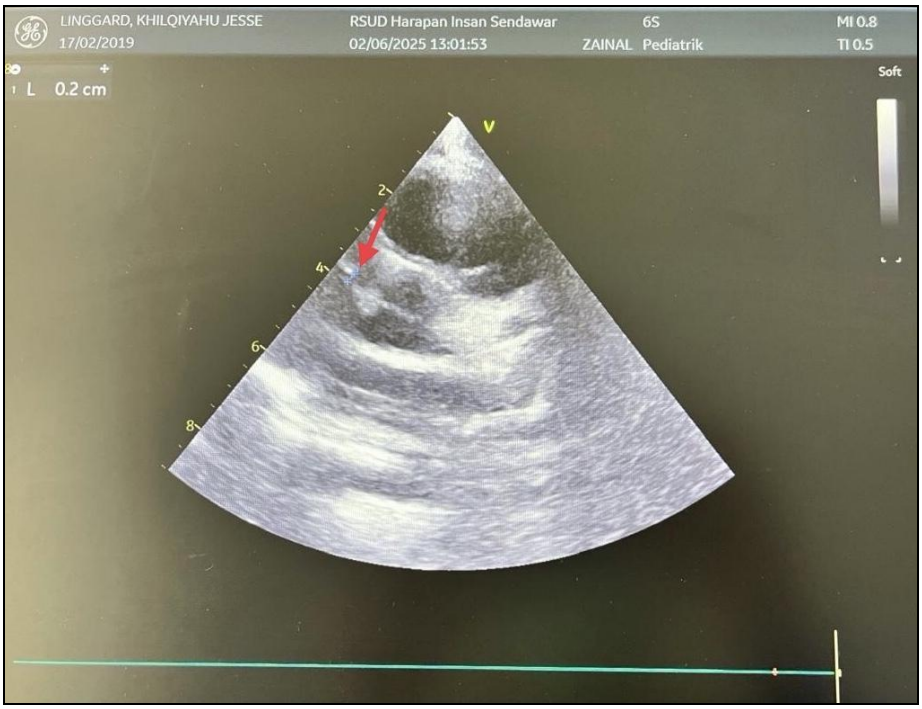

Figure 2. (A) size of the right coronary artery

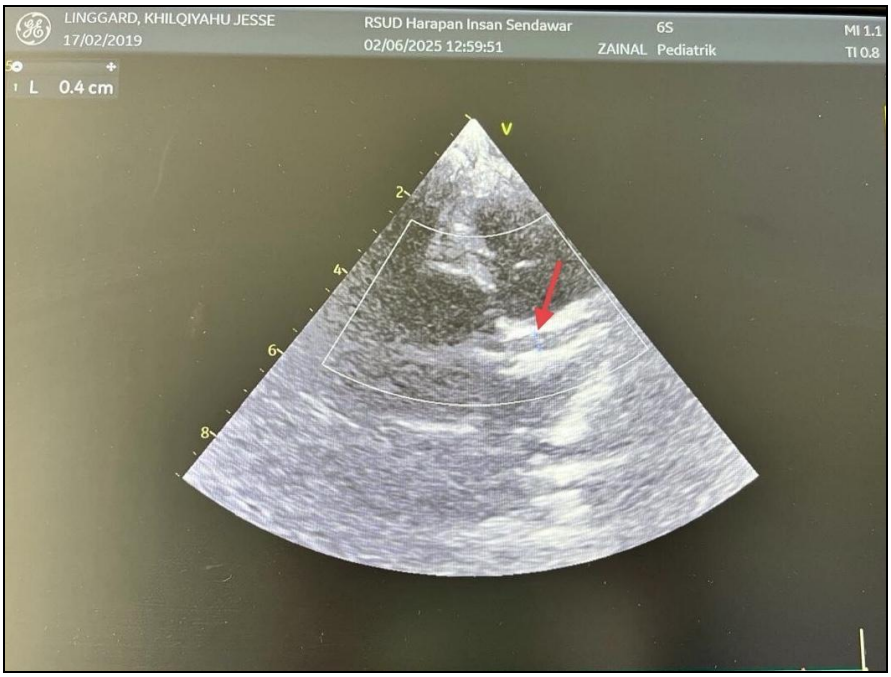

Figure 2. (B) Size of the left coronary artery

RESULTS AND DISCUSSION

This case involves a 6-year-old boy. Consistent with disease prevalence, Kawasaki disease generally occurs in children under the age of five, though cases have been reported in patients as old as 16 years. It is also more common in males than females, with a ratio of approximately 1.6:1. [1] The incidence of Kawasaki disease is higher in Asia, which is thought to be related to genetic factors influencing both the risk of aortic aneurysm and the response to intravenous immunoglobulin (IVIG) therapy. [6] Cases in Kutai Barat are considered rare, and treatment options remain limited in this region.

The child in this case presented with symptoms including fever lasting more than one week, red eyes, red tongue, dry lips, and mild skin peeling. These symptoms fulfil the cardinal criteria for Kawasaki disease diagnosis: rash, conjunctivitis, arthritis, red and cracked lips, and cervical lymphadenopathy. [5]

In acute cases, echocardiography is essential for assessing coronary artery involvement, such as aortic aneurysms. However, a normal echocardiogram cannot completely rule out coronary artery pathology, which may require more advanced imaging techniques like CT angiography. It is important to differentiate aneurysms from aortic ectasia, where dilation occurs without segmental aneurysm formation. [7] In this case, echocardiography confirmed aortic aneurysm consistent with Kawasaki disease suspicion. [8]

Laboratory findings revealed anaemia, leukocytosis, and elevated erythrocyte sedimentation rate (ESR), although C-reactive protein (CRP) levels were not measured. These lab results align with typical inflammatory markers observed in Kawasaki disease, which include elevated CRP, anaemia, thrombocytopenia, and hyponatremia. [8]

Pathologically, coronary artery changes in Kawasaki disease begin with acute necrotising arteritis, usually within the first two weeks from onset. Necrosis affects all arterial layers and is triggered by neutrophils, followed by a subacute vasculitis phase and later chronic myofibroblastic luminal proliferation. The acute phase may resolve spontaneously, but can persist and lead to rupture within the first month. [9] The patient in this case was still within the acute phase, being less than two weeks from symptom onset.

Aneurysms may regress depending on various factors, including a smaller initial size during the acute phase, age under one year, a fusiform aneurysm shape, and a more distal aortic location. Early administration of IVIG also supports aneurysm regression. [10]
The size of the aortic aneurysm is a prognostic factor that guides further therapy and clinical monitoring. Over time, aneurysms may persist, progress to stenosis, or develop thrombosis. [11] Aneurysms are classified as small if the diameter is under 0.4 cm, medium if between 0.4 and 0.8 cm, and giant if larger than 1 cm. [12] According to this classification, this case qualifies as a small aneurysm. The left coronary artery measured 0.4 cm, with dilation most evident in the mid-section of the aorta.

To categorise aneurysm size more precisely, Z-scores adjusted for body surface area (BSA) are used. [5] This scoring system guides ongoing management, classifying small aneurysms as Z-scores between 2.5 and <5, medium between 5 and 10, and giant above 10. [13] This patient falls within the small aneurysm category.

Besides Kawasaki disease, aortic aneurysms can arise from other conditions such as connective tissue disorders, congenital coronary abnormalities, infections, including endocarditis, trauma (including surgical history), or iatrogenic causes. [14] Typically, aneurysms in Kawasaki disease are located in the proximal left anterior descending artery, followed by the proximal right coronary artery and the left main coronary artery. In this patient, dilation was noted in the left coronary artery, consistent with these predilection sites. [14]

Electrocardiography (ECG) can reveal signs of ischemia, including ST-segment and T-wave changes, as well as arrhythmias. [15] However, an ECG was not performed in this case, and the patient did not report palpitations or chest pain.

Recommended treatment for Kawasaki disease includes IVIG and aspirin, ideally administered within the first 10 days of illness to reduce the risk of aneurysm formation and limit aneurysm size. [5,16] The recommended IVIG dose is 2 g/kg given over 10 to 12 hours. [17]

Aspirin can be administered at high doses (80–100 mg/kg/day) or lower doses (30–50 mg/kg/day), with no significant differences in outcomes observed between the two dosing regimens. High-dose aspirin is typically administered for the first two weeks, followed by low-dose therapy for up to eight weeks. In this case, a low dose of 30 mg/kg/day was given for two weeks, consistent with guideline recommendations. The patient experienced improvement in fever and other symptoms after aspirin therapy. [18] For medium-sized aneurysms, adjunctive antiplatelet treatment such as P2Y12 inhibitors may be considered. [19] For giant aneurysms, combined antiplatelet and anticoagulant therapy with warfarin or low-molecular-weight heparin (LMWH) is recommended. [20]

IVIG resistance is associated with a higher risk of aortic aneurysm development. In this case, the pediatric specialist at RSUD Harapan Insan Sendawar referred the patient to Samarinda for IVIG therapy. However, IVIG was unavailable at hospitals in Samarinda at that time, so the patient received only aspirin. Although the patient's symptoms improved and fever resolved, aortic dilation had already developed. Continued clinical and echocardiographic follow-up is necessary to monitor symptoms and assess aneurysm progression under ongoing aspirin therapy. [5]

Each patient's risk of cardiac complications, such as myocardial ischemia and thrombosis, can be evaluated through serial follow-up of coronary artery Z-scores and aneurysm size progression. Risk stratification guides the timing of follow-up and the need for additional therapy. [5]

CONCLUSION

The occurrence of aortic aneurysms in Kawasaki disease warrants careful attention through appropriate supportive examinations tailored to the patient's characteristics. It is also essential to differentiate this condition from other differential diagnoses that may present with aortic aneurysms. In cases where diagnostic and therapeutic facilities are limited, patients should be referred to specialised centres to ensure a better prognosis. In remote areas, management can be optimised through additional pharmacological treatments, early supportive examinations, and patient education to encourage continued follow-up, thereby improving control over disease progression.

REFERENCES

Mayasari, R., -, F. and K, S. (2019) "PENYAKIT KAWASAKI", *Media Dermato-Venereologica Indonesiana*, 46(2).

Srinivasan R, Weller R, Chelliah A, Einstein AJ. Multimodality cardiac imaging in a patient with Kawasaki disease and giant aneurysms Case Rep Pediatr. 2016;2016:4298098

Hoshino S, Tsuda E, Yamada O. Characteristics and Fate of Systemic Artery Aneurysm after Kawasaki Disease. J Pediatr 2015;167(1):108–112. e1–e2.

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;80:526–8

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(17):e927–e999

Onouchi Y. Genetics of Kawasaki disease: what we know and don't know. *Circ J*. 2012;76:1581–1586.

JCS Joint Working Group. Guidelines for diagnosis and management of cardiovascular sequelae in Kawasaki disease (JCS 2008): digest version. *Circ J*. 2010;74:1989–2020.

Newburger JW, Takahashi M, Burns JC. Kawasaki Disease. J Am Coll Cardiol 2016;67(14):1738–1749.

Kanai T, Ishiwata T, Kobayashi T, et al. Ulinastatin, a urinary trypsin inhibitor, for the initial treatment of patients with Kawasaki disease: a retrospective study. Circulation 2011;124(25):2822–2828

Friedman KG, Gauvreau K, Hamaoka-Okamoto A, et al. Coronary Artery Aneurysms in Kawasaki Disease: Risk Factors for Progressive Disease and Adverse Cardiac Events in the US Population. J Am Heart Assoc 2016;5(9):e003289.

McCrindle BW, Li JS, Minich LL, et al. Coronary artery involvement in children with Kawasaki disease: risk factors from analysis of serial normalized measurements. Circulation 2007;116(2):174–179.

de Zorzi A, Colan SD, Gauvreau K, Baker AL, Sundel RP, Newburger JW. Coronary artery dimensions may be misclassified as normal in Kawasaki disease. *J Pediatr*. 1998;133:254–258.

Marchesi A, Tarissi de Jacobis I, Rigante D, et al. Kawasaki disease: guidelines of the Italian Society of Pediatrics, part I - definition, epidemiology, etiopathogenesis, clinical expression and management of the acute phase. Ital J Pediatr 2018;44(1):102.

Onouchi Z, Shimazu S, Kiyosawa N, Takamatsu T, Hamaoka K. Aneurysms of the coronary arteries in Kawasaki disease. An angiographic study of 30 cases. Circulation 1982;66(1):6–13.

Sumitomo N, Karasawa K, Taniguchi K, Ichikawa R, Fukuhara J, Abe O, Miyashita M, Kanamaru H, Ayusawa M, Harada K. Association of sinus node dysfunction, atrioventricular node conduction abnormality and ventricular arrhythmia in patients with Kawasaki disease and coronary involvement. *Circ J*. 2008;72:274–280.

Kuwabara M, Yashiro M, Ae R, Yanagawa H, Nakamura Y. The effects of early intravenous immunoglobulin therapy for Kawasaki disease: The 22nd nationwide survey in Japan. Int J Cardiol 2018;269(334):338.

Oates-Whitehead RM, Baumer JH, Haines L, Love S, Maconochie IK, Gupta A, Roman K, Dua JS, Flynn I. Intravenous immunoglobulin for the treatment of Kawasaki disease in children. *Cochrane Database Syst Rev*. 2003;(4):CD004000.

Baumer JH, Love SJ, Gupta A, Haines LC, Maconochie I, Dua JS. Salicylate for the treatment of Kawasaki disease in children. *Cochrane Database Syst Rev*. 2006;(4):CD004175.

Gerschutz GP, Bhatt DL; Clopidogrel in Unstable Angina to Prevent Recurrent Events study. The Clopidogrel in Unstable Angina to Prevent Recurrent Events (CURE) study: to what extent should the results be generalizable? *Am Heart J*. 2003;145:595–601.

Sugahara Y, Ishii M, Muta H, Iemura M, Matsuishi T, Kato H. Warfarin therapy for giant aneurysm prevents myocardial infarction in Kawasaki disease. *Pediatr Cardiol*. 2008;29:398–401.