Systemic Artery Aneurysm in Two Children with Kawasaki Disease: A Frequently Overlooked Lesion

Shaikh Habiba Ahmed, Shakeel Ahmed, Shafiq Ur Rehman Farooq and Saleem Akhtar

Department of Paediatrics and Child Health, The Aga Khan University Hospital, Karachi, Pakistan

ABSTRACT

Kawasaki disease (KD) is a multisystem vasculitis affecting medium-sized arteries. While coronary involvement is a well-documented complication, extensive disease is often associated with systemic arterial involvement. Multiple studies have reported the occurrence of systemic and pulmonary arterial involvement. This atypical presentation underscores the importance of a thorough diagnostic evaluation, including early echocardiography, radiological imaging, and CT angiography, when KD is suspected. Early identification of atypical presentations can significantly improve the disease prognosis by reducing the risks of vascular stenosis, obstruction, and thrombosis. This case series highlights the underexplored atypical presentations of KD, emphasising the necessity for timely recognition, investigation, and management of systemic and pulmonary arterial involvement alongside coronary complications.

Key Words: *Systemic artery aneurysm, Atypical Kawasaki disease, Giant coronary artery aneurysm.*

How to cite this article: Ahmed SH, Ahmed S, Farooq SUR, Akhtar S. Systemic Artery Aneurysm in Two Children with Kawasaki Disease: A Frequently Overlooked Lesion. *JCPSP Case Rep* 2025; **3**:398-401.

INTRODUCTION

Kawasaki disease (KD) is an inflammatory disease of unknown aetiology mainly affecting children less than 5 years of age.[1]

A classic case of KD is characterised by fever of more than five days and four out of the following five criteria: Mucocutaneous changes (cracked lips and strawberry tongue), conjunctivitis, rash, extremity changes, and lymphadenopathy.[1] However, some cases do not fulfil the classic criteria and are classified as atypical or incomplete KD. The most serious complication associated with untreated KD is coronary artery involvement, which can occur in approximately 20-25% of cases if appropriate treatment is not initiated within the acute phase of 7-10 days.[2] Recommended treatment with aspirin and intravenous immunoglobulin (IVIG) significantly reduces the likelihood of coronary artery involvement to about 5%.[2] Since KD involves vasculitis of medium-sized arteries, systemic arterial aneurysms may develop in approximately 2.2% of children with complete KD.[3] Systemic artery aneurysms (SAAs) associated with the incomplete KD are rare and poorly described. This report presents two cases of incomplete KD with SAAs, both of which demonstrated regression following appropriate therapy.

Correspondence to: Dr. Saleem Akhtar, Department of Paediatrics and Child Health, The Aga Khan University Hospital, Karachi, Pakistan
E-mail: saleem.sadqani@aku.edu

Received: March 11, 2025; Revised: May 06, 2025; Accepted: July 12, 2025
DOI: https://doi.org/10.29271/jcpspcr.2025.398

CASE 1:

A three-month and ten-days male infant presented to the Emergency Department with fever and irritability for one month.

On examination, the child had normal anthropometric measurements but appeared irritable, febrile, and tachycardic. The rest of the examination was unremarkable, and laboratory findings indicated anaemia and raised inflammatory markers (CRP and ESR). However, despite initial investigation, a firm diagnosis could not be established. He was initially managed on the lines of sepsis, and treatment included broad-spectrum antibiotics along with supportive care. However, the persistent fever and raised inflammatory markers warranted further evaluation. CT of the abdomen and chest with contrast was done, which revealed multiple aneurysms in large and medium-sized vessels and infarcts in both kidneys and spleen, raising suspicion of systemic vasculitis (Figure 1, 2). Considering this, echocardiography was done, which showed giant coronary artery aneurysms and demonstrated severe dilation of the left main coronary artery, right coronary artery, left anterior descending artery, left circumflex artery, and partial thrombus in the right coronary and left anterior descending arteries. However, cardiac function was normal, and there was no valve regurgitation. Hence, a diagnosis of incomplete KD was made with systemic and coronary artery aneurysms. The child was treated with IVIG at a dose of 2 g/kg and high-dose aspirin. However, due to no response and persistent fever, a second dose of IVIG was administered along with the high pulse methyl prednisolone (30 mg/kg) for 3 days, followed by oral steroids and azathioprine. With this treatment, the fever subsided, inflammatory markers improved, and the child was later discharged in stable condition on aspirin (5 mg/kg), a tapering dose of prednisolone, and a ther-

apeutic dose of enoxaparin for treatment and prevention of coronary thrombosis. At subsequent follow-up in the cardiology and haematology clinics, the child remained afebrile; hence, his immunosuppressive medications were weaned off. However, anticoagulation therapy was continued.

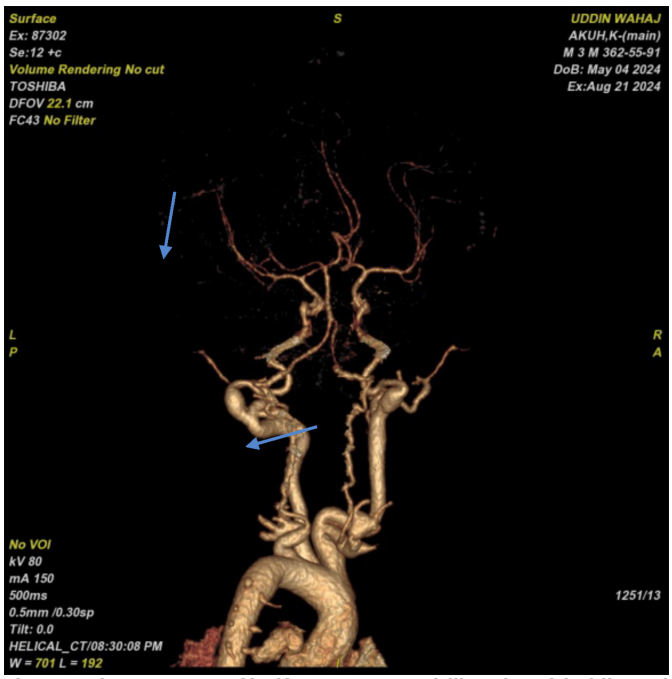

Figure 1: Short segment of fusiform aneurysmal dilatation of the bilateral internal and external carotid arteries (blue arrows).

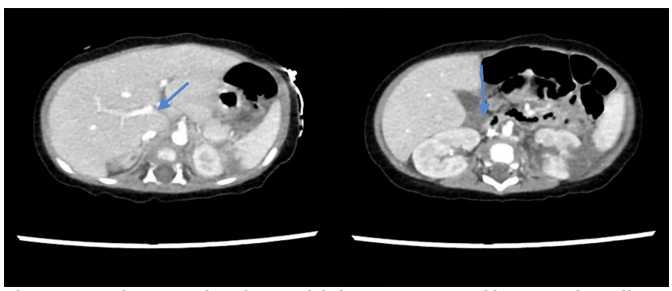

Figure 2: CT images showing multiple aneurysms of large and medium-sized vessels, *i.e.*, coeliac and superior mesentery artery (blue arrows).

CASE 2:

A two-month and twenty-one-day male infant presented to the Emergency Department with complaints of fever and rash lasting one month. This was accompanied by mild redness of the eyes, raising suspicion of measles. He was admitted to a hospital for workup and treatment of fever. He received broad-spectrum antibiotics but failed to resolve the symptoms. He also had an extensive workup done, which did not establish a diagnosis. Due to persistent fever, the patient was brought to this hospital for further evaluation. During his hospital admission, he had an echocardiogram done, which showed a small atrial septal defect. The rest of the study was reported as normal. The infant was admitted for seven days and was treated on the lines of pneumonia and discharged on intravenous antibiotic therapy, but later returned to the ED due to persistent fever. A repeated echocardiogram was done to rule out infective endocarditis. It revealed a small secundum atrial septal defect which was insignificant. However, the coronary arteries were severely dilated with a giant aneurysm affecting both the right main coronary and the left anterior descending and circumflex branches of the left coronary arteries. There was suspicion of a partial thrombus in the right coronary artery with a smaller thrombus in the left circumflex artery. Treatment for KD was initiated with high-dose aspirin and IVIG. Enoxaparin was also administered due to extensive coronary artery dilation and thrombus.

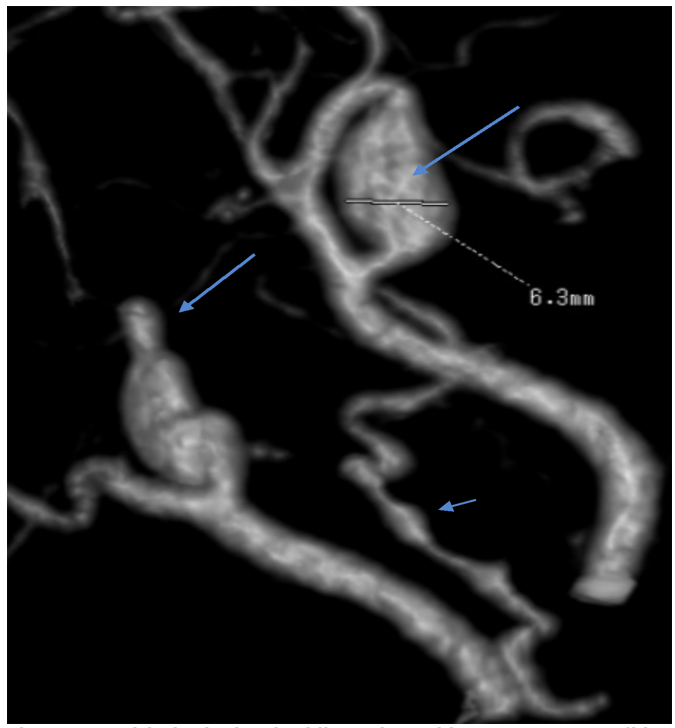

Figure 3: CT of the brain showing bilateral carotid artery aneurysms (blue arrows).

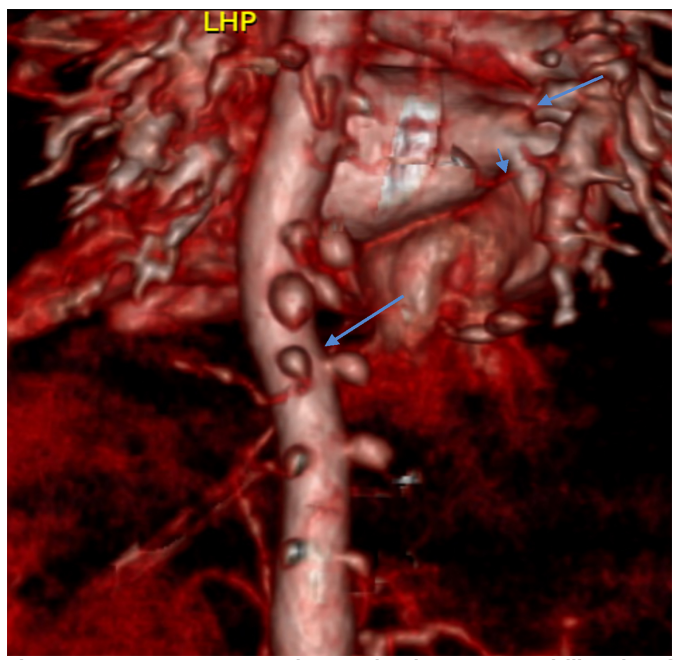

Figure 4: Pre-treatment CT-angiogram showing aneurysmal dilatation of bilateral intercostal arteries.

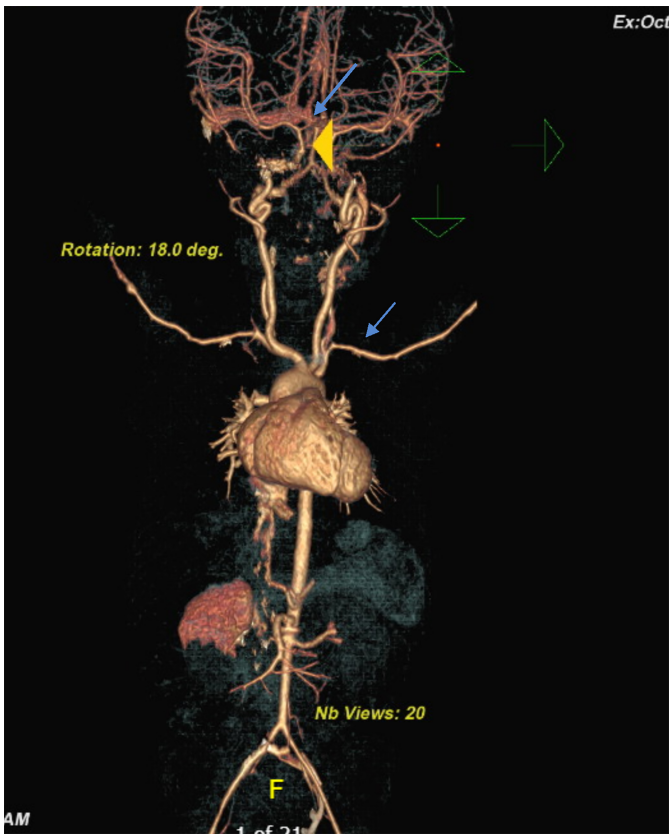

Figure 5: Post-treatment follow-up CT-angiogram showing reduction in the size of systemic artery aneurysms.

Due to persistent irritability, CT of the brain with contrast was done, which revealed aneurysmal dilatation of the bilateral internal carotid arteries, just after the bifurcation of the common carotid arteries (Figure 3). CT of the chest and abdomen showed aneurysmal dilatation at the origins of bilateral intercostal, coeliac, superior mesenteric, and renal arteries. A pre-treatment CT angiogram was done, showing bilateral intercostal artery involvement (Figure 4).

Due to extensive vasculitis and elevated inflammatory markers despite the 1st dose of IVIG, the child received a second dose of IVIG along with the pulse-dose methylprednisolone. This was followed by immunosuppressive therapy with azathioprine and hydroxychloroquine. After the addition of the second line immunosuppressive medication, the fever settled and inflammatory markers improved. The child's condition improved, and he was discharged in stable condition on immunosuppressants and anticoagulants. A follow-up visit in late 2024, included contrast-enhanced CT neck and chest imaging with angiography, which demonstrated interval improvement in previously noted aneurysmal dilatation of the bilateral internal carotid, intercostal, left circumflex, and bilateral renal arteries (Figure 5), while aneurysmal prominence of the right coronary, left anterior descending, and bilateral renal arteries remained unchanged.

DISCUSSION

Coronary artery aneurysms are the most feared complication of KD, which can have catastrophic consequences if not picked up properly. However, the recent studies also report the involvement of the systemic arteries as well. According to Yasuda *et al.*, the severe systemic vasculitis in younger infants shows multiple symmetrical SAAs.[3]

The prevalence of SAAs indicates a malignant course of KD. Both of these patients also demonstrated a protracted disease course that did not respond to two doses of IVIG and required 2nd line immunosuppressive medication to control the disease activity. Systemic aneurysms mostly affect the subclavian, brachial, axillary, and iliac arteries. In both of these cases, there was an extensive involvement of the systemic arteries, including renal, intercostal, and carotid arteries.[4] Patients have also been reported with stenosis in the bilateral common iliac arteries as a delayed outcome in disease progression.[5] However, neither of our cases had stenosis of any major blood vessel. In case of atypical presentation, the involvement of lobar and segmental pulmonary arteries has also been reported, which sparks a question on the extensive workup of giant and diffuse cardiac artery aneurysms and SAAs.[6] However, in both of these cases, no pulmonary artery involvement was found.

In both aortic and SAAs, early anticoagulation is the mainstay of treatment.[7] In acute and subacute giant and newly detected coronary aneurysms and thrombosis, aggressive thrombolytic therapy is required.[8]

Both the cases represent the extensive involvement of systemic arteries in KD along with coronary involvement. Undue delay in diagnosis and delayed treatment pose a major risk of systemic aneurysms, stenosis, obstruction, and thrombosis.[9]

SAAs are rare in KD but should be suspected in cases presenting with giant coronary artery aneurysms. These aneurysms tend to regress over time with appropriate therapy. A comprehensive diagnostic workup, including pulmonary artery imaging, is essential to ensure early identification of KD and its atypical presentations. Long-term follow-up with extensive cardiac evaluation, serial echocardiography, and routine cardiovascular assessment is recommended based on the risk of coronary involvement.

PATIENTS' CONSENT:

Written informed consent was obtained from the patients' guardians.

COMPETING INTEREST:

The authors declared no conflict of interest.

AUTHORS' CONTRIBUTION:

SHA: Literature search, drafting of manuscript, interpretation, reviewing, and proofreading of the draft.
SA: Review regarding case presentation.
SURF: Literature search and proofreading.
SA: Diagnosis, conceptualisation and critical revision, data interpretation, and final drafting of the manuscript.
All authors approved the final version of the manuscript o be published.

REFERENCES

1. Kuo HC. Diagnosis, progress, and treatment update of Kawasaki disease. *Int J Mol Sci* 2023; **24(18)**:13948. doi: 10.3390/ijms241813948.
2. Rife E, Gedalia A. Kawasaki disease: An update. *Curr Rheumatol Rep* 2020; **22(10)**:75. doi: 10.1007/s11926-020-00941-4.
3. Hoshino S, Tsuda E, Yamada O. Characteristics and fate of systemic artery aneurysm after Kawasaki disease. *J Pediatr* 2015; **167(1)**:108-12.e1.2. doi: 10.1016/j.jpeds.2015.04.036.
4. Kadiyani L, Mohankumar SP, Arumugam P, Gupta SK, Bagri NK, Pandey N, *et al*. Systemic artery aneurysms–A marker of high-risk Kawasaki disease. *Ann Pediatr Cardiol* 2024; **17(5)**:377-80. doi: 10.4103/apc.apc_156_24.
5. Tsuda E. Long-term results of large common iliac artery aneurysms caused by Kawasaki disease in four patients. *Cardiol Young* 2023; **33(9)**:1686-90. doi: 10.1017/S1047951112200316X.
6. Briceno-Medina M, Perez M, Waller BR, Sathanandam S. Systemic and pulmonary artery aneurysms in incomplete Kawasaki disease. *J Cardiol Cases* 2016; **13(6)**:185-8. doi: 10.1016/j.jccase.2016.02.003.
7. Chu C, He L, Lin YX, Xie LP, Liu F. Giant bilateral axillary artery aneurysms with left complete obstructive thrombus in intravenous immunoglobulin-sensitive Kawasaki disease: A case report. *Pediatr Rheumatol Online J* 2021; **19(1)**:157. doi: 10.1186/s12969-021-00643-w.
8. Gao L, Xie C, Zhang Q, Wang X, Fu S, Hu J, *et al*. Coronary thrombosis and myocardial ischemia in Kawasaki disease: A case report. *BMC Cardiovasc Disord* 2024; **24(1)**:473. doi: 10.1186/s12872-024-04148-8.
9. Yacoe ME, Dake MD. Development and resolution of systemic and coronary artery aneurysms in Kawasaki disease. *Am J Roentgenol* 1992; **159(4)**:708-10. doi: 10.2214/ajr.159.4.1529832.