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Case Report

Late-diagnosed Incomplete Kawasaki Disease Complicated by Giant Coronary Artery Aneurysms and Ischemic Heart Failure: A Pediatric Case Report

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ABSTRACT

Incomplete Kawasaki disease may present significant diagnostic challenges in young infants, frequently leading to delayed diagnosis and severe coronary complications. We report on a 3.5-month-old infant with a delayed diagnosis of incomplete Kawasaki disease who developed bilateral giant coronary artery aneurysms with progressive coronary obstruction. Despite appropriate medical management including immunoglobulin therapy and antithrombotic treatment, the patient developed ischemic heart failure and ultimately required coronary artery bypass grafting. This case highlights the diagnostic difficulty of incomplete Kawasaki disease in early infancy and emphasizes the importance of early echocardiographic evaluation in infants with prolonged unexplained fever.

Keywords: Kawasaki disease, incomplete Kawasaki disease, coronary artery aneurysm, ischemic cardiomyopathy, coronary artery bypass surgery

Introduction

Kawasaki disease is an acute, self-limited systemic vasculitis predominantly affecting infants and young children and represents the leading cause of acquired heart disease in developed countries (1). The disease is characterized by prolonged fever and mucocutaneous inflammatory findings. However, approximately 7-10% of patients, particularly infants younger than one year of age, present with incomplete Kawasaki disease, lacking the full set of classical diagnostic criteria (2,3).

In such cases, delayed diagnosis is common and it is associated with an increased risk of coronary artery abnormalities (1). Without treatment, coronary artery aneurysms develop in approximately 20-25% of patients (1). Early recognition and timely administration of intravenous immunoglobulin and acetylsalicylic acid (ASA) significantly reduce the risk of coronary complications (4). Delayed diagnosis is associated with severe outcomes, including giant coronary artery aneurysms, myocardial ischemia, and sudden death (5).

We report on a rare and severe case of delayed diagnosis of incomplete Kawasaki disease in a 3.5-month-old infant who developed bilateral giant coronary artery aneurysms, progressive coronary artery occlusion, and ischemic heart failure requiring coronary artery bypass grafting.

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Case Report

A previously healthy 3.5-month-old male infant presented with persistent fever (38 °C) and generalized rash. The chief complaint at admission was persistent fever accompanied by irritability and skin rash. Initial laboratory evaluation demonstrated marked leukocytosis (38,000/mm^3), elevated C-reactive protein (CRP) (94 mg/L), and thrombocytosis (720,000/mm^3). Empirical intravenous antibiotic therapy was initiated due to a suspicion of bacterial infection. Day 13 of illness: Fever persisted despite antibiotic therapy. Repeat laboratory tests revealed leukocytosis (31,000/mm^3), anemia (hemoglobin 8.1 g/dL), thrombocytosis (601,000/mm^3), elevated inflammatory markers (CRP 138 mg/L, erythrocyte sedimentation rate 78 mm/h), hypoalbuminemia (1.9 g/dL), and sterile pyuria. Due to meningismus, a lumbar puncture was performed. Cerebrospinal fluid analysis demonstrated pleocytosis with negative bacterial cultures, consistent with aseptic meningitis. Physical examination revealed persistent rash, hyperemic oropharynx and cracked lips, without conjunctivitis or cervical lymphadenopathy. Day 25 of illness: Due to persistent fever and elevated inflammatory markers, transthoracic echocardiography was performed. Echocardiography revealed giant coronary artery aneurysms, measuring 6 mm (Z-score 15.6) in the right coronary artery (RCA) and 10 mm (Z-score 23.0) in the left main coronary artery. Based on clinical findings, laboratory abnormalities, and coronary involvement, a diagnosis of incomplete Kawasaki disease according to the American Heart Association (AHA) diagnostic algorithm was established. Treatment with intravenous immunoglobulin (2 g/kg) and high-dose ASA (30-50 mg/kg/day) was initiated. Fever resolved rapidly and inflammatory markers normalized. Due to the presence of giant coronary artery aneurysms, antithrombotic therapy consisting of ASA and low-molecular-weight heparin was started in accordance with the current guidelines (4). At 16 months of age, systemic arterial involvement was detected, including bilateral axillary artery aneurysms and a focal aneurysm of the right internal iliac artery. The patient was followed with serial echocardiography and coronary computed tomography angiography. During long-term follow-up, electrocardiography demonstrated ischemic changes. Progressive left ventricular dilation and a decline in left ventricular ejection fraction from 65% to 55% were observed. Coronary angiography demonstrated complex coronary artery pathology. Aneurysmatic and calcified dilation was observed in the proximal and mid segments of the left anterior descending artery (LAD). Severe eccentric stenosis of approximately 80-90% was detected in the proximal circumflex artery. The distal circumflex artery was filled retrogradely through collateral circulation originating from the RCA posterior descending branch. The proximal RCA was found to be chronically totally occluded.

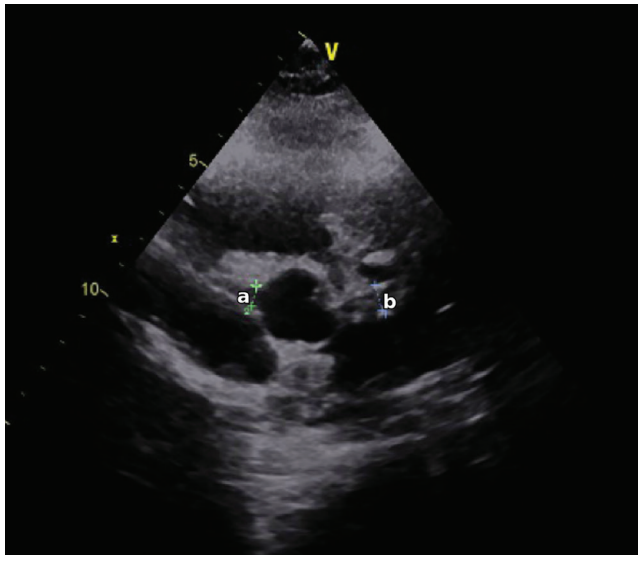

Figure 1. Image of aneurysms observed on transtrasic echocardiography at the time of diagnosis aneurysm (a) RCA aneurysm (b) LCA aneurysm
RCA: Right coronary artery, LCA: Left coronary artery

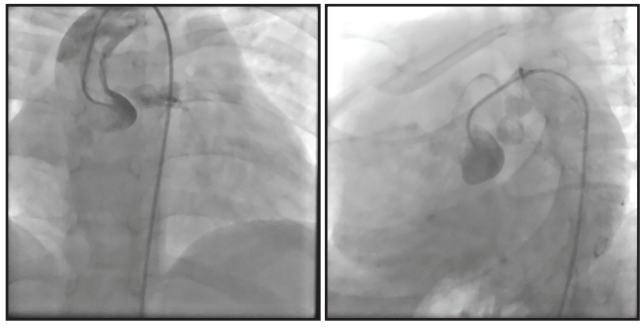

Figure 2. a) Angiographic imaging of coronary artery aneurysms, developing obstructions and collaterals: A distinct aneurysmatic calcified appearance is observed in the LAD proximal and mid segments. In the circumflex trunk/proximal segment, 80-90% eccentric stenosis is observed. It can be seen that the distal Cx is filled retrogradely from the RCA PD branch via collaterals. The RCA proximal is observed to have 100% chronic total occlusion. The collaterals originating from the sinus node branch are connected to the LAD proximal. b) Angiographic imaging of coronary artery aneurysms, developing obstructions and collaterals: A distinct aneurysmatic calcified appearance is observed in the LAD proximal and mid segments. In the circumflex trunk/proximal segment, 80-90% eccentric stenosis is observed. It can be seen that the distal Cx is filled retrogradely from the RCA PD branch via collaterals. The RCA proximal is observed to have 100% chronic total occlusion. The collaterals originating from the sinus node branch are connected to the LAD proximal
LAD: Left anterior descending artery, RCA: Right coronary artery, PD: Posterior descending artery

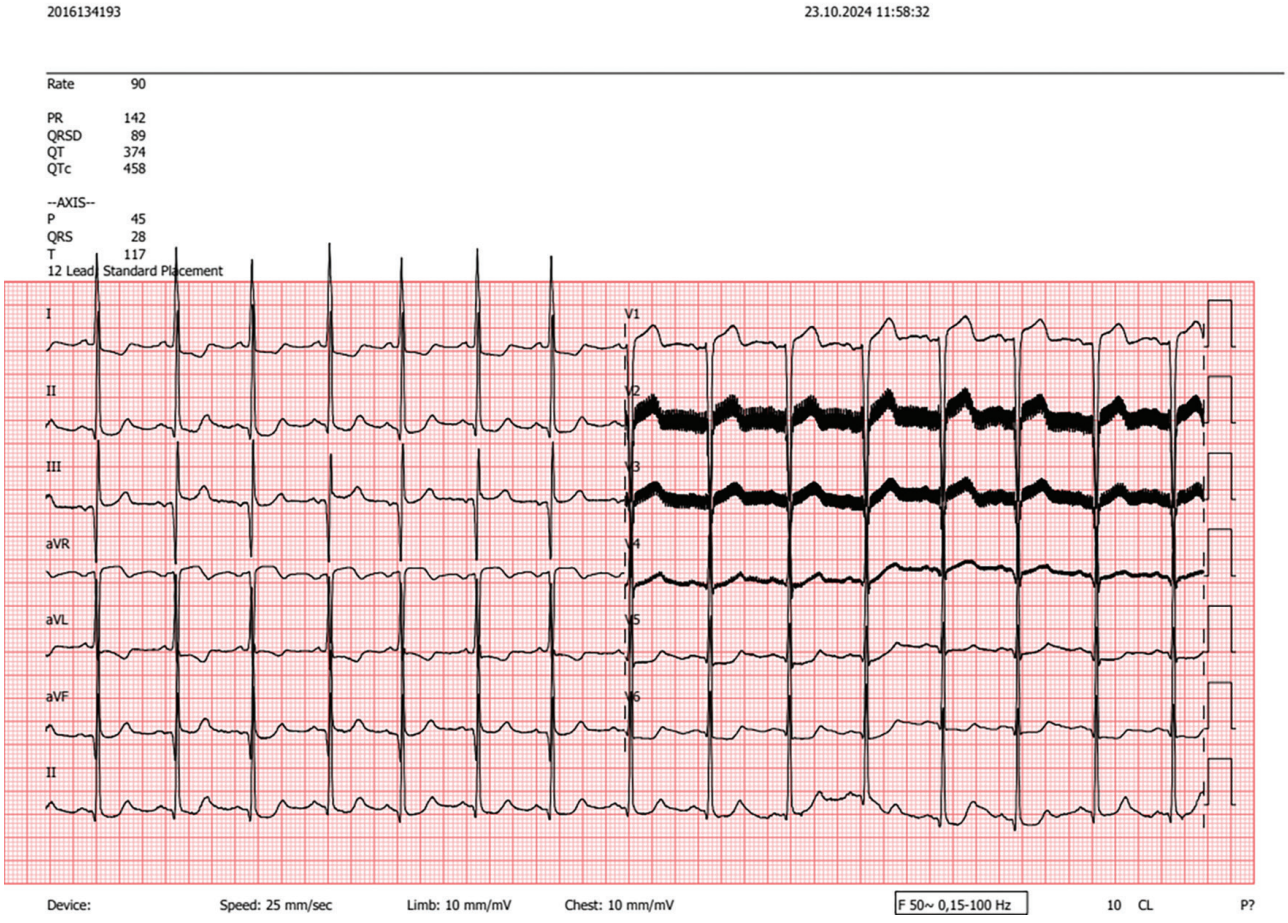

Figure 3. Ischemic ECG changes: The heart axis is slightly shifted to the left. 2 mm depletion is observed in the S-T segment in all versions
ECG: Electrocardiogram

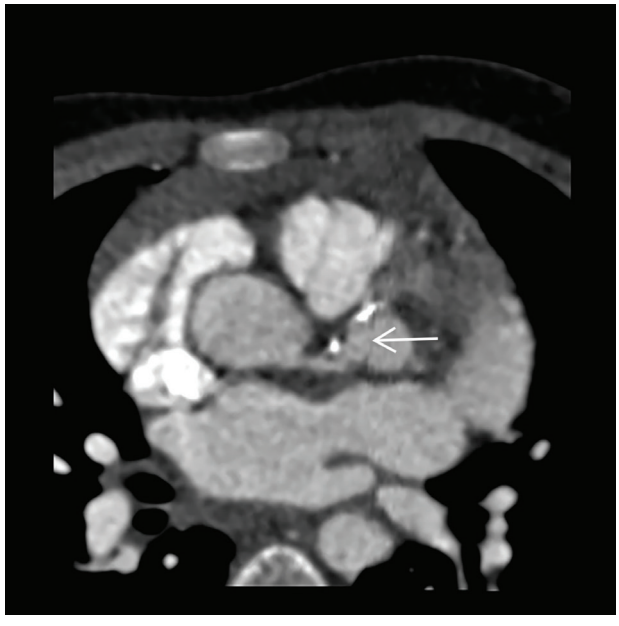

Figure 4. Left coronary aneurysm

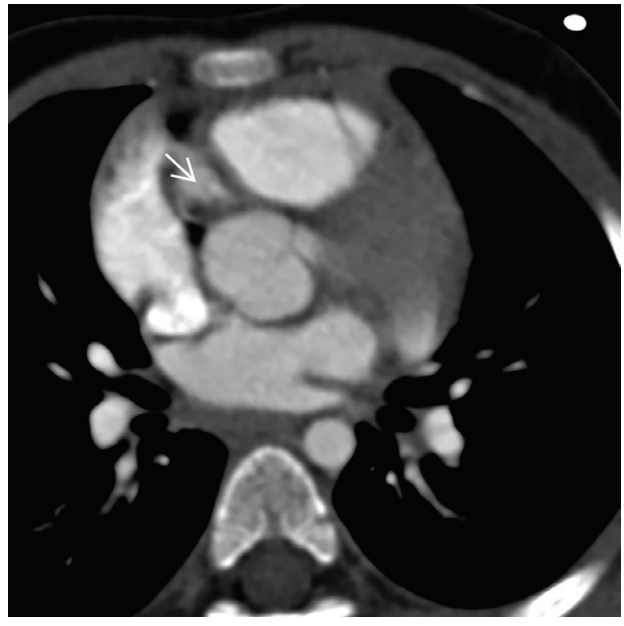

Figure 5. Right coronary aneurysm

In addition, collateral vessels arising from the sinus node branch were seen supplying the proximal LAD. These findings indicated severe multivessel coronary involvement with chronic occlusion and collateral circulation. Due to progressive coronary obstruction and objective evidence of myocardial ischemia, the patient underwent three-vessel coronary artery bypass grafting. Median sternotomy was performed and the left internal mammary artery (LIMA), right internal mammary artery (RIMA), and a saphenous vein graft were prepared. The LIMA was anastomosed to the LAD, the RIMA to the RCA, and a saphenous vein graft to the distal bifid LAD. The cardiopulmonary bypass time was 204 minutes and the aortic cross-clamp time was 104 minutes. The patient was successfully weaned from cardiopulmonary bypass and postoperative hemostasis was achieved without complications. Postoperative follow-up demonstrated improvement in left ventricular systolic function and clinical stabilization (Tables I-III).

Table I. Clinical timeline of illness, laboratory findings, imaging studies, and treatments in the reported patient

Illness Day	Clinical findings	Laboratory	Imaging	Treatment
Day 1	Fever, rash	↑WBC	-	Antibiotics
Day 13	Persistent fever	↑CRP, anemia	-	-
Day 25	KD suspected	Inflammatory markers	ECHO: giant CAA	IVIG + ASA
Follow-up	Ischemia	-	Angiography	CABG

WBC: White blood cell, CRP: C-reactive protein, KD: Kawasaki disease, ECHO: Echocardiography, CAA: Coronary artery aneurysm, IVIG: Intravenous immunoglobulin, ASA: Acetylsalicylic acid, CABG: Coronary artery bypass grafting

Table II. Application of the AHA incomplete Kawasaki disease diagnostic algorithm in the present case

AHA incomplete KD criteria	Findings in the patient
Fever ≥5 days	Fever for 12 days
Elevated CRP or ESR	CRP 138 mg/L, ESR 78 mm/h
Supplemental laboratory criteria	Anemia, thrombocytosis, hypoalbuminemia, sterile pyuria
Echocardiographic findings	Giant coronary aneurysms (RCA 6 mm, LMCA 10 mm)
Final diagnosis	Incomplete Kawasaki disease

AHA: American Heart Association, CRP: C-reactive protein, ESR: Erythrocyte sedimentation rate, RCA: Right coronary artery, LMCA: Left main coronary artery

Table III. Serial coronary artery measurements and imaging findings during follow-up

Time point	Coronary segment	Diameter (mm)	Z-score	Thrombus	Stenosis	Imaging modality
Day 25 (Diagnosis)	RCA	6 mm	+15.6	No	-	Echocardiography
	LMCA	10 mm	+23.0	No	-	Echocardiography
Follow-up	LAD	Aneurysmatic dilation	-	No	-	CT angiography
	Circumflex	-	-	No	80-90%	Coronary angiography
	RCA	-	-	No	100% occlusion	Coronary angiography

RCA: Right coronary artery, LMCA: Left main coronary artery, LAD: Left anterior descending artery, CT: Computed tomography

Discussion

Incomplete Kawasaki disease represents a significant diagnostic challenge, particularly in infants younger than six months, who frequently present without the full spectrum of classical diagnostic criteria (2,3). As a result, delayed diagnosis is common in this age group and may lead to severe coronary artery complications (5,6).

Cerebrospinal fluid pleocytosis has been reported in Kawasaki disease and it is considered a manifestation of Kawasaki-related aseptic meningitis. This finding may lead to an initial misdiagnosis of infectious meningitis, particularly in young infants presenting with fever and meningeal irritation. In such cases, negative bacterial cultures and the presence of systemic inflammatory findings should prompt consideration of Kawasaki disease in the differential diagnosis (2-5).

According to the AHA guidelines, echocardiographic evaluation should be performed in children with five or more days of unexplained fever and suspected Kawasaki disease (4). In infants younger than six months, echocardiography should be considered even more promptly when fever persists for seven days or longer without a clear source, as incomplete presentations are particularly common in this age group (7-9).

Giant coronary artery aneurysms rarely regress and carry a substantial risk of thrombosis, progressive stenosis, and myocardial ischemia. Current recommendations support anticoagulation therapy in those patients with giant aneurysms, with escalation to triple antithrombotic therapy when significant stenosis or thrombosis develops (4,9).

Although systemic arterial aneurysms are recognized complications of Kawasaki disease, particularly in those patients with giant coronary artery aneurysms, alternative etiologies should also be considered in the differential diagnosis. These include systemic vasculitides, monogenic autoinflammatory syndromes, and connective tissue disorders. Careful clinical evaluation and appropriate laboratory investigations are therefore important in order to exclude other causes of systemic aneurysmal disease (8,9).

Progressive coronary artery stenosis, chronic total occlusion, and objective evidence of myocardial ischemia may necessitate surgical revascularization. Coronary artery bypass grafting remains an important treatment option in selected pediatric patients with Kawasaki disease complicated by severe multivessel coronary involvement (8-10).

This case highlights the serious cardiovascular consequences of delayed diagnosis in incomplete Kawasaki disease and underscores the importance of early recognition, prompt echocardiographic evaluation, and close long-term cardiovascular surveillance.

Ethics

Informed Consent: Written informed consent for publication was obtained from the patient's legal guardians.

Footnotes

Authorship Contributions

Surgical and Medical Practices: Ş.Ş.Ö., E.D., B.B.A.A., H.K., B.K.B., O.N.T., Y.A., R.E.L., Concept: Ş.Ş.Ö., E.D., Z.Ü., B.B.A.A., H.K., B.K.B., Y.A., R.E.L., Design: Ş.Ş.Ö., Z.Ü., H.K., O.N.T., Y.A., R.E.L., Data Collection or Processing: Ş.Ş.Ö., Z.Ü., M.Y., B.K.B., R.E.L., Analysis or Interpretation: Ş.Ş.Ö., Z.Ü., M.Y., B.B.A.A., B.K.B., O.N.T., R.E.L., Literature Search: Ş.Ş.Ö., E.D., M.Y., B.B.A.A., H.K., B.K.B., O.N.T., Y.A., R.E.L., Writing: Ş.Ş.Ö., E.D., M.Y., B.B.A., B.K.B., H.K., Y.A., R.E.L.

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