

Giant LV Thrombosis and Thrombosed Coronary Aneurysm in an Infant with Kawasaki Disease

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Abstract

Background

Kawasaki disease (KD) also known as mucocutaneous lymph node syndrome is the febrile and self-limited vasculitis that occurs in children of all ages, especially younger than five years of age. Here, we describe a case of Kawasaki disease with coronary artery aneurysm and a big clot in LV with a diameter of 1.5 cm.

Case Presentation

In February 2019, a 10-month-old boy with a complaint of weakness and lethargy was referred to Imam Reza hospital in Mashhad (Iran). The patient has had coughs with no response to drug treatment for the past four months. Gallop rhythm was present in heart auscultation. Hepatomegaly was observed in physical examination. No lymphadenopathy was seen, but scalded skin was observed. On the second day of hospitalization in our center, he developed non-purulent bilateral conjunctivitis, strawberry tongue, and maculopapular rashes on the body and distal parts of the limbs. Echocardiography showed dilation of the heart chamber, blood clot in the left ventricle (LV) with the size of 1.5 cm, moderate to severe tricuspid regurgitation (TR), giant aneurysm (0.8 cm) in the left anterior descending coronary artery (LAD), right coronary artery (RCA) aneurysm (0.6 cm), and decreased ejection fraction (EF).

Conclusion

Kawasaki disease must be considered in the differential diagnosis of patients presenting symptoms of infection including fever and weakness, especially in infants. Early diagnosis of Kawasaki disease can lower the chance of complication including coronary artery aneurysms. Therefore, this disease must be permanently considered in patients (especially in infants) with long-term fever without a typical presentation of Kawasaki to avoid severe heart complications.

Key Words: Coronary artery aneurysm, Giant thrombosis, Kawasaki disease.

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1- INTRODUCTION

Kawasaki disease (KD) also known as mucocutaneous lymph node syndrome is febrile and self-limited vasculitis that occurs in children of all ages, especially those younger than five years of age (1, 2). KD is slightly more predominant in boys. In addition, its complication and mortality rate is higher in boys compared to girls (3). KD is the leading cause of acquired heart disease in developed countries and rheumatic heart disease in developing countries (4). Approximately 20% of children who do not receive intravenous immunoglobulin (IVIG) in acute phases develop coronary artery aneurysms (5). In this study, we describe a KD case with coronary artery aneurysm (the largest: 0.8 cm) and a big clot in LV with a diameter of 1.5 cm.

2- CASE REPORTS

A 10-month-old boy was referred to Imam Reza hospital in Mashhad, February 2019, with a complaint of weakness and lethargy. He was previously treated with antibiotics (vancomycin and ceftriaxone) due to suspicions of meningoenzephalitis following prolonged fever and drowsiness. He was a late preterm infant born at 36 weeks of gestation, having a successful natural delivery. He has had a healthy growth and development. Although he was 10 months old, he still had not received a pentavalent vaccine, which normally should be administered at six-month-old. He was feeding with cow milk since he was two months old and breastfeeding ended at six-month-old. The patient has had coughs with no response to drug treatment from four months ago. Gallop rhythm was present in heart auscultation. Hepatomegaly was observed in physical examination. No lymphadenopathy was seen, but scalded skin was observed. The boy underwent lumbar puncture, and the findings showed no infection and thus antibiotic administration was stopped.

On the second day of hospitalization in our center, he developed non-purulent bilateral conjunctivitis, strawberry tongue, and maculopapular rashes on the body and distal parts of the limbs. The administration of IVIG was done following the suspicion of Kawasaki disease. The fever stopped on day 5. Following chronic cough, the patient underwent a chest X-ray (CXR). Because the cardiomegaly was diagnosed on CXR (**Figure. 1**), and respiratory distress was detected after blood infusion, we asked for cardiologist consultation. Echocardiography showed dilation of the heart chamber, blood clot in the left ventricle (LV) with a size of 1.5 cm, moderate to severe tricuspid regurgitation (TR), giant aneurysm (0.8 cm) in the left anterior descending coronary artery (LAD), right coronary artery (RCA) aneurysm (0.6 cm), and decreased ejection fraction (EF) (**Figures. 2 and 3**).

On day 7 of hospitalization, he received a single dose of 25 gr intravenous immunoglobulin (IVIG) as suspicious of Kawasaki disease. The results of the laboratory investigation are presented in **Table 1**. ECG showed flat T waves in the v1 lead and inverted T waves in the aVL lead that may be attributed to the ischemia (**Figure. 4**). The patient received heparin infusion at the rate of 170 U/kg/h, Apowarfarin (1 mg every day (QD)), Lasix (20 mg for a 24-h infusion), Milrinone (8 mg for 24 hours' infusion), Aldactone (25 mg twice a day (BID)), Inderal (10 mg QD), ASA (50 mg QD), and Captopril (25 mg BID). After 11 days, heparin was replaced with subcutaneous enoxaparin 10 mg BID. As the international normalized ratio (INR) reached two, the enoxaparin administration stopped. Serial echocardiography was performed to evaluate the patient's condition. The last echocardiography showed ejection fraction was improved, the size of the right coronary artery (RCA) decreased, and a

blood clot in the left ventricle (LV) shrank to 1 cm. As soon as the situation was stabilized, the patient was discharged on Apo-warfarin (2 mg BID), aspirin (100 mg

QD), spironolactone (12.5 mg BD), captopril (3 mg BID), furosemide (5 mg BID), and propranolol (10 mg QD).

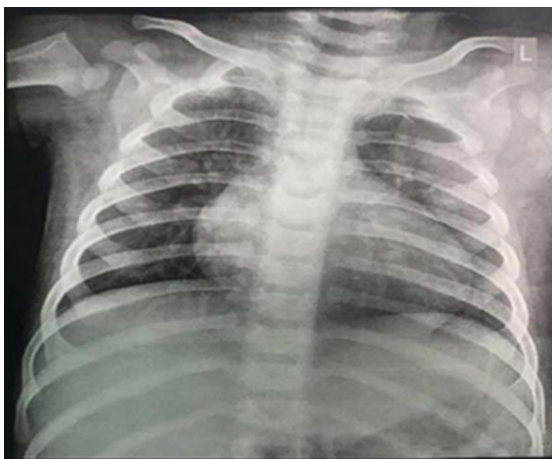


Fig.1. CXR showing cardiomegaly in the thorax. CXR: Chest x-ray.

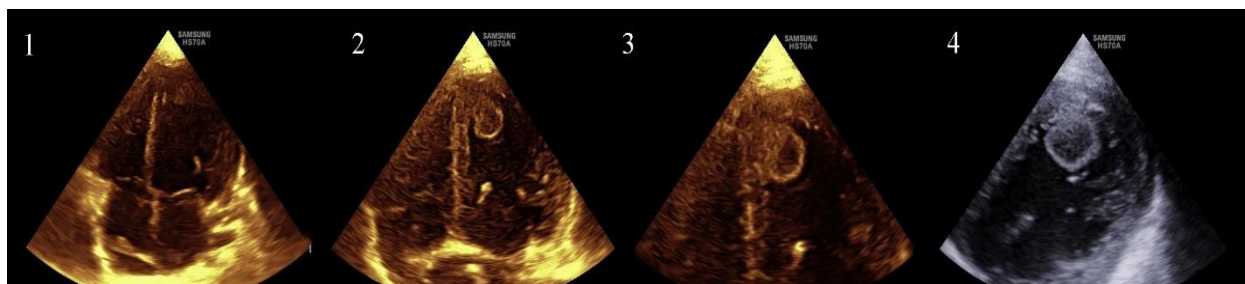


Fig.2. Echocardiography showing LV Enlargement and large Apical LV mass with central hypodensity (most probably, clot). LV: left ventricle.

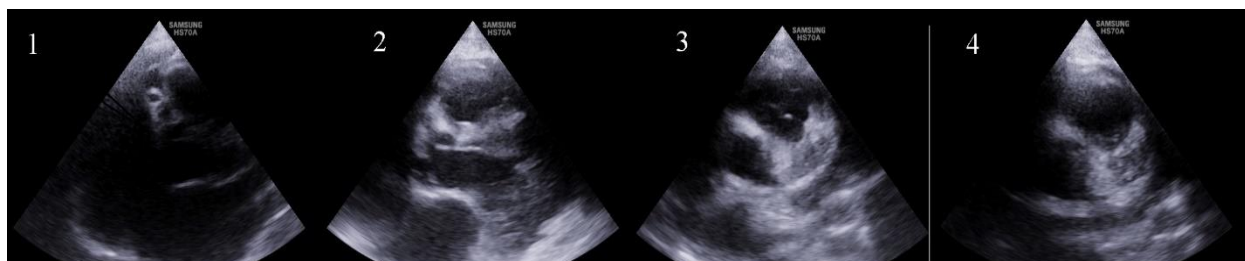
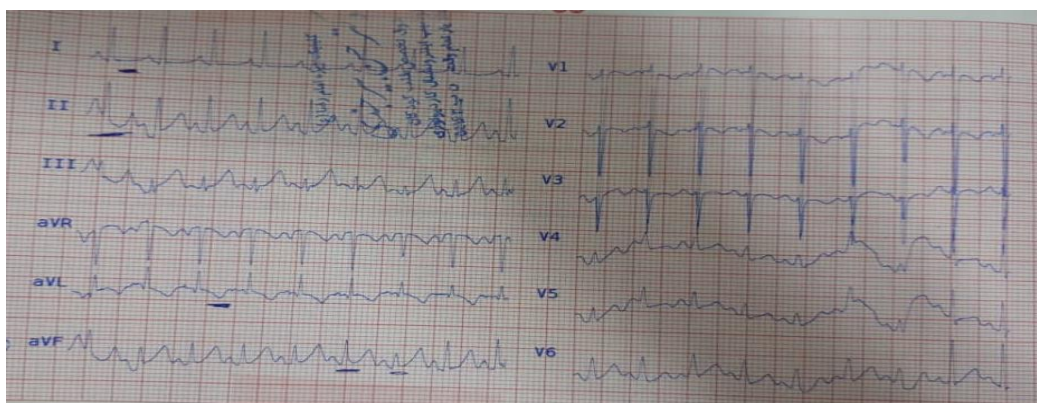


Fig.3. Echocardiography showing coronary dilation and large multiple aneurysms in LMCA, LAD, and RCA in addition to large clots in LMCA and LAD. LMCA: left main coronary artery, LAD: left anterior descending artery.

Table-1: Series of the patient's laboratory data at the time of disease onset, second, third and fourth weeks after admission.

Laboratory test	At the time of disease onset	2 weeks after disease onset	3 weeks after disease onset	4 weeks after disease onset
ESR (mm/h)	80	84	31	194
CRP (mg/l)	12	63	-	20
UA	Pyuria	-	normal	-
WBC (cells/ μ m)	15000	19000	15600	
PLT (cells/ μ m)	302000	646000	450000	
Hb (g/dl)	8.2	7.2	6	10.2
Albumin (g/dl)	-	-	3	0.4
Troponin I (ng/ml)	-	-	-	327.6
CPK MB (ng/ml)	-	-	-	19
AST (unit/ml)	-	-	-	213
ALT (unit/ml)	-	-	-	33
ALP (unit/L)	-	-	-	1303

ESR (erythrocyte sedimentation rate), CRP (C-reactive protein), UA (urine analysis), WBC (white blood cells), PLT (platelets), Hb (hemoglobin), CPK-MB (creatin kinase-MB), AST (aspartate aminotransferase), ALT (alanine aminotransferase), ALP (alkaline phosphatase).

**Fig. 4.** ECG showing flat T wave in v1 lead and invert T in aVL lead, which are suggestive of myocardial infarction.

3- DISCUSSION

In the present study, we reported a case of Kawasaki disease with coronary artery aneurysm and a big clot in LV with a size of 1.5 cm. According to the American heart association (AHA), the diagnosis of Kawasaki disease is based on clinical findings. AHA has defined classic KD as the presence of fever for at least five days in addition to the following 4 of 5 clinical criteria: 1) Erythematous oral mucosa, cracking lips, or strawberry tongue, 2) Non-purulent bilateral bulbar conjunctival injection, 3) Rashes, 4) Edema and erythema of the hands and feet, and 5) Cervical lymphadenopathy. Our case of KD represented non-purulent bilateral conjunctivitis, strawberry tongue, and

maculopapular rashes on the body and distal of the limbs, and thus he only met 3 of 5 clinical criteria. AHA has defined this condition as an incomplete KD. In such cases, there is high clinical suspicion to KD; however, the diagnosis of KD is confirmed by the presence of coronary artery abnormalities (CAA) (4). As we performed echocardiography, not only aneurysm was found in LAD and RCA, but also a big clot with a diameter of 1.5 cm was detected in LV. The presence of blood clots in the artery was previously described in several cases of KD (6-10). However, a big blood clot in the heart chamber is a rare condition. CAAs were observed on 53.1% of all KD patients (7). The most commonly involved arteries are

LAD and RCA. These developing aneurysms in coronary arteries, which are the desired environment for thrombus formation, can lead to thrombocytopenia in the acute phase of KD. Nevertheless, thrombocytopenia changes to thrombocytosis in the subacute phase (5). Studies showed a single high dose of IVIG and ASA administration during the first 10 days from the fever onset could reduce the risk of coronary artery aneurysm from 25% to 5% (5). This happens through the cessation of systemic inflammation by modulation of regulatory T cells and platelets (1, 5). However, recent findings questioned the safety of ASA. Although ASA can develop slow gastrointestinal absorption, hypoalbuminemia, and increase renal clearance (3), these side effects did not make the AHA put ASA aside from the treatment guideline of KD (4). Laboratory investigation is presented in **Table 1**. Erythrocyte sedimentation rate (ESR) raised in the fourth week of disease onset as the patient received incomplete treatment in the previous center before being referred. Therapeutic management in Imam Reza hospital for this Resistant-Kawasaki began the week 4 after the disease onset.

4- CONCLUSION

KD should be considered in the differential diagnosis of patients presenting symptoms of infection including fever and weakness, especially in infants. Early diagnosis of Kawasaki disease can lower the chance of complication including coronary artery aneurysms. Therefore, we should always consider Kawasaki disease in patients with long-term fever, without a typical presentation of Kawasaki to avoid severe heart complications.

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6- CONFLICT OF INTEREST: None.

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