

Kawasaki disease complicated by peripheral gangrene and a ventricular septal defect: An unusual association

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Kawasaki disease is an acute febrile illness associated with the vasculitis of small and medium size arteries. Peripheral gangrene is very rare but a known complication of Kawasaki disease mostly reported in the infantile age group. Early therapy is advocated for prevention of serious complications. We report a 10-month-old girl with Kawasaki disease and a small ventricular septal defect who developed peripheral gangrene despite initiation of early therapy.

Key words: infant, complications, Kawasaki disease, peripheral gangrene.

Kawasaki disease (KD) is an acute illness of unknown cause that chiefly affects infants and children and is characterized especially by fever, rash, conjunctivitis, inflammation of lips and tongue, and unilateral swollen lymph nodes of the neck and is the leading cause of acquired heart disease.¹⁻³ KD is associated with systemic vasculitis particularly affecting the coronary arteries, causing coronary artery aneurysms (CAA) in 15–25% of untreated patients while 2–3% of untreated cases die as a result of coronary vasculitis.⁴⁻⁶ Peripheral gangrene is a known but rare complication of KD that occurs almost exclusively in infants.⁷⁻¹⁵ Treatment with intravenous immunoglobulin (IVIG) before day 10 of illness has been shown to reduce the incidence of coronary artery aneurysm.⁴ It is speculated that early treatment might also reduce the incidence of peripheral ischemia and gangrene.¹⁰

In this report, we describe a recent case of infantile KD and ventricle septal defect associated with peripheral gangrene, in which the infant had been treated with IVIG at the earliest date of disease onset but still developed peripheral gangrene. This is the first such case to be reported from Pakistan.

Case Report

A 10-month-old girl presented to the emergency

department with history of fever for 5 days associated with a maculopapular rash, irritability, strawberry tongue, conjunctivitis and edema of the hands and feet. The fever was high grade, intermittent and associated with irritability. Four days later, the child developed a maculopapular rash which was generalized. It was associated with redness of the oral cavity and conjunctivitis. On examination, the child had a length of 75 cm and weight of 8.5 kg. She was irritable, febrile (101 F) with a heart rate of 120/min and blood pressure of 96/53 mm hg while maintaining oxygen saturation at room air. Generalized maculopapular rash was present with pallor and bilateral pedal and dorsal edema. The patient had a strawberry tongue and bilateral non-purulent conjunctivitis. Systemic examination did not reveal any other positive signs. The laboratory examinations were as follows: Hb 8.1 g/dl; WBC 32,100/mm³; platelet 2,91,000/mm³; ESR 81 mm/h; CRP 19.5 mg/dl; Albumin 3 g/dl and SGPT 70 IU/L. The child was admitted to the pediatric intensive care unit (PICU) and broad spectrum antibiotics were started. Echocardiogram showed a small 2 mm perimembranous ventricular septal defect (VSD) covered with tricuspid aneurysmal tissue and normal right coronary artery measuring 1.8 mm at origin with Z score value of +0.76 and a dilated left main coronary artery measuring

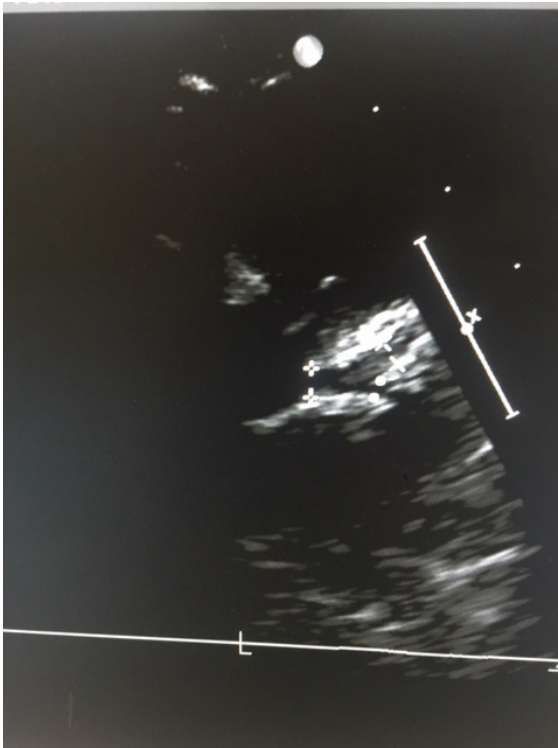


Fig. 1. Left coronary artery dilatation

3.0 mm at ostium with Z score value of +2.61 and 2.8 mm at bifurcation, LAD measuring 2.3-2.4 mm with Z score value of +3.02 (Fig. 1). No convincing coronary artery aneurysm was found. Under a diagnosis of KD, high-dose IVIG (2 g/kg, continuous intravenous infusion for 12 hours) and oral aspirin (100 mg/kg/day) were administered. On the 7th day of illness the child developed discoloration of left big toe (Fig. 2). Ultrasound Doppler showed normal flow in the left dorsalis pedis and posterior tibial arteries. Anticoagulation with low molecular weight heparin and local vasodilation with topical nitroglycerine paste was started after consultation with vascular surgery and monitoring for extension of gangrene was continued. On day 10 of the illness, it was well demarcated and only confined to the left big toe. Repeat echocardiogram on 14th day of illness ruled out coronary artery aneurysm. Fever subsides after the 1st dose of IVIG and further evaluation revealed a decreased ESR, CRP, total WBC and increasing platelet count. The patient was discharged home on low molecular weight heparin and aspirin in a stable condition.

During the course of illness she continued to



Fig. 2. Gangrene of left big toe

develop other features of peripheral vasculitis such as changes of the skin, changes on the BCG scar (Fig. 3) and changes on the eyelids (Fig. 4).

A written informed consent was taken from parents before submission of this case report.

Discussion

Kawasaki disease (KD) is an acute self-limiting inflammatory disorder, associated with vasculitis, affecting predominantly small to medium-sized arteries, particularly the coronary arteries. Very few cases of KD with peripheral gangrene and auto amputation have been reported in the literature.

Although the pathogenesis of this complication is not well understood, it likely includes some combination of local peripheral arteritis, arteriospasm, thrombosis peripherally and/or more proximally (e.g., in an axillary artery aneurysm), and cardiogenic shock.¹⁶ The previous cases of KD complicated with peripheral gangrene were infants except one adult patient.¹⁷ Our patient was also an infant. The possible explanation of peripheral gangrene with KD in the infantile age group



Fig. 3. Changes in BCG scar

is delayed diagnosis and treatment due to atypical presentation in this age group, but in our patient treatment was initiated early. To our knowledge the association of VSD with KD complicated by peripheral gangrene has never been reported in the literature.

Currently, there is no consensus regarding the ideal therapy for peripheral ischemia in the aforementioned case reports. There is a wide range of different medications which are used in the management of KD with gangrene. Some of the treatment agents that have been used are heparin, steroids, hydralazine, propranolol, warfarin, urokinase, Prostaglandin E1, dipyridamole, nitroprusside, tissue plasminogen activator, prostacyclin, glyceryl trinitrate, nifedipin, and even caudal block.⁸ However, von Planta et al.¹⁷ cited the concern of stealing perfusion from potentially compromised coronary arteries as their patient developed a myocardial infarction shortly



Fig. 4. Changes of eyelids

after the initiation of prostaglandin infusion. In our case, it appeared that IVIG and high dose aspirin were helpful in controlling the inflammation, as fever resolved soon after initiation of these therapies along with low molecular weight heparin for peripheral gangrene. The risk/benefit with prostaglandin infusion needs to be further evaluated.

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