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SHORT REPORT

Shock: an unusual presentation of Kawasaki disease

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Abstract Kawasaki disease (KD) is a common acute systemic vasculitis of childhood. Although KD has wide spectrum of clinical features, shock is not one of its common presentation form. We describe a 5-month-old female infant with severe shock syndrome requiring fluid resuscitation, inotropic support, and PICU admission. She was diagnosed retrospectively to have KD complicated by coronary artery aneurysms in spite of receiving early course of IV immunoglobulin. Conclusion: Diagnosis of KD could be missed in the pediatric intensive care unit because of its atypical presentation and the wide array of associated clinical symptoms. Subsequently, intensivists and emergency room physicians should maintain a high index of suspicion not to miss it or diagnose it at an advanced stage of the illness.

Keywords Kawasaki disease · Shock · Inotropes · Echocardiography · Coronary arteries aneurysm · Immunoglobulin

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Introduction

Kawasaki disease (KD) is an acute self-limiting vasculitis of childhood that may be complicated by coronary artery aneurysm in up to 25% of untreated patients [5]. Although there are established clinical criteria to diagnose KD, the diagnosis may be delayed due to the lack of clinical signs [4, 8]. The literature points out that the occurrence of atypical/incomplete presentations of KD is rising [1]. Hemodynamic instability and shock as the main presentation of KD is uncommon. In this report we describe an infant with shock as the initial presentation of KD and we review the literature, to increase the intensivists' and emergency room physicians' awareness of this uncommon cause of shock.

Case report

A 5-month-old previously healthy girl presented with history of high-grade fever for 3 days, decreased activity, and poor oral intake. There was no history of vomiting, diarrhea, or cough. She received 2 days course of cefuroxime without improvement.

On examination, the child was febrile (39.2°C), tachypneic, tachycardic (heart rate: 180/min), O_2 saturation was 95% in room air, BP of 80/33 mm/Hg with delayed capillary refill >5 s. She was irritable and oliguric.

Her arterial blood gas showed metabolic acidosis (pH 7.38; PCO_2, 23.9 torr, HCO_3, 16.7 meq/l; BE, −10.3; PO_2, 90 torr), glucose was 3.6 mmol/l. WBC count was 7,200/mm^3 (neutrophiles, 6,000/mm^3) with a normal platelet count of 277,000/mm^3. Her electrolytes and renal function were normal.

The patient required fluid resuscitation with normal saline up to 70 ml/kg and was started on vancomycin, ceftriaxone, and oseltamivir phosphate then she was transferred to pediatric intensive care unit (PICU).

In PICU, her vital signs were: BP, 70/35 to 80/40 mm/Hg; capillary refill>4 s; heart rate, 160 to 180/min; respiratory rate, 50–60/min; O_2 saturation, 99% on 1 L O_2 by nasal cannula; and the urine output was 2 ml/kg/h. Her extremities were cool and the peripheral pulses were weak and thready.

She had generalized red skin and lips, an injected throat, bilateral non-suppurative conjunctivitis, and BCGitis (inflammation and discharge from the BCG vaccine site). The red skin was thought to be a red man syndrome secondary to vancomycin infusion. No lymph node could be palpated. Chest, heart auscultation, and abdomen examination were unremarkable. Her neurological examination showed irritability. A repeated blood gas showed: pH 7.27; PCO_2, 28.9 torr; PO_2, 76 torr; HCO_3, 14.7 meq/L; and BE, −12.4. The glucose level was 7.6 mmol/l and the lactate was 2.2 mmol/l. Urine dipstick showed leukocyte 2+ with negative nitrite. Her CBC showed: WBC count increased to 21,800/mm^3 (neutrophiles, 19,000/mm^3), hemoglobin of 9.5 g/dl, and platelet count of 151,000/mm^3. The ESR was 42 mm at end of 1 h and the C-reactive protein (CRP) was high (184 mg/l). Urea was 9.6 mmol/l and the creatinine was 59 μmol/l. The coagulation profile showed: PT, 10.6 (control, 8.5); PTT, 63 (control, 32); INR, 1.3; and D-dimer, 3,930 μg/l. LFT showed AST, 77 U/l; ALT, 54 U/l; total bilirubin, 69 μmol/l (direct, 59 μmol/l) and albumin, 26 g/l.

The patient was started on dopamine which was titrated up to 20 mcg/kg/min and norepinephrine up to 0.5 mcg/kg/min. She also received hydrocortisone 1 mg/Kg/6 h for septic shock and started electively on mechanical ventilation. Antibiotics were changed to meropenem, flucloxacillin, and gentamycin.

The possibility of KD was raised and an echocardiography done on the day of admission, showed normal heart function, significant reversal flow in the descendant aorta (due to the severity of hemodynamic instability of the patient), and normal coronary artery diameters. Despite the incomplete KD features and the absence of coronary artery dilatation, Immunoglobulin infusion (1 g/kg for 2 days IV) was given.

The general condition and hemodynamic state of the patient improved gradually over the following 24–72 h. The metabolic acidosis resolved, with normalization of renal function, coagulation profile, and CBC together with improvement of her inflammatory markers (Table 1). All cultures and viral screening were negative. The vasopressor were discontinued after 5 days and she was successfully extubated. She remained afebrile during all her stay in PICU but she was noticed to be very irritable. Brain MRI and EEG were normal. Her skin redness improved but the conjunctivitis and the red lips remained almost the same with peeling of the extremities. Repeated echocardiography on day 4 of admission showed normal function, resolved reversal flow, but the coronary arteries were not checked. Patient was

Table 1 Main laboratory result during the first 7 days of PICU admission

	Day 0	Day 1	Day 2	Day 3	Day 4	Day 5	Day 6	Day 7
WBC (5–20/10^9 L)	21.8	39.7	54	42	26.6	21	20	19.4
Neutrophils (1–8.5·10^9/L)	19	38	47		22	15	16	
Lymphocytes (3.7–11·10^9/L)	1.6	1.9	1		2.9	3.8	4.6	
Hb (10–18 g/dL)	9.5	8.8	10.3	10.1	8.4	8.1	7.2	6.9
Platelet (150–450·10^9/L)	151	118	83	105	71	83	169	217
ESR (1–15 mm/h)	42							
CRP (<20)	184		178	160	90			65
Urea (2–7.5 mmol/L)	9.6	8.6	6.3	4.8	3.9	2.1	1.5	1.5
Creatinine (50–115 μmol/L)	59	54	45	38	29	27	23	29
PT (12–15 s)	10.6	9.8	9.6	9.5	10	9.9	10	10
PTT (32–43 s)	63	59	47	43	46	45	44	40
INR	1.3	1.2	1.1	1.1	1.1	1.1	1.1	1.2
D-dimer (<192 μg/L)			3,930					
AST (2–37 U/L)	77				66			
ALT (2–40 U/L)	54				47			
Gamma GT (7–32 U/L)					29			
Bilirubin total (2–22 μmol/L)	69		55		50	30		
Bilirubin direct (0–6 μmol/L)	59		50		49			
Albumin (36–51 g/L)	26	25	26	22	20	21	22	24

discharged to the ward on day 7 of admission. She was noticed to be afebrile, active, and in good condition, but CRP remained high (65 mg/l). Repeated echocardiography on day 16 of admission showed giant coronary aneurysms (>8 mm) of the right coronary artery and left anterior descending coronary artery. The child was started on low-molecular-weighted-heparin, and given a second course of IV immunoglobulin (IVIG); KD was considered as IVIG-resistant form. However her CRP remained high and did not improve after a pulse course of steroids. Infliximab was finally started which resulted in normalization of CRP.

Discussion

The salient features of this case were not only that not all diagnostic criteria of KD were fulfilled, which is known in the incomplete form of KD, but moreover that the main presentation was the presence of hypotensive shock. KD in our patient was an IVIG-resistant form complicated by coronary artery aneurysms.

Occasional patients with KD may present with severe manifestations leading to PICU admission. These include altered mental status, ischemic colitis secondary to mesenteric vasculitis [7], heart failure secondary to myocarditis [10], acute respiratory distress syndrome, and renal failure [6]. Recently, two case-controlled studies described patients with KD admitted to the PICU with hypotensive shock [2, 3]. Our patient and the series reported by Dominguez et al. [2] were fulfilling the criteria of KD shock syndrome (KDSS) established by Kanegaye et al. [3]. These criteria are an association of KD features with sustained decrease in systolic blood pressure from baseline of ≥20%, or clinical signs of poor perfusion.

Data from these reports describe the following common characteristics among patients with KDSS compared with patients with KD who were not admitted to the PICU: patients with KDSS were more likely to be female, and to have laboratory finding consistent with more inflammation, manifested by significantly more bands, lower platelet count, a higher CRP level, and a lower serum albumin level. These patients were more likely to have IVIG resistance and to require a second dose of IVIG or a second-line therapy (steroids or infliximab) and higher rate of coronary artery dilatation and aneurysm formation.

Features of KDSS can be strongly suggestive of toxic shock syndrome, in this context; echocardiography to search for coronary artery dilatation is of paramount importance to adjust the diagnosis and to start the appropriate treatment [9].

Questions remain concerning the precise mechanisms of hypotension in KD. It is most likely multifactorial. The high inflammatory markers and the consumptive coagulopathy suggest more intense vasculitis with capillary leak, cytokines dysregulation, and myocardial dysfunction with impaired systolic and diastolic function. In the study of Kanegaye et al., 31% of the patients with KDSS had low ejection fraction witch resolved promptly with therapy but abnormal ventricular diastolic function persisted during the follow-up. Our patient's heart function was normal indicating that the hypotension was mainly related to altered vasomotor tone and reduced vascular resistance. The majority of patients in the two studies met clinical criteria for KD; despite that KD was not the leading diagnosis in these patients, most likely because of the failure to consider KD in patients who present with hypotension requiring inotropic support and ICU admission.

Conclusion

This case report highlight that KD may present with severe shock, emphasizing the need for physicians to remain aware of the full range of clinical symptoms associated with the disease.

Statement This study is exempt from approval by our local ethics committee.

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