

Cardiogenic shock: do not forget the possibility of Kawasaki disease

Wei Wang, Wei-Hua Zhu, Yan-Qi Qi, Song-Ling Fu, Fang-Qi Gong

Department of Cardiology, Children's Hospital, Zhejiang University School of Medicine, Hangzhou, People's Republic of China

SUMMARY: Wang W, Zhu W-H, Qi Y-Q, Fu S-L, Gong F-Q. Cardiogenic shock: do not forget the possibility of Kawasaki disease. *Turk J Pediatr* 2012; 54: 86-89.

We report the case of a six-year-old boy who presented with cardiogenic shock due to Kawasaki disease (KD). He was misdiagnosed at first as septic shock. After careful examination, he was diagnosed as KD complicated with acute coronary syndrome, which leads to cardiogenic shock. Cardiogenic shock is often neglected as a complication of KD, and it tends to be misdiagnosed. We hereby call attention to KD, in some cases of which, it can lead to acute coronary syndrome in the acute phase.

Key words: cardiogenic shock, Kawasaki disease, acute coronary syndrome.

Kawasaki disease (KD) is a generalized vasculitis of unknown etiology that occurs predominantly in infants and young children. Although the cardiac complications of KD are well known, hemodynamic instability is unusual in the acute phase of the illness, except as an incipient complication. Patients tend to be misdiagnosed, leading to a delay in treatment with intravenous immunoglobulin (IVIG). We report such a patient, who suffered cardiogenic shock complicated with KD. He was treated with IVIG and high-dose glucocorticoid and suffered coronary artery aneurysm for an extended period.

Case Report

A six-year-old boy was referred to us with fever and abdominal pain lasting for six days, with dyspnea for three days. Extraordinary hypotension was noted on the physical examination, and his blood pressure was 65/27 mmHg on admission. He was diagnosed with shock, though on progressive examination, he was found afebrile with a heart rate of 122/min and respiratory rate of 52/min with respiratory distress. A systemic examination revealed muffling of heart sounds with no heart murmur. His liver was enlarged, while his spleen was not palpable. The chest film in the anteroposterior projection showed pulmonary edema, cardiac shadow enlargement and little amount of

pleural effusion. The cardiothoracic ratio was 0.55 (Fig. 1). Based on those conditions, his preliminary diagnosis was septic shock, and antibiotic was given to ward off infection. Large-volume resuscitation (injections of physiological saline at a total dose of 40 ml/kg) was also used. However, his situation did not improve after treatment, his blood pressure did not increase, and his liver augmented incessantly, so further examinations were made. Electrocardiography manifested extensive low voltage and ST-T segment depression. Enlarged left atrium and left ventricle were found through echocardiography, with decreased ejection fraction (EF) and left ventricular fractional shortening (LVFS) (Fig. 1, Table I). At the same time, it was also noticed that his heart sound was lower. He was therefore diagnosed as viral myocarditis complicated with cardiogenic shock, and was treated with Cedilanid, high-dose methylprednisolone (20 mg/kg/d) and oxygen supplementation. After his disease stabilized, a more detailed physical examination was conducted. He presented the following symptoms: subcutaneous edema of the hands, bilateral conjunctival hyperemia, bilateral cervical lymphadenopathy, and a strawberry-like tongue. His laboratory data on admission were as follows: white blood cell (WBC) of $17 \times 10^9/L$; platelet (PLT) of $620 \times 10^9/L$; C-reactive protein (CRP) concentration of 108

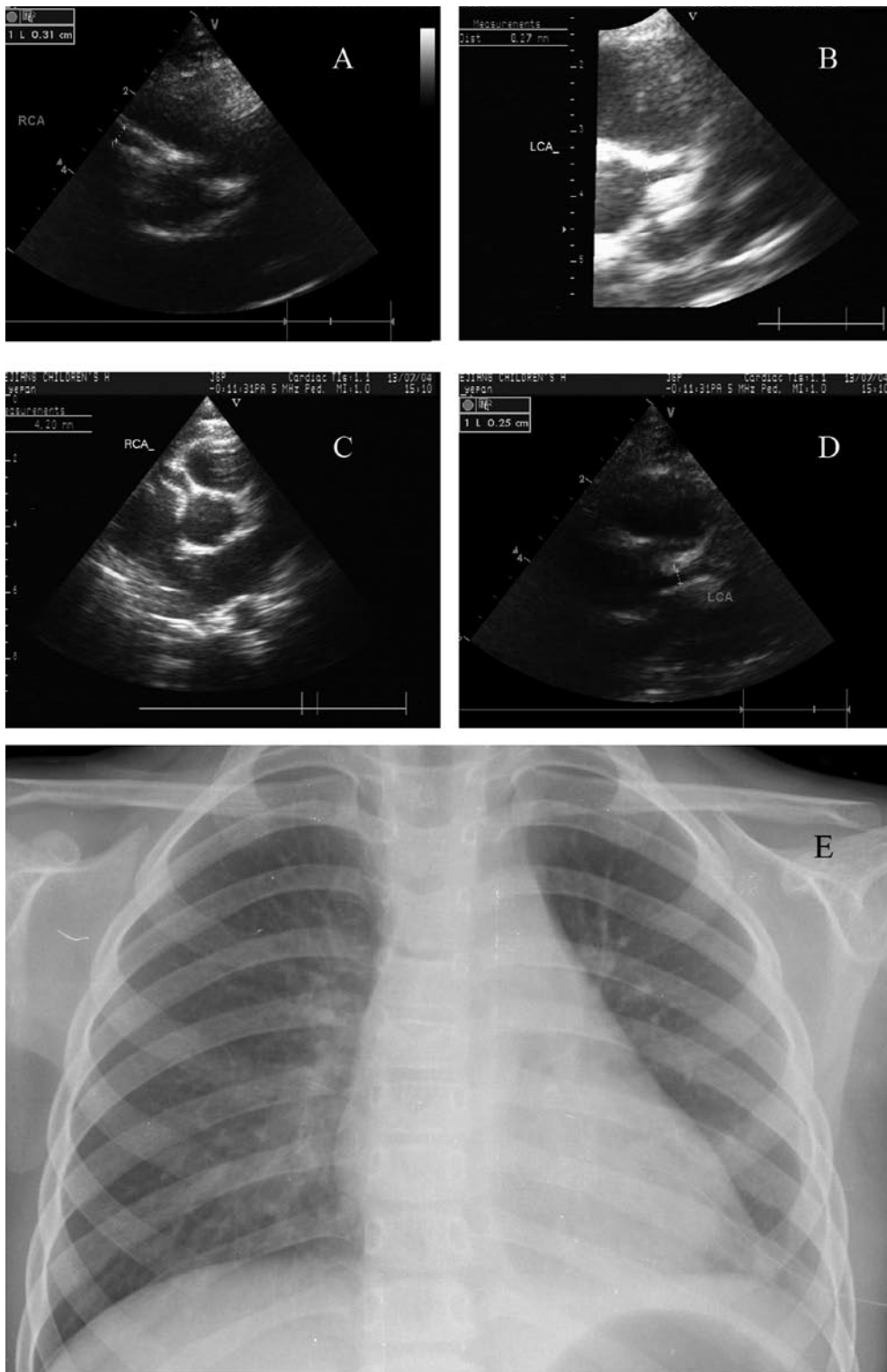


Figure 1. The echocardiography and X-ray of the patient. (A, B): Echocardiography demonstrated the right coronary artery (RCA) with some dilation (the extremity diameter was 3.1 mm at the widest point); the left coronary artery (LCA) was normal. (C, D): Echocardiography demonstrated development of the right coronary aneurysm (the extremity diameter was 4.2 mm at the widest point), while the LCA was still normal. (E): X-ray demonstrated pulmonary edema, cardiac shadow enlargement and little amount of pleural effusion. The cardiothoracic ratio was 0.55.

mg/dl; erythrocyte sedimentation rate (ESR) of 53 mm/h; cardiac troponin T (cTnT) of 0.49 mg/ml; and fibrin degradation product (FDP) level of 809 μ g/L. He was diagnosed as KD complicated with cardiogenic shock, and underwent IVIG (1 g/kg/d \times 2d) and oral administration of aspirin (30 mg/kg/day). The patient was discharged 18 days later. Physical examination results appeared normal at that time, as did the electrocardiography result. However, echocardiograms demonstrated further distention of the right coronary artery (RCA) (from 2.8 mm to 4.2 mm at the widest point) and normal left coronary artery (LCA) (Fig. 1, Table I). Following treatment with aspirin and dipyridamole, repeat echocardiograms demonstrated no further coronary artery aneurysm after six months. His status is good after follow-up for five years until the present.

Discussion

Cardiogenic shock is defined as persistent hypotension and tissue hypoperfusion due to cardiac dysfunction in the presence of adequate intravascular volume and left ventricular filling pressure. Clinical signs include hypotension, tachycardia, oliguria, cool extremities, and altered mental states¹. The cause of cardiogenic shock is manifold, including acute myocardial infarction, myocarditis, left ventricular outflow tract obstruction, and other conditions.

Kawasaki disease (KD) is a rare cause of cardiogenic shock. The cause of cardiogenic shock in KD could be arrhythmia, ischemic heart disease, valvular disease, endocarditis, and myocarditis². As is well known, KD is a vasculitis of medium-sized vessels and probably the vasculitis best known by cardiologists. We propose that acute coronary syndrome caused by cardiac vasculitis is the principal reason for cardiogenic shock in KD. Acute coronary syndrome is a term used for any condition brought on by sudden, reduced blood flow to

the heart. It is usually caused by two diseases involving the coronary arteries: myocardial infarction or unstable angina³. It is also possibly caused by KD in young adults, particularly when aneurysms are found⁴. In our case, the patient's cardiac function was decreased with RCA dilation, while cTnT was normal. Consequently, acute coronary syndromes might be the real reason for his cardiogenic shock.

Thus far, there is no diagnostic test or pathognomonic clinical feature of KD, and a KD patient with a manifestation of cardiogenic shock as the first or cardinal symptom is rare⁵. Thus, patients with the appearance of shock tend to be misdiagnosed, and are often mistaken for having septic shock or cardiogenic shock caused by viral myocarditis, leading to a delay in treatment with IVIG⁷. As acute coronary syndrome is the most common cause of cardiogenic shock complicated with KD, early use of IVIG treatment seems especially important. We report such a case, and the patient was admitted for the cause of cardiogenic shock. We misdiagnosed him at first as septic shock or viral myocarditis, and he was on the mend after use of Cedilanid and methylprednisolone, but his IVIG treatment was delayed. Those patients admitted for cardiogenic shock, just like our patient, are often misdiagnosed, leading to a delay in treatment with IVIG. On the other hand, high-dose glucocorticoid is an important treatment in patients with cardiogenic shock caused by viral myocarditis, and the proper use of glucocorticoid may be helpful since it is useful for the treatment of vasculitis, but it also has the potential risk of coronary artery aneurysm⁷. Therefore, the use of glucocorticoid should be carefully weighed.

In conclusion, cardiogenic shock is often neglected as a complication of KD, and it tends to be misdiagnosed. When a patient exhibits symptoms such as high fever and shock, and the therapy with antibiotic and large-volume

Table I. Cardiac Function and Width of Coronary Artery at Different Stages

Date	AAO (mm)	LA (mm)	LV (mm)	RCA (mm)	LCA (mm)	EF %	FS %	BP mmHg
27/6	2.18	30	45	2.8	2.7	0.50	0.23	65/27
3/7	1.9	22	42	3.1	2.7	0.59	0.31	105/60
13/7	2.1	22	40	4.2	2.5	0.63	0.33	108/65

AAO: Ascending aorta. LA: Left atrium. LV: Left ventricle. RCA: Right coronary artery. LCA: Left coronary artery. EF: Ejection fraction. FS: Fractional shortening. BP: Blood pressure.

resuscitation is ineffective, the possibility of cardiogenic shock complicated with KD should be considered. The early diagnosis is essential, because correct use of IVIG is most important to the prognosis. The use of glucocorticoid requires great consideration.

REFERENCES

1. Topalian S, Ginsberg F, Parrillo JE. Cardiogenic shock. *Crit Care Med* 2008; 36: 66-74.
2. Kanegaye JT, Wilder MS, Molkara D, et al. Recognition of a KD shock syndrome. *Pediatrics* 2009; 123: 783-789.
3. DeVon HA, Ryan CJ, Ochs AL, Shapiro M. Symptoms across the continuum of acute coronary syndromes: differences between women and men. *Am J Crit Care* 2008; 17: 14-24.
4. Knockaert DC. Cardiac involvement in systemic inflammatory diseases. *Eur Heart J* 2007; 28: 1797-1804.
5. Wilson N, Heaton P, Calder L, Nicholson R, Stables S, Gavin R. KD with severe cardiac sequelae: lessons from recent New Zealand experience. *J Paediatr Child Health* 2004; 40: 524-529.
6. Dominguez SR, Friedman K, Seewald R, Anderson MS, Willis L, Glode MP. KD in a pediatric intensive care unit: a case-control study. *Pediatrics* 2008; 122: 786-790.
7. Satou GM, Giamelli J, Gewitz MH. KD diagnosis, management, and long-term implications. *Cardiol Rev* 2007; 15: 163-169.