JCK Oral

JCK Oral 4 (II-JCKO4)

Kawasaki Disease/General Cardiology 1

Chair:Masahiro Ishii(Department of Pediatrics, Kitasato University School of Medicine, Kanagawa, Japan)
Chair:Min Huang(Shanghai Children's Hospital, China)
Chair:Seong-Ho Kim(Department of Pediatrics, Sejong General Hospital, Korea)
Sat. Jul 8, 2017 3:30 PM - 4:20 PM ROOM 3 (Exhibition and Event Hall Room 3)

3:30 PM - 4:20 PM

[II-JCKO4-03]Kawasaki shock syndrome complicated with macrophage activation syndrome

○Yao Lin, Lin Shi, Yanjun Deng, Yang Liu, Ping Lu (Department of Cardiology,Capital Institute of Pediatrics,Beijing,China)

We describe a case of Kawasaki shock syndrome (KDSS) in a 5-month infant who was complicated by macrophage activation syndrome (MAS). The boy had developed typical Kawasaki disease. After the first administration of intravenous immunoglobulin (IVIG) on the 6^{th} day at local hospital, he still had a persistent fever and elevated inflammatory markers 48 hours later without coronary artery dilation. He was admitted to our department on the 10^{th} day of illness and treated with IVIG (2 g/kg given as a single intravenous infusion) and aspirin (30–50 mg/kg/d). However, he still got a hypotension which showed a typical KDSS. Furthermore, blood cell (neutrophils, platelets and hemoglobin), fibrinogen and NK cell activity decreased, serum ferritin, cytokin and soluble CD25 concentration increased significantly, which showed signs of MAS. In addition, progressive coronary aneurysm formation was observed with the widest diameter of 9mm. On the basis of anti-shock therapy (including fluid resuscitation, human albumin intravenous infusion, and vasoactive agents), therapy for MAS was administrated immediately (methylprednisolone 4-6mg/kg/d tapering for 8 weeks and blood component transfusion). Clinical improvement was obtained 2 days later, and the patient was discharged 30 days later. Two months later, the boy was well developed with coronary aneurysm retraction to 7.3mm. KDSS and MAS are both severe and life-threatening complications of Kawasaki disease, therefore, early diagnosis and timely treatment is very important to save lives.