HK J Paediatr (new series) 2005;**10**:204-206

Case Reports

Unusual Presentation of Kawasaki Disease in an Adolescent Male Presenting With Hepatorenal Failure: Case Report

DML ***Wong,*** YM ***Ng,*** SH ***Lee***

Abstract Children older than ten years with Kawasaki disease accounts for less than one percent of all the cases. We report an adolescent male with Kawasaki disease presenting with unusual but serious hepatorenal failure. A twelve-year-old boy presented with high fever, rash, bilateral conjunctivitis, jaundice and developed acute renal failure, impaired liver function test and impaired conscious state and was suspected to have systemic infection. He required intensive care support and haemofiltration therapy. Sepsis workup was negative. He was treated with intravenous antibiotics but there was no response. Echocardiogram performed on day 15 showed dilated and irregular coronary arteries of RCA and LCA, fair ventricular contractility and mild pericardial effusions. Diagnosis of Kawasaki disease was made and intravenous immunoglobulin was given on day 15 of fever. Fever persisted after the first dose of immunoglobulin and subsided nine days after second dose of immunoglobulin. Serial echocardiogram showed the development of giant aneurysm of RCA and LCA. Cardiac catheterisation showed presence of giant aneurysm of RCA and moderate size aneurysm of LCA.

Key words Adolescent; Hepatorenal failure; Kawasaki disease

Introduction

Kawasaki disease typically affects young children less than five years old. Children older than ten years with Kawasaki disease accounts for less than one percent of all the cases.[1,2] Diagnosis of Kawasaki disease has been based on the presence of fever for more than five days and at least four out of five principal clinical features.[3] Patients who lack sufficient clinical signs to fulfill the classic criteria have been diagnosed as having incomplete Kawasaki disease.[3] We report an adolescent male with Kawasaki disease presenting with hepatorenal failure.

Department of Paediatrics & Adolescent Medicine, Queen Elizabeth Hospital, 30 Gascoigne Road, Kowloon, Hong Kong, China

DML **Wong** （黃美玲） *MBBS(HK), FRCP(Glasg), FHKAM(Paed)*
YM **Ng** （吳彥明） *MBBS(HK), FRCP(Glasg), FHKAM(Paed)*
SH **Lee** （李淑嫻） *MBBS(HK), FRCP(Edin), FHKAM(Paed)*

Correspondence to: Dr DML **Wong**

Received October 6, 2004

Case Presentation

A twelve-year-old adolescent male had fever and generalised erythematous rash on the second day and was admitted into private hospital. He developed jaundice and bilateral conjunctival congestion on the next day of fever. Presumptive diagnosis was that of sepsis. Fever persisted and there was progressive deteriorating liver and renal functions requiring continuous venovenohaemofiltration on day 13 of fever. He was transferred to tertiary care hospital for further management on day 14 of fever. Physical examination on day 14 showed jaundice of moderate severity, bilateral nonpurulent conjunctival congestion, generalised desquamation of skin. He was conscious with no neck rigidity. Examination showed signs of septic shock. The abdomen was distended and liver was enlarged to two centimeters below the costal margin with ascites.

Laboratory Findings

He developed progressive anaemia. There was marked leucocytosis. Leukocyte count rose from 10.7 x 10^9/L to a maximum of 56.9 x 10^9/L on day 13 of fever. The platelet

count was normal during the initial early phase of illness. Initial platelet count was 266 x 10^9/L on day 5 and started to rise on day 15 from 353 x 10^9/L to a maximum of 888 x 10^9/L on day 33 of course of illness. His erythrocyte sedimentation rate (ESR) was 50 mm/hr on day 9 and rose to a peak to 121 mm/hr on day 29 of fever and then it declined gradually. Creatinine level was increased to a peak of 583.8 μmol/L on day 13. Bilirubin level was increased and reached a peak of 510 μmol/L on day 16. ALT increased to a peak of 52 iu/L on day 14 of illness. Sepsis work up was negative. Autoimmune antibodies were negative. ASOT titre was negative. Viral study was negative. Cerebral spinal fluid was clear and there were lymphocytosis. Slidex test was negative. Polymerase chain reaction for tuberculosis, herpes simplex virus and enterovirus were negative. Fungal and viral cultures were negative.

Imaging Studies

CT scan of the brain was normal. Electroencephalogram showed diffuse encephalopathic changes compatible with hepatic encephalopathy.

Echocardiogram was performed on day 14 of fever. It showed bilaterally dilated and irregular coronary arteries. RCA measured 5.7 mm at origin while LCA measured 4.6 mm at origin. Ventricular contractility was fair and there was mild pericardial effusion.

Progress

He was diagnosed to have incomplete Kawasaki disease[3] complicated by coronary artery aneurysm. Intravenous immunoglobulin 2 gm/Kg was given on day 15 of disease. Second dose of immunoglobulin was given on day 20 for persistent fever.[4] Fever gradually subsided on day 29 and he developed generalised desquamation of skin again. Jaundice gradually subsided. Liver and renal function tests gradually returned to normal. Serial echocardiogram showed progressively enlarging aneurysm of both coronary arteries. RCA measured 8.5 mm at origin while LCA measured 7 mm at origin. Thallium scan showed reversible ischaemia at anterior, septal and inferior walls of myocardium. MRI gave no additional information.

He was put on aspirin and warfarin.[3,5] Coronary angiogram two years after onset of disease confirmed bilateral coronary aneurysm. Left coronary artery was irregularly dilated involving the origin of the left main coronary artery, left circumflex and the left anterior descending artery (Figure 1). Maximum size of aneurysm measured 7 mm and a length of 15 mm. Giant tubular aneurysm of right coronary artery measuring 9 mm was confirmed by coronary angiogram. There was no thrombus detected inside the coronary artery aneurysm.

Discussion

Kawasaki disease is uncommon in adolescent age group.[1,2] The diagnosis of Kawasaki disease in older children is often difficult and delayed because of atypical presentations. Acute Kawasaki disease is associated with hydrops gallbladder, intrahepatic bile duct damage and hepatobiliary dysfunction. Mild impairment of liver function test is a common finding in Kawasaki disease.[6,7] However jaundice or elevated bilirublin is uncommon in Kawasaki disease patient.[6,8] Our patient presented with hepatorenal failure with hepatic encephalopathy. This has never been reported. Jaundice may be caused by the extrahepatic problem like hydrops gallbladder or intrahepatic cause due to hepatobiliary dysfunction. The likely cause of hepatic failure in our patient is due to hepatocellular injury associated with Kawasaki disease.

Acute renal failure and pyelonephritis in Kawasaki disease had rarely been reported[8,9] but hepatorenal failure is never reported. Most cases of renal failure occurred in children older than five years old.

Our patient presented with fever of unknown origin and hepatorenal failure. This is a new and rare presentation.

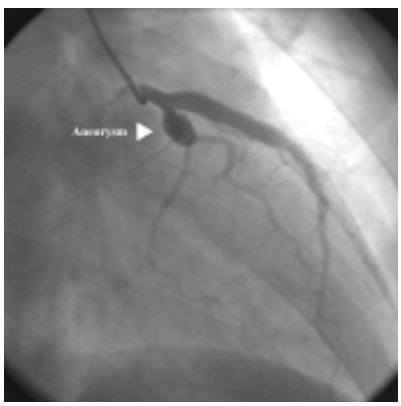

Figure 1 Coronary angiogram showing irregular and dilated left main coronary artery and aneurysm of left circumflex coronary artery.

The association between the two rare presentations is unknown. No virus could be identified in our patient and therefore the possibility of a viral etiology is not established. However this rare presentation should be alerted as prompt treatment with IVIG will help in controlling the systemic inflammation as well as decreasing the chance of development of coronary aneurysm.[10]

This case represents atypical case by age of presentation. There is higher risk of coronary giant aneurysm formation in adolescent age group.[1,2,5] The diagnosis was often delayed as paediatricians are less likely to consider Kawasaki disease at extremes of age with unusual presentations.[11] Hence paediatricians should be alert of the possibility of incomplete Kawasaki disease in adolescence with bizarre presentations as in this case and seek a second opinion. Follow-up echocardiogram is recommended in case of doubt.

Conclusions

Kawasaki disease presenting with hepatorenal failure is rare but serious. Adolescents with Kawasaki disease may have atypical presentation. They are at higher risk of coronary aneurysm formation. The diagnosis was late with a higher complication rate of coronary aneurysm. Clinician should have a high index of suspicion in evaluating patients presenting with fever of unknown origin in children beyond the typical age of Kawasaki disease.

References

1. Stockheim JA, Innocentini N, Shulman ST. Kawasaki disease in older children and adolescents. J Pediatr 2000 ;137:250-2.
2. Momenah T, Sanatani S, Potts J, Sandor GG, Human DG, Patterson MW. Kawasaki disease in the older child. Pediatrics 1998;102:e7.
3. Newburger JW, Takahashi M, Gerber MA, et al. Diagnosis, treatment, and long-term management of Kawasaki disease: a statement for health professionals from the Committee on Rheumatic Fever, Endocarditis and Kawasaki Disease, Council on Cardiovascular Disease in the Young, American Heart Association. Circulation 2004;110:2747-71.
4. Han RK, Silverman ED, Newman A, McCrindle BW. Management and outcome of persistent or recurrent fever after initial intravenous gamma globulin therapy in acute Kawasaki disease. Arch Pediatr Adolesc Med 2000;154:694-9.
5. Tatara K, Murata M, Itoh K, Kazuma N, Kondo C. Management of severe coronary sequelae of Kawasaki disease. Am Heart J 1996;131:576-81.
6. Kumar et al. Hepatobiliary involvement in Kawasaki disease 870. Pediatr Res 1998;43(4 suppl 2):150.
7. Ting EC, Capparelli EV, Billman GF, Lavine JE, Matsubara T, Burns JC. Elevated gamma-glutamyltransferase concentrations in patients with acute Kawasaki disease. Pediatr Infect Dis J 1998; 17:431-2.
8. Bonany PJ, Bilkis MD, Gallo G, et al. Acute renal failure in typical Kawasaki disease. Pediatr Nephrol 2002;17:329-31.
9. Atypical Kawasaki disease presenting as acute pyelonephritis. Nephrol Dial Transplant 2003;18 Supp 4:531-53.
10. Zhang T, Yanagawa H, Oki I, et al. Factors related to cardiac sequelae of Kawasaki disease. Eur J Pediatr 1999;158:694-7.
11. Pannaraj PS, Turner CL, Bastian JF, Burns JC. Failure to diagnose Kawasaki disease at the extremes of the pediatric age range. Pediatr Infect Dis J 2004;23:789-91.