CASE REPORT

Acute Complete Adult-onset Kawasaki Disease in a Middle-Aged Woman

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ABSTRACT

A 55-year-old Chinese lady was diagnosed with acute complete Kawasaki disease (KD) in January 2015. To our knowledge, this patient was the first described woman who was over 50 years and fulfilled all the characteristics of acute complete KD. The patient had a striking result on the 26th day since this disease onset after the initiation of treatment of immunoglobulin (IG) and aspirin; and did not have any complications up to now. The purpose of this case report was to raise the awareness of KD in the middle-aged persons and the therapeutic schedules, as there is no valid therapeutic therapeutic for adults with KD.

Key Words: Mucocutaneous lymph node syndrome, Adult, Kawasaki disease.

INTRODUCTION

Kawasaki disease (KD), which is also named as mucocutaneous lymph node syndrome, is an acute multi-systemic necrotising vasculitis. It affects middle and small sized vessels and occurs predominantly in young children of East Asia. Although KD is thought a kind of self-limited disease, it is also known as the most commonly acquired heart disease of children in a lot of developed countries, like Japan. It is also important to be aware of the diagnosis in adults, especially the old ones, although about 80% of patients occur at the age of 6 months to 4 years. The pathogenesis of KD is unclear and has been hypothesized that it is happened in the genetically predisposed individuals who at the same time are exposed to a possibly infectious trigger. Antigen-driven IgA response directed against cytoplasmic inclusions in KD tissues and the genes of inositol 1,4,5-triphosphate 3-kinase C (ITPKC), and genetic polymorphism of mothers against decapentaplegic homolog (SMAD), were thought to unlock the mysteries the pathogenesis of KD to some extent. As the KD’s etiology is still unknown, the diagnosis of KD is based on the clinical criteria as follows: fever, exanthema, conjunctivitis, erythema of lips and oral mucosa, and cervical lymphadenopathy. Timely treatment in the 10 days of the disease onset is so important that it can reduce the damage of coronary artery up to 75%. To the best of authors' knowledge, it is the first case reporting a female, complete KD, over 50 years. Without valid treatment standards for adults, the treatment of IG and aspirin were referred to the child therapeutic schedule, the patient had a striking result after the delayed initiation of treatment and did not have any artery complications up to now. The physicians should pay attention to the diagnosis of KD in the middle-aged persons and take actions as soon as possible to avoid deadly complications.

CASE REPORT

A 55-year Chinese woman presented to the hospital in January 2015 with high fever and right cervical lymphadenopathy for 18 days. The local node biopsy revealed no specific findings. Echocardiography showed the valvular vegetations of the heart. She was diagnosed with an atypical infection or subacute bacterial endocarditis (SBE) and had received a wide range of anti-inflammatory agents for more than 10 days, without any improvement. She denied animal exposure, new sexual partners, travelling recently, using alcohol or tobacco products or taking any medication. According to her medical history, she had hysteromyomectomy 10 years ago and fracture of her right knee 1 year ago. Contacts with similar symptoms or drug allergies were not identified. The history of the family was not contributory and no any family members had autoimmune diseases. The physical examination for the patient revealed that she has bilateral conjunctival chemosis, strawberry tongue, oedema of the face and the extremities, a skin rash of her trunk. The physical examinations for her chest, abdomen, neurological and musculoskeletal systems were not found obvious abnormal. The laboratory examinations disclosed normal white blood cells with neutrophilia; anaemia and thrombocytosis upon admission. Serum procalcitonin (PCT) and C-reactive protein (CRP) were all normal upon admission but CRP were later elevated obviously and decreased after treatment (Figure 1). The serum liver and renal function were normal. The analysis of
urine was normal. The results of three times of blood bacterial cultures were negative. All of the blood tests for autoimmune diseases, serum interferon-gamma release assays (IGRA) and serum tumor markers were negative. Serologic tests were negative for any kind of virus hepatitis. Cardiovascular physical examination and electrocardiogram (ECG) were normal. An echocardiogram showed a little pericardial effusion and no evidence of coronary aneurysms. Abdomen ultrasonography were normal. The enlargement of the cervical, axillary and inguinal lymph nodes were found in ultrasonography and positron emission tomography combined with computer tomography (PET/CT). Lumbar puncture was normal. The palms desquamated and the lips cracked on the 26th day since onset of the disease (Figure 2). With the suspicion of adult acute KD, she was given intravenous immunoglobulin (IVIG) at 25g per day and aspirin at 300 mg per day orally for the first 6 days, and then solely 300 mg of aspirin orally per day for another 3 months. As there was no any validated criteria for adult KD up to now, her diagnosis and treatment were referred to the criteria developed for child and considered the disorders and diseases with similar clinical manifestations. The women was in better condition and with normal temperature on the second day after the use of the drugs. In the days that followed, all clinical laboratory results tended to be normal inch by inch, except the reactive thrombocytosis and improved D-dimer remain. In total, she remained hospitalized for 20 days. Another echocardiogram was performed before discharge and no coronary aneurysms was found. On follow-up, about 12 months later, she had remained healthy.

**DISCUSSION**

KD is a self-limited multi-systemic necrotising vasculitis and occurs predominantly in young children of East Asia. The prompt treatment in 10 days of the disease onset is thought to avoid deadly complications. KD can also occur in adults, but the presentations to some extent are different from that can be founded in children. The representative clinical manifestations in both children and adults include fever, conjunctivitis, pharyngitis, and skin erythema which can progress to rash and desquamation, especially on the palms and soles. Cervical adenopathy, hepatitis and arthralgia are found more common in adults with 93%, 65% and 61%, respectively; while children are 15%, 10% and 24-38%, respectively. In contrast, children are more common in meningitis, thrombocytosis and coronary artery aneurysms with 34% vs. 10%, 100% vs. 55%, 18-25% vs. 5%, respectively compared with the adults. As the KD's etiology is still unclear, the diagnosis and differential diagnosis of adult-onset KD are all referred to the children’s criteria. The diagnosis of complete KD is based on the 4 of the 5 principal clinical manifestations including fever, exanthema, conjunctivitis, erythema of oral mucosa and lips, cervical lymphadenopathy and changes in the extremities are presented and at least 5 days of sustained fever. The “incomplete KD” is diagnosed, based upon coronary artery abnormalities in patients who are suspected of KD but do not fulfill all the diagnostic criteria. The timing of treatment initiation was thought the only significant risk factor of cardiovascular complications. Ideal treatment of KD for child should be administration of both IG and aspirin in the 10 days of the disease onset. The IVVG reduce the ratio of cardiac complications which may result in myocardial infarction and even sudden death. The mechanisms of IG and aspirin in treating KD are not clear. The possible mechanisms of IVVG may include neutralization of bacterial super-antigens, modulation of cytokine production and so on. The standard of treatment for child acute KD is 2g/kg infusion of IVIG and 80-100 mg/kg/day of aspirin in 4 divided doses. There were case reports describing the benefit in adults KD; but without controlled studies about the optimal timing, dose, or benefit of IVIG treatment existed in adults. Because the cases of acute adult KD are so rare, the IVIG plus aspirin treatment is based on the studies which are all performed in children. In this case, although the treatment was made on the 26th day of the onset of symptoms, the treatment also had
obviously clinical effect, although the serum PLT and D-dimer still at a high level when discharge. Due to the scarcity of literature regarding KD in adults, prompt treatment are so important for decreasing the ratio of cardiovascular morbidity and mortality. Attention should be paid to KD in adult patients, even the aged ones when the routine treatment cannot take effect. Further studies are needed to clarify the nature of adult KD.

REFERENCES