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Complete Kawasaki Disease in a 21-Year-Old Caucasian Patient

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Abstract

Kawasaki disease is a rare condition in adults and it may remain unrecognized because many patients do not fulfil all the diagnostic criteria for the diagnosis of "complete" Kawasaki disease. The authors report a case of 21-year-old male Caucasian patient complaining fever and presenting a diffuse maculopapular and erythema multiforme-like eruption, erythema and edema of the hands, feet and ears, non-exudative bilateral conjunctivitis, injected lips, strawberry tongue and symmetrical widespread lymphadenopathy. On the basis of clinical presentation, laboratory results and diagnostic criteria, a diagnosis of complete Kawasaki disease has been done. After treatment with systemic corticosteroids and aspirin (100 mg trice daily), the patient showed clinical improvement with typical large scale desquamation of the hands. The patient did not present any complication during one month follow-up. It is important to be familiar with the signs and symptoms of Kawasaki disease, because early diagnosis and adequate treatment is crucial for the prevention of cardiovascular morbidity and mortality.

Keywords

Kawasaki Disease, Adult Patient, Classification

1. Introduction

Adult Kawasaki disease (KD) is a very rare condition that may remain unrecognized. There is no specific diagnostic test available for KD. Diagnosis is based mainly on clinical features and etiology generally remains unknown. It is important to distinguish these forms from other potential diseases with similar symptomology because the management of the patient with KD is very different.

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2. Case Report

A previously healthy 21-year-old male Caucasian patient has been referred to the Emergency Departement for atypical acute exanthem initially interpreting as allergic eruption. On admission, the patient was febrile (39.0˚C) and he complained thrill which had lasted for 4 days despite amoxicillin treatment. Physical examination revealed a diffuse maculopapular and erythema multiforme-like eruption, erythema and edema of the hands, feet and ears, non-exudative bilateral conjunctivitis (**Figure 1**), injected lips, strawberry tongue and symmetrical widespread lymphadenopathy. Diagnosing an allergic disease, emergency physicians started the treatment with intravenous methylprednisolone (30 mg/kg).

Laboratory investigations showed neutrophilic leukocytosis (27.60×10^9/L), elevated C-reactive protein (29.1 mg/L) and erythrocyte sedimentation rate (120 mm/hour). Serological assay for human immunodeficiency virus (HIV) and hepatotropic viruses were negative. Serology for Epstein-Barr virus, morbillivirus, cytomegalovirus and Parvovirus B19 was indicative of past immunity. Pharyngeal swab was negative and antistreptolysin titer resulted within normal limits. Skin biopsy revealed no specific histological findings, demonstrating a mild hyperplasia of the epidermis and perivascular inflammatory infiltrates of lymphocytes in the dermis. Direct immunofluorescence showed deposits of IgM and C3 in dermal vessels.

After six days from admission, the patient showed clinical improvement but some lesions were still present on the trunk and large scale desquamation of the hands (**Figure 2**) and furfuraceous desquamation on the face was observed (**Figure 3**).

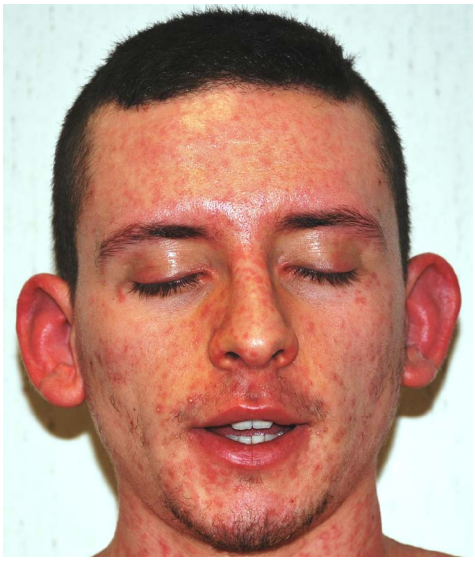

Figure 1. Maculo-papular eruption on the face and neck, marked edema of the ears, bilateral conjunctivitis and erythema of the lips.

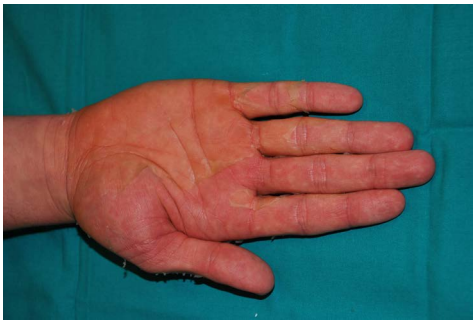

Figure 2. Lamellar desquamation of the palms 6 days after diagnosis.

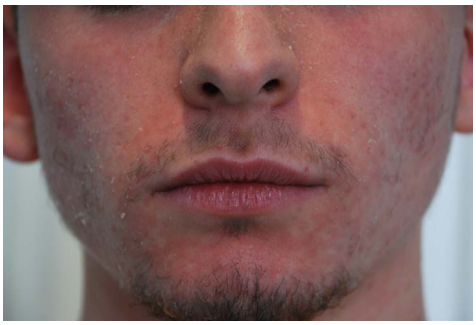

Figure 3. Slightly desquamation of the face 6 days after diagnosis.

The patient still complained fever (38.0˚C) and asthenia. Electrocardiogram, chest X-ray and echocardiography were within normal limits. On the basis of clinical presentation and diagnostic criteria a diagnosis of complete Kawasaki disease has finally been done. The patient was then treated with aspirin (100 mg trice daily) and systemic corticosteroids were gradually reduced in two weeks. He clinically improved after typical desquamation of palms and soles with complete resolution after 1 month.

3. Discussion

Kawasaki disease (KD) is an acute systemic vasculitis that occurs primarily in children and rarely in adults [1]. No specific diagnostic tests are available for KD, so the diagnosis is based on the presence of characteristic clinical findings [2]. When KD is defined "complete", the diagnosis is based on the presence of at least 5 days of fever, and 4 of the five principal clinical features (polymorphous exanthema, changes in extremities: acute phase with erythema of palms and soles, and edema of hands and feet, subacute phase with desquamation of fingers and toes, bilateral bulbar conjunctival injection, changes in lips and oral mucosa, cervical adenopathy), as in [3]. Some patients with suspected KD do not fulfill all the diagnostic criteria, so the diagnosis is made based on coronary artery abnormalities. These cases of "incomplete KD" are more frequent in children under one year of age [3]. In other studies [4] "incomplete KD" refers to patients with fever lasting ≥5 days and 2 or 3 clinical criteria (rash, conjunctivitis, oral mucosal changes, changes on extremities, adenopathy), without reasonable explanation for the illness. The term "atypical KD" should be reserved for patients who have atypical symptoms that are not common in classical KD, such as renal impairment, acute surgical abdomen, and pleural effusion [3]. In our case, on the basis of clinical presentation and diagnostic criteria, a diagnosis of "complete" Kawasaki disease was made.

The low diagnostic accuracy for adult-onset KD, in contrast to that in pediatric patients, can be attributed to the several differential diagnoses that are possible in adult, including drug hypersensitivity reaction and toxic shock syndrome. Other potential diseases with similar symptomology include streptococcal infection (scarlet fever), leptospirosis, Mycoplasma and Rickettsiae diseases, adenoviral and other viral illnesses (e.g. measles, rubella, Epstein-Barr virus infection, and fifth disease), toxoplasmosis, mercury poisoning (acrodynia), and rheumatologic disorders (Reiter syndrome, juvenile rheumatoid arthritis, and Still's disease) [2].

4. Conclusion

Early diagnosis and adequate treatment of Kawasaki disease is crucial for the prevention of cardiovascular morbidity and mortality. Treatment with aspirin and intravenous immunoglobulin has been seen to reduce the risk of developing coronary aneurysm from 20% to 25% to less than 5% if administered within the first 10 days of illness [5]. In KD, associated treatment within travenous immunoglobulin, aspirin and, possibly, pulse intravenous methylprednisolone is recommended [5]. The long-term follow-up of the patients is important for the prevention of cardiovascular morbidity and mortality.

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