Lupus-like onset of recurrent Kawasaki disease in an adolescent boy

Sirs,

A 15-year-old boy was admitted to our hospital for fever and rash. Five days before admission he had had fever and sore throat treated with clarithromycin without clinical improvement. At admission he was still febrile but his general condition was fair. A butterfly rash on his face was evident (Fig. 1), and he referred diffuse arthralgies, muscle weakness, headache and asthenia. Lymphadenopathy was not present.

Laboratory tests showed an elevated erythrocyte sedimentation rate (ESR), C reactive protein (CRP), firbrinogen, white blood cell number, and neutrophilia. Renal function was normal. Blood and urine cultures were negative. Rapid diagnostic test for group A β-haemolytic Streptococcus was negative. Serological tests for detection of antibodies IgM against *Epstein-Barr virus*, measles, mumps, chickenpox, herpes viruses, adenovirus, parvovirus, HIV, hepatitis C and B, cytomegalovirus, *Mycoplasma pneumonia, Leishmania* and blood culture were negative. Antinuclear autoantibodies (ANA), antineutrophil cytoplasmic antibody (ANCA) and anti-DNA titres were negative; anticardiolipin autoantibodies (ACL) IgM values were positive (50 MPL), IgG were absent. Chest radiograph was negative.

During the 4 following days, the patient presented high grade fever and developed cheilitis, strawberry tongue, bilateral non-exudative conjunctivitis with haemorrhages in the left eye and diffuse maculopapular rash. Kawasaki disease (KD) was then suspected. Electrocardiogram (ECG), echocardiogram and abdominal ultrasound scan were negative. The child was treated with intravenous immunoglobulin and acetil salicylic acid. Fever as well as systemic manifestations promptly disappeared. Blood tests gradually improved and the boy was discharged one week after admission with an acetil salicylic acid treatment. Six days later he was in general good condition, but presented hands and feet periungueal digital peeling. On 9 day from discharge, the patient presented again with fever, bilateral conjunctival injection without exudates, while extremity peeling was still present. All blood tests previously carried out on first admission including ACL were repeated. They resulted in normal range apart from ESR (90 mm/h), CRP (8.41 mg/dL), and fibrinogen (802 mg/dL). Cardiac evaluation, with ECG and echocardiography and abdominal ultrasound scan were normal.

Recurrent KD was then suspected (1). In the recurrent episodes of KD, which are more frequent in the adolescents than in younger children, steroid therapy or second cycle of IVIg are indicated (2-4). Thus one dose intravenous methylprednisolone was administered. Rapid improvement of signs and symptoms occurred and the patient was discharged one week later in good general condition with acetil salicylic acid treatment. At the last check-up in December 2003 a cardiologic evaluation, ECG and echocardiography were normal.

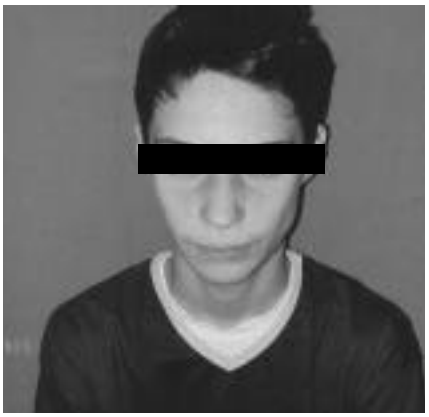

Fig. 1.

KD is rare in adolescents. The incidence reported is 0.2 cases for 100,000 subjects aged from 15 to 18 years (5). Furthermore, the signs and symptoms are often atypical (5). In these cases, the KD diagnosis may be difficult and consequently treatment may be delayed. Therefore, coronary complications are more frequent in adolescents than in younger children (6,7). In our case, the sudden and persistent fever, butterfly rash and systemic manifestations led us to consider systemic lupus erythematosus (SLE) diagnosis. However, the ACR diagnostic criteria were not satisfied. Otherwise, according to the revised diagnostic criteria for KD, the presence of bilateral non-exudative conjunctivitis, cheilitis, strawberry tongue and maculopapular rash suggested KD (9). The diagnosis was eventually confirmed by rapid improvement after IVIg therapy and subsequent extremity peeling. Positive IgM but negative IgG ACL are also reported in KD (10), possibly due to polyclonal B-lymphocyte activation. To our knowledge an adolescent with KD mimicking the onset of SLE has not been described so far. The diagnosis of KD in adolescents may be difficult, since the signs and symptoms are often atypical and can mimic other disease including SLE. A timely diagnosis is mandatory to reduce possible complications.

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References

1. HIRATA S, NAKAMURA Y, YANAGAWA H: Incidence rate of recurrent Kawasaki disease and related risk factors: from the results of nationwide surveys of Kawasaki disease in Japan. *Acta Pediatr* 2001; 90: 40-4.
2. HAN RK, SILVERMAN ED, NEWMAN A *et al.*: Management and outcome of persistent or recurrent fever after initial intravenous gamma globulin therapy in acute Kawasaki disease. *Arch Paediatr Adolesc Med* 2000; 154: 694-9.
3. SHULMAN ST: Is the role for corticosteroids in Kawasaki disease ? *J Paediatr* 2003; 142: 601-3.
4. SUNDEL RP, BAKER AL, FULTON DR *et al.*: Corticosteroids in the initial treatment of Kawasaki disease: report of a randomized trial. *J Paediatr* 2003; 142: 611-6.
5. CALLAWAY LK, BENNETT CJ, CALLAWAY JK: Adolescent Kawasaki disease with uveitis. *Intern Med J* 2002; 32: 421-2.
6. HIROSE K, NAKAMURA Y, YANAGAWA H: Cardiac sequelae of Kawasaki disease in Japan over 10 years. *Acta Paediatr Jpn* 1995; 37: 667-71.
7. DE ZORZI A, COLAN SD, GAUVREAU K *et al.*: Coronary artery dimensions may be misclassified as normal in Kawasaki disease. *J Paediatr* 1998; 133: 254-8.
8. RIUZ-IRASTORZA G, KHAMASHTA MA, CASTELLINO G *et al.*: Systemic lupus erythematosus. *Lancet* 2001; 357; 1027-32.
9. Diagnostic guidelines for Kawasaki disease. *Circulation* 2001; 103: 335-6.
10. GUPTA M, JOHANN-LIANG R, BUSSEL JB *et al.*: Elevated IgAand IgM anticardiolipin antibodies in acute Kawasaki disease. *Cardiol - ogy* 2002; 97: 180-2.

A case of Kawasaki disease accompanied by Henoch-Schönlein purpura

Sirs,

Cases of vasculitis occasionally have overlapping features with other forms of vasculitis (1); however, there have been few reports of such occurrences in Kawasaki disease (KD), a childhood form of vasculitis (2,3). We describe a patient who concurrently showed features of KD and Henoch-Schönlein purpura (HSP).

A 3-year-old boy was admitted with a history of 7-day fever, injected conjunctivae, red lips, a non-purpuric exanthema, puffy hands, and right cervical lymphadenopathy (1.8 cm in diameter). Laboratory data revealed leukocytosis (15,600/μl), mild thrombocytosis (445,000/μl), and an elevated C-reactive-protein level (CRP, 15.6 mg/dl). A diagnosis of typical KD was made, and in-