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.

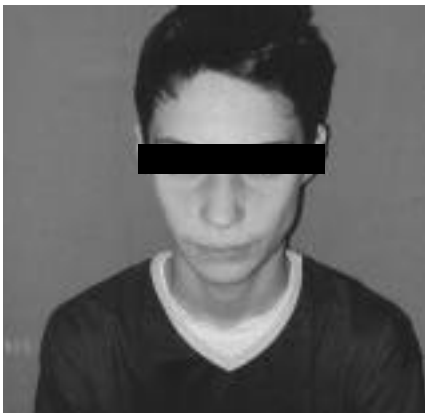

Fig. 1.

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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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-

Table I. Comparison between Kawasaki disease and Henoch-Schönlein purpura.

	Kawasaki disease	Henoch-Schönlein purpura
Nomenclature of vasculitis (2, 6)	Medium vessel vasculitis, involving large or small vessels	Small vessel vasculitis
Demographic features (2, 3, 6)		
Age distribution	Mainly under 5 years of age	Mainly 4 to 7 years of age
Male-to-female ratio	1.5–1.6 : 1	1.2 : 1
Symptom (frequency) (6-8)		
Fever	High grade fever (95-100%)	Fever preceding (occasionally)
Eye involvement	Conjunctival injection (90%)	Episcleritis and/or anterior uveitis (infrequent)
Changes of the oral mucosa	Erythematous lips (90-95%), strawberry tongue (70-80%)	Purpura (infrequent)
Skin lesion	Polymorphous exanthema (90%)	Purpura (50-100%), preceded by maculopapular lesions
Lymphadenopathy	Cervical (50-75%)	Celiac (infrequent)
Changes of the extremities	Swelling, redness, and/or periungual desquamation of the peripheral extremities (90-95%), arthralgia (25%)	Subcutaneous edema (35-70%), arthralgia (65-85%)
Gastrointestinal involvement	Diarrhea (35%), abdominal pain (occasionally)	Abdominal pain (45-75%), bleeding (35%)
Complication (frequency) (2, 6-10)		
Heart	Coronary lesions (15-25%), myocarditis (50%)	Myocardial infarction (rare), myocarditis (rare)
Kidney	Urethritis (30%), nephritis (rare)	Nephritis (25-50%)

travenous immunoglobulin and aspirin were administered, which was followed by improvement with periungual desquamation.

At the age of 4 years 5 months, the patient exhibited high fever for 3 days, a rash, arthritis on the knees, and left cervical lymphadenopathy (2.0 cm). The rash, beginning as maculopapules, progressed to crops of palpable purpura 3-5 mm in diameter on the legs. The eyes and lips were intact and subcutaneous edema was absent. The white-blood-cell count was 13,000/µl, the platelet count 334,000/µl, and CRP 7.4 mg/dl. Colicky abdominal pain occurred on the 5th day of illness. He received only supportive treatment and improved in 2 weeks. Six months later he was admitted because of high fever for 6 days and abdominal pain. He was noted to have non-purulent injected conjunctivae, right cervical lymphadenopathy (1.5 cm), an exanthema and purpura on the legs. Injected lips or edema on the peripheral extremities was not seen. The white-blood-cell count was 10,200/µl, platelet count 390,000/µl, and CRP 4.5 mg/dl. On the 7th day he was treated with intravenous immunoglobulin and aspirin, which were effective. In the second and third episodes, the non-thrombocytopenic purpura were compatible with HSP but not KD, whereas the high fever and cervical lymphadenopathy were consistent with KD but not HSP. Furthermore, the injected conjunctivae seen in the third episode is characteristic of KD, but not HSP. We therefore diagnosed these episodes, especially the third one, as overlapping atypical KD (2) and HSP.

At the age of 5 years 6 months, palpable purpura on the lower extremities, abdominal pain, arthralgia, and subcutaneous edema on the feet appeared. The white-blood-cell count was 9,200/µl, platelet count 365,000/µl, and CRP 0.8 mg/dl. These symptoms faded with supportive treatment, and reappeared 2 months later. The last episode was considered as typical HSP.

None of these 4 episodes showed coronary artery lesions, nephritis, abnormal clotting system, or infections of bacteria including group A ß-hemolytic streptococcus in throat and blood cultures. Serological tests for collagen diseases and viruses, urine or stool cultures, abdominal ultrasonography, or skin biopsies were not performed.

This patient is the second case of KD accompanied by HSPin the English literature; the first case (4) was accompanied by hemolytic uremic syndrome, different from our patient. A case showing symptoms compatible with KD at the onset and subsequently aortitis was reported (5). Similarly to polyarteritis nodosa (1), KD – a medium vessel vasculitis – may overlap small vessel vasculitis including HSPor large vessel vasculitis. The presence of vasculitis overlapping KD and HSPmay be supported by common clinical findings (Table I). A single trigger might concurrently induce KD and HSPin a patient genetically susceptible to both. The present patient recurrently suffered from KD, HSP or both on four occasions, although an apparent trigger was absent. Additionally, IgA may play a role in the pathogenesis, because IgA plasma cells are detected in several organs such as the coronary arteries and kidney in KD (2) and IgA deposition in the skin and renal mesangium in HSP (6,7). An etiologic link may be a clue to elucidate the unknown causes of KD and HSP.

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