Case report

MUCOCUTANEOUS LYMPH NODE SYNDROME (KAWASAKI SYNDROME)

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ABSTRACT

The authors present a Kawasaki syndrome in an adolescent, which is very rare. He failed to respond to antibiotic treatment with amoxicillin, but did respond to salicylates and antihistaminics. Thanks to the early diagnosis and the effective treatment, later complications could be avoided.

KEY WORDS

mucocutaneous lymph node syndrome, Kawasaki, early diagnosis, treatment with salicylates

INTRODUCTION

The mucocutaneous lymph node syndrome was first reported by Kawasaki in 1967. This disease occurs primarily in childhood. The typical clinical symptoms are high fever of the continua-type, indurative edema of the extremities, palmar and plantar erythema, polymorphous generalised exanthema, enanthema of the labial and oro-pharyngeal mucosa, bilateral conjunctivitis, acute non-purulent unilateral cervical lymphadenopathy and desquamation of the skin on the finger tips during the regression of the disease (1,2).

We present the classical Kawasaki syndrome in an adolescent, which is quite rare at this age.

CASE REPORT

A 16-year-old male patient was registered at our department 4 days after the appearance of the first clinical symptoms. He complained of an infection of the upper respiratory tract accompanied by high fever (38-39°C), lesions of the mucous membranes and an unilateral lymph node hypertrophy on the neck. He failed to respond to the treatment (amoxicillin, paracetamol, diphenhydramin) recommended by his family doctor and by the specialist for internal diseases. Additionally, two days later a disseminated exanthema and edema of the extremities appeared. The family-anamnesis was negative, his parents mentioned only a pneumonia in early childhood, till the present disease he enjoyed a good health.

***State at admission*:** numerous, disseminated, well defined maculopapules 0.2-2 cm in size, which extended to palms and soles. On the lower extremities there were point-like petechias. The lips were inflammed, red and swollen; there were some rhagades.The oral mucosa was inflammed too, the conjunctivae were injected (Fig.1-3).

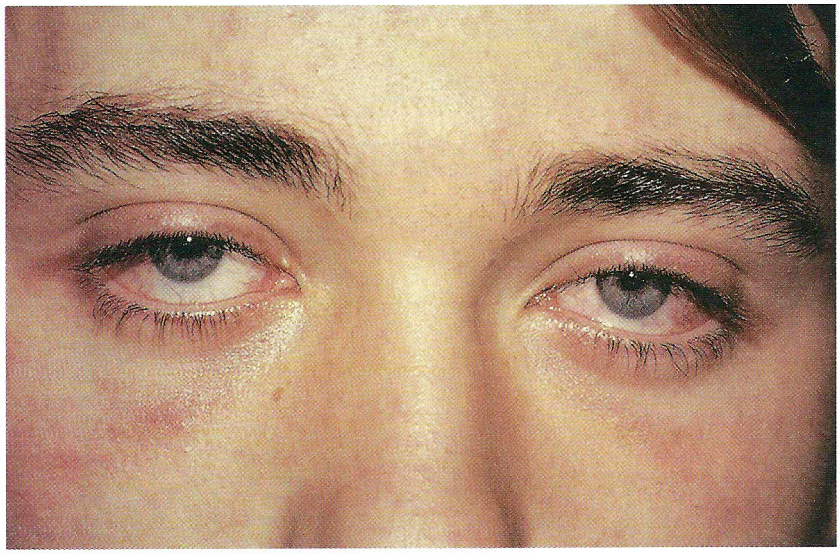

Fig. 1. Bilateral conjunctivitis

Other findings: covered right lung on the X-ray pictures, hepatomegalia, tachycardia and aching and restricted movements of extremities. In the right occipital region a painful hypertrophic and mobile lymph node was palpable.

Routine laboratory tests: ESR 20 mm/h, slight anaemia:hemoglobin 7.72 mmol/l, hematocrit 0.358, slight leukocytosis 98x 10^9/l, increased values of liver enzymes: SGOT 75 U, SGPT 54 U, decreased value of thrombocytes 89x10^9/l, and an increased value of fibrinogen 4.91x10^9/l. The differential blood count was (%): non-seg 0, seg 74, basophils 0, eosinophils 0, monocytes 0, lymphocytes 26. Total protein: 79 g/l. Protein electrophoresis: albumin 0.636, α_1 0.036, α_2 0.073, β 0.091, γ 0.162.

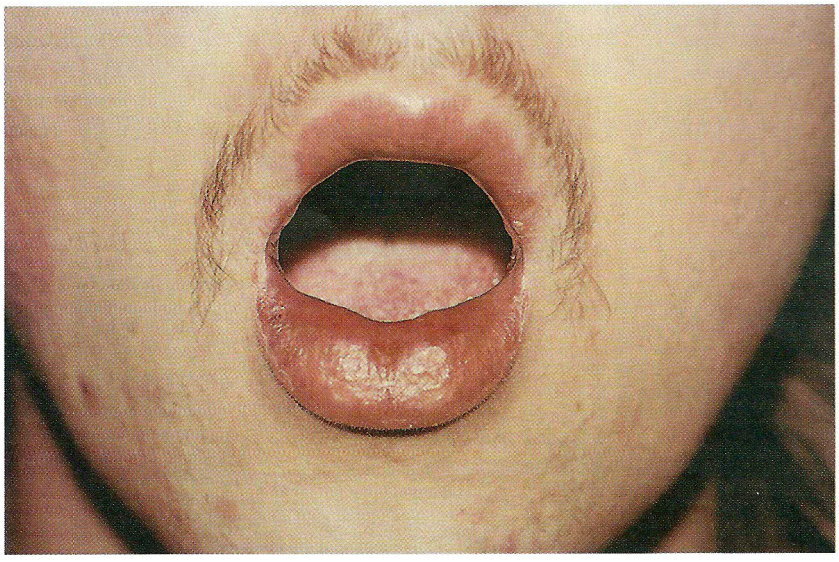

Fig. 2. Edematous, inflammed lower lip with fissures

Tests for viruses: HIV, parvo, adeno, morbilli, rubella, Epstein-Barr virus were negative. The Hbs antigens, hepatitis C virus antibody, monosticon tests and immunoelectrophoresis were also negative. Pathogenic agents from the larynx mucosa could not be cultivated. The abdominal ultrasound examination indicated hepatosplenomegalia and diffuse liver damage. The ECG showed no cardial complications and the result of the echocardiography examination was also negative.

Course of the disease and treatment: high fever (38-39°C) was registered during 3 days before and 3 days after the admission to our department. We induced the therapy with salicylates, antihistamines (clemastin, loratadin) and indometacin. As a result of the therapy, patient's continuous high temperature gradually receeded and his articular pain diminished significantly. Due to the externally applied antiphlogistic soothing treatment, the inflammatory symptoms of the skin and mucosa regressed. Our diagnosis was based on the existing symptoms at the admission. The diagnosis was confirmed also by the course of the disease. Further typical symptoms were the desquamation of the skin on the finger tips and the regression of the unilateral adenopathy on the neck in the second week of the disease (Fig 4).

Because of the early diagnosis and the immediate treatment with salicylates no cardial complications were found either by echocardiography during the stay in the clinic nor 8 weeks later.

DISCUSSION

Kawasaki was the first to describe the above-mentioned disorder as mucocutaneous lymph node syndrome. Today it is referred in the literature as the Kawasaki syndrome.

The number of reports the disease occurring at different ages has been increasing. It has been reported primarily in childhood, more often in boys than in girls (2:1). In some of these cases the disease was identified only in the late complicated stages (3,4,5). As far as we know in Hungary only pediatric cases and only one in an adult patient was published (6). Reports which describe an atypical course are also increasing (7).

The etiology of the disease is not quite clarified. On the basis of the frequent epidemic-like occurrence a biological agent as a cause has to be assumed (8).

The provocative roles of the viruses as Epstein-Barr, retro- and hepatitis B as well as Yersiniae, Rickettsiae, acarus and even of heavy-metal toxaemia were discussed but were not proven yet. The latest research tries to find the role of the parvovirus B 19 infection (1,2,9,10,11,12). Earlier it was believed to be a variant of polyarteritis nodosa juvenilis. Besides the immunological factors the cytokins seem to play a significant role in the pathomechanism of the disease. Thus the TNF-alpha can be blamed for the vascular changes. The production of the interleukin-2 and interleukin-6, the T-cell activity and the megakaryocyte proliferation seem to be on an increase. The interferon-gamma helps to present the new antigen to the macrophages.

During the disease the blood level of the immunoglobulins, the number and concentration of antiendothelial and antineutrophil antibodies increase. It is important to mention that the frequency of coronary complications shows correlation to concentration of cytokines in the tissues. (13). The recently drawn criteria help to diagnose the disease (14,15). Nowadays the superantigens are raised as the causal factor.

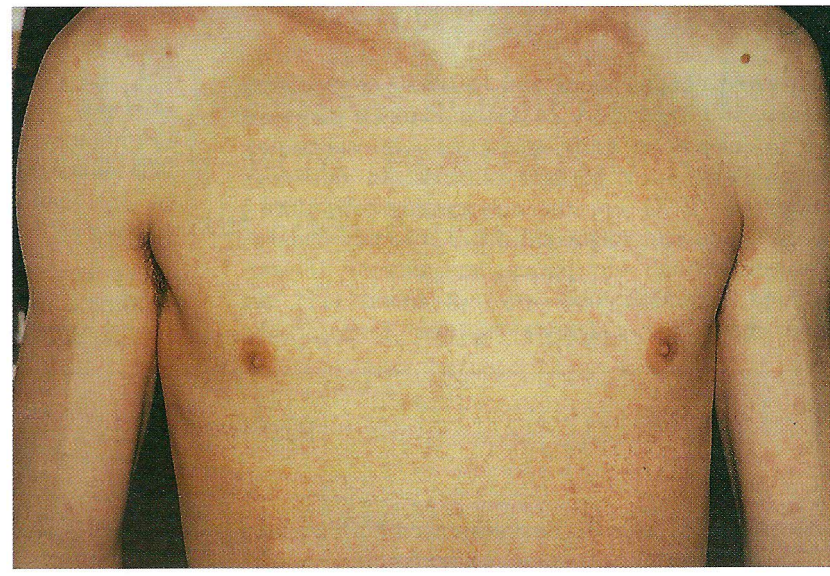

Fig. 3. Maculo-papular exanthema on the trunk

From differential diagnostic point of view, scarlatina, measles, rubella, erythema multiforme, hepatitis, juvenile rheumatoid arthritis, Steven-Johnson syndrome, infantile polyarteritis nodosa and toxic shock should be taken into consideration.

The serious complication of the syndrome are the coronary thrombosis and aneurysm. Death sets in due to aneurysm rupture or to infarction. Deviations, which can be registered by ECG can occur in the first or in the second week of the acute phase. Vasculitis can develop, the media of the vessels is destroyed and an inflammatory infiltration in the intima and in the surrounding connective tissue can be observed. On rare occasions cases of aseptical meningitis (5) and an obstruction of the arteria cerebri media were noted (16).

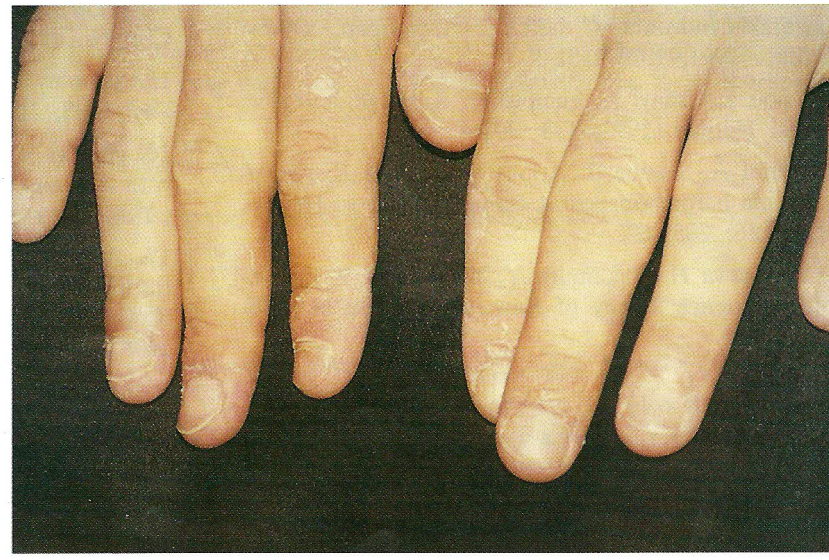

Fig. 4. Typical desquamation of the skin, localized on the finger tips

CONCLUSION

To avoid complications and to obtain a successful therapy, it is necessary to make a quick diagnosis and to induce an early treatment with salicylates, gamma-globulin or perhaps steroids. In our case, neither the ECG nor the echocardiography during the early and late stages did prove coronary damage. However, though our patient was already 16-year-old and thrombocytosis was not found (17), we could make the diagnosis on the basis of the following symptoms: high fever of the continua-type for more than 5 days, bilateral conjunctivitis, injected lips and pharynx, fissured lips, edematous erythema of hands and legs, a polymorphous and disseminated exanthema as well as cervical lymphadenopathy. The desquamation of the skin in phalangeal regions of the hands started in the 2nd week of the disease.

We could make the diagnosis of the Kawasaki syndrome in spite of the age (6) of the patient and of the moderate thrombocytopaenia.

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