

MUCOCUTANEOUS LYMPH NODE SYNDROME (KAWASAKI SYNDROME): A CASE REPORT*

*Asım Gültekin MD**, Ayşe Gökalp MD**, Aynur Oğuz MD**
Mehmet Marufi Hah MD****

Key words: Kawasaki syndrome, mucocutaneous lymph node syndrome

Kawasaki syndrome (KS), is an acute systemic illness of infancy and early childhood which was first described in Japan by Tomisaku Kawasaki¹ in 1967. Since then it has been recognized worldwide and similar cases from Korea, Canada, the USA, Greece, as well as Turkey have been reported²⁻⁶. Although it is widely believed to be microbial, the etiology of the disease remains unknown^{7,8}. Since this syndrome is very rarely encountered in Turkey, this case has been presented.

Case Report

A nine-year-old boy admitted to Cumhuriyet University Hospital presented with swelling of the hands, face and neck, a rash and a fever of ten days duration which was not responsive to antibiotic therapy.

Physical examination revealed a well-developed boy with body weight 22 kg. Bilateral bulbar conjunctival congestion, a polymorphous rash, diffuse reddening of the oral and pharyngeal mucosa with prominent tongue papillae, dry, cracked and fissured lips and moderately enlarged posterior cervical lymph nodes were observed. The ankle and knee joints were tender and feverish. Reddening of the palms and soles of the feet with non-pitting edema was also noted. There were no other important physical findings (Figs. 1,2).

Laboratory studies showed hemoglobin 12 g/dl, White Blood Cell count 42400/mm³ with a distribution of 78% neutrophils, 8% band forms, 12% lymphocytes, 2% monocytes and a normal platelet count. The sedimentation rate (ESR) was 62 mm/hr. BUN, creatinine, blood electrolytes, total protein and liver function studies were normal. Urinalysis showed pyuria (8-10 WBC per

* From the Department of Pediatrics, Cumhuriyet University Faculty of Medicine, Sivas.

** Associate Professor of Pediatrics, Cumhuriyet University Faculty of Medicine.

*** Assistant Professor of Dermatology, Cumhuriyet University Faculty of Medicine.



Fig. 1: The general appearance of the patient; indurative edema of the extremities and polymorphous rash.



Fig. 2: Typical appearance of the lips and bilateral conjunctival congestion.

high-power field) and trace proteinuria. Throat and blood cultures and C-reactive protein were negative. Antistreptolysin O (ASO), chest X-ray and electrocardiographic studies (ECG) were also normal.

Since the patient was mildly dehydrated and had all the clinical features of KS on admission, the antibiotic therapy which was started before admission was stopped. Intravenous fluid (1/4 serum physiologic and 3/4 5% dextrose) 2500 cc/m² and 100 mg/kg salicylate orally were started. Forty-eight hours after this therapy, the patient's fever was brought under control. On the eighth day after admission, desquamation of the skin, mostly of the fingers, soles of the feet and palms was noted. The patient became clinically normal at the end of the second week.

Discussion

The diagnosis of Kawasaki syndrome is based on the following clinical characteristics: prolonged fever of five days' duration or more which is unresponsive to antibiotics, bilateral bulbar conjunctival congestion, lip and oral mucosal changes, polymorphous rash, indurative edema and cervical lymphadenopathy⁸⁻⁹. Although five of the above-mentioned six criteria are sufficient for clinical diagnosis, our case had all of the six characteristics, including most of the minor ones such as increased ESR, negative ASO titer, leukocytosis with a shift to the left and pyuria. The manifestations of KS show a widespread inflammatory vasculitis¹⁰. The visible lesions on the skin are accompanied by involvement of the coronary vasculature, meningeal membranes, gallbladder, pancreas, liver, bowel, kidney and joints. The presence of ECG abnormalities in 25% of the children indicates that KS is commonly associated with cardiac involvement; in such cases the fever generally lasts for more than 14 days¹¹⁻¹². In our case, however, the ECG findings were normal and the fever lasted less than 14 days. This may indicate the absence of cardiac involvement.

The diagnosis of Kawasaki syndrome requires differentiation from measles, scarlet fever, infectious mononucleosis, Stevens-Johnson syndrome, and staphylococcal scalded-skin syndrome⁹⁻¹¹. Since very few cases of KS have been reported in Turkey, this may indicate that this syndrome might have been confused with the above-mentioned diseases. Likewise, the case presented had been diagnosed and treated as scarlet fever before admittance to our hospital.

Summary

Kawasaki syndrome (KS) is a rarely encountered disease in Turkey. To our knowledge, only three cases have thus far been reported. A nine year-old boy presenting with the clinical characteristics of a high fever unresponsive to

antibiotics, a polymorphous rash, reddening of the palms and soles of the feet, indurative edema, cervical lymphadenopathy and bilateral conjunctival congestion is diagnosed as KS and the relevant literature is reviewed.

REFERENCES

1. Kawasaki T. [Acute febrile mucocutaneous syndrome with lymphoid involvement with specific desquamation of the fingers and toes in children.] *Jap J Allerg* 16:178, 1967 (Jap).
2. Kim J, Yeo Y, Leo DB. Mucocutaneous lymph node syndrome: clinical observation of eight cases. *Kore New Med J* 16:1157, 1973.
3. Melish ME, Hicks RM, Larson EJ. Mucocutaneous lymph node syndrome in the United States. *Am J Dis Child* 130:599, 1976.
4. Valaes T. Letter: Mucocutaneous lymph node syndrome (MLNS) in Athens, Greece. *Pediatrics* 55:295, 1975.
5. Özsoylu S, Akgün NA. Akut febril mukokutaneus lenf bezi sendromu (MLNS). *Çocuk Sağlığı ve Hastalıkları Dergisi* 19:57, 1976.
6. Hiçsönmez G, Kanra G, Koçak H, et al. Acute mucocutaneous lymph node syndrome. *Clin Pediatr (Phila)* 16480, 1977.
7. Yanagihara R, Todd JK. Acute febrile mucocutaneous lymph node syndrome. *Am J Dis Child* 134:603, 1980.
8. Hicks RV, Melish ME. Kawasaki syndrome. *Pediatr Clin North Am* 33:1151, 1986.
9. Hansen RC. Staphylococcal scalded skin syndrome, toxic shock syndrome, and Kawasaki disease. *Pediatr Clin North Am* 30:533, 1983.
10. Meade RH, Braudt L. Manifestations of Kawasaki disease in New England outbreak of 1980. *J Pediatr* 100:558, 1982.
11. Koren G, Lavi S, Rose V, Rowe R. Kawasaki disease: review of risk factors for coronary aneurysms. *J Pediatr* 108:388, 1986.
12. Bitter JJ, Friedman SA, Paltzik RL, Mofenson HC. Kawasaki's disease appearing as erythema multiforme. *Arch Dermatol* 115:71, 1979.