



Kawasaki Disease川崎氏症(英文)

Kawasaki disease

Kawasaki disease (KD) or Mucocutaneous Lymph Node Syndrome (MCLS) is first reported in Japan in 1967 by Dr. Kawasaki. Eighty percent patients of the disorder are younger than 5 years of age.

Etiology and Pathogenesis

Specific etiologic pathogen has not been identified. The tendency of cases clustering in winter and spring months suggested a relationship with infectious disease. The fact that most cases occur on children younger than 5 years of age also indicates the patient' s abnormality in immunoregulatory function. However, the pathological findings show that it is a disease of systemic arteritis invading small to medium size arteries throughout body, especially the coronary arteries of the heart. .

Clinical Features

1. Fever for at least 5 days duration
2. Bilateral nonexudative conjunctivitis
3. Lesions on the lips and oral cavity, including redness, dryness, fissuring , bleeding, tenderness, and strawberry tongue
4. Lesions on the extremities, including erythema and/or indurative edema of palms and soles, which later convert to granular desquamation at the subungual regions (2nd week of illness)
5. Cervical lymphadenopathy, larger than 1.5 cm in width
6. Polymorphous exanthema with all kinds of skin eruptions, excluding bullous or vesicular lesions.

7. Lesions on previous BCG vaccination site : redness or scarring

8. perineal area redness or desquamation

Complication

Coronary artery aneurysm (5 - 15 %) and acute myocarditis (can be detected by echocardiography)

Treatment

1. Acute stage : Intravenous immunoglobulin (VIG) infusion by high dose regimen (2 gm/kg) with slowly dripping for 24 hours

2. Aspirin : 100 mg/kg/day, oral dose, during acute phase, then tapered to low dose of(5 - 10 mg/kg/day) after fever subsides. Afterwards, continue the low dose aspirin for at least 2 months.

3. Supportive care: control fever, give adequate hydration, and use cream or lotion to minimize lips fissuring or bleeding.

After discharge

1. Body temperature monitor

- When fever recurs : give adequate fluid and use warm water bath to control fever.
- Ice pillow is allowed for children older than 6-months -old.
- Return to OPD if the recurrent fever continues for 24 hours

2. Lips and oral protection

- Avoid hot, spicy or stimulating food
- Gentle cleaning of oral cavity with gauze
- Lipstick for lips fissuring

3. Comfortable environment and a temperate daily activity

- clean and quite environment
- observe any joint pain or gait limping
- Observe quality of sleep: interruption or being easily awakened
- any petechiae or echymosis?

4. Food : soft and balanced diet, more fresh fruits, more vitamin. C, low fat and low cholesterol

5. Cautions when taking aspirin

- Continue for at least two months

- Longer period may be required if complicated with coronary aneurysms
- When an epidemic of chicken pox or influenza occurs: withhold aspirin and return to OPD for other alternative medications

6. Changes of Vaccination Schedule

After high-dose of IVIG therapy, vaccination schedule should be postponed for at least 6-9 months. Please consult OPD Drs. for detail.

7. Consult OPD Drs. for individual planning of long term follow-up.

8. For patients complicated with moderate to giant coronary aneurysm, the family need to watch out for any earliest signs of heart failure or angina such as palor, dyspnea, or chest pain

9. Consult your doctor when the followings occur:

Any adverse effect of medications

10. Recurrence of Kawasaki disease (1 to 3 %), primarily presents with relapsing fever.

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