KAWASAKI'S DISEASE

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Abstract

Kawasaki's disease is otherwise called as mucocutaneous lymph node syndrome' is a rare childhood illness that harm the coronary arteries. Kawasaki disease is caused by streptococcal/staphylococcal infection. It is manifested by fever, non puritic rashes, conjunctival infection, strawberry tongue, erythematous hand and periungual desquamation. It is treated with IV immunoglobulin and NSAIDS.

Key Words: Kawasaki disease, IV immunoglobulin and NSAIDS.

Introduction

Kawasaki's disease was first described by Tomisaku Kawasaki in 1967, it isotherwisecalled as–Acute febrilemucocutaneous lymph node syndrome‖. It was initially thought to be a self-limiting benign illness, Kawasaki Disease (KD) has now been found in all parts of the world and in all races. It is a disease of unknown etiology that most frequently affects infants and children under 5 years of age. However, till very recently, reports from India had been few and farbetween.[1]

Definition

Kawasaki disease or mucocutaneous lymph node syndrome is a rarechildhood illness that affects the blood vessels. The symptoms can be severe for several days and can look scary to parents. But then most children return to normal activities.[1]

Kawasaki disease can harm the coronary arteries, which carry blood to the heart muscle. Most children who are treated recover from the disease without long-term problems.

Incidence

It primarily affects infants and young children, with 80-85% of the subjects being below 4 years of age. Boys are affected more commonly than girls and there is an ethnic bias of children belonging to Asian (Japanese andKorean)

It is reported that annual incidence is in the range of 10-100 per 100,000 children below 5 years of age. In Japan 5000-6000 new cases of Kawasaki Disease are diagnosed everyyear.[2]

Etiology

Kawasaki Disease may be triggered by a streptococcal/staphylococcal toxin which acts as a superantigen and causes widespread immune activation. Reports of rickettsiae, retroviruses, house-dust mites and skin commensals are possible causative agents. Other possible and triggering factors include exposure to rug shampoo, mercury poisoning and residence close to a body of water.[3]

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Clinical Manifestation

- A fever lasting at least 5days.
- Redeyes.
- Swollen, red, cracked lips andtongue.
- Swollen, red feet andhands.
- A non-pruriticrash.
- Painful brawny edema of palms andsoles
- Peripheralgangrene
- Cervical adenopathy
- Arthralgia/Arthritis
- Severe abdominalpain
- Sterileurethritis
- Hepaticdysfunction
- Asepticmeningitis
- Penumonia andDiarrhea[3-4]

Criteria for Diagnosis of Kawasaki Disease

- Fever lasting for at least 5days.
- Presence of 4 of the following 5conditions:

1. Bilateral conjunctivalinfection.
2. Changes in the mucosa of the oropharynx, including infected pharynx, dry fissured lips, strawberrytongue.
3. Changes of the peripheral extremities, such as edema or erythema of hands or feet, desquamation usually beginning periungually.
4. Rash, primarily truncal, polymorphous but non-vesicular.
5. Cervicallymphadenopathy.

- Illness not explained by other known disease process.[3-4]

Diagnostic Evaluation

Cardiovascular:

- On auscultation, gallop rhythm or distant heart sounds, ECG changes includes arrhythmias, abnormal Q waves, prolonged PR or QT intervals, occasionally ST-T–wavechanges
- Chest X-ray abnormalities(cardiomegaly)
- Echocardiography (pericardial effusion, coronary aneurysms, or decreased contractility) mitral or aortic valvular insufficiency and rarely, aneurysms of peripheral arteries, angina pectoris or myocardialinfarction

Gastrointestinal: Diarrhea, vomiting, abdominal pain, hydrops of gallbladder, paralytic ileus, mild jaundice, mild increase of serum transaminase levels and elevated liver enzymes.

Blood: Increased erythrocyte sedimentation rate, leukocytosis with left shift, positive C-reactive protein, hypoalbuminemia, and mild anemia in acute phase of illness (thrombocytosis in subacute phase)

Urine: Sterile pyuria of urethral origin and occasional proteinuria

Skin: Perineal rash and desquamation in subacute phase and transverse furrows of fingernails (Beau‘s lines) during convalescence

Respiratory: Cough, rhinorrhea, and pulmonary infiltrate

Joint: Arthralgia and arthritis

Neurological: Mononuclear pleocytosisin cerebrospinal fluid, striking irritability, rarely facial palsy, Anti-neutrophil cytoplasmic antibodies (ANCAs) andanti-

endothelial cell antibodies (AECAs) may be present.[4]

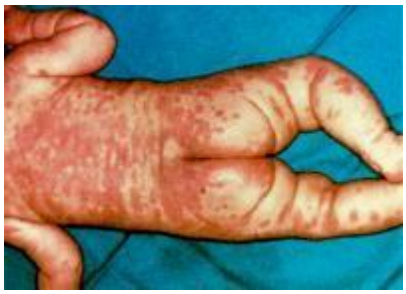

Fig1: Rash of Kawasaki disease

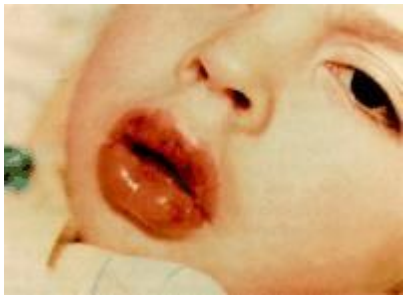

Fig 2: Conjunctival infection, lip edema

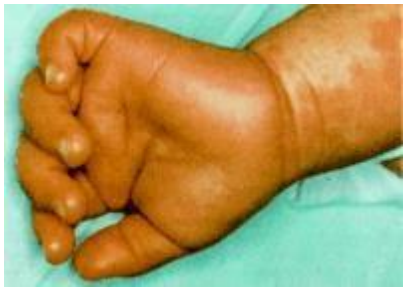

Fig 3: Erythematous and edematous hand

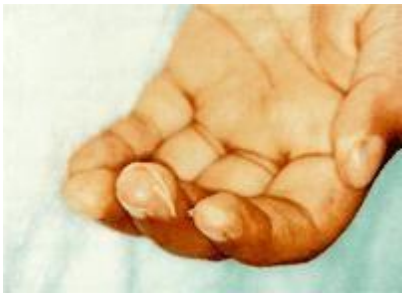

Fig 4: Periungual desquamation

Management

The goals of pharmacotherapy in Kawasaki disease are to reduce inflammation and platelet activation. Early and aggressive intervention improves outcome. Standard treatment includes intravenous immunoglobulin (IVIG) to treat inflammation and to prevent consequences of coronary artery disease and also it is an advantage to combine IVIG with corticosteroids, such as to reduce the incidence of formation of coronary artery aneurysms.

Immunoglobulin: IVIG is generally recommended as first-line therapy. It neutralizes circulating myelin antibodies by means of anti-idiotypic antibodies and promotes remyelination. It may increase cerebrospinal fluid IgG levels.[5]

Non- steroidal anti inflammatory drugs (NSAIDS): These agents inhibit prostaglandin synthesis, which prevents formation of platelet-aggregating thromboxane A2. Children with coronary artery disease have received aspirin for prolonged periods.

Aspirin :It is used to decrease inflammation, inhibit platelet aggregation and improve complications of venous stasis and thrombosis. It is first-line therapy, along withIVIG.

Anti-platelet: Dipyridamole is a platelet-adhesion inhibitor that possibly inhibits red blood cell uptake of adenosine, itself an inhibitor of platelet reactivity.

Corticosteroids: Corticosteroids have anti-inflammatory properties and cause profound

and varied metabolic effects. Corticosteroids modify the body's immune response to diverse stimuli. Prednisolone is indicated for the treatment of steroid-responsive inflammation of the palpebral and bulbar conjunctiva, cornea, and anterior segment. It decreases inflammation by suppressing migration of polymorphonuclear leukocytes and reversing increased capillary permeability.

Immunosuppressant: Infliximab may be added to treatment if steroids and other immunosuppressant drugs are ineffective in achieving or maintaining remission. It reduces the infiltration of inflammatory cells.[5-6]

Conclusion

On conclusion of Kawasaki‘s disease, it is difficult to diagnose in early stage for children and adults. In India, the cases of Kawasaki disease are rare but it causes acquired heart disease during adulthood. It is, therefore, imperative for the pediatrician to diagnose and treat Kawasaki Disease and should be considered in the differential diagnosis of all febrile illnesses in young children where the fever persists for more than 5-7days.[7]

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