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Case Report Kawasaki Disease

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Abstract: Kawasaki disease is a major cause of heart defects that are often found in children. In Indonesia, this disease is still rarely diagnosed because it is still considered rare and not yet widely known. The following two case reports are case reports of boys and girls aged 6 months and 1 year respectively. Both come with persistent fever for more than four days, rashes on the skin, diarrhea, cracked red lips. Patients with the first case got red eyes, peeled skin peeled, swollen neck and swollen legs, reddish tongue but not until like strawberry, the patient was later diagnosed as Kawasaki disease on laboratory examination to get an increase in C-reactive protein, Blood Sedimentation Rate and Thrombocytosis accompanied by a picture echocardiography in case 1 was Dilated A. Right coronary and minimal Pericard effusion. Whereas the second case was accompanied by nausea vomiting, cracked red lips and a strawberry-like tongue, the patient was later diagnosed as Kawasaki disease on laboratory tests to obtain an increase in C-reactive protein, Blood Sedimentation Rate and Thrombocytosis with echocardiographic features of TR Light and mild dilatation RCA . Both children were given intravenous immunoglobulin (IGIV) at a dose of 2 grams / KgBB single dose and aspirin 25 mg 1x1 while in the case of 2 aspilet a dose of 20 mg 1x1.

1. Introduction

Kawasaki disease is defined as a systemic inflammatory disease in children that causes coronary artery aneurysms, Miocardium infarction, and sudden death 4). Kawasaki disease is an acute vasculitic syndrome that predominantly affects small and medium arteries. Epidemiological data show that the genetic aspects of children underlie the pathogenesis of this disease 5). This disease is still very rarely diagnosed in Indonesia because it is considered still rare and not widely known. Kawasaki disease is diagnosed based on classical criteria that have existed since 1967 6).

Kawasaki disease (also known as lymph node syndrome, mucocutaneous node disease and pediatric polyarteritis) is a vasculitic disease and 80% of patients are toddlers, which affect many organs, including the skin, mucous membranes, lymph nodes, and blood vessel walls. The most serious effect is on the heart which can cause blood vessel dilatation or aneurysm. Without treatment, death can be close to 1%, which usually occurs within 6 weeks of illness 1).

2. Case Report

A child aged 6 months 4 days came to the Emergency Unit of RSI Jemur Sari Surabaya with complaints of continuous fever for 9 days accompanied by red lips, cracks, red tongue, red eyes, peeling skin, swollen neck, swollen feet and diarrhea. Redness on the skin or rash appears on the 10th day together with the heat beginning to fall. There was no history of complaints of reddish spots or blisters on the child, but a history of allergies was found in the mother and father. In the history of the history of the disease in getting the initial symptoms of the body feels hot, fussy children, have been brought to the doctor to get anti-inflammatory drugs. On the second day diarrhea arises, the body is still hot, red eyes, red spots come out in the area around the waist and buttocks and laboratory tests are still within normal limits. On the fourth day the eyes are still red but there is no dirt, the body is still hot, the red spots are still there, the skin of the foot has been peeled off, repeated laboratory tests have been found to increase neotrophils and eosinophils. On the tenth day the whole body appears red and the heat has begun to fall. On the eleventh day the patient entered the hospital with a physical examination found that the general condition of the child seemed fussy. On examination the subfebric temperature is accompanied by conjunctival injection, fissura lips, erythematous, and hyperemic pharynx. Other physical examinations show dorsum manic edema accompanied by exfoliation of the skin of the fingers and toes. Complete laboratory examination in these children showed an increase in basophils (1.564%), an increase in monocytes (8.840%), an increase in erythrocytes (5.10 jt / uL), a

decrease in erythrocyte index (MCV 68.9 fL, MCH 21.8 pg, MCHC 31 , 6%), thrombocytosis (820 thousand / uL), decrease in MDV (4,198 fL), increase in LED (52 mm / hour) and increase in CRP (22 mg / L). In these children no leukocytois, anemia, SGOT / SGPT is normal, Buns creatinine is normal, ASTO immunoserological examination is within normal limits.

On the 11th day of the day, electrocardiography was performed with the results of right coronary A dilatation and pericardic pleural effusion. Re-examination of the laboratory, found basophils had begun to decline from before (1.503%), monocytes had begun to decline from before (5.014%), erythrocytes were normal (4.90 jt / uL), erythrocyte index (MCV 68.7 fL had begun to fall than before, MCH 22.2 pg increased than before, but still not normal) MCHC was normal (32.3%), still thrombocytosis (802 thousand / uL), there was still a decline in MDV (4,235 fL increased from before, but still not normal), LEDs are increasing (53 mm / hour). this child then received intravenous immunoglobulin (IGIV) therapy 2gr / KgBB 6-8 hours, and aspirin 1x25 mg, MPS 18 mg 1x1 for 5 days.

On the 14th day of the illness, a repeat laboratory examination with basophils increased (1.995%), decreased neutrophils (5.87%), increased lymphocytes (87.790%). Decreased eosinophils (0.460%), normal monocytes (3.878%), normal erythrocytes (5.07 jt / uL), reductions in the erythrocyte index (MCV 68.2 fL decreased more than before, MCH 21.9 pg decreased from before) MCHC is normal (32.0%), thrombocytosis increases (946 thousand / uL), MDV decreases (4,147 fL), LED increases (62 mm / hour), CRP decreases (10 mg / L). Therapy is considered successful if the re-laboratory examination shows a decrease in CRP value, in this patient it drops to 10 mg / L and there is no fever until the 14th active day for 2 consecutive days, besides the normal leukocyte value is 9.26 thousand / uL. after treatment for 8 days the sufferer then returns home with repairs and planned routine controls.

3. Case Report 2

A 1-year-1-month-old boy comes to UGD Jemur Sari Surabaya Hospital with complaints of fever that lasts for 5 days continuously accompanied by the onset of rashes, vomiting and diarrhea. On physical examination, the subfebric temperature, strawberry tongue, reddish oral mucosa, and red lips are cracked. The results of the investigation showed an increase in basophils (1.845%), an increase in neutrophils (70.48%), a decrease in eosinophils (0.228%), an increase in LED (90 mm / hour) and an increase in CRP (116.7 mg / L). In this child there is no leukocytosis, anemia, and thrombocytosis. Complete urine examination found urine +1 leukocytes, microscopic examination (increase in 1-3pl leukocytes, increased epithelial cells 1-3), no negative proteinemia and bacteria were found.

Echocardiography of patients shows mild TR results and mild dilatation of RCA. Patients then begin treatment with Gamaras 2 g / kg body weight / 12 hours and aspilet 4x150 grams. On the 10th day of the illness a repeat laboratory examination was performed to assess the progress of therapy, a decrease in CRP values from previous (51 mg / L), basophils increased from before (2.004%), normal neutrophils (31.63%) previously increased, lymphocytes increased (50,060%) previously normal, normal eosinophils (3.675%) which had previously decreased, the monocytes increased (12.640%) which were previously normal, there was a decrease in the value of LEDs from before (60 mm / hour). Patients are then allowed to go home and control routinely to a pediatrician

4. Discussion

This disease was first described in 1967 by Dr. Tomisaku Kawasaki in Japan, so-called Kawasaki Disease. So far the highest incidence of Kawasaki disease has occurred in Japan (175 per 100,000 children). A hypothetical pathogenesis of Kawasaki disease is a "protein homeostasis system", that is, after an infection by an unknown pathogen, the resulting pathogen protein spreads and binds to endothelial cells of the coronary arteries as the main target cell 2). This disease affects boys with a ratio of 3: 2 and 76% of children under the age of 5 years 6). The incidence of Kawasaki disease has

increased in recent years, can affect all ethnicities and races in the world, but the high incidence in Asian races shows genetic predisposition and its interaction with the environment. In Japan the incidence of this disease is 218.6 per 100,000 in children aged 0-4 years 6), while data in Indonesia shows an estimated incidence of Kawasaki disease is 6,000 cases per year, but those diagnosed are less than 100 cases per year 7). Kawasaki cases in Indonesia are not small, every year there will be 3300-6600 cases of Kawasaki Disease. But in reality the detected cases are still very far under this angaka. Detected about 20% -40% of them have damage to the coronary arteries of the heart. Some will recover but others will be forced to live with a heart defective due to impaired coronary blood flow. A small percentage will die from heart damage.

The journey of Kawasaki disease can be divided into 3 phases, namely the acute, subacute and convalescent phase. The acute phase lasts 10-14 days, characterized by the onset of fever, conjunctivitis, lymphadenopathy, pleomorphic rash, erythema, and neck edema. This phase can also be accompanied by heart problems such as carditis characterized by tachycardia, S3 gallop, or signs of heart failure and liver disorders. The subacute phase takes place on the 2nd week until the 4th week. This phase is characterized by desquamation of the skin of the finger and perineum, and arthritis in one or more joints. Coronary artery aneurysms usually occur in this phase, but can appear earlier. Fever will decrease from the 3rd and 4th weeks accompanied by healing of the organs involved in the acute phase and the return of the normal platelet value. The last phase is the convalescent phase which lasts for months to years. Affected blood vessels undergo healing, remodeling, and scarring 6). Laboratory results on Kawasaki can be in the form of leukocytosis with neutrophilia. Increased erythrocyte sedimentation rate and CRP, anemia, abnormal plasma lipids, hypoalbuminemia, hyponatremia, thrombocytosis after the first week of illness, sterile pyuria, increased liver enzymes, increased γGT, cerebrospinal fluid pleocytosis, and leukocytosis in synovial fluid.

The diagnosis of Kawasaki disease is made when there is a fever that lasts for at least 5 days and is accompanied by at least four symptoms of nonpurulent conjunctival injection, lip and oral cavity changes such as eritrema, dry and cracked lips, pharyngeal injection, strawberry tongue, edema or hand erythema and foot in the acute phase, periungual desquamation, polymorphic rash, and unilateral ≥1.5 cm cervical adenopathy that is not caused by other diseases.

Patients who only meet 2-3 classic criteria, are included in incomplete or atypical Kawasaki disease. incomplete Kawasaki disease is Kawasaki disease with classic clinical symptoms that do not meet the criteria. Cervical lymphadenopathy is most often not found (90% of cases), rash (50% of cases), and changes in extremity (40% of cases), whereas changes in mucous membranes are most often found in atypical Kawasaki disease. the diagnosis of atypical Kawasaki disease is based on laboratory tests and echocardiography.

If the fever is accompanied by 2 or 3 typical symptoms that arise for 5 days or more and the characteristics of the patient indicate the possibility of Kawasaki disease, do a CRP and LED examination. If the CRP is <3 mg / dL, and the LED is <40 mm / hour, the child is monitored and therapy is carried out properly. If the CRP≥ is 3 mg / dL and LED is ≥ 40 mm / hour, additional laboratory tests should be carried out including albumin, alanine aminotransferase (ALT), thromboids, leukocyte type counts, and urine. Abnormal limits are albumin ≤ 3g / dL, anemia according to age, increase in ALT levels, platelets ≥ 450,000 mm3 (after 7 days), leukocytes ≥ 15,000 mm3, leukosituria ≥ 10 / lpb. If there are 3 additional positive laboratory criteria, a diagnosis of Kawasaki disease can be made. Children should be carried out with echocardiography and treated. If <3 additional laboratory criteria are positive, cardiac echocardiography needs to be done. If negative but the fever continues, repeat echocardiography needs to be done. If the echocardiography is negative and the fever decreases, Kawasaki disease cannot be established. If the positive echocardiography of the child is treated as Kawasaki disease. In the first case and history, there is persistent fever for 9 days, reddish eyes, changes in the lip mucosa, in the form of dry lips, cracking and redness, and changes in the swollen extremities, redness of the hands, feet, and joints of the fingers. . Physical examination found no strawberry tangue, lip mucosal eritrema, cracked lips, and palms eritrema and desquamation. In this patient there is no enlarged neck or red spots around the anus and genital area. Diarrhea patients and their parents have allergies. Investigation at the beginning was found to increase basophils, increase in monocytes, increase in erythrocytes decreased erythrocyte index, thrombocytosis, decrease in MDV,

increase in LEDs and increase in CRP. In this child not found leukocytois, anemia, SGOT / normal SGPT, normal Bunin creatinine, ASTO immunoserological examination within normal limits, the patient entered into the algorithm to do additional examiner examination and echocardiography. On the 11th day of the day, electrocardiography was performed with the right A coronary dilatation results and minimal pericard effusion. Re-examination of the laboratory, found that basophils had begun to decline, monocytes had begun to decline, erythrocytes were normal, the erythrocyte index had begun to fall, MCHC was normal, still thrombocytosis, there was still a decline in MDV, LEDs were increasing. Patients were then treated as Kawasaki disease with a combination of intravenous immunoglobulin (IGIV), aspirin and MPS for 5 days.

In the second case there was fever for 5 days with strawberry tongue, broken lips, nausea and vomiting and no edema in both extremities. New sufferers meet 3 classic criteria for Kawasaki disease. And from the laboratory results do not get anemia, leukocytosis, thrombocytosis, based on the algorithm, the patient is treated as incomplete Kawasaki disease.

Patients with Kawasaki disease who have been upright must be treated at the hospital with the aim of observation, monitoring heart function, and managing clinical manifestations. The goal of therapy to be achieved is to prevent long-term sequelae, especially coronary artery abnormalities 5). Outline management of Kawasaki disease includes acute phase management, management of acute phase therapy failure, and management beyond the acute period 4), 6). Acute management is given a combination of aspirin and intravenous immunoglobulin (IGIV). Aspirin is given at a dose of 80 mg / kg / day divided into 4 doses. The duration of aspirin varies, aspirin can be given until fever is free for 48-72 hours, but some experts give it until the 14th day and the fever has dropped for 48-72 hours 8). Intravenous immunoglobulin is given with a single dose of 2 g / kg for 8-12 hours 6), 8), 10). The best time for this combination is in the first 10 days of illness 4-6), 8-10). High-dose aspirin is given to obtain anti-inflammatory effects in addition to the effects of antithrombosis. Amping effects that may arise, namely hepatitis in drug induction, transient hearing loss, and Reye's syndrome 5). Intravenous immunoglobulin was first given to Kawasaki patients in 1984, but the exact mechanism of IGIV in Kawasaki disease is unclear 6), 10). It is thought to be an antibody for infectious agents, toxins, blockade of fc receptors, accelerate clearance of complement fragments, disrupt the solubility of immune complexes, increase suppressor T cells, inhibit cytokine formation, and induce lymphocyte and neutrophil apoptosis 6). Side effects vary for each individual, the most frequent effects are dizziness 6), 8). As many as 85-95% of patients respond to this combination, the patient then enters the stage of therapy beyond the acute phase, ie giving low-dose aspirin 3-5 mg / kg is given once a day for 6-8 weeks. In the absence of a response, the patient enters the management of acute phase therapy failure 8). Treatment of failure therapy can be given a second dose of IGIV, corticosteroids or monoclonal antibodies 4), 11). corticosteroids are the main therapy in vasculitis, so logically it can also play a role in Kawasaki disease but in fact the use of corticosteroids in the initial phase of Kawasaki disease therapy is still controversial 4). Preliminary studies of corticostreroids showed administration of oral prednisolone at a dose of 2-3 mg / kgBW / day for two weeks, the incidence of coronary artery aneurysms decreased, but the study was limited in terms of the stratification of subjects and methods carried out 4), 8). Newburger et al 8), in their study showed no significant differences in duration of fever, duration of treatment, incidence of therapeutic failure, and size of coronary artery with Kawasaki disease patients who were added with steroids in a combination of IGIV and aspirin regimens with no steroid added 12). A metaanalysis study concluded that the combination of IGIV and aspirin remained the standard therapy in Kawasaki disease. Corticosteroids can reduce the incidence of coronary artery aneurysm when combined with aspirin. But additional research is still needed to examine when corticosteroids are given in combination with IGIV and aspirin 12). At present corticosteroids are only given if Kawasaki disease does not respond to IGIV and aspirin 3).

In both cases, aspirin was administered at a dose of 25 mg / kg body weight / day in the second case with a dose of 25 mg / day. And IGIV 2 / kgBB, this combination can be given to patients with persistent fever without known other causes or ongoing aneurysms or inflammation, marked by increased LED or CRP 3) 7) 8). In one study reported the use of IGIV in children with Kawasaki disease accompanied by aneurysms during echocardiography 14).

After IGIV therapy, usually fever will go down followed by the disappearance of the rash, mucositis, conjunctivitis 6). For short-term evaluation, the success of acute therapy in Kawasaki disease is assessed by loss of fever and a decrease in the value of inflammatory markers within 48 hours after administration of immunoglobulin. The inflammatory markers used to evaluate are CRP, but LEDs cannot be used as a marker of improvement in inflammation in Kawasaki disease 11). In both cases on the 14th day of illness, when the patient has had 2 days free of fever since IGIV was given a CRP re-examination with decreased results which showed the response to acute phase therapy in this patient was very good. Patients then enter the stage of therapy outside the acute phase, which is giving aspirin 3-5 mg / kg for 6-8 weeks.

Long-term evaluation and monitoring of Kawasaki sufferers are primarily aimed at the possibility of coronary artery aneurysms and other cardiac complications. Coronary artery aneurysms occur most often in weeks 2 to 8 of the disease, so based on this the American Academy of Pediatrics recommends performing echocardiography when first diagnosed and repeated at 6 to 8 weeks from the first onset of illness 5), 8), 15). In patients who, at 1-2 months after the onset of illness, do not find a coronary aneurysm on echocardiography new coronary lesions are usually not found 6). Patients who experience improvement after administration of IGIV have repeated clinical examinations during the first 2 months to detect possible heart problems such as arrhythmias, heart failure, and myocarditis 6). After the first 2 months, then follow-up depends on the condition of the coronary arteries, in patients who have not found coronary artery aneurysms follow-up at intervals of 5 years 5), 6), 8). In both cases, routine control was planned for the first 2 months and the 6-8 week echocardiography examination was performed after illness. It was concluded that atypical Kawasaki disease is still very rarely diagnosed in Indonesia. Giving a combination of IGIV and aspirin responds well to Kawasaki disease.

5. Conclusion

There is no known method of prevention for Kawasaki disease. Prevention done to avoidworsening of coronary damage. Parents of children with disease Kawasaki with coronary abnormalities, stressed about the need for action further, namely taking medication regularly and monitoring of heart conditions. Monitoring of post-disease sufferers Kawa saki, especially with history severe coronary aneurysm, performed long term maybe even lifetime. The coronary aneurysm light in general will experience resolution in a few months.

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7. Referencess

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