Case report: corticosteroid as an alternative therapy for Kawasaki disease

Elok Izawati1*, Sandhi Parwata2, Pande Tiara Maharani3

ABSTRACT

Background: Kawasaki disease is an acute febrile illness with systemic vasculitis and predominantly causes acquired heart disease. The etiology is still unknown, and there is no specific diagnostic test. The diagnosis was made using clinical criteria, laboratory and echocardiographic findings. It is characterized by fever, bilateral non-exudative conjunctivitis, erythema of lips and oral mucosa, rash, cerebral lymphadenopathy, edema and erythema of the hand and foot. The initial treatment for both complete and incomplete Kawasaki disease is a single high dose of intravenous immunoglobulin (IVIG) and aspirin. The place of corticosteroids in the initial therapy is still controversial.

Case Description: A 3-year-old boy presented with a 7-day history of high fever and not relieved by antipyretic, which was followed by a maculopapular rash on his trunk and extremities, vomiting, reduced appetite, red on her lips and eyes who aspirin, IVIG and corticosteroid treated as initial therapy. He showed clinical improvement and did not have cardiovascular complications.

Conclusion: We report a case of Kawasaki disease in a 3-year-old boy. This case highlighted that adding corticosteroids to IVIG and aspirin as initial therapy might improve outcomes in Kawasaki disease and could be selected as a treatment modality to treat Kawasaki disease.

Keywords: Kawasaki disease, febrile, systemic vasculitis, corticosteroid.


INTRODUCTION

Kawasaki disease (KD) is an acute febrile illness with systemic vasculitis and is predominantly a cause of acquired heart disease in children. Kawasaki disease was first reported in Japan by Tomisaku Kawasaki in 1967 and became a worldwide problem.1 It mostly affects younger children than 5 years old, with a yearly incidence of 80 to 90 per 100,000 children in Japan and 10 per 100,000 children in the United States.2

The etiology of Kawasaki disease is still unknown, and there is a wide hypothesis to explain this disease. Some hypotheses suggest infection, autoimmune and genetic factors are causing KD.3 There is no specific diagnostic test. The diagnosis was made using clinical criteria, laboratory and echocardiographic findings. Kawasaki disease is characterized by high fever, remitting and unresponsive to antipyretics, and persisting at least 5 days, with minimal four of five classic criteria of Kawasaki disease including bilateral non-exudative conjunctivitis, erythema of lips and oral mucosa, rash, cerebral lymphadenopathy, and edema and erythema of the hand and foot. It is a self-limited disease, and most patients reported full recovery. However, if left untreated, it can lead to cardiovascular complications, including coronary artery aneurysms.4

The goal of therapy in the acute phase is to reduce inflammation, arterial damage and to prevent thrombosis in those with coronary artery abnormalities. The initial treatment for both complete and incomplete KD is a single high dose of IVIG together with acetylsalicylic acid. Intravenous immunoglobulin should be given within the first 10 days of illness onset, as soon as the diagnosis can be established.5 However, not every hospital in Indonesia has available IVIG.

Corticosteroids were used as the initial therapy for Kawasaki Disease long before the first report of IVIG efficacy suggested that steroid therapy was associated with the possibility of aggravation of coronary artery lesions.6 Shinohara et al. found that treatment regimens that included prednisolone were associated with significantly shorter fever duration and a lower prevalence of coronary artery aneurysms.7 A randomized controlled trial, involving eight trials and 1887 participants by Green et al. l 9 showed corticosteroid treatment reduced the subsequent occurrence of coronary artery abnormalities without serious adverse events and mortality. In addition, corticosteroids reduced the duration of fever, time for laboratory parameters (erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) to normalize and length of hospital stay.7,8 This case report describes a Kawasaki disease that responds to corticosteroids as an alternative treatment.

CASE REPORT

A 3-year-old boy presented with a 7-day history of a high fever that was not relieved by antipyretic, which was followed by a maculopapular rash on
illness with systemic vasculitis that causes acquired coronary heart disease in children. Kawasaki disease is diagnosed by clinical criteria, fever of at least 5 days followed by minimal four of five principal clinical features, and our patient presented with prolonged duration of fever and not responding to antipyretic and antibiotic. On physical examination, he looked irritable, and his temperature was 40°C with non-purulent conjunctivitis, cracked and red lips, redness on his palm and sores, strawberry tongue, a cervical lymph node measuring 0.5 cm and vomiting. In our patient, the diagnosis of complete Kawasaki disease was made based on the presence of > 4 principal criteria and with some laboratory support.

The treatment in the acute phase is to reduce inflammation and the risk of coronary aneurysms from 25% to 4%. Therapy is optional when IVIG is given within 10 days of the illness, and the dose is 2 gr/kg over 10-12 hours, together with aspirin. Many clinicians use high-dose aspirin (80-100 mg/kg/d orally in 4 divided doses) in the acute phase for anti-inflammatory effects. It continues until day 14 of illness or until the patient is afebrile for 48-72 hours and continues with a low dose of aspirin 3.5 mg/kg/day for 6-8 weeks until repeated echocardiographic testing is performed. We also treated our patient with high-dose aspirin followed by low-dose aspirin after his fever resolved. For patients with coronary abnormality or aneurysms based on echocardiographic, aspirin should be continued until the aneurysms resolve and our patient doesn’t have any cardiac abnormality on echocardiography.

Although corticosteroids are a treatment choice in other forms of vasculitis, their use has been controversial for children with KD. Corticosteroids were used as initial therapy for Kawasaki disease before the first report of IVIG efficacy. However, corticosteroids have been occasionally used more early as the second-line therapy for patients who is unresponsive to IVIG treatment, in combination with IVIG as the routine first-line therapy for all patients, or as the first-line therapy in combination with IVIG for selected patients at high risk of unresponsiveness to initial IVIG.

Treatment of Kawasaki disease on acute phase with methylprednisolone IV combinations with Aspirin and IVIG than with aspirin and IVIG only, resulted in faster resolution of fever, more rapid improvement in markers in children with acute KD. In contrast, a randomized study by Newburger et al. reported pulsed intravenous methylprednisolone to conventional therapy in the routine primary treatment of Kawasaki disease does not improve coronary artery outcome or reduce the numbers of adverse events, days in the hospital, or days of fever. Methylprednisolone intravenous therapy in our patients shows a beneficial and effective effect. But further studies are needed.
CONCLUSION
Corticosteroids could be selected as a treatment modality to treat Kawasaki disease. As reported in this case, the patient showed clinical improvement after administration of corticosteroid along with other medication.

ETHICS IN PUBLICATION
The patient’s parents agreed to share the clinical information and picture of the patient for publication and education purposes.

CONFLICT OF INTEREST
None declared.

FUNDING
None.

AUTHORS CONTRIBUTIONS
Author EI contributes to the literature review, patient examination, treatment follow-up, manuscript preparation, and publication. Author PTM contributes to patient follow-up, literature review, and translation.

REFERENCES

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