Kawasaki disease: giant aneurysm with a large thrombus of the left coronary artery

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Abstract

We report a six-month-old febrile infant presenting with stridor. Later on, he developed typical Kawasaki disease with giant aneurysm of the coronary artery with thrombosis that resolved with an aggressive anticoagulation therapy. The giant aneurysm still persisted a year later. Respiratory illness with stridor is an unusual presentation of Kawasaki disease.

Introduction

Kawasaki disease (KD) is an acute febrile illness affecting infants and young children, causing vasculitis of the small and medium size arteries, and its etiology is still unknown. Most concerns are the coronary arteries showing aneurysm formation, thrombosis with occlusion, rupture1 and myocardial infarction, which often cause death.

We report a six-month-old infant presented with croup from enlarged retropharyngeal lymphnodes, which is an unusual presentation. Later on, he developed symptoms of typical Kawasaki disease and formed a giant aneurysm and thrombosis of the left anterior descending and right coronary artery.

Case Report

A six-month-old male presented at the Comer Children’s Hospital (Chigaco, IL, USA) with history of fever, cough and stridor. He was treated in an outside hospital with antibiotics without any response. On transfer to our hospital he was noted to have stridor, barking cough and fever. He developed maculopapular rashes over the trunk, dry red swollen lips and bilateral non-purulent bulbar conjunctivitis. Neck x-ray showed retropharyngeal widening. The x-ray findings and the persistent fever required a computed tomography (CT) scan of the neck, which showed several enlarged retropharyngeal lymphnodes. He was treated with Dexamethasone and 3 doses of racemic epinephrine, then the fever defervesced within 12 h. He continued to have stridor at rest with substernal retractions, nasal flaring and remained febrile.

Pertinent laboratory findings are: erythrocyte sedimentation rate (westerngreen), 145 mm/h; C-reactive protein, 84 (nL<5); platelets, 1,244 million; serum albumin, 3.0 gm/dL (nL 3.5-5.0).

An echocardiogram demonstrated dilated coronary arteries with thrombus (Figures 1, 2 and 3); left anterior descending (LAD) artery measured 11×9.4 mm; right coronary artery (RCA) measured 4.1×4.8 mm.

Initially, he was treated with intravenous immunoglobulin (IVIG) 2 gm/Kg and a high dose of aspirin. Because of recurrence of fever and giant aneurysm of the LAD with thrombus, he was treated with a 2nd course of IVIG. He still remained febrile after receiving a course of Remicade (Infliximab), therefore Heparin infusion was started. Later on, he was started on Plavix (Clopidogrel), Warfarin (Coumadin) and Lovenox. During his hospitalization the thrombus within the LAD and RCA dissolved, but the LAD giant aneurysm still remained and the CT scan of the coronary arteries (Figure 4) confirmed the echo finding of giant aneurysm. He was then discharged home with prescription for Lovenox, Aspirin and Plavix.

The follow-up echocardiogram at the age of...
Discussion

In patients with KD, aneurysm of the coronary artery develops in 20% of the untreated patients and in 4-8% of children treated with gamma immunoglobulin. Giant aneurysm (>8 mm) of internal diameter of the coronary arteries is rare and has greatest risk of thrombosis and myocardial infarction, causing sudden death. Therefore, it is recommended a close follow-up of these patients and a very aggressive anticoagulation therapy with multiple agents. Surgical intervention (e.g. coronary artery by-pass) should be considered when the aneurysm size increases significantly in spite of maximal medical therapy.

References

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