Case report

Coronary artery aneurysm in a two-year-old child

Kemal Nisli[1], Murat Ugurlucan[2], Omer A. Sayin[2], Burcu Goker[2], Aygun Dindar[1], Emin Tireli[2], Enver Dayioglu[2]

[1]Department of Pediatric Cardiology, Istanbul Medical Faculty, Istanbul University, Istanbul, Turkey
[2]Department of Cardiovascular Surgery, Istanbul Medical Faculty, Istanbul University, Istanbul, Turkey

Submitted: 30 July 2007
Accepted: 8 October 2007

Arch Med Sci 2008; 4, 3: 342–344

Corresponding author:
Dr. Murat Ugurlucan
Bozkurt Caddesi, No 110-112
Benli Apt., Daire 6
80250 Kurtulus/Istanbul
Turkey
Phone: + 90 535 431 67 86
Fax: + 90 212 534 22 32
E-mail:
muratugurlucan@yahoo.com

Abstract

Kawasaki disease is an acute vasculitis affecting predominantly children. The disease leads to coronary artery ectasia and damage which further result in ischaemic heart disease, being the most common cause of myocardial infarction in childhood. The aetiology and guidelines for the definitive diagnosis are still not identified clearly and the aim of current treatment strategies is to reduce inflammation and prevent ischaemic myocardial damage. In this report, we present the perioperative cardiac images of a two-year-old boy who had giant coronary arteries. He was admitted with the diagnosis of fibrinous pericardial effusion and was referred for pericardiectomy. During surgery we found ectatic coronary arteries.

Key words: Kawasaki disease, coronary artery aneurysm, thrombocytosis, paediatric.

Introduction

Kawasaki disease is an acute vasculitis affecting predominantly children below five years of age. The aetiology and guidelines for the definitive diagnosis are still not identified clearly and the aim of current treatment strategies is to reduce inflammation and prevent ischaemic myocardial damage [1-5].

In this report, we present the perioperative cardiac images of a two-year-old boy who had giant coronary arteries. He was admitted with the diagnosis of fibrinous pericardial effusion and referred for pericardiectomy. During surgery we found ectatic coronary arteries.

Case report

A two-year-old boy was brought to the clinic with high fever, irritability, poor appetite and generalized pain. Physical examination revealed, in addition, enlarged cervical lymph nodes. His symptoms started suddenly and were present for one week. The erythrocyte sedimentation rate and leukocyte level were increased in blood analysis. Chest X-ray showed generalized infiltrations on both lungs and increased cardiothoracic index. Echocardiography indicated pericardial effusion of fibrinous character reaching up to 3 cm at the posterior pericardium.

The patient was referred for pericardiectomy following initiation of empiric antibiotics after obtaining first order blood cultures. Through standard median sternotomy, the pericardial sac was incised. On the epicardium, the coronary arteries appeared severely ectatic (Figure 1),

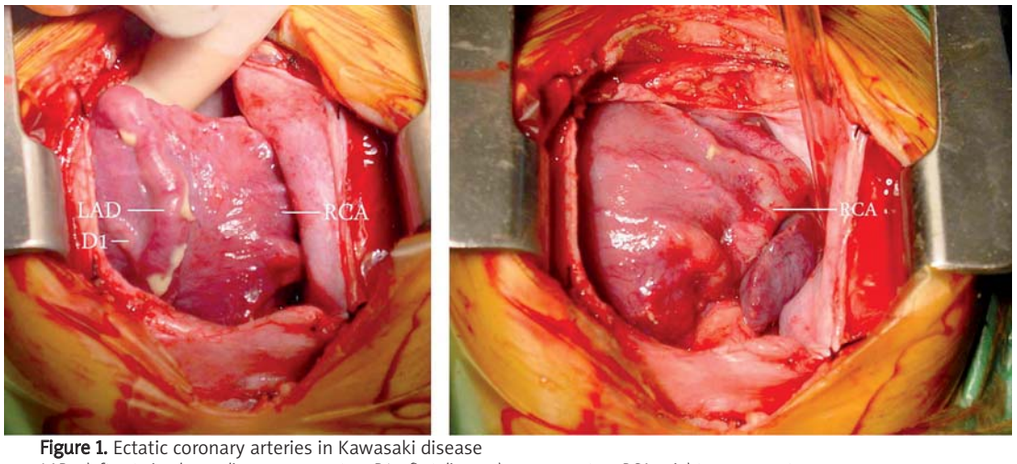

Figure 1. Ectatic coronary arteries in Kawasaki disease
LAD – left anterior descending coronary artery, D1 – first diagonal coronary artery, RCA – right coronary artery

suggesting possible Kawasaki disease. About 250 ml of fibrino-sero-haemorrhagic fluid was drained and pericardiectomy until the level of the phrenic nerves on both sides was performed. In the postoperative period the patient received aspirin in addition to prophylactic antibiotherapy. Postoperative course was uneventful from the surgical point of view and the patient was discharged on the fifth postoperative day to the paediatric cardiology clinic. During the follow-up period repeated complete blood count profile changed to normocytic anaemia as well as leukocytosis (12,300/ml) with polymorphonuclear cell predominance and thrombocytosis (654,000/ml). Erythrocyte sedimentation rate and C-reactive protein levels were also increased. Blood biochemistry (liver and renal functions, levels of lipids) and urinalysis were found to be normal. Blood cultures revealed negative results for microorganisms. Serial daily electrocardiography did not indicate ischaemic changes in the myocardium. All the diagnostic work-up of the patient together with the signs and symptoms and, most importantly, the coronary artery appearance and thrombocytosis indicated Kawasaki disease and the treatment strategy was planned accordingly with aspirin, intravenous immunoglobulin G and immunosuppressive medication with corticosteroids. Symptoms of systemic inflammation relieved with the therapy and the patient was discharged one month postoperatively. He has been followed with echocardiography and has been doing well without relapse for four months.

Discussion

Kawasaki disease, which is also known as lymph node syndrome, mucocutaneous node disease or infantile polyarteritis, is an acute vasculitis seen among children mostly below five years of age [2, 4, 5]. It was first described by a Japanese physician, Tomisaku Kawasaki, in 1967 [1]. The disease affects predominantly the small and medium sized arteries including coronary arteries in 10-18% of affected patients [2, 4, 5], leading to coronary artery giant aneurysms, and hence damage resulting in ischaemic heart disease [6], the most common cause of myocardial infarction in childhood [2].

Initial symptoms, mimicking viral infection, include remittent high fever (usually >39°C) with conjunctivitis, generalized rash, swollen tongue, palms and feet, sore throat and lymphadenopathy followed by peeling of skin on the hands and feet, abdominal and joint pain and gastrointestinal disturbances [3]. Later on complications start to develop.

Although bacteria, viruses, and environmental chemicals or pollutants have been blamed for the aetiology, none has been proven [1-5]. Additionally, it does not have a hereditary basis [1-5]. There are no definitive guidelines for diagnosis, which is based on the clinical signs and symptoms and exclusion of other diseases leading to similar symptoms such as streptococcal scarlet fever, juvenile rheumatoid arthritis, measles, toxic shock syndrome, Steven-Johnson syndrome, *Herpes simplex*, Epstein Barr or *Cytomegalovirus* infections [3].

The vasculitic basis of the disease may lead to severe complications mostly related to cardiac involvement; however, only in a small percentage of children does permanent damage occur. The aim is initiation of treatment as soon as possible in order to minimize the complications. The therapy regime includes aspirin, intravenous immunoglobulin and immunosuppressive agents. Cardiac involvement should be closely monitored; sometimes coronary interventions involving stenting or bypass grafting may be inevitable [3]. Symptoms usually rapidly

regress with therapy and most patients become free of symptoms in six months time. However, they should be followed regularly in order to prevent cardiac ischaemic risks.

The symptoms of our patient were not significant for Kawasaki disease; nor did he have increased thrombocyte levels on admission. However, the coronary artery appearance during pericardiectomy was reminiscent of vasculitis affecting the coronary arteries in early childhood and the whole clinical picture including the laboratory work-up indicated Kawasaki disease in the postoperative term. We believe early initiation of therapy provided better results; hence he was asymptomatic in the follow-up period.

References

1. Burns JC. Commentary: translation of Dr. Tomisaku Kawasaki's original report of fifty patients in 1967. Pediatr Infect Dis J 2002; 21: 993-5.
2. Freeman AF, Shulman ST. Kawasaki disease: summary of the American Heart Association guidelines. Am Fam Physician 2006; 74: 1141-8.
3. Celik U, Alhan E, Arabaci F. Incomplete Kawasaki disease: a pediatric diagnostic conflict. Anadolu Kardiyol Derg 2007; 7: 343-4.
4. Yeung RS. Lessons learned from an animal model of Kawasaki disease. Clin Exp Rheumatol 2007; 25 (1 Suppl 44): S69-71.
5. Newburger JW, Fulton DR. Kawasaki disease. Curr Opin Pediatr 2004; 16: 508-14.
6. Das S, Paul S. Additional openings of the coronary sinus and associated anomalies. Arch Med Sci 2006; 2: 128-30.