Case Report

Kawasaki’s Disease in Remission With Cardiac Involvement: Intrasacular Thrombus in a Giant Aneurism of Both Coronary Arteries. Case Report

Karla Mendiola Ramírez a,∗, Jorge Omar Osorio Díaz b, María del Rocío Maldonado Velázquez c, Enrique Faugier Fuentes d

a Second Year Pediatrics Resident, Hospital Infantil de México, Mexico City, Mexico
b Second Year Pediatric Cardiology Resident, Hospital Infantil de México, Mexico City, Mexico
c Chief of Servicio de Reumatología Pediátrica, Hospital Infantil de México, Mexico City, Mexico
d Attending Physician Reumatología Pediátrica, Hospital Infantil de México, Mexico City, Mexico

A B S T R A C T

Kawasaki disease is of relevance in pediatric practice because it is a systemic vasculitis of unknown origin and the most common cause of acquired heart disease in young patients. Its main complication is the formation of a coronary aneurism in 25% of the patients, unless they receive timely medical treatment.

We report the case of a 4-month-old male child with Kawasaki’s disease, who received treatment with gammaglobulin and acetyl-salicylic acid, in which the initial echocardiogram showed aneurisms. Admitted to our hospital with cardiogenic shock, we documented, by echocardiography, the presence of coronary aneurisms with intrasaccular thrombus and acute myocardial infarction. He received fibrinolytic therapy, with an adequate response: the size of aneurisms decreased, as did the intrasaccular thrombus. Currently the patient is asymptomatic and receiving treatment with warfarin and acetylsalicylic acid.

The diagnosis and treatment, as well as identification of risk factors can prevent fatal complications at the cardiovascular level. The treatment in the first 10 days of illness with gammaglobulin and acetyl-salicylic acid reduced cardiac complications from 4.7% to 25%.

© 2010 Elsevier España, S.L. All rights reserved.

Enfermedad de Kawasaki en fase de convalecencia con afección cardiaca: trombo intra-sacular en aneurisma gigante de ambas coronarias. Reporte de un caso

Resumen

La enfermedad de Kawasaki es de relevancia en la práctica pediátrica debido a que es la vasculitis sistémica de origen desconocido más común y la primera causa de cardiopatía adquirida en pacientes jóvenes. Su complicación principal es cardiaca, ya que el 25% de los pacientes sufre la formación de aneurismas coronarios si no reciben de manera oportuna tratamiento médico.

Reportamos el caso de un niño de 4 meses de edad, con enfermedad de Kawasaki, que recibió tratamiento con gammaglobulina y ácido acetilsalicílico. El ecocardiograma inicial no presenta ectasias o aneurismas. Ingresa en nuestro hospital con datos de choque cardiogénico, se documenta por ecocardiografía aneurismas coronarios, con trombo intra-sacular e infarto agudo al miocardio. Recibe terapia fibrinolítica, con respuesta adecuada: disminución del tamaño de los aneurismas y del trombo intra-sacular. Actualmente el paciente se encuentra asintomático y en vigilancia en la consulta externa, recibe tratamiento con warfarina y ácido acetilsalicílico.

El diagnóstico y el tratamiento oportuno, así como la identificación de factores de riesgo, pueden evitar complicaciones fatales a nivel cardiovascular. El tratamiento en los primeros 10 días de la enfermedad con gammaglobulina y ácido acetilsalicílico reduce del 4.7% al 25% de las complicaciones cardiacas.

© 2010 Elsevier España, S.L. Todos los derechos reservados.
Introduction

Kawasaki’s disease (KD) is of relevance in the pediatric practice because it is the most common systemic vasculitis of unknown origin and the first cause of acquired cardiac disease in young patients. The inflammatory process has a predilection for coronary arteries, leading to the formation of aneurisms, thrombus and stenosis. Aneurisms and coronary ectasia are detected in 15–25% of patients with KD who do not receive treatment and are the main cause of myocardial infarction, ischemia and sudden death.

The peak of mortality in KD occurs between days 15 and 45, after the resolution of fever; during this time there is vasculitis in the coronary arteries, platelet elevation and a state of hypercoagulability.

The American Heart Association (AHA) has published guidelines for the stratification of coronary risk; it divides the disease in 5 groups according to the size of the aneurisms.

Risk factors associated to the formation of coronary aneurisms (CA) are: age under 1 year or over 5, delay in the diagnosis and treatment of disease, an increase of inflammatory markers (ESR, CRP, procalcitonin) after the administration of gammaglobulin, leukocytosis over 30 × 10⁹/l, thrombocytopenia, an increase in liver enzymes and low albumin levels.

Treatment with gammaglobulin before day 10 of disease and aspirin reduces the risk of cardiac complications from 4.7% to 25%. Its objective is to inhibit the production of pro-inflammatory cytokines, metalloproteinases, TNF-α and platelet aggregation, reduction in the formation of CA and early identification of vasoocclusive cardiovascular risk factors may help reduce severe cardiac disease.

Timely diagnosis and treatment, identification of risk factors and close follow-up of patients may help prevent fatal cardiovascular complications.

Clinical Presentation

The patient is a 4-year-old male who was previously healthy. One month prior to his hospitalization he presented upper respiratory tract infection and difficult to treat fever, managed with antipyretics and antibiotics, without a good response.

KD diagnosis was established in a private hospital based on: (1) difficult to control fever lasting more than 5 days; (2) non-suppurative bilateral conjunctivitis; (3) changes in oral mucosa (raspberry tongue); (4) erythema polymorphus and (5) cervical lymphadenopathy. He presented desquamation of the site of BCG vaccine application. On day 8, an echocardiogram was performed showing a healthy heart, the right coronary measuring 1.8 mm and the left one measuring 2.6 mm; there were no aneurisms. Laboratory tests showed: Hb 9.3 g/dl (NR 12–15.5 g/dl), Hto 28% (NR 36%–47%), platelets 197,000 (NR 150,000–450,000), leukocytes 14,300 (NR 6,000–10,000). Harada Score: points. On day 8, the patient was treated with intravenous gammaglobulin at a dose of 2 g/kg/dose and aspirin 100 mg/kg/day for days. He was discharged without any further treatment due to improvement.

From day 12 to day 35 of the disease he presented no fever nor received medical treatment or attention.

He came to our hospital on day 36 of the disease with a 9-h episode of vomiting, abdominal pain, cyanosis, somnolence and shallow respirations. He presented cardiac arrest, metabolic acidosis and shock; after resuscitation for 5 min he achieved sinus rhythm.

He was hospitalized. An electrocardiogram showed sinus rhythm, heart rate (HR) of 136; P axis +45°; QRS axis +80°; PR: 0.11; cQT: 0.36; ST depression on V6; Q wave on DI, DII, aVL and aVR; and left ventricle hypertrophy (Fig. 1a). A chest X-ray showed Situs solitus; levocardia; ICT 0.62; normal pulmonary flow. The echocardiogram showed a mild pericardial effusion on the free wall of the left ventricle, subendocardial ischemia, dyskinesia of the lateral wall of the left ventricle, and paradoxical septal movement. The left coronary measured 2.8 mm; circumflex 2.3 mm, with a fusiform aneurism of 5.8 mm; anterior descending 1.7 mm with fusiform aneurisms measuring 7.5, 8.1 and 11 mm. The right coronary artery measured 2.4 mm, with a saccular aneurism measuring 7.5 mm and an intrasaccular thrombus 0.5 cm². Systolic function showed an ejection fraction of 55%, FA 27%, normal mitral EA and a Tie index of 0.26 (Fig. 1b).

Laboratory results showed: Hb 7.6 g/dl (NR 12–15.5 g/dl), Hto 23% (NR 36%–47%), platelets 379,000/mm³ (NR 150,000–450,000/mm³), leukocytes 7,900/mm³ (NR 6,000–10,000/mm³), albumin 3.1 g/dl (NR 3.5–5 g/dl), TGO 367 U/l (NR 15–50 U/l), TGP 107 U/l (NR 10–40 U/l), DHL 321 U/l (NR 110–295 U/l), procalcitonin 2.44 ng/dl (NR 2.0 ng/dl), ESR 40 mm/h (NR 0–10 mm/h), CRP 5.5 mg/l (NR 0–0.300 mg/l). Urinalysis presented albuminuria, hemoglobinuria, erythrocyturia 10–12/field, abundant leukocytes, granular casts 0–2/field, epithelial cells 2–3/field. Cardiac enzymes, 12 h after admission: AP 195 U/l (NR 150–420 U/l), DHL 1190 U/l (NR 110–295 U/l), CPK Mb 5.173 U/l (NR 0–25 U/l), TGP 352 U/l (NR 10–40 U/l), TGO 269 U/l (NR 15–50 U/l). Lipid profile:
Day 52

Figs. 2. (a) Electrocardiogram showing sinus rhythm, HR 125, aP +30°, QRS axis +60°, PR 0.12, cQT 0.40, Q waves on aVF, and V6. (b) Echocardiogram showing aneurism of the right coronary of 0.1 cm²; left anterior descending coronary of 6.8 mm; origin of both coronaries of 3 mm; EF 61%.

It is no longer possible to obtain the data from the image.
Diagnosis and treatment with gammaglobulin before day 10 of KD can prevent fatal cardiovascular complications.

Conflict of Interest

The authors have no conflict of interest to declare.

Acknowledgement

The authors wish to thank Dr. Abraham Galicia Reyes, cardiologist and electro physiologist, for sharing his expertise on these types of patients.

References