Revista Electrónica de Biomedicina
Electronic Journal of Biomedicine

ISSN: 1697-090X

INTESTINAL PSEUDO-OBSTRUCTION AND TRANSIENT CARDIOVASCULAR ABNORMALITIES IN KAWASAKI DISEASE

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Rev Electron Biomed / Electron J Biomed 2005;2:79-82

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SUMMARY
We describe a 2 year-old boy with severe vasculitis who presented with a typical Kawasaki disease complicated with an intestinal pseudo-obstruction, gallbladder hydrops, myocarditis and transient coronary abnormalities despite early administration of intravenous immunoglobulin treatment.

RESUMEN
Describimos el caso de un niño de 2 años con vasculitis grave que presentó un cuadro típico de enfermedad de Kawasaki complicada con una pseudo-obtrucción intestinal, hidrops vesicular, miocarditis y anormalidades coronarias transitorias, a pesar de la administración temprana de tratamiento con inmunoglulina intravenosa

Keywords: Kawasaki disease, cardiovascular abnormalities, intestinal pseudo-obstruction, abdominal pain, acute surgical abdomen

Palabras Clave: Enfermedad de Kawasaki, Alteracions cardiovasculares, pseudo-obstrucción intestinal, dolor abdominal, abdomen agudo quirúrgico

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INTRODUCTION

Kawasaki disease (KD) is the acute vasculitis responsible for most cases of acquired heart disease in children around the world. Surgical presentation of KD is not uncommon, including children who develop atypical forms of the disease. Recently, there have been reports of KD patients[1-3] who presented as intestinal pseudo-obstruction or acute surgical abdomen pictures, which resulted in delayed diagnosis and treatment. Many of these children consequently developed coronary aneurysms, the most common and fearful complication of KD.

We report a child with typical KD who developed intestinal pseudo-obstruction, gallbladder hydrops, myocarditis, and transient coronary abnormalities despite early intravenous immunoglobulin (IVIG) treatment.

Case Report

A 2-year-old boy presented to our children's hospital with a 5-day history of fever and 2 days prior to admission he progressively developed a maculopapular rash in thorax and genitals, had erythema and edema in palms and soles, bilateral conjunctival injection, swollen red lips, and a strawberry tongue. On the day of admission he developed a distended and painful abdomen, diarrhea, and vomiting.

On examination he was febrile (39.8ºC), heart rate was 180 beats/minute, met 5 major clinical diagnostic criteria for KD, and also presented a diffusely distended tender but soft abdomen, with no rebound sign. He received 1 dose of IVIG [2 g/Kg] and was started on acetylsalicylic acid (ASA) [100 mg/Kg/day]. Because of persistent abdominal distention and vomiting, a plain abdominal radiograph was performed which showed air fluid levels with diffuse small bowel loops distention, all of these consistent with an intestinal pseudoobstruction. An abdominal ultrasound revealed gallbladder hydrops, fluid-filled loops, and intestinal wall edema. He was seen by the pediatric surgeons who recommended conservative management with intravenous fluids and an open nasogastric tube, but no need for surgical intervention. Hemoglobin was 10.4g/dL, leukocytes 9930/mm3, (68% neutrophils), and platelets were 210,000/mm3; C-reactive protein was 322 mg/L, he had sterile pyuria, sodium was 129 mmol/L, potassium 3.5 mmol/L, and ALT and AST values were 68 IU/L and 163 IU/L, respectively. Abdominal distention persisted and the nasogastric tube drained 900 mL during a 24 hour period.

On day 2 of admission, an echocardiogram showed a mild dilatation of the left coronary artery, a moderate left ventricular dysfunction with a fractional shortening (FS) of 20%, for which oral furosemide (10 mg bid) and enalapril treatment were started. He improved dramatically on the same day and started to tolerate oral intake. A repeat echocardiogram on day 5 of admission showed a normal ventricular function with a FS of 30% and normal coronary arteries. He was discharged home with oral ASA, enalapril and furosemide. Two weeks later a new echocardiogram was normal and ASA was prescribed for 1 month more.

Discussion

Intestinal pseudo-obstruction occurs in 2-3% of children with KD[4]. Some of these children present early with dilated coronary arteries, that could be related sometimes with a delay of more than 10 days in IVIG administration[1,5]. However, other children as our patient develop coronary abnormalities despite early IVIG administration[2,4]. Some of the severe abdominal complications including intestinal pseudo-obstruction, may reflect severe vasculitis to some extent, and therefore clinicians should pay especial attention to coronary lesions in children with similar abdominal manifestations as ours[2,4].

This is of special concern in children in whom surgical manifestations develop prior to the onset of typical KD features, as recently described by Akikusa[1] and Zulian[2,3]. In a series by Miyake and colleagues[4], 5 of 7 children who developed intestinal pseudo-obstruction had coronary artery disease. Zulian et al.[2] reported 50% of their patients with surgical-onset KD having coronary artery abnormalities.

Our patient had evidence of multiple organ system inflammation and diffuse vasculitis, as suggested by the presence of myocarditis, coronary abnormalities, gallbladder hydrops, and intestinal pseudo-obstruction. The majority of these patients do not require surgery and improve dramatically after IVIG

treatment[6], although steroids have been used in some of these children[4]. Fortunately, our patient developed only transient coronary abnormalities as confirmed with the repeat echocardiograms.

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El caso clínico, su tratamiento y la evolución de la enfermedad están muy bien descritos y constituyen una buena contribución para los clínicos, pues alertan al Pediatra y al Cirujano infantil sobre una complicación infrecuente en esta enfermedad que es de manejo conservador como está señalado en el artículo.

La publicación de casos como el descrito permiten ir acumulando experiencias que a la fecha son escasas y tener presente esta complicación en niños con enfermedad de Kawasaki que presentan síntomas de abdomen agudo.

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The present paper is a report of a rare complication in Kawasaki disease. Intestinal pseudo-obstruction may occur up to 2-3% as reported in literature[1,2].

It is important to recognize this atypical clinical presentation in order to not to delay the diagnosis and treatment of the disease. Such delay may be responsible of increased rate in coronary complications when this onset is present.

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Received , August 5, 2005
Published August 25, de 2005