

Kawasaki's Disease: About an Unusual Case

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Abstract

Kawasaki disease is the second most common vasculitis in children after rheumatoid purpura. We report a case of a 7-year-old boy with typical Kawasaki disease occlusive syndrome and coronary artery disease. Echocardiography shows a giant aneurysm of the left coronary artery of 9 mm and the right coronary artery 7 mm. This observation highlights the interest of not disregarding Kawasaki's disease in front of all its complete incomplete or atypical forms to delay the diagnosis and the treatment. It emphasizes the importance of giving immunoglobulins as early as possible to prevent coronary involvement.

Keywords: *Kawasaki; Child; Aneurism; Occlusive Syndrome; Immunoglobulins*

Introduction

Kawasaki disease is an acute systemic febrile vasculitis that most commonly affects children under the age of five. It is an adenocutaneous mucosal syndrome which was first described in Japan in 1967. Its diagnosis is easy when it is a complete form, however it is so difficult for incomplete or atypical form. Cardiac involvement conditioned the prognosis. Here, we report an unusual case of a 7-year-old child who presented with giant coronary aneurysms and subocclusive syndrome.

Case Report

A 7 year-old boy, with no specific history, was referred for a prolonged fever associated with a rash. He presented 15 days before his admission a fever at 39°C not yielding to the antipyretics associated with erythematous lesions involving the trunk members and the perineum, abdominal pain associated with five episodes of vomiting and arthralgia. On admission, the examination noted a healthy child but hypersensitive. He had fever at 39°C, a negative urine strip, bilateral non-purulent conjunctivitis, cheilitis, morbilliform erythematous lesions of the trunk and scaly lesions of the perineum associated with edema in the feet. Cardiopulmonary examination was normal. The abdomen was sensitive to palpation in the epigastric region, there was no hepatomegaly or splenomegaly. The lymph node areas were free. There was no meningeal syndrome, no arthritis, and no change of BCG vaccination scar. The diagnosis of Kawasaki disease was done because we had a prolonged fever associated with 4 criteria, cheilitis a non-purulent conjunctivitis an exanthema on the level of the trunk and an attack of the extremities. The biological features showed hyperleukocytosis at 33,000 white blood cells/mm³ with neutrophilic polynucleosis at 30,000/mm³, hypochromic microcytic anemia at 9.1 g/dl with hyperferritinemia at 203 ng/ml of inflammatory origin. A platelet count was normal at 417,000/mm³. An inflammatory syndrome with a globular sedimentation rate increased to 103 mm at the first hour and a CRP reactive protein C increased to 316 mg/L. There was no hepatic cytolysis with AST at 17 U/l or gamma

cholestasis with GT at 10 U/l. Cytobacteriological examination of the urine showed leukocyturia at 48,000/ml without germs. The serum was normal at 135 mmol/l. The chest X-ray was without abnormalities and the ophthalmologic examination did not detect uveitis. Doppler echocardiography showing a giant aneurysm of the left coronary artery up to 9 mm and a giant aneurysm of the right coronary artery of 7 mm.(figure 1).

The management was acetylsalicylic acid 60 mg/kg into 4 doses for one week. Immunoglobulins were given at a dose of 2 g/kg in a single dose over 12 hours. Immediately after, the patient presented with a sub occlusive syndrome consisting of abdominal distension of bilious vomiting, stopping of the stool without stopping the gases and epigastric sensitivity to the abdominal examination. An unprepared abdomen did not notice hydroaeric levels and an abdominal ultrasound was normal. However, the patient was put on digestive rest with a nasogastric tube, an antispasmodic, an antiemetic and gastric protection. The evolution was marked by apyrexia, the disappearance of the occlusive syndrome, the regression of cheilitis and conjunctivitis however a significant palmar flaking of the hands in tatters was installed on day 21 (Figure 2). The dose of acetyl salicylic acid was reduced to 5 mg/kg once a day and an anti-vitamin K treatment was started to treat the giant aneurysm with an INR objective between 2 and 3. The therapeutic balance was obtained with difficulty because we resorted to stopping the anticoagulant treatment before an INR greater than 7 for 5 days and then resuming in small doses once the INR allowed it. At day 21, an echocardiography control showed a dilation of the two coronaries to 7 mm and the CRP decreased to 26 mg/l. The recovery was obtained and ultrasound control was planned after 3 months.



Figure 1: Doppler echocardiography showing a giant aneurysm of the left coronary artery up to 9 mm and a giant aneurysm of the right coronary artery of 7 mm.



Figure 2: Palmar flaking of the hands appeared after 3 weeks of the onset of clinical signs in our patient.

Discussion

Kawasaki disease (KD) is an acute vasculitis that affects the arteries of medium and small caliber with specific tropism for the coronary arteries, it represents the first cause of acquired heart disease in children. Indeed, it has been described all over the world, its incidence reaches respectively 134 cases and 5 cases per 100,000 children aged less than 5 years in Japan and in France [1,2]. In North Africa, only a few sporadic cases have been reported [3,4]. In Morocco, its incidence remains unknown. It prevails according to a seasonal mode with a predominance winter. The etiology of KD is unclear, the most likely hypothesis being the responsibility of an infectious agent as a trigger for the inflammatory reaction in a particular genetic area at risk leading to an inappropriate immunological reaction. Among the agents implicated are *Staphylococcus*, *Streptococcus* producing superantigenic toxins, coronavirus, adenoviruses, EBV virus and HIV [5]. It can also complicate a covid-19 infection. The incrimination of all these agents could be explained by the resemblance of KD with other pediatric infectious diseases and by the seasonal and often epidemic character [6]. KD is more common in boys with a sex ratio of 1.5 [7]. Our patient was male. The diagnosis of KD is clinical based on the following criteria: fever of more than 15 days an exanthema an enanthema an attack of the extremities or of the perineum bilateral conjunctivitis and cervical lymphadenopathies. The degree of association of these signs makes it possible to define three tables: typical KD (fever + 4 or 5 criteria), incomplete (fever + 1 to 3 criteria) and atypical (fever + coronary artery disease) [8-13]. In our case, it was a typical KD with cardiac involvement and an occlusive syndrome. The clinical signs do not all appear at the same time, they are based on the criteria of AHA [7]. Fever is a constant sign, flaking of the hands appears at the third week and means the end of the disease, that of the perineum is strongly suggestive. Our patient had all these signs that appeared during the course of the disease. Some differential diagnoses should be ruled out, namely scarlet fever, measles, staphylococcal shock, autoimmune diseases and drug allergies [3,5]. Cardiac involvement, in particular coronary aneurysms, occurs in the acute phase in 15%

to 25% of children without treatment [14]. Among the predictive factors for coronary artery disease have been identified: male, children aged 1 year or more than 8 years, delay in initiating immunoglobulin therapy or insufficient dose and hyperplaquectosis [15-18]. We had two factors in our case. We emphasize of administering immunoglobulins as soon as possible even before performing an echocardiography, this could reduce the risk of coronary artery disease. The digestive manifestations are variable, they can be of the type of vomiting of diarrhea of abdominal pain of jaundice and hydrocholecyst. In our patient, it was a subocclusive syndrome which joins the data of the Italian literature [19,20] where the disease was expressed by an acute surgical abdomen which required surgical intervention, we had not resort to surgery in our patient because these digestive signs have been attributed to KD. Therefore, we must think about KD if we have prolonged fever associated with a surgical abdomen even in the absence of typical clinical signs. The inflammatory syndrome is constant, there is no specific biological marker that supports the diagnosis. We had a high SV and CRP, which indicates the severity of the inflammatory syndrome and could be a risk factor for recurrence. In our case, CRP allowed monitoring of inflammation during evolution. Doppler echocardiography is the most sensitive examination for the acute phase of the disease, it allows the assessment of lesions to follow the evolution and to predict the prognosis. If there are no anomalies, a check will be made on the fifteenth day and the eighth week. If we have anomalies, it must be carried out weekly. Indeed, the proximal coronary arteries have been defined as normal in the case of an internal diameter less than or equal to 3 mm they are dilated if the diameter was between 3 mm and 4 mm a diameter greater than 4 mm corresponded to a moderate and greater aneurysm 8 mm to a giant aneurysm. Furthermore, Z-score can be used. We had two giant aneurysms of the right and left coronaries that appeared on day 21 of the onset of symptoms, which joins the literature since the aneurysms occur between the 10th and the 25th day and the frequency is between 15% and 25% in the absence of treatment [21]. In fact, coronary aneurysms expose to two types of complications, the first is linked to a myocardial infarction on thrombosis favored by the turbulence of the coronary blood flow and by hyperplaquectosis, the second complication is the rupture of aneurysm which is very fatal and constitutes a not uncommon cause of death. We did not identify any of these complications in our case. The electrocardiogram can detect rhythm and conduction disturbances caused by damage to the inflammatory system of the conductive system, myocarditis or ischemia. The treatment of Kawasaki disease is based on acetyl salicylic acid and immunoglobulins. Acetyl salicylic acid reduces inflammation, it is recommended at a dose of 80 mg/kg in 4 doses for a week to two weeks then this dose will be reduced to 5 mg/kg/day in a single dose for 6 weeks in the absence of coronary involvement and until the disappearance of dilation in the event of cardiac involvement [22]. Immunoglobulins have an immunomodulatory anti-inflammatory action and neutralize toxins. They should be administered as early as possible at a dose of 2 g/kg intravenously over 12 hours [9,23,24]. If KD is refractory, it is recommended to move after 36 hours to a second or even a third therapeutic cycle, a bolus of corticosteroid may be recommended as well as infliximab. Immunoglobulins were administered late because of their irregular availability and their very high cost. In front of the giant aneurysms, an anticoagulant treatment with heparin in a hypo coagulating dose was instituted as an emergency, which was followed by a long-term antivitamin K treatment to reduce the risk of thrombosis. The target normalized international INR ratio was 2 - 2.5. Monitoring must be strict and regular, it is both clinical biological and ultrasound. Ultrasound monitoring should be continued until one year after it is recommended to perform a coroscanner or a coronography depending on the size of the dilation. Some authors report that more than half of the aneurysms regress in the first two years by scarring and thickening of the intima during the remodeling phase [25,26]. If the aneurysms persist, beta-blockers will be discussed. Advice on physical activity has been given, if a vaccination has been scheduled it should be postponed for 3 months.

Conclusion

KD is a relatively common. It is underdiagnosed before incomplete forms, which exposes her to a diagnostic and therapeutic delay, hence the advantage of knowing how to evoke it before a prolonged fever associated with an occlusive syndrome. Its prognosis is conditioned by cardiac involvement, hence the importance of administering immunoglobulins urgently to reduce the risk of coronary aneurysms.

Conflict of Interest

None.

Authors Contribution

Author and authorship have read and approved the final version of the manuscript.

Patient's Consent

Informed consent has been obtained from the patient's parents for publication of the case report.

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