



CASE REPORT

CORONARY ARTERY ANEURYSMS IN KAWASAKI DISEASE DIAGNOSED IN ADULTHOOD: A CASE REPORT

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ABSTRACT

Kawasaki disease is an acute systemic vasculitis of children. In the adult, it's extremely rare, usually diagnosed by coronary artery aneurysms. We report the case of a 22-year-old patient with coronary artery aneurysms observed in coronary angiography evoking Kawasaki disease. We will discuss the peculiarities of this coronary disease with review of the literature.

Keywords:

Coronary artery aneurysms
Kawasaki disease, Adult.

INTRODUCTION

Kawasaki disease is a vasculitis that preferentially affects medium and small sized arteries, particularly the coronary arteries, mainly in children and exceptionally adults. The major risk of this pathology is related to the presence of coronary artery aneurysms with ischemic heart disease. We report a case of Kawasaki disease revealed in a young adult and we will detail the peculiarities of coronary artery abnormalities.

CASE REPORT

A 22 years old male, with history of fever at 40 °in August 2017, with truncal erythema, diffuse arthralgias, headache and abdominal pain with nausea and vomiting persisting despite treatment with amoxicillin-clavulanic acid, introduced by his doctor in front of a suspicion scarlet fever. On clinical examination, the rash is erythematous, maculopapular and non-pruriginous predominant on the trunk and extends to all four limbs with edema of the hands. Cervical lymphadenopathy, bilateral conjunctival injection with palpebral edema and strawberry tongue are also found. Biological examinations revealed a C-reactive protein (CRP) level of 106 mg / ml, neutrophil polynucleosis at 18 G / L and a disturbed hepatic assessment (ASAT = 58 U / L, ALAT = 131 U / L, GGT = 310 U / L, PAL = 328 U / L). The strepto-test is negative as well as ASLO / ASDOR. Blood cultures, the immunological status

and the different viral serologies (HIV-EBV-CMV-HBV-HCV-parvovirus) are also tested negative. The diagnosis of Kawasaki disease is based on all the above mentioned clinical criteria. A treatment combining intravenous immunoglobulin and aspirin with an anti-inflammatory dose is quickly effective, allowing a retrocession of fever and various symptoms in less than 48 hours. Classic peeling of the fingers is secondarily observed. Trans thoracic echocardiography showed normal valvular structures, and normal size of cardiac cavities, with good systolic left ventricular function (FE at 66%). A giant aneurysm involving the left main coronary artery with a diameter of 11 mm (Figure 1) and another aneurysm of left anterior descending coronary artery with a diameter of 12 mm (Figure 2). Coronary angiography revealed aneurysms on the proximal and distal segment of the left anterior descending coronary artery (Figure 3), staged aneurysms of the circumflex artery (Figure 4), ectasia on the right coronary artery with multiple aneurysms (Figure 5). The patient was treated by antiplatelet drugs and vitamin K antagonists with a favorable clinical evolution.

DISCUSSION

Kawasaki disease was first described in 1967 by Tomisaku Kawasaki (Kawasaki *et al.*, 1967), it is 10 times more common in Japan than in Western countries, and in the latter, more common in Asian subjects (Holman *et al.*, 2003) The etiology of this condition remains imprecise. It results from the conjunction of an excessive inflammatory response to infectious or inflammatory factors, in predisposed subjects (Wang *et al.*, 2005) the hypothesis of a genetic predisposition

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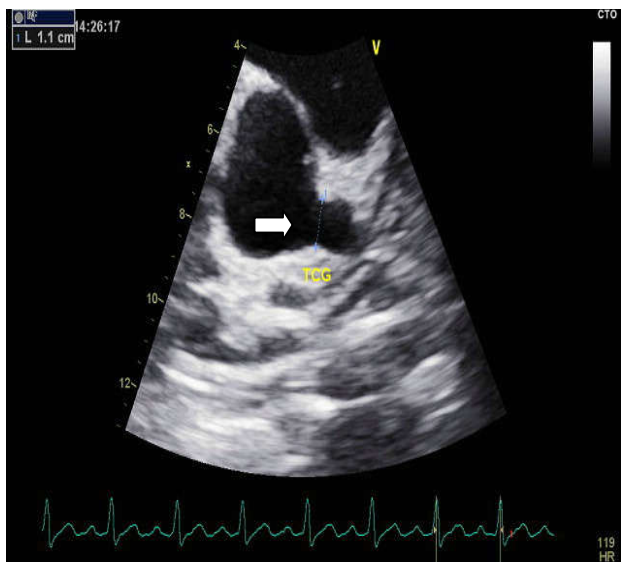


Figure 1. Echocardiography showing a giant aneurism involving the left main coronary artery with a diameter of 11 mm

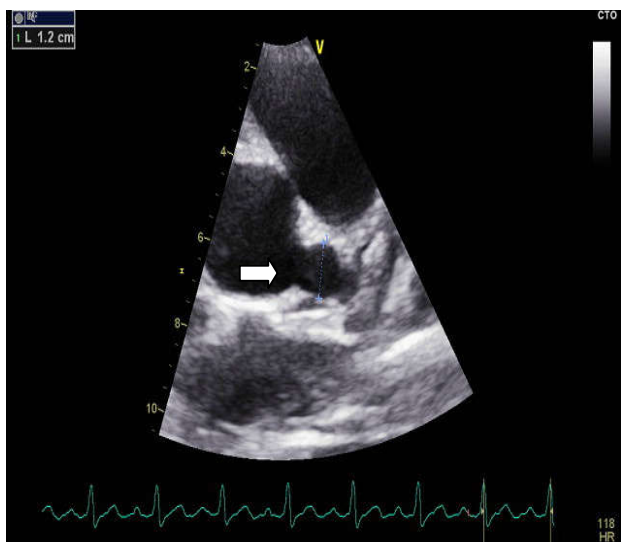


Figure 2. Echocardiography showing aneurism of the left anterior descending coronary artery with a diameter of 12 mm

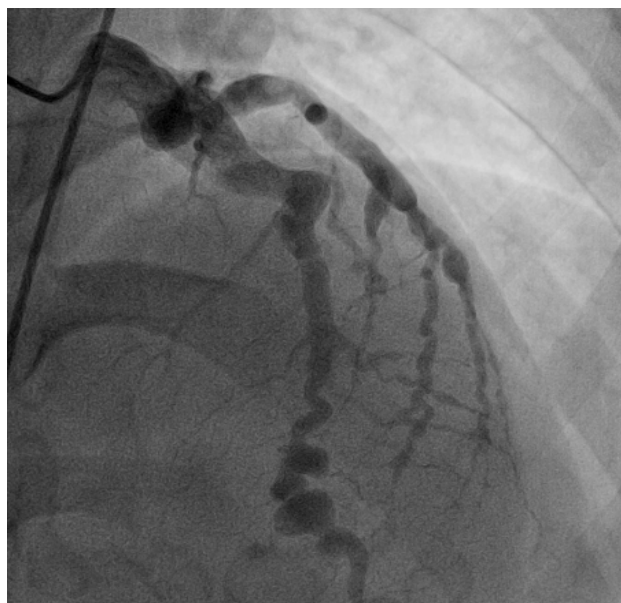


Figure 3. Right anterior oblique caudal view showing aneurysms on the proximal and distal segment of the left anterior descending coronary artery



Figure 4. Right anterior oblique caudal view showing staged aneurysms of the circumflex artery



Figure 5. Left anterior oblique view 30° showing ectasia on the right coronary artery with multiple aneurysms

has been evoked (Dimitriadis *et al.*, 2014). Its diagnosis is purely clinical, based on the association of a fever of 5 days or more without explanation and the presence of at least 4 of the following symptoms: polymorphous rash, modification of the extremities, bilateral conjunctival injection, cervical lymphadenopathy, oral mucosal changes (red and cracked lips, strawberry tongue) (Rauch *et al.*, 1985). Coronary aneurysms are a common mode of revelation in adults of this disease (Daniels *et al.*, 2012). An investigation done by (Kato *et al.*, 1992) identified 130 adult patients with coronary lesions attributable to childhood Kawasaki disease. The infiltration of the coronary artery wall by T lymphocytes and macrophages (Brown *et al.*, 2001) and the destruction of the elastic layer lead to the formation of coronary aneurysms, calcifications and later stenosis (Suzuki *et al.*, 2000) Pathological studies of coronary artery aneurysm in children showed remodeling, senescence and thickening of the coronary wall close to atherosclerosis of the adult (Fukazawa *et al.*, 2007) However, unlike atherosclerosis, the remodeling of aneurysms is marked

by an important production of growth factors such as transforming growth factor, platelet derived growth factor A, basic fibroblast growth factor and vascular endothelial growth factor (Gold *et al.*, 1997). Coronary artery disease is characterized by a significant evolutionary potential in the acute phase of the disease as well as in the late phase (Newburger *et al.*, 2004). The regression of the aneurysms occurs in 50 to 67% of cases during the first two years whereas stenosis on the aneurysm site develops in 5 to 19% of patients, usually after this period (Kato *et al.*, 1996). The size of aneurysms (> 5 mm), their shape (sacciform versus fusiform), involvement of proximal coronaries and the male sex are predictive factors for the persistence of aneurysms (Akagi *et al.*, 1992). Giant aneurysms, more than 8mm in diameter, and "sausage" dilatation are often complicated by infarction.

Local hemodynamic changes, endothelial lesions and slowdown circulatory promote thrombosis. Myocardial infarction occurs almost exclusively in late-stage coronary stenosis. About 40% of patients with stenosis have myocardial infarction during follow-up (Kato *et al.*, 1996). The first ischemic attacks of the myocardium are sometimes asymptomatic but subsequent accidents can be fatal or damage the myocardium by altering left ventricular function. Aneurysmal rupture is exceptional in adulthood (Satoda *et al.*, 1998). Coronary functional abnormalities like atherosclerosis are observed. The coronary reserve decrease on the late phase (Furuyama *et al.*, 2002). The most secondary coronary artery stenosis are developed in the first two years, but half may develop beyond that with a constantly ascending risk curve that will warrant prolonged systematic cardiac monitoring. The three "tunics" of the heart are also concerned by inflammatory involvement. Pericarditis, when the effusion is abundant, is easily evidenced by echocardiography to reveal coronary artery disease. In adults, transthoracic coronary examination is often difficult, (in our case this examination allowed the detection of coronary aneurysms) and the use of transesophageal echocardiography may facilitate the detection of coronary aneurysms (Habon *et al.*, 1998). Non invasive methods such as the multi-slice CT coronary angiography [20] allows the detection and monitoring of coronary artery disease in Kawasaki, to avoid the need for coronary angiography for many patients, it nevertheless requires a regular heart rate and low, a short apnea and a cooperation of the patient. MRI also allows functional analysis of the myocardium (Goo *et al.*, 2006). Patients with large, and especially giant, aneurysms have a risk of thrombosis or coronary stenosis greater than 40% (Kato *et al.*, 1996). As such, they must benefit from a regular biannual cardiologic follow-up with annual cardiac stress tests (Dobutamine ultrasound or cardiac scintigraphy with exercise test) (Newburger *et al.*, 2004). In case of proven ischemia, coronary angiography must be performed (Newburger *et al.*, 2004). These patients require long-term aspirin therapy often supplemented with long-term anti-vitamin K curative anticoagulation (INR target between 2 and 2.5) (Newburger *et al.*, 2004), especially for giant aneurysms. In the case of multiple or complex aneurysms, some experts use a double anti platelet with clopidogrel and aspirin (Newburger *et al.*, 2004). Cardiac catheterization is important to evaluate patients for thrombolytic therapy and catheter intervention to assess for possible coronary artery bypass surgery (Singh *et al.*, 2013). When transluminal coronary angioplasty is indicated, the difficulties are essentially of a technical nature, in particular the choice and the size of the stent because of the disparity of the caliber between the healthy

and the ectasic segment and the fear of deploying the stent when the lesion is included in an aneurysmal segment. The use of covered and self-expanding stents (Pastormerlo *et al.*, 2016) is of great help in the revascularization of these patients. In the absence of evidence for the long-term outcome of this group of patients, medical follow-up is recommended every two to five years with counseling and management of any additional cardiovascular risk factors (Newburger *et al.*, 2004).

Conclusion

Kawasaki disease rarely occurs in adulthood. However, it should be suspected at this age as in children with prolonged fever, and this clinical suspicion should lead to the realization of echocardiography searching for coronary complications. The patients must have a more severe control of the cardiovascular risk factors. The difficulty of ultrasonographic diagnosis of coronary abnormalities in adults should lead to repeat functional tests and perform complementary imaging, invasive or non invasive (MRI, CT, angiography)

Disclosure of Interests

The patient described in this case report has provided written consent for its publication. The author has no conflict of interests to disclose

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