INTRODUCTION

Kawasaki Disease (KD) is an acute vasculitis that involves mainly small- and middle-sized arteries, with predilection to coronary arteries. Coronary lesions show a broad spectrum of anatomic presentations varying from transient coronary ectasia to multiple giant and complicated aneurysms. Although the incidence and natural course of coronary aneurysms after KD are well documented in studies, related reports on peripheral arterial and aortic aneurysms are scarce. We report the occurrence of a giant aortic aneurysm involving the horizontal part of aortic arch in a 28-month-old boy diagnosed with KD. This complication was managed by steroids therapy in the beginning. Because of mechanical complication and potential risk of rupture, surgery was undertaken.

CASE REPORT

A 28-months-old boy, without any relevant past history, was admitted for fever evolving since 8 days, vomiting, and diffuse abdominal pain. On physical examination, we noticed a strawberry tongue, lips abnormalities (erythema, dryness, and cracking and bleeding of the lips), polymorphous skin rash, and edema of hands and feet. Cervical lymph nodes were not enlarged and there was no conjunctivitis. There was elevated levels of systemic inflammation markers: C-reactive protein level (102 mg/l), erythrocyte sedimentation rate (110 mm, 1st h), and platelet count (861.10^3 elements/mm^3). Blood cultures were negative and pro-calcitonin level was normal. Chest X-ray [Figure 1] showed homogenous and well-shaped opacity in the superior part of anterior mediastinum, displacing trachea to the right. The diagnosis of KD was very likely, given the association of persistent fever (>5 days) and evocative clinical features (three among the five diagnostic clinical features) in one hand and the absence of other potential etiologies in the other hand. A transthoracic echocardiography (TTE) was performed in order to look for coronary artery involvement. TTE did show a slight dilatation (4 mm) of right coronary artery, but there were no evidence for coronary aneurysms, valvular heart disease, myocarditis, or pericarditis. Electrocardiogram (ECG) was normal. Thoracic multislice computed tomography (CT) scan, performed in order to determine the exact nature of the opacity found on chest X-ray, showed a huge aneurysmal dilatation (40 × 40 × 45 mm) of the horizontal part of aortic arch between brachiocephalic artery and left common carotid artery [Figure 2]. Aneurysms at other potential locations were excluded by CT scan and Doppler studies. All observed manifestations were attributable to KD. Both intravenous immunoglobulins (IVIGs) and acetylsalicylate in anti-inflammatory doses were administered. Two days later, fever and markers of inflammation persisted and patient’s respiratory status worsened. Physical examination and chest X-ray [Figure 3] were consistent with atelectasis of the left lung, which was confirmed by bronchial fibroscopy showing compression of the left main bronchus. Corticosteroid therapy was then prescribed. Intravenous methylprednisolone (1 g/m^2 of body surface area/day) was given for 2 days followed by prednisone (1 mg/kg once daily orally). There was a favorable response. ECG monitoring remained normal.
and repeated echocardiographic studies showed total regression of the coronary artery dilation. The patient was kept on acetyl-salicylate at antiplatelet therapeutic dose and steroids till normalization of inflammatory markers.

Given the mechanical complication and the risk of aneurysm rupture related to its size, surgery was indicated. The aneurysm was surgically removed and replaced by prosthetic conduit (Dacron conduit of 14 mm of diameter). Histological findings of the removed tissue showed inflammatory infiltrates of the aortic wall with elastic fibers destruction. There was no necrosis of arterial media.

**DISCUSSION**

Although the incidence and the natural course of coronary aneurysms after KD are well-documented in several studies,[1,2] related reports on peripheral arterial and aortic aneurysms are rare. In the long-term follow-up study of 594 KD patients, published in 1996 by Kato and coworkers, 2.2% of the patients exhibited systemic artery aneurysms. Among these 13 cases, there were 11 axillary, nine common iliac, seven internal iliac, and four subclavian artery aneurysms. [3] Aortic aneurysms are rarely reported, although aortitis has occasionally been found in KD patients at autopsy.[4] Most of the KD-related aortic aneurysms were found in children.[5] Pathologic studies revealed thickened and calcified aneurysm wall, with atrophy of elastic media and the presence of the intraluminal thrombus.[6] In our patient, aged only 24 months, aneurysm localization in the horizontal part of aortic arch was quite uncommon. In fact, most of the KD-related aortic aneurysms seem to involve the abdominal aorta or descending thoracic aorta.[6,7] Because KD aortic aneurysms are extremely rare, standard treatment protocols are not established. Some reports attest that non-coronary artery aneurysm complicating KD has a tendency to regress.[8] On the other hand, severe complications as aneurysm thrombosis with distal ischemia or fatal femoral and hepatic aneurysms rupture were reported.[4,9] In our case, given aneurismal size and the presence of life-threatening mechanical compression, we opted for surgical treatment.

**CONCLUSION**

KD-related aneurysms commonly involve coronary arteries. Those arteries must be systematically investigated in any suspected case of KD. Aortic aneurysm are, however, quite rare. Even so, it is wise to look for them systematically in order to avoid catastrophic potential complications. Their rarity may be due in part to under-diagnosis.

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How to cite this article: Hakim K, Boussada R, Chaker L, Ouarda F. Giant aortic arch aneurysm complicating Kawasaki's disease. Ann Pediatr Card 2014;7:201-3.

Source of Support: Nil, Conflict of Interest: None declared