Cardiology in the Young

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Rare case of Kawasaki disease with cardiac tamponade and giant coronary artery aneurysms

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Brief Report

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Abstract

Kawasaki disease is an acute systemic vascular disease, generally self-limited and typically affecting children <5 years old, which leads to coronary artery aneurysms in about 25% of untreated cases. Cardiovascular involvement is characterised by transient pancarditis, in acute phase, while coronary illness, ranging from mild dilation to giant CAAs occurs late, rarely before the 10th day since fever onset. Here, we describe a peculiar case of KD, which occurred in a 4-month-old infant and presented with exudate cardiac tamponade and early giant aneurism of both the proximal right coronary artery) and the left circumflex coronary artery, in acute phase of the disease.

Case report

A 4-month-old infant was presented with high fever (>39°C, refractory to therapy, started 4 days before) and severe respiratory distress. During physical examination, he showed conjunctival injection, oral, and pharyngeal erythema, cervical lymphadenopathy, maculopapular rash, muffled heart sounds, and hypotension. Chest X-ray demonstrated marked enlargement of the cardiac outline and echocardiography showed a large pericardial effusion with cardiac tamponade. Thus, he was transferred to Pediatric Cardiac Surgery to undergo pericardiocentesis, and the pericardial fluid was citrine yellow, exudative, of inflammatory nature. At the post–procedure, ultrasound evaluation dilation of the proximal right coronary artery with a giant aneurysm at the origin (11 mm, z score +33.18) and giant aneurysm of the left circumflex coronary artery (4 mm, z score +12.50) were identified; therefore, diagnosis of KD was made and to reduce inflammation, immunoglobulin therapy (2 g/kg in 12 hours) and acetylsalicylic acid (40 mg/kg/daily) therapy was started, as well as anticoagulant therapy with Dicumarol (to obtain INR target levels 2–3).

Four months after the disease onset, the patient underwent a coronarography that confirmed the giant RCA aneurysm (Fig 1) in absence of downstream flow, and the giant LCX aneurism, partially filled by a collateral circulation. The patient underwent a regular cardiological follow-up and serial echocardiographic evaluations that documented a progressive size reduction of the coronary artery aneurysm. Three years later, a second coronarography was performed that confirmed the normal calibre of both the RCA and the LCX, and the absence of the aneurysmal formations previously reported. It was decided to suspend anticoagulant therapy and to continue with ASA at an antiaggregant dosage (3–5 mg/kg/daily).

Discussion

The onset of KD is generally characterised by acute and intense febrile symptoms, frequently associated with a general cardiac involvement (pancarditis), and is characterised, in the beginning, by symptoms of mild intensity. Although KD is generally self-limiting, 20–25% of patients develop severe complications, such as CAA (z score ≥10, or absolute dimension ≥8 mm),[1–2] myocardial infarction, and heart failure.[3]

Coronary artery involvement is a frequent complication of KD, and a giant aneurysm can occur, in particular in untreated patients during the subacute phase.[1] After the introduction of IVIG and high-dosage ASA for the treatment of KD, the occurrence and the regression of the CAA was improved. Recently, Kevin et al described the natural history of CAA in the United States of America KD patients and identified factors associated with CAA regression.[4] In their

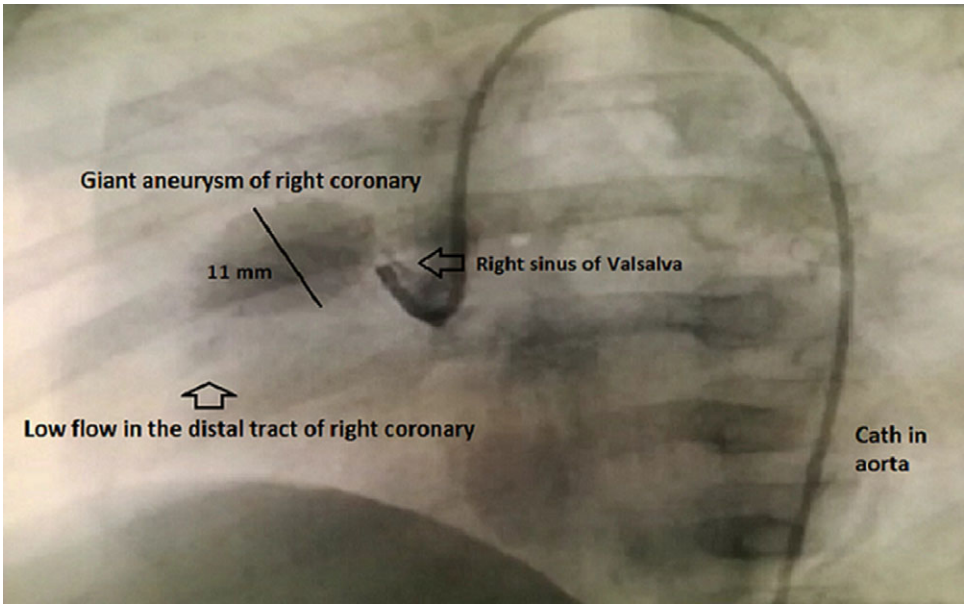

Figure 1. Coronarography scan shows the presence of a giant aneurysm and low flow in the distal tract of the right coronary artery.

large cohort, CAA regression occurred in 75% of patients. However, they found that lack of CAA regression was associated with larger CAA z score at diagnosis (in particular >10 z score), and bilateral CAA. Pericardial effusion represents another common complication of KD, reported in 6–24% of patients[5]; however, the cardiac tamponade is an exceptional event, which occurs principally in the convalescent phase of the disease, mostly due to an aneurysmal rupture.[6–7]

In our case, both the CAA formation and the cardiac tamponade, which was not related to CAA rupture, occurred in acute phase of KD. We believe that the early onset of this complication can be related to the young age of the patient. Indeed, in infants younger than 6 months of age, KD seems to have a rapidly fatal course in acute phase of disease, principally related to CAA complication, such as thrombosis and rupture that can lead to cardiac tamponade, myocardial infarction, and sudden cardiac death[8]; it was supposed that the susceptibility to coronary complication can be associated to the small thickness of the coronary walls in this age, which may favour the inflammation of the three arterial tunics (i.e., panarteritis).[8] Another important finding was the complete CAA regression, which is considered as an extraordinary event in young patients with bilateral CAA and giant aneurysm (z score >10).[4] This finding is consistent with previous studies showing that the incidence of CAA regression is higher in children less than 1 year of age compared to older patients.[9]

Conclusion

In patients <6 months of age, the onset of KD may be extremely aggressive, and a delayed diagnosis may lead to severe complications and death. In our case, the infant developed two important life-threatening complications of KD, the cardiac tamponade and the CAA formation; however, with an appropriate management, he showed a good long-term prognosis. These findings confirmed the importance of the early diagnosis and the proper treatment of KD, particularly in infants.

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Conflicts of interest. None.

Ethical standards. The authors assert that all procedures contributing to this work comply with the ethical standards of the relevant national guidelines on human experimentation and with the Helsinki Declaration of 1975, as revised in 2008, and has been approved by the institutional committees.

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