Case report - Cardiac general

Kawasaki disease presenting as cardiac tamponade with ruptured giant aneurysm of the right coronary artery

Madhankumar Kuppuswamy*, Philemon Gukop, George Sutherland, Chandrasekaran Venkatachalam

Department of Cardiology and Cardiothoracic Surgery, Atkinson Morley Wing, St. George's Hospital, Tooting, London SW17 0QT, UK

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Abstract

We report a case of a 22-year-old man with Kawasaki disease presenting with features of cardiac tamponade following rupture of giant aneurysm of his right coronary artery. He underwent an emergency operation. Aneurysmal sac was of size 4 × 4 cm. The entry point of the aneurysm was sutured. Right coronary artery was grafted with left radial artery. He had an uneventful recovery in the postoperative period.

Keywords: Kawasaki disease; Giant aneurysm; Cardiac tamponade

1. Case report

A 22-year-old gentleman presented with a history of dull aching chest pain of 6 h duration. He was diagnosed to have Kawasaki disease at the age of five years. He was known to have patent ductus arteriosus with aneurysm of the right coronary artery and was being followed-up in a different hospital. On examination his heart rate was 96 beats per min and blood pressure was 96/70 mmHg. His ECG showed Q wave with T wave inversion in the inferior leads. It also showed features of right bundle branch block. His troponin was negative. Echocardiography showed a mass over the right heart of size 4 cm with trivial pericardial effusion (Fig. 1). CT-scan showed a large mass of size 8 × 7 cm compressing the right atrium with calcified left coronary artery aneurysm. As the CT-scan was being performed his hemodynamics deteriorated with increasing size of the mass. Hence, he was taken for emergency surgery before completing the CT-scan.

During surgery his left femoral artery was cannulated. Then the chest was opened by median sternotomy. Approximately 750 ml of blood with blood clots were evacuated from the pericardial cavity. Bleeding was initially controlled with the finger. Ruptured giant right coronary artery aneurysmal sac was identified. The aneurysmal sac was just adjacent to the right coronary artery ostium. Entry point of the right coronary artery aneurysm was identified and the bleeding was then controlled by introducing Foley’s catheter into the ascending aorta through the entry point of the aneurysmal sac (Fig. 2). Two-stage single venous cannula was inserted to the right atrial appendage and the patient was connected to cardiopulmonary bypass.

Patient’s temperature was maintained at 34 °C throughout the procedure.

The entry point of the aneurysmal sac was at the origin of right coronary artery (RCA) which was closed with two Teflon pledgetted sutures. The aneurysmal sac was measuring 4 × 4 cm. Distal opening of the aneurysmal sac was also identified and sutured. Patent ductus arteriosus was identified looped and ligated. In the meantime left radial artery was harvested. Distal right coronary artery was opened and endarterectomy was done (specimen sent for histopathological examination) as the posterior descending artery was of small calibre and not graftable. Distal anastomosis of the left radial artery to right coronary artery was done with heart in ventricular fibrillation. Proximal anastomosis completed. Calcified aneurysm of size 1.5 cm was noted in the proximal left anterior descending artery. There was no evidence of ischemia in the left anterior descending artery territory. Hence, left anterior descending artery was not grafted. The patient came off cardiopulmonary bypass without difficulty. He had an uneventful recovery in the postoperative period and was discharged on the fifth postoperative day. Postoperative echocardiogram done before discharge showed moderately impaired function of the right ventricle. No pericardial collection was noted.

2. Discussion

Kawasaki disease or mucocutaneous lymph node syndrome is a systemic disease with generalised vasculitis predominantly affecting infants and young children [1]. It affects primarily medium sized muscular arteries [2]. Damage caused by the disease in the pediatric age group can progress and present in the adolescent or adult age group. The involved arteries develop aneurysmal formation,
thrombotic occlusion and premature atherosclerosis leading to ischemic heart disease [3]. Kato et al. showed the incidence of coronary aneurysm in acute Kawasaki disease was 25%, 55% of which showed regression. Giant coronary artery aneurysm was noted in 4.4% of the patients. During follow-up, ischemic heart disease developed in 4.7% and myocardial infarction in 1.9%. Death occurred in 0.8%. Death due to rupture of coronary artery aneurysm is extremely rare [3].

In untreated group, incidence of coronary artery aneurysm is 20% compared to 4–8% in the group treated with IV gamma immunoglobulin [4]. Hiroyuki et al. graded coronary artery aneurysm from grade 0 to grade 3, grade 3 being giant aneurysms with maximal diameter 8 mm. These grade 3 aneurysms are less likely to reduce in size and will often thrombose or rupture. Giant coronary artery aneurysms are very rare presentation [5].

In the case presented only the femoral artery was cannulated. Femoral vein was exposed but not cannulated. As the mass was compressing almost the entire right atrium, positioning the femoral cannula in the right atrium would have been difficult. Possibility of the femoral cannula disturbing the mass in the right atrium was also considered. As the clots were being removed from the pericardial cavity, the bleeding point was noted. It was controlled with the finger initially and the structures were identified. Then the patient was connected to cardiopulmonary bypass without significant hemodynamic deterioration. Fibrillatory arrest period was restricted to the distal anastomosis of the left radial artery to right coronary artery. All the other procedures were done without arresting the heart.

Calcification, fissuring, deposits of protein-like material and hyaline degeneration of the intima of the involved arteries in the Kawasaki disease were reported [6]. Histopathology of the endarterectomy specimen in the case reported showed marked fibrous scarring and calcification within the muscular wall. No active vasculitis or granulomas.

Giant coronary artery aneurysms which show increased rate of growth in size should be managed surgically in order to avoid acute complication like rupture as it happened in the case presented. Also, planning the surgery according to the need of the emergency situation like avoiding cardioplegic arrest in the case reported is to be considered for better outcome of these patients.

References


eComment: Coronary artery aneurysms in Kawasaki disease

Authors: Ioanna Koniar, Department of Cardiothoracic Surgery, University Hospital of Patras, 22500 Rion Patras, Greece; Efstratios Apostolakis, Nikolaos G. Baloussis, Grigorios Tsigkas
Your interesting case report concerning the rupture of a giant right coronary artery aneurysm constituting a really rare complication of Kawasaki disease is worth to be further discussed [1]. In fact, aneurysmal coronary artery disease should be considered a variation of coronary artery disease, with which it usually coexists. The mechanism of coronary artery stenosis in Kawasaki disease is not well-defined, depending on acute occlusion by massive thrombus formation in the coronary aneurysms or progression of marked intimal thickening often associated with calcification, that is similar to an arteriosclerotic lesion.

Coronary artery aneurysms occur often in the proximal coronary segments and at the bifurcations of the coronary arteries. They may involve multiple vessels. Complications from giant coronary artery aneurysms include thrombosis, rupture and long-term myocardial ischemia. The most important predictor of myocardial infarction is the aneurysmal size [2]. Especially, a long-term study in 1215 patients with Kawasaki disease found that 64 (5%) developed giant coronary artery aneurysms. Subgroup analysis revealed regression in 3 (5%), occlusion or stenosis in 30 (47%) and myocardial infarction in 15 (23%) [3].

Coronary angiograms have been the gold standard to assess coronary artery aneurysms. However, multislice computer tomographic (CT) coronary angiogram has been found to have an excellent correlation with coronary angiogram in terms of number, size and shape of coronary aneurysms [4]. Moreover, multislice CT coronary angiogram demonstrates the extraluminal components, such as thrombi and aneurysmal shape which could not be assessed by coronary angiogram [4]. Alternatively, giant coronary aneurysms can be detected by transesophageal echocardiography. The nuclear perfusion scan may be a more sensitive tool in the detection of myocardial ischemia and would complement the multislice CT coronary angiogram as a non-invasive method for follow-up of patients with significant risk of ischemia following Kawasaki disease [2].

Different surgical treatment, such as aortocoronary bypass surgery and percutaneous transluminal coronary angioplasty have been proposed. Intimal mammary arterial bypass graft results in better long-term patency and survival than saphenous vein grafts (77.1% ± 1.1% compared to 46.2% ± 6.3% for patency 85 months after operation) [5].

Finally, cardiac transplantation for severe ischemic heart disease as a sequela of Kawasaki’s disease has been proposed in the subgroup of patients who are not candidates for revascularisation because of distal coronary stenosis or aneurysms, and/or those with severe irreversible myocardial dysfunction.

References