CASE REPORT

Coronary artery aneurysm presenting as a large mass encroaching on the right atrium in an adult patient with untreated Kawasaki disease

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SUMMARY

A 26-year-old man presented with atypical chest pain and dyspnoea for several days before admission that was not related to exertion. The patient had no medical history other than a long duration of fever and rash as a child, which resolved after a few weeks. The initial blood investigations showed thrombocytopenia and raised inflammatory markers with a negative troponin level. The patient had positive antiplatelet antibodies and was diagnosed with immune thrombocytopenic purpura (ITP). Echocardiography showed a large cystic mass at the right atrium. Coronary multislice CT showed a huge aneurysm at the origin of the right coronary artery (RCA), which was confirmed by coronary angiography. The patient received intravenous immunoglobulin and platelet transfusion before coronary artery bypass surgery and he had a successful resection of the aneurysm with a saphenous vein graft to distal RCA. The patient had a good recovery and was discharged home.

BACKGROUND

Many cases of young adults presenting with angina or myocardial infarction with concomitant large coronary artery aneurysms, who may have had Kawasaki disease during childhood, have been reported. This is the most likely cause for our patient's presentation with a huge right coronary artery (RCA) aneurysm with previously untreated Kawasaki disease as a child. Our case report is to raise the awareness of Kawasaki disease complications that may present in adulthood with ischaemic cardiac events.

CASE PRESENTATION

A 26-year-old man presented with stabbing chest pain with referral to the left shoulder, unrelated to exertion or respiration. His chest was not tender to pressure. He had associated dyspnoea with minimal effort New York Heart Association (NYHA) functional class III for several days before admission. He had no lower limb oedema, orthopnoea or paroxysmal nocturnal dyspnoea. The patient had no medical history other than a long duration of fever and rash as a child, which resolved after several weeks. He had a family history for coronary artery disease as his father had myocardial infarction at the age of 54, with no history of aneurysms in the family. Clinical examinations showed a mild degree of exophthalmos and diffuse goitre, with no palpable masses and no tremors. Abdominal

examination showed mild splenomegaly with no other organomegaly or lymphadenopathy.

INVESTIGATIONS

The initial blood investigations showed thrombocytopenia and raised inflammatory markers with a negative troponin level. The patient had positive antiplatelet antibodies, mild splenomegaly by ultrasound and a bone marrow biopsy, by which he was diagnosed with immune thrombocytopenic purpura. Ultrasound scan revealed enlargement of the thyroid gland, with no evidence of nodules; the patient had a negative antithyroglobulin antibody.

ECG showed no significant ST-T wave changes and few ventricular ectopic beats. Transthoracic echocardiography showed a large extra-cardiac cystic mass near the right atrium, which was confirmed by a transoesophageal echocardiography (figures 1 and 2). Myocardial perfusion imaging showed an inferior wall defect (figure 3). Multislice CT coronaries showed a huge aneurysm at the origin of the RCA, which was confirmed by coronary angiography (figures 4 and 5).

DIFFERENTIAL DIAGNOSIS

Masses at the right atrium should be differentiated on the basis of location, shape, attachment and mobility, varying from intracardiac and extracardiac structures such as neoplasms, pericardial cysts,

Figure 1 Transthoracic echocardiography apical four-chamber view showing a cystic mass encroaching on the right atrium.

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Figure 2 Transoesophageal echocardiography short-axis view showing a large extracardiac cystic mass at the level of the right atrium.

aneurysms, diaphragmatic hernia or cystic lesions in abdominal organs.

TREATMENT

The patient received intravenous immunoglobulin and platelet transfusion before coronary artery bypass surgery and he had a successful resection of the aneurysm with a saphenous vein graft to distal RCA (figure 6).

OUTCOME AND FOLLOW-UP

The patient had a good recovery and was discharged home after 3 weeks.

DISCUSSION

Our patient presented with a large mass encroaching on the right atrium seen by transthoracic echocardiography, which was diagnosed by CT scan as a large aneurysm. There are multiple causes of right atrial masses and these should be differentiated on the basis of location, shape, attachment and mobility varying from intracardiac structures such as Eustachian valve, Chairi network, crista terminalis, Cor triatriatum dextrum, atrial myxoma, thrombus, vegetations and cardiac tumours, whether benign or malignant. The extracardiac causes vary from a large diaphragmatic hernia to neighbouring neoplasm, such as

Figure 3 Myocardial perfusion image showing inferior wall defect seen in the stress phase in the horizontal and short-axis cuts in comparison to the resting phases of these cuts.

Figure 4 Multislice CT showing a large aneurysm at the origin of the right coronary artery from the ascending aorta.

mediastinal or lung tumours, and pericardial fatty tissue may be seen as a mass near the right atrium. Aortic, coronary artery and saphenous vein graft aneurysms may compress the right atrium. Abdominal structures such as cystic lesion of other organs such as the liver and kidney may also compress the right atrium.¹

Our patient was presumed to most likely have had untreated Kawasaki disease as a child and he presented with an aneurysm as a complication as an adult. Kawasaki disease is an acute, self-limited vasculitis of unknown aetiology, although an infectious agent is suspected based on clinical and epidemiological features. A genetic predisposition is likely, based on varying incidences among ethnic groups, with higher rates in Asians.² Tomisaku Kawasaki first described it in Japan in 1967, and it occurs both in endemic and epidemic forms. Males are more susceptible than females with a ratio of approximately 1.5:1.^{3 4} There is no specific diagnostic assay for Kawasaki disease; therefore, diagnosis is based on clinical criteria.²

Figure 5 Coronary angiography showing a huge right coronary artery (RCA) aneurysm at the proximal RCA.

Figure 6 Intraoperative image showing a large right coronary artery aneurysm on the upper right side of the image.

Kawasaki disease can cause myocarditis in the first 3–4 weeks with affection of the mitral valve more than aortic valves and may involve the conduction system. It causes pericarditis with pericardial effusion in 30% of patients.^{3 4} Coronary artery aneurysms or ectasia develop in 15–25% of untreated children with the disease and may lead to myocardial infarction, sudden death or ischaemic heart disease. Inflammation persists in the walls of medium and large arteries, especially the coronary arteries, characterised by oedema, mononuclear cell infiltration and progressive fibrosis with disruptions in the internal elastic lamina. Such destructive changes lead to aneurysms that are more pronounced in the proximal segments and branching points of the coronary arteries, suggesting contribution of haemodynamic stress.³ Other arteries, such as iliac, femoral, axillary and renal, are less frequently involved. Intracranial arteries or intraparenchymal vessels within abdominal organs are almost never involved. More than 50% of aneurysms regress within 1–2 years, especially if small.³

Cardiac MRI and CT can accurately delineate coronary artery aneurysms in the proximal coronary artery segments.³

Initial treatment includes intravenous immunoglobulin (IVIG) plus high-dose aspirin followed by subsequent oral aspirin for 6–8 weeks. About 85–90% of patients respond to initial therapy.^{3 5} However, patients with large or giant coronary artery aneurysms (internal diameter greater than 8 mm) should be maintained on aspirin or clopidogrel and warfarin (target international normalised ratio 2–2.5) to prevent thrombosis within the aneurysm and myocardial infarction.² Surgical revascularisation for large aneurysms by internal mammary artery is

preferred over vein grafts, as they increase with growth of children in contrast to vein grafts, which shorten over time.⁴

The mortality rate is about 2% in untreated patients or in those who receive only aspirin treatment. Early treatment with aspirin and IVIG decreases the death rate to less than 1% and decreases the coronary artery dilation to 5%. The prognosis is quite good for patients with no coronary artery abnormalities. Small coronary artery aneurysms are very likely to undergo regression and tend to have good prognosis, at least through childhood. Large or complex aneurysms are less likely to undergo regression.³ Many patients with large aneurysms are at risk for ischaemic heart disease. There have been a number of young adults with angina or myocardial infarction with concomitant large coronary artery aneurysms, who may have had untreated Kawasaki disease during childhood.³

Learning points

- ▶ Right atrial masses should be differentiated on the basis of location, shape, attachment and mobility, whether intracardiac or extracardiac.
- ▶ Coronary artery aneurysms or ectasia develop in 15–25% of untreated children with Kawasaki disease, but the percentage decreases to 5% with treatment.
- ▶ Many young adults with angina or myocardial infarction with concomitant large coronary artery aneurysms may have had Kawasaki disease during childhood.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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