CASE REPORT

Non-atherosclerotic multiple coronary artery aneurysms

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SUMMARY

A 52-year-old man underwent two-dimensional echocardiogram which showed moderate to severe aortic regurgitation (AR) and dilated ascending aorta. CT angiography (CTA) showed dilated ascending aorta (5 cm) and transoesophageal echocardiogram revealed bicuspid aortic valve. He underwent cardiac catheterisation which revealed triple vessel aneurysmal disease of the left anterior descending, left circumflex and right coronary artery. The patient underwent aortic graft placement for ascending aortic aneurysm and aortic valve replacement with a Saint Jude valve for severe AR. There was no history or stigmata of Kawasaki disease and workup for coronary artery aneurysm including vasculitis and connective tissue disorders was negative. Histopathology did not reveal evidence of active aortitis or dissection. His aneurysms are being observed by a yearly coronary CTA. We present a rare case of multiple coronary artery aneurysms associated with bicuspid aortic valve and ascending aortic aneurysm.

BACKGROUND

Although coronary artery aneurysms (CAAs) are not uncommon, multiple aneurysms are rare and giant multiple aneurysm are rarer.¹ The majority are atherosclerotic in origin.^{2–4} Other causes include connective tissue disorders,⁵ trauma, vasculitis, congenital, mycotic, idiopathic and they are the hallmark of Kawasaki disease.² Most of the coronary aneurysms (CAs) are usually identified incidentally during angiography. A systematic review of

the literature suggests CAAs range from 0.15–4.9% in patients undergoing coronary angiography.^{3–6} We report an interesting case of multiple CAs without any evidence of atherosclerosis, associated with bicuspid aortic valve (BAV) complicated with ascending aortic dilatation. There is a need to report the present case to review the literature, understand the management options for CA and be cognisant of the association of CA as part of BAV syndrome (BAVS).

CASE PRESENTATION

A 52-year-old Caucasian man underwent two-dimensional echocardiogram (2D echo) for evaluation of heart murmur. His medical history was pertinent for dyslipidemia and hypertension. He did not recall any history of Kawasaki disease in childhood. Family history was negative for any history of sudden cardiac death, valvular heart disease or coronary artery disease. Physical examination was benign except for diastolic murmur in aortic area. His 2D echo showed moderate to severe aortic insufficiency, moderate dilation of the ascending aorta and mild dilation of the aortic sinuses. On further evaluation via an aortic CT angiography (CTA), the aorta was measured at 5 cm. He subsequently had a transoesophageal echocardiogram which showed BAV (figure 1) with systolic doming and there was no significant aortic stenosis. However, there was moderate to severe eccentric aortic incompetence. The aortic root at the level of the sinuses with Valsalva measured

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Figure 1 Transoesophageal echocardiogram showing bicuspid aortic valve. BV, bicuspid aortic valve; LA, left atrium; RA, right atrium; RV, right ventricle; PA, pulmonary artery.

Figure 2 Coronary angiogram showing aneurysmal disease of left anterior descending (LAD; A), left coronary artery (LCA; B) and right coronary artery (RCA; C).

3.9 cm and beyond the sinotubular junction, it measured 4.9 cm. He underwent a cardiac catheterisation that revealed severely aneurysmal disease of the left anterior descending (7 mm; figure 2A), left circumflex (7–8 mm; figure 2B) and the right coronary (9 mm; figure 2C) in the proximal segments. The aorta was severely dilated with moderate to severe aortic insufficiency. Ejection fraction was normal at 60%. The coronary angiography did not reveal significant coronary artery disease. The patient underwent aortic valve replacement with a St Jude mechanical valve and ascending aortic graft placement for ascending aortic aneurysm for severe aortic regurgitation of BAV and ascending aortic aneurysm. Later, the patient was worked up for CAAs, no apparent cause was found. He did not have any history or stigmata of Kawasaki disease. His workup was negative for vasculitis and connective tissue disorders: Venereal Disease Research Laboratory test, normal; rheumatoid factor, 10 IU/mL (normal range (nl) <14 IU/mL); hepatitis B surface antigen, negative; erythrocyte sedimentation rate, 4 mm/h (nl 0–20 mm/h) and C reactive protein, 0.5 mg/dL (nl 0–0.7 mg/dL). Pathology of aortic valve and ascending aorta from sinotubular ridge did not reveal any evidence of active aortitis, atherosclerosis or dissection. Currently, the patient is being monitored closely for his CAAs with coronary CTA. He is having coronary CTA on yearly basis (figure 3) which revealed similar findings with no change or increase in size of aneurysms. He is being managed conservatively with β -blockers, lisinopril, aspirin, lansoprazole and simvastatin in addition to anticoagulation with

coumadin in view of his St Jude mechanical valve with a goal international normalized ratio (INR) of 2.5–3.5.

OUTCOME AND FOLLOW-UP

Currently, the patient is being monitored closely for his CAAs; he had a repeat coronary CTA that revealed similar findings with no change or increase in size of aneurysms.

DISCUSSION

CAA refers to any coronary artery dilation exceeding the diameter of normal adjacent segments by 1.5 times⁷ and giant CA is defined with a diameter larger than 5 cm.⁸ Aneurysms can be sacular (transverse larger than the longitudinal axis) or fusiform (longitudinal at least twice the transverse axis). Coronary Artery Surgery Study (CASS) registry studied 20 087 consecutive patients undergoing coronary angiography and demonstrated a 4.9% incidence of CAs (978 patients), most commonly in the right coronary artery.³ The pathophysiology of aneurysm formation has been attributed to be weakening of the medial layer of the vessel wall due to chronic overstimulation of the vasodilator nitric oxide.² Other studies suggest that an excessive proteolysis in the arterial wall may act as a risk factor for development of CA in patients with coronary atherosclerosis.⁹ The majority of CA are associated with atherosclerosis. However, whether atherosclerosis predisposes to aneurysm or vice versa is not clear.⁷ Simultaneous aneurysms of the abdominal aorta and coronary arteries in a patient suggest mutation in MMP genes.⁹ Recently,

Figure 3 Coronary CT angiography showing aneurysmal disease of left anterior descending (LAD; A), left circumflex (LCX; B) and right coronary artery (RCA; C).

an increased number of cases of CAA after implantation of drug-eluting stent have been described.^{10–13} The exact mechanism remains unknown but may be related to the active drug and the polymer coating, might cause gradual luminal dilation, vascular remodelling and subsequent aneurysmal formation.¹⁰ Potential complications of CA include thrombosis¹⁴ and distal embolisation,^{15 16} rupture¹⁷ and vasospasm.¹⁸ The majorities of CAAs are asymptomatic; however, up to one-third of them present with angina, myocardial infarction and sudden cardiac death. Spontaneous rupture is rare but is associated with Kawasaki disease.¹⁹ A coronary angiography is the mainstay of diagnosis¹⁹ but it cannot differentiate a true aneurysm from a pseudoaneurysm.²⁰ A true aneurysm has an intact vessel wall in contrast to a pseudo aneurysm. An intravascular ultrasound (IVUS) can be used to differentiate true from pseudoaneurysm²⁰ as it can provide information regarding arterial wall structure and dimensions. Although, there were no clear data regarding safety of IVUS to detect CAAs; Maehara *et al*²⁰ reported no major complications in a series of 77 patients with aneurysmal dilation of coronary arteries. A CT of the coronary artery angiography²¹ and an MRI scan²² can diagnose CA and can also be used to monitor the aneurysm while continuing the conservative management. There is no optimal management strategy for patients with CAAs. CAs are associated with high mortality, irrespective of the flow limiting coronary obstructive disease.²³ Mortality rate in aneurysmal disease significantly higher, equals with three vessel coronary disease.^{23 24} Owing to the rarity of these aneurysms, particularly of the giant form, it is difficult to establish a standardised treatment.²⁵ Thrombosis is one of the most frequent complications with CAs, so antiplatelet therapy (aspirin and clopidogrel) or anticoagulation with coumadin (goal INR 2–3) is commonly used in the management of patients with close observation.^{26 27} Surgery is indicated for rupture, symptomatic patients and large aneurysms of more than 2 cm or if the CAAs are associated with other associated conditions like fistula.²⁸ Surgical management requires aneurysmectomy and cardiopulmonary bypass and myocardial revascularisation coronary artery bypass grafting.²⁹ CAAs need close monitoring because of potential complications and timely management.

BAV can be associated with vascular complications as part of BAVS.³⁰ Although, the exact mechanism of vascular events associated with BAV is unclear, some evidences suggest that it could be related to aortopathy³¹ or extracellular matrix dysregulation.³² Our case raises a very intriguing point about association of BAV with CA as part of BAVS; which could be vascular matrix dysregulation in the coronary arteries.

Learning points

- ▶ Bicuspid aortic valve can be associated with coronary artery aneurysm besides aortic aneurysm.
- ▶ Coronary aneurysms are most often incidental findings on coronary angiography.
- ▶ Thrombosis is one of the most frequent complications with coronary aneurysms.
- ▶ Patients need to be monitored closely with anticoagulation therapy in case of small aneurysms as mortality rate equals to three vessel coronary disease.
- ▶ CT coronary artery angiography and MRI scan are non invasive techniques to diagnose as well monitor the coronary aneurysm for complication and progression of aneurysmal disease.

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