CASE STUDY

Non-atherosclerotic coronary artery aneurysms: two case reports

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Abstract
Non-atherosclerotic isolated coronary artery aneurysm is not common. Two cases of non-atherosclerotic isolated coronary artery aneurysm, with similar presentations but different management strategies were presented. The patients were well four and six years later, respectively. The definition, incidence, causes, presentation, complications, investigations, management, and prognosis of coronary artery aneurysms are discussed. The difficulties of determining pathogenesis and different management strategies are highlighted. An isolated coronary artery aneurysm should be managed on its merits.
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Aneurysmal coronary artery disease is characterised by abnormal dilatation of a localised or diffuse segment of the coronary artery tree. Before the advent of coronary angiography, all reported cases were based on postmortem findings. The largest angiographic series is the Coronary artery surgery study (CASS) registry where 4.9% of 20 087 patients were observed at angiography to have coronary artery aneurysms.[1] In this series, a coronary artery aneurysm was defined as a dilatation with a diameter of ⩾ 1.5 times the adjacent normal coronary artery. Ninety per cent of these were associated with atherosclerotic coronary artery disease, where vessel ectasia and poststenotic dilatations are not an infrequent finding. Tunick *et al* in their series,[2] used the term "discrete aneurysm" to mean a localised spherical or saccular shaped dilatation of ⩾ 1.5 times. They excluded fusiform aneurysms as they felt that these may be confused with vessel ectasia. In their series of 8422 patients an incidence of 0.2% was observed. Others have suggested a diameter of at least three times normal as the definition of coronary artery aneurysm so that the incidence will vary depending on definition.[3]

The pathogenesis of coronary artery aneurysm involves underlying destruction of the vessel media. This thinning of the media together with increased wall stress causes progressive dilatation of the segment of coronary artery resulting in the angiographic appearance of diffuse coronary ectasia as well as localised ectatic segments. These changes can also be described as fusiform or saccular aneurysms. It is therefore reasonable to surmise that coronary ectasia and coronary artery aneurysm may represent either end of the spectrum of a common pathological process.

The most common cause of coronary artery aneurysm is atherosclerosis as demonstrated in the CASS registry.[1] Other causes of coronary artery aneurysms are not common and can be classified as: inflammatory—Kawasaki disease that occurs mainly in infants and children,[4 5] Takayasu's disease, polyarteritis nodosa, and systemic lupus; abnormal connective tissue synthesis—Marfan's syndrome and Ehlers Danlos syndrome; infectious—septic emboli, syphilis, and Lyme borreliosis[6]; tumour (rare)—primary cardiac lymphoma[7]; congenital—coronary artery aneurysm[8–11]; and trauma (iatrogenic)—percutaneous transluminal coronary angioplasty,[12 13] intracoronary Cook stents,[14] and directional coronary atherectomy.[15–17]

Coronary artery aneurysms may be detected in the absence of symptoms[10]; however, patients sometimes present with angina, myocardial infarction,[18 19] or sudden death.[5] If rupture into a heart chamber occurs, a continuous murmur may be apparent and decompensation may ensue. Fistula formation into the right atrium or right ventricle has been reported.[18 20]

Coronary artery aneurysms can be detected non-invasively using echocardiography,[8 18 20] computed tomography,[8 11 20] and magnetic resonance imaging.[8] However, definitive diagnosis with coronary arteriography is usually needed accurately to delineate the anatomy and to ascertain the absence or presence of atherosclerosis elsewhere.

Because of the rarity of coronary artery aneurysm, the management of this condition is not clearly defined. The true incidence and natural history is uncertain as some patients may be entirely asymptomatic and the reported mortality may not reflect actual overall prognosis. Coronary artery aneurysms that present with life threatening complications such as heart chamber compression or fistula forma-

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tion, require prompt surgical intervention.[18][20] Some authors[21] argue that coronary artery aneurysm of at least three to four times the size of the original vessel diameter is an absolute indication for surgical intervention because of the propensity for complications such as compression, rupture or thrombosis. Others[2] argue that discrete coronary artery aneurysms do not appear to rupture, and these patients appear to have a favourable long term prognosis; therefore, elective resection in asymptomatic patients is not warranted. This has been supported by a case report of a patient with asymptomatic coronary artery aneurysm who remained free from complications eight years after initial diagnosis.[10] In asymptomatic cases, the risks of surgical intervention should be balanced by any potential risks if the aneurysm is left alone. In such cases, the benefits, if any, of antiplatelet or anticoagulation therapies are unknown.[19]

The proximal and middle segments of the right coronary artery (RCA) are the most common sites for coronary artery aneurysm, followed by the proximal left anterior descending (LAD) and the left circumflex arteries.[1] Coronary artery aneurysm of the left main stem is rare.[13][22]

We report two unusual cases of coronary artery aneurysm arising from different arteries but with similar presentations.

Case 1
A 33 year old man with no previous history of ischaemic heart disease presented with acute central chest pain. He was a non-smoker, with no history of diabetes or hypertension. There was a positive family history of ischaemic heart disease. Total fasting cholesterol was 5.7 mmol/l. There was no history or clinical features suggestive of acute inflammatory or connective tissue disease at presentation.

ECG on presentation revealed ST segment elevation across the anterior chest leads, and he was thrombolysed with streptokinase. Sequential cardiac enzymes showed a rise of creatine

kinase to a maximum of 785 U/l. Screening for autoimmune and inflammatory disorders were negative.

Coronary angiography revealed a large coronary artery aneurysm arising from the mid-LAD (fig 1), subtending an area of apical infarction, and flow distal to the aneurysm was noted to be slow. The rest of the coronary tree was normal.

The patient was initially managed conservatively with aspirin; however, he presented a month later with further chest pains and elevated ST segments across the anterior chest leads. He was thrombolysed with tissue plasminogen activator and this time the creatine kinase rose to a maximum of 761 U/l. A repeat study of coronary anatomy revealed similar findings to the first angiogram. Because of the slow flow of contrast to the distal LAD, it was postulated that the aneurysm was acting as a capacitance to blood flow, with stasis leading to thrombus formation and resulting distal emboli. He was therefore fully anticoagulated with warfarin, keeping the International Normalised Ration (INR) at ~ 3.

In view of the two successive presentations of myocardial infarction and the site of the aneurysm, he underwent surgical excision of the coronary artery aneurysm and an internal mammary artery was anastomosed end to side to the proximal and distal origins of the aneurysm, thereby preserving the diagonal branch.

Sections of the coronary artery aneurysm examined histologically showed no evidence of atherosclerosis, active inflammation or necrosis. There was marked intimal thickening by fibromyxoid tissue with reduplicated elastic fibres and a hypocellular, hyalinised focus. The appearances were not specific of Kawasaki disease but could have represented the late stage of an inflammatory disorder or a focus of fibromuscular dysplasia.

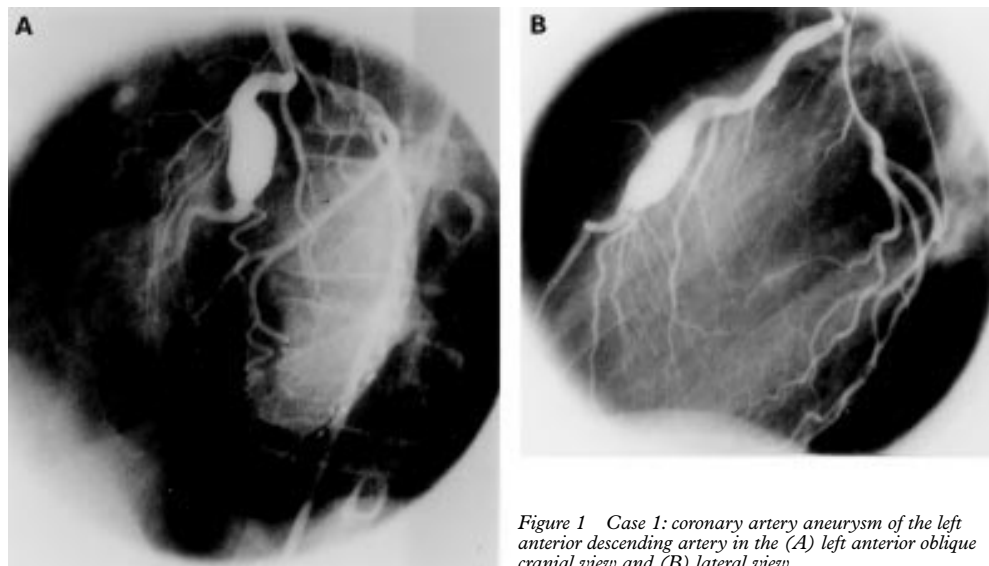The patient made a good recovery and has remained well four years after presentation.

Figure 1   Case 1: coronary artery aneurysm of the left anterior descending artery in the (A) left anterior oblique cranial view and (B) lateral view.
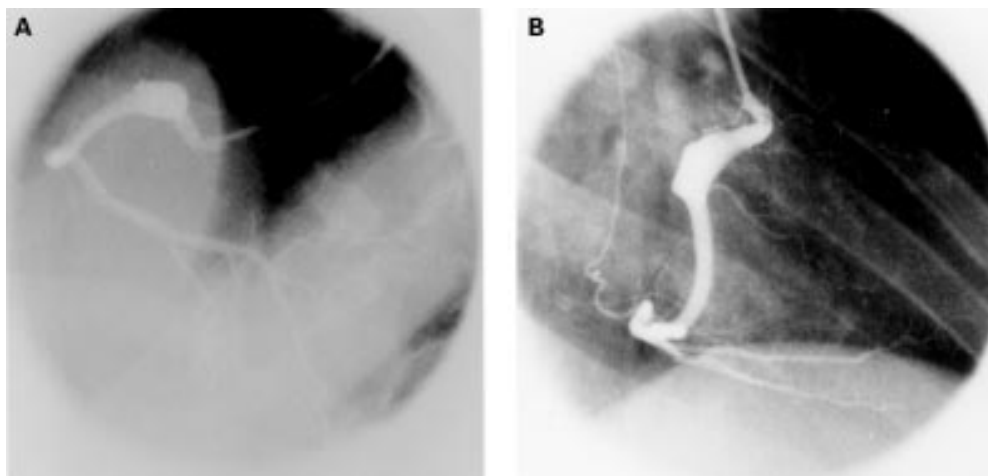
Figure 2   Case 2: coronary artery aneurysm of the left coronary artery in the (A) right anterior oblique cranial view and (B) the right anterior oblique view.

Case 2

A 37 year old woman presented with severe retrosternal chest pains 12 days after giving birth to her sixth child. She was previously fit and well, and her recent labour was uncomplicated. She did not experience any chest pains during labour. She was an ex-smoker of seven years, with no history of diabetes or hypertension. Both her parents had a history of myocardial infarcts in their later years. Her fasting total cholesterol was 6.3 mmol/l. There was no history or clinical features suggestive of acute inflammatory or connective tissue disease.

Initial ECGs showed T wave inversion in leads II and aVL only but an ECG two days later showed deep T wave inversion with Q waves in the inferior leads. Serial cardiac enzymes showed a slight rise of creatine kinase to a maximum of 297 U/l on day 2. She was not thrombolysed. Screening for autoimmune and inflammatory disorders was negative.

Coronary angiography revealed a normal left coronary system, with no evidence of atheroma. The RCA, however, had a large aneurysm arising proximally (fig 2). Left ventriculography revealed mild inferior wall hypokinesia with otherwise normal left ventricular function.

It was felt that the coronary artery aneurysm accounted for her symptoms; therefore, the patient was anticoagulated with warfarin. At follow up two months later a stress thallium study did not show any evidence of reversible ischaemia and the patient was well. Because the coronary artery aneurysm arose from the RCA, which was supplying only a small part of the left ventricle, it was felt that in the absence of symptoms or evidence of ischaemia, conservative management would be appropriate. The patient remained well six years after presentation.

Discussion

The diagnosis of congenital coronary artery aneurysm implies the exclusion of other acquired causes. In both the present cases, there was no evidence of atherosclerosis in the coronary arteries. Neither case had childhood histories suggestive of Kawasaki disease, nor was there evidence of acute inflammatory disease or other acquired causes. The histological specimen of case 1 was not conclusive. Although no histological specimens were available from case 2, it is possible that this case, as well as case 1, was a congenital abnormality. It is however impossible to rule out Kawasaki disease, as it can present undetected as a mild febrile episode in childhood, although this is uncommon. Coronary complications in Kawasaki disease usually occurs six to 12 months following the acute illness.[4 5]

Both our cases presented with evidence of infarction in the area of myocardium supplied by the culprit vessel. In the absence of atherosclerosis, it is likely the infarction was caused by thrombus formation within the coronary artery aneurysm resulting in embolisation to the distal artery.

Case 1 had surgical intervention because it was felt that the risk of further embolisation down the LAD was significant despite anticoagulation, and that there was potential for further damage to the left ventricle. In case 2, in view of the location of the coronary artery aneurysm in the RCA, it was felt that the risk of significant damage to the left ventricle was less than the risk of surgery, therefore, the patient was treated conservatively. Although little is known about the prevalence of distal embolisation or the benefits, if any, of antiplatelet or anticoagulation therapy,[19] we felt that case 2 would benefit from long term anticoagulation because of her presentation with a myocardial infarction.

The prognosis of coronary artery aneurysm appears to be dependent on the presence or absence of associated stenotic coronary artery disease. The greater incidence of infarction in patients with coronary artery aneurysm may only be a reflection of an overall increased prevalence of atherosclerosis. This is underscored by the fact that myocardial infarction is reported to be no more prevalent in patients with non-atherosclerotic coronary artery aneurysm.[1] Coronary artery aneurysms in Kawasaki disease are known to progress to obstructive lesions in later life.[4] The prognosis of congenital coronary artery aneurysm how-

ever is more uncertain because of its rarity. Those reported in the literature tend to be cases that present with complications although there has been one case report of asymptomatic congenital coronary artery aneurysm that remained free from complications for eight years.[10] Both our patients remain well four (case 1) and six (case 2) years after initial presentation.

In summary, we present two cases of non-atherosclerotic coronary artery aneurysm with similar presentations but different management strategies, both with favourable long term outcomes. These cases demonstrate the difficulties in determining the pathogenesis, and highlight the different management strategies, of isolated non-atherosclerotic coronary artery aneurysms. We believe the management of isolated coronary artery aneurysms should be individualised depending on their location and the clinical context.

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