Coronary artery aneurysms and myocardial infarction: adult sequelae of Kawasaki disease?

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SUMMARY Coronary artery aneurysms developed in a 43 year old man who had suffered an acute myocardial infarction at the age of 30. In childhood he had had an illness that was consistent with Kawasaki disease, and it is suggested that the proximal discrete aneurysms and myocardial infarction may be the adult sequela of this.

The arteriographic appearance of a coronary artery aneurysm is of localised dilatation of the vessel between two segments that are of normal calibre. Coronary artery aneurysms are not uncommon: they were found in 1.4% of necropsies performed in patients over the age of 16 years. They are usually arteriosclerotic in origin, but may be congenital, occur after injury, dissection, or infection, or be caused by polyarteritis nodosa. Mycotic and syphilitic coronary aneurysms are well described but more recently an additional possibly infective source has been recognised. Kawasaki disease was first described in 1967 in South West Japan as an acute febrile mucocutaneous syndrome with lymphoid involvement and desquamation of the fingers and toes in infants and young children. Coronary artery involvement is a prominent feature of Kawasaki disease and an important cause of death. The underlying pathological finding is an arteritis; and coronary aneurysms, ectasia, stenosis, and occlusion are all well recognised sequela.

Case report

A 43 year old male ambulance driver was referred for investigation of atypical chest pain. At the age of eight he had suffered a systemic, febrile illness with generalised lymphadenopathy, a mucocutaneous eruption, and alopecia. He had been in hospital for two months but no further details of this illness were available and no clear diagnosis was made.

He remained well until 1974 when, at the age of 30, he was admitted to hospital with an acute anteroseptal myocardial infarction confirmed on serial electrocardiograms and by cardiac enzyme activity. He made an uncomplicated recovery, and subsequent concentrations of fasting serum lipids were normal.

He remained well until 1986, when he was referred to the Brompton Hospital for investigation of atypical chest pain. He had been adopted and could give no family history, had never smoked, and had no history of venereal disease. He was normotensive and there were no abnormal physical signs.

An electrocardiogram was consistent with an old anteroseptal myocardial infarction. He completed 12 minutes of the standard Bruce protocol with a normal pulse rate and blood pressure response and no changes suggestive of myocardial ischaemia. A chest x ray and routine haematological, biochemical, and serological profiles were entirely normal. Cross sectional echocardiography showed two circular echo-dense areas approximately 1 cm in diameter in the region of the proximal left and right coronary arteries.

Cardiac catheterisation was performed and fluoroscopic screening demonstrated a circular thin rim of calcification just distal to the tip of the coronary catheter when this was engaged in the left coronary ostium (fig 1). Selective coronary arteriography in different projections further defined this lesion and showed that it was closely associated with the proximal left anterior descending artery and moved with it during the cardiac cycle. The left anterior...
Fig 1  X-ray in the left anterior oblique projection showing brachial coronary catheter engaging the left main stem coronary artery. There is a circular ring of calcification (arrowed), just distal to the tip of the catheter.

Fig 2  Selective contrast injection of the left coronary artery via a brachial catheter in the left anterior oblique projection. Contrast is seen to run through the circular ring of calcification (arrowed).
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oblique view (fig 2) was selected because it demonstrated the calcification most clearly, although in this view it seemed to overlie the left main stem. There was a slight irregularity of the left anterior descending artery at the same site, but otherwise the left coronary artery was normal. There was a similar fine, eggshell rim of calcification around the proximal right coronary artery that was clearly seen on fluoroscopy but was difficult to reproduce photographically. This lesion was also attached to the artery, which was itself mildly irregular at the same site. Apart from some slight distal ectasia the remaining right coronary artery was normal. Left ventricular wall movement was normal on cineangiography, with no evidence of previous infarction.

No specific treatment was started because the patient’s symptoms had largely resolved; at follow up after six months he remained symptom free.

Discussion

The aetiology of this patient’s coronary aneurysms cannot be determined with certainty, but there was strong circumstantial evidence of a childhood illness suggestive of Kawasaki disease. This is supported by the unusual appearance of the aneurysms which are not typical of those occurring in atherosclerosis. The irregularly narrowed lumen together with the surrounding eggshell rim of calcification resembles lesions found in Kawasaki disease. In addition, this patient had no risk factors for atherosclerosis, and there was no evidence for trauma, dissection, polyarteritis nodosa, syphilis, or congenital abnormalities being the underlying aetiology.

We suggest that the likely sequence of events in this patient was that coronary aneurysms formed after Kawasaki disease in childhood and that subsequent thrombus formation may have occurred within the sac of the left anterior descending artery aneurysm. Subsequent temporary occlusion of the artery at this point, or distal embolisation from the thrombus, could have caused the myocardial infarction.

Although it was first reported in 1967, Kawasaki disease is thought to have occurred endemically before then and is now known to occur both endemically and epidemically in children of all races in Asia, North America, and Europe. The coronary arteries are affected in 15–25% of cases, resulting in the development of aneurysmal or ectatic segments. Coronary insufficiency and myocardial infarction are known to occur after Kawasaki disease.

The prevalence of Kawasaki disease is increasing, with over 67 000 cases reported in Japan. A nation-wide epidemic was observed in Japan in April 1979, a second in 1982, and the third was reported during the winter of 1985–6.11 The epidemiological patterns of the disease suggest an infectious aetiology by an unknown agent, possibly a retrovirus.12

Many types of treatment have been used in Kawasaki disease but none is entirely effective. A standard treatment for the associated arteritis has been aspirin. Newburger et al13 concluded from a multicentre randomised trial that high dose intravenous γ globulin is effective in reducing the incidence of coronary artery abnormalities if given early in the course of the illness.

We believe that our patient had Kawasaki disease as a child and that his early myocardial infarction and coronary aneurysms are a consequence of this. Similar cases have been reported,14 but never with strong circumstantial evidence of the childhood illness, or such unusual angiographic appearances. If the prevalence of the condition rises in the United Kingdom, as has happened in the United States and Japan, then such sequelae may become more familiar.

References
512

Brecker, Gray, Oldershaw


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