

Sudden death in a young adult due to coronary artery aneurysm secondary to suspected Kawasaki disease

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Abstract

A healthy 17-year-old Chinese male suddenly collapsed and died during a game of badminton. The autopsy examination revealed a solitary calcified aneurysm of the left common coronary artery with marked stenosis of the orifices of the anterior descending and **circumflex** branches. Histology of the aneurysm was non-specific with hyalinised scar tissue and foci of calcification. The only illness of significance in the past was an episode of 'pyrexia of unknown origin' at the age of 8 months. A review of the notes of that hospital admission revealed that the illness was most probably Kawasaki disease.

Key words: Sudden death, young adult, coronary artery aneurysm, Kawasaki disease.

CASE REPORT

During a game of badminton, a 17-year-old Chinese male became suddenly dyspnoeic and collapsed soon after. Despite prompt admission to hospital and cardio-pulmonary resuscitation, he did not survive. The deceased had shown no symptoms of any ill health; in fact he had been quite active, playing regularly strenuous games of badminton. In the past he had had no significant illness except an episode of high fever at the age of 8 months (in 1970) for which he was admitted to the University Hospital Kuala Lumpur. The medical records revealed that at admission, in addition to the fever he had bilateral conjunctivitis, rhinorrhoea, injected pharynx, generalised maculo-papular rash and significant tachycardia.

The fever was not responsive to antibiotics but remitted spontaneously after 20 days of illness.

The autopsy examination revealed that the deceased was a fairly well-nourished 17-year-old Chinese male, 172.0 cm in height and 52.6 kg in weight. The abnormal anatomic pathology seen was in the cardiovascular system. The heart weighed 250.0 gm and was neither hypertrophied nor dilated. There was no evidence of valvular or myocardial abnormalities. There was a fusiform, hard (calcified) aneurysm, 1 cm x 1 cm x 1 cm of the left common coronary artery (Figs 1 and 2). The orifices of the left anterior descending and the left circumflex arteries were markedly stenosed. The right coronary artery, and the rest

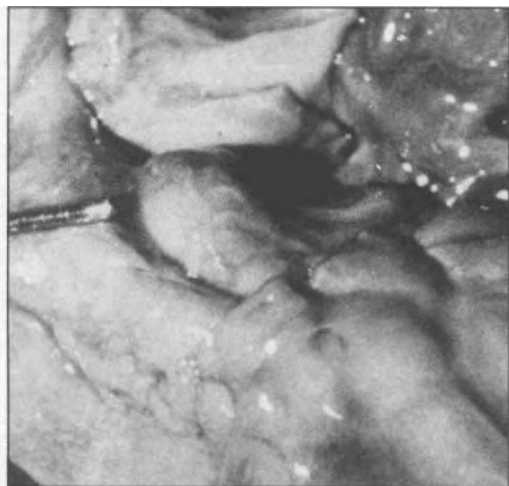


FIG. 1: Aneurysm of the left anterior descending coronary artery at its origin, indicated by the probe.



FIG. 2: Cut surface of the aneurysm shows a hard calcified wall, indicated by the probe.

of the left anterior descending and the circumflex branches did not show any aneurysms or other abnormalities. No thrombi were seen in the aneurysm or in the coronary arteries. The coronary ostia were unremarkable. Histology of the aneurysm showed hypocellular, hyalinised fibrous tissue with areas of calcification (Figs 3 and 4). The intimal and external elastic laminae were not present. There was no inflammatory cellular infiltrate. Microscopic examination of the myocardium did not reveal any abnormalities. There was no significant atheroma of the aorta or other major arteries. All the other internal organs were unremarkable.

DISCUSSION

Though uncommon, coronary aneurysms are now being increasingly recognised and diagnosed. Based on the aetiological classification generally used,^{1,2} the most common cause is atherosclerosis. This occurs, as expected, in the older age group. However, post-inflammatory aneurysmal dilatation is the more common cause in the younger age group. The inflammatory conditions include polyarteritis nodosa, bacterial endocarditis and the more recently recognised Kawasaki disease. In the case under discussion, the most probable cause of the coronary aneurysm is coronary arteritis due to Kawasaki disease. Kawasaki disease or Mucocutaneous Lymph Node Syndrome (MCLS) was first described by Kawasaki in Japan in 1967.³ Since then the number of cases diagnosed has increased both in Japan as well as in other countries. The principal clinical features useful in the diagnosis of this disease, as outlined by the Japan MCLS Research Committee⁴ are:

1. High spiking fever lasting at least five days
2. Bilateral non-exudative conjunctivitis
3. Dry, red, cracked lips; red oral and pharyngeal mucosa; and 'strawberry' tongue
4. Initially red and swollen hands, then characteristic desquamative skin lesions, particularly around the fingers, toes, palms and soles
5. Widely distributed polymorphous rash without vesicles or crust
6. Lymphadenopathy - the cervical lymph nodes are commonly involved; at least 1.5cm in diameter

Based on the clinical signs and symptoms recorded (Table 1),⁵ this patient's illness at the age of 8 months was most probably Kawasaki disease. It is not surprising that no definite diagnosis was made at that time as Kawasaki disease was then not well known.

One of the important complications during the acute phase of this illness is arteritis especially of the coronary and iliac arteries.^{3,4,6,7} Coronary artery involvement has been reported to occur in about 15% of all patients of Kawasaki disease.^{3,8} A small percentage (<2%) of these patients die during the acute illness, from acute myocardial infarction resulting from thrombosis in the coronary aneurysmal dilatation.^{4,7} Many of the surviving patients die of this cardiovascular complication during the first year after the onset of the illness. With improved 2-dimensional echocardiography and angiography, as well as from autopsy data, it is now known that there is a high rate of persistence of coronary artery lesion (aneurysm) secondary to Kawasaki disease. Early studies in Japan showed that about 50% of these coronary artery lesions resolved within 6 to 18 months.^{9,10} How-

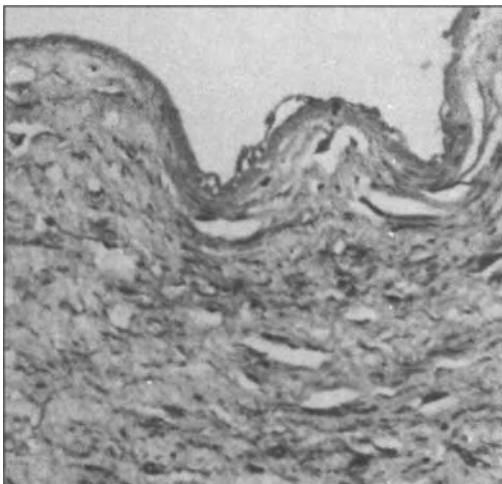


FIG. 3: Histology of the aneurysmal wall showing mainly fibrous tissue. H&E x 100.

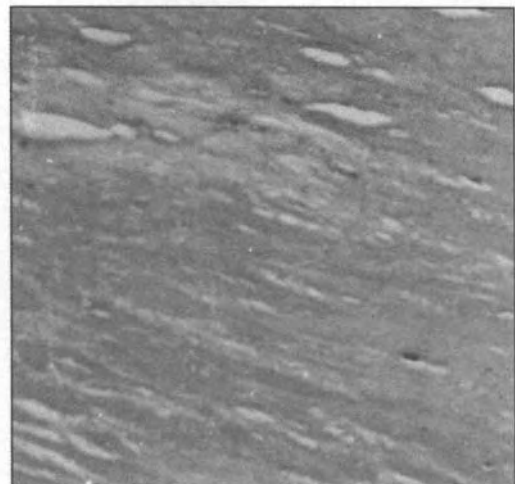


FIG. 4: Hyalinised fibrous tissue of the aneurysmal wall. H&E x 200.

TABLE 1: Features of Kawasaki disease noted in this patient

Principal diagnostic features	Symptoms and signs noted
1. high spiking fever >5 days	high fever >20 days
2. bilateral non-exudative conjunctivitis	bilateral conjunctivitis
3. dry, red cracked lips; red oral and pharyngeal mucosa; 'strawberry' tongue	injected pharyngeal mucosa
4. polymorphous rash without vesicles or crust	generalised maculo-papular rash
5. lymphadenopathy - neck	not stated
6. red and swollen hands; then desquamative lesions in the palms and soles	not stated
Other important feature: coronary artery aneurysm(s) demonstrated by 2-D echo-cardiography or coronary angiography	a single aneurysm of the left coronary artery demonstrated at autopsy

Note: At least 5 out of 6 of the principal diagnostic features should be present to diagnose Kawasaki disease. However, patients with 4 of them can be diagnosed if presence of coronary aneurysm(s) is demonstrated either by 2-D echocardiography or coronary angiography or at autopsy.⁵

ever, a more recent study in Canada showed that the resolution rate was only about 15%.³ The reason for this high persistence rate is unclear, but it was suggested that ethnic differences and longer duration of high fever could be important factors involved. Some of these patients suffer years later, in their teens, from acute myocardial ischaemia or infarction. Very often these patients are asymptomatic until they develop acute myocardial infarction or sudden death, and this usually follows some strenuous physical activity.⁶

The histology of the aneurysm in this case is compatible with a healed arteritis. Atherosclerosis as a cause can be excluded because no demonstrable atheromatous lesion is present in the aneurysm or the coronary arteries. Formerly, many cases of coronary aneurysms in young adults were thought to be congenital in origin.¹ However, it is now believed that the majority of them result from previous inflammation, as the histology in these cases is quite non-specific consisting mainly of fibrous tissue.^{2,3,5,6}

This case report, like that in a few others,^{2,5,11} reestablishes that (1) coronary artery aneurysm causes sudden cardiac death in young adults, (2) Kawasaki disease is an important cause of coronary aneurysm(s) and (3) the fatal, late sequelae of Kawasaki disease is due to the persistence of the coronary aneurysm(s). Hence, the detection and treatment of coronary aneurysm(s) in Kawasaki disease should not only be in the acute stage of the illness, but also in the long term follow up of such cases.

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