Short reports

Sudden death in incomplete Kawasaki's disease

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SUMMARY A boy of 12 years died suddenly of myocardial infarction, which was due to coronary artery aneurysm typical of Kawasaki's arteritis. Two years earlier he had an obscure illness, recognised retrospectively as incomplete Kawasaki's disease. We recommend routine echocardiography even if only one or two features of the syndrome are present.

Kawasaki's disease is typically a disease of young children and the diagnostic criteria have been clearly defined.¹ Most children present between the ages of 1 and 5 years and the disease is rare after 7 years.² The association with coronary artery aneurysms and sudden death is also well established.³ Recently incomplete Kawasaki's disease with coronary artery involvement has been described in five young children aged between 4 months and 2½ years, one of whom died in intractable heart failure.⁴ We describe a case of a child of 12 years who died suddenly of myocardial infarction due to thrombosis of an abnormal left coronary artery which showed the features of Kawasaki's disease. Two years before his death he had had an illness, which retrospectively may have been incomplete Kawasaki's disease.

Case report

This boy, who had previously been well, was admitted at the age of 10 years with a 10 day history of pain in the region of the left ear and throat, and fever. He had been treated with cotrimoxazole without effect. On examination he looked ill and pale, he was holding his head still, and had difficulty in opening his mouth. He was afebrile, and there was no obvious swelling in the neck. He had a few slightly enlarged lymph nodes in the groin. Initial investigation showed a neutrophil leucocytosis (total white count $23.2 \times 10^9/l$, neutrophils 91%) with toxic changes, a thrombocytosis (platelets $753 \times 10^9/l$), and a raised erythrocyte sedimentation rate of 107 mm in the first hour. Biochemical investigations and examination of the cerebrospinal fluid gave normal

results. A radiograph of the neck showed increased lucency of the left mastoid process; ultrasound examination of the neck was normal.

A diagnosis of osteomyelitis of the mastoid was made and he was treated with intravenous fusidic acid and erythromycin. Over the next seven days he had a moderate intermittent fever, which then settled. His erythrocyte sedimentation rate on the third day after admission was 118 mm in the first hour and thereafter slowly declined. His platelets peaked at $1206 \times 10^9/l$ on the fifth day but remained raised throughout his admission. All cultures, virological investigations, tests for antistreptolysin O titre, antinuclear factor, anti n-DNA, mitochondrial antibody, smooth muscle antibody, and parietal cell antibody gave negative results. He developed a hard tender mass at the insertion of the sternomastoid, which gradually declined. On the ninth day after admission it was noted by the parents and recorded by the nurses that the tops of his fingers were peeling. He remained rather pale and miserable throughout his stay in hospital.

He was followed up in outpatients for several months; a fibrotic mass at the sternomastoid insertion gradually resolved; he remained easily tired and prone to viral infections. On one occasion he described a single episode of acute muscular pain in the chest and shoulder, which had resolved with paracetamol. Thereafter he remained very well. He had a normal exercise tolerance, took part in all sport, and did not ever complain of chest discomfort. Twenty three months after the illness he had been out walking with a friend, felt tired and went to lie down. A little later he complained of chest discomfort, then suddenly turned over, clutched at his chest, and died almost immediately. Attempts at resuscitation by the ambulance team and in the accident and emergency department were of no avail.

PATHOLOGY FINDINGS

At postmortem examination the heart weighed 220 g and showed aneurysmal dilatation of the left stem and proximal left anterior descending artery (fig 1). The dilated segment measured 2.5 cm in diameter

and contained old and recent thrombi. The distal left anterior descending artery showed complete obliteration of the lumen by firm greyish tissue. The proximal portion of the right coronary artery showed similar luminal occlusion; the circumflex artery remained patent. Microscopically the left coronary artery in its distended portion showed a loss of normal smooth muscle and replacement by fibrous connective tissue. The distal left anterior descending artery (fig 2) showed luminal occlusion by cellular fibrous connective tissue, representing an organised thrombus. Recanalisation had occurred as evidenced by vascular channels of various size.

There was extensive fibrosis of the media and only a few disrupted remnants of the internal elastic

lamella were noted. The lungs showed oedema and congestion in keeping with acute heart failure after myocardial infarction due to thrombosis of an abnormal left coronary artery.

Discussion

Kawasaki's disease is a generalised vasculitis of unknown aetiology, which differs from the other vasculitic disorders by its propensity to effect the coronary arteries. The macroscopic and microscopic features of the coronary arteries in this case are typical of late stage Kawasaki's disease. This is well described by Hamashima as occurring between 40 days and four and a half years after the disease onset, and characterised by arterial scarring and recanalisation, in addition to the presence of aneurysms.³

In this case the acute attack occurred two years before the fatal thrombosis. A similar case of sudden death has been described in a 12 year old boy four years after the initial illness, with coronary artery pathology identical to this case.⁵ This presentation was unusual in that there were no specific signs of Kawasaki's disease apart from finger desquamation. We conclude that this 'incomplete form' of Kawasaki's disease still carries the risk of coronary artery involvement.

There is now evidence that long term treatment with antiplatelet therapy may prevent secondary coronary thrombosis in adults.⁶ The management of aneurysms of the coronary arteries in Kawasaki's disease is still controversial; currently low dose aspirin treatment is recommended for six months after the acute illness, but it may be appropriate to continue this treatment for much longer. Furthermore, coronary artery bypass surgery has been undertaken successfully in children with severe coronary artery abnormalities after Kawasaki's disease.⁷

We suggest that any child presenting with an illness associated with a raised platelet count and peeling of the fingers should have echocardiography to exclude lesions of the coronary arteries.

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Fig 1 A suture needle points to an aneurysm of the proximal left anterior descending artery.

Fig 2 The distal left anterior descending coronary artery shows evidence of old thrombosis and recanalisation. The internal elastic membrane is disrupted and frayed as a sequela of the initial vasculitis (elastic Van Gieson $\times 25$).

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A new chart for weight control in Duchenne muscular dystrophy

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SUMMARY Weight control is desirable in the muscle wasting conditions. A new chart is presented to allow the prediction of an ideal weight, free of excess fat, specifically for boys with Duchenne muscular dystrophy.

Patients suffering from muscular dystrophy often put on weight as the disease progresses. It is not uncommon for boys with Duchenne muscular dystrophy to have weight values greater than the 90th centile on normal growth and development charts, despite having a smaller muscle mass than normal boys of the same age. It has been shown that therapeutic weight reduction is possible in these patients,¹ and loss of unnecessary fat lessens the burden of weakened muscles and aids mobility.

Prevention of excessive weight gain in the younger patient is preferable to severe dietary restriction in the already obese subject. Normal growth and development charts make no allowance for the progressive loss of muscle that is occurring as the muscular dystrophy progresses throughout childhood. A child with pronounced muscle wasting looks 'cachectic' without the excess fat filling out his body contours, something parents find hard to accept. The loss of muscle coupled with reduced activity implies accumulation of fat tissue if weight gain approximates that of normal growth charts.

Method

The 24 hour excretion of creatinine in the urine as a breakdown product of phosphocreatine in muscle, assuming 1 mmol creatinine equals 2.2625 kg of muscle, provides a reliable estimate of the contribution of muscle to body weight in the absence of renal disease.²

Theoretical weight centiles for zero muscle mass can be drawn from data on normal creatinine excretion¹ and from standard weight and height centile charts.³ From a knowledge of the patient's creatinine excretion the muscle bulk may be derived and added to the zero muscle weight chart to produce an ideal weight.

Results

A new chart is presented, which gives ideal weight guidelines for weight control in boys with Duchenne muscular dystrophy (figure). From observations of body composition and creatinine excretion in boys with Duchenne muscular dystrophy we have found a close relationship between the reduction in total body potassium and that of 24 hour urinary creatinine excretion.⁴ Both total body potassium and urinary creatinine excretion, when related to the predicted values for the individual's height, declined with age at an average rate of 4% per year from 6 to 17 years.⁴ A similar 4% per year loss in muscle strength has been observed in boys with Duchenne muscular dystrophy.⁵ At 6 years a boy with Duchenne muscular dystrophy has only 60% of his predicted muscle mass, and at 16 years only 20%. The reduction in creatinine excretion can be calculated and from this the muscle bulk can be derived and added to the zero muscle weight charts. Based on this assumed 4% per year decline in muscle mass the 90th, 50th, and 10th 'ideal' weight centiles have been calculated to allow for progressive loss of muscle.

Discussion

This chart can be used in the normal way by selecting the appropriate height related centile. This chart is **only** applicable for Duchenne muscular dystrophy. In other muscle wasting conditions