Sudden Cardiac Death in a normally developed male Infant: An atypical presentation of Kawasaki disease?

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Introduction

Kawasaki disease is a medium vessel vasculitis of childhood associated with the mucocutaneous lymph node syndrome. It affects medium and small arteries with a predilection for coronary arteries, leading to acute and long-term complications – stenosis, dilatation and aneurysm formation. 85% of cases occur in the under 5’s, with males more commonly affected than females, 3:2. Most reported cases are in children of East Asian origin with the highest incidence in Japan, 138/100,000, compared to 8.1/100,000 in the UK. An increasing incidence is reported in East Asia, despite decreasing birth rate.

The aetiology of Kawasaki Disease is, as yet, unknown. The age distribution, seasonal variation and endemic nature of the disease support an infectious cause, whilst male predominance, racial susceptibility and familial cases point to a genetic susceptibility. Diagnosis is not straightforward as there is no diagnostic test. The disease is characterised by fever, lasting for more than five days, plus 4 out of 5 diagnostic criteria as established by The Japanese Ministry of Health Research Committee (table 1). Clinical features appear sequentially, so Kawasaki Disease should remain in the differential diagnosis in any child with ongoing fever. Prompt diagnosis and treatment can reduce coronary complications; 15-25% of untreated patients will develop coronary artery aneurysms compared to <10% in treated patients. Incomplete Kawasaki Disease is a recognised variant in which fewer than 4 diagnostic criteria are met, there is a 15-20% risk of coronary artery dilatation in these children.

Case

We report the case of a previously healthy male of mixed ethnicity who died unexpectedly at home, aged 3 years and 4 months. Relevant past medical history included two episodes of eye inflammation; shingles; and hand, foot and mouth disease. Most recently, 2 months prior to death, dry eczema of the face and trunk, chesty cough and fever was diagnosed (see table 2). Other diagnoses included recurrent dermatophytosis at 5, 7 and 18 months of age; upper respiratory tract infection with fever at 9 months, requiring observation in hospital for one morning. There were no overnight hospital admissions and no reports of prolonged fever.

Findings at Autopsy

- A well nourished, normally developed male infant; facial features suggesting South East Asian origin.
- Several crusted lesions present on the face.
- Pericardial effusion and enlarged heart, weight 114 g (normal 59 g).
- Giant aneurysm of the left coronary artery, with thrombus.
- Intimal thickening of the right coronary artery with a pinpoint lumen through the first 40 mm.
- Subendocardial pallor of the left ventricle.
- Heavy, congested, oedematous lungs, weight 384 g (normal 168 g).

Histological findings

- Acute myocardial infarction of the left ventricle.
- Myocardial fibrosis of the left ventricle.
- Intimal thickening and smooth muscle hypertrophy of the left anterior descending and right coronary arteries, with intra-luminal collagenisation, recanalization and dystrophic calcification.
- No necrotising vasculitis.

Discussion

The diagnosis of Kawasaki Disease is based on autopsy findings. Giant aneurysm, coronary artery remodelling with recanalization and dystrophic calcification together with a lack of necrotising vasculitis support a more chronic course as opposed to acute illness. Myocardial fibrosis is highly suggestive of previous myocardial infarct.

Review of the past medical history (table 2) raises questions about whether the infant’s previous illnesses form part of an emerging spectrum of Kawasaki Disease but with an atypical presentation. Many of the clinical diagnoses could almost fit into current diagnostic criteria (table 1), albeit well separated in time, occurring over a 15 month period. There are two separate reports of eye inflammation, possibly conjunctivitis although it appears that only one eye was affected where the diagnostic criteria state bilateral, without exudate. A diagnosis of shingles is incredibly unusual in an infant and should be questioned. Was the diagnosis of hand, foot and mouth disease criteria 4 and 5? Or was the final illness that warranted a visit to the GP (dry eczema, fever and chesty cough) actually the first presentation of Kawasaki Disease?

With increasing globalisation, racial profiling is particularly important in modern medicine. A high index of suspicion is warranted in febrile children of East Asian origin, in whom a family history of Kawasaki Disease should also be routinely sought.

References: