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Kawasaki disease in siblings

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SUMMARY We report a case of mucocutaneous lymph node syndrome, which was diagnosed in a 13 month old boy two years after the death of his sister from the same disease.

The aetiology of mucocutaneous lymph node syndrome, or Kawasaki disease, remains elusive, although it was first reported in 1967.¹ Environmental, genetic, and infective causes have all been postulated. We describe the non-contemporaneous occurrence of the disease in two children within the same family.

Case reports

CASE 1

In April 1983 a 6 month old girl was admitted for investigation of a prolonged fever. She had a two week history of ill health including coryzal symptoms, cervical lymphadenopathy, mild conjunctivitis, and a macular rash on her trunk. There had been no contact with infectious disease or travel abroad. Results of investigations are shown in the table.

On day 28 of her admission she developed mucousy diarrhoea and adenovirus type 5 was isolated from the stool. An adenovirus titre before this was negative but subsequently there was a significant rise to a titre of 64.

Her clinical condition deteriorated and nine days later she had a cardiac arrest. Resuscitation was successful and subsequent examination showed cardiac failure and gross cardiomegaly. A diagnosis of viral myocarditis was made. Despite symptomatic treatment there was further deterioration and the child died seven weeks after admission.

Postmortem examination found widespread

coronary artery aneurysms with antemortem thrombi and extensive infarction. There was no evidence of pericarditis or myocarditis. The final diagnosis was therefore mucocutaneous lymph node syndrome.

CASE 2

A boy, born to the same parents two years after the death of his sister (case 1), was well until 13 months of age. He was seen with coryza, prolonged fever, cervical lymphadenopathy, conjunctivitis, and a macular rash over the trunk. Results of investigations are shown in the table. Echocardiography showed an area of echodensity at the origin of the

Table Results of investigations

Investigation	Case 1	Case 2
Haemoglobin (g/l)	100	113
White cell count ($\times 10^9$ g/l)	16.8	15.3
Neutrophil (%)	60	54
Lymphocytes (%)	29	44
Platelets ($\times 10^9$ /l)	414	430
Erythrocyte sedimentation rate (mm in first hour)	116	66
Alanine aminotransferase (IU/l)*	264	30
Blood cultures	Negative	Negative
Complement fixation test:		
Respiratory syncytial virus	Negative	Negative
Adenovirus	Positive	Negative
Mycoplasma	Negative	Negative
Herpes simplex	Negative	Negative
Measles	Negative	—
Rubella	Negative	—
Neutralisation test:		
Coxsackie B	Negative	—
Paul-Bunnell	Negative	Negative
Throat swab	Negative	Negative

*Normal range 0-45 IU/l.

Figure Echocardiogram of case 2 (a) showing cross sectional view of aortic root at presentation and thickening at the origin of the right coronary artery (RCA); (b) showing cross sectional view of aortic root at six months with partial resolution of the thickening at the origin of the right coronary artery. AO, aortic root; LA, left atrium, PA, pulmonary artery.

right coronary artery (figure (a)), which was consistent with an inflammatory process. No aneurysms were found.

A diagnosis of mucocutaneous lymph node syndrome was therefore made and the child successfully treated with aspirin and gammaglobulin. Follow up echocardiography at six months showed partial resolution of the echodense area but there remained a residual thickening of the coronary artery walls (figure (b)).

Investigation of the parents and the other sibling was not undertaken.

Discussion

There have been previous case reports of siblings and twins with mucocutaneous lymph node syndrome but these have all occurred at the same time and thus lent support to the infectious aetiology.² Data on genetic factors such as HLA similarities have been inconsistent despite some evidence of association with linkage between HLA-A2, B44, and CW5 antigens.³ A Japanese twin study failed to find any evidence of an excess of cases associated with genetic HLA identity.⁴ The possibility of

an environmental trigger—microbial or toxin mediated—precipitating an immune cascade and causing the release of platelet derived mediators in genetically predisposed individuals seems the most attractive hypothesis. Our case lends clinical support to this suggestion.

We thank the late Dr CR Tribe for his histopathological opinions on case 1.

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