Mucocutaneous lymph-node syndrome (Kawasaki disease): probable soluble-complex disorder

The first description in English of the mucocutaneous lymph-node syndrome (MLNS)[1] was followed by reports from Hawaii, Canada, the United States, and, recently, Britain. We report a further case.

Case report

A previously healthy 22-month-old Japanese boy born in Osaka had lived in England for 17 months. He became ill with fever, redness of the hands and feet, and a rash on the trunk and limbs and then developed facial oedema with diarrhoea and vomiting. He was admitted to hospital on the eighth day because of pain and swelling of the neck and inability to swallow.

The patient was feverish, drowsy, and dehydrated, and had neck stiffness, facial oedema, conjunctivitis, cracked, bleeding lips, a red pharynx with prominent tongue papillae, and pronounced cervical adenitis. As the rash faded peripheral oedema became evident and was followed by a striking desquamation of the fingers spreading to the hands and arms. He returned home on the 22nd day, and five days later similar desquamation began on the toes and feet. During the second month we noted a transverse groove on the nails of the fingers and toes. At no time could we detect any abnormality of the cardiovascular system. He was given intravenous fluids and benzylpenicillin for 48 hours followed by cloxacillin for 48 hours until a diagnosis was made on the fourth day. Numerous investigations on blood, urine, cerebrospinal fluid, and faeces failed to yield a pathogenic organism. The table gives the results of haematological studies.

Comment

The findings in this case were identical with those described in MLNS. Many features of the disease suggest an acute infective aetiology but no consistent pathogen has been isolated. Several immunological changes have been recorded. Increases in serum IgA and IgM have been observed but no specific immunoglobulin changes were noted until Kusakawa and Heiner reported that the serum IgE was increased in 20 patients during the acute illness and then declined over the next two months.[2] Raised concentrations of IgE also occur in infantile polyarteritis, and IgE may play a part (by increasing vascular permeability, thus permitting leucocyte migration and antigen-antibody deposition) in the development of the vasculitis. There is no evidence that children with pre-existing high concentrations of IgE are particularly susceptible to MLNS, and family studies have not shown an increased incidence of allergic disorders. Our patient's IgE concentration was in the upper normal range during the acute phase of the illness and declined on recovery.

Changes in serum C3 have been noted in two patients. A reduced concentration was recorded on day 7 in one[3] and a raised value at an unspecified time in another.[4] Transient changes in serum complement concentrations may be expected in MLNS as in other acute illnesses, and repeated estimations are required to determine their significance. Our patient had low concentrations of both haemolytic complement (CH_{50}) and C4 on day 12 of the illness and increased concentrations of CH_{50}, C4, and C3 during recovery. In addition the platelet aggregation test for detecting circulating immune complexes[5] gave a strongly positive result on day 12, the activity declining to normal during recovery.

Although these observations require confirmation, the changes were similar to those found in other immune-complex disorders. Circulating immune complexes and increased concentrations of IgE possibly play a part in the pathogenesis of MLNS and may be responsible for the major complications.

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[1] Kawasaki, T, *et al*, *Pediatrics*, 1974, **54**, 271.
[2] Kusakawa, S, and Heiner, D C, *Pediatric Research*, 1976, **10**, 108.
[3] Melish, E, Hicks, P M, and Larson, E, *American Journal of Diseases of Children*, 1976, **130**, 599.
[4] Brown, J S, *et al*, *Journal of Pediatrics*, 1976, **88**, 81.
[5] Myllylä, G, *Scandinavian Journal of Haematology*, 1973, suppl No 19.

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Case of Kawasaki disease

In 1967 Kawasaki described 50 Japanese children with a febrile condition that he called the mucocutaneous lymph-node syndrome (MLNS). Though it resembles the Stevens-Johnson syndrome, genital lesions do not occur and the stomatitis and conjunctivitis are not associated with vesiculation or pseudomembrane formation. In 1974 Kawasaki *et al*[1] detailed the clinical and epidemiological features. The disease appeared to be confined to Japan until Melish *et al*[2] reported nine cases in multiracial children in Hawaii. Further reports have come from the USA, South Korea, Greece, Australia, and, recently, Britain. Japanese workers regard the condition as a disease entity and refer to it as Kawasaki disease.

I report a possible case of the disease in a child admitted to this hospital in July 1975.

Case report

A 5-year-old girl of Italian parentage who had lived in England since birth was seen with a 12-hour history of abdominal pains and vomiting. Fever and bilateral ocular congestion were noted. The vomiting settled within 24 hours. On the second day she started a course of amoxycillin. She then had circumoral redness, which, together with fever, persisted until admission two weeks later. A week before admission pronounced redness of the hands and feet developed, with desquamation starting the next day.

On admission she was found to be a well-nourished, febrile girl with a pulse rate of 136/min. The circumoral skin was erythematous and cracked, and yellow crusting was noted. She had stomatitis, and her tongue was partly coated with white plaques. Upper cervical lymph nodes were enlarged and tender. There was palmar and volar erythema, with desquamation occurring

Results of haematological investigations during illness

Day of illness:	8	10	12	17	23	40	109
Haemoglobin (g/dl)	11·3	10·3	10·1	9·8	9·3		
White cell count × 10^9/l (% neutrophils)	26·3 (90)	27·9 (91)	28·3 (88)	11·0 (80)	11·3 (68)		
Platelet count × 10^9/l	Normal	Normal	510	600	600		
Sodium (mmol/l)	127	136	135				
Urea (mmol/l)	2·4	1·4	3·0	4·0			
Albumin (g/l)		21		21	29		
IgG (g/l; normal 5-13)	9·20				10·75	13·82	7·65
IgA (g/l; normal 0·2-1·6)	1·82				1·41	1·28	0·29
IgM (g/l; normal 0·35-1·50)	0·90				1·12	1·80	1·26
IgE (μ/ml; normal 50-200)			130			200	85
CH_{50} (μ/ml; normal 26-40)			10			>40	
C3 (g/l; normal 0·78-1·61)			1·25		2·44	2·25	1·38
C4 (g/l; normal 0·15-0·45)			0·16		0·70	0·44	0·32
Platelet aggregation (normal <1 in 20)			1 in 320		1 in 40	1 in 20	Negative

Conversion: SI to traditional units—sodium: 1 mmol/l = 1 mEq/l. Urea: 1 mmol/l ≈ 6 mg/100 ml.