Acute renal failure in Kawasaki disease[1]

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Since Kawasaki (1967) described an acute mucocutaneous lymph node syndrome, a large number of cases have been reported, mainly in Japan but also in the United Kingdom (Lyen & Brook 1978, Morgan & Lynch 1978). A clinical spectrum has evolved, and many unusual features reported: myocarditis, arthritis, and hydrops of the gallbladder (Kawasaki *et al.* 1974, Yanagihara & Todd 1980). This paper describes a child with Kawasaki disease who developed acute renal failure during the first week of his illness. A renal biopsy on the tenth day revealed a patchy 'immune' type infiltrate, a finding not previously described in this syndrome.

Case report

GH, a previously well 2-year-old Caucasian male, was admitted to hospital in January 1981 with a 24-hour history of marked pyrexia, profuse diarrhoea with vomiting, and delirium. He was extremely ill with hyperventilation, central cyanosis, a marked tachycardia and unrecordable blood pressure. He was unresponsive to painful stimuli and had generalized twitching. There was notable conjunctival injection, but no evidence of neck stiffness or papilloedema.

Investigations revealed a haemoglobin of 14.6 g/dl, white cell count 9.6×10^9 with 86% neutrophils, platelets 314×10^9. A lumbar puncture yielded clear fluid, with no cells, protein 0.3 g/l and glucose 3.7 mmol/l. Blood gas analysis showed a marked metabolic acidosis, bicarbonate 10 mmol/l, and an associated respiratory alkalosis. His biochemistry was disturbed; urea 22 mmol/l, sodium 128 mmol/l, potassium 4.8 mmol/l, amylase 965 mmol/l, calcium 1.7 mmol/l, total protein 61 g/l, no salicylate was detected.

His initial management included fluid replacement, correction of acidosis, and elective ventilation with muscle paralysis. There was some improvement in circulating volume and urine output. However he became more hypertonic, with evidence of a right hemiparesis. A computerized brain scan was normal. At forty-eight hours he developed intense redness of his lips and buccal mucous membranes, and a generalized erythematous rash, which desquamated during week two; his limbs had a characteristic woody type of sclerema, with no evidence of oedema. His urea had risen to 32 mmol/l, potassium 6.6 mmol/l, there was a fall in haemoglobin to 9.6 g/dl, white cell count 13.4×10^9, platelets 93×10^9; there was no evidence of a microangiopathy on the blood film, and fibrin degradation products were normal. The urine output was negligible and was positive to stix testing for blood and protein. Treatment for hyperkalaemia was commenced, and the patient was transfused with packed red cells. Early on the third day while being ventilated he had a cardiac arrest, but was swiftly resuscitated. His urea had risen to 37 mmol/l, potassium was 6.6 mmol/l. He was given 12 units of soluble insulin in 20 ml of 50% dextrose over 20 minutes to control hyperkalaemia, but his urine output failed to increase following a frusemide injection. Peritoneal dialysis was therefore commenced, and after eight days there was a diuresis, with return of his biochemistry to normal.

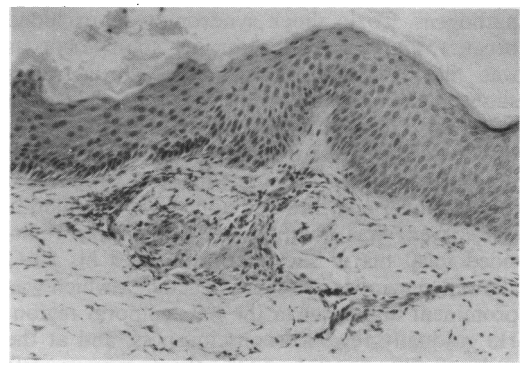

Figure 1. Photomicrograph of a skin biopsy showing a resolving leukocytoclastic vasculitis in a small vessel high in the dermis. (H & E ×375)

A renal biopsy on the tenth day contained 18 normal glomeruli and showed a patchy infiltrate of the 'immune' type containing plasma cells and eosinophils, with evidence of a recovering tubular necrosis. There was no conclusive evidence of a vasculitis or disseminated intravascular coagulation. Immunofluoresence studies were negative. The combined features were similar to those seen in acute transplant rejection and connective tissue disorders, and were compatible with complete recovery. A skin biopsy taken on day 11 demonstrated a resolving vasculitis, involving small arterioles with some residual fibrin (Figure 1).

Further investigation excluded heavy metal and drug intoxication, and repeated examinations of swabs, urine, faeces, blood and cerebrospinal fluid did not reveal any bacterial or viral

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pathogens. Toxic shock syndrome was excluded because an exotoxin-producing staphylococcus was not isolated. Serial electrocardiographs showed changes consistent with hypocalcaemia on day 1, and hyperkalaemia and day 3, otherwise they were normal. There was evidence of liver and pancreatic dysfunction, with raised levels of liver enzymes and amylase in the blood.

Recovery was complicated by multiple right-sided focal fits of short duration, and his EEG showed generalized slow-wave activity, which was prominent and focal in the left temporal region. He gradually regained consciousness, and at the end of the second week there was a left hemiparesis. During the next four weeks he steadily improved; on discharge from hospital he could sit unsupported and feed himself, and his left hemiplegia had become less dense. The patient is now 3 years and 6 months of age; his residual disabilities are a mild paraplegia and slight speech delay, but otherwise his psychomotor development is normal.

Discussion

The patient reported here fulfilled five of the principal diagnostic criteria for Kawasaki disease (KD): fever for more than five days, erythema of the lips and buccal mucosa, induration of the hands and feet, erythematous rash, and conjunctival injection (Melish 1981). He showed histological evidence of a vasculitis in the skin. We had no specific proof of cardiac involvement, his liver and pancreatic function were abnormal, and we speculate that his neurological symptoms could have been caused by a vasculitis.

Several renal complications have been documented in KD – pyuria, proteinuria and haematuria (Yanagihara & Todd 1980) and haemolytic-uraemic syndrome (Ferriero & Wolfsdorf 1981). Renal biopsy in the present case failed to demonstrate a vasculitis, but did show immune changes reminiscent of those seen in acute transplant rejection and connective tissue disorders, in both of which conditions a vasculitic process is involved. An earlier renal biopsy may have shown a perivasculitis, which is characteristic of the acute phase of KD.

The aetiology of KD remains obscure, although a 'microbial' source would still seem to be the most likely cause of the initial clinical picture. The finding of an 'immune' process on renal biopsy during the second week of illness in the present case would seem to stengthen the hypothesis suggested by Melish (1981) that the subacute phase of the illness is an immune-mediated reaction in response to an initial 'microbial' insult.

Management of KD is unclear, but close monitoring of cardiac function is imperative, using electrocardiograms, chest radiology and, more recently, echocardiography. Follow up must be on a long-term basis in view of the increased mortality. Patients given corticosteroids in the acute phase have been found to have an increased incidence of coronary aneurysms (Kato *et al.* 1979). Currently the recommended therapy is aspirin in standard anti-inflammatory doses during the acute phase; and if cardiac involvement is certain, then long-term low-dose aspirin is indicated. Until the aetiology of this mysterious condition is known, effective therapy is unlikely to be instituted.

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Epidermotropic eccrine carcinoma[1]

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Malignant sweat gland tumours are rare. A particular type of eccrine duct carcinoma which has distinctive clinical and histological features is now reported.

Case report

The patient, a male Caucasian aged 59 years, first noted a warty area on the left buttock in 1974. This gradually increased in size and in 1979 he presented with multiple, confluent, hyperkeratotic verrucous nodules and marked surrounding erythema over the left buttock and upper thigh.

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