

## SHORT REPORT

## Classical Kawasaki disease in a neonate

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A case is reported of classical Kawasaki disease in an infant younger than 2 weeks of age. Echocardiography detected a coronary artery aneurysm on the fifth day of the illness. Administration of intravenous  $\gamma$  globulin resulted in rapid improvement. Kawasaki disease is rare in neonates, but it may follow a rapid and severe course.

Kawasaki disease (KD) is unusual in newborn infants. Of 105 755 patients with KD registered in Japan over 25 years, only six were neonates, with the youngest aged 20 days.<sup>1</sup>

**CASE REPORT**

A full term, breastfed male infant, the only child of healthy, non-consanguineous parents, presented at 12 days of age after 48 hours of fever, irritability, and poor feeding. A generalised rash had developed 12 hours earlier. Pregnancy and delivery were normal, birth weight was 3080 g, and he had been well post partum. His temperature was 38.5°C, pulse 166/minute, respiration 52/minute, capillary refill less than two seconds, and weight 3400 g. He was drowsy and irritable with a high pitched cry. The fontanelle was soft and muscle tone was normal. A generalised non-tender, non-blistering, erythematous rash was present. The remaining examination was unremarkable.

Laboratory investigations included: haemoglobin 132 g/l, white blood cells  $5.4 \times 10^9/l$  (neutrophils 39%, lymphocytes 39%, monocytes 22%), platelets  $251 \times 10^9/l$ , sodium 135 mmol/l, potassium 4.7 mmol/l, calcium 2.6 mmol/l, creatinine 52  $\mu\text{mol/l}$ , glucose 4.8 mmol/l, total bilirubin 101  $\mu\text{mol/l}$ , albumin 30 g/l, C reactive protein 0 mg/l. Cerebrospinal fluid contained  $3 \times 10^6/l$  white blood cells,  $1 \times 10^6/l$  red blood cells, protein 0.43 g/l, and glucose 2.1 mmol/l. Catheter urine was analysed as follows: pH 5.0,  $< 10 \times 10^6/l$  white blood cells; no red blood cells, protein, or bacteria were detected.

The infant received amoxicillin and gentamicin, but eight hours later developed apnoea and multifocal clonic seizures. Phenobarbitone, phenytoin, flucloxacillin, and acyclovir were added, and assisted ventilation was started. Chest radiographs and computed tomography brain scans were normal, as were arterial pH, serum ammonia, lactate, and urine metabolic screening tests.

The rash spread next day to involve the palms and soles. The hands and feet became swollen. By the third hospital day, bilateral non-exudative conjunctivitis, erythematous dry lips, and a "red raspberry" tongue appeared. Echocardiography showed a dilated (3.5 mm) left coronary artery with a small aneurysm in its proximal segment. An electrocardiogram was normal. Intravenous  $\gamma$  globulin (2 g/kg) was given. Within 24 hours, the patient became afebrile and less irritable, the rash faded, and he was extubated. Low dose aspirin (5 mg/kg) was started. By the 5th day he was more alert. His hands and feet were no longer swollen, the rash and oral changes had resolved, and his conjunctivitis had improved. After seven days, the platelet count reached  $677 \times 10^9/l$ , while neutrophil numbers fell to  $0.4 \times 10^9/l$  before recovering over two weeks.

Periungual desquamation began on day 10 and continued for two weeks.

Bacterial cultures from blood, urine, and cerebrospinal fluid were sterile. The antimicrobial agents were discontinued within 24 hours of the start of  $\gamma$  globulin. Viral cultures from the nasopharynx, urine, and faeces and polymerase chain reaction tests for herpes simplex, enteroviruses, Epstein-Barr virus, and parvovirus from blood and cerebrospinal fluid were negative. Epstein-Barr virus and parvovirus IgG, but not IgM, antibodies were detected. His mother's syphilis serology was non-reactive.

The patient was discharged on aspirin after nine days in hospital. The aneurysm resolved by six weeks. Two and a half years later he is well, without cardiac or neurodevelopmental symptoms. The left coronary artery dilatation persists (3.5 mm) and he continues to take aspirin.

**DISCUSSION**

Two other neonates have presented with KD before age 2 weeks.<sup>2,3</sup> The first collapsed and died within four hours of birth. At necropsy, acute coronary artery vasculitis, vessel obstruction by fresh thrombus, and a recent myocardial infarction was found.<sup>2</sup> In the second case, the patient became febrile at five days and rapidly developed heart failure. He had myocarditis, dilated left coronary arteries, and raised C reactive protein. After being given  $\gamma$  globulin, he recovered, with resolution of the echocardiographic coronary artery abnormalities.<sup>3</sup>

Neither infant had classical features of KD (fever for five days or more with conjunctivitis, mucositis, extremity changes, rash with or without cervical lymphadenopathy). The diagnosis was based on cardiac abnormalities at autopsy or by echocardiography. Similar circumstances are seen in older infants with fever and less than four diagnostic features who develop coronary artery aneurysms. These patients have incomplete KD, which is a challenging diagnosis in young infants who are among the most susceptible to cardiac complications.<sup>4</sup>

In contrast, our patient fulfilled the clinical diagnostic criteria and also had echocardiographic coronary artery abnormalities. No alternative disorder was identified and he improved rapidly with high dose  $\gamma$  globulin. His lethargy, irritability, abnormal cry, seizures, and severe apnoea suggested involvement of the central nervous system. He did not have aseptic meningitis, which is often reported in patients with KD. Other neurological complications such as encephalopathy, seizures, cerebrovascular events, and isolated cranial nerve deficits are uncommon. Transient cerebral hypoperfusion has been observed during the acute phase of the illness, and it is postulated that vasculitis may cause these rare neurological manifestations.<sup>5</sup>

Although raised serum acute phase reactants are almost universal, more than 50% of patients with KD do not have increased C reactive protein at diagnosis.<sup>6</sup> Transient neutropenia is uncommon. In this patient it preceded the administration of  $\gamma$  globulin and aspirin, both of which have been implicated in a causal role. Presumably his young age, early diagnosis, and treatment may explain these findings.

The rarity of KD in neonates is consistent with epidemiological evidence that infectious agents cause this disease. Paediatricians and neonatologists should be aware that KD occurs in neonates, that the presentation may be atypical, and that it can follow a rapid and severe course.

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