

TEACHING FILES (GRAND ROUNDS)

AN INFANT WITH FEVER, RAISED INFLAMMATORY MAKERS AND NO APPARENT FOCUS FOR FEVER

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Clinical Problem

A 4 month old male infant presented with fever for 4 days. He had no localizing signs on examination. He was irritable and suspected to have meningitis. Investigations showed white blood cells 28,100/cumm (neutrophils 21,500/cumm), platelets 554000/cumm and CRP 312 mg/dl. Chest X-ray was normal and urine showed 25 pus cells/hpf but urine culture did not grow any organism. Cerebrospinal fluid (CSF) showed 8 cells/cumm, 1 red blood cell. CSF cultures were negative. Abdominal ultrasound (USG) and CT brain was normal. He was treated with intravenous antibiotics cefotaxime, amoxicillin and acyclovir. After 2 days of admission, he had a swollen ankle and slightly red lips. X-ray of the ankle was normal. Radioisotope bone scan showed no osteomyelitis. CRP was 212 mg/dl after 7 days of antibiotic treatment. Echocardiogram was done on day 8 which was normal. He remained irritable throughout this period

What is the diagnosis?

Expert Opinion :

This is a case of atypical Kawasaki with giant aneurysm of left coronary artery. Criteria for diagnosis of Kawasaki include fever persisting at least 5 days and presence of at least 4 principal features: 1) Changes in extremities: a) acute: erythema of palms, soles; edema of hands, feet and b) subacute: Periungual peeling of fingers, toes; 2) Polymorphous exanthema; 3) Bilateral bulbar conjunctival injection without exudate; 4) Changes in lips and oral cavity: Erythema, lips cracking, strawberry tongue, diffuse injection of oral and pharyngeal mucosae; 5) Cervical lymphadenopathy (>1.5 cm diameter), usually unilateral.¹ In the presence of ≥ 4 principal criteria, the diagnosis of Kawasaki disease can be made on day 4 of illness.¹ Supplemental laboratory criteria include albumin ≥ 3.0 g/dL, anemia for age, elevation of alanine aminotransferase, platelets after 7 days $\geq 450,000$ /cumm, white blood cell count $\geq 15,000$ /cumm, and urine ≥ 10 white blood cells/high-power field.¹ Children with Kawasaki disease often are more irritable than are children with other febrile illnesses.¹ Our patient did not fulfil all the criteria of

typical Kawasaki disease but on day 8 after admission IVIG (intravenous immunoglobulin) and aspirin were commenced. This was associated with immediate cessation of the fever and the parents reported that irritability improved almost immediately. CRP dropped from 212 to 48 mg/dl with 48 hours of starting IVIG. Repeat echocardiogram at 6 weeks follow-up showed giant coronary aneurysm. Because young infants may present with fever and few, if any, principal clinical features, echocardiography (and atypical Kawasaki) should be considered in any infant aged < 6 months with fever of ≥ 7 days' duration, laboratory evidence of systemic inflammation, and no other explanation for the febrile illness.¹ The incidence of atypical Kawasaki appears to be greater in infants younger than six months of age^{2,3} and are at substantial risk of developing coronary abnormalities.⁴

Coronary artery aneurysms are classified as small (<5 mm internal diameter), medium (5 to 8 mm internal diameter), or giant (>8 mm internal diameter).^{5,6} Coronary artery aneurysms or ectasia develop in 15%-25% of untreated children and may lead to ischemic heart disease or sudden death.^{5,7} Aneurysms rarely form before day 10 of illness¹, as noted in our patient with a normal initial echocardiogram. Even when treated with IVIG regimens within the first 10 days of illness, 5% of children with Kawasaki disease develop at the least transient coronary artery dilation and 1% develop giant aneurysms^{5,8,9} as was also seen in this case. A low platelet count at illness presentation is a risk factor for coronary aneurysms¹ however this was not a feature in this case.

Thus we conclude that incomplete Kawasaki disease should be considered in all children with unexplained fever for ≥ 5 days associated with 2 or 3 of the principal clinical features of Kawasaki disease.¹

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Compliance with Ethical Standards

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