

Case Number 2

Kawasaki Disease

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Case summary:

Demographic details:

Patient: YB, Male.

Resident in Attard.

Age: 2 years

Referred by: GP

Patient YB is a two-year-old boy who presented to A&E with a 7 day history of fever up to 101.8°F associated with symptoms of an upper respiratory tract infection with cough, vomiting and diarrhoea and bilateral non-purulent conjunctivitis and fissuring of the lips. Based on clinical findings the patient was diagnosed with Kawasaki Disease and was treated accordingly.

Presenting complaint:

Pyrexia up to 101.8°F: 7 days

Non-Purulent conjunctivitis: 7 days.

Cracked lips: 7 days.

History of presenting complaint:

The patient developed coryzal symptoms and cough associated with a temperature which persisted for 7 days. He was also vomiting and passing loose stools. Concomitantly he developed a non-purulent conjunctivitis with fissuring and crusting of his lips. He was seen by the GP who prescribed an antibiotic, a mucolytic agent and antipyretics. However, the fever persisted with no clinical improvement and the patient was admitted to hospital.

Past medical and surgical history:

Past medical history:

Bronchiolitis: 7 weeks prior to current admission

Drug history:

Drug	Dosage	Frequency	Type	Reason
Augmentin 457mg/5ml Syrup	2.5ml	BD	Penicillin Antibiotic	Treatment of secondary bacterial infection
Paracetamol Suppositories	125-250mg	4-6 hourly	Antipyretic and Analgesic	Brings about a reduction in fever

Ambroxol hydrochloride Syrup	2.5ml	TDS	Mucolytic-cough preparation	Secretolytic agent and Mucolytic agent which helps in the breakdown and thinning of mucus.
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Table 1¹ Treatment on admission

The patient has no known drug allergy.

Family history:

Both parents are unrelated and healthy. There is no significant family history.

Social history:

The boy has a fraternal twin sister, lives with his family and attends playschool.

Systemic inquiry:

- General Health: the patient was feverish and irritable.
- Cardiovascular System: nil of note
- Respiratory System: nil of note
- Gastrointestinal System: few episodes of vomiting, an episode of loose stools.
- Genitourinary System: the patient was passing normal volumes of urine.
- Central Nervous System: nil of note
- Musculoskeletal System: nil of note
- Endocrine System: nil of note
- Others: non-purulent conjunctivitis, mild cervical lymphadenopathy.

Discussion of results of general and specific examinations:

The patient was lying down in bed, miserable and irritable. He was not pale, jaundiced or cyanosed and was well-hydrated. There were no evident rashes. He had a normal respiratory rate. Mild cervical lymphadenopathy was noted. His temperature was 101.8°F (febrile). There was no generalised oedema but the hands were noted to be swollen. The tonsils were enlarged with no pus but a generalised erythema of the oral mucosa was noted. Healing perioral excoriations and satellite lesions on the nose were evident.

Cardiovascular examination revealed normal heart sounds S1+S2+0 and a pulse of 148 beats per minute of good volume.

Respiratory examination showed equal air entry in both right and left lungs with normal vesicular breath sounds. No added sounds were noted.

The abdomen moved with respiration and there were no swellings or scars. It was soft, with no guarding or rebound tenderness. No masses were felt and there was no organomegaly. Normal bowel sounds were auscultated and stools were normal.

Differential diagnosis:

- Kawasaki Disease
- Stephen Johnson Disease
- Measles
- Scarlet Fever
- Drug reactions
- Other febrile viral exanthems
- Toxic Epidermal Necrolysis
- Rocky Mountain Spotted fever
- Staphylococcal Scalded skin Disease
- Juvenile Idiopathic Arthritis
- Leptospirosis
- Mercury Poisoning²

Diagnostic procedures:

*Laboratory exams:*³⁻⁴

Test: Complete blood count, including platelet count.

Justification for test: May help with differential diagnosis specifically assessing white blood cell count and differential, which is likely to be elevated and the presence of anaemia. A marked thrombocytosis in the second week of illness is a typical finding.

Result: Normocytic anaemia, with an elevated white cell count and thrombocytosis.

Conclusion: These results are often seen in Kawasaki Disease.

Test: Urea & Electrolytes

Justification for test: These tests help assess state of hydration and electrolyte imbalances.

Result: Normal

Conclusion: This excludes any Renal Disease.

Test: C- Reactive Protein

Justification for test: Acute phase reactant.

Result: Elevated

Conclusion: This is usually elevated to a degree not typically found in common viral infections and can be an indication of Kawasaki disease or other invasive bacterial conditions.

Test: Erythrocyte Sedimentation Rate

Justification for test: Nonspecific marker of inflammation.

Result: Elevated

Conclusion: This is usually elevated to a degree not typically found in common viral infections and can help differentiate between connective tissue diseases and other bacterial infections.

Test: Blood cultures

Justification for test: This is done to detect the presence of actively multiplying bacteria or fungi in the bloodstream, to identify the microorganism(s) present and to guide antimicrobial treatment.

Result: Negative

Conclusion: Bacterial sepsis is excluded.

Instrumental exams:

Test: Echocardiogram (ECHO) ^{2-3,5}

Justification for test: Kawasaki Disease affects the coronary arteries in 1/3 of affected children within the first 6 weeks of illness leading to aneurysm formation. These are best visualised by echocardiography. In the acute phase of illness, coronary artery abnormalities include lack of tapering, perivascular brightness and ectasia. Echocardiography may also reveal decreased ventricular function, mild valvular regurgitation and pericardial effusion.

Result: The ECHO was found to be normal. Mitral, coronary or cardiac involvement was excluded.

Conclusion: The disease did not involve the coronary arteries in this patient.

Therapy:

Drugs:¹

Drug	Dosage	Frequency	Type	Reason
Immunoglobulin	2g/kg IVI	2 g/kg over 10 hours; preferably within the first 10 days of the illness	Human pooled Immunoglobulin	Suppresses inflammatory response which may lead to coronary artery damage
Paracetamol Suppositories	250mg PR	Every 4-6 hours	Antipyretic and analgesic	Provides symptomatic relief
Aspirin Tablets	7.5 -12.5 mg/kg/dose = 150 mg PO	QDS for 2 weeks or until afebrile; dose reduced to 2-5mg/kg od for 6-8 wk	NSAID Anti -Inflammatory	Antiplatelet and anti-inflammatory effect preventing coronary artery damage
Ranitidine Syrup 25mg/ml	2-4 mg/kg/dose = 4mls PO	BD	Histamine H2-receptor antagonist	This is used as prophylaxis against dyspepsia and risk of gastrointestinal bleeding caused by high dose aspirin

Diagnosis:

Kawasaki Disease is a systemic vasculitis which predominantly affects children under the age of 5 years. The etiology of Kawasaki disease remains unknown, although an infectious agent is strongly suspected based on clinical and epidemiologic features. The diagnosis of Kawasaki Disease cannot be made by a single laboratory test or combination of tests. Physicians make the diagnosis after carefully examining a child, observing signs and symptoms and eliminating the possibility of other, similar diseases.

To diagnose Kawasaki Disease the child must have a persistent fever of 5 days and 4 out of 5 of the criteria below:

- Polymorphous rash
- Bilateral (non-purulent) conjunctival injection
- Mucous membrane changes, e.g. reddened or dry cracked lips, strawberry tongue, diffuse redness of oral or pharyngeal mucosa
- Peripheral changes, e.g. erythema of the palms or soles, oedema of the hands or feet, and in convalescence desquamation

- Cervical lymphadenopathy (> 15 mm diameter, usually unilateral, single, non-purulent and painful). In this case the child presented with a 7 day history of fever, a bilateral conjunctival injection, cracked lips, swollen hands and cervical lymphadenopathy and therefore fulfilled the criteria of diagnosis of Kawasaki Disease^{2-3,5-7}.

Final treatment and follow up:

The patient was treated as shown in Table 2. He was also prescribed intravenous fluids - 5% dextrose in 0.9% saline at maintenance rate. These intravenous solutions are indicated for use in paediatric patients as sources of electrolytes, calories and water for hydration⁸. Oral fluids were encouraged and the patient was placed on a soft diet.

Renal profile tests were carried out and repeated as necessary. Parameters including fluid input and output were monitored at regular intervals.

Since no coronary artery abnormalities were noted at presentation, an echocardiogram is to be repeated at two weeks and at six to eight weeks after diagnosis². The risk of developing significant heart disease is less once the fever subsides³.

The patient was kept in hospital until the fever subsided. He was then discharged and reviewed at regular intervals.

The patient should be re-evaluated one week post-discharge and then have a repeat echocardiogram³.

Fact Box 2:

Name of Condition: Kawasaki Disease^{5,6,7}

Also known as: Mucocutaneous lymph node Disease; Infantile polyarteritis

Risk factors:

- Age: Children under 5 years old are most at risk of Kawasaki Disease.
- Sex: Boys are slightly more likely than girls are to develop Kawasaki Disease.
- Ethnicity: Children of Asian descent, such as Japanese or Korean, have higher rates of Kawasaki Disease.

Symptoms and Signs:

Children with Kawasaki Disease usually present with:

- A persistent fever of 5 days and 4 out of 5 of the criteria below:
- Polymorphous rash
- Bilateral (non-purulent) conjunctival injection
- Mucous membrane changes, e.g. reddened or dry cracked lips, strawberry tongue, diffuse redness of oral or pharyngeal mucosa
- Peripheral changes, e.g. erythema of the palms or soles, oedema of the hands or feet, and in convalescence desquamation
- Cervical lymphadenopathy (> 15 mm diameter, usually unilateral, single, non-purulent and painful)

Prevention: Kawasaki Disease cannot be prevented, but usually has tell-tale symptoms and signs that appear in phases.

Complications: Heart complications include:

- Inflammation of the heart muscle (myocarditis)
- Heart valve problems (mitral regurgitation)
- Abnormal heart rhythm (dysrhythmia)
- Inflammation of blood vessels (vasculitis), usually the coronary arteries, that supply blood to the heart

For a small percentage of children with Kawasaki Disease, this can result in death in spite of treatment.

Treatment:

Immunoglobulins: Infusion of an immune protein (gamma globulin) through a vein (intravenously) can lower the risk of coronary artery problems⁹.

Aspirin: High doses of aspirin may help treat inflammation. Aspirin can also decrease pain and joint inflammation, as well as reduce the fever. Kawasaki treatment is a rare exception to the rule against aspirin use in children.

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