Kawasaki Disease: Typical and Atypical Presentation

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Disclosure

Neither I, Michael Breunig, nor any family member(s) have any relevant financial relationships to discuss, referred to or illustrated with or without recognition within this presentation.
Objectives

- Recognition of typical Kawasaki Disease presentation
- Understanding of additional labs and imaging to aid in diagnosis of incomplete Kawasaki Disease
- Treatment of Kawasaki Disease
Definition

- Formerly called Mucocutaneous Lymph Node Syndrome
- Fever (>38°C) for at least 5 days plus minimum of 4 of 5 signs of mucocutaneous inflammation without another cause
- Unknown etiology
- Medium vessel vasculitis
  - All 3 vessel layers
Kawasaki Disease

- Very common pediatric illness
  - 95% < age 10
- Late winter/early spring
- Self-limited
- Asians > African Americans/Hispanics > Caucasians

Continuum of disease
- Acute (Day 1-11)
- Subacute (Day 12-21)
- Convalescent
Why do we treat?

- Low mortality
- Morbidity
  - #1 cause of acquired heart disease in patients <5yo
  - 15-25% of untreated patients develop cardiac manifestations
    - Aneurysm, MI, myocarditis, pericarditis, valvular disease, dysrhythmias
Case presentation #1

- HPI - 4yo Hispanic male presents to ED with fever (102-4F) for 7 days, bilateral conjunctivitis, decreased PO intake, fissured lips, diffuse rash started 2 days ago on trunk, vague leg pain and myalgias. Taking acetaminophen and ibuprofen but fever returns in 2-3 hours. Seen in Urgent Care 4-5 days ago for allergy symptoms and prescribed cetirizine and azithromycin.
Differential Diagnosis

- Extensive rule out other causes
- Infectious exanthems of childhood
- Exudative conjunctivitis
- Exudative pharyngitis
- Influenza
- General lymphadenopathy
- Stevens-Johnson syndrome
- Drug reaction
Case #1 (cont)

- **PMH**
  - Intraventricular hemorrhage at birth (MRI wnl)
  - Hospitalized with RSV at age 1
- **Birth Hx**
  - Twin born at 33 WGA SVD
  - 3 weeks in NICU
- **Immunizations**
  - UTD
- **Medications**
  - Acetaminophen, ibuprofen, cetirizine, azithromycin
Case #1 (cont)

- Allergies- NKDA
- Family Hx- HTN and asthma brother, possible thrombophilia mother
- Social Hx- non-contributory
- ROS
  - Positive – Fever, myalgias, bilateral conjunctivitis without exudate, rash, cracked lips, leg pain, decreased PO intake
  - Negative- CP, abd pain, sore throat, meningeal signs
Case #1 (cont)

- **PE**
  - VSS except T 101.3F
  - Gen- very ill appearing
  - HEENT- B injected conjunctiva, fissured lips, erythematous tongue, dry mucus membranes
  - Neck- Left Cervical lymphadenopathy
  - Pulm- CTAB, no increased WOB
  - CV – RRR, systolic murmur I/VI
  - Abd- Soft, NT, ND, BS +
  - Skin- Erythematous, moribiliform on trunk, buttocks, and extremities
  - Neuro – age appropriate, CN II-XII intact
Case #1 (cont)

- **Lab**
  - CBC, CMP, ESR, CRP, UA, rapid strep with cx, BCx, RVP
  - WBC 10.8, Na 131, ESR 78, CRP 2.8, AST 1623, ALT 955, UA Protein tr, ketones 1+, urobil 12.0, Rapid strep negative, RVP - + rhinovirus and H1N1

- **Imaging**
  - CXR - normal
Kawasaki Disease

- Fever >38 C for minimum 5 days with 4 or more mucocutaneous inflammation without other cause.
- Infants may present differently
  - Fever >7 days, consider workup
  - Higher risk of cariac aneurysm/dysfunction
Mucocutaneous Inflammation

- Bilateral conjunctival injection
  - Reported 90% cases
  - Non-exudative
- Mucus membrane changes
  - Fissured lips, strawberry tongue
- Polymorphous rash
  - Starts perianal but seldom reported
- Peripheral extremity changes
  - Arthritis, palmer erythema, desquamation
- Minimum 1 anterior cervical lymph node >1.5cm
  - Least reported
Case presentation #2

- 2yo Caucasian male presents to ED with fever 103F for 7-8 days, myalgias, diffuse rash, fissured lips, decreased PO intake, injected conjunctiva bilateral, and cough. Acetaminophen and ibuprofen improve fever but has not resolved. Sick contacts at daycare.
Case #2 (cont)

- PMH- None
- Birth Hx- FT 38 WGA svd, no complications
- Surg Hx- None
- Immun- up to date
- Meds- Acetaminophen, ibuprofen
- Allergies- NKDA
- Family Hx- non-contributory
- Social Hx- non-contributory
Case #2 (cont)

- **ROS**- Fever, myalgias, rash, fissured lips, cough, injected conjunctiva, decreased PO intake
- **PE**
  - **Gen**- Ill appearing
  - **HEENT**- Fissured lips, EOMI, PEERLA, TM intact, MM- dry, bilateral injected conjunctiva
  - **Pulm**- CTAB, no wheeze, rales or rhonchi
  - **CV**- RRR, no murmur
  - **Abd**- soft, NT, ND, +BS
  - **Extrem**- No c/c/e, moves all extremities equally
  - **Skin**- Diffuse maculopapular, erythematous rash
  - **Neuro**- CN II-XII intact, age appropriate
Case #2 (cont)

- **Lab**
  - CBC, CMP, ESR, CRP, UA, BCx, RVP
  - WBC- 18K, Platelets- 600K, CRP- 3.2, ESR- 61, ALT- 178, Alb- 2.8
  - UA- WNL, BCx- neg, RVP- neg

- **Imaging**
  - CXR- No acute findings
Atypical (incomplete) Kawasaki

- Does not meet criteria of typical (less than 4)
- Incidence - 10-60% of diagnosed cases
- Diagnosis
  - Need laboratory and/or 2D echo
  - Elevated CRP/ESR + 3 or more specific abnormal lab values or abnormal 2D echo
    - Abnormal echo - aneurysm, coronary arteritis, valvular regurgitation, pericardial effusion, abnormal LV function
Fever ≥5 days and 2 or 3 clinical criteria*

Assess patient characteristicsΔ

Consistent with KD

Inconsistent with KD

Assess laboratory tests

CRP <3.0 mg/DL and ESR <40 mm/hr

Follow daily

Fever continues for 2 days

No peeling

No follow up

Fever resolves

Typical peeling**

Echo¥

CRP ≥3.0 mg/DL and/or ESR ≥40 mm/hr

KD unlikely

Persistent fever

<3 supplemental laboratory criteria◊

Echo

Echo -

Fever persists

Repeat echo consult KD expert

KD unlikely

≥3 supplemental laboratory criteria◊

Treat and echo§

Echo +¥

Fever abates

Treat*
Supporting laboratory data

- CRP $\geq$ 3mg/dL
- ESR $\geq$ 40mm/hr
- WBC $\geq$ 15,000
- HgB $<$ 2 Standard deviations for age
  - Normocytic, normochromic anemia
- UA- WBC $\geq$ 10hpf
  - Sterile pyuria (-LE)
- Albumin $\leq$ 3
- ALT $>$ 50 U/L
Indications for 2D echocardiogram

- Anyone meeting criteria for typical or atypical Kawasaki disease
- Fever for 7 or more days in infant (without other cause) and elevated CRP/ESR
- Fever >=5 days with elevated CRP/ESR and less than 3 abnormal lab findings
- Desquamation (periungual) after fever resolves
Treatment

- IVIG- 2g/kg x 1 over 8-12 hr
  - Earlier treatment=less inflammation and fewer cardiac problems
  - May retreat if:
    - Continued fever of unknown origin after 10 days
    - Elevated inflammatory markers (ESR, CRP)
    - Coronary artery aneurysm
- Complications
  - Potentially toxic
  - Possible blood borne pathogens
  - Large amount of IVFs
Treatment

- IVIG (cont)
  - Postpone vaccinations with live vaccines for 11mo
  - Vaccinations for Influenza and Varicella especially those patients on long term ASA use
Treatment

ASA- 80-100mg/kg/day split in 4 doses
- Anti-inflammatory and anti-platelet
- Do not exceed 4g/day
- Confirm negative Influenza or Varicella (Reye Syndrome)
- Switch to low dose (3-5mg/kg/day) when afebrile 48 hr or acute inflammation improves (CRP normal)
Treatment

- Glucocorticoids
  - 2mg/kg/day split into 3 doses
  - Consider if refractory to IVIG and high risk of coronary artery aneurysm
  - Improves fever/inflammation
    - No improvement in prevention of coronary artery aneurysms
Outcomes

- **Mortality**
  - Very low

- **Morbidity**
  - Dependent on coronary artery involvement
    - No change on echo - Little risk but unknown LT
    - <8mm dilatation - Usually regress but need lifelong follow up
    - >=8mm - Greatest risk for MI, arrhythmia; need follow up
    - Physical activity restrictions - only with coronary artery involvement (cardiologist recommendations)
Case #1

- Typical Kawasaki disease with Influenza and rhinovirus
  - Treated with IVIG 2g/kg x 1 on hospital day #2
  - ASA 81mg daily
  - Oseltamivir 45mg bid x 5 days
  - Consult to peds cardiology and infectious disease
- 2D echo – Normal. Coronary arteries <2mm
- Discharge to home hospital day 5
- Follow up with cardiology and repeat echo in 1 and 6 weeks
Case #2

- Atypical (incomplete) Kawasaki disease
  - IVIG 2g/kg x 1 on hospital day #3
  - ASA 30mg/kg q8 hr for 2 days then 40mg per day
  - Cardiology consult
  - 2D echo – Normal without coronary artery thickening
  - Discharged to home hospital day #6
  - Follow up with cardiology and repeat echo in 1 and 6 weeks
References