Common Complaint with an Uncommon Diagnosis: From Trauma to Kawasaki Disease

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Introduction

Kawasaki Disease (KD) is a rare vasculitis that affects the coronary arteries, veins, and capillaries of children typically under the age of 5.

20 per 100,000 children younger than five years of age develop KD in the United States; East Asians and Pacific Islanders have the highest incidence (30/100,000) while Caucasians have the lowest incidence (12/100,000) [2].

The cause of KD is unknown. However several theories exist. The stimulus for inflammation in KD is unknown, but particularly dilates and destructs the walls of coronary arteries. Patients with KD can have a high count of neutrophils, CD8 T cells, IgA, eosinophils, and macrophages. The symptoms of KD present very similar to other pediatric infections, which may suggest KD is caused by an unknown transmissible agent. Some data suggest KD has a seasonal occurrence, specifically winter and summer. Genetic factors appear to contribute as well, as Asians and Asian-American are mostly affected [4].

Diagnostic Criteria for KD:

Presence of at least 5 days of fever and 4 out 5 following symptoms:

1. Bilateral bulbar conjunctival injection
2. Oral mucous membrane changes
3. Peripheral extremity changes, including erythema of palms/soles, edema of hands/feet
4. Polymorphous rash
5. Cervical Lymphadenopathy (at least 1 lymph node >1.5cm)

To diagnose KD, patient must have a **fever of 5 days or greater** and have **4 out of 5 symptoms**[1]. An elevated WBC, platelet count, transaminase, acute phase reactants, pyuria, and anemia, may also support KD diagnosis. An echocardiogram is taken to assess the patient's risk of developing vasculitis and to determine further management. The most significant complications from KD are Coronary artery aneurysm (CA), depressed myocardial contractility, and shock[3].

Treatment includes IVIG within the first 10 days of KD to reduce risk of CA aneurysms, and Aspirin.

Case Report

We present a case of a **3-year old** British Caucasian female with a history of repaired TOF, who initially presented to the ER with a 2 day history of fever. Patient had recent history of neck trauma on two separate occasions where the neck was forcibly flexed forward. The traumas occurred within 7 days of fever. She was seen in the ER for a total of 3 visits.

Visit 1: Patient had fever of 38.7 C, HR 163, RR 26, BP 98/66, and O2 99%. No LOC, N/V, cough, diarrhea, rash. Rapid strep test, UA, Urine culture were negative. She was discharged same day with Tylenol and Motrin.

Visit 2: Next day, patient presented to ER with neck pain and decreased appetite. She developed a lower lip blister and neck was tender to palpation. Laboratory studies showed elevated CRP and WBC. Patient was requested to come back next day for reevaluation.

Visit 3: The following day, patient presented with eye pain and headache. WBC decreased from 17k to 13k and CRP was constant. Neck U/S showed 2.2 cm Left cervical lymph node which was erythematous. C-spine was negative. CXR showed dilated heart and multiple coils. Patient was given IV Rocephin and clindamycin and was admitted to the hospital.

IV antibiotics were continued and viral studies showed negative results. Wound culture of left neck showed no organisms. Patient developed bilateral conjunctivitis and generalized rash consistent with KD during stay. ECG was done and exhibited normal coronary arteries. IVIG and high-dose ASA were administered. After IVIG course, patient became afebrile and returned to baseline function with a decreased CRP of 9.7. She was discharged with high-dose ASA for 2 more days and then switched to low-dose ASA.

Discussion

The unusual course of events beginning with trauma to the patient's head and neck on multiple occasions, followed by a fever and suspected viral illness, culminating with KD symptoms prompts us to question the cause of KD in this case.

Trauma, specifically in the cervical region, could be a possible stimulus for inflammation of the vascular tissue. Many lymph nodes, nerves, arteries, veins run through the cervical region. When trauma is induced, there may be a potential immunologic response, which may cause fevers, increased neutrophil/macrophage, eosinophil, and plasma cell release.

Although etiology of KD is relatively unknown, this case may provide insight that certain factors such as congenital malformations (i.e. TOF) and trauma put children at a greater risk for KD and must be considered in the evaluation of young children presenting to the ER with routine complaints.

Graphic 1

Left-sided Cervical Lymphadenopathy. 2.2 cm

References

1) Ayusawa M, Sonobe T, Uemura S, et al. Revision of diagnostic guidelines for Kawasaki disease (the 5th revised edition). Pediatr Int 2005; 47:232.
2) Holman RC, Belay ED, Christensen KY, et al. Hospitalizations for Kawasaki syndrome among children in the United States, 1997-2007. Pediatr Infect Dis J 2010; 29:483.
3) Orenstein JM, Shulman ST, Fox LM, et al. Three linked vasculopathic processes characterize Kawasaki disease: a light and transmission electron microscopic study. PLoS One 2012; 7:e38998.
4) Takahashi K, Oharaseki T, Yokouchi Y. Pathogenesis of Kawasaki disease. Clin Exp Immunol 2011; 164 Suppl 1:20.