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Case Report

Diagnostic Challenges Encountered When Predicting Patterns of Adult Kawasaki Disease: A Case Report

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ABSTRACT

The study of medicine has always awed researchers and allowed for quality scientific discoveries. According to several studies, Kawasaki Disease (KD) typically presents in both adults and children with signs and symptoms of conjunctivitis, fever (for ≥ five days), pharyngitis as well as skin erythema that advances to desquamatory rash that does not spare the palms and soles in many cases. However, marginally minimal amount of studies has investigated symptoms particularly in adults. The purpose is to report diagnostic challenges that happened our way as we tried diagnosing a rare adult case of Kawasaki Disease in an 18-year-old male with no significant past medical history or physical features to suggest KD. The aim of reporting this case was to highlight the difficulty in diagnosing KD but stressing the importance of managing inflammatory symptoms first especially in young adults with ≥ 5 days of high fevers.

Key words: Adult kawasaki disease; Inflammation; Small-medium-sized arteries; High fevers

INTRODUCTION

Kawasaki Disease (KD) is an acute necrotizing vasculitis of medium and small sized vessels that has predilection for mucocutaneous lymph nodes and coronary arteries. KD occurs mainly in Korean and Japanese infants and children aged 6 months to 5 years. Approximately 15%-20% of all untreated patients develop coronary aneurysms and 2%-3% dies of coronary vasculitis [1]. Complications are usually preventable when the right management is initiated early in the course of disease [2]. The challenge faced during diagnosis of KD is that the diagnostic criteria are non-specific. Therefore, clinical features may be present in other infectious and autoimmune pathologies. Also, as the etiology remains unknown, there is no specific diagnostic test to confirm disease. This presents a diagnostic and therapeutic challenge for the treating physician [3]. With about a little over 100 cases described worldwide, the patterns of disease manifestations and complications continue to pose as a conundrum [4]. This 18-year-old male presented with diagnostic challenges which included; fever, rash, throat pain and myalgia. The diagnostic criteria included fever for ≥ 5 days, in addition to any 4 of the 5 following symptoms:

- Non-exudative conjunctival injection,
- Erythema of hands and feet and periungual desquamation,
- Oral-mucosal changes like; strawberry tongue,
- Polymorphic rash, desquamation and
- Cervical lymphadenopathy.

It did not occur in order as some of the symptoms did not show at all like the strawberry tongue [3].

There is no specific diagnostic test therefore presenting as a formidable challenge to the treating physician and the diagnosis is often one of exclusion. No. of cases reported per 100,000 children under the age of 5 years is>130 in Japan, 90 in Korea,

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50: Taiwan, 20-30 Hong Kong and China, 4-15 in the US, 8: UK, 4: Australia and 3: South America (in descending order). Greater recognition of this condition in specific countries may reflect an increased awareness and lower threshold of suspicion. It may also indicate a strong genetic susceptibility as the number of cases identified in Japanese American children in Hawaii (135) is similar to that found in Japan.

Therefore, though it is still considered a rare disease in the US it is the most common cause of acquired heart disease in children in the US [5].

CASE PRESENTATION

An 18-year-old young man presented to the hospital in early January of 2019 with thrombocytosis and high fever (up to 40°C) unresponsive to anti-inflammatory drugs for the first two days but with a good response to aspirin after day four. The high fever continued through day six of admission along with other symptoms like persistent headache, malaise, arthralgia and signs of cervical adenopathy. Upon further investigation, lab results also showed inflammation of the liver (hepatitis) with high liver function tests. It was not until day 10 before the desquamatory rash, reddening of eye and throat pain.

This young man did not travel recently either within the country or abroad and he denies intravenous drug use or having new sexual partners or body tattoos as well as no animal exposure. After day ten, he remembered he took some Amoxicillin pills when he was feverish and had itchy skin. On general examination, he was morbidly obese, appeared anxious but was fully oriented to time, place and person.

BP was 135/65 mm hg all other vitals were stable. EKG (Electro Cardio Gram) showed normal sinus rhythm, no ST elevations or T wave inversions. Deep palpation of the abdomen was limited due to body habitus. Labs were drawn, and he was sent home after day 11 on oral and topical corticosteroids for possible allergic reaction to Amoxicillin with advice to follow up at either the clinic or the emergency department if symptoms persisted or worsened.

RESULTS AND DISCUSSION

The labs drawn showed WBC (White blood cells) of 14,200 (neutrophil dominant), ALT: 126 U/L, AST: 51 U/L, ALP: 207 U/L Albumin: 3.4 gm/dl, Total bilirubin: 0.6 mg/dl, ESR: 61 mm/hr, CRP (C-Reactive Protein): 6.9 mg/dl. Urinalysis showed traces WBC with no nitrites. Therefore, he was asked to follow up at the clinic as soon as possible (Figure 1).

On revaluation at the clinic three days later, all symptoms still persisted. He additionally also had redness of mouth and fissuring of lips. Examination revealed erythematous tongue and oral mucosa. There was mild periungual and facial desquamation. He was sent to the ER with a strong suspicion of Kawasaki disease.

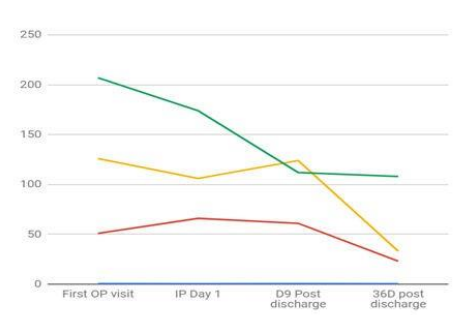

Figure 1: Shows the liver function tests numbers for our 18-year-old KD patient. Note: Total bilirubin (mg/dl) AST ALT (U/L) ALP (U/L)

At the hospital, repeat labs revealed lower WBC of 13,900 cell bil/L (neutrophil predominant). Liver markers had also reduced: ALT (Alanine Transaminase): 106 U/L, AST (Aspartate Transaminase): 66 U/L, ALP (Alkaline phosphatase): 174 U/L, Albumin: 3 gm/dl, Lactic acid: 1.5 mmol/L, ESR (Erythrocyte Sedimentation Rate) was increased at 76 mm/hr, Urine routine: No WBC's (Figure 2). EKG showed normal sinus rhythm and no change from previous, 2D transthoracic echocardiogram showed an ejection fraction of 65% with mild tricuspid regurgitation, right ventricular systolic pressure was 51 mmHg, right upper quadrant ultrasound was unremarkable. He was given acetaminophen 650 mg PO Q6H, diphenhydramine 25 mg PO Q6H, Intra Venous Immuno Globulin (IVIG), Aspirin 975 mg PO QD. Rapid strep test was negative; however ASO and DNase-B antibody was mildly positive at 480 IU/ml and 282 U/mL respectively.

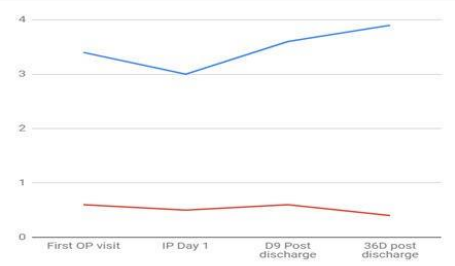

Figure 2: shows albumin levels increasing from the initial as KD progresses. Note: Albumin (gm/dl) Total bilirubin

Lab test later revealed a neutrophilic leukocytosis, WBC count was 14,200 cells/cumm. There was also a mild hepatitis and markers of inflammation (ESR and CRP) were elevated. Leukocytosis and hepatitis were a possibility in IMN, TSS (Toxic Shock Syndrome) and Kawasaki, however neutrophilic predominance favored Kawasaki and TSS (Figure 3). Patient was re-evaluated at the hospital three days later. By now, he complained of multiple spikes of fever on a daily basis. There was diffuse erythema of tongue and oral mucosa. There was also mild desquamation of rash over face and periungual desquamation which is fairly specific for Kawasaki. Our adult 18-year-old patient had persistent headache that subsided only after the administration of IVIG and ASA (Acetyl Salicylic Acid). Given the patient's age group, history of throat pain and generalized rash worsened with intake of amoxicillin, Infectious Mono Nucleosis (IMN) was high on our differential. Other

differentials considered were; generalized allergic or contact dermatitis reaction and complications of Group A Streptococcus (GAS) pharyngitis such as Toxic Shock Syndrome, Acute rheumatic fever and Scarlet fever. Appropriate labs done ruled these other conditions out.

While at the hospital, labs were repeated this showed persistence of neutrophil leukocytosis. Ejection Fraction (EF) was 65%, there was mild Left Ventricular Hypertrophy (LVH), tricuspid regurgitation and however, there were no coronary artery aneurysms. Patient was immediately started on an IVIG infusion, high dose aspirin, diphenhydramine and acetaminophen. Patient started to feel much better after the first infusion. Periungual desquamation soon progressed to a generalized desquamation of both hands and feet. CMV (Cyto Megalo Virus), EBV (Epstein-Barr Virus) and Parvovirus B19 IgG antibody was positive, however IgM was negative. In addition, EBV early Ag and nuclear Ag were also positive. This ruled out Infectious mononucleosis and Parvovirus as causes. ASO (Anti Streptolysin O) and Anti-DNase B were elevated which proved prior strep infection and was likely the cause for this presentation. Blood cultures showed a preliminary result and later reviewed Gram-positive rods; *Corynebacterium* manumission, *Corynebacterium amycolatum* and Diphtheriods which was considered to be a contaminant from the IV line.

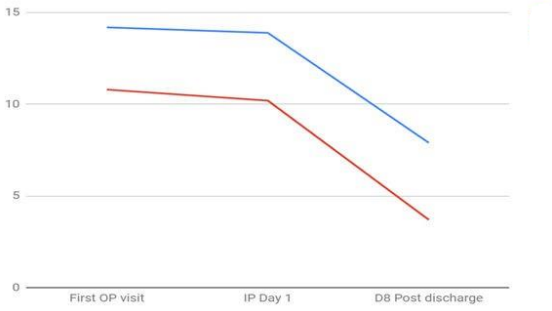

Figure 3: shows levels of granulocytes; neutrophils in our 18-year-old patient. Note: WBC Neutrophils

Despite 50 years of research in this condition, there is only a small body of literature describing new onset Kawasaki disease in adults. Coronary artery aneurysm occurs in 15%-25% of untreated children up to 5% of treated ones [6]. Short term sequelae of coronary aneurysms; death due to coronary vasculitis have been identified and long-term consequences of endothelial dysfunction, coronary artery disease and myocardial infarction have been described in survivors [7,8]. There is also a drastic reduction in complications if identified and treated within 10 days of onset of symptoms [9, 10].

After discharge, desquamation progressed to involve both nails in hands and feet as well. Onychomadesis has been described in Kawasaki only twice before [11]. Ten days post discharge; WBC had normalized. However, LFT (Liver Function Tests) normalized only within 1 month of discharge. EBV IgG was positive as>750 and EBV early antigen at 52.1 U/mL, however EBV IgM was <10 U/ml. Parvovirus B19 IgG and CMV IgG were positive however CMV and Parvovirus B19 IgM were negative. Patient continued to have spikes of fevers on the first day despite the IVIG infusion. By the 5th day after IVIG patients conjunctival injection, myalgia and polymorphous rash had improved. Periungual desquamation had progressed to a severe desquamation of both hands and feet. A retrospective diagnosis of Kawasaki was then made, and the patient was discharged on Keflex, Cetirizine and continued on low dose aspirin for 6 weeks.

CONCLUSION

This report of an adult case of Kawasaki disease may be beneficial to doctors of several medical specialties; like primary care physicians, Internists and intensivists as well as cardiologists, rheumatologists just to name a few. This case shows prolonged fever with thrombocytosis after day five with thrombocytosis improving after aspirin administration on day five. In the absence of other diagnoses, an incident of Kawasaki disease cannot be ruled out and it is worth noting that such a diagnosis should be highly suspected even as an adult patient without Asian descent.

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CONSENT TO PARTICIPATE AND PUBLICATION

Written consent was given by the patient

AUTHOR'S CONTRIBUTIONS

- References list and proof reading; Dr. Silas Y. Debrah
- Patient seen as inpatients/outpatients in one hospital in the city of Detroit (Dr. Christina Reji, Dr Asha Shajahan, Dr. Eric Ayers and Dr Donald Tynes)
- Abstract, discussion, conclusion, figures; (Dr. Smart Asare, Dr. Aubin Sandio,).
- Introduction, Case Presentation; (Dr. Asha Shajahan, Dr. Smart Asare, Dr. Christina reji and Dr. Aubin Sandio, Dr Eric Ayers).

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