CASE REPORTS

SYSTEMIC ONSET JUVENILE IDIOPATHIC ARTHRITIS (SOJIA) MIMICKING INCOMPLETE KAWASAKI DISEASE IN A 5-YEAR-OLD

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ABSTRACT

A 5-Year-Old male presented with 9 days of fever, rash, and poorly described muscle/joint pain despite multiple outpatient visits and treatment with antibiotics for presumed streptococcal pharyngitis, and acute otitis media. In the emergency department he appeared ill with cracked red lips, delayed capillary refill, joint and muscle pain, tachycardia, and a blanchable maculopapular rash coalescing into plaques. Laboratory tests demonstrated leukocytosis, elevated C-reactive protein, thrombocytosis, and hypoalbuminemia. Given the labs, prolonged fever, and mucocutaneous symptoms, he was treated for incomplete Kawasaki Disease with intravenous immunoglobulin (IVIG) and aspirin. Upon admission, echocardiogram and electrocardiogram were both within normal limits. The patient had continued fevers despite IVIG and was given a second dose, which he was also refractory to. Systemic steroids were started and relieved the fever, rash, and joint pains. He clinically improved and was discharged on a corticosteroid taper and aspirin.

He was readmitted two weeks later with fever, weight loss, malaise, migratory arthralgias/myalgias, and splenomegaly. Given the persistent fevers and weight loss, hemophagocytic lymphohistiocytosis and malignancy were excluded. He was started on NSAIDs and markedly improved. Given this improvement the diagnosis of juvenile idiopathic arthritis was made.

This case highlights the diagnostic overlap between Kawasaki disease and systemic-onset juvenile idiopathic arthritis, particularly early in the disease course when fever pattern, rash characteristics, and arthritis may be non-specific. Persistent fever and musculoskeletal symptoms despite IVIG treatment should prompt consideration of other diagnoses causing systemic inflammation, including systemic-onset juvenile idiopathic arthritis.

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Introduction

Systemic-onset Juvenile Idiopathic Arthritis (SoJIA) is a subtype of JIA characterized by systemic inflammation, prolonged fever, and arthritis for at least 6 weeks.[1,2,3] The fever should be present for at least 2 weeks (often ≥38.5°C) and is often quotidian in nature.[1,2,3] A salmon-colored, non-pruritic rash may be present and most evident during fever spikes. Other symptoms include lymphadenopathy, hepatosplenomegaly, and serositis.[2,3] Initial manifestations of SoJIA closely resemble Kawasaki disease (KD) including the presence of fever, rash, thrombocytosis, and increased inflammatory markers. There is no single definitive test to distinguish or confirm the two diseases.[1]

We describe a patient with delayed diagnosis of SoJIA complicated by fulfillment of criteria for incomplete KD. The patient presented during the night of his ninth consecutive day of fevers and was treated for incomplete KD to prevent disease sequelae. Symptoms initially improved but then worsened as the underlying disease revealed itself.

Case Report:

A 5-year-old male was admitted from the emergency department (ED) with rash, arthralgias and/or myalgias (patient could not differentiate bone vs. muscle pain), and 9-days of fever despite treatment with antibiotics and acetaminophen. 9 days prior to admission the patient experienced hip and knee pain, especially when ambulating and developed a fever of 38.3°C. He was evaluated at an urgent care, found to be streptococcus pyogenes positive, and prescribed a course of amoxicillin. The next day the patient developed an erythematous, maculopapular truncal rash on his abdomen, back, and extremities. He re-presented to urgent care two days later with persistent fever, malaise, and pain with ambulating. He was diagnosed with an ear infection, and the antibiotic was switched to amoxicillin-clavulanic acid. Despite three days of antibiotics the patient's fever and rash persisted, which prompted additional visits to urgent care and eventually the ED.

In the ED, the patient was ill-appearing with red, cracked lips, dry mucus membranes, a murmur, tachycardia with delayed capillary refill of 3-4 seconds, and a rash described as pink and blanchable with papules coalescing into plaques consistent with erythema multiforme. Labs demonstrated leukocytosis of 23.8 × 103 µL (normal 4.3-11 × 103 µL), elevated erythrocyte sedimentation rate (ESR) 67 mm/hr (normal 0-15 mm/hr), elevated C-reaction protein (CRP) 120 mg/L (normal ≤8 mg/L), thrombocytosis of 460,000 × 103 µL (normal 206-369 × 103 µL), and hypoalbuminemia of 3.0 g/dL (normal 3.5-4.5 g/dL). Serum electrolytes, hemoglobin, hematocrit, aspartate

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aminotransferase (AST), alanine transaminase (ALT), bilirubin, total protein, alkaline phosphatase lipase were all within a normal range, There was high suspicion for incomplete KD given the prolonged fever, elevated inflammatory markers, leukocytosis, thrombocytosis, and hypoalbuminemia, necessitating admission and prompt initiation of intravenous immunoglobulin (IVIG) and aspirin to prevent disease sequelae.

Upon admission, an electrocardiogram (EKG) and echocardiogram (ECHO) were obtained and were within normal limits. The rash started improving, but the patient continued to spike high fevers (>40°C) and had bilateral leg and ankle pain more than 36 hours after IVIG. A second round of IVIG was administered but fevers persisted. Therefore, high dose corticosteroids were started. Additional labs were obtained prior to starting corticosteroids to rule out hemophagocytic lymphohistocytosis (HLH) and included the following: hyperferritinemia of 2,421 ng/mL (normal 20-300 ng/mL), hypertriglyceridemia of 91 mg/dL (normal <75 mg/dL), elevated interleukin-2 receptor of 2,009.1 pg/mL (normal 175.3-858.2 pg/mL), fibrinogen elevated of 521 mg/dL (normal 120-450 mg/dL), and negative Anti-dsDNA antibody.

The patient reported improvement pain upon initiation of corticosteroids. He remained afebrile for >24 hours with improvement of his inflammatory markers. He was discharged on aspirin, famotidine, and a 5-day corticosteroid taper.

Two weeks later, the patient was readmitted with fever, weight loss, malaise, and migratory pain described as myalgias in the arms, legs, and lower back. He was found to have splenomegaly on ultrasound. Scheduled non-steroidal anti-inflammatory drugs (NSAIDs) were started for suspicion of JIA (10 mg/kg every 8 hours). Fever and pain resolved on NSAIDs. Given significant improvement with NSAIDs, elevated ferritin, and mild splenomegaly, a diagnosis of JIA made. Multisystemic inflammatory syndrome in children (MIS-C) was unlikely given the patient's lack of preceding illness and multiple negative respiratory viral panels. Despite not having six weeks of arthritis the patient had MSI-C, Kawasaki disease, malignancy, tick-borne illness, and HLH excluded making JIA the most likely diagnosis. The patient was discharged home with down trending labs, famotidine, and NSAIDs.

Discussion/Conclusions:

Incomplete KD and systemic-onset JIA (SoJIA) have been postulated to be part of the same disease spectrum.[4] They are both childhood systemic inflammatory diseases with similar cytokines activated, overlapping clinical features, and no specific diagnostic tests, making it difficult to distinguish between the two.[4,5] This can lead to delayed diagnosis and treatment of JIA, resulting in numerous rounds of IVIG and increased financial burden.[2,4,5]

Our patient presented with vague systemic symptoms which originally left our differential consisting of incomplete KD, serum sickness-like reaction, HLH, and JIA after ruling out infection, trauma, and malignancy.[2] Serum sickness-like reaction seemed less likely due to the rash being non-pruritic and presenting shortly after initiation of antibiotic. Our patient did not meet criteria for an HLH diagnosis and biopsy obtained showed no hemophagocytosis.[6]

The patient presented to our emergency department on day 9 of fever. The treatment for KD is IVIG which should be initiated within first 10 days of fever onset to reduce incidence of coronary artery aneurysms.[7] The patient met criteria for incomplete KD (fever ≥ 5 days, a arthritis, oral erythema and truncal rash, CRP ≥3 mg/L, platelet count ≥460,000 × 103 μL after the seventh day of fever, albumin ≤ 3 g/dL, and WBC ≥ 15,000/mm^3). Our patient received two rounds of IVIG and continued to fever, leading to the possibility of IVIG resistance and the initiation of corticosteroids or biologic agents.

Upon readmission, persistent fever ≥ 2 weeks, myalgias, hyperferritenemia, splenomegaly, and improvement with first steroids and then NSAIDs were all suggestive of SoJIA.[3,8] However, limitations to making this diagnosis sooner include:

1. Although the fever was persistent and occasionally ≥ 38.5°C, the quotidian pattern was not present, and the rash did not intensify with fever.
2. Due to the age of this patient, he was unable to localize the pain and had difficulty distinguishing between myalgias, arthralgia, and bone pain.
3. Patient had ≤ 6 weeks of arthritis.[3]
4. Patient met criteria for IKD with 9 days of fever, necessitating treatment with IVIG to prevent coronary artery aneurysms.[6]

Because of these limitations, we highlight the difficulty distinguishing incomplete KD and SoJIA, especially in the early course of SoJIA. Various reports demonstrate SoJIA being initially diagnosed as KD leading to multiple doses of IVIG and hospital readmissions before SoJIA was diagnosed.[9] Prolonged fever that presents with a rash and is non-responsive to IVIG, should raise suspicion for other inflammatory disorders like SoJIA, especially when accompanied with arthritis.

Compliance with Ethical Standards
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