Kawasaki Disease in a Postpartum Patient

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Kawasaki disease, also known as mucocutaneous lymph node syndrome, is a multisystem disease. It usually affects children below the age of five, but it occasionally affects adults. There are less than 50 English-reported adult cases in the literature, and only five reported cases of Kawasaki disease and pregnancy, as of 2003. The cases associated with pregnancy involved patients who had a history of Kawasaki disease during childhood and addressed how the complications of the illness (i.e,. coronary artery aneurysms) were managed during pregnancy and delivery.[1-5] There are no reported cases of Kawasaki disease in postpartum patients. This article presents a case of Kawasaki disease in a 21-year-old, four-week postpartum patient who initially responded to intravenous (IV) antibiotic therapy. This paper will review the diagnosis and treatment of Kawasaki disease as well as the multiple outside variables that impact the management of adult postpartum patients with Kawasaki disease.

Key words: adult ■ Kawasaki disease ■ mucocutaneous lymph node syndrome ■ postpartum ■ pregnancy

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Initial Presentation and History

A 21-year-old gravida 3 para 1111 African-American female returned to a community hospital emergency department one week postdischarge after having been hospitalized for five days with the working diagnosis of toxic shock syndrome, but she never fulfilled the criteria of multisystem involvement. At the time of initial presentation, she was four weeks postpartum, having experienced a normal vaginal delivery without complications. She presented with complaints of a one-week history of nausea, persistent fever, and a generalized painful, pruritic rash. Subsequently, the rash spread to her neck, arms, hands, abdomen, back, buttocks, and thighs.

During this admission, the patient presented with a reoccurrence of her previous complaints of persistent fever and the same painful, pruritic rash which had not resolved since the first admission. She denied sore throat, cough, shortness of breath, chest pain, vomiting, diarrhea, constipation, dysuria, vaginal discharge, a history of sexually transmitted diseases, or recent sexual activity. She also denied recent travel, insect bites, sick contacts, or allergies.

Physical Examination

The examination on admission revealed a thin female in no acute distress. She had a temperature of 101.9°F, blood pressure of 97/55 mmHg, pulse of 142 bpm, respiratory rate of 22, and pulse oximetry of 99% on room air. Her skin had a maculopapular rash with confluent pustular lesions located on her eyelids, neck chest, axillae, arms, back, thighs, and buttocks. The rash was erythematous, warm, and more prominent in the intertrigenous areas. She had tender lymphadenopathy involving the cervical, axillary, and inguinal areas. There was no conjunctival injection or oropharyngeal erythema. The cardiovascular examination revealed tachycardia with normal heart sounds. Lungs were clear to auscultation bilaterally. The abdomen was flat with striae, normal bowel sounds, nontender to palpation and without hepatosplenomegaly. The extremities were

without swelling and there was no rash or erythema on the palmar or plantar surfaces. A pelvic exam, which was unremarkable, was performed during the first admission.

Laboratory Evaluation

During the first admission, the potassium hydroxide and saline wet-mount slides, and the gonorrhea and chlamydia cultures were all negative. A review of her prenatal records revealed a negative prenatal work-up, including a negative HIV test.

The laboratory evaluation from this admission revealed a leukocyte count of 26,800/mm^2 with 24% bands. Other laboratory measurements are as follows; hemoglobin 8.4 gm/dl, platelets 460,000 cu mm, sedimentation rate 40 mm/hour. The basic metabolic panel was within normal limits. The urinalysis revealed a specific gravity of 1.020, 20–25 white blood cells, 0–4 red blood cells, few epithelial cells, 4+ bacteria, and 4+ mucus. Wound culture from a pustule on her arm had light growth of gram-negative bacillus. Blood and urine cultures were negative. The viral profiles which included hepatitis, herpes simplex virus, coxsackie A and B, echovirus and the Epstein-Barr virus were all negative. The anitstreptolysin titer, antinuclear antibody, and double stranded DNA antibody, and HIV antibody were all negative.

Imaging Studies

Pelvic ultrasound revealed a small amount of fluid posterior to the cervix and anterior to the fundus and hyperechoic areas within the endometrium. (In the postpartum state, these are common findings on pelvic ultrasound.) The chest radiograph revealed a small pericardial effusion and interstitial edema.

The echocardiogram revealed trace mitral valve regurgitation with thickened mitral leaflets, an ejection fraction of 55% and confirmed the small pericardial effusion seen on the chest radiograph.

Medical Intervention and Patient Outcome

The patient was started on IV antibiotics (vancomycin and piperacillin/tazobactam). Soon thereafter, a dopamine drip was started due to the drop in her blood pressure from 97/55 mmHg to 76/25 mmHg. The patient also received two units of packed red blood cells for the continued decrease in hemoglobin to 8.0. Within 36 hours after her admission, she began to develop conjunctivitis, strawberry tongue, red lips, erythema and swelling of her hands and feet, and desquamation of her palmar and plantar surfaces. Subsequently, as a result of the events above, she was diagnosed with Kawasaki disease and started on high-dose aspirin—3 grams/day—and a single dose of intravenous immunoglobin (IVIG)—108 grams. Within 36 hours, the patient's condition improved, and she was afebrile throughout the remainder of her hospital stay. The patient was discharged home the eighth day on low-dose aspirin, oral antibiotics, and instructed to follow up for her scheduled appointment with her primary care physician in one week.

DISCUSSION

Kawasaki disease is an acute febrile illness, also known as mucocutaneous lymph node syndrome. It predominantly affects children between the ages of six months and two years, and usually less than five years of age. Asian children are at higher risk due to genetic predisposition.[6] The male–female ratio is 1.5:1.[7] Males are more frequently affected and are also at a greater risk of developing coronary artery aneurysms.[7]

Kawasaki disease has been associated with infectious agents (i.e., Epstein-Barr virus, herpes virus, and streptococcus), dust mites, carpet shampoo, and septic water.[6] There are no specific diagnostic tests for Kawasaki disease. (Table 1).

There are three phases in the clinical course of untreated Kawasaki disease. First is the acute febrile phase, which lasts one-to-two weeks and consists of the diagnostic criteria along with anorexia, irritability, hepatic dysfunction, and aseptic meningitis.[7] Second is the subacute phase, which lasts two-to-four weeks and consists of thrombocytosis, arthritis and desquamation of the hands and feet.[7] Fever, rash, and lymphadenopathy resolve, and conjunctivitis and irritability may persist.[7] At this time, patients are at greatest risk for coronary artery thrombosis. Finally, the convalescent phase begins when the clinical signs have resolved and the platelet count and sedimentation rate returns to normal. This phase usually occurs within six-to-10 weeks.[7]

The case presented is typical of Kawasaki disease in the way the disease evolved with the clinical signs

Table 1. Diagnostic Criteria for Kawasaki Disease[6,7]

Fever for at least five days plus four of the following:

1. Nonexudative bulbar conjunctival injection
2. Erythematous mouth and pharynx, and/or strawberry tongue, and/or red, cracked lips
3. Generalized erythematous rash
4. Swelling and erythema of the hands and feet followed by desquamation
5. Cervical lymphadenopathy with at least one node greater than 1.5 cm

and symptoms but atypical with respect to the patient's postpartum status and her response to IV antibiotic therapy. This appears to be the first perinatal case of Kawasaki disease.

There have been five reported cases of Kawasaki disease and pregnancy in the English literature. All of these cases described patients who had Kawasaki disease during childhood and addressed how the sequelae of coronary artery aneurysms were managed during pregnancy and delivery. The first case described the successful pregnancy and delivery of a patient on low-dose aspirin who had two coronary artery aneurysms.[1] Another report described a case of a successful pregnancy and delivery nine years after coronary artery bypass grafting for Kawasaki coronary artery disease.[2] The third case described a successful pregnancy and delivery with a patient who had a coronary artery aneurysm treated with heparin anticoagulant therapy during pregnancy.[3] The fourth case described the successful use of epidural anesthesia for the caesarean section of a patient with a childhood history of Kawasaki disease.[4] Finally, the fifth case describes a patient who had a peripartum myocardial infarction from presumed Kawasaki disease.[5]

The patient presented in this case was 21 years old and four weeks postpartum, with an initial presentation of a one-week history of fever and rash, which had initially responded to intravenous antibiotic therapy. During her first hospitalization, several conditions were considered. The differential diagnosis included toxic shock syndrome, septic shock, scarlet fever, Stevens Johnson syndrome, hypersensitivity reaction, folliculitis, endometritis, and Kawasaki disease. The patient was treated with the working diagnosis of toxic shock syndrome, but the lack of multisystem involvement and the laboratory results did not fulfill the criteria to make the diagnosis of toxic shock syndrome. She responded to one week of supportive care and IV antibiotic therapy, followed by one week of remission until the fever returned, the rash progressed, and the common features of Kawasaki disease began to evolve. Subsequently, she fulfilled more than the minimum criteria needed to make the diagnosis of Kawasaki disease.

Generally, the clinical signs of Kawasaki disease tend to evolve with time and do not respond to antibiotics. Due to the patient's response to IV antibiotics, the diagnosis of Kawasaki disease appeared to be less likely. Physicians who consider Kawasaki disease in the differential diagnosis of patients should maintain close follow-up to assure that the patients' clinical signs and symptoms resolve.

MANAGEMENT

The current treatment for Kawasaki disease includes IVIG and aspirin. Adults who are treated with IVIG have demonstrated similar responses and recovery times as children.[6] High-dose IVIG has proven to reduce the morbidity and mortality that is associated with Kawasaki disease. As mentioned by Saulsbury, when IVIG is given within the first 10 days of the illness, the risk of coronary artery aneurysms is reduced from 20% in untreated patients to 4%.[7] IVIG also aids in the resolution of established coronary artery aneurysms. The most effective dose of IVIG is a single infusion of 2 g/kg given over 10 hours.[7]

Aspirin is used as an adjunct to IVIG. The initial dose is 80–100 mg/kg/day divided every six hours or until defervescence. Salicylate levels should be monitored during high-dose therapy. After 14 days, decrease the dose to 3–5 mg/kg/day until the platelet count returns to normal. Patients with coronary artery aneurysms should remain on the low-dose aspirin indefinitely or until one year after the aneurysm resolves.[7]

Unfortunately, 10% of the cases are unresponsive to IVIG. In these cases, plasmapheresis may be used. In refractory cases, corticosteroids have been used. Corticosteroids may increase the risk of coronary artery aneurysms; therefore, they are not recommended.[10] Patients with coronary artery aneurysms are to have regular follow-up by a cardiologist. For aneurysms that are greater than 6.5 mm in diameter, warfarin is indicated. Catheter intervention with stent implantation may be necessary in those patients with cardiac complications.[10]

The management of Kawasaki disease in an adult postpartum patient has multiple outside variables that usually do not impact the management of a child. Specifically, this patient had a newborn infant at home; therefore, there was the issue of demanding a premature hospital discharge, avoidance of medications in order to continue breastfeeding, and the lack of desire and/or understanding for the need to comply with recommended tests and/or studies, medications, and long-term follow-up.

CONCLUSION

According to the literature, Kawasaki disease is more common in adults than we have previously believed. It tends to present with the same physical findings and have the same sequelae as children. But it is very important to be sensitive to the fact that adults may have other complicating circumstances (i.e., pregnancy, postpartum, breastfeeding, noncompliance, etc.) that may have an impact on our final diagnosis and/or medical management. The postpartum patient, in this case, initially responded to IV antibiotics before more of the diagnostic clinical signs of Kawasaki disease began to appear. Is it possible that IV antibiotics can prolong the acute

phase of Kawasaki disease? This is a question that needs further investigating. To date, there is no documented evidence of antibiotics prolonging the acute phase of Kawasaki disease. In the meantime, it is crucial that physicians follow adult patients very closely until the diagnosis of Kawasaki disease has been ruled out.

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