A 5-year-old Boy with Fever and Rash

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A 5-year-old, previously healthy, white boy presented to the clinic because of a nonpruritic, generalized erythematous rash and fever. He was seen in the clinic on day 2 of fever and was started on cefalexin for a presumptive diagnosis of scarlet fever. However, he remained febrile despite the antibiotic, even to day 5 of illness. His rash had then spread over the entire body.

He denied any sore throat, headache, cough, arthralgias, dysuria, or hematuria. He had bilateral conjunctival redness but no photophobia. His parents reported that no one else in his family had been sick.

Physical examination showed a sick-looking boy with temperature of 98.9°F. A diffuse nonpruritic erythematous rash was present on his face, neck, trunk, and extremities, including the palms of the hands and soles of the feet. The affected skin felt like sandpaper. He also had an erythematous peeling rash in the groin and genital area. His bulbar conjunctivae were red. He had pharyngitis, strawberry tongue, and chapped lips with mucosal irritation. Further laboratory tests confirmed the diagnosis.

On day 5 of illness, laboratory tests were WBC = 18.2 X 10^3/μL with 67% neutrophils, 22% bands, 5% lymphocytes, 3% monocytes; Hgb 12.8 g/dL; platelet count 467 X 10^3/μL; serum albumin, 3.3 g/dL; ALT 374 IU/L; AST 302 IU/L, alkaline phosphatase, 382 IU/L; total bilirubin 2.9 mg/dL, C-reactive protein 5.8 mg/dL, ESR 63 mm/hr. Urinalysis: WBCs 30-50/HPF; RBC 3-5/HPF, leukocytes +, nitrite negative.

Urine and blood culture were negative. CMV panel, immunoglobulins, DFA for adenovirus, and hepatitis panels all were negative. Fungal and bacterial cultures of the groin were negative.

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Editor’s note: Each month, this department features a discussion of an unusual diagnosis in genetics, radiology, or dermatology. A description and images are presented, followed by the diagnosis and an explanation of how the diagnosis was determined. As always, your comments are welcome via e-mail at editor@pediatricsupersite.com.
DISCUSSION
Fever for 5 days, generalized morbilliform rash, conjunctival injection, mucositis with strawberry tongue (See Figure), and genital excoriated rash with high CRP and ESR confirmed the diagnosis of Kawasaki disease. Initial echocardiogram on day 6 of fever revealed no coronary involvement, and IVIG was started.

He was afebrile within 36 hours of IVIG infusion and was discharged on the fourth hospital day on high-dose aspirin (80 to 100mg/kg/day).

The patient was followed up in the clinic 3 days after the hospital discharge. At this time, his rash and fever were completely resolved. He had peeling of the finger tips and groin area. During the second week of the illness, his platelet count was 627x10^3/mcL.

Kawasaki disease (KD) is an acute systemic inflammatory illness that can result in coronary artery aneurysm, myocardial infarction, and sudden death in previously healthy children. It is the number-one cause of acquired heart disease in developed countries. If left untreated, there is a 20% to 25% risk of coronary artery aneurysms. The etiology of KD is unknown, but a viral agent is strongly suspected.1,2 Diagnosis of classic KD is based on the presence of fever of 5 or more days and four or more of the five principal clinical features: 1) changes in the hands and feet (erythema, edema, peeling); 2) polymorphous exanthema; 3) bilateral bulbar conjunctival injection without exudates; 4) changes in lips and oral cavity (erythema, strawberry tongue); and 5) unilateral cervical lymphadenopathy.

Changes in the extremities include erythema of palms and soles, sometimes painful induration of hands and feet, and later desquamation of fingers and toes. Desquamation begins in the periungual region within 2 to 3 weeks and may involve palms and soles.

An erythematous maculopapular rash usually appears within 5 days of fever. Bilateral painless conjunctival infection usually involves bulbar conjunctivae and is typically nonexudative. Changes in lips and oral cavity include cracking and bleeding lips, “strawberry tongue” with erythematous prominent papillae, and diffuse oropharyngeal mucositis. Unilateral cervical lymphadenopathy is the least common of the principal clinical features. At least one node should be more than 1.5 cm in diameter. Hepatomegaly and jaundice and sometimes acalculous distension of the gall bladder can develop.

Laboratory evaluation shows leukocytosis in the acute stage of the illness and includes predominantly immature and mature granulocytes. Thrombocytosis is a characteristic of subacute stage of illness with the platelet count ranging from 500,000 to 1 million or more. Thrombocytosis usually peaks in the third week and gradually returns to normal by 4 to 8 weeks after the onset of illness.

Mild to moderate elevation in serum transaminases occurs in about 40% of patients, and mild hyperbilirubinemia occurs in approximately 10%. Hypoalbuminemia is common and is associated with more severe and prolonged acute disease. Urinalysis reveals mild-to-moderate sterile pyuria in about one-third of patients. A moderate to marked elevation of CRP (> 3 mg/dL) and/or ESR more than 40 mm/hr is nearly universal in KD.

Some patients do not fulfill the clinical criteria for KD and are diagnosed using an algorithm developed by a committee of the American Heart Association for atypical or incomplete KD. The term “incomplete KD refers to greater than 5 days of fever with fewer than 4 of the classic symptoms. This is more common in infants than older children, and these patients are at higher risk for developing coronary abnormalities detected by echocardiography. Coronary aneurysms are rarely present before day 10 of the illness, but some evidence of cardiac involvement or arteritis can be present in the acute stage of illness before the formation of aneurysms.
TREATMENT

The current standard of therapy for KD is a combination of aspirin and IVIG. In children treated with a single-dose of IVIG and high-dose aspirin, the risk of coronary abnormalities can be reduced by 85% to 90%. In the acute phase of illness, aspirin is administered at 80 to 100 mg/kg/day in four doses along with IVIG. High-dose aspirin is usually continued through illness day 14.

When high-dose aspirin is discontinued, therapy is continued with low dose aspirin (3 to 5 mg/kg/day) until the patient shows no evidence of coronary changes by 6 to 8 weeks after the onset of illness.

For children who manifest coronary abnormalities, aspirin or clopidogrel (Plavix) may be continued indefinitely. Because Reye’s syndrome is a risk for children receiving aspirin while infected with varicella or influenza, parents should be educated to contact their child’s physician if they have been exposed to varicella or influenza. These children should receive influenza immunizations routinely and be up to date with varicella immunizations.

CONCLUSIONS

The differential diagnosis of maculopapular red rash with fever can be extensive and includes not only scarlet fever but also drug reactions and infection, such as meningococcus; Staphylococcus aureus; adenovirus; enteroviruses; roseola; measles; rubella; toxic shock syndrome; and parvovirus infections.

Although great progress has been made in the treatment of KD, much is left to learn about the etiology. Until an etiological agent is identified and a definitive diagnostic test devised, children who have KD still will be underdiagnosed or misdiagnosed. Some who have KD will be untreated, suffering potentially serious morbidity and mortality. Correct diagnosis and prompt treatment will reduce the serious complications, such as coronary aneurysms, to much less than 5%.

REFERENCES