

# Fever and Rash in a 3-Year-Old Girl: Rocky Mountain Spotted Fever

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## GOAL

To explain the presentation and treatment of Rocky Mountain spotted fever (RMSF)

## OBJECTIVES

Upon completion of this activity, dermatologists and general practitioners should be able to:

1. Understand the etiology and epidemiology of RMSF.
2. Recognize the clinical manifestations of RMSF.
3. Describe the diagnosis and management of RMSF.

CME Test on page 174.

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*Initial symptoms of Rocky Mountain spotted fever (RMSF), a tick-borne illness caused by Rickettsia rickettsii, are nonspecific and include headache, gastrointestinal disturbances, malaise, and myalgias, followed by fever and rash. The classic triad of fever, rash, and history of tick exposure is uncommon at presentation. Clinical manifestations of RMSF range from virtually asymptomatic to severe. Because of the potentially fatal outcome of RMSF, presumptive clinical diagnosis and empiric*

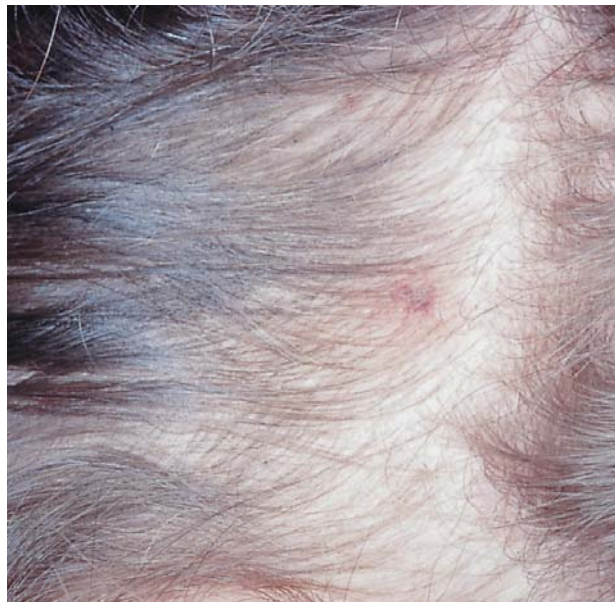
*antimicrobial therapy can be critical. We present the case of a 3-year-old girl from New York State who presented with fever and rash.*

**R**ocky Mountain spotted fever (RMSF) is the most common rickettsial illness in the United States. It is a tick-borne illness caused by the gram-negative intracellular bacterium *Rickettsia rickettsii* and is endemic in the Southeastern and South Central United States. Most cases occur between April and September, coinciding with the active season of the vector tick species. The disease is reported most frequently in children aged 5 to 9 years. An incubation period ranging from 2 to 14 days precedes a short prodrome of headache, malaise, myalgias, nausea, vomiting, and anorexia, with fever and rash following. Early diagnosis is critical because untreated RMSF may progress to fulminant systemic illness. We describe the case of a 3 year-old girl from New York State who presented with a fever and rash.

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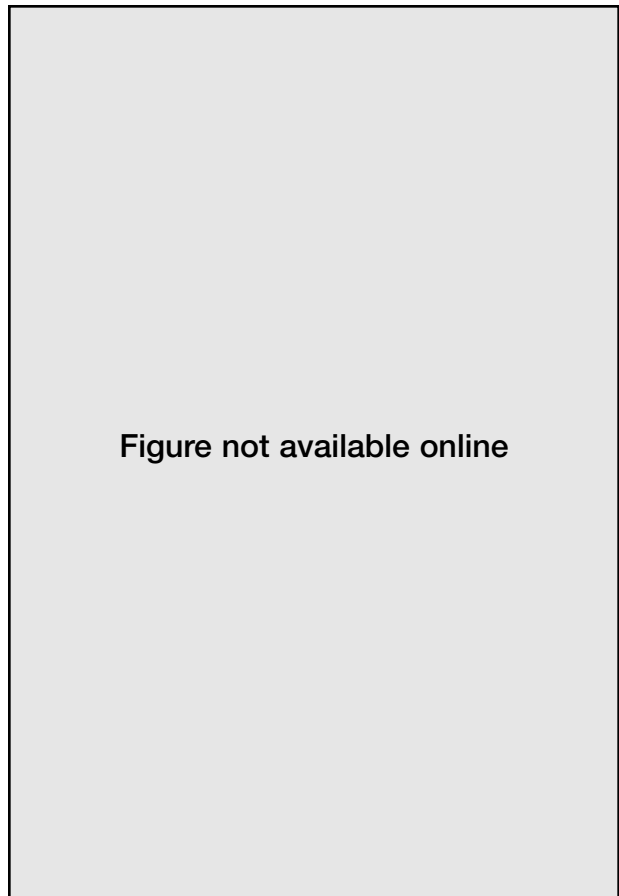
**Figure 1.** Inflamed site of tick attachment on the patient's scalp.

At presentation, a tick was attached to her postauricular scalp but was concealed by her hair and therefore not discovered at that time.

### Case Report

A previously healthy, 3-year-old white girl developed low-grade fever, vomiting, and general malaise 10 days after vacationing in Montauk, New York, during the month of May. The patient subsequently developed a morbilliform eruption on her face and proximal extremities. The patient's pediatrician diagnosed enteroviral infection and prescribed acetaminophen. However, the patient remained febrile, and the eruption became generalized over the following week. On reassessment by the pediatrician, an engorged tick was discovered and removed from the patient's right postauricular scalp (Figure 1). At this time, the patient's mother recollected that she had removed a second tick from her daughter's scalp 2 weeks earlier. The following day, the patient presented to the emergency department with increased lethargy, irritability, and anorexia.

On physical examination, the patient was resting comfortably and was not in acute distress. She was febrile (temperature, 103.7°F). The patient was normotensive at 110/45 mm Hg. Other vital signs were stable. A diffuse, mildly pruritic, blanching, morbilliform eruption was noted on her face and extremities, with lesser involvement of the trunk and abdomen (Figure 2). Scattered petechiae were present on the palms (Figure 3) and soles. The



**Figure 2.** A diffuse, blanching, morbilliform eruption, with rare petechiae, is present on the face and extremities. Note the relative sparing of the trunk and abdomen.

patient denied any myalgia, arthralgia, or headache. There was no evidence of photophobia, nuchal rigidity, or other meningeal signs. Lymphadenopathy and hepatosplenomegaly were absent.

Significant laboratory studies yielded the following: erythrocyte sedimentation rate of 72 mm/h (reference range, 0–20 mm/h) and albumin level of 2.9 g/dL (reference range, 3.5–5.0 g/dL). Results of complete blood count, hepatic function tests, and chemistries were otherwise unremarkable. The laboratory was unable to speciate the tick. A presumptive diagnosis of RMSF was made, and the patient was started on doxycycline (25 mg orally each day). Results of the serologic evaluation subsequently revealed elevated *R. rickettsii* antibody titers of 1:1024 and 1:128 (IgM and IgG, respectively). The findings from the Lyme and ehrlichiosis titers were normal. The patient defevered within 24 hours of therapy and was discharged on hospital day 2, with significant improvement of her eruption and general condition.



**Figure 3.** Scattered petechiae on the palms.

### Comment

There are 600 to 1200 cases of RMSF reported annually, with 90% of cases occurring during the active season of the vector tick species, early spring to late fall.<sup>1</sup> RMSF first was recognized more than a century ago; however, because of the long incubation period (2–14 days), lack of a reliable diagnostic test early in the disease, and nonspecific initial symptoms, diagnosis remains a challenge.<sup>1</sup>

*Etiology and Epidemiology*—The tick is both the vector and the main reservoir of the disease. The most common species in the United States are *Dermacentor variabilis* (dog tick) in the eastern two thirds of the country and parts of California, Oregon, and Washington and *Dermacentor andersoni* (wood tick) in the Rocky Mountain states. *Rickettsia* in ticks is maintained by transovarian transmission among successive tick generations.<sup>2</sup>

Autochthonous cases (native to the region) of RMSF have been reported in all states except Hawaii and Vermont.<sup>2</sup> RMSF first was reported in the Eastern United States in the 1930s and has since predominated there. In 1998, there were 365 cases of RMSF reported in the United States, reflecting a rate of 0.14 per 100,000, the lowest infection rate in nearly 2 decades. Thirteen of the reported cases occurred in New York State, including 2 in New York City.<sup>3</sup> The incidence of RMSF is higher in males; whites; children (aged 5–9 years); and individuals with exposure to dogs, wooded areas, or areas with tall grass.<sup>1</sup>

*Clinical Manifestations*—Clinical manifestations of RMSF range from virtually asymptomatic to severe. A 2- to 14-day incubation period precedes

a short prodrome of headache, malaise, myalgia, nausea, vomiting, and anorexia, with fever and rash following. The rash typically begins between days 3 and 5 of illness, first appearing peripherally (on the hands, wrists, feet, and ankles) and then spreading centrally to the trunk and face. Palm and sole involvement is common (50%–75%).<sup>4</sup> The rash develops earlier in younger and more severely ill patients and may be more difficult to recognize in dark-skinned patients. Lesions begin as discrete, blanching 1- to 4-mm macules and papules, but within days become petechial, hemorrhagic, and nonblanching. In severe cases, lesions may coalesce and form ecchymoses. Small gangrenous areas may appear over the fingers, toes, nose, ears, and genitals. Untreated RMSF may progress to systemic and pulmonic hemorrhage, edema, neurologic manifestations, anemia, and thrombocytopenia. Mortality rates in untreated patients range from 20% to 80%.<sup>4,5</sup>

Rickettsiae preferentially invade and multiply within the endothelial and smooth muscle cells of blood vessels. Infected cells proliferate, swell, and degenerate, resulting in partial or complete thrombosis of the vascular lumen of affected capillaries, arterioles, and venules.<sup>5</sup> Skin lesions coincide with the site and extent of vascular damage.

*Diagnosis and Management*—The classic triad of fever, rash, and history of tick exposure is present in fewer than 20% of patients when medical attention is first sought.<sup>1</sup> During early stages, the differential diagnosis includes enteroviruses, infectious mononucleosis, measles, scarlet fever, ehrlichiosis, and leptospirosis. When the rash becomes petechial,

more serious illnesses (eg, *Neisseria meningitidis* infection) should be considered.<sup>4</sup>

In general, definitive diagnosis cannot be made until 6 to 10 days after symptom presentation. Serologic diagnosis depends on a 4-fold increase in antibody titer. During the acute phase, the only diagnostic test is direct immunofluorescence or immunoperoxidase staining of a skin biopsy. Although not universally available, this test is 70% to 90% sensitive and 100% specific and can be used to confirm RMSF in patients who present with a rash.<sup>4</sup> If clinical and epidemiologic data suggest RMSF, empirical antimicrobial therapy should be initiated without waiting for test results, to avert severe morbidity or death.<sup>6</sup>

Chloramphenicol and the tetracyclines are the only antibiotics with proven clinical efficacy in RMSF. Until this decade, treatment of young pediatric patients with tetracyclines was avoided because of the risk of permanently staining teeth. However, tooth staining is dose and duration dependent and unlikely to occur during the short treatment course of RMSF.<sup>6</sup> Furthermore, tetracyclines are the only agents with proven efficacy against the life-threatening disease human ehrlichiosis, which is often difficult to differentiate clinically from RMSF. Doxycycline is the recommended therapy for pediatric patients because of its documented effectiveness, broad margin of safety, low incidence of gastrointestinal disturbances, and convenient dosing schedule. It also binds less strongly to calcium and therefore is less likely to stain teeth.<sup>6</sup>

Doxycycline (4 mg/kg of body weight per day in 2 divided doses for children <45 kg or 100 mg every 12 hours for children >45 kg) currently is recommended for children of all ages with presumed or proven RMSF and should be administered for at least 3 days after fever subsides. When initiated during the first 4 to 5 days of illness, treatment leads to defervescence within 24 to 72 hours. A total of 5 to 10 days of therapy is typical; however, severe or complicated cases may require more protracted courses.<sup>7</sup>

**Prevention**—Until a vaccine becomes available, prevention strategies are of utmost importance for

populations at risk. Individuals with potential tick exposure routinely should search themselves, their children, and outdoor pets for ticks. Particular attention should be paid to the scalp, as this case illustrates. Because ticks require at least 6 hours to engorge with the host's blood and transmit infection, there is sufficient time to check for and properly remove ticks before infection sets in. Additionally, long sleeves and pants, as well as tick repellants (permethrin applied to clothing and DEET [N,N-diethyl-*m*-toluamide] applied to exposed skin), are recommended.<sup>1,8</sup> Adverse reactions to DEET, such as dermatitis, allergic reactions, and neurologic and cardiovascular toxicities, are more common in children than adults. However, preparations containing less than 10% DEET concentration are both safe and effective for children.<sup>6</sup> The use of prophylactic antibiotics for asymptomatic individuals following a tick bite has not been proven to be beneficial and may make diagnosis more difficult by prolonging the incubation period.<sup>1</sup>

## REFERENCES

1. Thomer AR, Walker DH, Petri WA Jr. Rocky Mountain spotted fever. *Clin Infect Dis*. 1998;27:1353-1360.
2. Walker DH. Tick-transmitted infectious diseases in the United States. *Annu Rev Public Health*. 1998;19:237-269.
3. Centers for Disease Control and Prevention. Summary of notifiable diseases, United States, 1998. *MMWR Morb Mortal Wkly Rep*. 1998;47(53).
4. Abramson JS, Givner LB. Rocky Mountain spotted fever. *Pediatr Infect Dis J*. 1999;18:539-540.
5. Schaffner W. Rickettsial diseases. In: Fitzpatrick TB, Eisen AZ, Wolff K, et al, eds. *Dermatology in General Medicine*. 4th ed. New York, NY: McGraw-Hill; 1993:2500-2503.
6. Cale DF, McCarthy MW. Treatment of Rocky Mountain spotted fever in children. *Ann Pharmacother*. 1997;31:492-493.
7. Centers for Disease Control and Prevention. Rocky Mountain spotted fever. Available at: <http://www.cdc.gov/ncidod/dvrd/rmsf>. Accessed April 3, 2002.
8. Brown M, Hebert AA. Insect repellants: an overview. *J Am Acad Dermatol*. 1997;36:243-249.

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