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A 6-YEAR-OLD GIRL WITH FEVER, RASH, AND INCREASED INTRACRANIAL PRESSURE

Matthew E. Ravish, MD,* Daniel P. Krowchuk, MD,* Michael Zapadka, MD,† and Avinash K. Shetty, MD*

*Department of Pediatrics and †Department of Radiology, Wake Forest University School of Medicine and Brenner Children's Hospital, Winston-Salem, North Carolina

Reprint Address: Avinash K. Shetty, MD, Wake Forest University Health Sciences, Department of Pediatrics, Medical Center Boulevard, Winston-Salem, NC 27157

□ **Abstract—Background: Rocky Mountain spotted fever (RMSF) is a well-described, potentially lethal, tick-borne zoonotic infection and has very effective therapy. However, the diagnosis might not be made early enough, often leading to worse outcomes. Objective: Our aim was to discuss the diagnostic dilemmas facing the physician when evaluating patients with suspected RMSF. Methods: We report a case of RMSF in a 6-year-old girl who presented to our hospital with a 7-day history of fever, headache, and a petechial rash. After blood cultures were obtained, the patient was treated empirically with doxycycline, vancomycin, and ceftriaxone. During the next 24 h, her clinical status worsened, with acute onset of altered mental status, posturing, and fixed and dilated pupils. A computed tomography scan of the brain demonstrated diffuse cerebral edema with evidence of tonsillar herniation. She died 24 h after admission. A serum specimen tested positive for immunoglobulin G to *Rickettsia rickettsii* at a titer of 128 dilutions, confirming recent infection. Conclusions: We present this case to raise awareness of RMSF in patients who present with a nonspecific febrile illness in tick-endemic areas in the United States. Early diagnosis and treatment with doxycycline before day 5 of illness is essential and can prevent morbidity and mortality. © 2013 Elsevier Inc.**

□ **Keywords—Rocky Mountain spotted fever; raised intracranial pressure; fever; rash**

INTRODUCTION

Rocky Mountain spotted fever (RMSF) is a potentially lethal tick-borne zoonotic infection caused by *Rickettsia rickettsii,* an obligate intracellular gram-negative pathogen (1). Even though RMSF is well described and has very effective therapy, making an early diagnosis of RMSF often poses a diagnostic dilemma for the clinician, often leading to worse outcomes (2). We report a fatal case of serologically proven RMSF in a 6-year-old girl and discuss the diagnostic dilemmas facing the physician when evaluating patients with suspected RMSF. This case underscores the need for physicians to diagnose early and treat appropriately this potentially lethal infectious disease.

CASE REPORT

A 6-year-old white girl was transferred to our hospital for management of possible sepsis. The patient was well 7 days earlier and then began to have fever and headache. She was evaluated by her pediatrician and diagnosed with a viral illness and prescribed acetaminophen. A throat swab for rapid group A Streptococcus antigen testing was negative. During the subsequent 2 days, fever persisted and abdominal pain and a maculopapular rash

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developed. On day 7 of illness, the patient was admitted to a community hospital with continued fevers, headache, vomiting, and a petechial rash. A blood culture was obtained and she received intravenous ceftriaxone. On day 8 of illness, she was transferred to our hospital secondary to worsening clinical status. The patient resided in eastern North Carolina. Two weeks before admission, she had a tick bite, but denied sick contacts or recent travel.

On admission, she was ill-appearing with a temperature of 100°F, heart rate of 138 beats/min, respiratory rate of 20 breaths/min, and blood pressure of 87/58 mm Hg. A diffuse maculopapular rash was noted, with petechia noted on her face, chest, trunk, and extremities, including palms and soles (Figure 1). Abdominal examination was notable for hepatomegaly and right upper quadrant tenderness. Her extremities demonstrated a 1+ bilateral pretibial and pedal edema. The rest of the examination was unremarkable.

Laboratory tests showed a hemoglobin level of 9.7 g/dL, white blood cell count of 10,300 g/dL (with 53% neutrophils and 39% band neutrophils) and a platelet count of 35,000/mm^3. A comprehensive metabolic panel showed hyponatremia (serum sodium, 134 mEq/L), hypoalbuminemia (2.3 g/dL [normal 3.8–5.4 g/dL]), elevated aspartate aminotransferase (453 U/L [normal 15–50 U/L]) and alanine aminotransferase (258 U/L [normal 15–50 U/L]), but normal renal function. A C-reactive protein was 272 mg/L (normal 0–10 mg/L). Prothrombin time was 14.2 s (normal 8.9–12.1 s), activated partial thromboplastin time was 69.4 s (normal <30 s), and quantitative D-dimer was 12.42 mg/L (normal <2.30 mg/L). Chest and abdominal x-ray studies were normal. Blood cultures were obtained.

Given the clinical presentation of an ill-appearing child with high fever and petechial rash, the differential diagnosis considered on admission included bacterial sepsis due to *Neisseria meningitidis*, community-acquired *Staphylococcus aureus*, *Streptococcus pneumonia,* and rickettsial diseases, such as Rocky Mountain spotted fever (RMSF) and Ehrlichiosis. The patient was treated empirically with doxycycline, vancomycin, and ceftriaxone. During the next 24 h, her clinical status worsened, with acute onset of altered mental status, posturing, and fixed and dilated pupils. A computed tomography scan of the brain demonstrated diffuse cerebral edema with evidence of tonsillar herniation (Figure 2). She died 24 h after admission. Blood culture was sterile. A serum specimen tested positive for immunoglobulin G to *R. rickettsii* at a titer of 128 dilutions, thereby confirming recent infection.

Figure 1. Petechial skin rash.

DISCUSSION

RMSF is a potentially lethal zoonotic infection caused by *R. rickettsii,* an obligate intracellular gram-negative pathogen and is transmitted to humans via the bite of the American dog tick (*Dermacentor variabilis*), the Rocky Mountain wood tick (*Dermacentor andersoni*), and the brown dog tick (*Rhipicephalus sangineus*) (1). Most cases occur in the south Atlantic, southeastern, and south central states, although recently, a new vector for RMSF (*Rickettsia sangineus*) was identified in the southwestern United States (US) (2–4). The disease can affect people of any age, but most cases occur in children 1–9 years of age (1,3). The highest incidence occurs April through September, correlating with peak tick activity (5).

The clinical manifestations of RMSF are secondary to the invasion of endothelial cells of small vessels by the organism (1). After an incubation period of 5–10 days, the

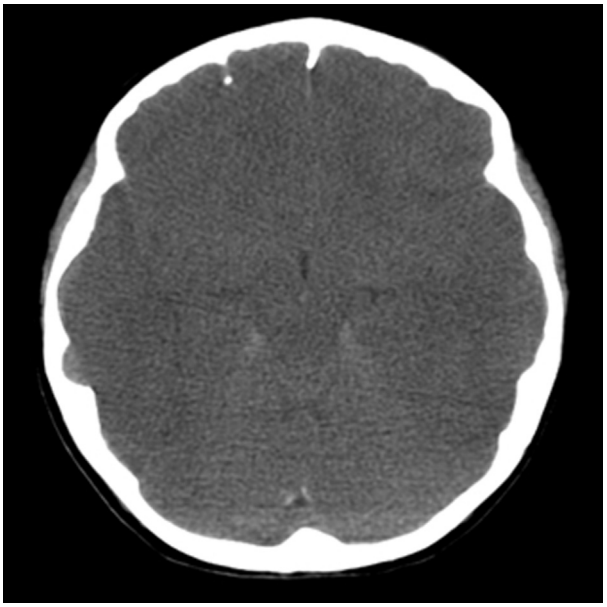

Figure 2. Noncontrast computed tomography of the brain demonstrates diffuse cerebral edema with evidence of transtentorial herniation.