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## Rocky Mountain Spotted Fever Encephalitis: MR Findings

Mark D. Baganz, Peter E. Dross, and John A. Reinhardt

**Summary:** We describe a case of Rocky Mountain spotted fever encephalitis and present the associated findings of an MR examination of the brain, which showed increased signal intensity in the distribution of perivascular spaces. Resolution of the MR abnormalities coincided with clinical improvement.

**Index Terms:** Encephalitis; Brain, magnetic resonance

Rocky Mountain spotted fever is an acute, often severe, tick-borne infection caused by the bacterium *Rickettsia rickettsii*. The most common symptoms include fever, rash, and headache. The infection spreads hematogenously, producing a vasculitis that directly damages the blood vessel walls of virtually any organ. Encephalitis, which may be fatal, is a frequent manifestation in cases escaping early diagnosis and treatment. The case reported here of Rocky Mountain spotted fever encephalitis is accompanied by magnetic resonance (MR) demonstration of the associated perivascular inflammatory response, as well as resolution of the MR abnormalities coinciding with clinical improvement.

### Case Report

A 45-year-old previously healthy woman was admitted to an outside community hospital after 1 week of progressive symptoms consisting of dizziness, myalgias, fever, confusion, and disorientation. Several days before admission, she had been treated by her family physician for fever, dizziness, and an earache; oral ticarcillin-clavulanic acid was prescribed. A few days before admission, a red punctate rash developed on her arms and legs. She had sustained a tick bite about 1 week before the onset of symptoms.

On admission to the outside institution, physical examination revealed a fever of 40°C and a maculopapular rash with petechial areas on her torso and the flexor surfaces of her upper and lower extremities (Fig 1). Laboratory studies revealed a white blood cell count of 9300/mm<sup>3</sup>, hemoglobin of 11.6 g/dl, hematocrit of 34%, and platelet count



Fig 1. The flexor surface of the patient's forearm soon after admission shows the petechial rash that is characteristic of Rocky Mountain spotted fever.

of 49 000/mm<sup>3</sup>. Liver function tests showed total bilirubin of 7.0 mg/dl, direct bilirubin of 5.2 g/dl, aspartate aminotransferase of 256 U/ml, and lactate dehydrogenase of 848 U/ml.

Because of high fever and jaundice, she was presumed to have cholangitis, and broad-spectrum antibiotic treatment was initiated with imipenem. There was no response to this treatment, and the patient became increasingly lethargic and hypotensive. She was transferred to our institution for further evaluation and treatment after 4 days of hospitalization.

She had a grand mal seizure during transport to our institution. On admission, she was unresponsive, except for assuming decerebrate positioning in response to pain-

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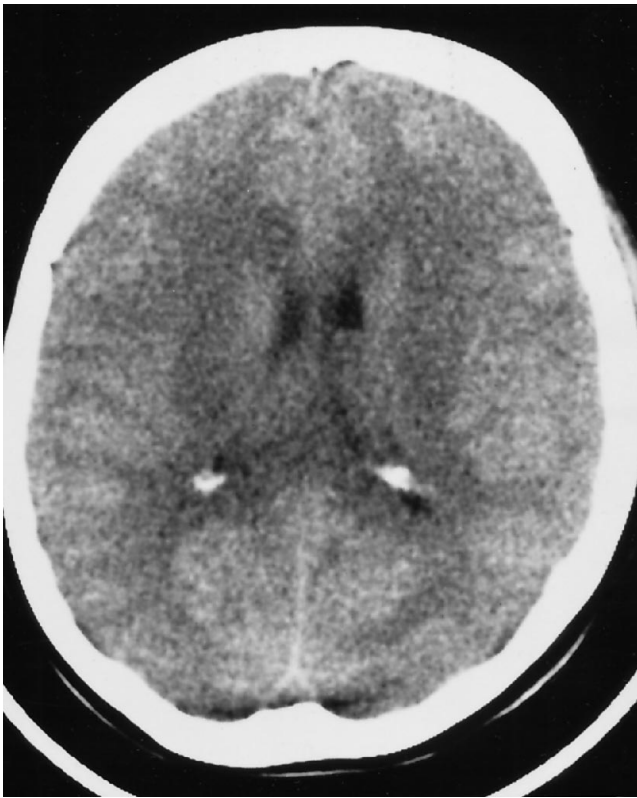


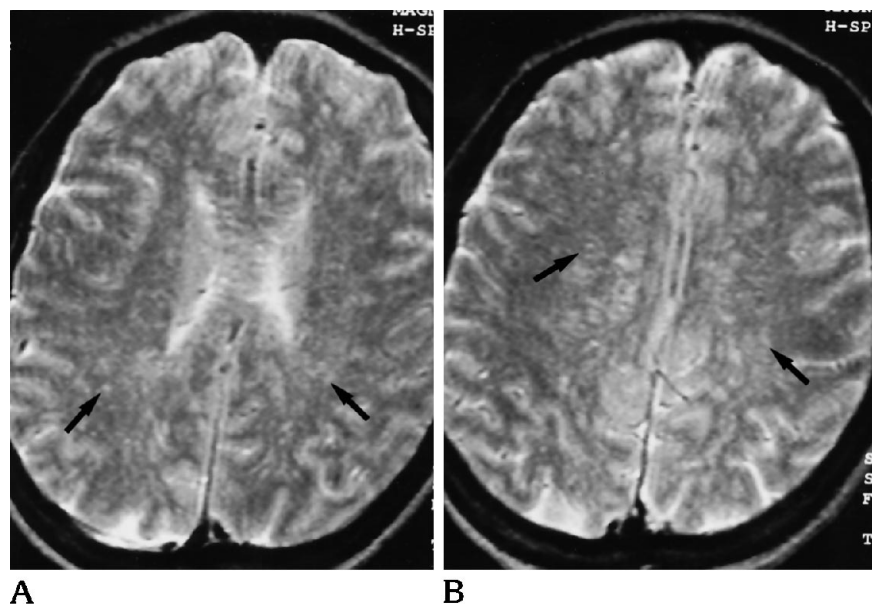
Fig 2. Unenhanced CT scan of the brain at admission shows a subtle decrease in the size of the lateral ventricles and cortical sulci, with heightened gray matter/white matter differentiation, suggestive of a diffuse white matter process.

ful stimuli. She had a blood pressure of 58/44 mm Hg and a platelet count of 48 000/mm<sup>3</sup>. Cerebrospinal fluid cell count was normal, and Gram-stained specimen and cultures were negative for organisms. An electroencephalogram demonstrated diffuse generalized slowing with occasional sharp paroxysmal discharges consistent with a generalized encephalopathy.

Computed tomography (CT) of the brain on the second day of hospitalization at our institution suggested mild diffuse white matter hypodensity with small-caliber ventricles and sulci thought to be consistent with diffuse cerebral edema (Fig 2). MR examination of the brain was performed with a 1.0-T magnet 1 day after admission to our institution. T2-weighted images showed multiple punctate areas of increased signal throughout the cerebral white matter in the distribution of perivascular spaces, corresponding to Virchow-Robin spaces or possibly small end-artery lesions (Fig 3). No abnormalities were identified on T1-weighted images, either before or after administration of gadopentetate dimeglumine. Another MR examination of the brain, performed on the 19th day of hospitalization after clinical improvement, showed a marked decrease in the prominence of the punctate areas of increased T2 signal (Fig 4).

Because of the thrombocytopenia, encephalopathy, history of a tick bite, and maculopapular rash, the diagnosis of Rocky Mountain fever was suspected, and the patient was treated with chloramphenicol. Presumptive serologic evidence of Rocky Mountain spotted fever was provided by a positive Weil-Felix test, which demonstrated a *Proteus vulgaris* Ox-2 titer of 1:20, an Ox-19 titer of 1:2560, and a negative Ox-K titer. Definitive diagnosis of Rocky Mountain spotted fever was provided by positive acute and convalescent serum antibody levels to *Rickettsia rickettsii*. The patient's clinical status improved with treatment, and she was discharged in good condition 22 days after admission.

Fig 3. T2-weighted MR images (2000/80/2 [repetition time/echo time/excitations]) of the brain obtained at admission show numerous punctate regions of increased signal (arrows) in the distribution of perivascular (Virchow-Robin) spaces.



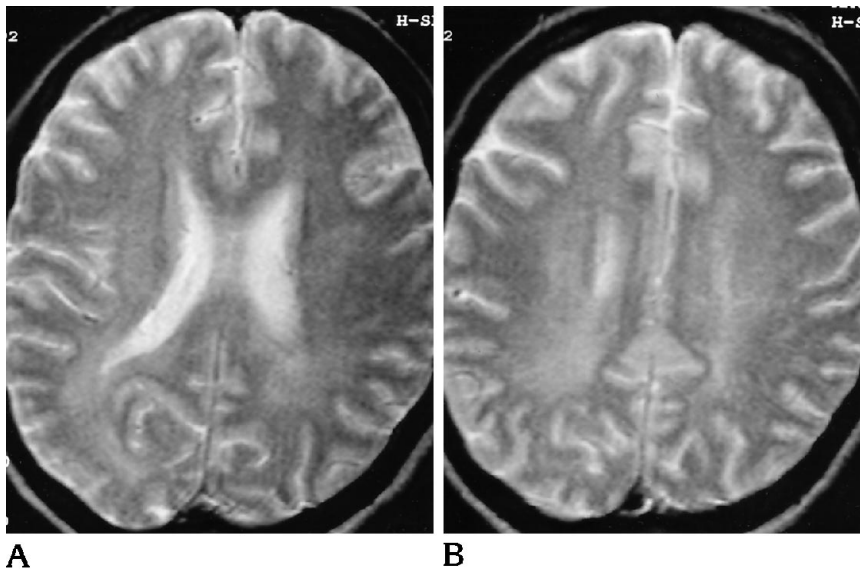


Fig 4. T2-weighted MR images (2000/80/2) of the brain obtained after clinical improvement show substantial resolution of the high signal, corresponding to resolution of the perivascular inflammation.

## Discussion

Rocky mountain spotted fever, first documented at the end of the 19th century among the residents of the Bitter Root and Snake River valleys of Montana and Idaho, is an acute, tick-borne, often fatal disease caused by the obligate intracellular bacterium, *Rickettsia rickettsii* (1, 2). The causative organism was named after Howard Ricketts, who was the first to demonstrate the role of the tick (*Dermacentor andersonii*) as the vector for the disease in western Montana in 1906 (3). Over 1000 cases are reported each year in the United States, primarily in rural areas and mainly in late spring and early summer during periods of maximal tick activity (4).

The most frequent presenting symptoms of the illness include fever, headache, rash, and myalgias (5). The rickettsiae spread via the blood stream, proliferate and injure endothelial and vascular smooth muscle cells, and therefore cause damage to the microcirculation of virtually all organs (6). The vascular damage is the pathophysiologic basis for the rash and meningoencephalitis found with Rocky Mountain spotted fever.

The rash, occurring in as many as 90% of patients with the disease, begins on the cooler parts of the body (ie the extremities) and is the finding that most often leads physicians to consider the disease (7). First maculopapular in nature, the rash usually begins on the flexor surfaces of the hands and feet and then spreads centripetally to involve the entire body, features

clearly demonstrated by our case. Later in the disease, the skin lesions become petechial and eventually purpuric and ecchymotic and can rarely lead to skin necrosis or gangrene (1, 7).

Early recognition of the central nervous system involvement is critical, as such involvement is often a causative factor in deaths (8, 9). Less serious central nervous system symptoms include lethargy, confusion, disorientation, and most commonly, headache. More severe neurologic findings are ataxia, aphasia, paralysis, seizure, stupor, and coma (9). Unresponsiveness in a patient with Rocky Mountain spotted fever carries a poor prognosis, with up to 88% mortality for patients in stupor (5), making our patient an unusual survivor of this serious neurologic state.

The brain lesions in a Rocky Mountain spotted fever patient with this disease assume a characteristic pathologic appearance, the "typhus nodule," consisting of perivascular accumulations of mononuclear cells (6). Other pathologic lesions include white matter microinfarcts and a mononuclear cell-rich leptomeningitis. The perivascular inflammatory response seen with Rocky Mountain spotted fever the disease is similar to that of typhus and other rickettsial infections, and may mimic the necrotizing vasculitis of collagen vascular diseases (10).

Descriptions of head CT findings in Rocky Mountain spotted fever include normal CT findings (2) and cerebral infarction (9). Benhammou et al (11) reported ischemic internal cap-

sules on a head CT scan of a patient with Mediterranean boutonuse fever, another rickettsial disease that has a similar pathophysiology. Brooks et al (12) reported a case of acute Q-fever encephalitis attributed to the rickettsial organism *Coxiella burnetii* in which findings on head CT were normal. Cerebral CT in our case showed no indication of infarction, although a diffuse white matter process was suggested.

The initial brain MR examination in our case showed a pattern of abnormal signal intensity on T2-weighted images in the distribution of perivascular spaces indicative of the perivascular inflammatory response that is central to the pathophysiology of Rocky Mountain spotted fever vasculitis. Similar MR findings described in central nervous system cryptococcosis in immunocompromised patients (13, 14) and in Lyme disease encephalitis (15) represent the perivascular inflammation that is common to all these disease processes. The follow-up MR study of our patient showed clearing of this perivascular high signal, suggesting the perivascular inflammation is reversible if treated in time.

In summary, a case of Rocky Mountain spotted fever encephalitis was documented with MR in a rare survivor of severe neurologic deficits caused by this rickettsial disease. The perivascular inflammatory response in the vasculitis manifests itself by prominent perivascular spaces, representing dilated Virchow-Robin spaces or small end-artery lesions. Similarities are drawn to cryptococcosis and Lyme disease, also known to cause perivascular inflammation pathologically and dilated Virchow-Robin spaces on MR imaging. Reversibility of these MR findings is also documented.

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