

Case 5

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Clinical abstract: The patient was a 41-year-old white female with a two-day history of headache, nausea and photophobia. She was admitted to the emergency room and rapidly became unresponsive and unarousable, and suffered a generalized seizure with decorticate posturing five hours after admission. Repeated lumbar punctures revealed opening pressures of 320-326 mm/Hg, 1000 mg of protein and a mild pleocytosis (10 WRC/mm³). A CT scan showed diffuse edema. The patient was started on antiviral therapy in view of the presence of oral fever blisters; however, she rapidly deteriorated and died 32 hours after admission.

At autopsy, the brain was grossly swollen, and on coronal sectioning showed marked vascular congestion of the subcortical white matter and diffuse white matter edema. Congestion was also found in the thalami and basal ganglia. There was a segment of cortex, about 2 cm long, along the lateral border of the left posterior parietal lobe which showed marked hemorrhagic discoloration. Aside from that lesion, no hemorrhages or necrosis were seen in the cerebrum, cerebellum or mid-brain.

Within the same week, three other patients were admitted with the same clinical symptoms, but the disease was less fulminating. All three patients had a brain biopsy performed from the frontal region, and in two of these patients the microscopic findings were similar to those demonstrated in the slide submitted here.

Following various negative investigations for the demonstration of a bacterial, viral, toxic or metabolic disorder, all three patients admitted within a week of the present case showed increased titers in their serum against OX proteus (Weil-Felix reaction) and significant raises in the latex agglutination test for Rocky Mountain spotted fever. A fifth patient, admitted two weeks later, has shown a similar rise in the latex agglutination test. In none of the patients was there any history of having been bitten by a tick. Two of the four patients developed a rash after admission for their neurological disease, and in one of them the rash was so unremarkable as to be originally attributed to trauma.

Material submitted: One H&E stained section. The slide submitted is representative of the most severe lesions found microscopically in the deep white matter and in the basal ganglia, especially the thalamus.

- Points for discussion:
1. Diagnosis
 2. Specificity of the neuropathological lesion
 3. Are we dealing with a new type of Rickettsial organism, possibly with special neurotropic virulence?