
I read with great interest the article by Zu-Rhein et al (1), which presented 3 cases that closely resembled a case I had encountered in 2004 and presented at The Canadian Association of Neuropathologists’ 2010 annual meeting. A 50-year-old woman without previous pertinent medical or surgical history presented with a subacute progressive neurologic syndrome evolving during a 4- to 5-year period. She initially presented with tremor, gait difficulty, and disequilibrium along with motor deficits characterized by hyperreflexia, clonus, and rigidity. During the next few years, she developed a frontal dementia with psychotic elements. Weeks before her death, she displayed episodes of syncope and falls followed by seizures, culminating in status epilepticus.

Laboratory investigations, including computed tomographic scan, and magnetic resonance imaging, were normal. A positron emission tomographic brain scan showed a slight bilateral frontal deficit. Cerebrospinal fluid leak analysis revealed a slight increase in protein but was otherwise normal, including a search for oligoclonal bands, infectious workup, and 14-3-3 protein analysis. An electroencephalogram showed slight cortical dysfunction. Complete metabolic workup was negative as was heavy metal screening. Family history was negative for neurologic disease. The postulated clinical diagnosis was Creutzfeldt-Jacob disease.

The lesions in the patient’s brain were identical, in most respects, to those reported by Zu-Rhein et al (Figs. 1, A–F). The only exceptions were an even greater number of abnormal endothelial cell nuclei and a lack of mineralization, perhaps reflecting an earlier stage. Our pontine section, however, was more rostral than theirs.

I thank Drs Zu-Rhein and Powers for reviewing my case, comparing it with theirs, and advising me on a suitable venue to document this case.

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FIGURE 1. (A) Atypical (arrows) to clustered (circle) endothelial cell nuclei in cerebral white matter. Hematoxylin and eosin. (B) Mildest myelin lesion in center of field consisting of a loss of myelin staining and oligodendroglial nuclei. A necrotic microvessel (arrowhead) lies within the lesion; there are atypical to clustered endothelial cell nuclei (arrows). Hematoxylin and eosin. (C) Some myelin lesions are spongy because of macrophages with myelin debris (center of field; blue granules). Atypical endothelial cell nuclei (arrows). Luxol fast blue–hematoxylin and eosin. (D) Two necrotic and thickened microvessels (arrowheads) within a lesion displaying a mild loss of myelin staining, a few macrophages, and reactive astrocytes. Luxol fast blue–hematoxylin and eosin. (E) Multiple dilated, telangiectatic-like, microvessels (bv) in cerebral white matter. Hematoxylin and eosin. (F) Pleomorphic circular to elliptical electron opaque mycoplasma-like particles (MLPs; top center) lie between 2 nuclear lobes (N) of an atypical endothelial cell. A cluster of empty MLPs is at the bottom of the field near a split basal lamina. Most MLPs have diameters of approximately 141 to 237 nm and trilaminar limiting membranes (not shown). Uranyl acetate–lead citrate.