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CASE 2011-6

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Clinical History:
The deceased was a 54-year-old gentleman with a history of mental disability, functioning at a level of a 5-year-old child. He presented with a history of staggering, difficulty moving legs and incontinence progressing to spastic quadriparesis in less than a week. The initial MRI revealed central and cortical atrophy, with no evidence of structural disease, stroke or trauma, but revealed significant cervical spinal stenosis at C5-6. Initial CSF studies were normal. He was then placed on high dose steroids and underwent a decompressive laminectomy, but failed to improve. He was then given IVIg for a possible stiff man syndrome. Repeat CSF revealed mild protein elevation (73 g/dl), but no pleocytosis. Repeat MRI showed several scattered bilateral 1-2 mm T2W/FLAIR bright foci scattered in the corona radiata, subcortical white matter and thalami. He then developed mild headache and low-grade fever, eventually became unresponsive and died two months after his initial presentation. A brain-only autopsy was performed and the brain was sent to the National Prion Disease Pathology Surveillance Center to rule out an atypical prion disease.

Autopsy findings:
The brain weighed 1,318 grams post-fixation and had no external abnormalities. On section, numerous poorly defined, partially confluent areas of yellow-tan to red-tan discoloration were identified, involving predominantly the deep cerebral cortex and the superficial subcortical white matter. These areas occurred within all lobes of the cerebrum as well as in the cerebellar cortex. These same lesions were identified bilaterally in the thalami and surrounding the fourth ventricle.

Material submitted: One H&E section of the brain

Points for discussion:
1. Differential diagnosis.
2. Pathogenesis