

49th ANNUAL DIAGNOSTIC SLIDE SESSION 2008

CASE 2008 - 2

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Clinical History: Seven years (year 2000) prior to her death this 79-year-old woman developed progressive walking difficulties accompanied by muscle cramps. A year later she required the use of a walker. Initially, she had spasticity in the left arm and leg in a pyramidal distribution with brisker DTR's and an up-going toe; later, she manifested difficulties while manipulating small objects. Her gait became wide-based and spastic, and she could only take small steps. Of note were the absence of dysphagia notwithstanding some dysarthria, lack of sensory abnormalities and that spasticity had extended to the right side of her body. The patient displayed no ataxia and power in the sternomastoid and trapezius muscles remained normal. The attending physicians remarked about the unfortunate circumstances at home, with the patient having physical impairment while being able to make decisions for her husband's care as he was developing cognitive dysfunction because of strokes. There was no family history of neurodegeneration.

Over the ensuing years the patient's condition had a protracted course. She remained ambulatory and resided in a chronic care facility with periodic visits to the amyotrophic lateral sclerosis clinic with the diagnosis of primary lateral sclerosis; clinical notes were sparse but noted that the patient had developed a hand tremor. Minor lower motor neuron dysfunction was interpreted in one EMG study. The nurses' report from January 2007 indicated that the patient was fully alert, aware, and able to converse coherently and fluently; she fully understood commands and made decisions when asked about options for supportive treatment. Death, cause by respiratory failure, occurred on May 3, 2007.

Necropsy findings: The brain showed slight frontotemporal atrophy and weighted 1100 grams. The corticospinal tracts were slightly pale. Using routine preparations and UBQ and TDP-43 protein immunostains, no changes of ALS or of FTLD "U" were demonstrable. Additional immunolabeling (Tau, AT8) and silver stains evinced Alzheimer neurofibrillary change Braak stage I/VI

Materials submitted:

1. LFB-H&E-stained section of sensorimotor area
2. Unstained section of sensorimotor area

Points for discussion:

1. Diagnosis
2. Pathogenesis