

Joint Event
12th International Conference on
Vascular Dementia and Dementia
&
8th International Conference on
Neurological Disorders and Stroke

March 14-16, 2019 | London, UK

Alzheimer's disease and severe cerebral Amyloid Angiopathy; (CAA), CAA related Leukodystrophy leading to synergistic rapidly progressive Dementia

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We present an informative case of a female with a rapidly progressive dementing illness clinically, initially thought to represent a form of prion encephalopathy. The patient succumbed to the illness after an approximate two month course and came to a neuro autopsy. Examination of the brain showed no changes of prion disease, but rather typical Alzheimer's disease neuropathology with a severe cerebral amyloid angiopathy (CAA). No other significant small vessel disease such as hyalinizing arteriosclerosis was present. The severe CAA was widespread, was unassociated with infarction, but lead to a marked

generalized secondary leukodystrophy. We emphasize the interrelationship of Alzheimer's disease and CAA, in particular severe CAA which can produce a vascular based leukodystrophy. This in turn can synergize with and contribute significantly to the dementia. This can contribute to the underlying pathogenesis of the dementia through a white matter disconnection syndrome with acceleration of progression of the Alzheimer-type pathology. Furthermore, the accelerated clinical course due to the synergism can mimic other dementias such as prion disease.

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