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Case 1

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Clinical Abstract:

This male patient developed, around the age of 54, intermittent tremor, first of the right hand, then of the left, that progressed to the point where he had great difficulty with handwriting. This was followed by unsteadiness of gait, mild slurring of speech, impairment of memory, mild urinary urgency, and declining sexual function, although he denied any visual or hearing difficulty.

An older brother, 20 years his senior, had suffered from head tremor, although he was fully ambulatory until his death in his 80's from "old age". His brother's daughter, now aged 65, suffers from the same ailment.

Examination at age 64 showed him to be slightly dysarthric. Gait was wide-based, and there was mild unsteadiness when attempting to tandem-walk. There was a moderately severe intention tremor of the right arm and a mild to moderate tremor on the left side. Heel-to-shin testing revealed clumsiness and mild intention tremor in the legs. Muscle strength, bulk, and tone were normal throughout. Muscle stretch reflexes, however, were barely perceptible at the biceps and knees and absent elsewhere. Plantars were flexor. Sensory exam was normal to pin, touch, vibration, and joint position testing in the hands and feet.

Hemogram, blood chemistries, folate, vitamin B12, vitamin E, tests of thyroid function, and ceruloplasmin were normal. LP returned clear fluid (OP 150) with 2 WBC, a protein of 42, a glucose of 78, an IgG/albumin ratio of 0.17, no oligoclonal bands, and a negative VDRL. CT of the head showed diffuse bilateral lucency in the deep white matter above and below the tentorium. MRI showed severe white matter abnormality that was judged to be compatible with either subcortical arteriolar encephalopathy or demyelination. The cerebellar hemispheres were also judged to be affected.

The patient died of myocardial infarction at the age of 65.

At autopsy, the brain in coronal section showed the presence of irregular, roughly symmetrical, sharply circumscribed, partially confluent foci of tan-grey darkening and (especially rostrally) retraction within the dorsolateral frontal, parietal, and occipital white matter. These lesions were separated from the overlying cerebral cortex by a slender band of well preserved subcortical white matter and from the margins of the lateral ventricles by a distance of 0.5 cm or more.

Material Submitted: One H&E- and one unstained section of cerebral cortex and white matter

Point for Discussion: Diagnosis