The pivotal sign of CANVAS

A 75-year-old woman complained about insecure gait since age 55. Clinical examination revealed signs of cerebellar ataxia, bilateral vestibulopathy, and peripheral sensory impairment. Sensory nerve action potentials were absent. The visually enhanced vestibulo-ocular reflex (VVOR) was impaired (video on the Neurology® Web site at www.neurology.org, figure 1) and the diagnosis of cerebellar ataxia (figure 2) with neuropathy and bilateral vestibular areflexia syndrome (CANVAS) was made.1 CANVAS is considered to be a recessive disorder with a mean age at onset of 60 years.2 VVOR impairment is its characteristic clinical sign.2 It can only be elicited if both smooth-pursuit eye movements and the vestibulo-ocular reflex are deficient. Normally, both are redundant at low head velocities.2

Jens Alexander Petersen, MD, Werner W. Wichmann, MD, Konrad Peter Weber, MD

From the Departments of Neurology (J.A.P., K.P.W.), Neuroradiology (W.W.W.), and Ophthalmology (K.P.W.), University Hospital Zurich, Switzerland.

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Marked cerebellar atrophy involves the vermal lobules VI, VIIa, and VIIb. Note also the spinal atrophy. CANVAS = cerebellar ataxia with neuropathy and bilateral vestibular areflexia syndrome.

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