The pivotal sign of CANVAS

Figure 1 Video-oculography of impaired visually enhanced vestibulo-ocular reflex and pathologic head impulse test

(A) Slow head rotations demonstrate an impaired visually enhanced vestibulo-ocular reflex with multiple catch-up saccades (arrowheads) instead of smooth compensatory eye movements. (B, C) The video head impulse test shows bilaterally reduced vestibulo-ocular reflex gains with overt catch-up saccades (arrows) after head rotation to either side.

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

Supplemental data at
www.neurology.org

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Author contributions: J.A. Petersen recorded the video and wrote the manuscript. W. Wichmann acquired the MRI and revised the manuscript. K.P. Weber designed the study, acquired the eye movement data, and revised the manuscript.

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Figure 2 T2-weighted sagittal MRI illustrates the cerebellar atrophy in CANVAS

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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