Central neurogenic hyperventilation

Figure Extensive pontine lesions in CLIPPERS resulting in central neurogenic hyperventilation

Axial (A) and coronal (B) T1 postgadolinium brain MRI shows characteristic punctate and curvilinear enhancement peppering the brainstem in chronic lymphocytic inflammation with pontine perivascular enhancement responsive to steroids syndrome (CLIPPERS). Presumed medial pontine nuclear involvement resulted in central neurogenic hyperventilation (D) with inability to breath-hold (C).

A 79-year-old woman developed subacute progressive gait instability. Brain MRI demonstrated patchy nodular enhancement within the brainstem and cerebellum (figure). Brain biopsy was consistent with chronic lymphocytic inflammation with pontine perivascular enhancement responsive to steroids syndrome (CLIPPERS).¹ Questioning revealed shortness of breath (figure, video). Blood gas on room air revealed pH 7.59, PaCO₂ 16, PaO₂ 103, and HCO₃ 15.

Central neurogenic hyperventilation, often associated with brainstem lymphoma, rare in noncomatose patients, and never described in CLIPPERS, results from released respiratory inhibition from medial pontine nuclear damage.² Being comfortably tachypneic can be due to a pontine lesion.

Christopher L. Kramer, MD, Eelco F.M. Wijdicks, MD, PhD

From Mayo Clinic, Rochester, MN.

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Correspondence to Dr. Kramer: kramer.christopher1@mayo.edu

Supplemental data
at Neurology.org

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