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Researches Classified – Panels Award – Case Report

Cervical spondylotic myelopathy with concomitant ex vacuo hydrocephalus in the differential diagnosis of idiopathic normal pressure hydrocephalus

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Introduction: Idiopathic normal pressure hydrocephalus (iNPH) is a syndrome of gait disturbance with enlargement of the cerebral ventricles, not justified by another cause. It frequently occurs with cognitive dysfunction and overactivity of the bladder's detrusor muscle, resulting in dementia and urinary incontinence. We report a case in which the patient´s gait and initial image suggested iNPH, posteriorly found to be a cervical spondylotic myelopathy (CSM).
Objectives: To report a CSM with concomitant ex vacuo hydrocephalus case resembling iNPH and to discuss the biases of the initial diagnosis.
Case presentation: A 78 year-old woman, previously diagnosed with hypertension and type 2 diabetes mellitus, presented to our ambulatory with a long history of progressive gait disturbance, radiating pain to the right arm and several episodes of falls. The relatives referred possible cognitive deficits in attention and memory, as well as urinary urgency. Given the peculiar unstable wide-based gait and history, an iNPH was suspected. At the outpatient unit, a transcranial ultrasound examination revealed large cerebral ventricles, compatible with hydrocephalus. A brain magnetic resonance imaging (MRI) obtained at another facility suggested iNPH. The patient was then hospitalized at our hospital for investigation of iNPH and had a lumbar tap test scheduled. The complete neurological examination revealed: an unstable wide-based gait, with head and neck leaning forward, marche à petit pas, and prolonged Timed Up and Go Test; positive Romberg's test; spasticity of the lower limbs; globally reduced muscular strength, especially at distal lower limbs and right hand (compatible with C6 and C7 myotomes); global preserved or enhanced reflexes; Hoffman and Tromner signs bilaterally; inversion of the right bicipital due to enlargement of the reflexive area of the ipsilateral tricipital; cutaneous plantar reflexes in flexion; impaired sensations on the right hand, especially on the index and middle fingers, with normal sensations on the left hand; loss of vibratory sensation at distal lower limbs; positive Spurling's test; normal cranial nerves; normal coordination. Frontal Assessment Battery, Mini Mental State Examination and Montreal Cognitive Assessment showing no signs of cognitive impairment. As the neurological examination added a possible spinal cord syndrome differential diagnosis, the tap test was postponed and both brain and cervical MRIs ordered. The cervical MRI revealed a serious and extensive CSM compressing the spinal cord at C3 through C5. The brain MRI obtained at our institution showed hydrocephalus with no ventricular or acqueductal flow void, normal callosal angle and diffuse signs of brain atrophy, suggesting ex vacuo hydrocephalus. The patient then awaits for cervical decompressive neurosurgery. We bring this case to shed light on the differential diagnosis between these conditions, a case infrequently found in medical literature. The atypical presentation

of CSM with longstanding parkinsonian-like gait, urinary disturbance and vague cognitive complaints, aside with the first mistaken MRI report, allowed the attending clinicians to search for iNPH. Whilst, notwithstanding the importance of complementary exams, its high accountability may lead to diagnostic error and unnecessary invasive procedures. This report emphasizes the importance of a complete neurologic examination to highlight a correct topographic and nosological diagnosis.

Keywords: Case report; Cervical spondylotic myelopathy; Ex vacuo; Hydrocephalus; Normal pressure hydrocephalus.