









Case Report 305

A Rare Presentation of Functional Movement Disorder Mimicking Normal Pressure Hydrocephalus

Halil Onder¹ Aycan Cemil Ulker¹ Selcuk Comoglu¹

¹ Department of Neurology, Neurology Clinic, Diskapi Yildirim Beyazit Training and Research Hospital, Ankara, Türkiye

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Address for correspondence Halil Onder, MD, Department of Neurology, Neurology Clinic, Diskapi Yildirim Beyazit Training and Research Hospital, Şehit Ömer Halisdemir Street. No: 20, Altındag, Ankara 06110, Türkiye (e-mail: halilnder@yahoo.com).

Abstract

Keywords

- ► functional movement disorder
- ► gait
- ► CSF tap test
- hydrocephalus
- ► normal pressure hydrocephalus

Herein, we describe a 55-year-old female patient with a functional movement disorder (FMD) who presented with normal pressure hydrocephalus (NPH)-like clinic. The neuroimaging data and positive response to the tap test initially suggested NPH. However, a detailed investigation of the clinic features yielded a final diagnosis of FMD. Via the presentation of this patient, we expand the phenomenology of FMD. To our knowledge, this is the first presentation of a patient with FMD mimicking NPH. Therefore, we think this rare illustration is interesting and may provide valuable perspectives for clinical practice.

Introduction

History: A 55-year-old female patient was admitted to our movement disorder clinic due to gait difficulty that had started 3 weeks before. It was learned that the clinic had initiated as follow: The patient had fallen in the bath and hit her head on the floor. She was conscious at the time of the fall and there was no cut or laceration to the skin. However, she had been suffering from headache and tinnitus after the fall and therefore applied to the emergency service. The cranial computed tomography, performed at emergency service, showed communicated hydrocephalus. Therefore, the patient was informed about the appearance of chronic hydrocephalus and a neurology polyclinic admission was suggested. Interestingly, her relatives defined a deterioration in her gait soon after this briefing. Of note, the patient had been suffering from mild symptoms related to urge-type urinary incontinence over the past 10 years.

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Physical Examination: On neurological exam at admission, the patient was orientated and cooperative. However, her speech was evaluated as dysphonic, and the phenomenology of speech fluctuated during the interview. Her speech accelerated sometimes during the interview, whereas the meaningless repetition of some specific words compatible with palilalia was also observed at times. The sensory and motor examinations were within normal limits. However, she had a widebased gait with short strides and freezing episodes during turns were apparent. On the other hand, moderate fluctuations were also observed in her gait and movements throughout the day. The standardized mini-mental test score was 27 points. The cranial magnetic resonance imaging (MRI) showed communicated hydrocephalus (>Fig. 1). Some specific features such as the tight medial and high convexity subarachnoid space, narrowing of the callosal angle, and bulging of the lateral ventricular roof were also present suggesting an underlying normal pressure hydrocephalus (NPH).

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Management: At this point, the patient and her relatives were informed about the cerebrospinal fluid (CSF) tap test procedure, and its transient efficiency. Thereafter, the CSF tap test was performed (opening pressure 150 mm H2O, 40 cc CSF was drained) that yielded a marked improvement in the clinic. After the tap test, the patient could walk without support and her stride lengths had improved significantly. Besides, the patient stated that her thoughts were clearer, and feeling like a weight had been lifted from her brain. No improvement was defined in the urinary symptoms. Based on the dramatic response to the tap test, the patient was referred to the neurosurgery department. In the following course, the clinic of the patient deteriorated, and the tap tests (two more times with an interval of 2 days) were reperformed that did not provide significant and consistent improvements in the clinic this time. The patient was reevaluated in the movement disorders council and some clinical points were found atypical for NPH. First, the clinical presentation was very rapid for NPH. Second, some clinical features such as fluctuations of stance and gait, and excessive slowness of both movements and speech were considered to suggest a functional movement disorder (FMD). To confirm the possible FMD, a placebo effect was tested (150 mL intravenous physiological serum was administered) that also provided a dramatic improvement in the patient's gait and mobilization (►Videos 1-2). Taken together, the diagnosis of documented FMD¹ was established, and the hydrocephalus was evaluated as asymptomatic chronic hydrocephalus. A psychiatric evaluation yielded the diagnosis of depression and hypochondriasis. The Minnesota Multiphasic Personality Inventory was also performed that revealed abnormal results in the subscales of hypochondriasis, depression, and psychasthenia. Medical therapy (sertraline 50 mg) was initiated for depression and psychotherapy was initiated for hypochondriasis. In the third month of follow-up, the symptoms were completely resolved (>Video 3).

Video 1

The images of the neurological exam before placebo therapy showing wide-based gait and short-stepped gait. However, excessive slowness of gait and cautious manner were observed that were atypical for an organic gait disorder. Online content including video sequences viewable at: https://www.thieme-connect.com/products/ejournals/html/.

Video 2

The images after the placebo therapy showing marked improvement in gait features. Online content including video sequences viewable at: https://www.thieme-connect.com/products/ejournals/html/.

Video 3

The images of the neurological exam after 3 months of psychotherapy and medical therapy showing completely resolution of the gait disease. Online content including video sequences viewable at: https://www.thieme-connect.com/products/ejournals/html/.

Discussion

FMDs represent a challenging issue in clinical practice. The timely diagnosis of FMD is critical for avoiding unnecessary, and potentially harmful, interventions as well as initiate the appropriate treatment to provide optimal results. The most pronounced subtype of FMDs is the psychogenic tremor that may represent up to 55% of them, with the following types of psychogenic dystonia and psychogenic parkinsonism, respectively. Gait disorder is also a crucial subtype of FMDs and includes several subtypes such as astasia-abasia, pseudotabetic, pseudopolyneuritic, and tightrope walker.¹ To our knowledge, there is no published report describing a patient with FMD clinically mimicking NPH. The specific clinical features including momentary fluctuations of gait and stance, and excessive slowness raised the suspicion of psychogenic etiology. The dramatic response to placebo test and the complete resolution of symptoms with psychiatric treatments confirmed the diagnosis of FMD.

Communicated hydrocephalus is a condition where there is no structural lesion blocking the passages connecting the ventricles, and the CSF is able to leave the ventricular system. The most important subtype among the communicated hydrocephalus is NPH.³ NPH is a diagnosis of communicated ventriculomegaly that is supported by the clinical symptoms of gait difficulty, urinary problems, and cognitive difficulties in response to CSF drainage.4 However, there are many controversies regarding the diagnosis of this entity that has no definitive pathological findings. For instance, the differential diagnosis between NPH and hydrocephalus ex vacuo, which may both display hydrocephalus and brain atrophy on cranial MRI, may be extremely challenging.⁵ However, the hydrocephalus is rather secondary to encephalic volume loss in the latter, whereas the primary pathology is the emerging hydrocephalus and secondary neural effects leading to clinical symptomatology in NPH. Although there are some clinical clues, acute hydrocephalus developing due to secondary causes such as subarachnoid hemorrhage and meningitis may also mimic NPH. In this regard, another subtype of chronic hydrocephalus, called "longstanding overt ventriculomegaly," which emerges sub clinically in early childhood, but manifests in late adulthood, has contributed substantially to our understanding of the physiology of communicated hydrocephalus. More recently, a terminology of "Asymptomatic ventriculomegaly with features of idiopathic normal pressure hydrocephalus" was proposed to define a prodromal phase of NPH that further

Fig. 1 The cranial magnetic resonance imaging of the patient showing communicated hydrocephalus (A-C), tight medial and high convexity subarachnoid space (double arrows) (D), narrowing of the callosal angle (single arrow) (D), and bulging of the lateral ventricular roof

complicates the differential diagnosis.⁷ Such that, the appearance of NPH-like MRI findings was demonstrated to occur even 13 years prior to the clinical manifestation of NPH.⁸ In light of these observations, several authors remarked on the brain's ability to maintain normal mental status and function in spite of profound chronic hydrocephalus.⁹ Taken together, recognition of the emergence of associated clinical features constitutes critical points for the appropriate and timely diagnosis. The illustration of our patient with severe hydrocephalus on MRI and NPHlike clinic also draws attention to the diagnostic challenges in this entity. Interestingly, the patient had also benefitted from the CSF tap test initially that further complicated the diagnostic process. Although the neuroimaging clues are important, the appearance of a NPH clinic is essential for the diagnosis of NPH. However, a prodromal phase of NPH cannot be excluded in our case.

In conclusion, via the presentation of this case, we remark on the importance of a multimodal clinical approach, particularly the detailed clinical assessment, to establish the appropriate diagnosis and avoid unnecessary, potentially harmful interventions. The clinicians should keep in mind FMD as a crucial etiology that may mimic every subtype of neurological manifestations that was the NPH-mimic in our patient.

Authors' Contributions

H.O. was involved in conception, organization, execution of research project and writing of the first draft and review of the manuscript. A.C.U. helped in organization of the research project and review of the manuscript. S.C. contributed to conception of the research project and review of the manuscript.

Data Availability Statement

The authors confirm that the data supporting the findings of this study are available within the article and its supplementary materials.

Ethical Approval

The authors confirm that the approval of an institutional review board was not required for this report. A written informed consent was obtained from study participants. We confirm that we have read the journal's position on issues involved in ethical publication and affirm that this work is consistent with those guidelines.

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Conflict of Interest None declared.

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