Acta Scientific Neurology (ISSN: 2582-1121)

Volume 6 Issue 4 April 2023

Case Report

Normal Pressure Hydrocephalus in a Known Multiple Sclerosis Patient: A Rare but Considerable Comorbidity

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Received: December 15, 2022

Published: March 09, 2023

Abstract

Multiple sclerosis (MS) is a common chronic neurological disease with pathophysiology consisting of both chronic neuroinflammatory and degenerative process. MS affects people mostly from young age with disabling neurological deficits like impairment memory, gait, psychomotor retardation, and sphincter incontinence. All mentioned symptoms also present as Hakim's triad in another well-known neurologic disease, i.e., normal pressure hydrocephalus (NPH).

Here, we present a rare case of communicating hydrocephalus in a relapsing-remitting MS patient. The patient admitted with a complaint of gait imbalance and memory problems, which could not be attributed to new MS attack and rather showed a good response to shunting surgery. Further, we focused on concurrency of two diseases NPH and MS in the current literature.

Keywords: Normal Pressure Hydrocephalus; Multiple Sclerosis; Shunt-Responsive Hydrocephalous

Introduction

Today, multiple sclerosis (MS) is known as a chronic and disabling demyelinating disease that involves different parts of the central nervous system (CNS) [1]. The neuroinflammatory processes in the pathophysiology of MS dominate the research field and also the treatment approaches [2]. Although immunomodulatory drugs may reduce the severity and number of the MS attacks, they could not effectively reduce the long-term progressive and neurodegenerative features of the disease [3]. This disease may present with various manifestation and mimic many syndromes. The Hakim's Triad described in normal pressure hydrocephalus (NPH) cases including gait problems, urinary dysfunction, and/or dementia could be seen in long-standing MS. It is of note that such symptoms plus non-obstructive hydrocephaly have been reported by various degrees of severity in MS cases. Short steps with apraxia of gait, cognitive problems, psychomotor retardation, impairment of executive memory, and urinary urgency and/or incontinence could be seen in several cases of chronic MS [4,5]. Furthermore, MS patients could be involved by other diseases as comorbidities. Here, we presented a rare disorder of shunt-responsive hydrocephalous in an MS patient and also narrative review of the current literature focusing on concurrency of two conditions, normal pressure hydrocephalus, and MS in a patient.

Case Report

A 33-year-old female was admitted to our hospital. She was a known case of relapsing-remitting MS (RRMS) in the last 15 years and was on Fingolimod as a disease-modifying drug from two years ago. She had not developed any clinical relapse within the last 2 years but there were some steady progressive complaints such as fatigue, gait problems, falling attacks, impaired memory, and headaches. The neurological examination revealed quadriparesis (3-4/5, which was more prominent in lower extremities muscle strength), diffuse hyperreflexia, normal sensory examination ex-

cept impaired position sense in legs, and also apraxic gait. In her recent imaging, there was a non-obstructive hydrocephalous without prominent brain atrophy or the presence of enhanced active plaques (Figure 1).

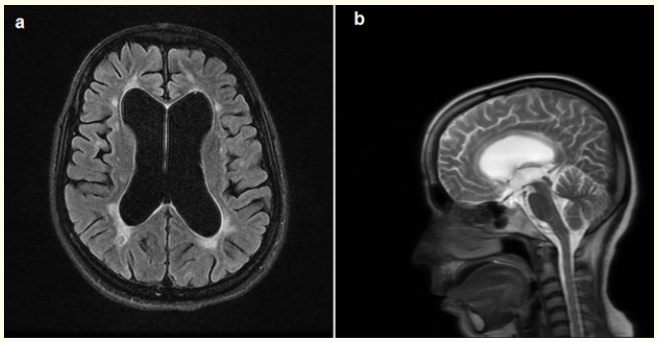

Figure 1: a: Axial FLAIR- T2-weighted MRI and b: Sagittal T2-weighted MRI image showing widening of brain ventricles, disproportionate to brain atrophy.

Regarding clinical examination and imaging findings, CSF study and Fisher tap test were planned. The opening CSF pressure and analysis were within normal limits. The walking time and mini-mental test were recorded (Table 1 and Video 1), at baseline, 30 min, and 24h after CSF tap test. Based on the significant improved of gait and cognitive function following the tap test, ventriculoperitoneal shunting surgery was planned for the patient.

	Before test	30 minutes after tapping	24 hours after tapping
MMSE	21	21	24
Walking time(second)	45	42	27

Table 1: MMSE (Mini Mental State Examination) and gait improvement after fisher's Tap test, distance of 40 meters.

In subsequent examinations and follow-ups, gait and memory problems were improved notably without any major complications related to shunting surgery.

Discussion

As described, our patient was an RRMS case with progressive and disabling walking and memory problems without any obvious clinical attack. Magnetic resonance imaging (MRI) study showed non-obstructive hydrocephalous, which has a significant response to VP shunting. To our knowledge, there are only two reports of concordance of NPH in chronic MS cases. O'Brien., *et al*. (1993) described two MS cases with Hakim's triad. In this study, neuroimaging was compatible with NPH in both but fisher tap test was positive in one of them. Both of them showed considerable improvement of symptoms after shunting surgery [6]. Another case of shunt-responsive hydrocephalous with detailed MR imaging and CSF flow and velocity data compatible to NPH cases was ascribed by Algin., *et al.* [7]. They described a 28-years-old female, known case of MS from 8 years ago, who admitted with a complaint of paraparesis, headache, and loss of appetite, nausea, vomiting, gait apraxia, and urinary incontinence. All the symptoms started from 10 days before admission day. Presence of a new attack was considered, and she received 1g/day of methylprednisolone for 5 days. All the symptoms presented after pulse therapy except gait disturbance and urinary problems. Shunting surgery was planned based on neuroimaging properties and normal opening pressure and CSF study. As a result, remnant symptoms were fully resolved with no surgical complications. Clinical and imaging features of NPH have also been reported in other chronic CNS inflammatory conditions such as neurosyphilis [8,9], systemic vasculitis with CNS involvement such as rheumatoid arthritis [10], systemic lupus erythematosus [11,12], and Wegner disease [13].

NPH cases are divided into two etiological subgroups described as primary (idiopathic) and secondary NPH. The prevalence of idiopathic NPH is more frequent in old ages. There has been shown that the disease is 10 times more in 70-90-years old people compared to previous decades. In comparison, the prevalence of secondary NPH has been more common in younger ages [14]. Pulse wave encephalopathy has been proposed according to recent studies to be a pathophysiology MS [15], a mechanism that has been considered in NPH. This may partially describe the concordance of two diseases is pulse-wave encephalopathy. This theory has been suggested as a causal mechanism that leads to dementia in neurodegenerative diseases such as Alzheimer's disease, vascular dementia, and normal pressure hydrocephalus [3,16]. Juurlink., *et al*. claimed that intracranial compliance may decline with an increase in CSF volume and velocity of flow through the cerebral aqueduct in MS patients [3]. Moreover, it has been shown that there is a pulse wave alteration of CSF flow within the craniospinal cavity in MS

patient, similar to the findings in NPH. In MS patients, these changes include 35% reduction in arteriovenous delay, 5% reduction in the arterial inflow returning via the sagittal sinus, 26% increase in arterial stroke volume, and 30% reduction in sagittal and straight sinus stroke volume. The same changes in arteriovenous delay and blood have been reported in NPH patients [15]. Other common CSF characteristics suggested in both NPH and MS cases in different studies include increased ventricular [17] and Virchow-Robin space size [18], with reduced net flow of CSF through the same structure and decreased CSF pulsation at C2 level [19].

Another considerable aspect that links MS and NPH is the presence of some neuro-inflammatory process in pathophysiology of both diseases. Czubowicz., *et al*. also reported that the concentration of IL-6 and IL-8 in NPH patients were significantly higher compared to patients with arrested hydrocephalus or excavated hydrocephalous related to brain atrophy. All the mentioned cytokines also played major roles in immune-modulating features of MS [20].

Conclusion

In conclusion, comorbidities, especially with treatable disorders, should be considered in MS patients. In this regard, normal pressure hydrocephalus (NPH) may be a comorbid condition in patients with gait disorder, forgetfulness, and urinary incontinence, which are the cardinal manifestations in long-standing multiple sclerosis.

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