Spontaneous cerebrospinal fluid leakage in a patient with normal pressure hydrocephalus: "a miracle"

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Introduction

Normal Pressure Hydrocephalus (NPH) is one of the causes of potentially treatable dementia and presents with characteristic triad of gait apraxia, cognitive impairment \RL and urinary incontinence. Typically, it is associated with normal cerebrospinal fluid (CSF) pressure and ventriculomegaly that is disproportionate to amount of cerebral atrophy. Relkin et al. proposed the diagnostic criteria for idiopathic NPH (iNPH) which was classified into probable, possible, and unlikely categories (1).

There is a transient improvement in symptoms after a large-volume lumbar tap or a short-term lumbar drain. In clinical point of view, this technique is applied for prediction of patient who may respond to shunt surgery. Furthermore, serial lumbar tap may be as a therapeutic option for patients with NPH who are not good candidate for surgery.

CSF leak have been classified into traumatic and spontaneous (2). Spontaneous (non traumatic) CSF leak is an uncommon, but well documented presenting feature of a wide variety of intracranial pathologies including hydrocephalus, tumor, increased intracranial pressure, congenital anomalies or unknown causes (2, 3). Rarely, it may be a presenting feature of NPH, which was reported previously in the context of aqueductal stenosis (4, 5).

Here, we report an adult patient with NPH suffering from dementia and gait disturbance who developed spontaneous CSF leakage from his nose that led to dramatic improvement in gait and cognitive function. To our best knowledge, it is the first report of spontaneous CSF rhinorrhea in patients with NPH.

Case History/examination:

On 22 December 2019, a 79 years-old Iranian gentleman was referred to our hospital for diagnostic lumbar tap. He was a building painter and had an educational level of 6 years Primary School. The symptoms were begun since one year ago, primarily urinary incontinence and then gait disturbance. In addition, his family members noticed gradual decline in cognitive function, in the form of repeated questions and statements, word finding difficulty and time and place disorientation for recent several months. There was also some history of visual hallucination and rapid eye movement (REM) sleep behavior disorder. The past medical history was positive for diabetes mellitus, hypertension and cardiac arrhythmia that underwent permanent pacemaker. His medications included rivaroxaban, insulin, bisoprolol, valsartan, simvastatin, clonazepam and duloxetine. The physical examination revealed positive Myerson sign, absent ankle reflex, stocks and gloves hypoesthesia, and mild tremor in left hand. There was no rigidity, bradykinesia, ideomotor and ideational apraxia, and grasp and snout reflexes. Standing posture was slightly stooped with prominent postural instability. The walking was laborious needs help and consisted of wide based gait, very short steps and shuffling, in favor of frontal lobe gait. Neuropsychological assessment showed impairment in memory, attention and visuospatial domains. The total score of Mini-Mental Status Exam (MMSE) and of Montreal Cognitive Assessment (MoCA) were 19 and 14 out of 30, respectively.

Methods:

According to clinical features, assessment for the cause of cognitive decline and gait difficulty was performed. The brain CT scan showed significant ventriculomegaly (i.e. Evans index of 0.37) including third and fourth ventricles which was disproportionate to the amount of cortical atrophy (Figure 1). The other laboratory tests were in normal range including complete blood count, thyroid function tests, HIV antibody titer and vitamin B12 level. The clinical and imaging features were in favor of probable iNPH. Consequently, lumbar tap was performed two times that revealed significant change in gait after removal of approximately 30 ml of CSF. After the first trial of tap test, he can walk independently, which was more pronounced after the second trial. The speed and cadence of gait after first and second trial of tap test were noted in Table 1. The analyses of CSF markers were in normal range but the opening pressure was slightly elevated in two times of four lumbar puncture (Table 1).

Conclusion and Results:

Due to dramatic response to lumbar tap test, the patient would be a good candidate of surgery for ventriculoperitoneal or ventriculoatrial shunt insertion. However, we refuse the referral of patient for shunt insertion after considering prominent comorbidity and negotiation with family members. In addition, the significant improvement after removal of CSF helped us to give the decision of serial lumbar tap as the subsequent treatment procedure. The third lumbar tap performed on 21 January 2020. The family members reported significant improvement in daily activities after each episode of lumbar tap. At February 2020, we encounter with the emergence of COVID-19 pandemic. Consequently, the serial lumber taps were stopped temporarily. Although the gait deteriorated gradually again, but the situation was not ideal for performing new lumbar tap.

On the last days of March 2020, an interesting event occurred. The patient had several sudden sneezing which result in spontaneous CSF rhinorrhea with the amount of approximately 30 to 40 ml CSF. With respect to the reports of family members, this event led to significant improvement in gait and cognition, described as a "miracle" and lasted for about 3 months. Afterwards, the symptoms were returned gradually that he could not walk independently.

Discussion

Normal pressure hydrocephalus is a cause of potentially reversible dementia. In 2019, an old man was referred to our hospital with classic triad of NPH. Serial CSF removals led to dramatic improvement in symptoms. The emergence of COVID-19 pandemic has stopped the planned treatment program. Meanwhile, a "miracle" event of spontaneous CSF leakage occurred which led to gradual improvement in cognitive and gait profiles.

Cerebrospinal fluid leakage have been classified into traumatic and spontaneous by Ommaya et al at 1968 (2). The "spontaneous" is applied to indicate that the leakage is not an immediate or delayed result of trauma. Interestingly, their article had other amassing features including a comprehensive diagram contains subdivisions for nontraumatic CSF rhinorrhea (2). In this regard, nontraumatic CSF leakage can be categorized into high-pressure and normal-pressure leaks. Hydrocephalus, either obstructive or communicating, was classified as a type of high-pressure CSF leakage (2). In 1978 , Oblu classified the different causes of nontraumatic CSF leak into four groups: congenital, tumoral, infectious and vascular. Hydrocephalus may be classified under the group of congenital (i.e. congenital stenosis of aqueduct of Sylvius) or tumoral (i.e. indirect effect of tumor by providing intracranial hypertension) (3).

In the published literatures, several cases with sudden leakage of CSF have been reported. In most of these cases, there is an evidence of raised intracranial pressure. For instance, there are some reports of choroid plexus papilloma (6, 7), colloid cyst of third ventricle (8), and Idiopathic Intracranial Hypertension (IIH) (9). Yang et al. collected 21 patients with spontaneous CSF rhinorrhea whose mean preoperative CSF pressure was 17.6 cmH2O. Surprisingly, all of the patients had increased postoperative ICP with mean ICP of 25.5 cmH2O (range from 21 to 32 cmH2O) (9). Schlosser et al. had performed postoperative lumbar puncture in ten patients with spontaneous CSF leak that all of them had elevated ICP (mean: 26.5 cmH2O) (10).

In the same way, the CSF pressure was slightly elevated in our patient. To our knowledge, it is the first case of NPH reported with spontaneous CSF leakage. Interestingly, this event was associated with marked improvement in clinical features. There are some reports of spontaneous CSF leakage secondary to aqueductal stenosis (4, 5). Satyarthee reported a 16-yers old girl with headache, vomiting and chronic CSF rhinorrhea. She had an obstructive hydrocephalus due to aqueductal stenosis and underwent a ventriculo-peritoneal shunt surgery, but the CSF rhinorrhea persisted despite functional shunt and need to surgical repair (5). In contrast, Muzumdar et al. reported a 30-year-old male with 9-monthd history of CSF rhinorrhea as a result of aqueductal stenosis who had favorable response to shunting (4). The authors claimed that the fragile anatomy of cribriform plate and daily variations of CSF pressure might be a contributing factor for spontaneous CSF rhinorrhea in a patient with aqueductal stenosis. Alongside, any causes of transient rising of intracranial pressure (ICP) may open forcibly an occult anatomical dural defect and led to CSF leak (4). Sudden sneezing, coughing or Valsalva maneuver may have a similar mechanism for CSF leak. As the same way, our patient reported several times of sneezing prior to emergence of CSF rhinorrhea. Beside of these physiologic factors, we assume that daily variations of intracranial pressure may also a contributing factor in transient increment of ICP that consequently lead to spontaneous CSF leak. This was evident in two out of four times of measuring CSF opening pressure in our patient.

The interesting point of this case was the marked improvement of gait and cognition after spontaneous CSF leakage. It has been reported that the symptoms of IIH may improve after spontaneous CSF leakage. In the study of Yang et al., 18 out of 21 patients had presented with symptoms of IIH that disappeared after occurrence of spontaneous CSF rhinorrhea (9). In this clinical feature, the possible mechanism is sudden decrement of CSF pressure. Weather this mechanism can also explain the improvement of gait and cognition in NPH, is not clear.

Non-traumatic spontaneous CSF leakage may occur in the context of high or normal intracranial pressure. We report a patient suffered from NPH who had spontaneous CSF rhinorrhea that led to marked improvement in gait and cognitive function.

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Author contribution:

MAD: diagnosis and management, conception of study idea, manuscript clarification and submission

SF: manuscript template writing, conception of study idea

TK: MAD: manuscript clarification and submission

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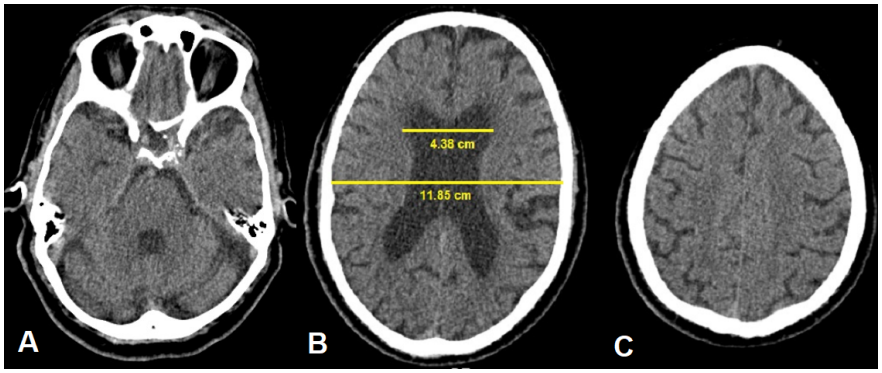
Table 1: The characteristics of gait indices, MoCA score and CSF markers before and after four trials of lumbar tap.

Figure 1: Brain CT scan, axial view, revealed increased the size of 4th (A), 3rd (not shown) and lateral ventricles (B) with the Evans index of 0.37 (B). The ventriculomegaly is disproportionate to the amount of cortical atrophy (C). These findings are compatible with the diagnosis of NPH.



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