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Case Report

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Idiopathic Normal Pressure Hydrocephalus Presenting with Oculomotor Palsy as a Predominant Symptom: A Case Presentation

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Abstract

Though symptoms in patients with idiopathic normal pressure hydrocephalus are well characterised, there are a lot of case reports presenting other predominal symptoms, reinforcing the complexity and the multi-dimensional "universe" in brain networks. We are presenting an unusual case presentation of an adult patient who fulfilled the radiogical criteria for iNPH and suffered from paroxysmal unilateral oculomotor palsy as a cardinal symptom.

Keywords: Idiopathic Normal Pressure Hydrocephalus; Oculomotor Palsy; Water-Hammer Effect; Third Nerve; Magnetic Resonance Imaging

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Introduction

Idiopathic normal pressure hydrocephalus (iNPH) is a disorder characterized by gait and balance disturbance, cognitive dysfunction, and urinary symptoms caused by an impaired turnover of the cerebrospinal fluid (CSF) [1]. The diagnosis of possible iNPH is based on clinical symptoms, brain imaging, and altered CSF dynamics according to the International guidelines from 2005 and the second edition of the Japanese guidelines from 2012 [2,3]. Brain imaging in typical cases shows enlarged ventricular system with an Evans' index more than 0.3, enlarged Sylvian fissures, tight medial and high convexity sulci, a callosal angle between <50° and >90° and disproportionately enlarged subarachnoid-space hydrocephalus (DESH) [4-6]. Although the disease is characterized by the classic triad, many other symptoms have been described in patients with possible iNPH such as epilepsia [7], amyotrophic lateral sclerosis [8], neuropsychiatric symptoms [9], parkinsonian symptoms [10], depression, olfactory palsy [11] and schizophrenia [12]. Cultrera F, et al. (2009) reported an unusual case of a 16-year-old boy with paroxysmal unilateral oculomotor nerve paralysis associated with ventriculomegaly and Dandy-Walker variant cystic malformation which went to totally regress after a ventriculo-peritoneal shunt operation [13]. Herein, we report a case of an adult patient with possible iNPH who presented with oculomotor palsy as a first symptom.

Case Report

A 51-year-old Caucasian woman with hypertension, single kidney after kidney tumor operation at the age of 23 years and repeated pneumothorax because of unspecified lung emphysema for which she operated at the age of 24 proceeded to the Emergency Room (ER), after have woken up with right-sided headache (visual analog scale 5) and right monocular blurred vision. Neither trauma nor infection was mentioned. The patient described that the night before when she went to sleep felt as usual and she woke up with the above named symptoms. The patient's respiration rate was stable. The blood pressure was 154/88 mm Hg and the electrocardiogram (ECG) showed sinus rhythm with some premature ventricular extrasystoles (VES). The neurological assessment showed anisocoria with right-sided mydriasis with poor papillary reflex, inability to move the right eye inward and absence of ptosis. According to National Institutes of Health Stroke Scale the patient had 0 points. Blood samples with C - reactive protein, erythrocyte sedimentation rate, international normalized ratio, creatinine and liver status were normal. The blood glucose was 5.2 (normal 4.2-6.0 mmol/L), thyroid-stimulating hormone 1.8 (0.3-4.2 mIE/L) and cholesterol 3.2 (3.9-7.8 mmol/L). An acute brain computer tomography (CT) showed neither infarction nor bleeding but obvious ventriculomegaly disproportionate to cerebral atrophy, an Evans index of 0.35, corpus callosum thinning and elevation with a callosal angle of 80°, widening of the temporal horns (without hippocampal atrophy) and widening of the third ventricle to 13 mm, narrowing of the sulci and subarachnoid spaces over the high convexity and midline surface of the brain, and ballooning of the frontal horns (Figure 1). A per oral medication with clopidogrel 75 mg once a day was started as transient ischemic attack couldn't be ruled out as a possible diagnosis.

The patient was admitted in the neurological department for further monitoring, observation and investigation.

Ophthalmological consultation ensured the diagnosis of oculomotor palsy without any ocular aetiology. An acute duplex of carotid and vertebral arteries was without arteriosclerosis or dissection. A brain magnetic resonance image (MRI) showed no infarction, tumor or dissection. Regarding the ventricular system demonstrated an aqueductal flow void (Figure 2). There was no microangiopathy in the periventricular white matter. Radiological imaging fulfilled the criteria for iNPH.

A 24-hour ambulatory blood pressure monitoring showed normal levels. Telemetry showed episodes of ventricular extrasystoles. A second ECG showed slight ST-T changes inferiorly and laterally which were unchanged compared to a previous ECG (Figure 3).

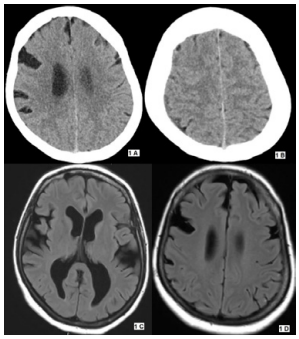

Figure 1: In figures 1A and 1B, the brain CT shows narrowing of the sulci and subarachnoid spaces over the high convexity and midline surface of the brain. In figures1C and 1D the brain MRI shows neither infarction nor bleeding but obvious ventriculomegaly disproportionate to cerebral atrophy, an Evans index of 0.35.

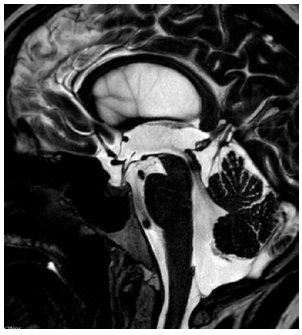

Figure 2: Brain MRI with T2 sequences demonstrated an aqueductal flow void.

After consultation with a cardiologist, a heart ultrasound showed a normal left ventricle with normal systolic and diastolic function and slight septal hypokinesis. Furthermore, coronary CT angiography was without arteriosclerosis or stenosis. An exercise ECG showed reduced physical working capacity, the patient managed to cycle only 61% of expected for her age and interrupted because of muscle fatigue, without any angina or ST depressions. During the rest time, the patient expanded pronounced ventricular arrhythmia with recurrent VES. The patient dismissed from the clinic with planned follow-up.

Four months after the discharge, the patient presented to our outpatient department for iNPH investigation. Despite the obvious radiological picture which pointed for iNPH the patient missed the clinical traid for probable iNPH diagnosis. The gait parameters evaluated by our physiotherapist (JR) and the patient needed 8.5 seconds to complete a Timed Up and Go Test (TUG), 12 steps for TUG step-test, 9 seconds for the 10-metre walking time test and 15 steps for the 10-metre walking step test. The cognitive evaluation by the iNPH team's occupational therapist (KO) showed normal results on a Mini Mental Statement Test (MMSE 30/30) without any cognitive impairment. A lumbar puncture (LP) was performed with the patient placed in the lateral decubitus position and the opening pressure was 23 cm H20. We noticed no pleocytosis, intrathecal immunoglobulin (Ig) G synthesis was lacking, IgG and IgM for borrelia burgdorferi were negative. The neurodegenerative markers showed neurophilament at 500 ng/L (normal <890 ng/L), glial fibrillary acidic protein at 270 ng/L (normal <750 ng/L), tau at 217 (normal <479 ng/L), phosphorylate tau at 14 ng/L (normal <61 ng/L), β-amyloid 42/40 at 0.87 ng/L (normal >0.61 ng/L) but the β-amyloid 42 was low at 268 ng/L (normal >620 ng/L).

Despite the obvious radiological imaging the iNPH team decided to wait with shunt operation for this patient and to plan a follow-up every sixth month.

Discussion

Oculomotor palsy is a neurological condition characterized by the sudden onset of binocular horizontal, vertical, or oblique diplopia and a droopy eyelid and/or enlarged pupil. Pain accompanying the onset of third nerve palsy is common [14].

The third nerve nucleus is located in the midbrain. The parasympathetic pupil nucleus (Edinger-Westphal nucleus) controls the pupil constriction. Both of them innervate the muscles which adduct, depress, and elevate the eye. There are many possible triggers of oculomotor palsy. Through the anamnesis, there was no trauma in patient's history. Ophthalmological migraine was considered

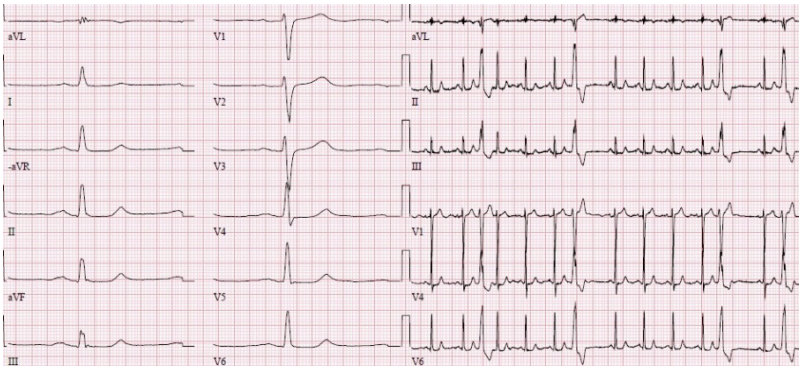

Figure 3: ECG shows slight ST-T changes inferiorly and laterally with some premature ventricular extrasystoles.

as a possible cause but the brain MRI showed lack of gadolinium enhancement to the cisternal segment of the right third nerve [15]. Diffusion-weighted imaging (DWI) together with qualitative and quantitative assessment of the apparent diffusion coefficient (ADC) are particularly sensitive to detection of acute ischemic stroke within a few minutes after arterial occlusion and to differentiation of acute stroke from other processes that manifest with sudden neurologic deficits [16,17]. In our case both sequences were normal and ischemia was excluded as a cause for the oculomotor palsy. The susceptibility weighted imaging (SWAN) is an MRI sequence sensitive to visualize parenchyma changes caused by blood products. This aspect was not investigated in our patient. Nonetheless, by using time of flight angiography (TOF) sequences we can visualize blood flow within vessels, without the administration of contrast agents. In our case we couldn't notice any aneurysmatic change. By using the duplex technique, we achieved to exclude the extracranial dissection as a probable reason of oculomotor palsy.

The ophthalmological investigation was without retinal diseases and no papilledema could be seen. The blood controls could exclude diabetes, hypothyreosis, infection and inflammation. The LP was with lack of pleocytosis, no IgG band and all viruses and bacterial proves were normal. Above named couldn't lead us to a concrete cause of the acute oculomotor palsy.

A theory which could explain patient's oculomotor palsy could be the water-hammer effect by the impairment of the CSF flow. The nucleus of third nerve is placed near the aqueduct and altered CSF dynamics could lead to the paroxysmal dysfunction of the nucleus. The lumbar CSF pressure was at 23 cm H20 but elevated abrupt to 27 cm H20 when we asked the patient to cough or to tighten the abdominal muscles. Maybe when the patient investigated from our department was the turning point for obstructive hydrocephalus. Nevertheless, retinal control with optical coherence tomography and direct ophthalmoscopy couldn't show any pressure-related findings [18]. The radiological approach was compatible to iNPH. In iNPH many CSF biomarkers have been analyzed but the reduced Aβ42 with concomitant normal or reduced t-tau and p-tau levels is common finding [19]. In our case the patient had a lowered β-amyloid. After the iNPH investigation the patient was not found eligible for a shunt operation at that moment. The spontaneous regress of oculomotor palsy remains an unanswered question.

Conclusion

Although iNPH is a disease with well characterized symptoms, in this rare case iNPH should be considered as a probable cause of oculomotor palsy, and this is an issue that merits further research.

Declarations

List of Abbreviations Used

iNPH: Idiopathic normal pressure hydrocephalus

CSF: Cerebrospinal Fluid

ER: Emergency Room

CT: Computer Tomography

MR: Magnetic Resonance Imaging

ECG: Electrocardiography

Authors' Contributions

Andreas Eleftheriou was the neurologist who performed the clinical and neurological evaluation. Andreas Eleftheriou was the major contributor in writing the manuscript. Martin Nilsson was the neurosurgeon who performed the neurosurgical approach of our case.

Conflicts of Interest

The authors have no conflicts of interest to disclose.

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