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Case Report

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An atypical presentation of normal pressure hydrocephalus: a detailed case study of Hakim-Adams syndrome in a young adult

Ayşe Şule Akan

Department of Emergency Medicine, Kayseri City Hospital, Kayseri, Turkiye

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Corresponding Author: Ayşe Şule Akan, draysesuleakan@gmail.com

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ABSTRACT

Normal pressure hydrocephalus (NPH), historically termed Hakim-Adams syndrome, is predominantly diagnosed in older adults. This case report details an atypical presentation in a 32-year-old male, underscoring the importance of differential diagnosis in younger patients exhibiting NPH-like symptoms. The patient presented with dizziness and mild ataxia, without the classical triad of gait disturbance, dementia, and urinary incontinence typically associated with NPH. Computed tomography (CT) and magnetic resonance imaging (MRI) revealed ventricular dilation and a suspected fourth ventricle lesion, leading to a provisional diagnosis of Hakim-Adams syndrome. This report highlights the need for heightened clinical vigilance and comprehensive imaging in atypical presentations of NPH.

Keywords: Ataxia, dizziness, Hakim-Adams syndrome, normal pressure hydrocephalus, young adult

INTRODUCTION

Normal pressure hydrocephalus (NPH), first defined in 1965 by Hakim and Adams, is a form of chronic hydrocephalus characterized by ventriculomegaly without increased cerebrospinal fluid (CSF) pressure.[1] The condition is predominantly observed in the elderly and is marked by a symptom triad of gait disturbance, cognitive impairment, and urinary incontinence.[2] NPH is categorized into idiopathic NPH, which arises spontaneously, and secondary NPH, often following trauma or hemorrhage.[3] This case report illustrates an uncommon presentation of NPH in a young patient, challenging the conventional demographic and symptomatic parameters typically associated with the syndrome.

CASE

A 32-year-old male with no significant medical history presented to the emergency department with complaints of dizziness. Upon examination, the patient was alert, oriented, and cooperative. Vital signs were within normal limits, except for mild ataxia observed during physical examination. Blood tests and CT imaging of the brain were conducted. The CT scan indicated dilation of the third and lateral ventricles, along with a possible space-occupying lesion at the level of the fourth ventricle. An MRI was performed for further evaluation, which confirmed tetra ventricular dilation and raised suspicion of Hakim-Adams Syndrome (Figure 1). Consultations with neurosurgery and ophthalmology were sought for papilledema and optic nerve assessment. The patient was admitted to neurosurgery for further management and potential surgical intervention.

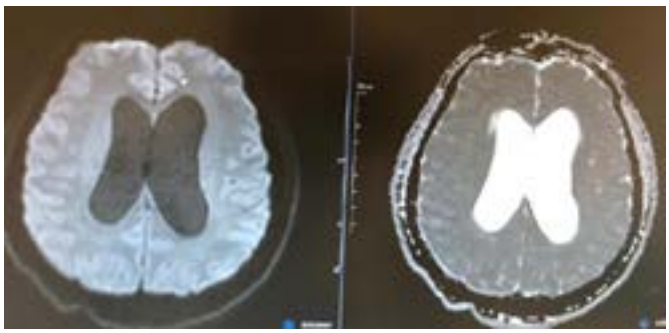

Figure 1. The patient's brain MRI axial image

DISCUSSION

NPH is typically a condition of the elderly, with the first sign often being gait disturbances.[2] In contrast, our patient's initial symptom was dizziness, a less common presentation that paralleled Shamov's findings of ataxia and apraxia as early symptoms of NPH.[4] The pathophysiological mechanisms behind the ventricular dilation characteristic of NPH remain under investigation.[5]

The increasing prevalence of NPH in older populations, and its potential co-occurrence with psychiatric disorders, further complicates the diagnostic process.[6,7] The presentation of NPH in a younger adult, as in our case, emphasizes the need for a comprehensive diagnostic approach, even in atypical patient demographics.

Recent studies have delved into the underlying pathophysiology of NPH. One theory suggests that

altered CSF dynamics, particularly impaired absorption at the arachnoid granulations, play a central role in its development.[2] The condition's progression is insidious, often leading to misdiagnosis or late diagnosis, especially in cases where classic symptoms are absent or subtle.

The diagnostic challenge is further compounded by the similarity of NPH symptoms to other neurodegenerative conditions, particularly in the elderly. Distinguishing NPH from diseases like Alzheimer's or Parkinson's requires careful clinical evaluation and often, reliance on imaging studies. CT and MRI are pivotal in diagnosing NPH, with MRI providing more detailed information on ventricular size and morphology, as well as the identification of potential comorbid conditions.[8]

Treatment options for NPH typically involve surgical intervention, most commonly ventriculoperitoneal shunting, which has shown varying degrees of success in alleviating symptoms.[9] The decision to operate is based on a careful evaluation of potential benefits versus risks, particularly in younger patients where the presentation and progression of the disease may differ from the elderly population.

CONCLUSION

This case underscores the complexity of diagnosing NPH in younger individuals, particularly when the presentation deviates from the classic symptom triad. Early recognition and appropriate management of NPH, even in atypical cases, can significantly alter patient outcomes. This report contributes to the growing understanding of NPH as a condition with a broader clinical spectrum than previously recognized and highlights the importance of clinical vigilance and comprehensive imaging in its diagnosis.

ETHICAL DECLARATIONS

Informed Consent

The patient signed the free and informed consent form.

Referee Evaluation Process

Externally peer-reviewed.

Conflict of Interest Statement

The author has no conflicts of interest to declare.

Financial Disclosure

The author declared that this study has received no financial support.

Author Contributions

The author declared that she has approved the final version.

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