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Obstructive Hydrocephalus due to Aqueductal Stenosis (web) Presenting with Cerebellar Ataxia: A Case Report and Review of the Literature

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Abstract

Obstructive hydrocephalus is a condition marked by the accumulation of Cerebrospinal Fluid (CSF) in the brains ventricular system, which leads to increased intracranial pressure. Aqueductal stenosis, a rare cause of obstructive hydrocephalus, involves the narrowing of the aqueduct of Sylvius and may result from congenital malformations, tumors, or inflammatory processes. This condition can present with various symptoms, such as headache, nausea, vomiting, and cognitive impairment. However, presentations can sometimes be subtle, as with cerebellar ataxia. In this case report, we discuss a patient with obstructive hydrocephalus due to aqueductal stenosis who exhibited cerebellar ataxia, a neurological disorder characterized by coordination and balance difficulties. The patient's presentation included abnormal body movements affecting the right side of her upper and lower extremities. MRI findings disclosed a linear band of tissue (web) at the aqueduct of Sylvius's inferior part, with funneling at the superior part leading to severe ventricular dilation and an associated transependymal leak. This case underscores the necessity of considering obstructive hydrocephalus in the differential diagnosis of cerebellar ataxia and the critical nature of early intervention and meticulous monitoring. Furthermore, it highlights the need for a multidisciplinary approach to management.

Keywords: Aqueductal stenosis; Cerebellar ataxia; Endoscopic third ventriculostomy; Hydrocephalus

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Introduction

Aqueductal Stenosis (AS) is a condition responsible for inappropriate enlargement of the lateral and third ventricles due to abnormal Cerebrospinal Fluid (CSF) accumulation, accounting for 20% of hydrocephalus cases [1]. Hydrocephalus attributable to AS occurs in 6%–66% of children and 5% to 49% of adults, with peak incidence in adolescence and the first year of life [2]. Idiopathic Aqueductal Stenosis (IAS), where the cause remains unclear, is seen in approximately 75% of patients [3]. The remaining cases are linked to factors such as genetics, infections (bacterial and viral), hemorrhage (intraventricular hemorrhage in prematurity, subarachnoid hemorrhage), and central nervous system malformations [4]. Aqueductal stenosis is not considered stable and is often long-tolerated. Various theories have been proposed to account for this. Xian Z et al. [5] suggest that head injuries, subarachnoid hemorrhages, or viral infections can worsen occlusion over time. They also posited that functional processes could complete partial stenosis: fluid buildup in the supratentorial ventricular system may cause brain stem deformation and aqueduct kinking, thus aggravating stenosis in a self-perpetuating cycle [6]. Subjective symptoms in acute or subacute stages may include episodic or continuous headaches, vomiting (especially with posterior fossa tumors), and decreased consciousness levels. In compensated stages, morning exacerbation of headache and nausea, diplopia from abducens nerve palsy in intracranial hypertension syndrome, dizziness, and somnolence may occur. Surgical management for AS focuses on [7-9].

Addressing the root cause: removing etiologic tumors, implementing a ventriculo-peritoneal

shunt with a valve, and dilating the prestenotic aqueduct in patients with obstructive triventricular hydrocephalus (ETV is considered the optimal surgical method for obstructive hydrocephalus due to AS). Aqueductal Plasty (AP) may be performed alone or in addition to third ventriculostomy, with or without a silastic stent. However, it carries significant risks of serious complications such as diplopia, dysconjugate eye movement, and trochlear palsy due to trauma to the dorsal tectal plate and ventral midbrain tegmentum. Retrograde aqueductoplasty with endoscopic trans-fourth ventricle approach is considered especially in cases of supratentorial slit ventricles.

Case Presentation

A 14-year-old female patient presented to the University of Gondar Hospital with complaints of Abnormal Body Movement (ABM) affecting the right side of her upper and lower extremities. She described the ABM as rhythmic tonic contractions followed by jerky releases. She was conscious and could recall these episodes. Four months prior to her current presentation, the patient experienced non-bloody, non-bilious, and non- projectile vomiting. Behavioral changes such as irritability, emotional lability, and forgetfulness were also reported. Preceding these symptoms, she suffered from intermittent frontal headaches that were position- dependent and exacerbated by bending down, coughing, and straining. The headaches, severe enough to disrupt her education and daily activities, had been occurring for a year before her presentation. Additionally, she reported urinary incontinence but no increased frequency, and fecal incontinence. Pertinent negative histories include no trauma, chronic medical conditions, or drug abuse. Physical examination showed a communicative patient with a blood pressure of 120/70 mmHg, a pulse rate of 84 beats per minute, a respiratory rate of 20 breaths per minute, a temperature of 36.6 degrees Celsius, and an oxygen saturation of 96%. The examination was normal except for the neurological findings.

Her Glasgow Coma Scale (GCS) score was 15/15. She was oriented, but her gait was significantly affected and staggering (as seen in the video). Rapidly alternating movements were slightly irregular. The heel-to- shin test was abnormal, and the Romberg test was positive. Her Scale for the Assessment and Rating of Ataxia (SARA) score was 8. Cranial nerve examination, muscle power, and deep tendon reflexes were otherwise normal. Laboratory investigations are summarized in the Table 1. Pre and post contrast MRI showed a linear band of tissue (web) at the inferior part of the cerebral aqueduct of sylvius with funneling of the superior part of the aqueduct resulting in upstream severe ventricular dilation with associated transependymal leak (Figures 1 and 2). The patient was loaded and maintained on the anti-convulsant Phenytoin. Then an endoscopic third ventriculostomy was done and the patient showed significant improvements in terms of symptomatology.

Table 1: Summary of laboratory investigation done for the patient.

	Patient value	Normal laboratory range
WBC	6.8	
Neutrophil percentage	50	
Hemoglobin	15	
platelets	327	
sodium	139	
Potassium	4.32	
Chlorine	103	
SGOT	28	
SGPT	16	
DIRECT BILIRUBIN	0,1	
Total bilirubin	0.15	
Creatinine	0.57	
BUN	21	
Alpha fetoprotein	1.08	

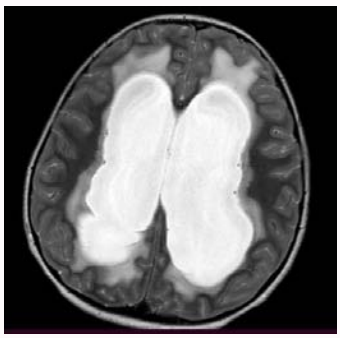

Figure 1: MRI showing dilation of the bilateral right and left lateral ventricles with associated transependymal leak.

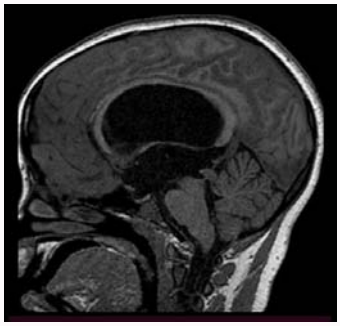

Figure 2: MRI of the patient showing dilation of the lateral and third ventricles with cerebral aqueduct obstruction by a linear band of tissue.

Discussion

Obstructive hydrocephalus is a condition characterized by the blockage of Cerebrospinal Fluid (CSF) flow, resulting in fluid accumulation within the brain's ventricular system. Aqueductal Stenosis (AS), a narrowing of the aqueduct of Sylvius which connects the third and fourth ventricles, is one such cause. Factors like congenital malformations, tumors, and inflammation may lead to AS [4]. Symptoms of obstructive hydrocephalus from AS vary in severity and duration, with common ones including headache, nausea, vomiting, and vision changes. A notable symptom is cerebellar ataxia, a disorder impacting movement coordination and balance, caused by pressure on the cerebellum from CSF accumulation [5]. Cerebellar ataxia results from damage to the cerebellum, the brain region essential for coordinating and regulating movement. In individuals with obstructive hydrocephalus due to Aqueductal Stenosis (AS), CSF accumulation within the ventricular system may increase pressure on the cerebellum, causing ataxia. Symptoms can include difficulties with walking, fine motor skills, and coordination [6]. Various theories exist about hydrocephalus's impact on the cerebral aqueduct. Williams et al. [7] suggested that hydrocephalus expansion may compress the brainstem, leading to AS, rather than AS causing hydrocephalus.

This pressure could close the aqueduct more firmly. Nugent et al. [8] proposed that expanding cerebral hemispheres might displace the third ventricle downward, narrowing the aqueduct. AS impedes the CSF's pulsatile movement, transmitting pressure to the ependymal walls, leading to periventricular edema, tissue damage, and ventricular enlargement [9]. Our patient exhibited a transependymal leak in MRI Figure 1, likely due to this process. CT and MRI are instrumental in diagnosing and managing hydrocephalus from AS. They help identify the presence and severity of aqueduct stenosis and fluid buildup, contributing to ataxia development [4,5].

Our patient's AS was caused by a web at the cerebral aqueduct's inferior part. Symptoms emerged after a long asymptomatic phase due to CSF flow compensation [10]. She experienced a gradual rise in intracranial pressure, leading to intermittent headaches, vomiting, behavioral changes, abnormal movements, and cerebellar ataxia. The management of obstructive hydrocephalus due to Aqueductal Stenosis (AS) typically includes both medical and surgical interventions. Medical management consists of administering medications to lower intracranial pressure and alleviate symptoms. Diuretics may be used to decrease CSF volume, while anticonvulsants are prescribed to control seizures [4,6]. Endoscopic Third Ventriculostomy (ETV) is the preferred surgical treatment, reducing reoperation rates and hospital stays. It creates an opening in the third ventricle floor for CSF bypass [11]. In some cases, a shunt may divert CSF to the peritoneal cavity [4,6]. A meta-analysis found ETV associated with fewer infections and repeated surgeries in Europe and Africa, though mortality and CSF leakage rates were comparable to shunt procedures [3]. Our patient underwent ETV. The approach to managing AS-induced obstructive hydrocephalus is multi-disciplinary, depending on symptom severity, stenosis cause, and patient factors. Proper management can significantly enhance life quality. The prognosis varies by cause and condition severity, with early treatment helping to mitigate long-term complications and improve outcomes [5,6].

Conclusion

Cerebral Aqueductal Stenosis due to a web is rare, with initial subtle signs and symptoms that become prominent with ventricular dilatation from stenosis in adolescence or later. This case illustrates the variety of possible symptoms, including but not limited to headache, nausea, vomiting and cognitive impairment together with cerebellar ataxia and movement disorders. T1 & T2 weighted MRI sequences are effective for aqueduct evaluation. Early detection and timely neurosurgical intervention, particularly endoscopic third ventriculostomy, are critical for limiting long-term effects.

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