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CASE REPORT

A Rare Presentation of Dandy-Walker Syndrome with Meningitis Symptoms in a 3-Month-Old Female: A Case Study From Somalia

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Video Abstract

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Abstract: Dandy-Walker Syndrome (DWS) is a rare cerebellar malformation characterized by the underdevelopment of the cerebellar vermis and associated complications. This case report presents a 3-month-old female from Somalia who exhibited fever, loss of consciousness, vomiting, and developmental delays. Neurological examination suggested signs of meningitis, and MRI revealed an enlarged posterior fossa consistent with DWS. Despite initial antibiotic treatment for suspected bacterial meningitis, CSF cultures yielded no growth, raising concerns about prior antibiotic use. Unfortunately, the patient's condition deteriorated, leading to her death. This case highlights the diagnostic challenges of DWS in resource-limited settings and emphasizes the need for improved healthcare access, early diagnosis, and intervention for rare neurological conditions.

Keywords: Dandy-Walker syndrome, cerebellar malformation, macrocephaly, developmental delay

Introduction

Dandy-Walker Syndrome (DWS) represents a rare neurological condition characterized by hypoplasia of the cerebellar vermis, dilation of the fourth ventricle, and an enlarged posterior fossa. It is the most prevalent malformation of the cerebellum, with its pathophysiology still inadequately understood.[1] These morphological irregularities often lead to a wide range of neurological and developmental disorders, including motor impairments and cognitive difficulties. The global incidence is approximately 6.79 per 100,000 births, while in the United States, it is estimated at 1 in 30,000 live births.[2]

The etiology of Dandy-Walker Syndrome has been linked to deletions in a specific chromosomal region, namely 3q24q25.1. This region encompasses the ZIC1 and ZIC4 genes, which are thought to play crucial roles in cerebellar development. Deletions of these genes have been associated with DWS-like symptoms in mouse models, indicating their importance in normal neurodevelopment.[3] Additionally, abnormal collections of cerebrospinal fluid (CSF) within the posterior fossa, identified as the Dandy-Walker complex (DWC) and arachnoid cysts (AC), contribute to the clinical manifestations of DWS. Symptoms may include psychomotor retardation, ataxia, episodes of apnea, muscular weakness, intermittent muscle spasms, seizures, nystagmus, and macrocephaly.[4,5]

Diagnosis is typically established through prenatal sonography, although challenges remain in achieving a definitive diagnosis before 18 weeks of gestation.[6] Prognostic outcomes depend on the severity of the condition and the presence of concomitant abnormalities.

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Materials and Methods

Case Presentation

A 3-month-old female from Somalia presented to our facility with fever, loss of consciousness, vomiting, and developmental delays. The clinical assessment revealed the patient to be in poor general condition, with an overall score of 10. Vital signs included a pulse rate of 119 bpm, a temperature of 39.7°C, oxygen saturation at 97%, and a respiratory rate of 40 bpm. Upon examination of the head, eyes, ears, nose, and throat, oral thrush was observed. Neurological evaluation indicated signs consistent with meningitis, including cervical rigidity, a positive Kernig's sign, a bulging fontanelle (which developed during treatment), and macrocephaly, suggestive of increased intracranial pressure. Cardiovascular and abdominal examinations were unremarkable. Nutritional assessment indicated Severe Acute Malnutrition (SAM), with a recorded weight of 3 kg and height of 52 cm (z-score < -3).

Results

Investigations: A complete blood count demonstrated a white blood cell count of 15,900, with a lymphocyte count of 7,600 (48%), granulocyte count of 45%, and platelet count of 545,000. Malaria rapid diagnostic tests were negative on two occasions. Due to suspected increased intracranial pressure, a lumbar puncture was initially delayed. MRI imaging, performed after seven days of antibiotic therapy, revealed an enlarged posterior fossa consistent with Dandy-Walker Syndrome (Figures 1–3). Neurosurgical consultation concluded that surgical intervention was not necessary at that time. A cerebrospinal fluid (CSF) analysis was recommended to rule out ventriculitis. Pending CSF analysis showed turbidity, a significant elevation in protein concentration (31 g/dl), and reduced glucose levels (37 g/dl). The white blood cell count was markedly elevated at 250 cells/μL, with 89% being polymorphonuclear neutrophils. Additionally, Gram-positive diplococci were identified, suggesting bacterial meningitis, with Streptococcus pneumoniae as the likely causative agent. CSF cultures, incubated for 48 hours, yielded no detectable microorganisms, indicating an absence of bacterial proliferation.

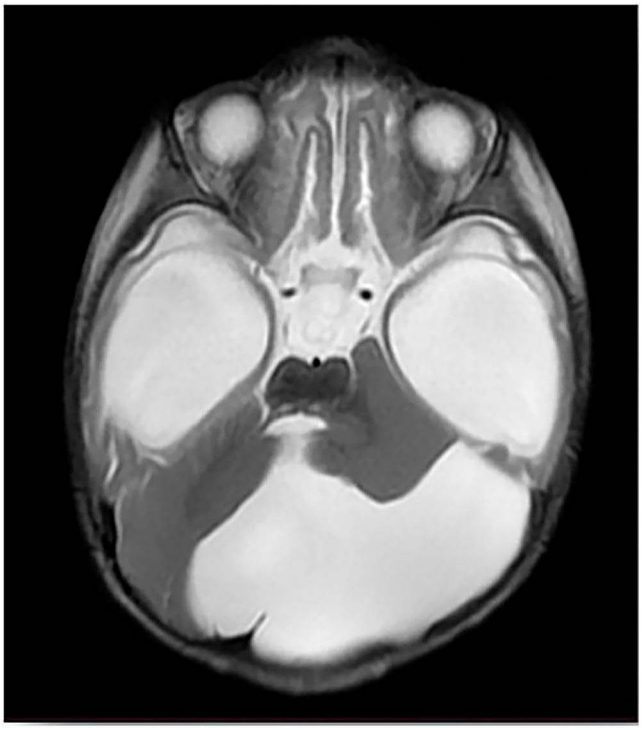

Figure 1 Axial T2 magnetic resonance imaging reveals an enlargement of the posterior fossa accompanied by cystic dilation of the fourth ventricle. Cerebellum and vermian hypoplasia with bilateral rudimentary remnant of cerebellum which are pushed anterolaterally by the cystic mass.

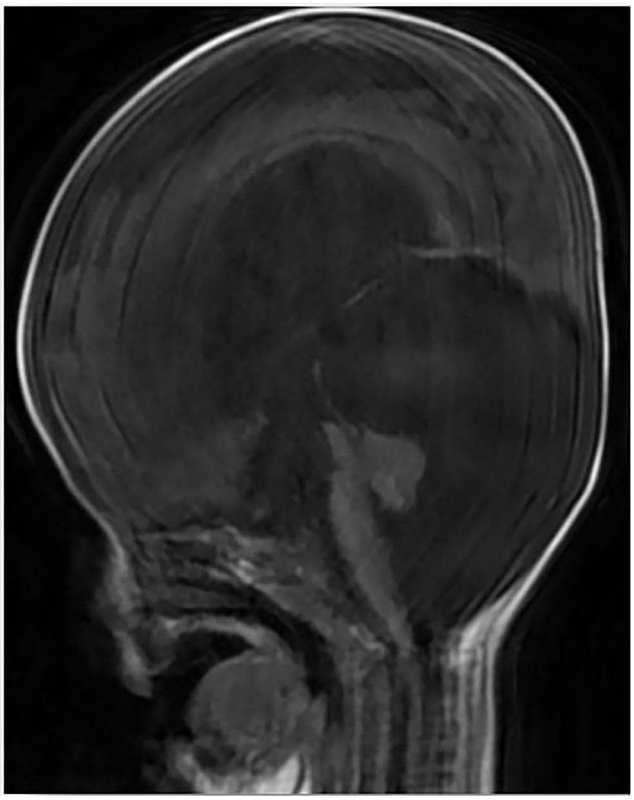

Figure 2 Sagittal T1 MRI showing: upward sloping tentorium cerebelli with Cerebellum and vermian hypoplasia. Enlarged posterior fossa with cystic dilation.

Treatment and Outcome

Initial treatment included empirical antibiotics for bacterial meningitis (Ceftriaxone and Vancomycin) and Dexamethasone. Supportive care included Ondansetron for vomiting and Paracetamol for fever. Despite an initial favorable therapeutic response, the patient's clinical condition subsequently declined, ultimately resulting in her death due to the advancement of the disease.

This case highlights the complexities of diagnosing rare neurological conditions such as Dandy-Walker Syndrome in resource-limited settings. The initial presentation mimicking meningitis necessitated careful consideration and management. The challenges encountered underline the importance of adhering to ethical research protocols and university review board guidelines. The case illustrates the critical need for heightened awareness and diagnostic insight when faced with atypical presentations in pediatric patients, particularly in environments with limited resources. Further studies are warranted to improve diagnostic pathways and treatment strategies for rare neurological conditions in similar contexts.

Discussion

This case presents an atypical manifestation of Dandy-Walker Syndrome (DWS) in a three-month-old female patient from Somalia, resulting in adverse clinical outcomes. The patient's clinical symptoms included fever, developmental delay, and signs of meningeal irritation, ultimately leading to a diagnosis of bacterial meningitis. Despite aggressive treatment, the patient's clinical deterioration underscores the complexities involved in diagnosing and managing DWS, particularly in resource-limited settings. The absence of positive cerebrospinal fluid (CSF) cultures, despite the presence of clinical signs indicative of infection, raises significant concerns regarding the potential impact of prior antibiotic administration on diagnostic results. This case highlights the urgent need for timely diagnosis and intervention, especially in low-resource environments where access to advanced diagnostic tools is severely restricted.

The etiology of DWS remains inadequately clarified; however, overlapping deletions on chromosome 3q24q25.1 have been implicated in a limited number of documented cases.[4] The condition, initially reported by medical practitioners

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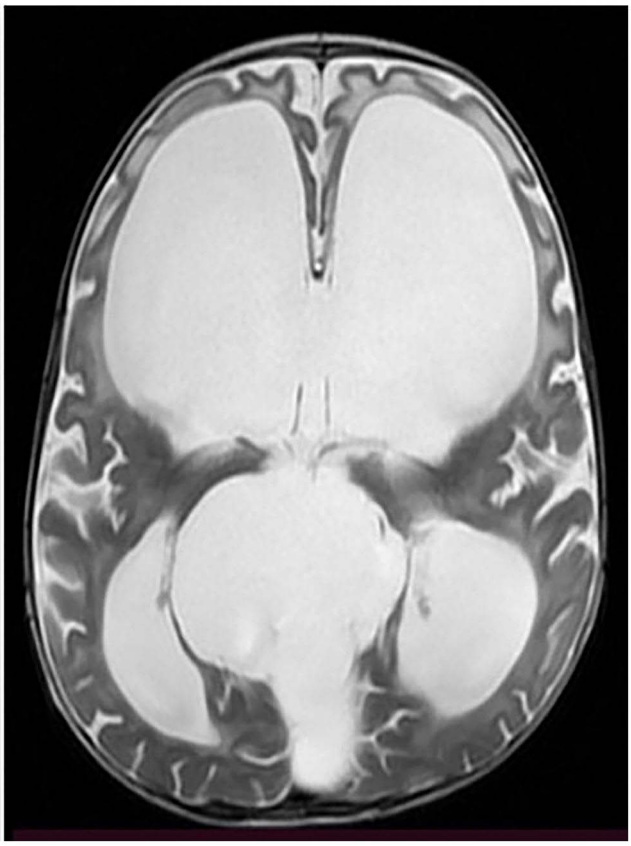

Figure 3 Marked dilation of the bilateral lateral ventricular and 3rd ventricular (obstructive hydrocephalus).

Walter Dandy and Arthur Walker in the early 20th century, encompasses a spectrum of congenital anomalies that affect the cerebellum and its adjacent structures. Despite nearly a century of recognition, the medical community continues to encounter significant challenges in comprehending the etiology, progression, and clinical manifestations of Dandy-Walker Syndrome (DWS).[2] Cystic dilation of the fourth ventricle, either agenesis or hypoplasia of the cerebellar vermis, and an increased posterior fossa are indicative of Dandy-Walker Syndrome.[6] Although the current case did not explore genetic determinants, the clinical features correspond with recognized characteristics of DWS, including cerebellar hypoplasia, an enlarged fourth ventricle, and macrocephaly.[2]

The patient's clinical presentation, characterized by developmental delay, signs of meningeal irritation, and macrocephaly, strongly suggests increased intracranial pressure (ICP) associated with hydrocephalus. This observation is consistent with previous reports indicating that DWS frequently manifests as hydrocephalus in pediatric patients.[1] Imaging via brain CT confirmed markedly dilated third and fourth ventricles, as well as bilateral lateral cerebral ventricles, thereby reinforcing the diagnosis of hydrocephalus. Additionally, the presence of an infratentorial cystic lesion communicating with the dilated fourth ventricle strongly suggests the existence of a Blake's pouch cyst, which is often associated with DWS.

The diagnosis of bacterial meningitis was substantiated by the examination of a turbid CSF specimen, which revealed elevated protein concentrations, reduced glucose levels, and increased leukocyte counts predominantly consisting of polymorphonuclear neutrophils. Gram-positive diplococci were identified, suggestive of Streptococcus pneumoniae. However, the CSF culture yielded no bacterial growth after 48 hours of incubation, likely attributable to prior antibiotic therapy administered before CSF analysis.

While this case report emphasizes a rare presentation of Dandy-Walker Syndrome, the neurological complications observed align with findings from other studies regarding bacterial meningitis in young infants. Investigations have shown that over 75% of bacterial meningitis cases in neonates result in neurological complications, often culminating in adverse

prognoses.[7] This underscores the necessity for rigorous monitoring of neurological sequelae in neonates afflicted with meningitis and highlights the importance of prompt cranial imaging to identify those exhibiting neurological deficits.

The primary etiological agents implicated in bacterial meningitis among infants are Group B Streptococcus and Escherichia coli, which together account for 65–78% of cases, with mortality rates ranging from 13% to 25% among both term and preterm infants.[8,9] Research focusing on bacterial meningitis in adults indicates that approximately three-fourths of affected individuals experience significant intracranial complications, including hydrocephalus, subdural empyema, infarction, and ventriculitis, all contributing to enduring neurological deficits and mortality.[5,10,11] The current investigation reveals that neurological complications manifest in more than 75% of bacterial meningitis cases in young infants, which are linked to a considerable incidence of unfavorable outcomes.

The results from this case underscore the critical importance of prompt diagnosis and intervention for Dandy-Walker Syndrome, particularly in resource-constrained environments. Despite the challenges associated with diagnosing DWS, prenatal sonography plays a vital role in its detection, although establishing a definitive diagnosis prior to 18 weeks of gestation can be problematic. Timely diagnosis and immediate intervention may significantly enhance prognosis and mitigate the long-term neurological repercussions associated with both Dandy-Walker Syndrome and bacterial meningitis.

This case contributes to the limited literature on Dandy-Walker Syndrome, particularly in resource-limited settings. The patient exhibited atypical symptoms, including signs of meningitis, complicating the clinical picture and highlighting the diagnostic challenges faced by healthcare providers in such environments. By documenting this case, we aim to raise awareness about DWS and its potential presentations among healthcare professionals, particularly in regions where access to advanced diagnostic tools is limited. Improved awareness can facilitate earlier recognition and intervention, potentially improving patient outcomes. This case underscores the importance of strengthening healthcare infrastructure and training in the diagnosis and management of rare neurological conditions, emphasizing the need for comprehensive healthcare strategies to address disparities in access to care, which is vital for achieving better health outcomes.

Conclusion

This case highlights the critical need for improved awareness and early intervention in rare neurological conditions such as Dandy-Walker Syndrome (DWS). In resource-limited settings, healthcare providers must be trained to recognize the signs and symptoms of DWS to facilitate timely diagnosis and management. Enhanced access to diagnostic tools and the establishment of comprehensive care protocols are essential for better patient outcomes. Furthermore, investing in healthcare infrastructure and encouraging research initiatives will contribute to a more effective response to rare conditions, ultimately improving the quality of care for affected individuals.

Implications

1. Healthcare Training: There is a need for enhanced training programs for healthcare professionals in recognizing and managing rare neurological conditions, ensuring better patient outcomes.
2. Access to Diagnostic Tools: Improved access to essential diagnostic tools, including prenatal sonography and advanced imaging techniques, is crucial for timely intervention in cases like DWS.
3. Research and Policy Development: The findings emphasize the necessity for ongoing research to understand DWS better and to inform policy decisions that enhance healthcare infrastructure and resources in low-resource environments.

Contextualizing to Sustainable Development Goals

This case report corresponds with multiple Sustainable Development Goals (SDGs), specifically SDG 3 (Good Health and Well-being) and SDG 10 (Reduced Inequalities). SDG 3: The case underscores the significance of enhancing access to high-quality healthcare, particularly in settings with limited resources, for the early detection and management of intricate conditions such as DWS and bacterial meningitis. SDG 10: The case emphasizes the necessity of mitigating health disparities and bolstering access to healthcare services for all individuals, irrespective of geographic location or socioeconomic standing. This becomes particularly imperative for rare conditions like DWS, which may be devoid of specialized healthcare infrastructure in developing nations.

Additional research is requisite to explore the enduring ramifications of DWS and bacterial meningitis on neurological development, especially in resource-constrained environments. This understanding is vital for formulating effective interventions and enhancing the overall quality of life for those impacted by these conditions.

Parental Consent

Written informed consent was secured from the patient's guardians for the purposes of publication as well as for any supplementary imagery.

Ethical Considerations

The study protocol, case investigation, and consent documentation were subjected to rigorous evaluation by the institutional review board of the College of Health Sciences at Amoud University. The study was approved in collaboration with the Ministry of Health and Borama Regional Hospital located in the Awdal Region of Somaliland (BRH-160/2024).

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Author Contributions

All authors made a significant contribution to the work reported, whether that is in the conception, study design, execution, acquisition of data, analysis and interpretation, or in all these areas; took part in drafting, revising or critically reviewing the article; gave final approval of the version to be published; have agreed on the journal to which the article has been submitted; and agree to be accountable for all aspects of the work.

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Disclosure

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