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CASE REPORT

The "White Cerebellum Sign", a Striking Computed Tomography Scan Finding in a Critically Ill Infant: A Case Report

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Abstract: The "white cerebellum sign" is a striking neuroradiological finding, resulting from diffuse hypodense brain lesions in both cerebral hemispheres while sparing the cerebellum. This indicates widespread ischemic damage or expanded cerebral edema associated with the loss of white-gray matter differentiation. It is commonly reported in pediatric cases with severe neurological conditions, usually resulting in a poor prognosis. We present a case of a three-month-old female infant who was managed for confirmed acute bacterial meningitis complicated by status epilepticus, where the Computed tomography scan (CT scan) showed the "white cerebellum sign" in relation to diffuse cerebral hypoxic-ischemic lesions. However, the patient had a favorable outcome following treatment. This case underscores the importance of early recognition in cases presenting with the "white cerebellum sign". It highlights the potential for a positive prognosis even in severe neurological conditions when appropriate interventions are promptly administered.

Keywords: white cerebellum sign, CT scan, meningitis, case report

Introduction

The "white cerebellum sign" describes the typical sparing of the cerebellum, which appears relatively hyper-attenuating compared to the affected supratentorial brain due to the loss of gray-white matter differentiation, diffuse cerebral hypoattenuation, and sulcal effacement.[1] The "white cerebellum sign" is an unusual yet worrying characteristic finding in neuroradiology and is mainly documented in paediatric patients with hypoxic brain damage with poor outcomes.[2] Children from birth to two years of age are the most affected, with a meager prognosis.[3] It has been associated with several cases of cerebral infections such as meningitis, encephalitis, severe head trauma, perinatal asphyxia, drowning, hypothermia, status epilepticus, post-anoxic encephalopathy, and other cases of general brain hypoperfusion.[4] Radiologists are particularly required to recognize this typical appearance of the "white cerebellum sign" to establish an early correct diagnosis, which is crucial for clinicians to plan appropriate management.[3]

The concrete mechanism is still debated; however, one study suggested that cerebral blood flow reorganizes predominantly into the posterior circulation. This may be explained by cerebral edema in hypoxic-ischemic cases.[4–6] We present this case in view of the rarity of this sign (especially in the context of infection or malaria), its classic radiological appearance, and its prognostic significance. The patient was managed in a hospital located in a resource-limited country, where cerebral infections, including meningitis and cerebral malaria, are devastating to children.

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Case Presentation

Patient Description and Case History

A previously healthy three-month-old female infant presented with a one-week history of fever. She was initially diagnosed and treated for malaria with oral quinine syrup and paracetamol at a peripheral clinic for four days. Despite this treatment, the baby continued to have persistent fever. Three days later, she developed an acute-onset seizures characterized by jerky movements of the right hand and leg, eye blinking, altered consciousness, and difficulty in breathing. The patient was promptly transferred to a tertiary hospital for further management. This marked the infant's first admission, and she had no history of head trauma since birth. Her medical records revealed a normal and uneventful pregnancy, with delivery occurring at 38 weeks and 5 days of gestation, as confirmed by antenatal obstetric ultrasound. Since birth, she has been exclusively breastfed, exhibiting normal age-appropriate growth and achieving developmental milestones within expected ranges. Moreover, her parents diligently adhered to the recommended vaccination schedule, ensuring she received all essential immunizations appropriate for her age.

Physical Examination Results

Upon admission, the infant was comatose. She responded to pain stimulation by opening her eyes and moaning and exhibited withdrawal from nailbed pain. According to the Pediatric Glasgow Coma Scale (pGCS), she scored 9 of 15. The patient also presented with high-grade fever (38.8°C) and exhibited jerky movements of the right hand and leg, as well as blinking of the eyes, lasting more than five minutes. Notable findings included marked pallor and moderate-to-severe respiratory distress (Silverman score of 4), including nasal flaring, intercostal and subcostal retraction, and chest indrawing. Oxygen saturation levels were measured at 87–89% in room air. Physical examination revealed neck stiffness and positive Kernig's and Brudzinski's signs. The infant had a weight of 5.6 kg and a length of 53 cm, which suggests normal growth and nutritional status. The infant was admitted to the Pediatric Acute Critically Ill Patients Special Care Unit where emergency treatment and life support were initiated. Oxygen therapy was administered at a rate of 2 L/min via nasal prongs, and sequential doses of anticonvulsant agents were administered to manage the status epilepticus.

Results and Interpretations of Investigations

The baseline complete blood count revealed a leukocyte count of 14,300/μL, with granulocytes accounting for 53.6% (7.7109/L). Anemia was also evident, with a hemoglobin level of 6.9 g/dL and a hematocrit of 23%. The random blood sugar level was measured at 4.2 mmol/dL, and the peripheral blood smear for malarial parasites was negative. Cerebrospinal fluid (CSF) analysis revealed turbidity and purulence, elevated protein levels (11.1 g/L), and low glucose levels (0.02 mmol/liter). CSF samples were collected for culturing.

A brain Computed tomography scan (CT scan) of the patient revealed a "white cerebellum sign" in relation to diffuse hypodensity in both cerebral hemispheres with loss of white-grey matter differentiation, indicative of widespread hypoxic-ischemic lesions (Figures 1 and 2).

Treatment Plan

The patient received a loading dose of phenobarbital (20 mg/kg), followed by two doses of phenobarbital (10 mg/kg) after 30 min. Subsequently, the patient was switched to intravenous phenytoin, starting with a dose of 15 mg/kg, administered slowly over 60 min. Maintenance therapy with phenobarbital was continued at a dose of 5 mg/kg every 12 hours for four days. Additional supportive care included the intravenous maintenance of fluids, antipyretics, and maternal breastfeeding via a nasogastric tube. Vital signs and urine output were closely monitored, and intravenous ceftriaxone at a dosage of 100 mg/kg per day was initiated as empirical antimicrobial therapy.

On the fourth day of admission, CSF culture results showed the isolation of oxacillin-sensitive *Staphylococcus aureus*, which was also susceptible to trimethoprim-sulfamethoxazole and chloramphenicol, but resistant to azithromycin, ampicillin, and erythromycin. The diagnosis of Methicillin-Sensitive *Staphylococcus aureus* (MSSA) Meningitis was made, and intravenous Oxacillin was initiated at a dose of 150 mg per kg per day.

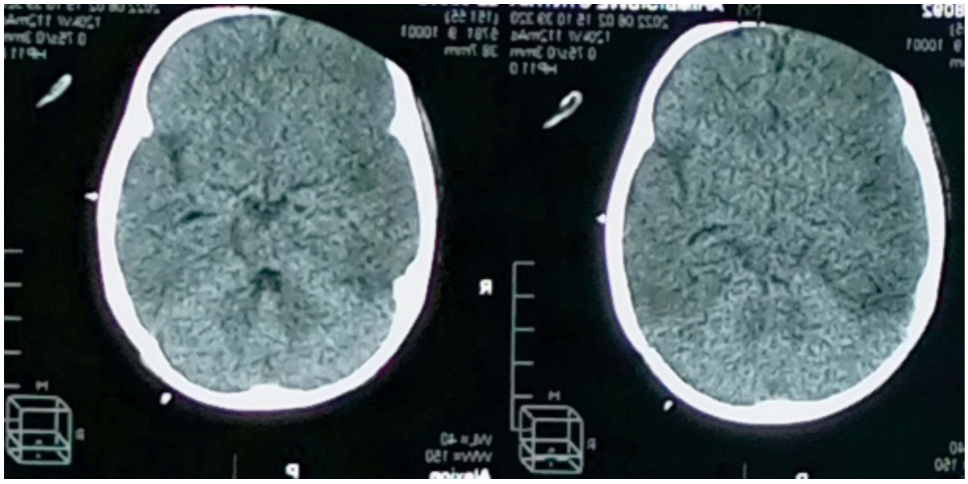

Figure 1 Computed Tomographic scan of the head in axial view demonstrates hypo-density of both brain hemispheres with loss of white-grey matter differentiation and the "White Cerebellum" sign in relation to hypoxic-ischemic lesions.

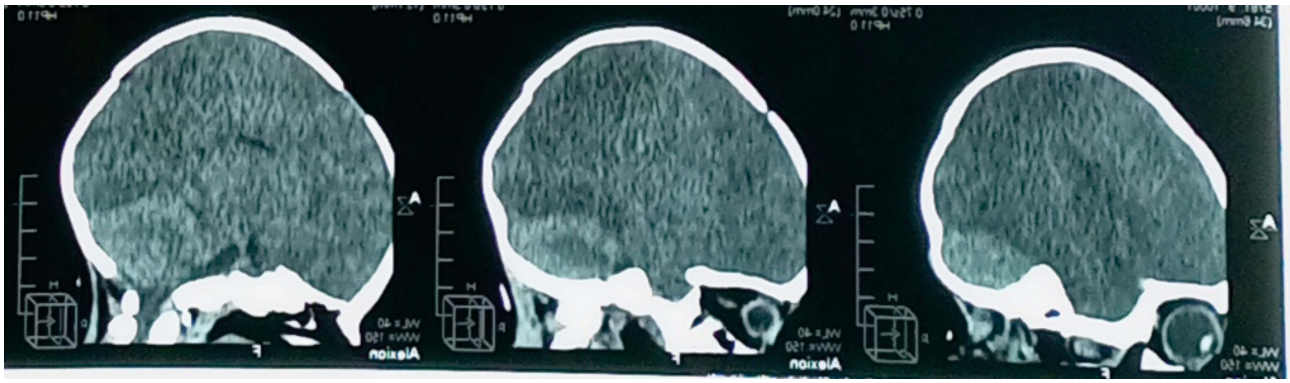

Figure 2 Computed Tomographic scan of the head in sagittal view, demonstrates hypo-density of both brain hemispheres with loss of white-grey matter differentiation and the "White Cerebellum" sign in relation to hypoxic-ischemic lesions.

Outcome

Within two weeks, the infant exhibited remarkable clinical improvement. She fully recovered consciousness and progressively regained the ability to breastfeed. The fever and other symptoms resolved completely. The baby was discharged, and parents were provided with detailed instructions regarding further follow-up. During the follow-up visits, she was reviewed two months later, revealing normal, age-appropriate developmental progress, with no further episodes of convulsions reported.

Discussion

The "White cerebellum sign" is often associated with severe permanent brain damage and a poor prognosis. One-third of the patients die, while the remaining patients suffer from severe irreversible brain damage.[2] The case presented here is a rare occurrence, in which the patient showed improvement, demonstrating a remarkable short-term favorable outcome and good developmental progress without notable sequelae three months after discharge. The current literature distinguishes between the "white cerebellum sign"" and the "reversal sign".[7,8] However, some authors have argued that the two radiological signs are similar.[9,10] To elucidate the imaging discrepancies between these signs, MOOSA and ANDRONIKOU presented two cases of pediatric patients with neurological injuries, one demonstrating the "white cerebellum sign" on CT scan and the other exhibiting the true "Reversal Sign".[8] In the first case, CT scan images revealed a "white cerebellum sign", characterized by global hypodensity of the supratentorial parenchyma with effacement of the

gray–white interface. However, the cerebellum and other infratentorial structures maintained their density and appeared to be relatively hyperdense. In contrast, the second case displayed features of the true "reversal sign", including hypodensity of the peripheral cortex with sparing of the central white matter, gray matter nuclei, and posterior fossa structures, accompanied by a reversal of the gray–white difference.[8] The description of CT scan appearance of the reversal sign in neuroradiological texts remains inconsistent and confusing.

This paper presents a case illustrating the occurrence of the "white cerebellum sign" in the CT images of a 3-month-old infant who was treated for Staphylococcus meningitis and status epilepticus. The patient had no history of asphyxia or trauma. The CT scan revealed hypodensity in both hemispheres, resulting in loss of white-grey matter differentiation. Additionally, a "white cerebellum sign" was observed, indicating the presence of hypoxic-ischemic lesions. However, the patient had a favorable outcome following treatment. It's crucial to recognize the limitations of this case report, primarily stemming from financial constraints in the resource-limited healthcare setting where the patient was treated. We could not conduct repeated CT scans or advanced neurological tests like Magnetic resonance imaging (MRI) during follow-up, hampering our ability to provide continuous radiological assessment. Nevertheless, the observed favorable clinical improvement indicates a positive general and neurological prognosis despite these constraints.

Conclusion

The "white cerebellum sign" is a rare radiological finding observed on CT scans and is mainly reported in critically ill pediatric patients with severe brain injuries. Although uncommon, this sign typically indicates severe widespread anoxic-ischemic brain injury or expanded cerebral edema. Therefore, it is usually associated with a poor prognosis. Consequently, recognizing and identifying this sign promptly is of utmost importance for both physicians and radiologists.

Abbreviations

CBC, Complete Blood Count; CSF, Cerebrospinal Fluid; CT-Scan, Computed Tomography Scan; GWD, Grey-white difference; HB, Haemoglobin; HTC, Haematocrit; kg, kilogram; mg, Milligram; MRI, Magnetic resonance imaging; pGCS, Pediatric Glasgow Coma Scale; WBC, White Blood Cells.

Data Sharing Statement

All data generated or analyzed during this study are included in this case report.

Ethical Approval

Not applicable. The institutional approval was not required.

Patient Consent and Right of Anonymity

The parental informed consent for publication was obtained.

Disclosure

The authors report no conflicts of interest in this work.

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