

The Uncommon White Cerebellum Sign in Three Patients in Different Settings

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Abstract**Case Report**

The white cerebellum sign is a rare, unusual but striking radiological sign encountered commonly in patients with severe and often irreversible anoxic-ischemic brain injury, usually due to extra-cranial causes. It is not frequently described and its pathophysiology is still controversial. Radiologically, it is characterized by cerebellar hyperdensity, associated with diffuse cerebral hemispheres hypoattenuation. Its identification by the radiologists is imperative as it is often associated with an unfavorable prognosis and poor outcome. In this paper, we report three cases of children with full clinical and imaging workup, in whom brain computed tomography (CT) scan revealed the white cerebellum sign. We discussed the relevance of this sign as it affects diagnosis, treatment, prognosis and equally reviewed the pertinent literatures.

Keywords: The white cerebellum, anoxic-ischemic brain injury, cerebellar hyperdensity.**Copyright © 2023 The Author(s):** This is an open-access article distributed under the terms of the Creative Commons Attribution 4.0 International License (CC BY-NC 4.0) which permits unrestricted use, distribution, and reproduction in any medium for non-commercial use provided the original author and source are credited.

INTRODUCTION

White cerebellum sign is a classic yet uncommon radiological finding mainly reported in pediatric patients and presents a wide variety of causes. It is also known as inversion sign or dense cerebellum sign, and is often an indicator of severe irreversible anoxic-ischemic brain damage with a poor prognosis [1, 2]. It indicates diffuse decrease in the density of the supratentorial brain parenchyma, with relatively increased attenuation of the thalami, brainstem, and cerebellum [3, 4]. The white cerebellum sign has drawn the attention of scientists over the years but rarely documented in the literature. Therefore, it is very important to be aware of this sign and recognize it at the earliest for diagnostic, therapeutic and prognostic purposes. We report three cases of children with full clinical and imaging workup, in whom brain CT scan revealed the white cerebellum sign.

CASE REPORTS

Patient 1

We report a case of a 3-year-old boy with no historical medical or surgical background, who was admitted to the hospital with a 2-day history of fever, vomiting, and lethargy. On initial exam, the patient was febrile, lethargic with neck stiffness, and had normal brainstem reflexes with a 11/15 Glasgow. The biological assessments showed hyperleukocytosis at $19.10^3/l$ with polymorphonuclear neutrophils. Before performing the lumbar puncture, CT of the head demonstrated the white cerebellum sign denoting a relative hyperdensity of the thalami, brainstem and cerebellum, when compared to the supratentorial compartment (Figure 1). Further cerebrospinal fluid findings have proven acute bacterial meningitis. The patient was treated with antibiotics, steroids, and osmotherapy and made a good recovery.

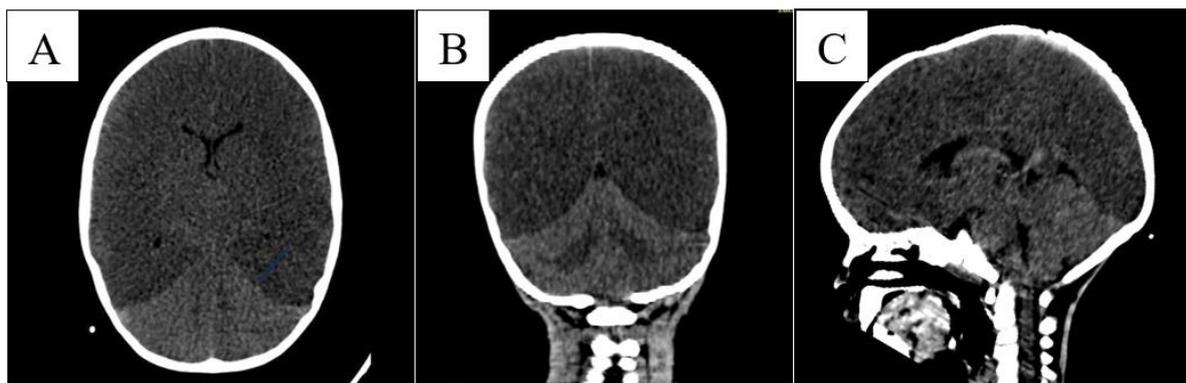


Figure 1: Non-contrast axial (A), coronal (B) and sagittal (C) CT scan images showing supratentorial hypodensity with deletion of sulcus and fissures in the cerebral hemispheres, associated with spontaneous hyperdensity of the thalami, brainstem and cerebellum

Patient 2

This patient is a 3-year-old male with history of epilepsy (complex partial seizures type), referred for brain CT on account of deteriorating conscious level and focal fits one day before presentation. Patient had complex partial seizures that includes recurrent focal fits of right side of body for 40 minutes duration associated with decreased conscious level. There was no history of up-rolling of eyes or frothing from mouth. At presentation, patient had a Glasgow scale of 9/15, pupils were fixed and dilated. His complete blood picture, liver

function tests, renal function tests, serum electrolytes, calcium, magnesium, phosphate, albumin, C-reactive protein and urine routine examination were within normal limits. CT scan brain plain was done which showed diffuse hemispheric cerebral edema marked in the left side, with relative increased attenuation of the thalami and cerebellum (Figure 2). The diagnosis of white cerebellum sign caused by status epilepticus was made, and patient was managed accordingly. Patient's family was counseled in detail.

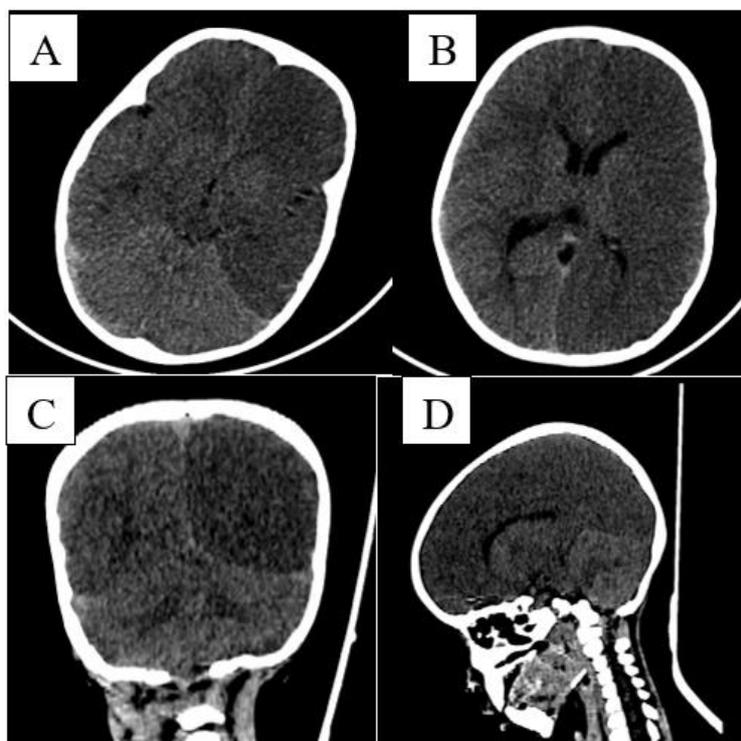


Figure 2: Head CT scan in axial (A, B), coronal (C) and sagittal (D) sections revealing diffuse hemispheric cerebral edema marked in the left side, with increased attenuation of the thalami and cerebellum

Patient 3

A 4-year-old child with cerebral palsy and history of birth asphyxia presented with episodes of generalized tonic seizures. Patient was hypotensive and

bradycardic with shallow respiration. His pupils were bilaterally dilated and not reacting to light. After initial stabilization, intubation and ventilation, an urgent head CT scan revealed diffuse reduction in the density of the

supratentorial brain parenchyma with loss of normal grey-white matter interface in keeping with severe oedema. This was associated with comparatively increased density of brainstem and bilateral cerebellar hemispheres and was consistent with white cerebellum

sign (Figure 3). There was no evidence of infection and patient was then managed symptomatically. Despite mechanical ventilation, anti-cerebral edema measures and supportive care in the intensive care unit, the child succumbed after 24 hours of admission.

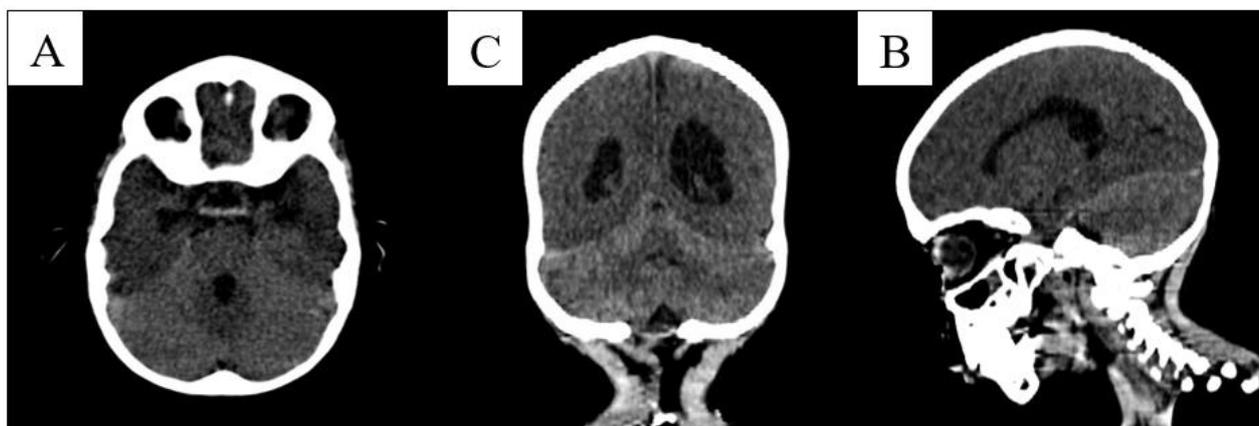


Figure 3: Cerebral CT scan in axial (A), coronal (B) and sagittal (C) sections demonstrating diffuse reduction in the density of the supratentorial brain parenchyma with loss of normal grey-white matter interface in keeping with severe oedema, associated with comparatively increased density of brainstem and bilateral cerebellar hemispheres

DISCUSSION

White cerebellum sign, also known as dense cerebellum sign or the inversion sign, is a rare neuroimaging finding. It usually represents severe and irreversible anoxic neuronal damage that results in global cerebral ischemia [5,6]. It is a radiological sign most often described in children with hypoxic brain injury, but it has also been described in patients with severe head trauma, status epilepticus, drowning, bacterial meningitis, encephalitis, neonatal asphyxia, child abuse, smoke inhalation, hypothermia, postpartum, Wernicke's encephalopathy, post-cardiac arrest asphyxia and other causes of global hypoperfusion, and is rare in adults compared to children [3,7]. The exact mechanism and pathogenesis remain unclear and is not fully understood, though various and several hypothesis has been put regarding as the cause of this rare condition, which some includes distention of deep medullary veins due to partial obstruction by raised intra-cranial pressure resulting in cerebral oedema.

Another proposed theories are the relative preference of posterior circulation blood flow, transtentorial herniation partially relieving the increased intracranial pressure and thus increase perfusion of central structures [3, 5, 7], hypoxia disrupts the sodium pump and limits ATP (Adenosine Triphosphate) production and the influx of water and sodium into cells causes cytotoxic oedema. Finally, anoxia and ischemia elevate brain glucose leading to damage of the cortex and basal ganglia [1, 4, 8, 9]. Specific appearance of this sign, on CT as in Magnetic resonance imaging (MRI) brain, is due to a diffuse oedema leading to a generalized hypodensity of the cerebral hemisphere with the loss of distinction between gray and white matter as compared

to cerebellum which retains its normal density and appear hyperdense [5, 7]. Other signs of diffuse cerebral edema are the absence of cortical grooves, erasure of basal cisterns, especially the peri-mesencephalic cisterns, and bilateral compression of the lateral ventricles [10]. Although the white sign may be discrete on CT imaging, the MRI findings appear to be conspicuous on apparent diffusion coefficient (ADC) maps and becomes the most sensitive modality for the early diagnosis of cerebral hypoxia. Metabolic differences between supra and infratentorial compartments and shifts in compartment perfusion with rising intracranial pressure have been postulated as possible explanations for this finding [10,11]. This sign is important from diagnostic, therapeutic and prognostic side as treatment is basically symptomatic and aimed to reduce intracranial hypertension and reversing ischemic injury [1, 10]. White cerebellum sign, when present, is a poor prognostic indicator [3]. It has been documented that about one-third of patient with this sign on CT scan will die while the remaining population will suffer severe and permanent brain damage with subsequent development of diffuse atrophy and cystic encephalomalacia [1, 9].

CONCLUSION

White cerebellum sign is a subtle but classic radiological imaging finding mostly seen in pediatric patients with history of birth asphyxia or trauma. Neuroimaging features are important as they inform about the extent and severity of the injury and can predict the outcome in long term. Early diagnosis of this sign is imperative to avoid unnecessary investigations and prolonging the patient's misery. Clinicians and particularly radiologists must take cognizance of the

classic appearance of the white cerebellum sign for the bleak and grim prognosis it carries.

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