WHITE CEREBELLUM SIGN AS A DARK PROGNOSTIC INDICATOR OF CEREBRAL INJURY: A CASE REPORT

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Submission Date: 6th Oct., 2023
Date of Acceptance: 1st April, 2024
Publication Date: 30th April, 2024

ABSTRACT

Introduction: **The white cerebellum sign (WCS) is a classical but rare radiological finding usually associated with irreversible diffuse hypoxic-ischemic cerebral injury. Very few cases exist in the literature globally, especially from the West African region, as a potential hallmark of poor prognostic outcome. We describe the white cerebellum sign in a Nigerian pediatric patient, managed for severe head injury.**

Case Presentation: **A fourteen-year old boy presented to our emergency department with loss of consciousness following a pedestrian road traffic accident. Physical examination revealed a critically ill boy with fever, hypotension, tachycardia, gasping respiration, GCS 3, bilateral dilated unreactive pupils, absent corneal, gag and oculocephalic reflexes. He was thus diagnosed of severe traumatic brain injury and brainstem dysfunction. He had endotracheal intubation, ventilatory and inotropic support. Cranial computerized tomography scan of the patient showed radiological features in keeping with the WCS. His clinical status remained poor until he suffered a cardiac arrest about twelve hours after admission.**

Conclusion: **WCS has been reported in relation to child abuse, anoxic-ischemic brain injury, inflammatory and metabolic brain disorders and trauma. It is a classical radiological description of diffuse cerebral edema alongside relatively normal cerebellar hemispheres and brainstem. Management of this pathology is symptomatic, and aims to ameliorate the associated raised intracranial pressure, control seizures and prevent cerebral infarction. The index patient, who presented 24 hours after severe head injury with associated early post-traumatic seizures, respiratory failure and brainstem dysfunction, had an unfavourable outcome consistent with previous reports of WCS.**

We have reported the rare but classical white cerebellum sign. It remains a grave prognosticator of cerebral injury and should be sought for in the neuroimaging of patients with acute brain insults.

Keywords: Hypoxic-Ischemic encephalopathy, Inversion sign, Pediatric head injury, Case report

INTRODUCTION

The white cerebellum sign is a characterization of relative hyperdensity of the cerebellar hemispheres and brainstem against a backdrop of diffuse hypodensity of the cerebrum, on cranial computerized tomography scan.[1-3] Very few cases exist in the literature globally, especially from the West African region as a potential hallmark of poor prognostic outcome. These have been reported mostly in association with hypoxic-ischemic encephalopathy and few ascribed to trauma.[1-4] Whereas the sign has been widely associated with irreversible brain damage, few cases of survival have been documented, with most of these having debilitating neurological sequaelae.[1,3-5] However, rare instances of good outcome have been reported by previous authors.[3] Given that the cerebral edema-induced raised intracranial pressure is central to the resultant global cerebral ischemia, it stands to reason that early identification of this anomaly on neuroimaging and prompt institution of the appropriate therapy is crucial to the survival of the affected individuals and their long-term neurological status.

We describe the white cerebellum sign in a fourteen-year-old Nigerian boy, who was managed for severe head injury in a resource poor neurosurgical facility. The authors hope to bring more awareness to this rare poor prognostic indicator of brain injury to

clinicians. This case report has been written in accordance with the CARE guidelines.[6]

CASE PRESENTATION

A fourteen-year-old right-handed boy presented with persistent loss of consciousness following pedestrian motor vehicular accident, which occurred a day earlier. He had two episodes of generalized tonic-clonic seizures prior to presentation, and had received intravenous fluids and analgesics at a private facility where initial care was sought. There was no past history of seizure disorder and no family history of epilepsy.

Clinical Findings

Physical examination revealed a critically ill boy with fever, tachycardia, hypotension, and gasping respiration. He had a Glasgow coma scale score of 3 with bilaterally blown unreactive pupils, absent corneal, gag, oculocephalic reflexes and a flaccid quadriplegia. He had no other associated injuries. We made a diagnosis of severe traumatic brain injury with brainstem dysfunction.

Timeline

He had immediate endotracheal intubation in the emergency department, following which he became apneic. He was manually ventilated with an AMBU bag, via the endotracheal tube, and was commenced on intravenous normal saline solution, intravenous acetaminophen, Dopamine infusion and oxygen.

Diagnostic Assessment

Cranial computerized tomography (CT) scan done about an hour after admission (Figure 1) showed diffuse hypodensity of the supratentorial structures indicative of generalised cerebral edema, with differential relative hyperdensity of the cerebellum and brainstem. There was an associated effacement of all the ventricles and basal cisterns. The prognosis was deemed to be guarded based on the clinico-radiological evidence, and this was discussed with the patient's parents.

Therapeutic Intervention

Intravenous Phenytoin was prescribed for seizure control at 200mg 8 hourly for the first 24 hours, then subsequently 200mg daily. However, medical decompression with intravenous 20% Mannitol could not be instituted because of the patient's persistent hypotensive state.

Follow-Up And Outcome

The patient suffered a cardiac arrest after twelve hours of hospitalization and was certified dead following failed resuscitative efforts.

DISCUSSION

The white cerebellum sign (WCS) is also known as the inversion sign. It is a pathological neuroimaging finding relating to the cranial CT scan appearance of diffuse cerebral edema with preservation of the infratentorial structures, and consequently, relative but not actual hyperdensity of this brain compartment.[1-4] The term, in the strict sense, is a misnomer given the fact that the abnormality is in the cerebrum rather than the cerebellum. This radiological appearance may therefore be more appropriately described as, '*the dark cerebrum sign*'. Although, used interchangeably with the 'reversal

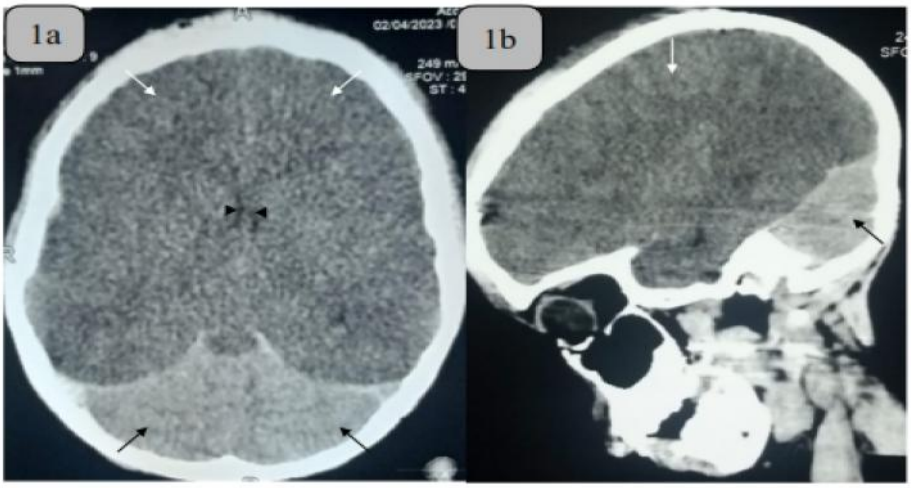

Figure 1: Unenhanced axial cranial CT (a) with sagittal reconstruction (b) showing hyperdense cerebellum (black arrows) relative to the density of the cerebral hemispheres (white arrows) aka 'the white cerebellum sign'.

Note the effacement of the frontal horns of the lateral ventricles (black arrow heads), generalized cerebral sulcal effacement (white arrows), and poor gray-white matter differentiation as evidence of severe diffuse cerebral edema.

sign' by some authors, these two signs are distinct and should be recognized as such. The latter refers to reversal of the normal gray/white matter densities, with the central cerebral structures, thalamus, brainstem and cerebellum appearing denser compared to the peripheral cerebral cortices.[7-9] While the WCS was first described within the context of child abuse, it has been reported in other conditions associated with anoxic-ischemic brain injury, inflammatory and metabolic brain disorders and trauma.[1,3,4,10] These include birth asphyxia, meningoencephalitis, acute cerebellitis, metabolic encephalopathy, drowning, status epilepticus, and severe traumatic brain injury (TBI).[1,3,7,10] Reports of WCS following cardiac arrest, in the immediate post-partum period, as well as post-revision of ventriculo-peritoneal shunt exist in the literature.[1,5,10] In the retrospective review of 20 cases of WCS by Han *et al.*, nine of the patients had hypoxia/anoxia, while only two were associated with TBI.[9] Conversely, in a series of five patients reported by Pereira *et al.* over a three-year period, the predominant aetiology was trauma (three cases).[7] About 10-20% of the documented patients with the condition were aged 18 years and above.[4] The index case, which fell within the paediatric age-group, had severe TBI with associated brainstem dysfunction and respiratory failure. It is important to note that this patient was not declared brain dead at presentation because of hypotension (a confounding condition) which was persistent until he was certified clinically dead upon cessation of his cardiac activity.

The exact pathogenesis of the white cerebellum sign is yet to be clearly elucidated, but previous authors have postulated several mechanisms for the condition. Some of the proposed mechanisms equivocate the white cerebellum and reversal signs and are thus controversial. They include hypoxia-induced preferential blood flow through the infratentorial compartment, cerebral hyperglycaemia from hypoxic-ischaemic brain injury with preferential damage to the basal ganglia and cerebral cortex, and intracranial hypertension-related obstruction of the venous outflow resulting in dilation of deep medullary veins, hence hyperdensity of the central brain structures.[1,3-7,9] Hypoxia-induced sodium ATPase pump failure causing cytotoxic cerebral oedema in brain regions susceptible to hypoperfusion, and transtentorial herniation from raised intracranial pressure (ICP) with resultant partial ICP reduction, which improves perfusion of the central structures have also been proposed as possible pathophysiological mechanisms for the WCS.[11-13] The observed brain anomaly in our patient may be due to hypoxia and ischemia from respiratory failure and hypotension-induced poor cerebral perfusion respectively. He may also have sustained some hypoxic brain insults during the episodes of post-traumatic seizures he experienced.

The diagnosis of white cerebellum sign is largely made from cranial CT scan findings, as done in the index patient. However, other investigative modalities such as brain magnetic resonance imaging (MRI) and angiography may be employed in the evaluation of WCS. These were not carried out in our patient. Diagnostic brain MRI features of the condition are a hyperintense signal in the caudal cerebellum on diffusion-weighted imaging (DWI) and corresponding restricted diffusion on apparent diffusion coefficient (ADC) sequences, and relatively normal cerebral hemispheres on both DWI and ADC sequences.[14] Absent blood flow in the internal carotid arteries distal to the petrous segment of the vessels are classical cerebral angiographic findings associated with the white cerebellar sign.[12] Worthy of mention is the rarer concept of a "*dark cerebellum sign*" wherein hypodensity of cerebellum and brainstem on cranial CT scan contrasts with the normal density of the supratentorial cerebral structures due to diffuse parenchymal cerebellar oedema and or infarction.[5,15] It has been reported in preterm neonates and tricyclic anti-depressant overdose.[14]

Management of patients with WCS is essentially symptomatic or supportive, aimed at reducing intracranial hypertension, seizure control and reversal of ischemic brain injury.[4] These include administration of intravenous 20% Mannitol, anti-seizure medications (where indicated), and the use of steroids. Craniotomy and evacuation of the offending intracranial bleed (if any exists) may be beneficial in patients with traumatic brain injury, while decompressive craniotomy may be useful in controlling cerebral edema-induced elevated intracranial pressure when medical decompression fails. Few cases of survival documented in the literature include patients with meningitis who received appropriate antimicrobial therapy and also a pediatric patient treated surgically for acute extradural and subdural hematoma.[3,10,14,16] The white cerebellum sign is usually a poor prognostic indicator with mortality in about one-third of patients, while the remaining survivors may have severe longterm neurological deficits with diffuse supratentorial encephalomalacia on neuroimaging.[1,3,5,13] Poor outcome is a consistent finding in the literature, and appears to be commoner amongst patients with anoxic encephalopathy, severe TBI, and status epilepticus.[1-8,10,12-16] For example, four of the five patients reported by Pereira *et al* died (3 TBI-related and 1 following drowning), while the only surviving patient (with metabolic encephalopathy) recovered with a Glasgow outcome score of 3.[7]

Similarly, seven out of the 20 patients reviewed by Han *et al.* died within seven days on admission, while the remaining 13 patients survived with debilitating neurologic deficits and developmental delay.[9] The index patient, who presented 24 hours after severe TBI with associated early post-traumatic seizures and brainstem dysfunction, also had an unfavourable outcome.

CONCLUSION

We have reported the rare but classic white cerebellum sign. It remains a grave prognosticator of cerebral injury and should be sought for in the neuroimaging of patients with acute brain insults.

Patient Perspective

This is not available as the patient died shortly after his admission.

Informed Consent

Not applicable.

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