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Case Report

Coarctation of the Aorta Presenting as Haemorrhagic Stroke in a 3-Year-Old Boy- A Case Report

***Efe Abolodje[1], Francis Chukwuebuka Campbell[2], Stellamaris Nwanneka Etumudor[1], Fred Ofakpo Ejumudo[2], Chukwuemeke Ovili[1]**

[1]Department of Paediatrics, Delta State University Teaching Hospital, Oghara, Nigeria.
[2]Department of Surgery, Delta State University Teaching Hospital, Oghara, Nigeria.

Abstract

Coarctation of the aorta is an obstructive form of congenital heart defects that presents with upper limb hypertension. If untreated, Coarctation of the aorta can lead to left ventricular dysfunction and cerebral vasculopathy. Missed diagnosis due to its subtle presentation is common with attendant complications. This is a case report of an apparently healthy 3-year-old boy who lapsed into a coma after a trivial fall. He had upper limb hypertension and the pulses in the lower limbs were barely palpable. Brain computed tomography (CT) revealed non-traumatic haemorrhagic stroke and echocardiography showed severe left ventricular hypertrophy and severe coarctation of the aorta. Coarctation of the aorta can manifest as chronic upper limb hypertension in children with complicated non-traumatic haemorrhagic stroke. Routine blood pressure measurement in young children can serve as a screening tool for early diagnosis of the condition.

***Correspondence:** Abolodje Efe, Department of Paediatrics, Delta State University Teaching Hospital, Oghara, Nigeria.
Email: efexabo@gmail.com

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Introduction

Coarctation of the aorta (CoA) is any constricted segment of the aorta that is due to localized medial thickening, with some in-folding of the medial and superimposed neo-intimal tissue.[1] The localized segment may form a shelf-like structure or may be a membranous curtain-like structure with a central opening. Coarctation of the aorta (CoA) is among the obstructive congenital cardiac defects, with a prevalence of 5-7% compared to other cardiac defects.[2] This condition can present as heart failure and failure to thrive in infancy or a subtler manner with chronic upper limb hypertension. Diagnosis of the latter form can be overlooked, resulting in complications such as severe left ventricular (LV) hypertrophy, LV dysfunction, aneurysmal transformation, and cerebrovascular disease. This case involves a 3-year-old boy who slipped into a coma after a minor fall, later confirmed to have a haemorrhagic stroke on a CT scan and CoA on an echocardiogram.

Ethical Declaration: The report was carried out after obtaining informed consent from the parents of the patient. The identity of the patient has also been removed to protect their privacy.

Case Summary

The 3-year-old boy had been in good health until he fell while attempting to climb a staircase, becoming unresponsive. He was rushed to a peripheral hospital, where he later experienced multiple episodes of generalized tonic seizures. Subsequently, he had episodes of haematemesis and melena stool. He had no history of sickle cell disease or prior seizures, having reached developmental milestones appropriately before falling ill. On examination, he was deeply comatose with a Blantyle Coma Score of 2/5, pale, febrile with a temperature of 39.0°C, he had pinpoint, poorly reactive pupils, left CN 6 palsy, and reduced tone and reflexes in all limbs.

Upper limb pulses were full, with blood pressure in the upper arm measuring 144/110mmHg, exceeding the 95th percentile for his age, height, and sex. Lower limb pulses were weak, with a blood pressure of 112/45mmHg. The apex beat was on the 4th left intercostal space, laterally displaced from the midclavicular line, accompanied by normal heart sounds without murmurs.

A brain CT scan displayed extensive left frontoparietal intracranial haemorrhage (about 120 mls) with associated edema, midline shift, and periventricular/intraventricular haemorrhage (figure 4). There was no evidence of skull fractures therefore ruling out the possibility of traumatic haemorrhage. Echocardiography revealed severe left ventricular hypertrophy with an ejection fraction of 61.0%, left ventricular mass index of 97.56gm/$m^2$and severe CoA with reduced flow on Colour Doppler interrogation (figure 1, figure 2 and figure 3). Other investigations showed clotting parameters, electrolytes and platelets within normal limits. The overall features were in keeping with spontaneous left Parieto-occipital Hypertensive Haemorrhage (ICH score 4).

Neurosurgeons opted against surgical intervention due to the risk of exacerbating bleeding and poor clinical state. The patient received conservative treatment which included hyperventilation with oxygen therapy, diuretics, anti-seizure medications, sedatives, beta-blockers, and ACE inhibitors. Despite intensive care and ventilator support, the patient succumbed to the illness after experiencing cardiac arrest on the 7th day of management.

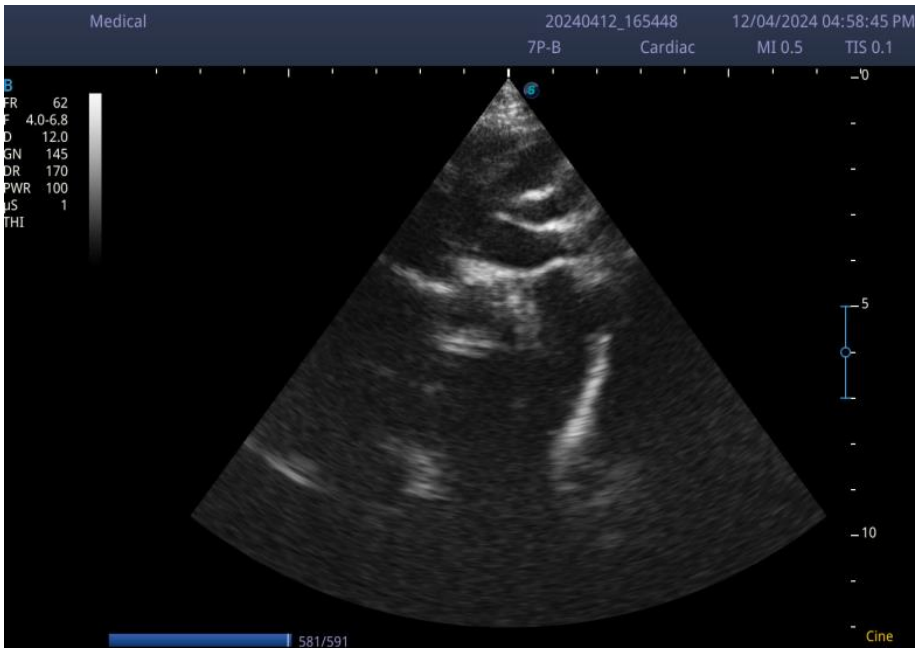

Figure 1: Suprasternal view of Aorta with CoA demonstrated

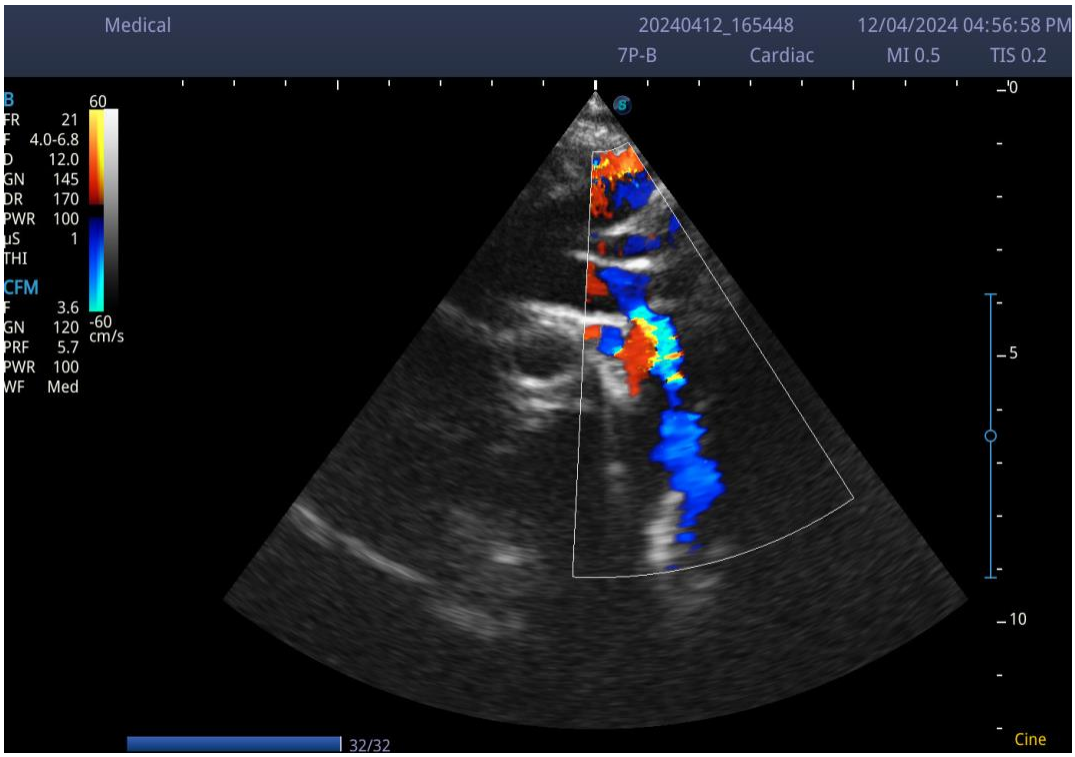

Figure 2: Colour Doppler of Supra-sternal View with Minimal flow across CoA

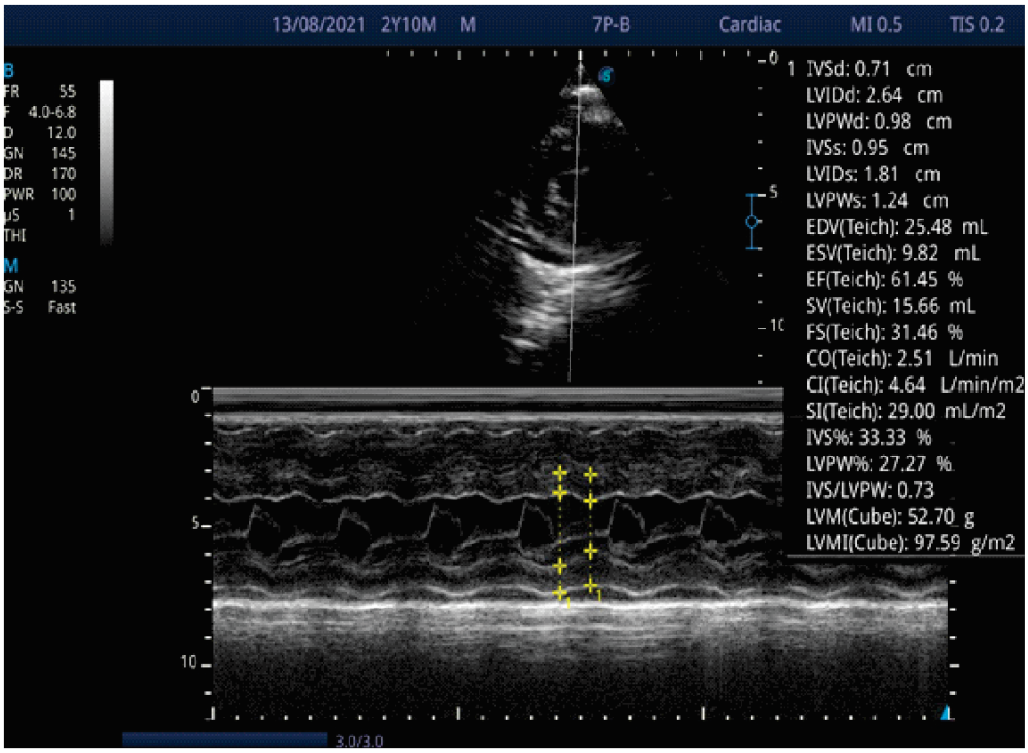

Figure 3: Parasternal Short Axis View with Left Ventricular Hypertrophy

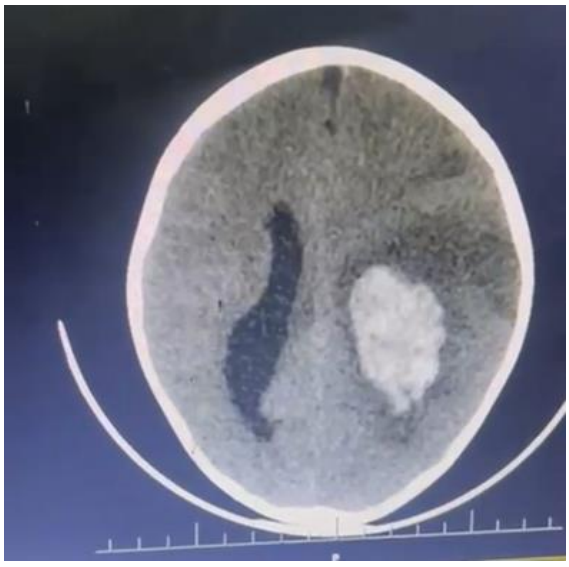

Figure 4: Axial view of Computed Tomography with Intraventricular Haemorrhage

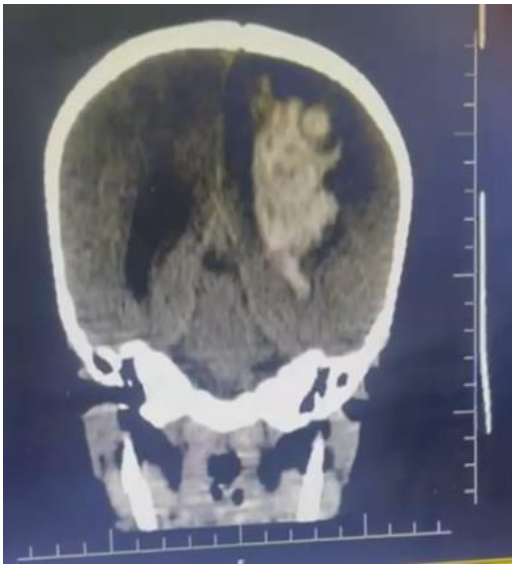

Figure 5. Coronal views of Computed Tomography with Periventricular/Intraventricular Haemorrhage

Discussion

The incidence of paediatric stroke has been reported to range from 1.2 to 13 cases per 100,000 children under 18 years of age.[3] The infrequency of occurrences and subtle presentation often lead to misdiagnosis and delayed diagnosis, as observed in the index case. Delays in making a diagnosis of paediatric stroke can result in poor clinical outcomes.[4] While hypertension is a well-known risk factor for stroke in adults, contributing to over 50% of cases, its role is less observed in paediatric strokes.[3,4]

Sickle cell anemia is highlighted as a significant risk factor for stroke in Nigerian children, whereas cerebral arteriopathies are identified as the leading cause among Caucasian children.[5-7] Coarctation of the aorta (CoA) is a less common cause of paediatric stroke, often presenting as a haemorrhagic stroke when it occurs.[8] Coarctation of the aorta may manifest in infancy as acute heart failure or circulatory collapse, or in older children as chronic upper limb hypertension.[9] It can also present in association with other cardiac defects or as an isolated finding, as seen in the index case.[9] Coarctation of the aorta poses a diagnostic challenge, particularly when presenting with upper limb hypertension due to its subtle nature, necessitating a high index of suspicion. More challenging is the fact that routine blood pressure check in children is not a common practice. Untreated, chronic hypertension associated with CoA can progress to left ventricular hypertrophy, left ventricular dysfunction, and heart failure.[9] In less severe cases of CoA, collateral arteries develop from the internal thoracic arteries and subclavian arteries to supply the lower part of the body. These collaterals are responsible for rib notching seen in CoA typically after 10years of age.[10] The rib notching is an important finding that aids the diagnosis of CoA but it was not seen in the index case because of the early presentation of the case.

Chronic hypertension may lead to fibrenoid necrosis, lacunar infarcts of cerebral vessels, predisposing them to rupture and resulting in haemorrhagic stroke.[11] It can also cause cerebrovascular aneurysmal changes (Charcot-Bouchard aneurysms), which may rupture and lead to haemorrhagic stroke.[11] In the index case, extensive intraventricular haemorrhage was observed with associated brain parenchymal involvement. The presentation of haemorrhagic stroke often includes persistent fever, seizures, and evidence of raised intracranial pressure, as seen in this case.[12] The patient also experienced melena stool and vomiting of blood, likely attributed to stress ulcers, managed with proton pump inhibitors.

Though the median age for development of haemorrhagic stroke in CoA is reported to be 23 years [13], there are reported cases of children with CoA presenting with haemorrhagic stroke. The condition has been reported in a 31-day-old male infant, in a 14-year-old male, in a 15-year-old male, in an 8-year-old female and in a 22-year-old male. [14-17] The index case represents one of the youngest reported cases with CoA-related haemorrhagic stroke. Gender differences may play a role, with males exhibiting a higher prevalence as seen in the reported cases. A review of the admitting blood pressure of the above patients showed that the 31-day old male infant had an admitting blood pressure of 174/95 mmHg, the index case, a 3-year-old male presented with an admitting blood pressure of 144/110mmHg and the 8-year-old female had an admitting blood pressure of 140/90mmH. Also, the 14-year-old boy had an admitting blood pressure of 225/120mmHg, the 15-year-old boy had an admitting blood pressure of 190/50 mmHg, and the 22-year-old boy had an admitting blood pressure of 180/110mmHg. The severity of hypertension seems to correlate with an earlier onset of stroke in preadolescent cases but not necessarily in the adolescent cases.

Paediatric stroke carries a high mortality rate, and the volume of haemorrhage relative to brain volume is deemed an essential predictor of outcome. The estimated haemorrhage volume in the index case was approximately 12% of the brain volume, potentially contributing to the poor outcome. A report indicated that a haemorrhage volume exceeding 4.0% of brain volume is associated with unfavourable clinical outcomes.[18]

Conclusion:

Coarctation of the aorta can present chronic upper limb hypertension in children with complicated non-traumatic haemorrhagic stroke. Routine blood pressure in young children can be a screening tool for early diagnosis.

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