Article 6

PAMJ Clinical Medicine

Case report

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Received: 12 Aug 2023 - **Accepted:** 07 Oct 2023 - **Published:** 18 Oct 2023

Keywords: Invasive intracranial pressure monitoring, craniosynostosis, blindness, case report

Cite this article: Uchenna Ajoku et al. Intracranial hypertension complicated with blindness in a child with non-syndromic sagittal craniosynostosis: a case report. PAMJ Clinical Medicine. 2023;13(18). 10.11604/pamj-cm.2023.13.18.41428

Available online at: https://www.clinical-medicine.panafrican-med-journal.com//content/article/13/18/full

Intracranial hypertension complicated with blindness in a child with non-syndromic sagittal craniosynostosis: a case report

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Abstract

Craniosynostosis is the premature ossification of a cranial suture. Demonstration of intracranial hypertension in neglected craniosynostosis is an important guide to intervention to halt the progression of neurological complications. This case report aims to demonstrate intracranial hypertension in neglected sagittal craniosynostosis with the neurological sequela of blindness. A 9-year-old male with sagittal craniosynostosis and monocular blindness was reported. Clinical assessments were consistent with morbidity associated with neglected craniosynostosis. His Cranial CT showed radiological features of chronic intracranial hypertension such as sulci effacement, flattening of the gyri, and copper-beaten appearance of the calvarium. He had intraparenchymal intracranial pressure monitoring over a 30-hour period. The result of continuous monitoring of intracranial pressure (ICP) was consistent with intracranial hypertension. Although visual loss from intracranial hypertension can be a catastrophic complication of non-syndromic craniosynostosis, invasive intracranial pressure monitoring can act as an objective confirmation of raised ICP which can guide surgical decision-making.

Introduction

Craniosynostosis is the premature ossification of one or more calvaria sutures in an infant. It can occur in isolation or as part of a syndrome complex. Most cases of craniosynostosis occur in an isolated, non-syndromic fashion. About 8% of the time, it occurs as part of a familial syndrome, which may be transmitted as an autosomal dominant trait [1]. The underlying genetic defect being a disruption of the fibroblast growth factor receptor pathway, with the up regulation of the tyrosine kinase activity [2]. The incidence of craniosynostosis is estimated to be 1 in 2000-2500, live births in most Western literature [3]. The exact incidence in sub-Saharan Africa is not known, although the condition is not as uncommon as once believed. Cosmetic concerns are the main reasons parents bring their children to the clinician. However, calvarial growth restriction with resultant intracranial hypertension is a major indication for surgical intervention in single-suture craniosynostosis [4]. The pathophysiologic basis for intracranial hypertension has been described. Venous outflow obstruction at the jugular foramina, increased central venous pressures from obstructive sleep apnoea, and obstructive hydrocephalus from aqueductal stenosis or fourth ventricular outflow obstruction have all been described as contributory factors. The benefit of early decompressive surgery for craniosynostosis is improving neurocognitive outcomes has been debated for over 5 decades. Optic nerve dysfunction and visual loss caused by intracranial hypertension have been documented in the literature and are a rare complication of non-syndromic craniosynostosis [5]. We present a case of neglected sagittal craniosynostosis with monocular blindness and intracranial hypertension which was demonstrated with a 30-hour continuous intraparenchymal Intracranial pressure (ICP) monitoring to emphasize the rationale for early evaluation of ICP, and surgical treatment when necessary.

Patient and observation

Patient information: a 9-year-old male was referred to our clinic with a history of loss of vision in the left eye for 2 years duration. There was an initial complaint of occasional vomiting after eating at the onset of symptoms occurring once to thrice a week, which resolved spontaneously after 3 months. There was also a history of headaches at the time but no seizures, limb weakness, or cognitive impairment. He had no history of trauma, or previous cranial or ophthalmic surgery. He was delivered preterm at 26 weeks and was nursed in the special care baby unit and managed for 12 weeks prior to discharge. He achieved neurological milestones at the appropriate time. However, during his routine follow-up with the

paediatricians, he was noted to have microcephaly, unfortunately, he was lost to follow-up until about 2 years ago when he noticed a loss of vision in his left eye. His academic performance was reported as average. Following a review by the ophthalmologists he was referred to our unit for further evaluation.

Clinical findings: at presentation, he was noted to have obvious scaphocephaly with ridging of the sagittal suture. His vital signs were within normal ranges. Glasgow's coma score was 15 and a mini-mental state assessment was normal. The pupils were 4mm briskly reactive bilaterally. Visual acuity assessment was 6/12 in the right eye but no light perception in the left. His cranial computed tomography scan showed copper beaten appearance of the skull (Figure 1), flattened gyri, and effacement of the sulci and basal cisterns.

Diagnostic assessment: diagnostic testing was done using computed tomography. No challenges were encountered during the diagnostic assessment.

Diagnosis: a diagnosis of intracranial hypertension and monocular blindness from untreated sagittal craniosynostosis was made. His parents were counselled for a 48-hour invasive intracranial pressure monitoring to confirm the diagnosis.

Therapeutic interventions: following admission into the intensive care unit, he was in the supine position, a Raumedic intraparenchymal ICP probe (Raumedic, Germany) was placed through the right Kocher´s point under sedation and local anaesthesia. The distal end of the probe was tunneled through a separate stab incision and then connected to a multiparameter patient monitor (Figure 2). The wound was closed in layers and a dressing was applied. Hourly recording of the ICP values during sleep and when awake was done for the 30 hours of monitoring and are presented in Table 1, while intracranial pressure trends are presented in Figure 3, Figure 4.

Follow-up and outcome of interventions: the results of ICP monitoring were discussed with the patient´s parents, and the decision to proceed with cranial decompression was discussed. However, parents have yet to give consent for surgery.

Patient perspective: the invasive ICP monitoring will help surgeons confirm intracranial hypertension and predict the effect of surgery.

Informed consent: the patient´s parents gave informed consent.

Discussion

In this patient, we were able to objectively demonstrate intracranial hypertension using invasive intraparenchymal ICP measurement done over a 30-hour period. Absolute ICP values showed increased mean ICP while awake and during sleep over the study period (Figure 3, Figure 4). However, the mean ICP value was significantly higher during sleep than while awake (20.64mmHg vs 16.31mmHg, p=0.001). An increase in ICP during sleep has been demonstrated by several studies, while the value of what constitutes normal ICP is still debated, studies that performed overnight ICP monitoring in craniosynostosis all defined ICP of 15 mm Hg or greater as abnormal [6]. One of the consequences of chronic intracranial hypertension is visual loss. This grim sequalae justifies the rationale for early cranial decompressive surgery. Visual loss, although rare, may be a result of direct developmental involvement of the optic nerve or eyeball from deficient bony development (in syndromic cases) or indirect effect from intracranial hypertension. The pathophysiology of papilloedema and blindness from raised ICP have been well described. The two theories that have been proposed are the venous hypertension theory and the axoplasmic stasis theory [7]. The venous hypertension theory (mechanical theory) proposes that intracranial hypertension causes compression of the central retinal vein due to increased pressure within the subarachnoid space. This transmitted pressure progressively causes

venous obstruction and, with time, optic nerve ischaemia. In the axoplasmic theory, raised ICP is transmitted to the axon through the subarachnoid space. This raised pressure causing stasis of intra-axonal fluid and results in the leakage of intra-axonal contents into the extracellular space of the optic disc. Optic nerve swelling and reduced perfusion of axons result in a secondary phenomenon of venous obstruction and dilation and nerve ischemia [7]. The insidious nature of these changes makes visual deterioration go unnoticed, particularly in children. There is first, gradual loss of peripheral nerve fibres which results in field constriction. Subsequently, there is an enlargement of the blind spot, which may then progress to total blindness.

Another consequence of intracranial hypertension, particularly in children, is reports of impairment of brain growth with attendant neurocognitive, and motor dysfunction. These neurobehavioral deficits have also been shown to be mitigated or even prevented by corrective surgery. The consequences of intracranial hypertension were initially thought to be low in non-syndromic craniosynostosis. However, a 2014 study by Wall *et al*. from Oxford found a 44% incidence of raised ICP among 39 patients with isolated non-syndromic sagittal craniosynostosis [8]. A similar study in 2012 study by the same group found a high or borderline ICP in five of seven patients with isolated non-syndromic unilateral coronal craniosynostosis. These findings of intracranial hypertension were obtained through invasive measuring of ICP in the patients´ cohort. In the clinical setting however, ICP evaluations of these patients are done through indirect means using clinical symptoms and signs such as headache, nausea, and vomiting, as well as examination findings of increased occipitofrontal circumference and papilledema on endoscopy. Very recently, optic nerve sheath diameter (ONSD) has been shown to correlate with raised intracranial pressure. Plain skull X-ray and CT scan may show a “copper-beaten” appearance to the skull (this is imprinting of the cerebral gyri on the inner table of the skull caused by chronic intracranial hypertension (Figure 1). Unfortunately, clinical features alone as markers of raised ICP are notoriously unreliable, hence the need for a more objective assessment of intracranial hypertension whenever possible.

As part of surgical evaluation, some institutions perform invasive monitoring of ICP for patients with craniosynostosis. Why this may appear as overkill, Deopujari, and Samantray proposed 4 scenarios for the rationale use of invasive ICP monitoring in patients with craniosysnostosis [9]. The first is when non-operative management is being considered irrespective of the severity of the condition, a recommendation initially put forward by Wall *et al*. The reason for this was the high incidence of raised ICP (44%) in their patient cohort even among patients with mild symptoms [8]. The second is in patients with delayed diagnosis of craniosynostosis as was in our index patient. This formed the basis for our proposed treatment plan. Also, when deciding between a posterior expansive cranioplasty and a frontal-orbital advancement, the use of ICP values has been reported by Tamburrin *et al*. [10] to guide this decision-making. The former procedure was employed in their patients when raised ICP was demonstrated, and the latter procedure was only for cosmetic reasons in patients with normal ICP. Finally, in post-operative evaluation of patients, ICP monitoring may help identify patients with intracranial hypertension after surgery.

Conclusion

Visual loss from intracranial hypertension can be a catastrophic complication of non-syndromic craniosynostosis. When in doubt, invasive intracranial pressure monitoring as an objective confirmation of raised ICP can guide surgical decision-making.

Competing interests

The authors declare no competing interests.

Authors' contributions

Patient management: Uchenna Ajoku, Bright Uche Nwekeala. Data collection: Uchenna Ajoku. Manuscript drafting: Bright Uche Nwekeala. Manuscript revision: Uchenna Ajoku. All the authors have read and agreed to the final manuscript.

Table and figures

Table 1: intracranial pressure values while awake and during sleep during the 30-hours period

Figure 1: copper beaten appearance demonstrated on cranial computed tomography scan

Figure 2: insertion of raumedic intraparenchymal Intracranial pressure probe

Figure 3: intracranial pressure trend when patient was awake

Figure 4: intracranial pressure trend when patient was sleeping

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Table 1: intracranial pressure values while awake and during sleep during the 30-hours period	
ICP Awake(mmHg)	**ICP Asleep (mmHg)**
15	18
17	24
12	23
13	23
23	23
23	24
18	23
17	19
16	17
17	16
11	20
21	17
18	21
13	21
14	_
13	_
Mean ICP	
16.31	20.64
ICP: intracranial pressure	

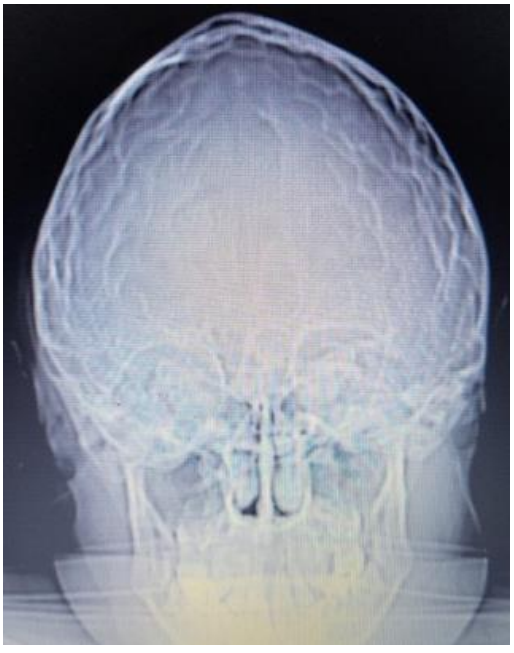

Figure 1: copper beaten appearance demonstrated on cranial computed tomography scan

Figure 2: insertion of raumedic intraparenchymal Intracranial pressure probe

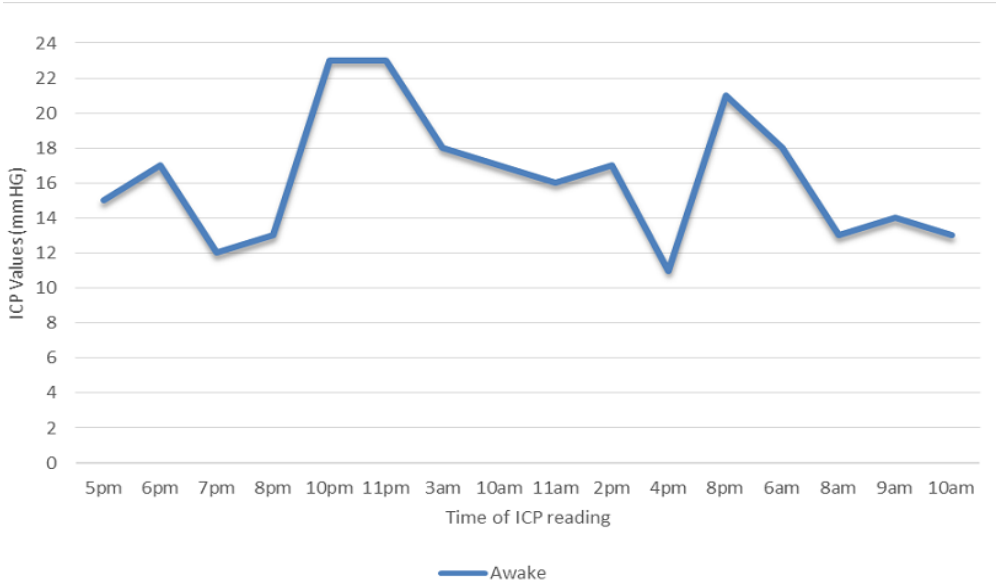

Figure 3: intracranial pressure trend when patient was awake

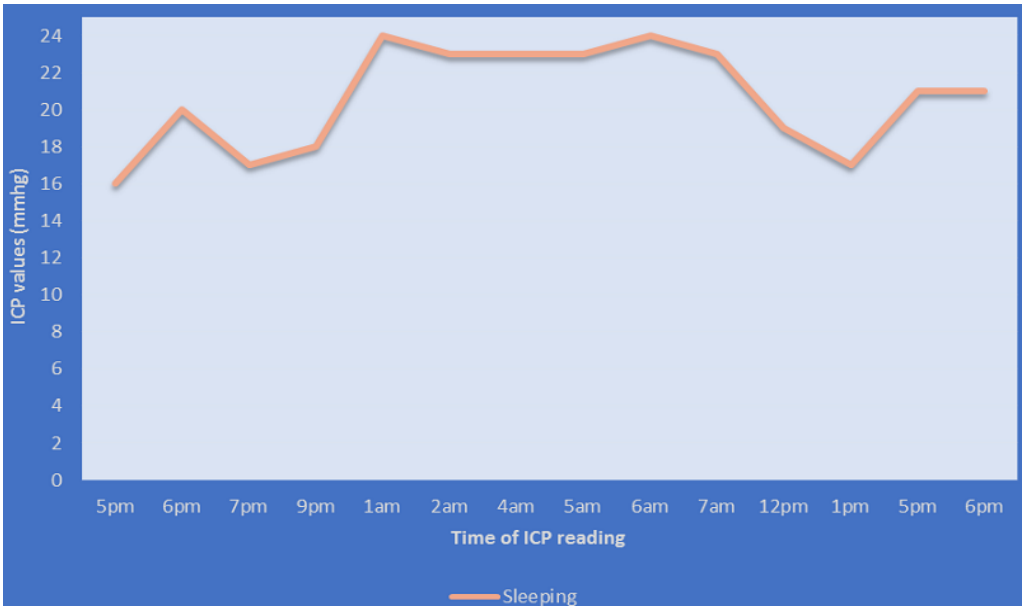

Figure 4: intracranial pressure trend when patient was sleeping