CASE REPORT / CAS CLINIQUE

CRANIAL EXPANSION FOR SHUNT-INDUCED CRANIOSTENOSIS. CASE REPORT

EXPANSION CRANIENNE POUR UNE CRANIOSTÉNOSE SHUNT - INDUITE. A PROPOS D'UN CAS

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RESUME

Introduction

Les dérivations ventriculo-péritonéales sont compliquées par ventricules fendues symptomatiques dans 1-5 % des cas. Quelques patients atteints de fentes ventriculaires développent une craniosténose shunt induite provoquant une augmentation de la pression intracrânienne pendant que le cerveau se développe peuvant être mal interprété comme un problème de dysfonctionnement de shunt. L'expansion de voûte crânienne a été décrit pour traiter cette craniosténose shunt-induite, mais il n'y a pas eu de rapports à ce jour de l'expansion crânienne dans les milieux pauvres en ressources .

Présentation de cas

Nous rapportons un enfant de trois mois qui a subi la reparation d'un myéloméningocèle et l'insertion d'un shunt ventriculo-péritonéal . Quand elle avait quatre ans , elle a développé des symptômes d'hypertension intracrânienne avec une circonférence de 8 cm de la tête en deça de la normale. Les radiographies du crane ont démontré une apparence cuivre battu. Une opération d'expansion crânienne a été effectuée. En post-opératoire, les symptômes d'hypertension intracrânienne ont été résolus et ne se reproduirent plus avec deux ans plus tard des étapes normales de développement , une circonférence supérieure de 5.4cm par rapport au statut préopératoire , et l'amélioration des constats radiologiques .

Conclusion

Les craniosténose shunt induites doivent être pris en charge chez les enfants présentant les symptômes de syndrome ventricule fente. Le diagnostic peut être fait avec des critères cliniques et radiographiques , sans surveillance de la pression intracrânienne , dans les milieux pauvres en ressources , et traité efficacement avec l'expansion de la voûte crânienne .

ABSTRACT

Introduction

Ventriculoperitoneal shunts are complicated by symptomatic slit ventricles in 1-5% of cases. A few patients with slit-ventricles develop shunt-induced craniostenosis, which causes increased intracranial pressure as the brain grows, and may be mis-interpreted as a primary shunt problem. Cranial vault expansion has been described previously to treat shunt-induced craniosynostosis, but there have been no reports to date of cranial expansion in resource-poor settings.

Case presentation

We report a three-month old child who underwent myelomeningocele repair and insertion of a ventriculoperitoneal shunt. When she was four years old, she developed symptoms of increased intracranial pressure, had a head circumference 8cm below normal, and skull radiographs demonstrated a beaten-copper appearance. A cranial expansion operation was performed. Post-operatively, her symptoms of increased intracranial pressure resolved and did not recur. Follow-up two years later revealed normal developmental milestones, no symptoms of increased intracranial pressure, a head circumference 5.4cm greater than preoperatively, and improved radiologic findings.

Conclusion

Shunt-induced craniostenosis should be considered in children with symptomatic slit-ventricle syndrome. The diagnosis can be made with clinical and radiographic criteria, without intracranial pressure monitoring, in resource-poor settings, and treated effectively with cranial vault expansion.

INTRODUCTION

Ventriculoperitoneal shunts (VPS) often result in over-drainage of cerebrospinal fluid (CSF), particularly shunts with differential pressure valves. Approximately 1-5% of children with slit ventricles become symptomatic and present with a spectrum of symptoms that range from intermittent chronic headaches to seizures and coma (1-4). Their symptoms usually develop several years after shunt insertion and are often treated with several shunt revisions (1). Some children with slit-ventricle syndrome have patent, functioning shunts and their symptoms of increased intracranial pressure (ICP) are caused by neither shunt over-drainage nor by under-drainage, but rather by raised pressure secondary to shunt-induced craniostenosis, the inability of the skull to expand and accommodate brain growth because of functional fusion of cranial sutures

We present the case of a four-year old child who had increased intracranial pressure secondary to shunt-induced craniostenosis and her treatment in a resource-poor setting.

CASE REPORT

A four-year old girl was brought in by the mother to Kijabe Hospital, Kenya. She was born with a lumbosacral myelomeningocele and had repair of the myelomeningocele and insertion of a medium pressure Chhabra shunt when she was three months old. The mother reported that the child had worsening irritability and generalized malaise during the previous three weeks. She had regression of her developmental milestones, with difficultly speaking, refusal to crawl and difficulty going from a lying position to a sitting position. She had no vomiting but her oral intake had markedly reduced and she only took liquids. There was no history of fevers or convulsions.

On examination, she was microcephalic; her head circumference of 43.6cm was 8cm below the 2nd percentile. She had mild right esotropia and her upgaze was normal; ophthalmoscopy was not performed. Other cranial nerves were normal. Her neck was supple but in slight hyperextension. Her upper limbs were normal and she was paraplegic. Skull x-rays revealed no abnormality of the shunt but demonstrated severe diffuse beaten-copper changes, indicative of chronically increased intracranial pressure (ICP). Figures 1a and 1b show the preoperative imaging. The Chhabra shunt valve was punctured with a 22 gauge needle and CSF could be aspirated easily into a syringe, confirming patency of the ventricular catheter. A manometer was not available to measure ICP. Her skull x-rays demonstrated severe diffuse beaten-copper changes, indicative of chronically increased ICP.

A diagnosis of shunt-induced craniostenosis was made and a cranial expansion was recommended. She was positioned supine with her head supported on a horse-shoe headrest. A bicoronal scalp incision was made and a subgaleal dissection was performed anteriorly to the orbital rim and posteriorly to the lambdoid sutures.

Strip craniectomies were performed with a Midas Rex drill, removing 6-8mm strips along both coronal sutures and a 12mm strip along the sagittal suture. Burr holes were made at the pterion bilaterally and were connected with a saw cut just above the orbital rim. The opening resulted in bilateral parietal bone segments and a bifrontal segment. The dura was 'paper-thin' and it tore at places as it was being separated off the skull. Barrel-stave osteotomies were made in each segment of parietal bone. The dura was opened widely in each parietal area and enlarged with Duragen. The scalp was closed in two layers. Minimal bleeding occurred intraoperatively and no transfusion was given. The shunt was left in situ and not altered.

Postoperatively, the child's symptoms resolved immediately and she had no complications. Two years post-operatively, she remained asymptomatic, developmentally normal, and her head circumference was 49cm, 5.4cm greater than pre-operatively. The skull radiographs at two years postoperative demonstrated a more normal globoid appearance (figures 2a and 2b).

DISCUSSION

Shunt-induced craniostenosis is characterized pathologically by the presence of cartilage and islands of bone within fibrous cranial sutures, in contrast to the complete bony union of adjacent cranial bones, without intervening cartilage, that characterizes craniosynostosis (1,2).

The development of craniostenosis in children with slit ventricles is attributable to functional shunts causing chronic dampening of normal intracranial pressure waves, which are a fundamental cause of skull enlargement. Without those waves, cranial bones adjacent to sutures do not separate normally; they continue to ossify along sutures and then within them. In time, the ossified sutures lose their ability to allow separation of adjacent bones. Later, as brain enlarges with growth, the non-expansile skull causes increased intracranial pressure (ICP) and the radiographic hallmarks of raised pressure develop (1,3).

Shunt-induced craniostenosis must be differentiated from symptomatic slit-ventricle syndrome, which is far more common and a consequence of chronic overdrainage of CSF because of siphoning. The poor brain compliance associated with slit ventricles further diminishes the normal brain expansion forces on the skull (2). This chronic overdrainage of CSF explains the choice of treatment by some neurosurgeons to change the valve pressure or to insert anti-siphon valves as initial treatment in some patients (1, 2). Sandler et al (4,5) describe this entity as craniocerebral disproportion (CCD) when the available intracranial space is less than the volume of brain. They described need to create space for the developing brain. These patients have slit-ventricles with functioning shunts and have increased intracranial pressure evidenced by continuous ICP monitoring (4,5).

Patients with shunt-induced craniostenosis usually have shunts done at age of less than 6 months and present with symptoms later in childhood (1,9). Chronic severe headache is a common symptom in older children (4) and Rekate (1,6) described five conditions causing headaches associated with small ventricles in patients with shunts as (1) intracranial hypotension caused by overdrainage, (2) intermittent proximal obstruction secondary to overdrainage, (3) shunt failure without ventricular enlargement (4)increased ICP with a functional shunt and (5)shunt-related migraine(controversial). Other signs and symptoms include seizures and blindness which may rarely be present. Some may present acutely with coma or lethargy. On examination, bradycardia, papilloedema and microcephally may be present (1, 3). Children may also present with bizzare behavior and neuropsychiatric behavior with reduced performance in school as the main symptoms (4).

Investigations done to check features of increased ICP include lateral view skull radiographs, computed tomography of the head, isotope shuntogram. Confirmation of increased ICP may be done with manometry via a shunt tap or with ICP monitoring, which may require longer than five days of monitoring (4). The use of ICP monitoring is useful in centres where available. Patients with low ICP, chronic headaches and slit-ventricles may benefit from anti-siphon devices (4). In our case, shunt-induced craniostenosis was suggested by the clinical presentation, the sub-normal head circumference, the patent ventricular catheter, and the radiographic beaten-copper appearance of the skull.

Various surgical techniques of cranial vault expansion have been described to manage post-shunt craniostenosis. These include traditional decompressive techniques, cranial-facial advancement techniques and spring-assisted remodeling methods (1,3,4, 8-11). The earliest benefit of increasing intracranial volume was shown by Epstein et al (7) who reported bilateral temporal decompression in patients who had slit ventricles and had multiple shunt revisions. Biparietal decompressive craniotomy techniques have also been used as a technique of cranial vault expansion (11). The spring-assisted (10) or distraction osteogenesis techniques (4) have been reported to have less blood loss and less morbidity but they need strict monitoring and continued adjustments. They also need ICP monitoring, are expensive and not available in most countries. A combination of cranial vault expansion and changing shunt to programmable valves in patients with post-shunt craniosynostosis and increased ICP on the monitors has been described (9).

Cranial expansion has been shown to have good results as evidenced in the literature. All reported patients had improved neurologic function and all improved cranial vault shape (1,3). Cranial expansion also reduces

the rate of shunt revisions. Albright and Kabara (1) reported five children in their series who underwent cranial expansion for shunt-induced craniostenosis. Two children had symptomatic Chiari 1 demonstrating high intracranial pressures. All did well post-operatively except one child who underwent a bi-frontal advancement two years later. Weizweig et al (3) reported on twelve children who underwent cranial expansion. Of these, two had previously undergone subtemporal decompression. All patients had symptoms recover and only three out of the twelve patients underwent secondary expansion, one having it nine years after the initial. Sandler et al (4) also described a case of a child who had a functioning shunt and developed symptomatic Chiari 1 and had posterior fossa craniectomy at age of 3 years and cranial expansion at 5 years with evidence of increased intracranial pressures on clinical evaluation, imaging and ICP monitoring. The post-operative CT imaging showed cranial volume increase with cerebral volume increase. This shows the benefit of cranial expansion in these subset of patients who develop craniostenosis post-shunting which limits brain volume expansion as they grow.

CONCLUSION

Cranial expansion is an effective surgical technique in treating patients with shunt-induced craniostenosis and can be used by neurosurgeons even in developing countries with few resources. The clinician should be aware of the signs of slit ventricles and craniostenosis in patients with functioning shunts. The case demonstrates that simple investigations like radiographs can be used in resource-poor setttings without ICP monitoring facilities.

ACKNOWLEDGEMENT
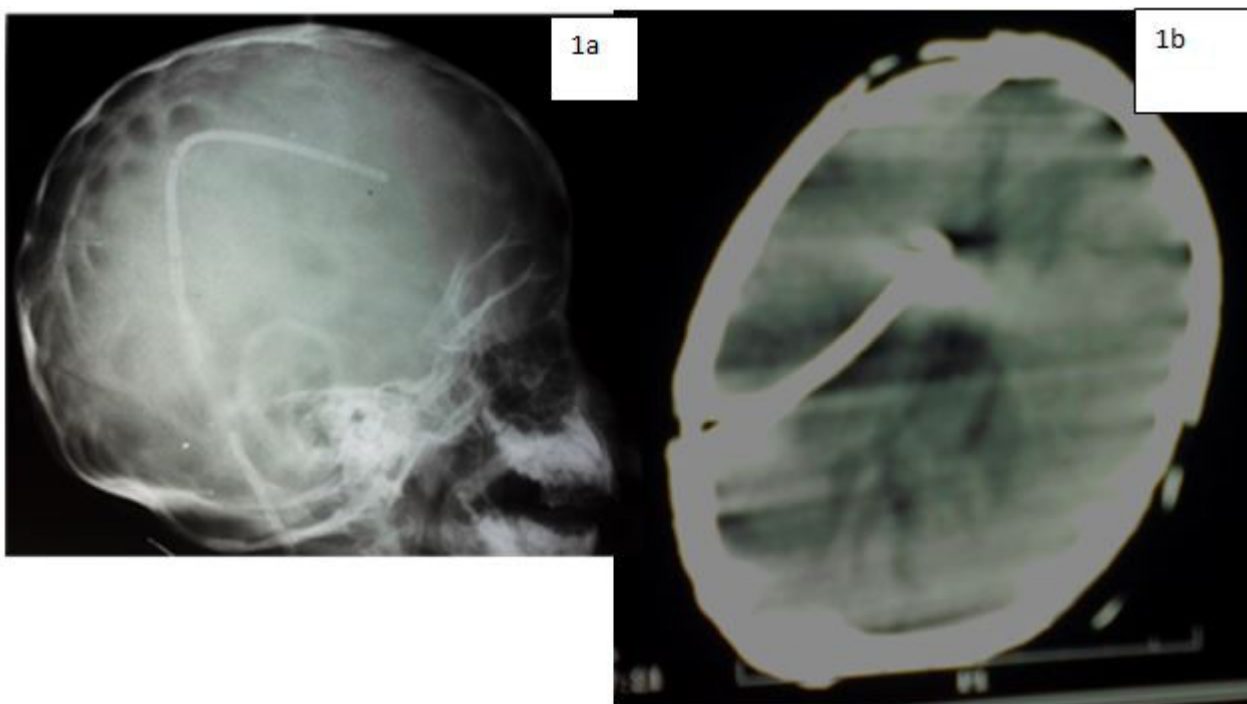
Figure 1

Figure 1a: Preoperative lateral radiograph demonstrating marked beaten-copper appearance of the skull and the ventricular cathether in situ

Figure 1b: Axial CT scan demonstrating slit ventricles with the ventricular catheter tip in the right frontal horn
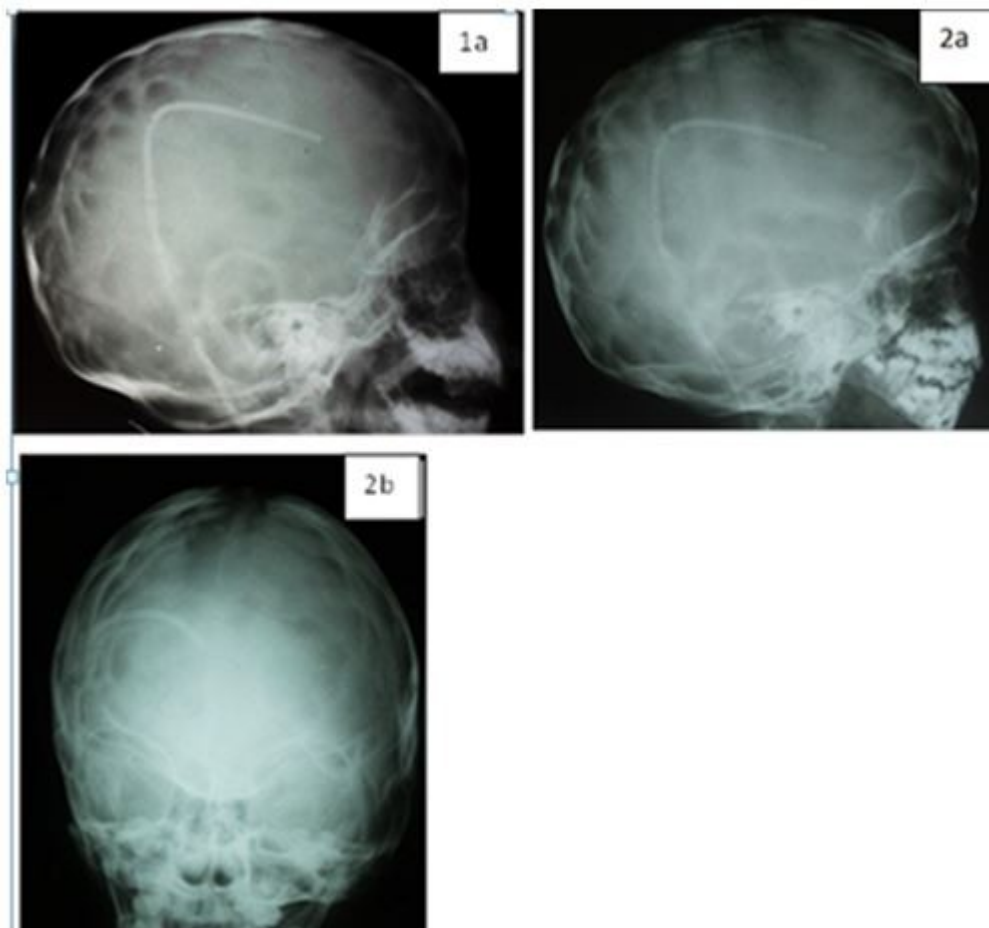
Figure 2

Figure 2a: Postoperative lateral radiograph demonstrating a more globoid head shape, reduction of the beaten copper appearance, and residual craniectomy defects. (compared with Figure 1a - preoperative)

Figure 2b: Postoperative anterior-posterior radiograph demonstrating the parasagittal craniectomy defect

Conflit d'intérêt	Aucun

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