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ORIGINAL PAPER

The evolution of cerebellar tonsillar herniation after cranial vault remodeling surgery

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Abstract
Purpose We sought to examine the pre- and postoperative changes of cerebellar tonsillar herniation by MR imaging in asymptomatic pediatric patients with nonsyndromic, single-suture craniosynostosis (N-SSSC), who underwent cranial vault remodeling surgery without suboccipital decompression. We required cerebellar tonsillar herniation through foramen magnum ≥3 mm for Chiari type I malformation (CMI). We hypothesized that the increase of intracranial volume by cranial vault remodeling would correct the asymptomatic CMI.

Methods We identified 9 patients among 121 N-SSSC children undergoing craniofacial surgery from January 2004 to October 2010 with CMI. However, two of them were excluded from the study due to missing postoperative MR images. In the final study population, six were males, five were scaphocephalic, while two were diagnosed with coronal synostosis.
Results In four of the cases, the CMI was decreased in postoperative MR imaging varying from 6 to 12 mm. In three cases, the herniation remained stable. The median change of cerebellar tonsillar herniation was −6.5 mm.
Conclusions We conclude that asymptomatic patients with existing CMI may benefit from cranial vault remodeling surgery alone increasing the intracranial volume.

Keywords Decision making · Chiari I malformation · Craniosynostosis · Operative treatment

J. Leikola and A. Hukki contributed equally to this study.

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Introduction

Chiari type I malformation (CMI) is coined to reflect cerebellar tonsillar herniation downward through the foramen magnum into the upper cervical spinal canal [21]. It is a common incidental finding, with estimates of about 0.75 % of the population [15, 27]. Etiology of cerebellar tonsillar herniation is most likely multifactorial, in many cases as a result of hind brain growth and small posterior fossa [7, 17, 18]. The acquired variants of CMI have been described in connection with many different conditions including shunt overdrainage, increased intracranial pressure, and chronic hydrocephalus [24]. CMI has also recently been recognized as a potential sequel of cranioplasty [26]. Currently, there is no true consensus as to what constitutes a cerebellar tonsillar herniation regarded as CMI. Many define 5 mm of tonsillar

descent as a minimum criterion [2, 16] for CMI, whereas others suggest that 3 mm, or less, might still be consistent with pathological CMI [4, 21].

CMI is commonly associated with craniosynostosis, both multisuturic and syndromic [7]. In addition, single-suture nonsyndromic craniosynostosis associated with CMI has been reported. Most of these cases involve the lambdoid suture [5, 26]. CMI has been reported even in conjunction with sagittal, metopic, and unilateral coronal synostosis [11, 22, 29]. In both multi- and single-suture craniosynostosis, the cranial base growth is distorted, even in the absence of premature fusion of a cranial base synchondrosis [25].

The optimal management of CMI in the setting of craniosynostosis is not well established as some advocate a simultaneous correction of craniosynostosis and CMI decompression, while others recommend that CMI should only be treated surgically if symptomatic or associated with a syrinx [7, 9]. Craniofacial disorders that lead to a small or deformed posterior fossa may result in the congestion of posterior fossa and tonsillar herniation [6]. We hypothesized that the increase of intracranial volume by cranial vault remodeling surgery would correct the asymptomatic CMI.

Patients and methods

Patients with nonsyndromic, single-suture craniosynostosis (N-SSSC) and CMI were identified by a retrospective analysis of brain magnetic resonance imaging (MRI) of children undergoing craniofacial surgery from 1 January 2004 to 15 October 2010 at the Cleft Palate and Craniofacial Centre, Department of Plastic Surgery, Helsinki University Hospital, Finland. During the study period, altogether 121 patients had brain MR imaging before craniofacial surgery [10]. We routinely schedule all our craniosynostosis patients for MRI. Majority (78 %) were male, with a mean age of 21 months. The most common craniosynostosis type was sagittal synostosis (74 %), followed by coronal (9 %), metopic (11 %), and lambdoid (6 %) synostosis.

We required for cerebellar tonsillar herniation through foramen magnum ≥3 mm descent. This resulted in nine patients. In this study population, there was only one (11 %) female patient. All patients were and had previously been asymptomatic of their cerebellar tonsillar herniation.

All the patients in this study underwent expansive cranial vault remodeling surgery. Based on preoperative neurosurgical evaluation, none of the patients was thought to require decompressive surgery for CMI. Asymptomatic CMI is generally not an indication for suboccipital decompression in our institution.

The MRI studies were performed with a GE Signa HDx 1.5 Tesla MR unit. We obtained T1-weighted spin echo (SE) images in sagittal and axial planes and T2-weighted SE images in axial and coronal planes. Patients were sedated during both procedures. The pre-and postoperative MR images were compared from sagittal planes, and the alteration of tonsillar herniation was expressed in millimeters. An experienced pediatric neuroradiologist (LV) confirmed the cerebellar tonsillar herniation diagnosis in all the MR images. For study purposes, all the images were re-evaluated consecutively in a single session by one researcher (AK) to ensure similar and comparable measurements.

Results

Postoperative MRI was not performed for two patients, reducing our study population to seven patients. In one case, the postoperative MR imaging was prevented by a metallic cranial fixation material used in a previous surgery and, in the other case, by continuous upper airway infection that hampered with general anesthesia. Table 1 presents the demographic data of these seven patients. None of the patients was diagnosed with other associated abnormalities.

The detailed review of the clinical files revealed that psychomotor abnormalities, mostly abnormalities in speech, behavior, and motor skills, were documented in five of the seven patients included in this study. These psychomotor symptoms were not regarded as a manifestation of CMI. In all cases, the psychomotor disturbances were diminished or normalized after the cranioplasty operation.

In preoperative MR images, the cerebellar tonsillar herniations ranged from 6 to 12 mm (Fig. 1). In four of the cases, the cerebellar tonsillar herniation decreased and in three remained the same. The median change of cerebellar tonsillar herniation was −6.5 mm. No adjoining syrinx was recorded in the postoperative MRIs.

Discussion

Herein, we analyzed the outcome of seven nonsyndromic single-suture craniosynostosis patients with CMI that were managed with cranial remodeling surgery solely. Based on radiological imaging, the cranial vault remodeling surgery enlarging the intracranial volume ensured advantageous effect on cerebellar tonsillar herniation on all the seven patients included in this study. We have previously recognized 5.6 % prevalence of CMI malformation in N-SSSC patients (CMI) [11]. In our previous article, we identified seven patients with CMI, the additional 18 months in this current study [10], added two cases of CMI and both pre- and postoperative MR images. The preoperative brain MRI is routinely used in our department; however, the postoperative imaging is not.

Very recently, Levitt et al. reported a case of CMI and cervical syrinx in a syndromic craniosynostosis patient,

Table 1 Clinical data of the seven patients with preoperative Chiari I malformation who underwent cranialvault remodeling surgery

Patient number	Age at operation, months/gender	Time between operation and postoperative MRI, months	Craniosynostosis	Cranioplasty technique	Cerebellar tonsillar herniation change, mm
1	13/F	12 and 81	Coronal synostosis	Fronto-orbital advancement	11–11 (±0)
2	42/M	54	Scaphocephaly	Posteriorly increased intracranial volume cranioplasty	6–3 (−3)
3	63/M	35	Scaphocephaly	Posteriorly increased intracranial volume cranioplasty	12–0 (−12)
4	11/M	60	Coronal synostosis	Fronto-orbital advancement	10–4 (−6)
5	15/M	27	Scaphocephaly	Posteriorly increased intracranial volume cranioplasty	8–8 (±0)
6	74/M	33	Scaphocephaly	Posteriorly increased intracranial volume cranioplasty	12–7 (−5)
7	48/M	55	Scaphocephaly	Posteriorly increased intracranial volume cranioplasty	9–9 (±0)

managed only with posterior cranial vault surgery without suboccipital decompression [12]. The follow-up revealed complete resolution of CMI and decreased syrinx. In our experience, the volume gain due to fronto-orbital advancement is inferior in comparison with a total cranial vault remodeling to correct scaphocephaly in young patients. In this small series, only two patients had fronto-orbital advancement. In the postoperative MR images, one was recorded with stable herniation and the other with improved tonsillar location. In light of our findings, it seems that posteriorly focused cranioplasty is more beneficial in comparison with fronto-orbital advancement for patients with cerebellar tonsillar herniation.

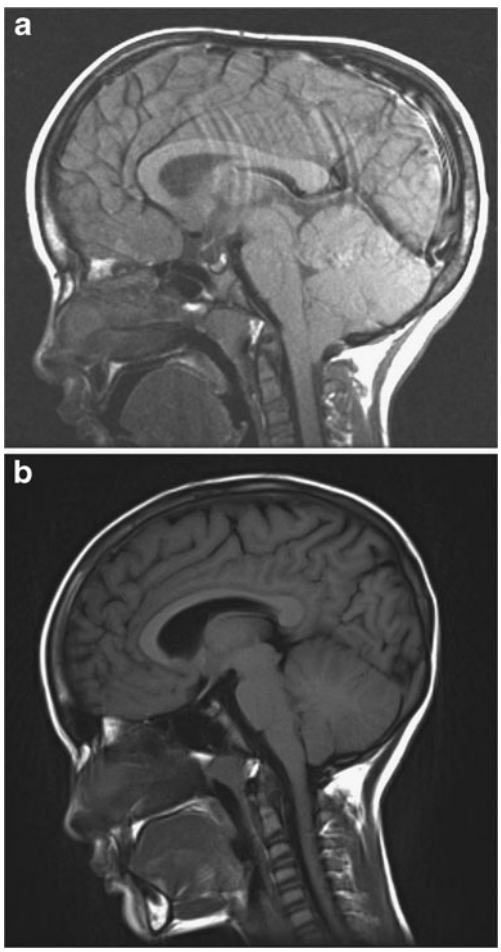

Fig. 1 **a** Preoperative image of Chiari I malformation with a 12-mm descent. **b** Postoperative picture of the same patients, with no Chiari I malformation

The natural history of CMI has been clarified in series in which the entity was managed without surgery [14, 19, 20, 28]. Regarding pediatric patients with CMI, Novegno et al. reported 22 children with incidentally detected CMI, of whom 17 were asymptomatic or improved over follow-up from 3 to 19 years [20]. Radiologically, only four patients showed a decrease (one complete resolution and three improved) of their CMIs. Aitken et al. identified 51 children with CMI among 5,248 head and spine MR images, providing prevalence of 1 % [1]. Of these, 19 were asymptomatic on presentation, and during the 6.4 years of mean follow-up, only 4 developed new neurologic problems. They did not include radiological follow-up in their study. Massimi et al. analyzed 16 asymptomatic children with incidental CMI findings [14]. During the follow-up, three patients showed appearance of the symptoms. Radiologically, the majority of CMIs were stable, with only one case of ascending tonsils.

All preoperative MRIs were performed according to our routine protocol, and the subsequent cerebellar tonsillar herniation findings can therefore be considered as incidental. It is to be noted that all our patients were asymptomatic preoperatively and to our knowledge have remained so during the postoperative follow-up period. CMI may be asymptomatic [30]. The percentage of asymptomatic patients is relatively high; Saletti et al. presented a series of 65 patients, with 58 % asymptomatic [24]. The increased use of neuroimaging tools makes the diagnosis of CMI more

frequent with high rate of asymptomatic patients. There is no consensus on what is the true natural history of patients with asymptomatic CMI [14]. Spontaneous resolution of CMI is an uncommon but possible phenomenon [20].

On the other hand, CMI can present with a variety of symptoms and signs ranging from a slight headache or neck pain [1, 24] to other neurologic symptoms including visual disturbances, vertigo, and ataxia [17, 31] or even severe myelopathy and brainstem compromise [23]. The psychomotor abnormalities recognized in this study can be also addressed to craniosynostosis. Nonsyndromic craniosynostosis is often associated with cognitive, speech, and/or behavioral abnormalities [3, 13].

None of the patients in this series presented with syrinx. Based on the literature, syrinx is associated with symptomatic CMI, up to 76 % [8]. Among asymptomatic patients, syrinx remains infrequent. Meadows et al. identified only 1 case of CMI and syrinx among 21 patients, resulting in a prevalence of 4.8 % [15]. It appears that no relationship exists between the extent of tonsillar herniation and the dimensions of the syrinx or remaining spinal cord [15]. Although no theory has gained full acceptance, it seems clear that altered CSF dynamics play a significant role in the development of syringomyelia.

In conclusion, asymptomatic patients with diagnosed, existing cerebellar tonsillar herniation appear, in the light of MR imaging, to benefit from cranial vault remodeling surgery with an increased intracranial volume. As the radiological resolution of CMI is rare, the effect of cranioplasty operation is plausible. It seems that especially posteriorly focused intracranial volume increase has a superior impact on the resolution of cerebellar tonsillar herniation, and CMI decompression can be reserved only for cases refractory to cranioplasty. However, the role of cranioplasty as an etiological or escalating factor remains unresolved.

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