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CLINICAL ARTICLE

Asymmetric laterality of Chiari type I malformation in patients with non-syndromic single-suture craniosynostosis

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Abstract
Background Chiari type I malformation is a frequent incidental finding commonly associated with craniosynostosis. However, there seems to be a paucity of literature concerning the asymmetry of tonsillar herniation in patients with non-syndromic single-suture craniosynostosis.
Methods To study the asymmetry in this cohort, measurements of the right and left tonsils were made from sagittal images from both pre-operative and post-operative images from 11 patients with non-syndromic single-suture craniosynostosis.
Results Pre-operatively, the mean difference between the caudal descent of all tonsils ranged from 0 to 7 mm, with a mean difference between sides of 2.45 mm. In three cases, cerebellar tonsils were symmetrically herniated. Post-operatively, the mean difference between caudal descent of all tonsils ranged from 0 to 4 mm, with a mean difference between sides of 1.45 mm. Four were symmetrically herniated. In patients with non-syndromic single-suture craniosynostosis, the tonsillar herniation is asymmetric in the majority of cases.
Conclusions Asymmetry of cerebellar tonsil herniation is a frequent finding in this cohort. The right tonsil is more inferiorly located in majority of cases, with predominance to the synostotic suture side in asymmetric craniosynostosis cases.

Keywords Chiari I malformation · Tonsillar ectopia · Asymmetry · Right

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Introduction

Chiari type I malformation (CMI) is characterized by a downward descent of the cerebellar tonsils through the foramen magnum into the upper cervical spinal canal with displacement of the cerebellar tonsils more than 5 mm [1]. It is a common incidental finding, especially in the pediatric population [2]. Moreover, CMI is commonly associated with craniosynostosis, in single-suture, multisuturic and syndromic forms of craniosynostosis [4, 8]. CMI is caused by hind-brain growth and small posterior fossa [4, 10, 12]. The acquired variants of CMI have been described with increased intracranial pressure and chronic hydrocephalus [15]. In syndromic craniosynostosis, CMI is considered secondary to a disproportion between the posterior fossa and the growing hind-brain structures [12, 16]. The same pathogenesis of CMI is supposed to apply to a single-suture craniosynostosis [13]. We have previously shown that the

morphometric measurements of the foramen magnum in children with CMI are smaller as compared with children without CMI in pediatric non-syndromic single-suture craniosynostosis (N-SSSC) cohort (Leikola et al. 2012*, in press* Acta Neurochirurgica) (Fig. 1).

Magnetic resonance imaging (MRI) has become the standard in the radiological diagnosis of CMI patients. In most cases, the assessment of CMI is done by measuring a mid-sagittal distance from the plane of the foramen magnum to the inferior pole of the cerebellar tonsil from sagittal images. However, cerebellar tonsils are a bilobed structure with distinct left and right sides. To the best of our knowledge, only one previous publication has assessed both tonsils separately in a pediatric CMI population [18].

We hypothesized that the disproportion between foramen magnum and hind-brain structures and the distortion of the skull and skull base as seen in patients with craniosynostosis would result in asymmetrical herniation of the cerebellar tonsils through the foramen magnum. Therefore, our aim was to assess the descent of right and left cerebellar tonsils in a cohort of patients with N-SSSC and CMI.

Patients and methods

Our current study cohort is extracted from our previous study setting; retrospective analysis of 121 children with N-SSSC were examined using brain MRI and operated for expansive cranial vault remodeling surgery, during 1.1.2004–15.10.2010 in Cleft Palate and Craniofacial Centre, Department of Plastic Surgery, Helsinki University Hospital, Finland. We required for pre-operative CMI diagnosis herniation of ≥5 mm. Based on the brain MRI studies, 11 children with both N-SSSC and CMI were recognized. All patients were and had previously been asymptomatic of their CMI. The detailed description of the demographic data of these patients is presented elsewhere [6]. The mean at the time of the imaging in patients with CMI was 44 months. These 11 children underwent expansive cranial vault remodeling surgery.

In our department, pre-operative brain-MRI is routinely used to recognize those patients needing other surgical intervention. Brain MRI has been included in our routine clinical protocol since 2004. All N-SSSC patients are subjected to MRI, none is deliberately left without imaging. The additional information pro-vided by MRI, especially concerning vascular anatomy and intracranial anomalies are better detected from MRIs than CTs. All parents have so far complied with this protocol. Our current study is based on those brain MRI images obtained through our routine clinical protocol. The post-operative brain MRI is not in routine use. In nine cases, a post-operative brain MRI examination was done and the measurements of the tonsils were preformed from those images as well. Regarding the missing post-operative images, the postoperative MR imaging was prevented by metallic cranial fixation material used in previous surgery and, in the other case, by persistent upper airway infection that contraindicated the general anesthesia.

For study purposes, all images were re-evaluated consecutively in a single session by one researcher (AK) to ensure reproducibility with similar and comparable measurements. The extent of cerebellar herniation was measured from the tips of the cerebellar tonsils to a line drawn between the basion and the opisthion separately for the right and left tonsils using GE Centricity Radiology RA 600 v6.1 software (GE Healthcare, Little Chalfont, Buckinghamshire, UK) from sagittal images. The institutional review board approved the study protocol.

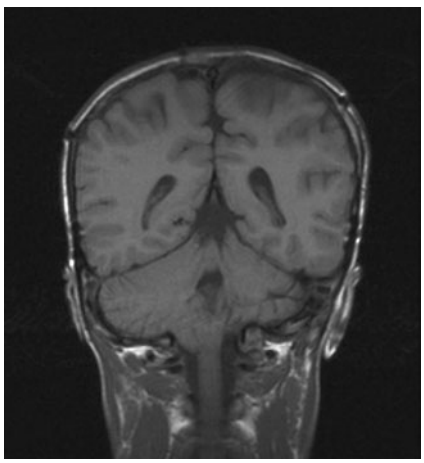

Fig. 1 Coronal image showing an asymmetrical herniation of cerebellar tonsils. The right cerebellar tonsils shows 7-mm descent, while the left cerebellar tonsil is at the level of the foramen magnum with descent of 0 mm in a patient with right coronal synostosis

Results

Craniosynostoses

Of the 11 patients, seven had scaphocephaly affecting the sagittal suture, three had anterior plagiocephaly affecting the right coronal suture, and one patient had posterior plagiocephaly affecting the right-sided lamboid suture.

Symmetrical craniosynostosis

Seven (63 %) of the 11 patients presented with symmetrical craniosynostosis, scaphocephaly. In both pre- and post-operative imaging, three patients of this cohort presented with symmetrical descent. The mean difference between the sides in preoperative imaging was 1.4 mm and in postoperative imaging 1 mm. In preoperative imaging, the mean right

descent was 8.14 mm and on the left 7 mm. In post-operative imaging, the mean right descent was 4.5 mm and on the left 3.5 mm.

Asymmetrical craniosynostosis

Four (36 %) of the 11 patients presented with asymmetrical craniosynostosis either with anterior or posterior plagiocephaly. In this subcohort, two patients presented with left cerebellar tonsil at the level of the foramen magnum. The mean difference between the sides in preoperative imaging was 4.25 mm and in postoperative imaging 2.33 mm. In preoperative imaging, the mean right descent was 8.25 mm and on the left 4 mm. In post-operative imaging, the mean right descent was 6 mm and on the left 3.66 mm.

Pre-operative imaging

The mean difference between the caudal descent of all tonsils (both left and right sides) ranged from 0 to 7 mm, with a mean difference between the sides of 2.45 mm. The largest difference was 7 mm in one case, in a patient with anterior plagiocephaly, followed by 5 mm difference in two cases, in patients with posterior plagiocephaly and scaphocephaly.

The descent of right tonsils had a mean of 8.1 mm (range 5–12 mm). The descent of left tonsils had a mean of 5.9 mm (range 0–12 mm).

In seven (63 %) cases, the right tonsils were found to be more inferiorly located in comparison with those on the left side. In one case (10 %), the left tonsils were found to be more inferiorly located versus those on the right side in a patient with scaphocephaly. Three (27 %) cerebellar tonsils were symmetrically herniated, all in scaphocephalic patients.

Two patients (11 %) had a left cerebellar tonsil that was at the level of the foramen magnum yet the right tonsil was ectopic (5 and 7 mm).

Post-operative imaging

The mean difference between the caudal descent of all tonsils (both left and right sides) ranged from 0 to 4 mm, with a mean difference between the sides of 1.45 mm. The largest difference was 4 mm in one case in a patient with anterior plagiocephaly, followed by 3 mm difference in two cases in patients with posterior plagiocephaly and scaphocephaly.

The descent of right tonsils had a mean of 5 mm (range 1–11 mm). The descent of left tonsils had a mean of 3.5 mm (range 0–8 mm).

In five (56 %) cases, the right tonsils were located more inferiorly versus those on the left side. Four (44 %) of cerebellar tonsils were symmetrically herniated, all these patients had scaphocephaly. None of the left tonsils were located more inferiorly in comparison with the right side.

Similarly, two patients (22 %) had a left cerebellar tonsil that was at the level of the foramen magnum with the ectopic right tonsil (3 and 2 mm).

Pre-operative vs. post-operative imaging

Comparing the pre- and post-operative images showed that the mean difference between pre- and post-operative images was in the right 4.37 mm and in the left 3.5 mm. In two patients, the herniation was symmetrical in pre-operative mages, and in post-operative images, the symmetrical herniation was noted to have preserved in these two patients. Further, in two additional patients, the herniation was symmetrical in post-operative images.

Discussion

Herein, an asymmetrical herniation of CMI malformation in patients with non-syndromic single-suture craniosynostosis was observed. In both pre-operative and post-operative MRI images, the right-sided tonsils were located more inferiorly in comparison with the left side. This finding is in agreement with a previous study by Tubbs et al. [18]. Asymmetrical descent of the cerebellar tonsils is not an uncommon discovery during surgery in both pediatric and adult patients with CMI [5, 7, 11, 14, 17]. Further, it seems that the inferior location of the left tonsil is less common [7, 14]. In those studies addressing the asymmetric herniation of cerebellar tonsils, symmetrical herniation seems to be more uncommon [3].

There is a paucity of data documenting the possible asymmetry or symmetry in cerebellar tonsillar herniation. Extension of the tonsils below the foramen magnum is considered normal up to 3 mm and borderline between 3 and 5 mm [1]. The widely accepted criteria in the literature for CMI call for one or both cerebellar tonsils herniating more than 5 mm below the basion-opistion line [1, 9] although false-positive or -negative results cannot be excluded [9]. Two patients in both image series in this study did not have a left cerebellar tonsil herniation (descent 0 mm) and therefore did not qualify for CMI, yet the other tonsil was undoubtedly ectopic. In this study, the mean difference in tonsillar descent was only 2.45 mm, smaller than recognized by Tubbs et al. [18]. It must be noted that all the descent measurements in this study were also clearly smaller compared with that study. One possible explanation could be that in our study, the patients were considerably younger, with a mean age of 44 months vs. 10.5 years.

The etiology of cerebellar tonsillar herniation is multifactorial. In many cases, it is a result of a hind-brain growth and small posterior fossa [4, 10, 12]. The acquired variants of CMI have been described in connection with many different

conditions [15] including decreased intracranial volume, as seen in craniosynostosis [4]. This study was set out on the premise that the distortion of the cranial vault and cranial base due to craniosynostosis with narrower foramen magnum would result in an asymmetry of cerebellar tonsillar herniation. We indeed identified asymmetric descent in the cerebellar tonsillar herniation. Further, all the patients with asymmetric craniosynostosis, i.e., synostotic suture locating laterally, were right-sided with an increased descent on the right cerebellar tonsil in comparison with symmetrical synostosis patients and with all patients. Therefore it seems plausible to suggest, based on the results of this current study, that N-SSSC is associated with asymmetric descent of the cerebellar tonsils, with predominance to the synostotic suture side. We underline that larger-scale studies are needed to verify this preliminary finding.

The value of this asymmetry is an open question. Tubbs et al. postulated that morphological alteration in the posterior fossa leads to an asymmetrical descent of tonsils that may manifest in a patient's clinical presentation [18]. All the patients included in this study were asymptomatic in regard to their CMI, so we cannot make an assumption on this connection. This theory has not been proven by other studies. Another unresolved issue concerning this finding is whether it affects the diagnostic accuracy. However, when analyzing the images form coronal planes, several images are usually reviewed for determining the maximal descent.

The pathology of CMI is much due to the actual physical compression of the herniation towards the medulla causing clinical symptoms. So far, the extent of CMI has conventionally been evaluated from sagittal images, and the maximal downward cerebellar herniation has been regarded as the degree of CMI. The results of this study erect another perspective to these measurements. It appears that the asymmetry of the tonsils presented here can have a great impact on the actual volume of the tonsillar soft tissue herniation. After all, it is the total of the descending herniating mass that occludes the flow of cerebrospinal fluid and creates the pressure causing clinical symptoms. To further evaluate the pressure and severity caused by the CMI, one must therefore measure the size of both of the herniating tonsils in a 3D format providing the precise volumetric figure of the CMI.

We chose not to perform statistical analysis. Given the limited number of patients included in this study (11/9), we feel that statistical analysis would not have been appropriate due to the fact that the analysis would have been underpowered, and thus no definitive conclusions should be drawn. Further, since this is an observational study, and to the best of our knowledge the first one to recognize this phenomenon in N-SSSC patients, we did not want to set biased premises for future studies.

In conclusion, asymmetry of cerebellar tonsil herniation is a frequent finding in this cohort and the right tonsil is more inferiorly located in majority of the cases, with predominance to the synostotic suture side in asymmetric craniosynostosis cases. Further studies are needed to evaluate the pressure and severity caused by the asymmetric presentation of CMI to cerebrospinal fluid flow.

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Conflicts of interest None.

Author contributions All authors were equally involved in the study design, data analysis, and interpretation. VK wrote the first draft of the manuscript, and all authors were involved in critically revising the manuscript for important intellectual content. All authors approved the final version for publication.

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