Morphometric Factors affecting Functional Outcome in Symptomatic Chiari I Malformation and Syrinx

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ABSTRACT

Objective: To find out the morphometric factors predicting outcome in Chiari I malformation (CM I) associated with syringomyelia.

Materials and methods: In a series of 73 patients with CM I and syrinx who underwent posterior fossa decompression (PFD) between August 2013 and October 2015, a total of 54 subjects with sufficient clinical data and imaging suitable for morphometric measurements were evaluated. The parameters analyzed were posterior fossa volume (PFV), tonsillar descent (TD), foramen magnum (FM) diameter, supra-occiput length, clival length, the syrinx, and cord diameter. Patients were divided into two groups: With or without improvement. Improvement at follow-up was assessed with the Chicago Chiari Outcome Scale (CCOS).

Results: Mean PFV was significantly higher in the improvement group (219.90 ± 30.20 vs 187.95 ± 12.51 cm³, p = 0.047). Syrinx cord ratio was lower in the improved group (0.54 ± 0.21 vs 0.64 ± 0.27, p = 0.081). The cut-off value of preoperative PFV for prediction of improvement was found to be 198.58 cm³ (sensitivity 77.8%, specificity 100%).

Conclusion: To the best of our knowledge, this is the largest series to evaluate the role of morphometry in prediction of surgical outcome in patients with CM I associated with syringomyelia. The PFV is the only radiological factor that differs significantly in patients with and without improvement.

Keywords: Chiari malformation, Chicago Chiari Outcome Scale, Functional outcome, Morphometry, Syringomyelia.

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INTRODUCTION

Chiari I malformation is a fairly common anomaly of hind brain that was first described over a century ago by Hans Chiari during a postmortem examination. Chiari I malformation represents a heterogeneous group of conditions characterized by a >5 mm downward displacement of cerebellar tonsil into FM which may lead to compression, central cord, and cerebellar symptoms. Clinical features are usually due to the combined pathophysiological effect of CM I and syringomyelia.

The patients usually require surgery for long-term presenting symptoms. No definitive criteria are available for surgery. Treatment for asymptomatic patients remain controversial, however surgical decompression is highly recommended in symptomatic patient with cerebrospinal fluid flow obstruction.

Long-term outcome studies are becoming more prevalent, and patients are commonly operated on with generally favorable results. It is difficult to predict who might benefit from Chiari decompression surgery, and as such careful patient selection is perhaps most important to achieve successful outcomes for this population. As per literature, 9.2 to 25% of patients do not show any improvement after surgical intervention. The criteria used to document improvement also varies substantially among various studies. The CCOS is a standardized, Chiari specific, and recently validated tool for evaluating clinical outcome in CM I patients.

In the present study, we measured bony elements of posterior cranial fossa in patients with CM I and evaluated the role of morphometry in predicting surgical outcome of CM I using CCOS. We discuss our findings in the light of pertinent literature.

MATERIALS AND METHODS

In a series of 73 patients with CM I and syrinx who underwent PFD at our institute between August 2013 and October 2015, 54 subjects with sufficient clinical data and imaging suitable for morphometric measurements were evaluated. The parameters analyzed were PFV, TD, FM diameter, supra-occiput length, clival length, the syrinx, and cord diameter. Patients were divided into two groups: With or without improvement. All patients underwent PFD without duroplasty. Improvement at follow-up was assessed with the CCOS. The mean duration of follow-up
was 10.23 ± 5.8 months. The outcomes were evaluated based on the last available follow-up.

**Radiological Evaluation**

The diagnosis of CM I was made using sagittal T1-weighted magnetic resonance imaging (MRI) brain studies, and was defined as tonsillar herniation of at least 5 mm below the level of the FM.

**Magnetic Resonance Imaging Protocol**

Magnetic resonance imaging examinations were performed using 1.5 Tesla scanners. The MRI of the craniovertebral junction and the cranium was used to evaluate TD, to obtain linear dimensions of the posterior fossa, and to estimate PFV.

Out of 73 patients, 19 patients did not have usable MRI for varying reasons (e.g., MRI performed at other hospital and artifacts on MRI).

**Cerebellar Tonsillar Descent**

The line between the basion and opisthion (i.e., McRae’s line) was assumed to represent the planum of the FM. The TD was then evaluated by measuring the distance to the most caudal aspect of the tonsils on a line running perpendicular to the opisthion basion line.

**Measurement of Posterior Fossa Volume**

The method for determining the PFV was based on previously reported methods of measuring posterior fossa. Calculation of PFV was based on a spheroidal formula:

\[ PFV = \frac{4}{3} \pi \times \left(\frac{x}{2}\right) \times \left(\frac{y}{2}\right) \times \left(\frac{z}{2}\right) \]

where \( x \) is the anteroposterior measurement from the posterior clinoid process to the torcula, \( y \) is the height of the posterior fossa measured from the basion to the peak of the tentorium cerebelli, and \( z \) is the maximum width of the posterior fossa.

**Length of the Clivus and Supra-occiput**

The length of the clivus was defined as the distance from the top of the dorsum sellae to the basion, and the length of the supra-occiput was measured between the internal occipital protuberance and the opisthion (Fig. 1).

**Assessment of Syrinx and Cord Dimensions**

Sagittal and axial images of the spine were used to calculate the diameter of the syrinx and cord. Quantitative evaluation of the syrinx diameter was done on an axial image at the level of the maximum diameter of the syrinx. A ratio of the syrinx to the cord diameter was calculated by dividing the maximum anteroposterior diameter of the syrinx by the anteroposterior diameter of the spinal cord at the same level to indirectly quantify the syrinx size in relation to cord diameter as described earlier (Fig. 2).

**Assessment of Functional Outcome**

We used the CCOS originally provided by Aliaga et al for postoperative clinical evaluation. The CCOS is a standardized, recently validated tool for evaluating clinical outcome in CM I patients. This is a total 16-point scale consisting of four categories of postoperative outcome (pain, nonpain symptoms, functionality, and complications) that are graded 1 to 4, which is further categorized in improved (score 13–16), not improved (score 9–12), and worsened group (score <9). To avoid interobserver variability, all patients were graded by the same investigator.

**Statistical Analysis**

All statistical tests were performed using Statistical Package for the Social Sciences, version 22 software (Chicago, Illinois, USA). Quantitative variables were described as
means and standard deviations, qualitative variables as percentages and variables on ordinal scale as medians and interquartile range. Qualitative variables were analyzed between the groups with χ² test or Fisher’s exact test as appropriate. Normally distributed quantitative variables were analyzed using independent samples Student’s t-test between the groups and paired samples t-test for within-group analysis across time points. Non-normally distributed quantitative data and ordinal data were analyzed using Mann–Whitney U test for between-group comparison and Wilcoxon signed ranks test for within-group comparison across time points. Receiver operating characteristic (ROC) curve was created to observe predictability of outcome from baseline PFVs. A p-value of <0.05 was taken as the level of statistical significance.

RESULTS

Demographics

The mean age at diagnosis was 34 years (34.50 ± 13.82, 4–60 years); 7.4% (n = 4/54) were under 18 years of age and 51.85% (n = 28/54) were male.

Neuroradiological Studies

Mean TD below FM was 12.99 ± 5.54 mm (6–27.20 mm).

Linear Measurements of Posterior Fossa

The mean length of the supra-occiput was 3.74 ± 0.59 cm (2.6–6.9 cm), and the clival length was 3.61 ± 0.59 cm (2.2–6.2 cm). The mean FM length was 3.67 ± 0.48 cm. The mean PFV was 218.19 ± 33.98 cm³ (146.60–309.76 cm³).

Difference in Mean Values in the Improved vs Not Improved Groups

Postoperative functional improvement occurred in 83.33% (n = 45) of patients with CM I with syrinx, 16.66% (n = 9) were unchanged and none of them deteriorated (Table 1). The median CCOS was 14 ± 1.34 (11–16). Age, sex, and duration of symptoms had no significant bearing on outcome as assessed by the CCOS (Table 2). Of the various radiological factors analyzed in the two groups, the differences in the mean values of the PFV were statistically significant (219.90 ± 30.20 vs 187.95 ± 12.51 cm³, p = 0.04). Syrinx to cord ratio was lower in the improved group (0.54 ± 0.21 vs 0.64 ± 0.27, p = 0.081). The cut-off value of preoperative PFV for prediction of improvement was found to be 198.58 cm³ (sensitivity 77.8%, specificity 100%; Graph 1).

DISCUSSION

Factors affecting Outcome and Influence of PFV on the Treatment Response

Recently, morphometric studies have revealed the significantly smaller posterior cranial fossa volume in patients
with CM I\textsuperscript{12,14-16}. Milhorat et al\textsuperscript{3} have noted a smaller PFV in adult CM I patients (measured by Cavalieri method). Nishikawa et al\textsuperscript{15} found that the PFV in adult Chiari patients and the PFV in the control group were 186 and 195 cm\textsuperscript{3} respectively. They noted a smaller PFV in the CM I patients. Tubbs et al\textsuperscript{17} found that the mean PFV was 208.5 cm\textsuperscript{3} in Chiari patients. They did not find any statistically significant difference between controls and patients with CM I. Furtado et al\textsuperscript{18} found that the PFV (spheroidal formula) in childhood patients with Chiari malformation was significantly lower than in the control group (p = 0.002).

In our series, mean PFV before decompression was 214.91 ± 30.40 cm\textsuperscript{3}. The mean preoperative PFV, calculated using a spheroidal formula, was significantly smaller in unimproved group than in the improved group (187.95 ± 12.51 vs 219.90 ± 30.20), pointing to an overcrowded posterior fossa. We used an ROC curve to measure cut-off value of preoperative PFV for prediction of improvement. It was 198.58 cm\textsuperscript{3} (sensitivity 77.8%, specificity 100%; Graph 1).

Noudel et al\textsuperscript{18} in their series of 11 patients undergoing decompression without duroplasty measured the increase in PFV (measured by Windows Volume Analysis software, version 4.3) and found that the degree of PFV increase positively correlated with symptomatic improvement (p = 0.014). The authors further suggest that an optimal PFV increase could be predicted to improve patient symptoms, and an increase of at least 15% was needed for a complete recovery, whereas partial recovery is obtained when the PFV is increased by only 7%. The measured preoperative PFV in this study was 156.68 ± 16.0 cm\textsuperscript{3} in patients with complete response (n = 4 patients), while it was 185.12 ± 24.2 cm\textsuperscript{3} (p-value = 0.09) in patients with partial response (n = 7). Our study is strikingly different from the study by Noudel et al in terms of sample size (n = 54 vs n = 11), method of PFV measurement (spheroidal formula vs Windows Volume Analysis software), and functional outcome assessment scale (Chiari-specific CCOS vs functional grading system). Demographic confounders also could be attributed to this difference as suggested by Roller et al.\textsuperscript{19}

In our study, postoperative PFV could not be calculated as we used spheroidal formula to calculate PFV.

Attenello et al\textsuperscript{20} showed no significant correlation between the length of symptoms and symptom resolution. In our study, demographic variables including age, sex, and duration of symptoms did not predict clinical outcome. In a study considering multiple radiological variables, including TD, odontoid retroflexion, and maximal axial width of the fourth ventricle, Tubbs et al found no preoperative factors associated with subsequent radiological treatment failure. Furtado et al\textsuperscript{21} reported that among the different radiological factors, change in cord diameter was the only factor noted to be significantly different in patients who improved and in those who did not, while functional improvement with respect to syrinx size and syrinx/cord ratio was appreciable but not statistically significant. In their study of 20 pediatric patients, mean PFV in not improved group was smaller (213 ± 30 cm\textsuperscript{3}) as compared with improved patients (215 ± 41 cm\textsuperscript{3}); however, it was not statistically significant and result could be influenced by small sample size or only pediatric cohort (p = 0.96).

Of the various radiological factors we analyzed in the improved and unimproved group, the differences in the mean values of the PFV were statistically significant (p = 0.04), while the change in syrinx/cord ratio in the two groups was considerable but not statistically significant (p = 0.08). Future implications of this study can be significant and a lower preoperative PFV could be taken as an important factor in choosing surgical strategy, either duroplasty could be added to this group or larger craniectomy could be planned. This idea should be pursued in future studies.

CONCLUSION

To our knowledge, this is the largest series to evaluate role of morphometry in prediction of surgical outcome. The PFV is the only radiological factor that differs significantly in patients with and without improvement. Although syrinx to cord ratio is not significant, trending might be significant. Outcome does not correlate with demographic and other cranial morphometric measurements or with change in any of the syrinx-related factors. We believe that these correlations and measurements will facilitate in prediction of surgical outcome.

REFERENCES

Kumar et al have done a very nice study examining the value of morphometric variables in prediction of outcomes of Chiari I patients with syrinx. They have used a spheroidal formula to calculate posterior fossa volume (PFV). This is not the first attempt to search for the value of PFV and other morphometric measurements in Chiari type I patients, but this is the most reliable one. They have evaluated patient outcomes using Chicago Chiari outcome scale.

Although the groups are not similar (45 improved, 9 not improved patients), the term “clinically not improved” is not very objective, and this is a retrospective cohort, the surgical outcomes are significantly better with larger PFVs. I believe, the findings of this study will be quite useful in our practice: patients with smaller PFVs, patients with larger syrinx does not benefitted much from surgery. However, the amount of tonsil herniation does not predict the outcomes of surgery. Those statements will be helpful to tell the patients expected outcomes before surgery.

The authors have done a conservative surgery, foramen magnum decompression without duraplasty. The main issue arising after those statements should be the effect of different surgical techniques to outcomes and also to morphometric parameters. Do the volumes of posterior fossa increase after bony decompression, simple duratomy, or with duraplasty? Do we need to perform different surgeries for patients with smaller posterior fossa? It will be necessary to conduct new and larger studies to look at those variables and PFVs described in this study.

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