Adult Chiari I malformation

Clinical presentation and surgical outcomes

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I. Swallowing dysfunction in adult patients with Chiari I malformation; Fawaz S. Almotairi, Mats Andersson, Olof Andersson, Thomas Skoglund, Magnus Tisell. Submitted.

II. Chiari I malformation - neuropsychological functions and quality of life, Fawaz S. Almotairi MD, Per Hellström PhD, Thomas Skoglund MD, PhD, Åsa Lundgren Nilsson PhD, Magnus Tisell MD, PhD, Submitted.

III. Acute deterioration of adults with Chiari I malformation associated with extensive syrinx, Almotairi FS, Magnus Tisell, Submitted.

IV. Cerebrospinal Fluid Disturbance In Overweight Women After Occipitocervical Decompression In Chiari Malformation, Almotairi FS, Magnus Tisell , Acta Neurochirurgica: 2016 Mar; 158(3): 589-94; discussion 594.

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ABSTRACT

Background

Chiari I malformations (CMIs) are hindbrain anomalies that are characterized by cerebellar tonsillar ectopia. The typical presentation is an occipital headache that worsens with exertion. In this thesis, I focus on the following three atypical presentations of CMI: swallowing difficulty, neuropsychological (NP) dysfunction and acute deterioration. In addition, I address the impact of CMI on patient quality of life (QOL). The preoperative factors that predict cerebrospinal fluid disturbance (CSFD) after surgery for CMI have rarely been reported and will be discussed and investigated in this thesis.

Patients and methods

In the first two studies presented in this thesis (studies I & II), patients were prospectively included over a two-year period. Patients underwent both subjective and objective assessments of swallowing function in Study I and of NP functions and QOL in Study II. The total number of patients included were 11 and 14, respectively. All patients were assessed both before and at 3 months after surgery.

In the last two studies presented in this thesis (studies III & IV), patients were retrospectively included over two overlapping 10-year periods. The total number of patients included were 52 and 65, respectively. In Study III, I explore the preoperative radiological factors that indicate a risk of acute deterioration in CMI patients. In Study IV, I investigate preoperative patient characteristics that might determine the risk of postoperative CSFD.

Results

In Study I, four patients (36%) reported varying degrees of swallowing complaints (mean Watson Dysphagia score, 16). In two of these patients, there was substantial penetration of contrast on videofluoroscopy, and in the other two patients, minor disturbances were observed. None of the patients reported remaining symptoms after surgical decompression.

In Study II, the majority of included patients demonstrated cognitive functions within the normal range. However, their postoperative performance on some of the tested NP assessment tools significantly improved.

There was a lower level of satisfaction with QOL both before and after surgery in patients than in healthy subjects In addition, the 5-level Euroqol-5 dimensional questionnaire (EQ-5D-5L) indicated that patient QOL was significantly better after surgery.

In Study III, three patients (4.6%) presented with acute deterioration of symptoms. Additionally, the length and size of the syrinx were higher and it was extended more rostral (above C1 level) in these acute patients than in non-acute patients with CMI.

In Study IV, six patients developed CSFD after occipitocervical decompression (OCD) and subsequently underwent cerebrospinal fluid (CSF) diversion procedures. All of these patients were females, and they had a mean body mass index (BMI) of 32.3, whereas the mean was 24.3 in patients without CSFD (p=0.0011). There was no difference between the two groups in other examined patient characteristics.

Conclusion

Symptoms of dysphagia are not uncommon in CMI patients. A preoperative NP assessment of adult patients with CMI showed that there was a statistically significant improvement in four of the nine tasks tested after surgery. Furthermore, preoperative QOL was poorer in CMI patients than in healthy individuals. Surgery can potentially remedy the causes underlying dysphagia and NP dysfunctions, thereby relieving their symptoms.

Study III shows that it is important to assess the preoperative size, length and rostral extension of the CMI-associated syrinx because changes in these parameters could indicate acute deterioration, and affected CMI patients should be assigned for early surgical decompression.

All patients with postoperative CSFD were female, and their mean BMI was significantly higher than that of patients without this complication. Pseudotumor cerebri must be excluded in this group of patients.

Keywords
Arnold-Chiari Malformation, Deglutition Disorders, Deglutition, Surveys and Questionnaires, Decompressive Craniectomy, Treatment Outcome, Cognitive Dysfunction, Executive Function, Patient Satisfaction, Psychological Tests, Quality of Life, Syringomyelia, Emergencies, Postoperative complications, Hydrocephalus, Body mass index.

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