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ALTERAÇÕES NA DEGLUTIÇÃO DE PACIENTES COM MALFORMAÇÃO
DE ARNOLD-CHIARI TIPO I PÓS NEUROCIRURGIA DESCOMPRESSIVA

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Trabalho para a conclusão da Residência Multiprofissional em Terapia Intensiva pelo Hospital Geral de Fortaleza como requisito para obtenção do título de especialista no caráter residência.

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SWALLOWING DISORDER IN PATIENTS WITH CHIARI MALFORMATION TYPE I AFTER DECOMPRESSION NEUROSURGERY

ABSTRACT

OBJECTIVE: The present study aimed to investigate postoperative swallowing disorders in patients diagnosed with CM I after decompression neurosurgery.

METHODS: This is a retrospective case series from the ICU of the Hospital Geral de Fortaleza. We selected 54 patients after decompression neurosurgery procedure for CM I confirmed by brain MRI, from January 2017 to December 2019. The patients were followed by the speech-language assessment (CSLSA) 24 hours after post-surgical extubation. The CSLSA observed aspects of posture, sensitivity, tone, and extra- and intraoral mobility of the phonoarticulatory organs and visualizing the functions of breathing, phonation, articulation, chewing, and swallowing. We correlated the severity of the dysphagia and the presence of associated malformations in the CM I patients. Moreover, we performed a Fisher's exact test and the chi-square tests and considered statistically significant at p<0.05.

RESULTS: In our study, 39 patients were female (72.3%) and 15 were male (27.7%). The mean age was 41.3 ± 47.1 years. Thirty-seven (68.5%) patients with CM I had associated malformations (p=0.0243): 21 patients had syringomyelia, 11 basilar invagination and 5 syringomyelia and basilar invagination. Oropharyngeal dysphagia was present in 37 of the cases (68.5%; p=0.0016), of which 18 cases were mild, 3 cases moderate, and 16 cases severe. Dysphonia was found in 13 cases (24.1%; p=0.4785) and dysarthria in 3 cases (5.6%; p=0.0042). In the CSLSA results related to swallowing disorders, the following changes were observed: orofacial hypomobility (29 patients–53.7%; p=0.0204), orofacial hyposensitivity (26 patients–48.1%; p=0.3023), reduced GAG reflex (24 patients–44.5%; p=0.1041), ineffective cough (19 patients–35.2%; p=0.0022), ineffective saliva swallowing (18 patients–33.3%; p=0.0396), dyspnea (13 patients–24.1%; p=0.4785), orofacial hypotonia (11 patients–20.4%; p=0.0004), altered cervical auscultation (10 patients–18.5%; p=0.0003), and dysarthria (3 patients–9.3%; p=0.0042). The presence of associated malformations significantly influenced the appearance of swallowing disorders, the severity of oropharyngeal dysphagia, the post-surgical complications (17 patients–31.5%; p<0.001), and the clinical outcomes (p=0.0029) of patients with CM type I.

CONCLUSION: Decompressive neurosurgery for Chiari Malformation type I had low rates of post-surgical complications overall. However, there was a high incidence of oropharyngeal dysphagia. The presence of associated malformations is associated with poorer prognosis and greater severity of oropharyngeal dysphagia in patients with Chiari Malformation type I.
INTRODUCTION

Chiari malformations (CM) are a group of congenital anomalies that involve the structures of the hindbrain and are characterized by herniation of the brainstem and cerebellum below the foramen magnum into the cervical part of the spinal cord and changes in the development of the central nervous system (CNS). Chiari malformation type I (CM I) consists of herniation of the cerebellar tonsils 3 to 5 mm below the foramen magnum and occurs when the base of the skull and the cervical spine are not properly formed. Although Chiari I is asymptomatic in most patients, clinical manifestation can occur in late adolescence and adulthood. Symptoms are related to compression of the craniocervical junction, including ataxia, dysphagia, nystagmus, headache, otalgia, dizziness, tinnitus, diplopia, sleep-disordered breathing, and other changes involving the functions of the cranial nerve pairs.

The diagnosis of CM I is made through clinical history and Magnetic Resonance Imaging (MRI), and the intervention of choice is decompressive craniectomy. Neurosurgical decompression enables functional rehabilitation and symptomatic improvement. However, given assessments of symptomatic improvement in these patients are heterogeneous, interest in developing validated standardized measures and scales to assess symptomatology after decompressive neurosurgery in CM I patients has increased. Dysphagia is one of the most common symptoms. Severe dysphagia may result in tracheal dysphagia, leading to pulmonary infections, malnutrition, prolonged hospital length of stay, and death.

At our center, CM I patients are managed with a multidisciplinary team including physiotherapist, nurse, occupational therapist, psychologist, and a speech therapist preoperatively and postoperatively. The speech therapist evaluates, diagnoses, and rehabilitates deficits in phonoarticulatory organs and the stomatognathic functions such as breathing, sucking, chewing, swallowing, phonation, and articulation. For patients with dysphagia, the speech therapist suggests dietary modifications, speech
therapy, and, if necessary, an alternative way of feeding. The present study aimed to investigate postoperative swallowing disorders in patients diagnosed with CM I after decompression neurosurgery.

**METHODS**

This is a retrospective observational study performed at the Intensive Care Unit (ICU) of a single hospital - Hospital Geral de Fortaleza (Ceará, Brazil). Patients from January 2017 to December 2019 with a CM I confirmed by Magnetic Resonance Imaging (MRI) were included. Patients were included regardless of the presence of associated malformations (syringomyelia and/or basilar invagination). Due to the elective nature of the neurosurgical intervention, patients with associated malformations were admitted to the ICU for postoperative monitoring and Clinical Speech and Language Swallowing Assessment (CSLSA). Patients with inconclusive CSLSA who were regularly using sedative medication at the time of the evaluation were excluded.

The patients underwent neurosurgical intervention with decompressive craniectomy after diagnosis on MRI, followed by the speech-language assessment 24 hours after post-surgical extubation. The same therapist performed the CSLSA on all patients. The CSLSA consisted of observing the aspects of posture, sensitivity, tone, and extra- and intraoral mobility of the phonoarticulatory organs (FAO) and visualizing the functions of breathing, phonation, articulation, chewing, and swallowing. The functional evaluation of swallowing occurred with the offering of foods in liquid (50 mL water), liquid-pasty (50 mL water plus a measure of Thick & Easy® instant food thickener), pasty (50 mL water plus two measures of Thick & Easy® instant food thickener), and solid (two crackers). The speech therapist offered the foods in increasing order of difficulty, liquid-pasty and pasty in a spoon (5 ml), solid directly into the oral cavity, and the liquids in a cup (50 ml). Initially, the patient was asked to keep the food in the mouth to allow assessment of oral containment capacity under command. Then,
the examination was conducted with free administration of other food consistencies according to the capabilities of each patient.

The exams were considered abnormal in the presence of one or more of the following dysphagic manifestations for each food consistency: previous escape (occurrence of food draining through the lips after being taken - mild dysphagia), increased oral transit time (time for complete formation of the bolus and beginning of the elevation of the hyolaryngeal complex exceeding five seconds for liquid and pasty and 30 seconds for solids - mild dysphagia), food stasis in the oral cavity (food accumulation in the anterior or lateral vestibule, floor of the oral cavity and/or surface of the tongue after swallowing - moderate dysphagia), cough (presence of cough reflex before, during or after swallowing - moderate dysphagia), and wet voice (bubbling sound during phonation after swallowing, indicative of tracheal aspiration - severe dysphagia).

After analyzing these variables and the sensitivity, tone, and mobility aspects of FAO, the main dysphagic manifestations were observed at each stage of the assessment with the different food consistencies. We correlated the severity of the dysphagia and the presence of associated malformations in the CM I patients and performed a Fisher's exact test and the chi-square tests. We considered the analyses statistically significant at p < 0.05 using SPSS software, version 21 (SPSS Inc., Chicago, IL, USA).

This work was approved by the Ethics and Research Committee (CAAE 24379019.2.0000.5040). The Informed Consent Term (ICT) was waved due to the nature of the study as a retrospective observational analysis using medical records and evaluation forms. The General Hospital of Fortaleza (Hospital Geral de Fortaleza - HGF) was asked to sign the Loyal Depository Term by the Customer Service Center (Núcleo de Atendimento ao Cliente - NAC) to consent to the data collection authorization, permit the use of data and information related for the purpose research,
and safeguard the rights ensured by Resolution 466/12 of the Brazilian National Health Council.

RESULTS

A total of 64 patients diagnosed with CM I were admitted to the ICU after decompressive craniectomy between 2017 and 2019. We excluded 10 patients because they had insufficient data for the documented inclusion criteria, medical records and/or progress notes. We included 54 patients in our study, of whom 39 were female (72.3%) and 15 were male (27.7%). Their age ranged between 18 and 68 years (mean age: 43.1 ± 47.1 years). Thirty-seven (68.5%) patients with CM I had associated malformations ($p=0.0243$): 21 patients (38.8%) had syringomyelia, 11 patients (20.4%) basilar invagination and 5 patients (9.3%) syringomyelia and basilar invagination (Table 1).

Oropharyngeal dysphagia was present in 37 of the cases (68.5%; $p=0.0016$), of which 18 cases (33.3%) were mild, 3 cases (5.6%) moderate, and 16 cases (29.6%) severe. Dysphonia and dysarthria were found less frequently, in 13 (24.1%; $p=0.4785$) and 3 (5.6%; $p=0.0042$) cases, respectively (Table 1).

In the CSLSA results related to swallowing disorders, the following changes were observed: orofacial hypomobility (29 patients – 53.7%; $p=0.0204$), orofacial hyposensitivity (26 patients – 48.1%; $p=0.3023$), reduced GAG reflex (24 patients – 44.5%; $p=0.1041$), ineffective cough (19 patients – 35.2%; $p=0.0022$), ineffective saliva swallowing (18 patients – 33.3%; $p=0.0396$), dyspnea (13 patients – 24.1%; $p=0.4785$), orofacial hypotonia (11 patients – 20.4%; $p=0.0004$), altered cervical auscultation (wet voice) (10 patients – 18.5%; $p=0.0003$), and dysarthria (3 patients – 9.3%; $p=0.0042$) (Table 2).

The presence of associated malformations significantly influenced the appearance of swallowing disorders, the severity of oropharyngeal dysphagia, the postsurgical complications, and the clinical outcomes of patients with CM type I. A total of
17 cases (31.5%; \( p<0.001 \)) had post-surgical complications with worsening of symptoms, which directly impacted the clinical outcome (\( p=0.0029 \)) of these patients. The presence of syringomyelia was related to the onset of mild oropharyngeal dysphagia in 10 cases (18.5%), while basilar invagination was related to the presence of severe oropharyngeal dysphagia in 12 cases (22.2%) and post-surgical complications in 81.8% of the procedures (Table 3).

The presence of other associated malformations was associated with structural changes of the phonoarticulatory organs and the CSLSA findings. The absence of associated malformations (41.2%) and syringomyelia (47.1%) were associated with normal swallowing. Syringomyelia was associated with a greater incidence of mild dysphagia (50%) and moderate dysphagia (66.7%). Basilar invagination was associated with a higher incidence of severe dysphagia (56.3%) (Table 4).

**DISCUSSION**

*Demographics*

CM I consists of herniation of the cerebellar tonsils through the foramen magnum, where the caudal protrusion of these tonsils in the spinal canal is observed up to the second vertebra. It is the most common and less severe type of CM, occurring in about 0.5-3.5% of the general population, with a slight predominance in females (1.3:1).12,15 We observed a greater female predominance was also identified in our sample, perhaps due to low sample size. The causes of CM type I include primary or secondary congenital hypoplasia or acquired morphological changes, such as premature suture closure, calvarial dysplasia or genetic/syndromic. Mutations in chromosomes 1 and 22 have been identified as possible causes of hereditary hypoplasia of the posterior fossa.16 Despite being a congenital anomaly, CM type I usually presents with a late diagnosis, and patients become symptomatic after the second decade of life. Arnautovic
showed that most patients become symptomatic at around 40 years of age. The mean age of patients (47.1) in this study is similar to that found in the literature.15

Associated Malformations

The skull base is underdeveloped in patients with CM type I resulting in a reduced volume of the posterior fossa, and the cerebellar tonsils are displaced through the foramen magnum due to the inadequate volume to contain the entire cerebellum. This may cause obstruction of cerebrospinal fluid flow due to crowding of the foramen magnum and, consequently, the formation of hydrocephalus and/or syringomyelia over time.17 In the present study, syringomyelia was the most frequent associated malformation in 48.1% of cases, followed by basilar invagination in 29.7% of the patients. Although the prevalence of syringomyelia is within the range reported by previous institutional studies, the prevalence of basilar invagination is far greater in our study.18,19,20 While the relationship between CM I and basilar invagination remains incompletely understood, both malformations are mesodermal, and surgical outcomes for patients with CM I with and without basilar invagination do not differ.20,21,22

Presentation

The most frequent symptoms are suboccipital and/or cervical headaches in 80% of the cases. While this is consistent with prior studies reporting headache as the most frequent manifestation, the proportion of patients presenting with headache was double that reported in previous studies.18,19 Other common symptoms include eye disorders, otoneurological symptoms (dizziness, hearing loss, vertigo), gait ataxia, and generalized fatigue. Although less common, case series have reported patients with isolated pain or weakness in the extremities.18,23 One of these reports included a presentation of unilateral shoulder pain with isolated muscle weakness in a sports medicine clinic.23 Other symptoms that were initially neglected, such as aspiration, regurgitation,
asphyxia, dysphagia, dysphonia, chronic cough, and sleep disorders, result from impaired oropharyngeal function or upper airway obstruction and can progressively become fatal.24

Postoperative Swallowing Disorders

In the present study, dysphagia was present in 68.5% of the cases, corroborating the literature that reported that aspiration and other abnormalities are observed in 36% of the post-surgical cases submitted to swallowing videofluoroscopy.25 The prevalence of dysphagia in patients with CM Type I has been reported to vary from 4 to 47%.12,26 Some reports have described cases in which dysphagia was the only symptom of CM type I presentation and improved after occipitocervical decompression.27 Thus, the current evidence on whether dysphagia is a manifestation of CM type I is based mainly on retrospective observations. The reasons why dysphagia is common in patients with CM I are not clear. Vascular ischemia, traction of the lower cranial nerves, and compression of the nearby brainstem and cerebellum are possible explanations.28,29 Syndromes associated with CM I, such as syringomyelia and basilar invagination, further affect the bulbar and upper cervical region, indicating worsening prognosis and promoting a greater risk of respiratory complications and a higher degree of dysphagia.30 Cerebellar signs can also be found, including ataxia, dysmetria, nystagmus, and lower cranial nerve deficits (IX, X, XI, XII).31 Injury to cranial nerves may explain the appearance of dysphagia. In the present study, the hyposensitivity of orofacial structures (48.1%) and GAG (44.5%) may be related to injury to the IX cranial nerve; hypotonia (20.4%) and hypomobility (53.7%) of the orofacial structures associated with injuries to the XII cranial nerve and; alteration in vocal quality (24.1%), altered cervical auscultation (18.5%) and ineffective cough (35.2%) related to injury to the X cranial nerve. Sleep apnea can also occur in CM type I patients due to a weakness in the pharynx muscles caused by the brain stem, upper spinal cord or compression of the lower cranial nerve.32
We emphasize that the sample in the present study was evaluated shortly after decompressive craniectomy, which may explain the higher incidence of dysphagia compared to previous studies. Although CM I generally portends a favorable prognosis, the prognosis depends on the presence of pre-existing neurological deficits. Most patients without neurological deficits have excellent post-surgical outcomes. Studies examining severe cases of CM I are still few and discouraging, indicating that survivors may experience increasing motor dysfunction over time. Individuals with chronic weakness or gait problems generally do not improve, and their prognosis is poor. Additional studies examining outcomes and strategies to improve the prognosis of these patients are necessary.

Limitations

Our study has limitations. The small single-center study population subjected the results of the study to the patient selection and management preferences of our department. Additionally, objective swallowing tests were not performed. However, the population submitted to the clinical speech-language assessment of swallowing, which was superior to the studies that used objective exams. The incidence of dysphagia was greater than in published studies, possibly due to the time of post-surgical evaluation. Findings during the oropharyngeal phases of swallowing were also highly variable. Therefore, it is difficult to define the specific limits that distinguish the normal variations in swallowing from the findings indicative of dysphagia. Large prospective studies are necessary to investigate these factors further.

CONCLUSION

Decompressive craniectomy leads to low rates of post-surgical complications in
patients with CM. However, there was a high incidence of oropharyngeal dysphagia in these patients. The presence of associated malformations is associated with a poorer prognosis and greater severity of oropharyngeal dysphagia in patients with CM I. Postoperative monitoring of patients with CM I is important in relation to stomatognathic functions, especially swallowing, due to the risk of potentially fatal bronchoaspiration. Clinical evaluation of swallowing by a speech therapist as part of a multidisciplinary approach is essential for the identification, diagnosis, and rehabilitation of these symptoms. Additionally, the speech therapist enables the determination of a safe feeding route and promotes improved outcomes at hospital discharge in CM I patients undergoing decompressive neurosurgery.

REFERENCES


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