· 论著 ·

Clinical characteristics of neurogenic dysphagia in adult patients with Chiari malformation type Ⅰ

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ABSTRACT **Objective**: To investigate changes of swallowing function and associated symptoms in Chiari malformation type Ⅰ (CMⅠ) patients with and without dysphagia by the analysis of their clinical and high-resolution manometry (HRM) parameters. **Methods**: A total of 42 patients diagnosed with symptomatic CMI without atlantoaxial dislocations which were confirmed by clinical manifestations and magnetic resonance imaging(MRI) findings between January 2010 and July 2015 at Peking University Third Hospital were included in this study. Twenty patients had a history of various dysphagia symptoms, or reported symptoms of choking, coughing after eating or drinking, while the other 22 patients denied symptoms of dysphagia. The data collected from the medical records of these patients included the patient's age, sex, date of diagnosis, duration of illness, symptoms, results of MRI and HRM, and date of surgery. **Results**: (1) Dysphagia group had 14 female patients, and no-dysphagia group had 8 female patients. Dysphagia usually occurred in female patients, and in addition to dysphagia, we recorded other symptoms and signs in the CMⅠ patients, including numbness, hypoesthesia, limb weakness, neck pain, muscle atrophy, ataxia, hoarseness, symptoms caused by posterior cranial nerve damage, pharyngeal reflex, uvula deviation, and pyramidal signs. A higher percentage of the CMⅠ patients with dysphagia (15/20) had symptoms of posterior cranial nerve damage compared with the control group (5/22; $P=0.01$). (2) HRM showed a significant difference in upper esophageal sphincter (UES) relax ratio measurement (75.3% *vs.* 63.1%, $P=0.023$) and UES proximal margin (17.2 cm *vs.* 15.7 cm, $P=0.005$) between the two groups. (3) The percentage of syringomyelia affecting the bulbar or upper cervical region on MRI was significantly higher in the dysphagia group (17/20 *vs.* 7/22, $P=0.001$). **Conclusion**: CMⅠ was usually accompanied by symptoms caused by posterior cranial nerve damage, ataxia, and positive pyramidal signs. Location of the syringomyelia affecting specifically the bulbar or upper cervical region was associated with dysphagia in CMⅠ patients. These findings suggest that the mechanism of dysphagia in CMⅠ may be due to a dysfunction in the neurological pathway of pharyngeal muscle movement. Dysphagia etiology work-up should include CMⅠ in the differential diagnosis.

KEY WORDS Dysphagia; Chiari malformation type Ⅰ; High-resolution manometry

Chiari malformation type Ⅰ (CMⅠ) is defined as cerebellar tonsillar herniation greater than 5 mm inferior to the foramen magnum[1]. Anatomically, this herniation is also associated with decreased posterior fossa volume, decreased cerebro-spinal fluid (CSF) in the posterior fossa, and variable skull base dysplasia[2-3]. Epidemiological studies of CMⅠ suggest that approximately 0.56% - 0.97% of the population shows > 5 mm tonsillar herniation on MRI imaging[4-6]. Either explicit or vague neurological symptoms attributable to hindbrain herniation is present in 63% - 69% of patients with radiologically proven CMⅠ. Presenting symptoms are highly variable and include headache syndromes, brainstem and/or cerebellar dysfunction, neck pain, and spinal cord dysfunction[2]. Classically, these symptoms are nonspecific, but can progress, resulting in the need for surgical management weeks to months after development of the first symptom[7-8].

Clinical presentation also varies with age. Several authors believe that young children tend to present with oropharyngeal symptoms, while older patients tend to present with headaches and progressive sensory or motor findings[9-12]. However, in our study population, we found that 26.9% of the adult patients (34/126) with CMⅠ without atlantoaxial dislocations suffered from varying severity of swallowing dysfunction. The incidence of dysphagia ranged from 8% to 28% according to large CMⅠ case series[13-15]. However, there only existed a few case reports of CMⅠ patients with swallowing dysfunction[16-18]. Paulig[19] and Achi-

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ron et al[20] reported two CMⅠ cases with severe dysphagia. The marked improvement in dysphagia after neurosurgical posterior fossa decompression suggested that CMⅠ was the principle cause of dysphagia. The prevalence of this type of neurogenic dysphagia in adult CMⅠ patients was not as rare as previously thought.

High-resolution manometry (HRM) is a promising technique for the evaluation of esophageal motor function. There are two main types of HRM recording systems—those that use intraluminal solid-state transducers, and those that use perfused assemblies connected to external transducers. Water-perfusion manometry (WPM) allows recording of multiple pressure channels from one catheter and is relatively inexpensive. To date HRM data about CMⅠ patients are very rare.

In this study, we report the clinical and high-resolution manometry characteristics of dysphagia in CMⅠ patients.

1 Materials and Methods

1.1 Patients and study design

A total of 42 patients who were diagnosed with symptomatic CMⅠ without atlantoaxial dislocations at Peking University Third Hospital between January 2010 and July 2015, were included in this study. The age range was from 16 – 70 years . Twenty of the patients reported a history of dysphagia, while the other 22 patients denied symptoms of dysphagia. All the patients suffered from numbness, hypoesthesia, limb weakness, neck pain, muscle atrophy, ataxia, hoarseness, symptoms caused by posterior cranial nerve damage (such as decreased sweating or hypoesthesia on the affected side of the face), pharyngeal reflex, uvula deviation, and pyramidal signs. The data collected from the medical records of these patients included the patient's age, sex, date of diagnosis, duration of illness, symptoms, results of MRI and HRM, and date of surgery.

The treatments for CMⅠ were surgery and management of symptoms, based on the occurrence of clinical symptoms rather than the radiological findings. All the patients underwent decompressive surgery, which involved removing the lamina of the first and sometimes the second cervical vertebrae and part of the occipital bone of the skull to relieve pressure. This surgery involved the opening of the dura mater and the expansion of the space beneath, a dural graft was usually applied to cover the expanded posterior fossa.

The improvement of dysphagia was evaluated within 7 days after surgery by the patient's self-evaluation.

All the patients gave informed consent in writing before commencement of the study.

1.2 HRM

All the patients underwent HRM. The water-perfused high resolution manometry system (Medical Measurement Systems, Enschede, Netherlands) was equipped with a catheter composed of 22 thin polyvinyl tubes (channel P1 – P22). All the patients underwent esophageal manometry according to the standard clinical protocol of the gastrointestinal motility center of Peking University Third Hospital. During the manometry process, the subjects lay supine and the catheter was passed transnasally into the stomach. After recording the lower esophageal sphincter (LES) and upper esophageal sphincter (UES) rest pressure and length, motility function analysis was performed using a series of 10 wet swallows with 5 mL water, each separated by 30 seconds. For each esophageal procedure, the parameters captured included the proximal LES and UES margins, rest pressure, relax ratio, contraction front velocity (CFV), distal contractile index (DCI).

1.3 Statistical analysis

The statistical analysis was designed to determine the degree of dysphagia associated with CMⅠ. The quantitative data that followed a normal distribution were expressed as mean ± standard deviation (SD). The quantitative data of nonnormal distribution were presented as medians (quartile range). Categorical variables were expressed as numbers of cases. We compared the characteristics and HRM variables between the groups using a two-sided Fisher's exact test as appropriate. The count data were tested using the Chi-squared test. Statistical analysis was performed with the use of SPSS software, version 19.0 (SPSS Inc., Chicago, IL, USA). The P value < 0.05 was considered a statistically significant difference.

2 Results

2.1 Clinical characteristics of dysphagia in patients with CMⅠ

Almost half of CMⅠ patients (20/42) included in this study had symptoms of dysphagia, choking, or coughing after eating or drinking. As shown in Table 1, dysphagia group had 14 female patients. And no-dysphagia group had 8 female patients. The difference was significant. The median age was 47.6 (range: 24 – 58) years in the dysphagia group and 48.9 (range 16 – 70) years in the non-dysphagia group. The time between the first symptom onset and diagnosis of CMⅠ was 78.6 (24 – 240) months in the dysphagia group and 107.6 (1 – 240) in the non-dysphagia group. No association was noted between the incidence of dysphagia and age, or the median time from the symptom onset to the diagnosis of CMⅠ.

2.2 Associated symptoms

In addition to dysphagia, we recorded other symptoms and signs in the CMⅠ patients, including numbness, hypoesthesia, limb weakness, neck pain, muscle atrophy, ataxia, hoarseness, symptoms caused by posterior cranial nerve damage (such as decreased sweating or hypoesthesia on the affected side of the face), pharyngeal reflex, uvula deviation, and pyramidal signs. A higher percentage of the CMⅠ patients with dysphagia (15/20) had symptoms caused by posterior cranial nerve damage compared with the control group (5/22, $P = 0.01$). The ratio of those with hoarseness, ataxia and positive pyramidal signs in the dysphagia group (4/20, 9/20, 9/20, respectively) was also higher than in the non-dysphagia group (0/20, 3/20, 3/20, respectively), which was significantly different ($P < 0.05$). Other associated symptoms were not statistically significant between the two groups (Table 1).

Postoperatively, 10/12 patients reported that headache/neck pain improved in dysphagia group, while 14/16 patients improved in non-dysphagia group. Four patients (2 patients in each group) reported no change. In dysphagia group, 11/15 patients who suffered from symptoms caused by posterior cranial nerve damage improved after surgery, while 3/5 improved in non-dysphagia group. Pyramidal signs improved in 6/9 patients in dysphagia group while 1/3 in control group.

Table 1 Characteristics of Chiari Ⅰ malformation patients with and without dysphagia

Characteristic	Dysphagia	No dysphagia	Total	*P* value[a]
Patients/*n*	20	22	42	
Female/*n*	14	8	22	0.029
Median age (range)/year	47.6(24 – 58)	48.9(16 – 70)	48.3(16 – 70)	0.774
Median time between symptom onset and diagnosis of CMⅠ (range) /month	78.6(24 – 240)	107.6(1 – 240)	93.8(1 – 240)	0.234
Associated symptoms				
Numbness/*n*	15	17	32	0.863
Hypoesthesia/*n*	17	17	34	0.527
Limb weakness/*n*	15	14	29	0.426
Neck pain/headache/*n*	12	16	28	0.382
Muscle atrophy/*n*	9	7	16	0.380
Hoarseness/*n*	4	0	4	0.027
Symptoms caused by posterior cranial nerve damage/*n*	15	5	20	0.010
Ataxia/*n*	9	3	12	0.025
Pharyngeal reflex abnormality/*n*	11	9	20	0.361
Uvula deviation/*n*	13	8	21	0.068
Pyramidal signs/*n*	9	3	12	0.025

a, *P* value were the results of dysphagia group *vs.* no dysphagia group.

2.3 HRM

Table 2 shows the main motility measurements of CMⅠ patients with or without dysphagia. There were significant differences in UES relax ratio measurement (75.3% *vs.* 63.1%, $P = 0.023$) and UES proximal margins (17.2 cm *vs.* 15.7 cm, $P = 0.005$) be-

tween the two groups. Although the average UES rest pressures differed (64.3 mmHg *vs.* 55.4 mmHg, 1 mmHg = 0.133 kPa), this was not significant (P = 0.18). No statistical significance was shown in LES proximal margins, rest pressure, relax ratio, DCI or CFV.

Table 2 High-resolution manometry parameters of Chiari malformation type Ⅰ patients with and without dysphagia

Characteristic	Dysphagia	No dysphagia	P value
UES proximal margin/cm	17.2(13.7 – 19.6)	15.7(15.2 – 18.4)	0.005
UES rest pressure/mmHg	64.3(18.8 – 151.2)	55.4(11.6 – 166.2)	0.180
UES relax ratio/%	75.3(6.0 – 92.0)	63.1(16.0 – 97.0)	0.023
LES proximal margin /cm	39.3(35.50 – 47.20)	43.2(37.2 – 45.0)	0.136
LES rest pressure /cm	14.8(4.4 – 37.40)	14.9(6.1 – 27.6)	0.872
LES relax ratio /%	52.6(34.1 – 93.0)	40.2(17.0 – 95.3)	0.576
DCI/(mmHg · s · cm)	711.0(350.0 – 2 456.0)	603.1(222.0 – 2 235.0)	0.655
CFV/(cm/s)	7.1(2.9 – 14.4)	17.8(3.5 – 100.0)	0.758

1 mmHg = 0.133 kPa; UES, upper esophageal sphincter; LES, lower esophageal sphincter; DCI, distal contractile integral ; CFV, contractile front velocity.

2.4 MRI

All the patients underwent MRI as a gold standard diagnosis for CMⅠ. Seventeen patients in the dysphagia group and 19 patients in the non-dysphagia group had MRI findings of syringomyelia. There were no significant differences in diameter of syringomyelia (8.7 cm *vs.* 9.6 cm) or involved segments (12 *vs.* 11) between the two groups . However, the percentage of syringomyelia affecting the bulbar or upper cervical region was significantly higher in the dysphagia group (15/17 *vs.* 7/19, P = 0.001). Posterior pharyngeal wall thickness was also measured between the two groups, which was not shown to be significantly different (3.45 mm *vs.* 3.11 mm, P = 0.572, Table 3). Postoperative improvement of syringomyelia was found for all the 36 patients with syrinx.

Table 3 Magnetic resonance imaging characteristics of Chiari malformation type Ⅰ patients with and without dysphagia preoperation

Characteristic	Dysphagia	No dysphagia	Total	P value[a]
Syringomyelia/n	17	19	36	0.900
Diameter (range) /mm	8.7(4 – 19)	9.6(4 – 14)	8.9(4 – 19)	0.335
Involved segments (range)/cm	12(4 – 17)	11(4 – 15)	12(4 – 17)	0.521
Affecting bulbar or upper cervical region/n	15	7	24	0.001
Thickness of posterior pharyngeal wall/mm	3.45(1.74 – 4.42)	3.11(2.10 – 3.60)	3.35(1.74 – 4.42)	0.572

a, P value were the results of dysphagia group *vs.* no dysphagia group.

3 Discussion

The most common presenting symptoms of CMⅠ include cervical neck pain, occipital headache, movement dysfunction, paresthesia, muscle atrophy, or unsteady gait. Comparatively, symptoms of cranial neuropathies, such as hoarseness, choking, and swallowing difficulty can often be ignored by patients and even neurosurgeons. In previous study, we reviewed the medical records of 140 CMⅠ patients from January 2002 to September 2006 in our hospital. There were only 15 cases (10.7%) which reported having "difficulty in swallowing", or "choking while drinking", while 72 cases (51.4%) reported "no difficulty in swallowing". The remaining 53 cases (37.6%) did not clearly indicate presence or absence of dysphagia. In order to study changes of swallowing function in CMⅠ patients, we surveyed swallowing function of 126 CMⅠ patients in our hospital from January 2007 to July 2010[21]. In the study, 34 cases (26.9%) clearly reported dysphagia of varying severity. Most patients admitted difficulty in swallowing or choking while drinking after careful history taking. The average interval between the onset of symptoms and the diagnosis was

(7.3±1.8) years, while the onset of dysphagia and choking was only (2.5±0.9) years. Dysphagia occurred more often in advanced CMⅠ, but was frequently overlooked by clinical practitioners.

Dysphagia may give rise to clinically relevant complications. Aspiration occurs in 50% of patients with oropharyngeal dysphagia, and is associated with a mortality of up to 50%[22]. Additionally, new studies show that dysphagia risk factor for low respiratory tract infections (LRTIs) and community-acquired pneumonia (CAP) in the elderly.

We analyzed other associated symptoms' relationship to dysphagia in CMⅠ patients. A higher percentage of CMⅠ patients with dysphagia (15/20) experienced symptoms caused by posterior cranial nerve damage, such as decreased sweating or hypoesthesia on the affected side of the face, as compared with the control group (5/22, $P=0.001$). The ratios of those with ataxia and positive pyramidal signs in the dysphagia group (9/20, 9/20, respectively) were also higher than those in the non-dysphagia group (3/22, 3/22, respectively), and there were significant differences ($P=0.025$). These associations suggest a pathophysiological mechanism of dysphagia in CMⅠ.

Manometry is a standard method of characterizing oropharyngeal and esophageal motor function by providing measurements of pressures, peristalsis, and coordination. There was a significant difference in UES relax ratio measurement (75.3% *vs.* 63.1%, $P=0.023$). The UES normally prevents food reflux into the pharynx. Surprisingly, the UES relax ratio was higher in the dysphagia group, suggesting that excessive relaxation of UES might cause food refluxing and coughing. On the other hand, UES rest pressures in the dysphagia group [64.3 (18.8 – 151.2) mmHg] were higher than those in the non-dysphagia group [55.4 (11.6 – 166.2) mmHg], though not significantly different ($P=0.180$). These findings suggest that dysphagia in CMⅠ patients may be a dysfunction in the neurological pathway of pharyngeal muscle movement. Unfortunately, other parameters, including UES relaxation time, cannot be measured using water-perfusion manometry, which is an important parameter in estimating UES relaxation function. Solid-state manometry (SSM) would better measure UES pressure in future studies.

From a neurological point of view, swallowing control is governed by three main elements: the afferent system, the brain stem swallowing center and the efferent system. Oropharyngeal afferents project to supramedular structures and to the brain stem swallowing center, in the medulla oblongata, to allow involuntary onset of the pharyngeal swallow response and modulate volitional swallowing. The afferent neurons of the oropharynx and larynx involved in swallowing are the maxillary branch of the trigeminal nerve (Ⅴcranial nerve), the pharyngeal branch of the glossopharyngeal nerve (GPNph, Ⅸ cranial nerve) and two branches of the vagus nerve (Ⅹ cranial nerve)—the pharyngeal branch and the superior laryngeal nerve (SLN). These afferents are formed in part by non-myelinated (type C) or thinly myelinated (type A) fibers, and are sensitive to mechanical stimuli (pressure, touch), temperature changes, and chemical stimuli (hydrogen ions, taste stimuli)[23]. As dysphagia associated with CMⅠ is found to frequently be associated with symptoms caused by posterior cranial nerve damage and ataxia and pyramidal signs, this suggests that the underlying pathophysiology is comprised of a set of sympathetic and parasympathetic nerve fiber injuries.

In our study, we found that the location of syrinxon MRI was highly associated with the presence of dysphagia symptoms, as the percentage of syringomyelia affecting the bulbar or upper cervical region was significantly higher in the dysphagia group 15/20 *vs.* 7/22, $P=0.001$). To our surprise, diameter and involved segments of syringomyelia did not show the same correlation. This may mean that injury of nerve fibers passing specifically through the bulbar and upper cervical region is important in the pathogenesis of dysphagia. Postoperative improvement of syringomyelia was found for all the 36 patients with syrinx which was consistent with mate analysis results made by Arnautovic et al[24]. In that study, the authors reviewed 145 English-language reports of pediatric, adult, and combined (adult and pediatric) surgical series of patients with CMⅠ published from 1965 through August 31, 2013. They concluded that 78% of the CMⅠ patients who had syringomyelia improved or resolved postoperatively.

Consistent with our previous study[24], dysphagia

was improved (according to the patients' self-evaluation) in half of the cases shortly after posterior fossa decompression with duraplasty. We believe that improvement of dysphagia may be a meaningful index to evaluate the effect of surgery in the early postoperative stage. According to our study, headache/neck pain, symptoms caused by posterior cranial nerve damage, and pyramidal signs may be associated with dysphagia. However, there were no differences in the outcomes of these symptoms between the two groups. In the study of Arnautovic et al[24], most (80%) reported postoperative neurological outcomes as follows: 75% improved, 17% showed no change, and 9% experienced worsening. Postoperative headaches improved or resolved in 81% of the patients[23]. We need further study to identify whether the dysphagia and other associated symptoms or signs outcomes correlate in these patients.

A limitation in this study is the small number of patients in each group. Evaluation of the total incidence, risk factors, assessment, intervention and postoperative change requires a systematic analysis of a larger sample size.

Neurogenic dysphagia is thought to be a relatively rare symptom in adult CMⅠ patients without atlantoaxial dislocations. However, it may cause fatal complications. Dysphagia associated with CMⅠ was usually accompanied by symptoms caused by posterior cranial nerve damage, ataxia, and positive pyramidal signs. Location of the syringomyelia affecting specifically the bulbar or upper cervical region was associated with dysphagia in CMⅠ patients. The difference of UES rest pressure and relax ratio in dysphagia patients *versus* non-dysphagia patients suggests that the pathophysiology of dysphagia may be a dysfunction in the neurological pathway of pharyngeal muscle movement. Dysphagia etiology work-up should include CMⅠ in the differential diagnosis. We hope that this study highlights the importance of dysphagia in the CMⅠ adult population, and shows how it can be a meaningful index for improvement in symptoms after posterior fossa decompression. In the future, we hope to further characterize mechanisms of dysphagia in CMⅠ adult patients using SSM in a larger patient population.

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Chiari 畸形Ⅰ合并神经源性吞咽功能障碍的临床特征

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[摘 要] **目的**:成年 Chiari 畸形Ⅰ型(chiari malformation typeⅠ,CMⅠ)患者中神经源性吞咽障碍的发生率并不少见,临床上罕有关于食管高分辨率测压(high-resolution manometry,HRM)技术对 CMⅠ患者吞咽功能的精确评价的报道。本研究拟通过 HRM 量化评价 CMⅠ患者吞咽功能,初步探讨影响吞咽功能的可能相关因素及其机制。**方法**:共纳入北京大学第三医院 2010 年 1 月至 2015 年 7 月收治的 42 例经临床和 MRI 检查、确诊不合并寰枢椎脱位的 CMⅠ患者,将患者分为吞咽障碍组 20 例和不合并吞咽障碍组 22 例。所有患者均接受 HRM 检查,收集所有患者的临床、影像学资料 和 HRM 评价参数,并做统计学分析。**结果**:(1)合并吞咽障碍的女性 CMⅠ患者比例明显高于不合并吞咽障碍组(14/20 *vs.* 8/22, $P = 0.029$),吞咽障碍组后组颅神经损伤的其他症状(包括声音嘶哑、咽部感觉减退、患侧面部感觉减退及汗液分泌减少等)发生率明显高于不合并吞咽障碍对照组(15/20 *vs.* 5/22, $P = 0.01$)。(2)HRM 显示吞咽障碍组食管上括约肌(upper esophageal sphincter ,UES)松弛比高于对照组(75.3% *vs.* 63.1% ,$P = 0.023$),UES 上缘亦高于对照组(17.2 cm *vs.* 15.7 cm,$P = 0.005$)。(3)吞咽障碍组 MRI 影像上延髓或上颈部的脊髓空洞比例明显高于对照组(17/20 *vs.* 7/22, $P = 0.001$)。**结论**:CMⅠ患者的吞咽障碍经常与后组颅神经损伤、共济失调和阳性锥体束征相关,HRM 显示平均 UES 松弛比例的差异亦有可能与延髓或上颈髓的脊髓空洞症有关,CMⅠ中的吞咽障碍的机制可能是由于神经源性咽肌运动障碍所导致,吞咽障碍病因学检查应包括 CMⅠ畸形鉴别诊断。

[关键词] 吞咽障碍;Chiari Ⅰ畸形;食管高分辨测压
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