INTRODUCTION

Chiari type I malformation, one of the anomalies of the cerebellum, is defined as descent of the cerebellar tonsils into the cervical spinal canal. This pathology may cause compression of the brain stem and is often associated with syringomyelia. Therefore, symptoms can result from the brain stem compression and/or syringomyelia. Such a variety of the symptoms sometimes makes the diagnosis difficult, especially when dysphagia is the singular symptom. Although the symptom may be rare, it can progress to significant impairment, and even to death by bronchial aspiration. We report a case of Chiari I malformation with persistent dysphagia as a long-term symptom. This paper reminds physicians that Chiari type I malformation should also be considered when encountering cases of persistent or progressive dysphagia.

CASE REPORT

A fifty-eight year-old woman had a past history of hypertension, diabetes mellitus, and hemifacial spasm which was surgically treated 14 years earlier. Since 1995, the patient had experienced difficulty in swallowing. Coughing and choking also occurred while eating and drinking. The course of the symptom had fluctuated with remissions and exacerbations. After developing pneumonia in January 1996, she was referred to our department for neurological evaluation because neurogenic tracheobronchial aspiration was suggested. On admission, her general condition was good. Neurological examination

Abstract: The authors describe a rare case of Chiari type I malformation presenting as persistent dysphagia. A fifty-eight year-old woman showing a nearly one-year history of swallowing difficulty was transferred to our department for neurological evaluation of aspiration pneumonia. Neurological examination revealed absence of bilateral gag reflexes, and mild weakness of the right upper extremity. Magnetic resonance images showed typical features of Chiari I malformation combined with spinal cord compression due to cervical spondylosis at four intervertebral levels from C3/4 to C6/7. Neither hydrocephalus nor syringomyelia was associated. Decompression of the foramen magnum combined with expansive laminoplasty of C2 through C7 was performed. Dysphagia and abnormal gag reflex improved within 1 week after surgery. Thus, awareness of the Chiari type I malformation as a cause of dysphagia in its adult symptomatology will lead to correct diagnosis of the pathology in its early stage.

Key words: Chiari malformation, Dysphagia, Surgery, MRI
revealed absence of bilateral gag reflexes, and mild weakness of the right upper extremity. Her voice was somewhat nasal. Left peripheral facial nerve palsy was a sequel of surgery for hemifacial spasm. Mild atrophy of right sternocleidomastoid muscle was suggested but a denervation pattern was not proven by electromyography. Impairment of tongue movement was equivocal. There was no objective confirmation of sensory disturbance though she complained of light numbness in the finger tips of both hands which appeared to increase with neck extension. Since deep tendon reflexes were not decreased, diabetic neuropathy seemed not to be likely. Headache or neck pain was not induced with head posture. Magnetic resonance images (MRI) showed caudal displacement of the cerebellar tonsil to the level of C1 lamina, but there was no evidence of hydrocephalus or syringomyelia. MRI also revealed developmental narrow spinal canal and degenerative disc disease causing spinal cord compression at the intervertebral levels from C3/4 to C6/7 (Fig. 1). Occipital craniectomy and laminectomy of the C1 posterior arch were performed with expansive laminoplasty of C2 to C7 in April 1996. Tight dura mater at the foramen magnum was opened and patched with artificial dura. Numbness of the fingers disappeared 4 days later, and the gag reflex normalized 1 week after surgery. There was no swallowing difficulty detected during the 28-month follow-up period. Postoperative MRI showed well-decompressed medulla oblongata and the cerebellar tonsil at the foramen magnum. The spinal cord also decompressed in the spinal canal (Fig. 2).

DISCUSSION

The major presenting symptoms of Chiari malformation without myelodysplasia consist of pain, weakness, and sensory disturbance in both adult and pediatric patients. Dysphagia is also noted in 6 to 27% of the patients, however, the complaint has always been associated with other major symptoms at the time of diagnosis. The main physical signs are a combination of motor deficit with hyperreflexia and muscle atrophy, sensory disturbance, and other miscellaneous signs including cerebellar ataxia or lower cranial nerve palsies. Lower cranial nerve dysfunction can be noted in 16 to 56% of the patients. The cranial nerves affected are the
glossopharyngeal and vagal nerves in many, and disturbance of those nerves result in abnormal gag reflex. Recently, progressive dysphagia has also been recognized as a preceding symptom suggestive of following severe brain stem dysfunction in approximately one third of the patients with Chiari malformations. Progression of the symptom seemed to be related to the age of the patients. Whereas infants, mostly with Chiari Type II, showed rapid deterioration after the initial presentation, adults show more gradual deterioration. Accordingly, there may be rare adult cases complaining only of long term dysphagia. A few such patients with dysphagia as a sole manifestation of Chiari malformation were recently reported.

Achiron and Kuritzky first reported 2 adult cases with dysphagia and recurrent aspiration pneumonia without any other complaints for several years. In one of the cases, horizontal nystagmus, truncal ataxia and hyperreflexia followed the initial symptoms 11 years after the onset. Patients similar to our case are listed in Table 1. All patients were adults without syringomyelia or hydrocephalus, and dysphagia as a sole symptom persisting for several months to 11 years. Dysphagia frequently presented as recurrent aspiration or pneumonia. Neurological examination revealed absent gag reflex, palatal hypesthesia or palatal hypomotility, but some showed no neurological deficits at the initial examination. Our patient had a one-year history of dysphagia without any other definitive neurological signs similar to other patients. MRI findings were also very similar. Decompression of the foramen magnum and upper cervical laminectomy was effective in all cases, and the symptom disappeared or markedly improved within 8 months after surgery. We performed laminoplasty of C2 to C7 combined with decompression of the occipital bone and C1 lamina, since the patient also had spinal cord compression due to cervical spondylosis combined with a narrow spinal canal. The necessity of a wide-ranged laminoplasty performed in this case may be controversial, because a narrow spinal canal with mild cord compression does not always cause symptoms. However, we decided to perform surgery because the precise origin of finger numbness was not clinically identified.

The swallowing reflex is under the control of the swallowing center in the medulla oblongata, the 5th, 7th, 9th, 10th, and 12th cranial nerves and their nuclei with other connecting nuclei. Though the swallowing mechanism is physiologically divided into three phases; oral, pharyngeal, and esophageal, all three can be affected by brain stem compression and the cranial...

Fig. 2. MR image one month after surgery showed the well-decompressed spinal cord and cerebellar tonsil.
nerve distortion seen in patients with Chiari malformation⁶,¹⁴. Pollack et al. described swallowing dysfunction in Chiari malformation as a combination of pharyngoesophageal dysmotility, cricopharyngeal dysfunction, nasal regurgitation, tracheal aspiration, and gastroesophageal reflux shown after esophageal manometry and on barium esophagogram. Cricopharyngeal dysfunction, especially failure of complete relaxation of the upper esophageal sphincter muscle (namely cricopharyngeal achalasia), had been emphasized as a manifestation of Chiari malformation in children⁷,¹⁵. Cricopharyngeal myotomy in such cases results in complete resolution of dysphagia². A case reported by Elta et al. showed simultaneous contraction of the upper and lower esophageal sphincter, in other words non-peristaltic wave forms of the esophagus in esophageal manometry¹⁰. Another case showed laryngeal influx, incomplete closure of the larynx, and pooling in the valleculae on fluoroscopy esophagogram¹⁶. It is suggested that ambiguous symptoms seen in adult patients may result from physiological attempts to compensate for the a functional loss of involuntary components in the oral phase of swallowing⁶. Though precise otopharyngological evaluation was not performed in our case, the gag reflex was severely disturbed as a dysfunction of the pharyngeal phase of swallowing. However, further investigations will be necessary to clarify the pathophysiological mechanism of dysphagia in adult Chiari I patients.

The problem of misdiagnosis in adult Chiari malformation presenting with ambiguous symptoms has already been pointed out³. Cases showing only dysphagia may be especially difficult to diagnosed before the MRI era. All cases listed above were reported in the 1990’s, and MRI led to the correct diagnosis in all cases.

<table>
<thead>
<tr>
<th>Authors</th>
<th>Age/Sex</th>
<th>Initial symptoms</th>
<th>Duration</th>
<th>Following symptoms</th>
<th>MRI findings</th>
<th>Operation</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Achiron (1990)</td>
<td>57/F</td>
<td>dysphagia pneumonia</td>
<td>11y</td>
<td>truncal ataxia</td>
<td>type I</td>
<td>no</td>
<td>no</td>
</tr>
<tr>
<td></td>
<td>74/F</td>
<td>dysphagia pneumonia</td>
<td>3.5y</td>
<td>no</td>
<td>type I</td>
<td>no</td>
<td>no</td>
</tr>
<tr>
<td>Michel (1990)</td>
<td>22/M</td>
<td>dysphagia pneumonia</td>
<td>9mo</td>
<td>ataxic gait</td>
<td>type II</td>
<td>no</td>
<td>no</td>
</tr>
<tr>
<td>Jeménez (1991)</td>
<td>34/F</td>
<td>dysphagia pneumonia</td>
<td>1y</td>
<td>no</td>
<td>type I</td>
<td>no</td>
<td>no</td>
</tr>
<tr>
<td>Elta (1996)</td>
<td>27/F</td>
<td>dysphagia pneumonia</td>
<td>3y</td>
<td>dizziness pain</td>
<td>type II</td>
<td>no</td>
<td>no</td>
</tr>
<tr>
<td>Ikusawa (1996)</td>
<td>63/F</td>
<td>dysphagia pneumonia</td>
<td>5y</td>
<td>weakness</td>
<td>type I</td>
<td>no</td>
<td>no</td>
</tr>
<tr>
<td>Present case</td>
<td>58y/F</td>
<td>dysphagia pneumonia</td>
<td>1y</td>
<td>no</td>
<td>type I</td>
<td>no</td>
<td>no</td>
</tr>
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Sagittal MR image is the key to resolving the problem. Clinicians must recognize dysphagia as a possible indicator for Chiari malformation. Complete resolution can be achieved by a relatively simple surgical procedure, namely decompression surgery\(^9,10,11,13\).

**REFERENCES**


