Oropharyngeal dysphagia associated with Chiari I malformation and syringomyelia

ABSTRACT

Background: Dysphagia associated with neurological disease is an important clinical manifestation in the diagnosis of injury that justifies compression of the brainstem and lower cranial nerves. We undertook this investigation to emphasize the study of dysphagia in a patient with Chiari type I malformation associated with syringomyelia in the absence of primary gastroenterological symptoms.

Clinical case: We describe the case of a 62-year-old woman with oropharyngeal dysphagia of 6 years evolution, cervicobrachialgia, ptosis and facial diplexia.

Conclusions: Magnetic resonance imaging is an essential element for establishing the etiologic diagnosis of neurogenic dysphagia.

Key words: oropharyngeal dysphagia, Chiari malformation, syringomyelia.


**ABSTRACT**

Dysphagia is difficulty in swallowing or transit of food from the mouth to the stomach. Its causes are classified into two clinical categories: oropharyngeal and esophageal. The first reflects the dysfunction in the different phases of the process of swallowing and can originate from lesions in the different structures of the central nervous system as well as neuromuscular and neurodegenerative diseases. In type I Chiari malformation (TICM), caudal displacement of the cerebellar tonsils through the foramen magnum may cause a lower cranial nerve injury and dysfunction of the brain stem, which in some patients manifests as dysphagia and may be concomitant with syringomyelia. The timely recognition and diagnosis of dysphagia can be achieved with the appropriate signs and symptoms along with a detailed physical examination and imaging studies as in this case.

**CLINICAL CASE**

A 62-year-old female patient was admitted in January 2010 to the General Regional Hospital, Morelia, Michoacan, with a history of systemic arterial hypertension and primary open angle glaucoma of long evolution. Her disorder began 6 years prior with nonprogressive dysphagia of solids and liquids, sense of choking and cough, and nonquantified weight loss. On directed questioning the patient noted no heartburn, sialorrhea or epigastric pain. Neurological examination was notable for bilateral palpebral ptosis (Figure 1), peripheral facial diplegia and loss of pharyngeal reflex. Muscular strength in the upper and lower extremities was 3/5 according to the Daniels scale and generalized hyporeflexia was diagnosed. Dysesthesias and progression to segmental anesthesia dissociative in character, as well as cervicobrachialgia of left dominance and occasional periods of dizziness, were observed. Based on the signs and symptoms it was suspected that she suffered from dyspha-
changes. An electromyogram was done with monopolar needle of the left pharyngeal, mas- 
seter, mentalis, deltoids, long supinator and 
adductor muscles of the thumb with prolonged
insertion, indicating generalized sensory and 
motor polyneuropathy with predominance of the 
latter and whose changes were compatible with 
mild to moderate myelin involvement without 
axonal damage. Immunological studies were 
positive for antinuclear antibodies (1:320) with 
homogeneous fluorescence pattern (Table 1).

### Table 1. Laboratory studies.

<table>
<thead>
<tr>
<th>Test</th>
<th>Result</th>
</tr>
</thead>
<tbody>
<tr>
<td>CRP</td>
<td>Negative</td>
</tr>
<tr>
<td>LE cells</td>
<td>Negative</td>
</tr>
<tr>
<td>Antinuclear antibodies</td>
<td>Positive</td>
</tr>
<tr>
<td>Smooth muscle antibodies</td>
<td>Negative</td>
</tr>
<tr>
<td>Antimitochondrial antibodies</td>
<td>Negative</td>
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CRP, C-reactive protein; LE, lupus erythematosus.

The patient underwent median suboccipital cra-
niectomy with resection of the posterior arch of 
C1, surgery of the dura mater and coagulation of 
the cerebellar tonsils, which were found down 
to C1. Management of the generalized sensory-
motor polyneuropathy was based on prednisone 
administration at a dose of 1 mg/kg/day.

In the following 12 weeks, mental functioning 
was noted to be without alterations, cranial 
nerves without lesion, muscle strength in the 
upper and lower extremities were 4/5 on the 
Daniels scale and sensory system with paresthe-
sias in the left upper extremity.

**DISCUSSION**

Oropharyngeal dysphagia is a common symptom 
in patients with neurological diseases that may 
be due to central or peripheral nervous system 
lesions or to muscular diseases and neuromus-
cular junction, which leads to sensory and motor 
deterioration of the oral and pharyngeal phases 
of the swallowing mechanism. The physiological 
process of swallowing can be divided into three 
phases: oral, pharyngeal and esophageal. Under 
normal conditions the swallowing mechanism is
based on the precise coordination of numerous afferent and efferent somatic and visceral pathways, which begins as the tongue guides the food towards the posterior part of the buccal cavity and bolus comes in contact with the posterior wall of the oropharynx. A defect at swallowing initiation may indicate a deficiency in the contraction of the elevator and tensor muscles of the soft palate, which close the nasopharynx and prevent nasal regurgitation as in the case of myasthenia gravis or of a disorder of cranial nerves IX and X, secondary to lesions due to elongation, as well as alterations due to compression in the region of the brain stem, especially in the swallowing center as in tumors of the posterior fossa, vascular lesions and TICM.

The Chiari malformation, described in 1891 by Dr. Hans Chiari, is characterized by changes in the development of the posterior fossa, craniovertebral junction, bulbomedullary junction and spinal cord. Four well-defined types are recognized, with type I being detected with greater frequency in adulthood. TICM may be congenital or acquired and is characterized by caudal displacement of the cerebellar tonsils at least 3-5 mm through the foramen magnum towards the cephalic portion of the cervical conduit, which could co-exist with syringomyelia or hydrocephalus.

The clinical presentation in patients with concomitant syringomyelia varies from the absence of symptoms to pain, weakness and sensory disorders. This type of malformation has been related with syringomyelia in 30-70% of cases and the most accepted explanation is based on the theory by Oldfield et al. in which herniation of the cerebellar tonsils propels the cerebral spinal fluid of the spinal subarachnoid space to the interior of the cord, causing a dilation that can be seen on magnetic resonance. With respect to the clinical manifestations, the patient demonstrated the typical description reported in the literature: thermoalgetic dissociation; however, pain is a nonspecific symptom, indicating that patients are not diagnosed in an early and timely fashion as in this case.

In a series described by Milhorat et al. who evaluated 364 symptomatic patients, the most common symptom was suboccipital headache (81%), whereas the clinical manifestations of the brain stem, cerebellum and lower cranial nerve alterations were corroborated in 52% of the subjects. In the group of TICM concomitant with syringomyelia, there were 100 patients with dysphagia. In the series reported by Guinto et al. from the Hospital de Especialidades, Centro Medico Nacional Siglo XXI, 41% of 48 patients had dysphagia secondary to hernia of the cerebellar tonsils and brain stem compression. Analysis of the case reported here suggests that TICM with syringomyelia is the cause of the dysphagia, and its physiopathological mechanism is defined principally by changes of the brain stem itself as well as by elongation of the lower cranial nerves due to caudal displacement of the spinal cord. In a study by Pollack et al. a mechanism was established based on the dysfunction of the brain stem secondary to compression, ischemia and intrinsic dysgenesis. This directly affects the swallowing center and is considered as the probable mechanism in this case. It is defined by a deficiency in the integrated sequence of the neuromuscular activity for swallowing in the medulla oblongata in the region of the nucleus of the solitary fasciculus and the adjacent reticular formation, the reason why the patient required medical attention. In adults with TICM with or without syringomyelia, the insidious advance of dysphagia leads to an important diagnostic delay because absence of other clinical findings of the brain stem could be mistakenly attributed to local pharyngoesophageal abnormalities such as a hiatal hernia, peptic esophagitis or stenosis, as in this case. Initially, these patients frequently experience loss of the involuntary component of the oral phase of swallowing, which may be compensated; however, if not
treated, patients may gradually develop cough and choking sensation with solids and liquids as described in this article. Lack of recognition of the characteristics of oropharyngeal dysphagia leads to the performance of gastroenterology and ENT procedures that may be unnecessary so that when diagnosis is established, patients already have difficulty in carrying out daily activities as well as experiencing secondary effects such as chronic malnutrition and aspiration pneumonia. The latter represents another factor related with respiratory function, believed to be one of the most important complications of dysphagia due to the fact that these patients cannot maintain a sufficient period of apnea while swallowing, therefore increasing morbidity and mortality. During clinical evaluation of these patients, differentiation with other diseases such as the lateral bulbar syndrome, amyotrophic lateral sclerosis, oculopharyngeal muscular dystrophy, Alzheimer’s disease and Parkinson’s disease should be considered. It is of note that the patient suffered bilateral peripheral facial paralysis, palpebral ptosis and electromyographic changes compatible with myelin involvement unexplained by TICM, which suggested the possible co-existence of a chronic inflammatory demyelinating polyradiculoneuropathy that was treated with corticosteroids. However, treatment with gammaglobulin or plasmapheresis is preferred over long-term prednisone administration. The systemic autoimmune chronic inflammatory demyelinating polyradiculoneuropathy component can, in turn, explain the presence of antinuclear antibodies in the patient. The relationship between facial paralysis and the disorder of the swallowing process has not been systematically studied. Cranial nerve VII contributes to the oropharyngeal phase of the swallowing process, and so the oral orbicular and buccinator muscles gain importance in the oral phase of the swallowing mechanism. Regarding the case described here, it is probable that this condition contributed to the severity of the dysphagia.

Magnetic resonance imaging is a fundamental method to evaluate abnormalities of the craniovertebral junction as well as diseases of the spinal cord and allows establishing objective anatomic criteria with clinical and prognostic significance. With respect to the radiological findings, displacement of the cerebellar tonsils and syringomyelia is highly variable. Radiological criteria on the degree of hernia of the cerebellar tonsils in TICM are not absolute and the indication for surgical management should be made based on an objective analysis of the entire clinical pathological context. The literature supports that the fundamental problem of TICM is that of a volumetrically small posterior cranial fossa, leading the clinical presentation of this disease to be diverse. Videofluoroscopy is considered the gold standard to study the oropharyngeal mechanisms of dysphagia, if and when this technological support is available for evaluating the swallowing function. Management of TICM is controversial because of the many available surgical options. In general, patients with concomitant symptoms of TICM and syringomyelia are considered surgical candidates, independent of the degree of herniation of the cerebellar tonsils. Asymptomatic patients with medullary cavity ≥ 50% of the transversal diameter of the spinal cord will also be considered. Occipital craniectomy or decompression of the posterior fossa combined with C1 laminectomy (LC1) plus surgery of the dura mater in its total thickness is an important procedure. Patients with symptoms of brain stem compression improve dramatically with decompression of the posterior fossa, whereas the symptoms of syringomyelia wane slightly or are stabilized. Reduction in the size of the cerebellar tonsils by means of bipolar coagulation allow for the cerebral spinal fluid circulation to be re-established and to decrease the size of the syringomyelia as well as to halt the advance of the disorder. As seen in this patient, clinical manifestations improved with this surgical procedure.
In conclusion, this case illustrates the importance of carrying out an adequate diagnostic evaluation of a patient with oropharyngeal dysphagia with neurological clinical data that should prompt suspicion for the existence of central and peripheral nervous system abnormalities and neuromuscular diseases. Dysphagia is only one part of the clinical context in numerous neurological disorders or may be the main characteristic, such as described in the TICM of this report. For this reason, physical examination should be directed, complete and specific on the topographical areas that govern swallowing. Only in this manner will earlier diagnoses and adequate timely treatment be achieved. Timely recognition of neurogenic dysphagia is essential for a favorable postoperative result.

REFERENCES


