Spontaneous improvement in syringomyelia in a patient with Chiari 1 malformation: illustrative case

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BACKGROUND “Chiari malformation” refers to a spectrum of hindbrain abnormalities characterized by impaired cerebrospinal fluid circulation through the foramen magnum. Syringomyelia is frequently found in patients with Chiari malformation type 1. Although many theories have suggested how cerebrospinal fluid enters and makes the cystic cavity in the spinal cord, the pathogenesis of syringomyelia remains controversial. This report documents a case with spontaneous resolution of syringomyelia followed up by 3-year serial magnetic resonance imaging (MRI). These kinds of cases support a more conservative approach.

OBSERVATIONS A 59-year-old female presented to the authors’ clinic in June 2019 with a history of Chiari malformation type 1. This symptomatic patient has been followed up with serial MRI. When the last MRI was performed in August 2022, compared with previous imaging, resolution of the syringomyelia was recognized.

LESSONS Because the natural evolution of mildly symptomatic/asymptomatic patients with syringes is unclear, these patients pose a treatment dilemma. Although surgical intervention is a widely accepted therapeutic method, a more conservative approach can be considered in cases with spontaneous resolution. Especially for patients without progressive symptoms, the surgical approach should not be considered as the first step. In view of relapses, follow-up with periodic neurological examinations and radiological imaging is preferable.

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KEYWORDS syrinx; syringomyelia; Chiari malformation type 1; magnetic resonance imaging; cerebrospinal fluid

Chiari malformations are characterized by the degree of herniation of the cerebellar tonsils through the foramen magnum, and they include congenital hindbrain herniation. The most common subtype among them is Chiari malformation type 1 (CM1). It was described by Arnold Chiari in 1891.1 The prevalence of CM1 in the population is between 0.24% and 0.9%.2 Radiographically, it is defined as the displacement of the cerebellar tonsils of 5 mm or more below the McRae line.3 Approximately 50% to 75% of patients with this condition may develop syringomyelia.4 There are still many theories about the development of syringomyelia, and it is not clearly illuminated. We present a case of the spontaneous resolution of syringomyelia in an adult patient with CM1 and associated syringomyelia.

Illustrative Case

A 59-year-old female presented to our clinic in June 2019 with a history of CM1. She had concomitant symptoms such as cervical and bilateral upper extremity pain, numbness, and hypoesthesia spread through the C6–T7 dermatomes for about 5–6 months. However, she did not have any headache, motor weakness, or gait and balance dysfunction. The patient followed up with serial magnetic resonance imaging (MRI) initially for 6 months then at 1-year periods.

The first cervical MRI performed in August 2019 at our clinic revealed a syrinx extending from C3 to C5 with a maximum diameter of 7 mm. Follow-up MRI in August 2021 demonstrated a reduced size of the syrinx in terms of both diameter and length. When the last MRI was performed in August 2022, as compared with previous imaging, resolution of the syrinx was recognized. But there was no apparent difference in terms of herniation of cerebellar tonsils (Fig. 1). Also, clinical symptoms and complaints of the patient had disappeared. Therefore, we did not suggest the patient for surgery because she did not fulfill the criteria for surgery.

ABBREVIATIONS CM1 = Chiari malformation type 1; CSF = cerebrospinal fluid; MRI = magnetic resonance imaging.

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Patient Informed Consent
The necessary patient informed consent was obtained in this study.

Discussion
Observations
Syringomyelia can be defined as a disruption of cerebrospinal fluid (CSF) flow in the subarachnoid space with a fluid-filled space in the spinal cord and progressive neurological impairment. This term was first used by Ollivier d’Angers in 1827. Syringomyelia can be associated with trauma, tumor, or congenital anomalies, or it can be seen as idiopathic. Symptoms include headache, weakness and numbness of the extremities, and nystagmus. Although the relationship between CM1 and syringomyelia has been accepted, the pathogenesis of syringomyelia is still controversial. Various theories have been put forward in the literature to explain the formation of the syrinx.

One of them is named the “craniospinal pressure dissociation theory,” which was proposed by Williams. It suggests that each coughing, straining, and Valsalva maneuver creates a pressure difference between the intracranial and intraspinal compartments. Because of the relatively higher pressure of the intracranial compartment than in the intraspinal one, intracranial CSF in the fourth ventricle is driven into the central canal; thus, the syrinx is formed. Gardner’s hydrodynamic theory proposed that when there is crowding or obstruction of the posterior fossa, the CSF is unable to pass out of the fourth ventricle and is redirected to the central canal through the obex by every systolic pulsation. It can be described as a “water-hammer effect” in the syrinx that creates the progression of cavitation. But the fourth ventricle foramina remains patent in many cases, so it causes a conflict with this theory. In addition, many radiological and anatomical findings are inadequate to confirm communication between the syrinx and the fourth ventricle. Milhorat et al. assert that when foramen magnum outflow is obstructed, CSF passes into the spinal cord via Virchow-Robin spaces due to high pressure created by subarachnoid obstruction. Another theory, which is named the “tonsillar piston theory,” proposed that cerebellar tonsils act as a “piston” on the spinal CSF space when there is an obstruction at the foramen magnum, so systolic pressure develops on the spinal cord. However, recent studies suggest that the syrinx fluid is not originated from the CSF but is composed of extracellular fluid from the spinal cord microcirculation.

About 21 adult patients have been reported in whom the resolution of spontaneous syringomyelia by MRI without surgical intervention has been documented (Table 1). Various theories have been proposed to explain this spontaneous resolution phenomenon as well. The communication between syrinx and subarachnoid space by tear or fissure formation during a Valsalva-like maneuver is another theory that causes resolution of the syrinx. Also, spontaneous rupture of arachnoid thickening or scarring at the foramen magnum or foramen of Magendie improves the flow of CSF and results in the resolution of the syrinx.

Lessons
Because the natural evolution of mildly symptomatic patients or asymptomatic patients with syringes is unclear, these patients pose a treatment dilemma. Although surgical intervention is a widely accepted treatment method, a more conservative approach can be considered with the occurrence of cases with spontaneous resolution. Especially for patients without progressive symptoms, the surgical approach should not be considered as the first step. In other words, the progressive deterioration of symptoms, such as cervicobrachialgia, sensory deficits, especially motor deficits that affect the quality of life, and the worsening appearance of the syringomyelia on radiological imaging, would warrant surgical intervention. Due to the poorly understood pathophysiological mechanisms and the possibility of relapse, these patients can be followed up with periodic neurological examinations and radiological imaging. Meanwhile,
patient selection for observation must be carefully weighed. The decision to perform either surgery or follow-up is critically important and based on risk–benefit analysis.

References


TABLE 1. Adult patients with spontaneous syringomyelia with CM1 reported in the literature

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Age (yrs), Sex</th>
<th>Follow-Up</th>
<th>Syringomyelia Resolution</th>
</tr>
</thead>
<tbody>
<tr>
<td>Jack et al., 1991</td>
<td>30, F</td>
<td>2 yrs</td>
<td>−</td>
</tr>
<tr>
<td>Santoro et al., 1993</td>
<td>39, M</td>
<td>37 mos</td>
<td>−</td>
</tr>
<tr>
<td>Pierallini et al., 1997</td>
<td>25, F</td>
<td>5 yrs</td>
<td>+</td>
</tr>
<tr>
<td>Sudo et al., 1998</td>
<td>34, F</td>
<td>7 yrs</td>
<td>+</td>
</tr>
<tr>
<td>Fukutake et al., 1998</td>
<td>40, F</td>
<td>2 mos</td>
<td>?</td>
</tr>
<tr>
<td>Klekamp et al., 2001</td>
<td>37, M</td>
<td>32 mos</td>
<td>+</td>
</tr>
<tr>
<td>Itoyama et al., 2001</td>
<td>54, M</td>
<td>1 yrs</td>
<td>−</td>
</tr>
<tr>
<td>Kyoshima et al., 2003</td>
<td>39, M</td>
<td>6 mos</td>
<td>−</td>
</tr>
<tr>
<td>Coppa et al., 2006</td>
<td>27, F</td>
<td>?</td>
<td>+</td>
</tr>
<tr>
<td>Sung et al., 2008</td>
<td>43, F</td>
<td>2 yrs</td>
<td>−</td>
</tr>
<tr>
<td>Deniz et al., 2009</td>
<td>30, M</td>
<td>11 yrs</td>
<td>−</td>
</tr>
<tr>
<td>Tortora et al., 2012</td>
<td>21, M</td>
<td>3 yrs</td>
<td>−</td>
</tr>
<tr>
<td>Vaquero et al., 2012</td>
<td>58, M</td>
<td>2 yrs</td>
<td>−</td>
</tr>
<tr>
<td>Muthukumar et al., 2013</td>
<td>24, F</td>
<td>3 yrs</td>
<td>+</td>
</tr>
<tr>
<td>Jain et al., 2017</td>
<td>32, F</td>
<td>30 mos</td>
<td>−</td>
</tr>
<tr>
<td>Yuan et al., 2019</td>
<td>36, F</td>
<td>10 yrs</td>
<td>+</td>
</tr>
<tr>
<td>Gallo et al., 2021</td>
<td>46, F</td>
<td>10 yrs</td>
<td>−</td>
</tr>
<tr>
<td>Present case</td>
<td>59, F</td>
<td>3 yrs</td>
<td>−</td>
</tr>
</tbody>
</table>

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Disclosures
The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions
Conception and design: Celtikci, Ozturk. Acquisition of data: Ozturk, Cindil. Analysis and interpretation of data: Ozturk, Cindil. Drafting the article: Celtikci, Ozturk, Cindil. Critical revising the article: Celtikci, Ozturk, Emmez, Kuzucu. Reviewed submitted version of manuscript: Celtikci, Ozturk, Kuzucu. Approved the final version of the manuscript on behalf of all authors: Celtikci. Administrative/technical/material support: Celtikci, Kuzucu. Study supervision: Celtikci.

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