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Syringomyelia intermittens: highlighting the complex pathophysiology of syringomyelia. Illustrative case

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BACKGROUND Chiari Type I malformation (CM1) is a disorder recognized by caudal displacement of the cerebellar tonsils through the foramen magnum and into the cervical canal. Syringomyelia is frequently found in patients with CM1, but the pathophysiology of syringomyelia remains an enigma. As a general consensus, symptomatic patients should be treated and asymptomatic patients without a syrinx should not be treated. Mildly symptomatic patients or asymptomatic patients with a syrinx, on the other hand, pose a more challenging dilemma, as the natural evolution is uncertain. For many surgeons, the presence of a syrinx is an indication to offer surgery even if the patient is asymptomatic or mildly symptomatic.

OBSERVATIONS The authors describe an illustrative case of a 31-year-old female with an incidental finding of a CM1 malformation and cervical syrinx in 2013. Conservative management was advocated as the patient was asymptomatic. Monitoring of the syrinx over a course of 8 years showed resolution, followed by reappearance and finally a complete resolution in 2021. A review of the literature and the possible pathophysiology is discussed.

LESSONS The unusual course of this patient highlights the importance of guiding treatment by clinical symptoms, not radiological findings. Furthermore it reflects the complexity of the pathophysiology and the uncertain natural history of syringomyelia.

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KEYWORDS intermittent; syringomyelia; Chiari 1

Chiari Type I malformation (CM1) is a disorder characterized by caudal displacement of 1 or both of the cerebellar tonsils through the foramen magnum and into the cervical canal. It was originally described by Arnold Chiari in 1891 as an "elongation of the tonsils and the medial part of the inferior lobes of the cerebellum into cone-shaped projections which accompany the medulla oblongata into the spinal canal."[1] Cerebellar tonsillar ectopia is generally considered pathological when greater than 5 mm below the foramen magnum or McRae line. The 5-mm limit originates from older studies comparing the descent of cerebellar tonsils in healthy volunteers with those in patients who had symptomatic CM1.[2,3] The estimated prevalence of CM1 is probably between 0.24–0.9% of the population, and the majority of these patients are asymptomatic.[2–5]

Despite CM1 being studied for 130 years, many questions regarding the pathophysiology and expected evolution still remain.[1] Symptomatic patients should be treated and asymptomatic patients without a syrinx should not be treated. As the natural evolution of mildly symptomatic patients or asymptomatic patients with a syrinx is uncertain, they pose a greater treatment dilemma, For many surgeons the presence of a syrinx is an indication to offer surgery even if the patient is asymptomatic or mildly symptomatic.[6] However, as this case illustrates, resolution or recurrence of a syrinx might occur spontaneously.

Illustrative Case

Eight years ago, a 24-year-old female was involved in a car accident, with brief loss of consciousness. As part of the radiological trauma

ABBREVIATIONS CM1 = Chiari Type I malformation; CSF = cerebrospinal fluid; CT = computed tomography; MRI = magnetic resonance imaging; PFD = posterior fossa decompression; PFDD = posterior fossa decompression with duraplasty; SAS = subarachnoid space.

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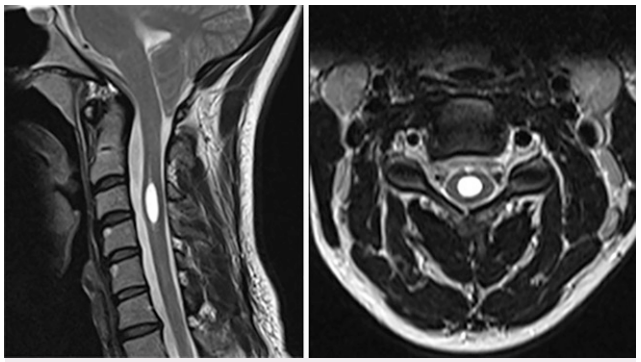

FIG. 1. Left: Sagittal T2-weighted spin-echo MRI February 2013. **Right:** Axial T2-weighted spin echo MRI in February 2013.

protocol, computed tomography (CT) of the brain and full spine was performed. No intracranial or spinal traumatic lesions were seen. However, a CM1 was seen with a suspicion of a syrinx at C3. Further investigations with magnetic resonance imaging (MRI) of the brain and full spine confirmed a CM1 with a tonsillar descent of 12 mm reaching down to the posterior arch of C1 and an associated syrinx at C3 measuring 6 $\times$ 18 mm (Fig. 1A and B). The MRI of the full spine did not show any other abnormality.

As the patient was completely asymptomatic, conservative management was recommended with follow-up MRI in 6 months. This follow-up MRI showed an almost complete resolution of the syrinx as well as a mild improvement of the tonsillar descent at the foramen magnum (Fig. 2).

Two years later, another MRI was performed, with the expectation of seeing a complete resolution of the syrinx; however, this MRI showed a recurrence of the syrinx with the cerebellar tonsils sitting again at the level of the posterior arch of C1 (Fig. 3). At the time, the patient was still asymptomatic, and the decision was made to repeat imaging if the patient developed symptoms.

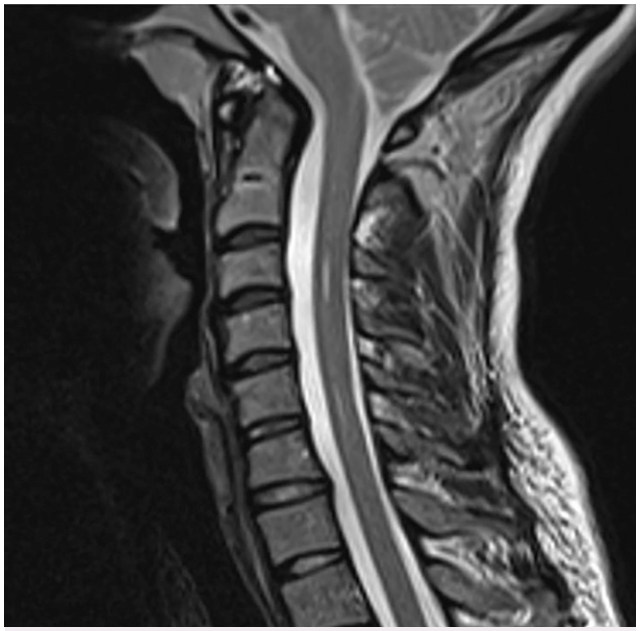

FIG. 2. Sagittal T2-weighted spin-echo MRI in July 2013.

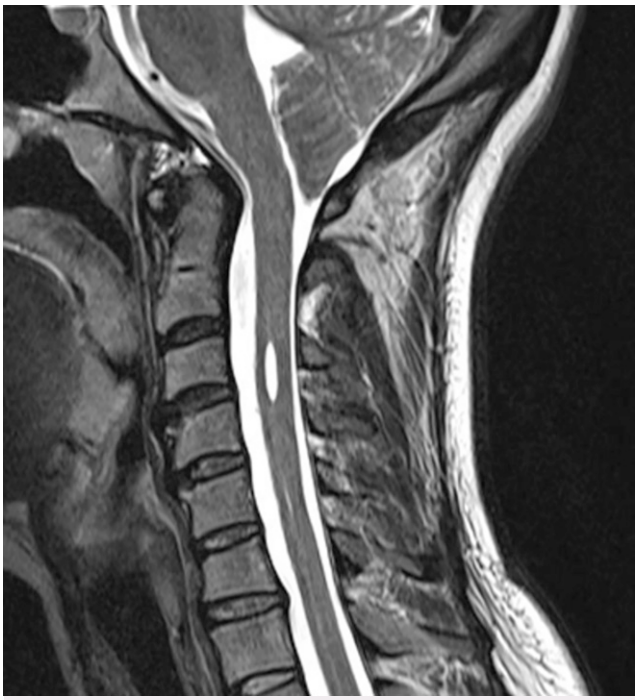

FIG. 3. Sagittal T2-weighted spin-echo MRI in June 2016.

Recently, the patient was referred back to the Neurosurgical Department by her primary care provider with severe persistent neck pain and occipital headaches following an awkward maneuver during housework. An MRI of the brain and full spine was performed, demonstrating complete resolution of the syrinx and significant improvement of the tonsillar descent (Fig. 4). The patient was seen in the outpatient clinic and mentioned a resolution of her symptoms after a short course of nonsteroidal antiinflammatory drugs. The anamnesis was most compatible with a muscular or facetogenic cause for these transient symptoms.

Discussion

Observations

The exact pathophysiology of syringomyelia due to CM1 remains frustratingly uncertain. Five theories have been proposed. The first theory was proposed by Gardner and Angel in 1958; the "water-hammer" theory states that "partial obstruction of the outflow of cerebrospinal fluid (CSF) from the fourth ventricle directs the systolic pulsations of CSF from the fourth ventricle through a patent central canal."[7] The second theory was the "cranial-spinal pressure dissociation" theory proposed by Williams in 1969, stating that a block in caudal, but not upward, CSF flow causes an increase of intracranial pressure over spinal intrathecal pressure and, as such, forces CSF from the fourth ventricle down into the central canal, producing "communicating" syringomyelia.[8] Therefore, both postulate communication between the fourth ventricle and the syrinx via the obex and the central canal; however, this communication is seldom seen on MRI or autopsy.[9]

A third theory published in 1972 by Ball and Dayan suggested that the tonsils obstruct the rapid rostral movement of CSF from the spinal subarachnoid space (SAS) to the cranial SAS during transiently increased thoracic venous pressure, causing sporadic increased

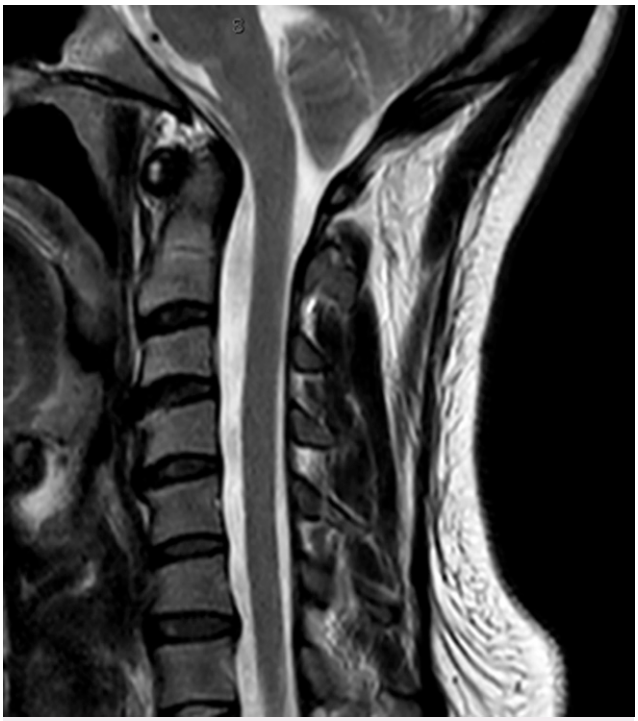

FIG. 4. Sagittal T2-weighted spin-echo MRI in February 2021, showing disappearance of the syrinx and clear CSF high T2 signal at the foramen magnum and rounder cerebellar tonsils.

spinal CSF pressure; this forces the CSF along extracellular paths through the spinal cord surface to initiate syringomyelia.[10] The work by Oldfield et al. showed compression of the upper part of the syrinx during systole; the theory was refuted.[11] However, the role of the dilated Virchow–Robin spaces to initiate syringomyelia has been confirmed.[12]

Based on this theory, a fourth hypothesis was introduced by Oldfield and colleagues, "the piston theory." This theory suggests that "syrinx formation is caused by the cerebellar tonsils acting as a piston to produce large pressure waves in the spinal subarachnoid space, and this action forces fluid through the surface of the spinal cord or the perivascular spaces into the central canal. Once formed, such syringes are believed to enlarge due to external compression of the cord."[11]

Stoodley et al. suggested a fifth theory stating that CSF normally flows along the perivascular spaces from the spinal subarachnoid space, but perturbations of this flow caused by Chiari malformations or other conditions are responsible for the initiation of a cyst formation and enlargement (due to increased inflow or reduced outflow).[12]

Despite more than half of century of research, it is still unclear why some patients with CM1 develop a syrinx and others do not. Leung et al. showed no difference in cerebellar tonsillar motion between patients with and without syrinx.[13] And although most patients with a CM1 have a "crowded" posterior fossa, a small number of patients have a spacious posterior fossa but nonetheless demonstrate herniation of their tonsils.[14]

Whatever the exact pathophysiology might be for the development of syringomyelia in patients with CM1, we know that surgically restoring CSF flow at the foramen magnum or bypassing the obstruction with a shunt alleviates the symptoms and the syrinx.[3,12,15,16]

Spontaneous resolution has been reported frequently in children due to growth of the skull and subsequent restoration of CSF flow.[17] In adults, on the other hand, the treatment for symptomatic syringomyelia is generally surgery.[14]

A wide range of techniques have been described from simple bony decompression to more extensive surgery with coagulation or aspiration of tonsils and duraplasty. In some cases, the extra room required at the foramen magnum for successful surgery may be only a fraction of a millimeter.[12]

A recent systematic review and metaanalysis compared the 2 most commonly used techniques: posterior fossa decompression with duraplasty (PFDD) and without duraplasty (PFD).[18] They evaluated 3618 pediatric and adult patients. The reported results with both techniques were excellent: a clinical improvement of 86.8% in the PFDD group (77.6% improvement in adults, 93.3% in children) versus a 69.8% in the PFD cohort (62.9% improvement in adults, 74.8% in children).[18] As simple bony decompression is not sufficient in about 10% of the adult patients, it again reflects the heterogeneity of the patient population.[13] When a duraplasty is performed, either an autologous graft (pericranium) or a nonautologous graft (synthetic, bovine pericardium, collagen-based, allograft) can be used. A recent metaanalysis shows superiority of autologous grafts versus nonautologous in terms of complications.[19]

Despite an estimated prevalence of CM1 between 0.24–0.9%, there is a paucity of data reporting on the natural history of adult CM1. Langridge et al. performed a systematic review on the natural history of patients with CM1 and could only include 1 article describing the natural history in asymptomatic patients and 2 articles following up symptomatic patients who were conservatively managed.[6] Five asymptomatic patients were followed for a mean of 2 years and remained asymptomatic.[6]

Twenty cases of spontaneous resolution of a syrinx in patients with CM1 have been previously described.[20–36] In about half of them, an association with improvement of the tonsillar herniation is described, similar to our case. Recurrence after spontaneous resolution has been described in only 1 other case.[26] This is the first report of a spontaneous resolution after a recurrence.

An improvement of CSF flow at the foramen magnum due to spontaneous rupture of the arachnoid adhesions, typically encountered at the foramen magnum, has been suggested by 4 different authors.[25,27,33,35] Others have reported spontaneous improvement of CM1 due to brain atrophy in ageing patients.[37,38]

Lifestyle changes, such as avoiding straining and breath holding and ceasing hard physical labor, have been described, leading to a resolution of syringomyelia due to reduced venous pressure.[6,11] A prospective trial was conducted by the group of Oldfield, which included 18 patients with CM1 and showed a significant improvement of CSF flow at the foramen magnum by simply flexing the head.[39] Apart from starting meditation, our patient did not have any significant lifestyle changes. Whether meditation caused a change in posture or reduced intrathoracic pressure is questionable, although it could explain the intermittent character.

Raised intracranial pressure due to a space-occupying lesion (chronic subdural hematomas, cavernoma), hydrocephalus, head injuries, and venoocclusive disease have been described as a cause for tonsillar herniation. None of these was present in our patient.[40–44] A CSF leak as a cause of reduced intracranial pressure and subsequent resolution of CMI has also been reported.[28]

Other pathophysiological mechanisms for spontaneous resolution have been proposed, when a change in tonsillar descent is absent or less clear. In 1991, Jack et al. published a hypothesis of spontaneous drainage of a syrinx into the spinal subarachnoid space, due to pressure necrosis of the spinal cord,[20] a hypothesis supported by Santoro et al. and Multani et al., who had MRI findings suggestive of a CSF channel between the syrinx and the subarachnoid space.[21,36] The efficacy of syringosubarachnoid shunts in the treatment of syringomyelia supports this.[14] However, this would be in conflict with the theory of Oldfield and Heiss that the mechanism of a syrinx origin and resolution arises outside, not inside, the spinal cord.[45]

It is possible that there are 2 distinct pathophysiological mechanisms leading to spontaneous resolution of a syringomyelia; however, as can be seen in our case, a subtle decrease in tonsillar herniation might easily be missed, and a subtle improvement in flow at the craniocervical junction might be sufficient to restore CSF flow and resolve the syrinx. Both mechanisms (spontaneous improvement of CSF flow at the foramen magnum and spontaneous drainage of the syrinx into the spinal subarachnoid space) support the hypothesis of Stoodley et al. that cyst volume and pressure are a function of the balance between inflow and outflow of CSF via the perivascular spaces.[11]

Lessons

The unusual course of this patient highlights the importance of guiding treatment by clinical symptoms, not radiological findings. Furthermore, it reflects the complexity of the pathophysiology and the uncertain natural history of syringomyelia.

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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: Van Der Veken, Hatami. Acquisition of data: Van Der Veken, Agzarian. Analysis and interpretation of data: Van Der Veken, Hatami, Agzarian. Drafting the article: Van Der Veken. Critically revising the article: Hatami, Harding, Agzarian, Vrodos. Reviewed submitted version of manuscript: Van Der Veken, Harding, Hatami, Vrodos. Approved the final version of the manuscript on behalf of all authors: Van Der Veken. Administrative/technical/material support: Van Der Veken. Study supervision: Harding, Vrodos.

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