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Successful treatment of unilateral facial nerve palsy in a pediatric patient with syringobulbia and Chiari malformation type I: illustrative case

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BACKGROUND Unlike syringomyelia, syringobulbia is not commonly observed in pediatric patients with Chiari malformation type I (CMI). Previous series have reported the incidence of syringobulbia as between 3% and 4% in these patients. Presentation is typically chronic, with the slow onset of neurological symptoms and cranial nerve (CN) palsies resulting from lower brainstem involvement. The authors report the first case of a pediatric patient with simultaneous CMI, syringobulbia, and unilateral CN VII palsy.

OBSERVATIONS A 7-year-old male presented with right facial weakness in addition to headaches, ataxia, urinary incontinence, and falls. Magnetic resonance imaging revealed CMI with a syrinx of the cervicothoracic spine and syringobulbia. Posterior fossa decompression with duraplasty was performed without complications, and the patient was discharged home on postoperation day 5. At the 3-week follow-up, the patient's neurological deficits had largely subsided. At the 3-month follow-up, his CN VII palsy and syringobulbia had completely resolved.

LESSONS Pediatric CMI patients with syringomyelia are at risk for developing syringobulbia and brainstem deficits, including unilateral facial palsy. However, craniocervical decompression can prove successful in treating such deficits.

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KEYWORDS Chiari malformation type I; syringobulbia; syringomyelia; cranial nerve palsy; facial nerve palsy; decompression

Chiari malformation type I (CMI), defined as caudal displacement of the cerebellar tonsils at least 5 mm below the foramen magnum, is the most common of the four types of Chiari malformation (I–IV).[1] Patients with CMI can present with a range of symptoms because of direct compression of nearby neural tissues. The most common of these include headache, neck pain, torticollis, oropharyngeal dysfunction, abnormal ocular mobility, and gait disturbances.[2] Radiographically, syringomyelia (a fluid-filled cavity, or syrinx) of the cervicothoracic spinal cord is a condition commonly observed on spinal imaging. Syringobulbia, a fluid-filled cavity in the brainstem resulting from extension of the syrinx from the upper cervical cord, is rarely (4%) observed in CMI, usually in conjunction with syringomyelia.[3] Common symptoms associated with syringobulbia include those expected from lower brainstem involvement, such as cranial nerve (CN) IX–X palsy, as well as sensory and motor deficits resulting from interruption of the ascending and descending tracts.[3] Patients will usually experience slowly progressive symptoms such as vertigo, loss of pain and temperature sensation in the face, atrophy and fibrillation of the tongue, dysphonia, vision changes, and ataxia.[3] Nystagmus can be observed on clinical examination.[3] Treatment for CMI and its various symptoms is posterior fossa decompression to relieve the crowding that occurs at the craniocervical junction.

In the present study, we discuss a rare case involving a pediatric patient who presented with CMI, syringomyelia, syringobulbia, unilateral CN VII dysfunction, and ataxia. This is, to our knowledge, the only reported case of CMI presenting with unilateral CN VII palsy and syringobulbia. Furthermore, to characterize the rarity of this presentation and provide a comprehensive summary of clinical symptomatology and surgical management strategies associated with CMI, we systematically and comprehensively reviewed the neurosurgical literature.

Illustrative Case

A 7-year-old male presented with unsteady gait, right-sided facial weakness, intermittent right parietal headaches, and emesis. The

ABBREVIATIONS CMI = Chiari malformation type I; CN = cranial nerve; CSF = cerebrospinal fluid; MRI = magnetic resonance imaging; SSS = syringo-subarachnoid shunt.
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patient was noted to veer to the right when walking and reportedly fell multiple times at school. He also had a few episodes of urinary incontinence. With respect to pertinent negatives, the patient had no changes in vision or hearing, numbness, tingling, weakness, or problems with swallowing. He had mild right nystagmus on physical examination. He had no history of surgery, trauma, or major illness. Magnetic resonance imaging (MRI) showed CMI, a visualized cervical and thoracic spine syringomyelia, and syringobulbia (Fig. 1A, B, and D).

Craniocervical decompression for CMI was performed with no complications. Intraoperatively, the cerebellar tonsils were compressed against the spinal cord. Bipolar cauterization was used to shrink both cerebellar tonsils up to the level of the obex. Membranes were found to cover the obex, and adhesions were found to cover the brainstem and floor of the fourth ventricle. These were removed using microscissors and microhooks. Adequate cerebrospinal fluid (CSF) flow was visualized. A pericranial graft was obtained and, using 4–0 Nurolon, was sutured in place. A Valsalva maneuver was performed by the anesthesiologist. There was no evidence of CSF leakage. The incision was closed without complications.

The patient was discharged 5 days postoperation. The 3-week postoperative follow-up revealed a drastic reduction in the preoperative symptoms of unsteady gait and right-sided facial weakness. The 9-week postoperative follow-up showed complete resolution of the preoperative symptoms including the right-sided facial weakness. Postoperative MRI showed significant improvement in the syringomyelia and syringobulbia (Fig. 1C and E).

Patient Informed Consent

The necessary patient informed consent was obtained in this study.

Discussion

Methods

To characterize the clinical presentations, treatments, and outcomes associated with syringobulbia in CMI better, we conducted a systematic literature search using the PubMed, Google Scholar, and Web of Science databases. We queried these databases for

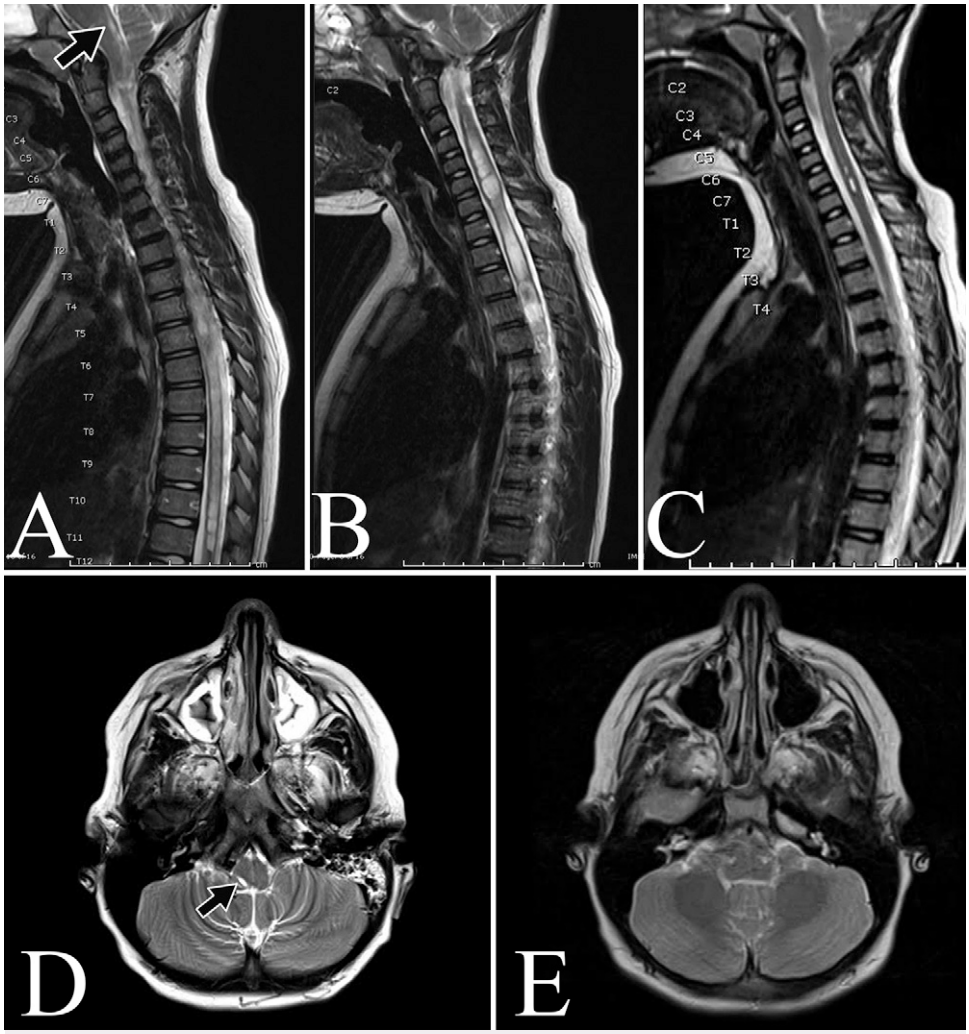

FIG. 1. A: Preoperative sagittal T2-weighted MRI demonstrating the extent of a syrinx (*arrow*) in the brainstem. **B:** Preoperative sagittal T2-weighted MRI. **C:** Two-month postoperative sagittal T2-weighted MRI. **D:** Preoperative axial T2-weighted MRI demonstrating the extent of the syrinx (*arrow*) in the brainstem. **E:** Two-month postoperative axial T2-weighted MRI.

the dates from inception to August 10, 2022, using the following Boolean search term "Chiari" AND "syringobulbia."

Observations

Our search yielded 12 relevant studies reporting 55 cases of CMI presenting with syringobulbia. Fifty-one (93%) of 55 cases involved syringobulbia in conjunction with syringomyelia.[2–13] Headaches and nystagmus were the most common presenting symptoms of CMI involving syringobulbia, with 33% of patients experiencing these symptoms.[2–13] Diplopia (24%), hyperreflexia (24%), ataxia (22%), and dysphagia (22%) were also common symptoms in this cohort.[2–13] Dysarthria (9%), dysesthesia (9%), and dyspnea (4%) were less commonly observed in these patients[2–13] (Table 1).

In total, 24 patients (44%) exhibited CN deficits. CN V, VI, VII, IX, X, and XII palsies were all reported.[3,4,6,9] CN IX and X palsies were reported in 15 (27%) of the 55 patients.[3,4,6,9] CN VI palsy was reported in 10 patients (18%), and CN V, VII, and XII were reported in 3 patients.[3,4,6,9] All CN VII (3, 5%) palsies were bilateral.[6] Altogether, more than 20 distinct symptoms were reported among the 55 patients in the literature review[2–13] (Table 2).

The actual causes of syringomyelia and syringobulbia, although attributed to fluid collection within the spinal cord and brainstem, respectively, are likely the result of a multitude of factors. The first theory that attempted to explain the cause of this fluid buildup was presented in 1958. This explanation proposed that a syrinx could be caused by the flow of CSF from the fourth ventricle being blocked, which would push the CSF through a patent central canal.[14] The second theory, proposed in 1969, stated that a block in caudal CSF flow causes increased intracranial pressure, forcing CSF from the fourth ventricle into the central canal.[14] Additionally, the third theory, termed the piston theory, presented the idea that a syrinx was caused by the cerebellar tonsils creating large pressure waves in the spinal subarachnoid space, forcing fluid through the surface of the spinal cord into the central canal.[14] Although many hypotheses have been produced, there is still no definitive answer regarding any single cause of syringomyelia and syringobulbia; each presentation is likely the result of one or more of the above pathophysiological processes.

TABLE 1. Literature summary of signs and symptoms associated with CMI and syringobulbia

Symptoms	Times Occurred	Frequency
Headache	18	33%
Nystagmus	18	33%
Diplopia	13	24%
Hyperreflexia	13	24%
Ataxia/gait disturbances	12	22%
Dysphagia	12	22%
Weakness in extremities	12	22%
Pain in extremities	10	18%
Paresis	10	18%
Numbness	9	16%
Horner's syndrome	7	13%
Dysarthria	5	9%
Dysesthesia	5	9%
Ptosis	5	9%
Neck pain	4	7%
Spasticity	3	5%
Muscle tone abnormalities	3	5%
Dyspnea	2	4%
Tetraparesis	2	4%
Vomiting	2	4%

TABLE 2. Literature summary of type and frequency of CN palsies

CN Palsies, All Bilat	Times Occurring	Frequency
CN V	3	5%
CN VI	10	18%
CN VII	3	5%
CN IX	15	27%
CN X	15	27%
CN XII	3	5%

At present, there are two prominent treatments for CMI: posterior fossa decompression with versus without duraplasty.[15] A posterior fossa decompression is performed to relieve elevated intracranial pressure in CMI by removing the bottom portion of the occipital bone but leaving the dura intact. A bony decompression with duraplasty is done the same as a bony decompression, except that the dura is opened to allow for better decompression. For cases refractory to posterior fossa decompression, a subsequent treatment option is to shunt the syrinx, thus providing a path for CSF to return into the subarachnoid space[15]; however, syringo-subarachnoid shunt (SSS) placement has some clinical risks. These risks include a secondary spinal cord injury from the posterior midline myelotomy. Additionally, the SSS catheters can obstruct and fail due to an inflammatory response and adhesion.[16]

Although the literature features one patient who presented with syringomyelia, syringobulbia, and left gaze paralysis,[14] our case is unique given the presentation of both syringomyelia and syringobulbia in conjunction with unilateral CN VII deficit. As previously stated, CN VII palsy is rarely observed (5%)[6] in patients with syringobulbia because of the location of the CN VII nucleus in the brainstem. It is not common for the syrinx to extend to the location of the CN VII nucleus within the pons; in fact, syringobulbia usually arises from upward extension of syringomyelia. Therefore, the cranial nerves closest to the craniocervical junction, such as CN IX and X, are most likely to be affected. Because of the location of the nucleus, syringobulbia must extend further into the brainstem to cause dysfunction in CN VII. Due to the pathophysiology of syringobulbia, the fluid cavity typically forms in the central canal, causing bilateral CN palsies. In the case presented here, the syringobulbia in our patient was found to arc unilaterally, which caused the CN VII palsy to present with right-sided facial hemiparesis.

Lessons

To our knowledge, we report the first pediatric case of a rare presentation of unilateral CN VII palsy secondary to syringobulbia in the setting of CMI. Treatment of the underlying craniocervical compression resulted in the successful resolution of the patient's most severe deficits. The differential for pediatric cranial neuropathies should remain broad with workup including MRI for thorough evaluation to guide surgical treatment.

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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: Olaya, Sherlock, Chan. Acquisition of data: Sherlock, Brown, Campos. Analysis and interpretation of data: Olaya, Sherlock, Chan, Campos. Drafting the article: Sherlock, Brown, Chan, Campos. Critically revising the article: Olaya, Brown, Campos. Reviewed submitted version of manuscript: Olaya, Brown, Campos. Approved the final version of the manuscript on behalf of all authors: Olaya. Study supervision: Olaya.

Supplemental Information

Previous Presentations

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