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A single-center experience with eccentric syringomyelia found with pediatric Chiari I malformation

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Abstract
Introduction Eccentric syringes associated with Chiari I malformation have received scant attention in the medical literature. Herein, we describe our experience and long-term outcome in patients with this finding.
Materials and methods A retrospective analysis of a Chiari I database was performed. Patients known to have an associated syringomyelia were then further analyzed for the type of syrinx present. When an eccentric syrinx was noted, the symptoms and postoperative course of these patients were analyzed.
Results Of well over 500 operative cases of Chiari I malformation, roughly 70 % (pre-syrinx and minimally dilated central canals were excluded) were found to have an associated syringomyelia. Of these, four patients were found to have an eccentrically positioned syrinx. Three of these cases presented with symptoms referable to the side of the eccentric syrinx. Postoperatively, cases with both a central and eccentrically located syrinx were found to have a greater decrease in the size of the central portion of their syrinx compared to the eccentrically located portion. Symptoms decreased in all patients.
Conclusions The minority of our patients with hindbrain-induced syringomyelia were found to have an eccentrically located syrinx. Of these, most will have symptoms localized to the abnormal fluid-filled cavity, and these may not decrease in size as much as centrally located syringes following posterior fossa decompression. However, all symptoms decreased in those operated. Based on the literature, non-hindbrain-induced syringomyelia is more likely to result in an eccentrically placed syrinx. The mechanism for this is yet to be elucidated.

Keywords Pediatrics · Children · Tonsillar ectopia · Syringomyelia · Hydrosyringomyelia · Chiari I malformation · Eccentric

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Introduction

Syringomyelia is a fluid-filled cystic dilation within the spinal cord. Milhorat [18] classified syringomyelia into three subgroups. Communicating syringomyelia was described as a dilation of the central canal that communicates with the fourth ventricle and non-communicating syringomyelia as a dilation without connection to the fourth ventricle. A third category of syringomyelia also exists in which the dilation in the spinal cord does not communicate with the central canal or the fourth ventricle, the so-called eccentric syringomyelia. Eccentric syringomyelia, also known as extracanalicular non-communicating syringomyelia or primary parenchymal cavitation [18], refers to a fluid-filled cystic dilation that originates in the gray and/or white matter of the spinal cord parenchyma [18–21, 26–28] and contains either cerebrospinal fluid (CSF) or proteinaceous fluid [19, 23, 31].

As these so-called eccenteric syringes associated with the Chiari I malformation have received scant attention in the medical literature, we review our experience and long-term outcome in patients with this finding.

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Materials and methods

A retrospective analysis of our hospital's (Children's Hospital, Birmingham, AL, USA) Chiari I database for the two senior authors was performed. Patients known to have an associated syringomyelia were then further analyzed for the type of syrinx present. When an eccentric syrinx was noted, the symptoms and postoperative course of these patients were analyzed by a review of their records and imaging.

Results

Of well over 500 operative cases of Chiari I malformation over a 20-year period, roughly 70 % (pre-syrinx and minimally dilated central canals were excluded) were found to have an associated syringomyelia. Of these, four patients (approximately 0.7 %, three females and one male aged 5–15 years, mean=11 years) were found to have an eccentrically positioned syrinx (Figs. 1 and 2). All cases presented with symptoms neurologically referable to the side of the eccentric syrinx. Symptoms included upper limb pain and parasthesias brought on with Valsalva maneuvers in all three patients. This occurred in the shoulder, elbow, and hand in one and in the arm and hand in the others. Each of the four cases had preoperative and postoperative imaging noting the status of their syringomyelia. All but one case underwent posterior fossa decompression with duraplasty. One female patient presented with left-sided weakness specifically in her serratus anterior muscle. She was unable to raise her arm above her head with scapular rotation and had lost the ability to protract the scapula. Her family chose not to have surgery, and her weakness persists at 10 years of follow-up. Postoperatively, cases with both a central and eccentrically located syrinx were found to have a greater decrease in the size of the central portion of their syrinx compared to the eccentrically located portion. Symptoms decreased in all patients. Cranially, these patients were found to have typical Chiari I malformations with tonsillar descent ranging from 9 to 22 mm below the foramen magnum. All syringes were cervicothoracic in location. No patient had a history of meningitis, hydrocephalus, or other spinal pathology including trauma to the back. One patient had a 12° levoscoliosis that has not progressed at long-term follow-up. The average follow-up for this small cohort of patients ranged from 1 to 10 years (mean=1.9 years). No change in the degree of tonsillar ectopia was noted at long-term follow-up imaging.

Discussion

We found a very small number of patients out of a large group of patients with hindbrain-induced syringomyelia to have an eccentric component to their syrinx. Interestingly, all of them presented with symptoms localized to the side of their syrinx. Also, most did not have the same degree of response for the eccentric component of their syrinx

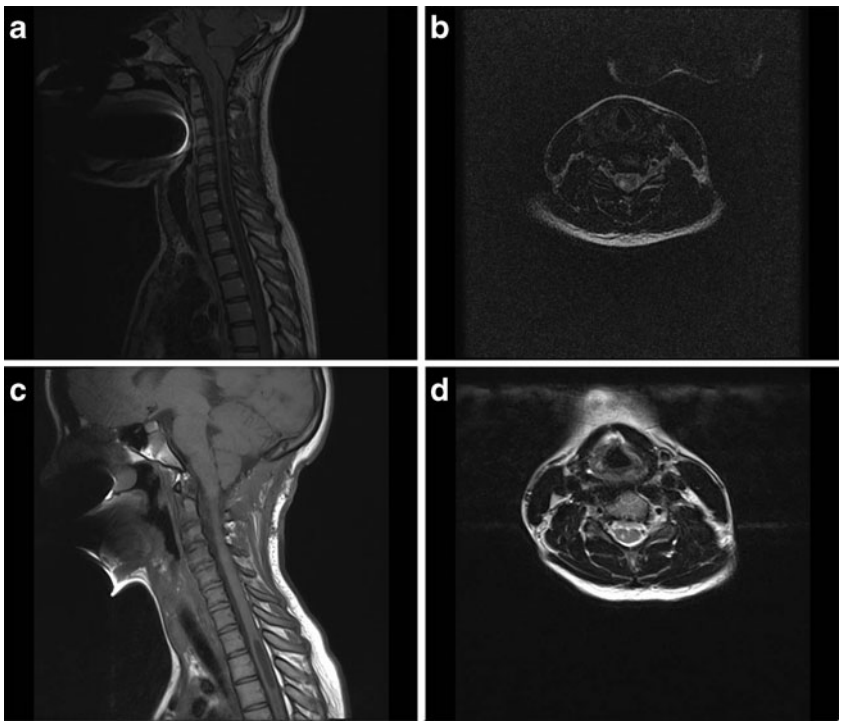

Fig. 1 Example of one patient found to have eccentric syringomyelia due to Chiari I malformation. Preoperative sagittal (**a**) and axial (**b**) images and postoperative sagittal (**c**) and axial (**d**) images. Note a significant decrease in syrinx size following surgery

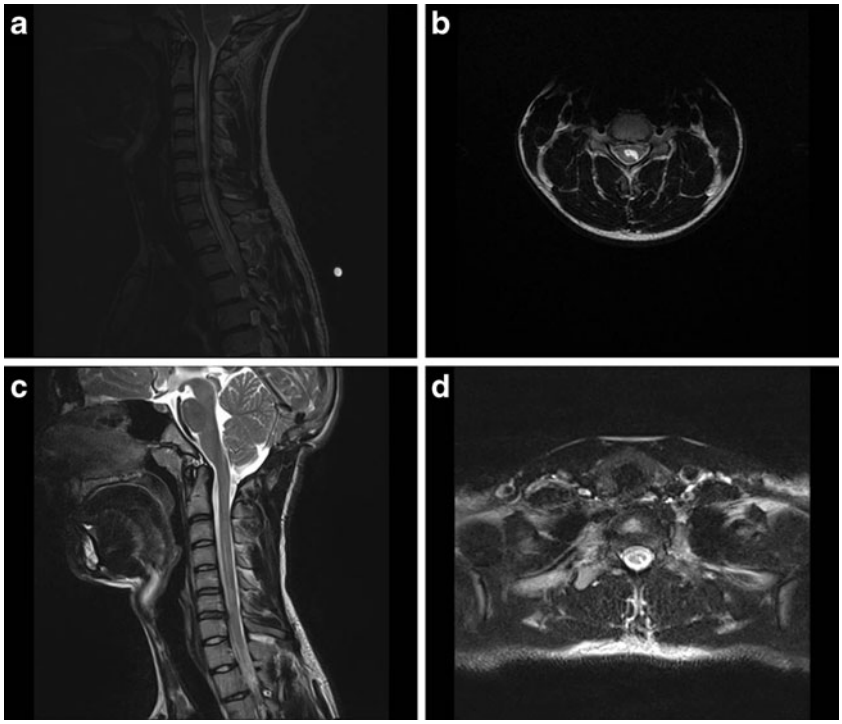

Fig. 2 An additional example of a patient in our series found to have eccentric syringomyelia due to Chiari I malformation. Preoperative sagittal (**a**) and axial (**b**) images and postoperative sagittal (**c**) and axial (**d**) images. This patient was found to have a significant decrease in the size of their centrally located component of the syrinx, but less decrease in the eccentrically located part

compared to the centrally located part of the syrinx. One theory for this may be that overdistension of the ependymally lined central portion of the syrinx may have a greater ability to return to a baseline size compared with the intraparenchymal portion (eccentric) of the syrinx. However, symptoms resolved in all operated patients.

Etiology

Etiologies of syringomyelia include conditions that damage the spinal cord and may or may not alter the normal CSF flow and pressure within and around the spinal cord [18–20]. Milhorat et al. [19] reported on 105 individuals with syringomyelia who underwent spinal cord autopsies; 35 (33 %) patients were found to have eccentric syringomyelia. The most common cause was trauma (13, 37 %) followed by ischemic infarction (11, 31 %), post-meningitic infarction (6, 18 %), spontaneous intramedullary hemorrhage (2, 5.5 %), transverse myelitis (2, 5.5 %), and radiation necrosis (1, 3 %). Notably, 45 out of 100 (45 %) intramedullary masses reported in a series were associated with syringomyelia [23]; 49 % of the syringes were above, 11 % below, and 40 % both above and below the tumor. In order of frequency, ependymomas, hemangioblastomas, cavernomas, and, less frequently, astrocytomas were the most common medullary tumors associated with syringomyelia. Some have advocated that tumor-associated syringes represent a distinct entity as it contains a portentous fluid rather than CSF and, occasionally, the spinal cord cavity is lined by tumor cells rather than glial/ependymal cells [18]. Our group of patients with hindbrain-induced eccentric syringomyelia made up <1 % of cases of syringomyelia in our institutional experience.

Anatomy

The location, extension, and diameter of the eccentric syringomyelia depend on its etiology. In a study by Sherman et al. [24], MRI was performed on 58 patients with syringomyelia due to trauma (16 patients, 28 %), tumor (9 patients, 16 %), Chiari malformation (24 patients, 41 %), and unknown causes (9 patients, 15 %). The post-traumatic syringomyelia mainly involved the cervicothoracic area, ranging from one vertebral segment to an entire spinal cord involvement (eight to nine vertebral segments). The diameter varied from 3 to 15 mm. Tumor-associated syringes also involved the cervicothoracic area and ranged from one vertebral segment to holocord syringomyelia. The diameter varied from 2 to 12 mm.

Histology

Eccentric syringomyelia is frequently associated with segmental (at the level of syrinx) or full-length myelomalacia without any evidence of whether this atrophy is the cause or the effect of the syrinx formation [19]. It involves primarily the central gray matter, dorsal and lateral to the central

canal, between the anterior and posterior spinal arteries (watershed zone), which might suggest vascular insufficiency [18, 19]. Large cavitations extend from the ventral or dorsal horns into the adjacent white matter and mainly along the posterior funiculus. Gray matter involvement is associated with central chromatolysis, neuronophagia, and gliosis. White matter involvement is associated with variable patterns of demyelination associated with gliosis and infiltration of foamy fat-laden macrophages [19, 21]. The interior of the syrinx is covered by glial or fibroglial cells containing large amounts of reticulin. Hemosiderin-laden macrophages and microglial cells are a result of trauma or hemorrhage [19, 21].

Pathogenesis

The pathogenesis of eccentric syringomyelia is still unknown. Since the first treatment attempt of syringomyelia by Brunner [7] in 1700, a wide variety of theories have been put forth. In 1867, Hallopeau [11] described the cause of syringomyelia as an inflammatory process that affects the ependyma and leads to sclerotic changes that obstruct the central canal. In the mid- to the late nineteenth century, Charcot and Joffroy [9] and Jofrroy and Achard [13] suggested the role of ischemia secondary to venous and arterial occlusion from inflammatory processes such as meningitis, which was later corroborated by experimental evidence [8, 12, 17, 29]. However, other experiments were able to produce syringomyelia without ischemic changes [3], or arterial [30] or venous occlusion [16] without inflammation.

In an attempt to explain the source of the fluid in the cysts, most of the early theories were based on the assumption that there is always communication between the syrinx and the fourth ventricle. However, recent studies have demonstrated that the majority of spinal cord syringes do not communicate with the fourth ventricle [18, 19]. In 1950, Brierley [5] demonstrated the movement of CSF tracers from the subarachnoid space into the central canal through the perivascular space for the first time. In 1972, Ball and Dayan [2] injected water-soluble contrast media into the subarachnoid space, which accumulated in the syrinx, and with further studies, they found that it entered the spinal cord by a transparenchymal flow rather than the fourth ventricle. In 1990, Rennels et al. [22], in an animal model, also described the flow of horseradish peroxidase-labeled CSF from the subarachnoid space through the perivascular space and interstitium. Others attributed this pathway to the lymphatic function of the spinal cord [19–21]. Following the injection of kaolin or quisqualic acid (QA) into the subarachnoid space in an animal model, Stoodley et al. [26–28] were able to produce spinal cysts, which did not communicate with the central canal or the fourth ventricle. QA injection produced focal neurological injury and cell death, which resulted in small cyst formation at the site of injection. Kaolin injection caused arachnoiditis and adhesions in the subarachnoid space. Arachnoiditis alone was not sufficient for cyst formation, while the combination of cell injury and inflammation resulted in an enhanced syrinx formation. The role of arachnoiditis was explained on the basis that it reduced the compliance of the subarachnoid space and increases the perivascular flow of CSF. As the CSF spreads through the spinal parenchyma before reaching the central canal, it results in extracanalicular cyst enlargement. CSF flow was driven by arterial pulsation, and thus, reduction in spinal pulse pressure by partial ligation of the bracheocephalic artery, with minimal alteration of the mean pressure, decreased the perivascular flow and cyst enlargement.

In 1994, Cho et al. [10], based on animal models of 38 rabbits, described the role of arachnoiditis in trauma-induced syrinx enlargement. These authors demonstrated that the trauma can induce cavity formation by hematoma, ischemia, or even the trauma itself and interrupt normal CSF flow, while the expansion of the syrinx is related to subarachnoid adhesions due to arachnoiditis associated with the trauma, and in the same mechanism described above [6, 10].

To explain syrinx progression and expansion, Williams [31] demonstrated the Slosh theory in 1980 in which increasing pressure around the spinal cord (e.g., epidural venous pressure) associated with cough, Valsalva maneuver, or even heartbeats, can be transmitted through and compress the wall of the syrinx, producing two types of fluid movements, upward and downward, leading to syrinx expansion.

In 1875, Simon [25] reported an association between syringomyelia and spinal cord tumor. Many theories were proclaimed since then to explain the role of intramedullary tumor in syrinx formation and expansion. One of the most popular theories is the transudation hypothesis. These stated that intramedullary tumors can compress and displace the spinal cord parenchyma, which in turn can lead to syrinx formation. Transudation from tumor blood vessels and secretion by tumor cells, with impairment of the normal CSF flow from the subarachnoid space into the central canal, can contribute to syrinx enlargement [14, 23, 31].

Clinical presentation

In addition to the initial injury that leads to syrinx formation, expansion of the eccentric syringomyelia causes direct compression on and damage to the surrounding spinal cord, and associated neurological deficits depend on its location and the nuclei and tracts affected. In a study by Milhorat et al. [19] involving autopsies of 105 patients with syringomyelia, 35 (33 %) patients were found to have eccentric syringomyelia. For these patients (the eccentric syringes were not culled out), the findings are the following: numbness in 25

(71 %), paraesthesias or dysesthesias in 16 (46 %), paraparesis in 14 (40 %), weakness or paralysis in the upper extremities of 8 (23 %), muscular atrophies in 8 (23 %), Brown–Sequard syndrome in 6 (17 %), dissociated sensory loss in 4 (11 %), impaired position sense in 4 (11 %), cranial nerve palsies in 4 (11 %), and quadriparesis in 3 (8.5 %). Pain is the most common presentation in post-traumatic syringomyelia. It can be localized or diffused, and it commonly presents as dull, aching, or burning pain. Other neurological symptoms can include any of the above, and its appearance varies from a few months to many years after the trauma [1, 4, 10, 15]. We too observed ipsilateral symptoms in our group with eccentrically positioned syringes.

Conclusions

We reviewed our clinical experience with eccentric syringes as found in the pediatric Chiari I malformation.

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