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CASE REPORT

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Surgical observation of the agenesis of the foramen of Magendie

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We report a young girl who presented with headache and back pain. Dynamic MRI revealed no cerebrospinal egress from the median aperture (Foramen of Magendie) of the fourth ventricle and syringomyelia. A posterior cranial fossa exploration was performed and agenesis of the median aperture was observed. Following surgical penetration of the posterior aspect of the fourth ventricle and at the most recent follow-up examination, this patient's syringomyelia had resolved, as had her symptoms. Agenesis of the foramen of Magendie may be a rare cause of inhibition of normal cerebrospinal egress from the fourth ventricle with resultant syringomyelia.

Key words: posterior cranial fossa, median aperture, cerebrospinal fluid, ventricle, anomaly

INTRODUCTION

Spinal cord cavitation (syringomyelia) may be secondary to processes such as multiple sclerosis, tumours, trauma, and infection [9, 10]. In each of these cases the syrinx fluid is dissimilar to the surrounding cerebrospinal fluid. Posterior cranial fossa pathology, arachnoid cysts and hindbrain herniation (Chiari malformations) for example, has been related to the development of syringomyelia. In the United States syringomyelia is most frequently found in the setting of congenital hindbrain herniation. Gardner [5] pursued this association of spinal cord cavitation and herniation of the posterior cranial fossa contents and developed his hydrodynamic theory. This theory removes the aetiology of syrinx formation from the spinal cord and places the pathological change at the craniocervical junction. The theory states that the pulsatile flow of cerebrospinal fluid is transmitted inferiorly through the central canal via a "water hammer" effect. This central canal flow is postulated to result from the blockage of normal cerebrospinal fluid egress from the fourth ventricle. The theory relies upon a persistent communication between the syrinx and the fourth ventricle. Embryologically, this requires a delayed or inadequate rupture of the rhombic roof and lack of communication of the fourth ventricle with the subarachnoid space. The main problem in utilising this theory is the inability to find consistently a communication with the syrinx in either clinical or pathological material. The hydrodynamic theory does not adequately address the formation of syringes in cases of spinal trauma or tumours.

Williams [12–15] has developed an adaptation of the Gardner theory that suggests that syrinx formation is the result of differential pressure between the intracranial and intraspinal compartments. This theory may explain focal syrinx formation in the presence of other spinal lesions. With Valsalva manoeuvres, intraspinal subarachnoid pressure rises due to increased venous pressure. This rise in spinal subarachnoid pressure is thus communicated to the intracranial space, which normally occurs with little impedance to flow. As the intraspinal pressure falls, cerebrospinal fluid again flows caudally back into the spinal canal. If this pathway is blocked by a process

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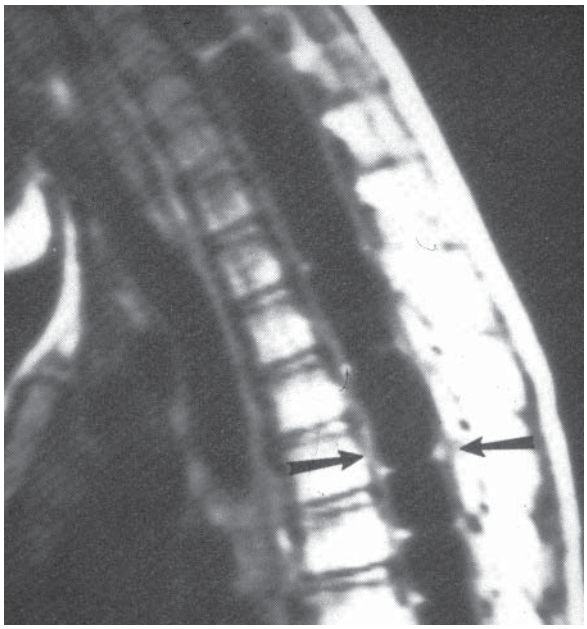

Figure 1. T1-weighted sagittal MRI noting the large syrinx found in our patient (between arrows).

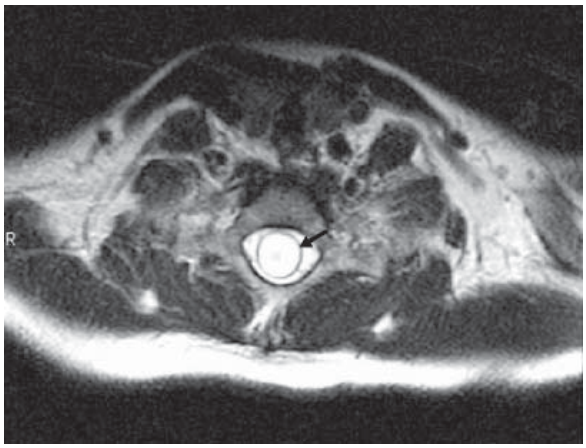

Figure 2. T2-weighted axial MRI of the large syrinx seen in Figure 1. The arrow is on the thin circular rim of the spinal cord with the interior of this structure representing the syrinx.

at the fourth ventricular outlet, such as tumour or tonsillar herniation (i.e. Chiari I malformation), then a momentary pressure differential between the intraspinal and intracranial compartments may develop. The spinal fluid then escapes the intracranial cavity and dilates the central canal, thereby increasing the block to caudal flow of cerebrospinal fluid and thus furthering development of the syrinx. Any process that blocks the normal to-and-fro motion of fluid between the intracranial and spinal subarachnoid spaces may act to force cerebrospinal fluid into the spinal cord, when there is a momentary vector of force out of the intracranial compartment. The resolution of this pressure differential may be the movement of tissue through the foramen magnum or the movement of cerebrospinal fluid into the spinal cord. Those patients with a constant discrepancy between the intracranial and intraspinal compartments rather than momentary dysequilibrium will have the most severe symptomatology.

CASE REPORT

We report a 12-year-old girl with the presentation of headache and back pain. Subsequent imaging of the spine revealed syringomyelia (Fig. 1, 2). No hydrocephalus was identified and there was mild (<15 degrees) of scoliosis. Dynamic MRI revealed no cerebrospinal egress from the median aperture (Foramen of Magendie) of the fourth ventricle. A posterior cranial fossa exploration was performed and agenesis of the median aperture was observed (Fig. 3). Following surgical penetration of the posterior aspect of the fourth ventricle (i.e. through the imperforate Foramen of Magendie) (Fig. 4) and at the most recent follow-up examination, this patient's syringomyelia had substantially improved, as had her symptoms. There was no history of meningitis, head trauma, or past medical or surgical history in this child. She had no signs of spinal dysraphism.

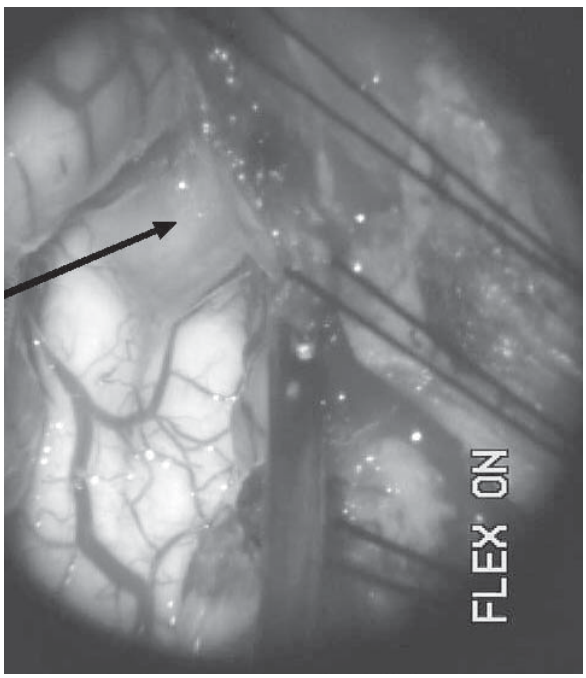

Figure 3. Intraoperative view of the atretic foramen of Magendie (arrow). The forceps have spread apart the left and right cerebellar tonsil. Note the posterior spinal artery and retention sutures along the left edge of the dura mater.

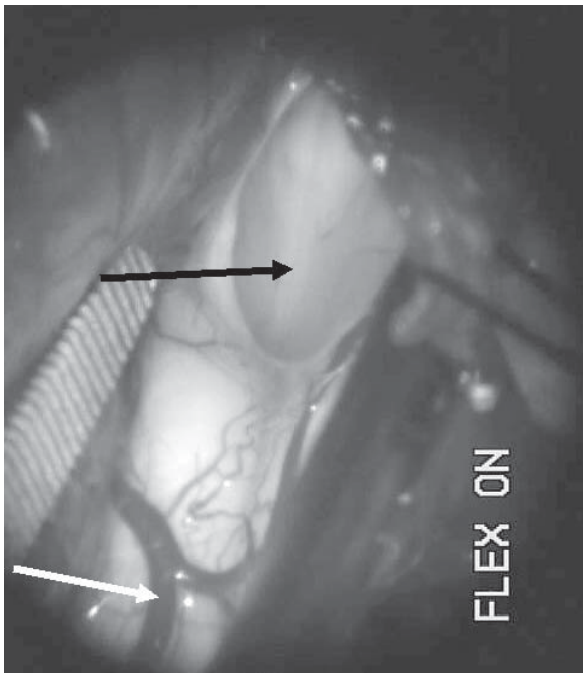

Figure 4. Following the penetration of the atretic foramen of Magendie. Note the avascular floor of the fourth ventricle (arrow). Again note the posterior spinal artery (arrow) for reference.

DISCUSSION

The foramen of Magendie forms between 12 and 20 weeks of gestation and is present not only in humans but also in canine, avian and ruminant species and in rodents. The foramen is normally formed by an evagination of the tela into the pia-arachnoid, which starts in man when the foetus is about 100 mm in length [4]. Gibson [6] found that this aperture is absent in the cat, rabbit, and goat. Interestingly, atresia of one or two cerebellar foramina is often detected in normal brains without hydrocephalus [2]. We would therefore assume that our patient had marginal egress from the fourth ventricle, but that over time this output of cerebrospinal fluid was insufficient and thus resulted in her syringomyelia. Hashish et al. [7] have stated that only eight cases of membranous obstruction of the foramen of Magendie exist in the literature. The authors further state that congenital imperforation of the foramen of Magendie must be considered as a possible aetiology of chronic hydrocephalus. One entity that may cloud observers' descriptions is a membrane that occludes the median fourth ventricular outlet but does not represent true atresia of this aperture [8].

MRI data has supported the Williams theory of syrinx formation [13, 14]. Using dynamic flow measurements, preoperative patients with hindbrain herniation demonstrated elevated systolic and diastolic pressure waves within the syrinx. These numbers tend to be higher in patients with larger syringes. After surgical decompression pressure in the subarachnoid space increases and pressure in the syrinx diminishes. From this study it would appear that diastolic pressures both pre- and postoperatively correlate with clinical outcome [1]. With each systole increased downward pressure forces the hindbrain inferiorly, thus creating a "piston" that increases the pressure further [1]. This would tend to support the hydrodynamic theory.

Syringes can be induced experimentally in dogs by the intra-cisternal injection of kaolin. This model has revealed more severe destruction of anterior horn cells in the spinal cord in the animals that develop a syrinx [3]. Syrinx cavities often do not communicate with the central canal in this model, supporting Williams' craniospinal dissociation theory [15]. It is likely that a combination of Gardner's hydrodynamic theory and Williams' craniospinal pressure differential theory currently explains the pathogenesis of syringomyelia. Whether delayed perforation of the rhombic roof is responsible for the initial development of syringomyelia or not will require more investigation. Certainly, there can be components of both theories in the development of syringomyelia, and further work will be required to understand the contributions of these two theories to the pathophysiological formation of syringomyelia.

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