Microsurgery of Arnold-Chiari malformation in adults with and without hydromyelia

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Microsurgical exploration of 15 adults with Arnold-Chiari malformation with and without hydromyelia using 3 to 20 × magnification has led to the following conclusions. Hydromyelia, associated with Arnold-Chiari malformation, is a progressive mechanical disorder that causes spinal cord deficits by pressure distention of the cord. Arnold-Chiari malformation causes slowly or suddenly progressive bulbar dysfunction by impaction of the malformation in the foramen magnum. Decompression of both can be achieved by a suboccipital craniectomy, upper cervical laminectomy, establishing an outlet from the fourth ventricle, and opening the distended cord in the thinnest exposed area, which is usually along the dorsal root entry zone. If Pantoque myelography in patients in the supine position shows the Arnold-Chiari malformation, hydromyelia can be established as a cause of central cord deficit even if myelography shows the cord size to be normal. Syringomyelia, traditionally considered a degenerative disease, is a less common cause of a slowly progressive central cord deficit than either hydromyelia or intramedullary tumor.

KEY WORDS • Arnold-Chiari malformation • hydromyelia • syringomyelia • microneurosurgery • spinal cord

HYDROMYELIA, a condition in which the central canal of the spinal cord communicates with the fourth ventricle and is distended by cerebrospinal fluid (CSF), often accompanies Arnold-Chiari malformation in the adult. The first manifestation is often a loss of pain sensation in the cervical dermatomes followed by atrophy and weakness caused by extension of the cavity into the anterior horns. The painless, slowly progressive cord deficits suggest a diagnosis of syringomyelia that has traditionally been regarded as an untreatable degenerative process. The location of the lesion has led to a constellation of neurological findings characteristic of a central cord syndrome, and the association of these deficits with syringomyelia has led to the designation of syringomyelic cord syndrome.

Careful radiological evaluation of the early patients in this series led the author to conclude that Gardner's mechanical concept of a pressure distention origin of syringomyelia cord syndrome is correct. This concept provided the rationale for the treatment outlined in this paper. Most of these patients had a previous diagnosis of untreatable degenerative disease and severe deficits had developed in some before treatment.

Clinical Material and Methods

Clinical Material

Fifteen adults operated on by the author for Arnold-Chiari malformation during the last 9 years provided the basis for this review. The patients ranged in age from 19 to 64 years. In one patient the onset of symptoms
occurred after the age of 60. Eleven patients had an Arnold-Chiari malformation with hydromyelia and four had Arnold-Chiari malformation only. All patients with hydromyelia had signs and symptoms referable to it; usually a typical central cord syndrome began with sensory loss around the shoulders extending to one or both upper extremities, and a lower motor neuron deficit initially appeared in the hands. Sensory loss usually preceded lower motor neuron signs. These deficits ranged in severity from a minimal subjective sensory loss to a widespread sensory loss with quadriplegia and marked atrophy of all extremities. Two patients with longstanding quadriplegia were admitted suffering respiratory difficulty because of loss of lower motor neurons supplying the respiratory muscles.

Four patients were followed without surgery after diagnosis because their deficits were minimal. The deficit increased in all four and all had surgery within 1 year of the initial diagnosis.

The four patients with Arnold-Chiari malformation without hydromyelia presented with a combination of progressive bulbar and cerebellar signs; in most cases there was a combination of upper and lower motor neuron signs in the bulbar musculature, gait difficulties due to cerebellar and long-tract involvement, and some nystagmus. Four of the 11 patients with hydromyelia also had bulbar signs and symptoms. Seven patients with Arnold-Chiari malformation and hydromyelia had symptoms referable only to the hydromyelia.

A 24-year-old man with Arnold-Chiari malformation without hydromyelia was admitted with severe respiratory stridor caused by bilateral vocal cord paralysis of sudden onset. A previous diagnosis of bulbar degenerative disease had been made. Bertrand describes the mechanism for sudden onset or change of symptoms with this condition.

No patients in this series of adults had hydrocephalus or evidence of increased intracranial pressure (ICP) even though defective ventricular drainage is thought to be important in development of the hydromyelia.

Radiological Findings

All patients had Pantopaque myelograms performed through the foramen magnum in the supine and prone position (Figs. 1 and 2). Those patients with central cord deficits were assumed to have hydromyelia, even if the cord size was normal or only minimally enlarged, if the spinal cord neurological deficits were typical and the Pantopaque myelogram done in the supine position showed the Arnold-Chiari malformation. Myelography done in the prone position demonstrated the Arnold-Chiari malformation in all cases, yet the prone study was frequently normal. Most of these patients had had a myelogram done previously only in the prone position, which was normal in each case.

The radiological diagnosis was somewhat more difficult to obtain if the patient had Arnold-Chiari malformation but not hydromyelia. In these patients, the diagnosis was often not suspected before contrast studies. These adults, because of the brain-stem findings, often underwent posterior fossa angiography or pneumoencephalography initially. The diagnosis of Arnold-Chiari malformation was suspected in several patients after angiography because of the low descent of the caudal loop of the posterior inferior cerebellar artery (PICA) (Fig. 3 upper left). After angiography, pneumoencephalography was often done rather than ventriculography because there was no evidence of increased ICP. The ventricles frequently did not fill at pneumoencephalography, and subsequently a ventriculogram was done that showed typical caudal displacement of the fourth ventricle (Fig. 1 center right). The pneumoencephalogram showing nonfilling of the ventricles often revealed the tonsillar herniation. No patient had significant hydrocephalus even though dense scar tissue over the foramen of Magendie frequently prevented filling of the fourth ventricle at pneumoencephalography and supine Pantopaque myelography.

Operative Findings

All patients had a suboccipital craniectomy and upper cervical laminectomy. All dissection after bone removal was done using the surgical microscope at 3 to 20 × magnification (Figs. 3, 4, and 5).

The cerebellar tonsils and fourth ventricle were displaced caudally into the foramen magnum or upper cervical area. The caudal part of the fourth ventricle often extended into the upper cervical area and the caudal loop of the PICA often descended to the level
Arnold-Chiari malformation and hydromyelia

of C-2, marking the lower margin of the cerebellar tonsils (Fig. 3 upper left).

The elongated cerebellar tonsils varied in appearance from those normal in color and consistency to those which were white and firm because of scarring and gliosis (Fig. 3 upper right). In the cases in which some Pantopaque passed into the area between the cerebellar tonsils and the dura, the cerebellar tonsils were near normal in consistency and covered with only mild arachnoidal adhesions. If no Pantopaque passed into the

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Fig. 1. Myelography and pneumoencephalography in Arnold-Chiari malformation. *Upper Left:* Normal lateral view of Pantopaque study with the patient in a supine position. Pantopaque fills the cisterna magna and fourth ventricle to the level of the aqueduct. *Upper Right:* Pantopaque myelogram done in the supine position outlines the tonsillar herniation with Arnold-Chiari malformation. *Lower Left:* Pantopaque study of the foramen magnum in the supine position showing an Arnold-Chiari malformation. No Pantopaque passes into the cisterna magna above the level of foramen magnum. At surgery, dense adhesions in the area of the cisterna magna bound the medulla and cerebellum to the dura. *Lower Right:* Lateral view of ventriculogram showing blunting of the lower margin of the caudally displaced fourth ventricle with Arnold-Chiari malformation.
FIG. 2. The upper pair are from one patient and the lower pair from another. In both patients previous myelograms to define the cause of progressive central cord deficits were reported to be normal. The myelograms (upper right and lower right) show no striking abnormalities in the cervical regions. A diagnosis of degenerative disease had been made because cord size appeared normal on previous myelograms. Lateral view of the Pantopaque study of the foramen magnum in the supine position in both patients (upper left and lower left) shows the obstruction to filling of the cisterna magna. The findings from supine Pantopaque myelography led to a diagnosis of Arnold-Chiari malformation with hydromyelia.
Arnold-Chiari malformation and hydromyelia

Fig. 3. Posterior views of Arnold-Chiari malformations through suboccipital craniectomy. The pictures were taken through surgical microscope at 6 × magnification and retouched and labeled to increase clarity. Only upper right and center left are from the same patient. Upper Left: Caudal loop of the posterior inferior cerebellar artery (PICA) displaced around caudal margin of herniated cerebellar tonsil. The lower margin of the tonsil was at the C-2 level. Upper Right: Lower margin of herniated cerebellar tonsil. Same patient as shown in Fig. 1 upper right. The pale color at the tip of the left cerebellar tonsil is caused by gliosis in herniated tonsillar tips. Center Left: Posterior view of the fourth ventricle after separation of the cerebellar tonsils shown in upper right. A small membrane at the lower margin of the fourth ventricle blocks the view of the upper end of the central canal. This membrane would retain a small muscle plug if one were placed in the central canal. Center Right: This patient did not have a hydromyelia because the tonsils did not extend far enough caudally to block the central canal. The tonsils were separated to expose the fourth ventricle. The obex lies slightly below the caudal level of the cerebellar tonsils. The elongated, narrow fourth ventricle is seen in the midline. Lower Right: A dense mat of scar tissue that includes the dura blocks the area of the foramen of Magendie and is adherent to surface of the medulla. The caudal loop of the PICA enters the scar.
FIG. 4. Posterior views in a patient with Arnold-Chiari malformation and hydromyelia through a suboccipital craniectomy and Cl-3 laminectomy. Pictures were taken at 6 X magnification through surgical microscopes and retouched for clarity. **Upper Left:** Incision of a dense mat of scar tissue over the foramen of Magendie. **Upper Right:** The opening into the fourth ventricle is completed, and a Silastic tube anchored to the dura below is threaded rostrally. **Center Left:** A needle is introduced into the right half of the spinal cord lateral to the posterior column near the C-3 dorsal root entry zone. Clear fluid was obtained. The cord was so thin in this area that the needle tip could be seen through the bulging surface. **Center Right:** An incision 1 cm in length is made in the dorsal root entry zone with a No. 11 knife blade. The cord was thinner than the dura. **Lower Right:** Silastic wick is attached to the dura above leading downward into the hydromyelic cavity.
Arnold-Chiari malformation and hydromyelia

Fig. 5. Left: View through a suboccipital craniectomy of the membrane occluding the foramen of Magendie. A hydromyelia was also present and is shown (right). The occluding membrane was incised with a No. 11 blade. Right: The cord was incised at C-3 dorsal root entry zone. The covering over the hydromyelic cavity was only as thick as a layer of dura. A Silastic wick was left in both the fourth ventricle and cord.

area between the cerebellar tonsils and the dura, an intense mat of scar which included the dura, arachnoid, cerebellar vermis and tonsils, and dorsal surface of the medulla was found at surgery (Figs. 3 lower right, 4 upper left, and 5 left). Intense scarring over the foramen of Magendie was commonly seen with hydromyelia. Those with less scarring were less likely to have hydromyelia.

Typically, there was a kink in the lower medulla or at just below the foramen magnum. In some cases, the lower medulla extending into the upper cervical region was so malformed that it had an irregular multinodular appearance. There often were multiple small cysts within the malformed area. Biopsies of these cystic areas revealed only gliosis.

In patients having Arnold-Chiari malformation without hydromyelia, the caudal margin of the fourth ventricle, the obex, and the upper end of central canal of the cord lay caudal to the lower margin of the cerebellar tonsils; because of this caudal fourth ventricular extension, the hind-brain hernia did not overlap and occlude the central canal (Fig. 3 center right). The fact that the tonsils did not block the central canal of the cord explains the lack of hydromyelia.

The hydromyelic cords in the upper cervical area varied from those with a normal surface appearance to those having an almost transparent covering over the fluid cavity. The thinnest part of the cord was the dorsal root entry zone between the lateral and dorsal columns. The thickness of the cord covering the cavity at the dorsal root entry zone varied from no thicker than a layer of pia-arachnoid to 4 mm. The posterior columns were relatively preserved in comparison to the remainder of the cord even when cord destruction was far advanced.

Operative Procedures

On the day before surgery, the patients sat for 20 to 30 minutes in the cervical semiflexed position to be used for surgery to make certain that the position would not increase their neurological deficit or cause any respiratory problems, as has been reported with the use of the flexed position in Arnold-Chiari malformation. A suboccipital craniectomy and Cl-3 laminectomy was done through a midline incision, thus widely decompressing the foramen magnum. The surgical microscope was brought into the field at this point. The dura was opened in the midline over the cervical cord and above this the incision was U-shaped with the lower margins of the U encompassing the mat of scar tissue often attached to the upper cord and lower medulla (Figs. 3 lower right, 4 upper left, and 5 left). If a plaque of arachnoid and dura was densely adherent to the dorsal medulla and cord, it was left attached because an attempt to disconnect this densely adherent tissue might injure the dorsal surface of the medulla and cord. The degree of adherence of dura to cord and medulla could usually be predicted from the myelogram. If no Pantopaque passed posterior to the cord and cerebellar tonsils at the foramen magnum, the dura was usually matted to the cord. If Pantopaque passed freely, although slowly, between the arachnoid and cerebellar tonsils, the
meninges were usually easily separated from the tonsils. If the foramen of Magendie was blocked, its lower part was opened in the midline, and care was taken to make certain that the dissection was far enough superior, usually over the lower vermis, to enter the fluid cavity of the fourth ventricle rather than the cord or medulla (Figs. 4 upper, and 5 left). After the fourth ventricle was opened, a Silastic wick was attached to the dura and threaded up into the new midline opening (Fig. 4 upper right). The opening at the outlet of the fourth ventricle should be large enough that it would not close even if no wick was threaded into it. The wick was used to maintain the patency of the outlet rather than as a conduit for drainage. Exploration was then directed to the cervical cord at the lower margin of the exposure. The hydromyelia was drained by incising in the dorsal root entry zone between the posterior and lateral columns because that was usually the thinnest area (Figs. 4 center right, and 5 right). A midline myelotomy in such cases could have led to a posterior column deficit in the leg that was nonexistent before surgery, but a lateral myelotomy near the dorsal root entry zone on the side of the most involved arm usually would not increase the already-existing deficit in that extremity. Before incising the cord, a needle was introduced into the cavity at the thinnest area and fluid collected for cell count and protein determinations (Fig. 4 center left). The spinal fluid protein in hydromyelia is normal. An elevated protein or colored fluid suggests the presence of an intramedullary tumor. A vertical incision at least 1 cm in length was made into the thinnest area along the dorsal root entry zone and a Silastic wick, anchored to the dura above, was threaded downward into the myelotomy (Fig. 4 lower right). Usually the cord was thinnest on the side of the greatest neurological deficit. At this point, CSF was draining from the outlet of the fourth ventricle and the cord was decompressed. The dura was loosely closed with a dural graft, with care taken not to constrict the area.

Results

Only one patient had an increased neurological deficit as a result of surgery, a mild proprioceptive sensory loss in the right thumb, which did not impair the patient in any work requiring moderate dexterity. Otherwise, there were no complications. Most patients noted some subjective improvement in sensory and motor deficit but the neurological signs remained about the same as before surgery. No patient has shown further progression of deficit during follow-up periods ranging from 1 to 6 years except one patient with an Arnold-Chiari malformation without hydromyelia who subsequently developed classical amyotrophic lateral sclerosis. It appeared that the Arnold-Chiari malformation was only incidentally related to the initial bulbar symptomatology of this degenerative disease.

Discussion

Barnett, et al., agree that a hydromyelic cavity is the most likely cause of a spontaneously appearing central cord syndrome, although they refer to the condition not as "hydromyelia," but as a "communicating syringomyelia." The author prefers to refer to the dynamic cord pathology associated with Arnold-Chiari malformation as "hydromyelia," as proposed by Gardner and Angel, rather than as communicating syringomyelia because of the traditional association of the term "syringomyelia" with an untreatable degenerative disease. Merritt's neurology textbook classifies syringomyelia as a degenerative process for which, "there is no satisfactory treatment."

In their excellent monograph, Barnett, et al., divide syringomyelia into a communicating and noncommunicating type. Their noncommunicating type developed as a late sequela of spinal cord trauma, arachnoiditis, or intramedullary tumor. This author has seen 19 cases of spontaneous central cord syndrome in which there was no prior history of spinal trauma, and of these 11 were caused by hydromyelia associated with Arnold-Chiari malformation and eight by intramedullary tumor.

The majority of syringomyelic cord syndromes are caused by treatable conditions, and an active investigational attitude should be adopted so that early cases of this disorder are recognized and treated, because patients beyond a certain stage of disability have little likelihood of useful recovery. A number of patients in this series were initially diagnosed as having untreatable degenerative disease because myelography showed the cord size was normal; in these patients the disease
Arnold-Chiari malformation and hydromyelia

Progressed and they developed crippling deficits before surgery. At surgery, some of these patients’ cords had areas no thicker than the pia arachnoid covering the fluid-filled cavity. In four patients the correct diagnosis was made, but surgery was deferred because the central cord deficit was mild; in all the disease progressed and required surgery within 1 year of diagnosis.

Spinal cord size on Pantopaque myelography may be normal even in the presence of hydromyelia causing a marked cord deficit. The diagnosis can be made in spite of a normal cord size by myelography in the supine position. The combination of abnormal findings on neurological examination of a central cord syndrome and tonsillar herniation on Pantopaque myelography establishes the diagnosis of Arnold-Chiari malformation with hydromyelia.

Hydromyelia usually produces a much less striking myelographic picture of intramedullary cord enlargement than does an intramedullary tumor (Fig. 6). Intramedullary tumor even when diagnosed early usually causes striking cord enlargement and frequently is associated with an almost complete block. The enlargement with hydromyelia is diffuse over many segments; that occurring with a tumor is more localized.

The author recommends that Arnold-Chiari malformation and hydromyelia be treated with suboccipital craniectomy and upper cervical laminectomy to decompress the malformation at the foramen magnum. In addition, the outlets of the fourth ventricle...
FIG. 7. Positive contrast ventriculograms from an infant previously treated with a shunt for hydrocephalus. Left: Lateral ventriculogram shows normal-sized ventricles. Right: Contrast medium enters a large cavity in the cervical cord that communicates with the ventricles.

should be reestablished if they are blocked, and, if there is a hydromyelia cavity, it should be drained. The increased accuracy obtained by the use of surgical magnification facilitates dissection through the scar over the fourth ventricle, inspection of the lower fourth ventricle, identification of the proper area to incise the cord, and the final incision into the spinal cord.

The cord incision should be in the posterolateral cord at the dorsal root entry zone because this is the thinnest area. The natural enlargement of the cavity along the dorsal root entry zone also leads to a proprioceptive deficit in the upper extremities, hence, incision here will not aggravate the patient’s deficit. The posterolateral myelotomy used in hydromyelia is different from the incision used in intramedullary tumors. A midline myelotomy is made between the fasciculi gracilis to expose and remove an intramedullary tumor. An incision approximately 1 cm long is made to drain a hydromyelia.

Percutaneous needleing of the hydromyelia cavity has been mentioned as a possible mode of therapy; however, the author has noted that removal of fluid by aspiration at surgery is followed by rapid refilling of the hydromyelic cavity from the ventricular system. In hydromyelia, the opening into the cord should be large enough to prevent its closure. It seems unlikely that a needle tract would remain patent.

Gardner initially recommended decompression at the foramen magnum and plugging of the upper end of the patent central canal at the area of the obex with a small piece of the muscle. There is often a membrane stretched across the fourth ventricle just posterior to the patent central canal, which would help retain a pledget of muscle (Fig. 3 center left); however, the author favors incision between the posterior and lateral cords to drain the hydromyelia rather than risking damage to the hypoglossal and vagal nuclei which are near the obex.

Recently Gardner, et al., have advocated a procedure called terminal ventriculostomy. The terminal ventricle is a dilated portion of the central canal extending below the tip of the conus medullaris into the filum terminale. A laminectomy is done over the caudal limit.
Arnold-Chiari malformation and hydromyelia

of the fluid sac and the filum is opened. This procedure does not decompress the malformation at the foramen magnum but may prove satisfactory if only hydromyelic symptoms are present.

Shunting from the lateral ventricle to the atrium or peritoneum has been recommended. However, the majority of these adults, even those with marked hydromyelia deficits, have only a minimal degree of ventricular dilatation and the small ventricles make shunting difficult. In addition, CSF pressure has been normal in our adult cases. Even if shunts were done, there remains a hydrostatic fluid column from the ventricles acting on the cord when the patient is in the upright position. Shunting would be indicated if the ventricles were large or intracranial pressure was increased.

The finding of normal-sized ventricles in these patients has led the author to conclude that intracavitary pressure too low to dilate the ventricles may cause progressive cord destruction by distention. A recent case in an infant supports this concept. This child had hydrocephalus after birth that required shunting; the shunt became infected, was removed, and not replaced because head and ventricular size remained normal. The child then developed a progressive, severe lower motor neuron deficit in the arms. Repeat contrast studies showed normal-sized ventricles but positive contrast ventriculography showed a hugely dilated cord in the cervical region (Fig. 7).

The common association of bulbar symptoms with the syringomyelic cord syndrome has often led to a diagnosis of syringobulbia. This experience has led the author to conclude that this bulbar dysfunction is not caused by a rostral bulbar extension of the syringomyelic degenerative process but by the Arnold-Chiari malformation.

References


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