Meningomyelocele and progressive hydromyelia

Progressive paresis in myelodysplasia

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Five cases of myelodysplasia with progressive paraparesis are presented. Three of the five patients developed spasticity, but dissociated sensory loss and loss of sphincter control was not a prominent feature. All were found to have compensated hydrocephalus and extensive communicating hydromyelia. The use of myelography and ventriculography in the diagnosis of hydromyelia is discussed. Ventricular drainage led to clinical improvement in two cases and radiological improvement in one. The relationship of compensated hydrocephalus, meningomyelocele, and progressive hydromyelia postnatally may support the hydrodynamic hypothesis of myelodysplasia.

Key Words • meningomyelocele • hydromyelia • syringomyelia • hydrocephalus • spina bifida • paraplegia

Progressive neurological deficit has long been recognized as a complication in patients with myelodysplasia. Bladder dysfunction has most frequently been documented but isolated cases of progressive spasticity or syringomyelic syndromes have also been reported. Such changes have generally been attributed to spinal cord tethering and compression at the meningomyelocele site. Surgical release of the cord in such cases has had equivocal success.

In our long-term follow-up of meningomyelocele patients, we have been impressed by the association of progressive paresis with compensated hydrocephalus. Five such patients have been evaluated and all have been found to have extensive communicating hydromyelia with mild to moderate compensated hydrocephalus. We will describe the clinical presentations of these five patients and propose various methods of evaluation and treatment.

Case Reports

Case 1

This 19-year-old woman had had a lumbosacral meningomyelocele repaired at the age of 2 months. A lumboperitoneal shunt was performed at the age of 5 months for
P. V. Hall, R. L. Campbell and J. E. Kalsbeck

marked flexor contractures and hyperreflexia in her arms. Hypesthesia with preservation of pain sensation was present below the right knee. Sphincter function was still intact. The meningomyelocele repair was tender and compression produced dysesthesia down both legs. The head circumference was 63 cm. A low-pressure Holter ventriculoperitoneal shunt was inserted on this occasion and by 2 weeks postoperatively, she could voluntarily extend both legs and could abduct her thighs. She is now able to raise her leg and wiggle her toes on the right side. Sensory and sphincter function have remained unchanged.

Case 2

This 7½-year-old boy had had a lumbo sacral meningomyelocele repaired on the day of birth. A ventriculocisternal shunt was inserted at the age of 1 month. He subsequently was able to walk in short leg braces, and had an L-5 motor and sensory level. Sphincter function was absent. At the age of 1 year, he was considered independent of shunt.

In January, 1974, the patient developed rapid deterioration of gait and within 2 days could not walk. Two weeks after onset, he was admitted to Riley Children's Hospital; he was alert with no signs of raised intracranial pressure. A flaccid paraplegia, with only minimal hip flexion on the left, was evident. Sensory examination revealed an L-5 level on the right and an L-3 level on the left with no sensory dissociation. There was generalized spinal tenderness with a tense, exquisitely tender meningo myelocele repair site.

A shunt injection confirmed nonpatency. A ventriculogram was performed showing moderate enlargement of the ventricles with a mean intraventricular pressure of 160 mm H₂O. A myelogram by lateral cervical puncture revealed enlargement of the lumbar cord, maximal at L-2 (Fig. 2 left). A diagnosis of hydromyelia was made and a ventriculoperitoneal shunt with a low-pressure Hakim valve was instituted. Within 24 hours, hip flexion improved bilaterally and the meningo myelocele site was sunken and nontender. Three days postoperatively, the patient could raise both legs, and at 1 week he was again walking in short leg braces. Repeat myelography revealed narrowing of the previously enlarged lumbar cord (Fig. 2 right). Improvement has been maintained over 6 months.
Meningomyelocele and progressive hydromyelia

Case 3

This 5-year-old girl was born with a sacral meningomyelocele, repaired shortly after birth. She developed with moderate intellectual retardation, but was able to walk well and was continent. Over 2 years prior to admission, she developed progressive spastic paraparesis for which she was evaluated in May, 1974. Examination revealed an irritable, retarded child with a head circumference of 57 cm. There was marked spasticity with clonus and hyperreflexia of all four extremities. The patient walked with a spastic gait. Sensory examination could not be assessed; sphincter function was intact.

Cervical x-ray films revealed the interpeduncular distance at the upper limits of normal. A myelogram by lumbar puncture showed marked dilatation of the spinal cord from C-4 to T-3. Above and below this, the cord was of normal size. A ventriculogram revealed moderate hydrocephalus. Pantopaque was introduced into the ventricles, filling the upper cervical central canal, which was markedly dilated. Films repeated 24 hours later revealed no evidence of Pantopaque within the ventricular system. Two days after the examination, the patient was found dead.

Autopsy confirmed the diagnosis of marked cervical thoracic hydromyelia. The hydromyelia was partly interrupted by a fenestrated membrane at midcervical levels, the end point of the Pantopaque. X-ray films taken postmortem showed the ventricular Pantopaque within the central canal as far as the lumbar region. The cervical cord was then injected with Conray and showed a hydromyelia extending from the upper cervical region to the lumbar region (Fig. 3).

Case 4

This 1½-year-old girl had had a lumbo-sacral meningomyelocele repaired at birth. A ventriculoatrial shunt for hydrocephalus was performed at the age of 1 month, and subsequently removed at 5 months of age when staphylococcal septicemia was found. She was left with bilateral L4-5 motor function. Sensory and sphincter function were well preserved. She was considered of above average intelligence at school.

At the age of 10 years, she developed a rapidly progressive kyphoscoliosis with weakness of the left leg. Because of the...
weakness, she was admitted to Riley Children's Hospital for reevaluation. Sphincter function was unchanged. Examination in June, 1974, revealed an alert girl with a head circumference of 60 cm. Good motor function was found to an L-5 level on the right side. On the left, moderate hip flexion with only a flicker of knee extension and dorsiflexion of the foot was found. There was hyperreflexia in the left arm. Hypalgesia was present below a T-4 level bilaterally with hypesthesia below the knee on the left side.

A lumbar Pantopaque myelogram revealed marked enlargement of the lumbar cord above the level of the meningomyelocele with a total block at about L1-2. A radioisotope ventriculogram with 3 mCi of isotonic technetium-99 labeled serum albumin demonstrated moderate hydrocephalus with rapid filling of the spinal area down to the sacrum (Fig. 4). No basal cisternal filling was identified. In view of the marked cord enlargement with spinal block, we feel that the radioisotope filled the central canal.

A low-pressure Holter ventriculoperitoneal shunt was performed on June 14, 1974, with no clinical improvement by 5 days postoperatively. However, the subarachnoid Pantopaque dye flowed freely after ventricular shunting.

Case 5

This 12-year-old boy had a lumbar lipomeningomyelocele repaired at the age of 1 year, and was left with an L-5 motor level bilaterally. He had some degree of sacral sensation, but no sphincter control, and had compensated hydrocephalus.

In 1970, lumbar fusion was performed for scoliosis secondary to an L-5 hemivertebra. Shortly after, the patient noted progressive weakness and wasting of his right hand. In November, 1972, he was found to have marked weakness and atrophy in a C-8 to T-1 distribution on the right side. The interpeduncular distance of the cervical spine was well above normal limits. A myelogram revealed marked enlargement of the cervical cord from C-3 to C-7. A pneumoencephalogram showed normal lateral ventricles with mild enlargement of the fourth ventricle.

In July, 1973, he noticed a marked exacerbation in his symptoms on the right and the development of clumsiness in his left arm. Examination at that time revealed a normocephalic boy with normal mental function. There were no signs of raised intracranial pressure. There was moderate weakness of the biceps, severe weakness of the triceps, and marked weakness and atrophy of the intrinsic on the right. The left triceps and intrinsic muscles also showed weakness. Normal hip flexion and knee extension with weak dorsiflexion of the foot was present in the left leg, with only weak hip flexion and a flicker of knee extension in the right leg. Hyperreflexia was present in the left arm. Sensory examination revealed hypalgesia below L-4 on the right.

An air myelogram (Fig. 5) revealed marked enlargement of the cervical cord, but no change with the Valsalva maneuver. A suboccipital cranectomy and laminectomy of C-1 to T-2 was performed July 23, 1973. An extensive communicating hydromyelia was found. Clear fluid was aspirated with a protein of 60 mg/dl. The dura was left open. Postoperatively, the patient's condition has remained stable without improvement.
Meningomyelocele and progressive hydromyelia

Summary of Cases

All five cases were considered to have compensated hydrocephalus and had been either without shunts or without functioning shunts for years prior to their neurological deterioration. Patients in Cases 1, 2 and 3 had low-level meningomyeloceles with good function, and presented with progressive spasticity or paraparesis. In Case 1, severe spastic paraparesis with hyperreflexia of the arms developed over a 3-year-period; the patient in Case 2 became flaccidly paraplegic over 2 weeks. In Case 3, marked spastic quadriplegisa developed over 2 to 3 years, while the patient in Case 4 presented with severe kyphoscoliosis over 1 year, with asymmetrical loss of function and flaccid weakness of the left leg. Case 5 was unusual in that extensive weakness and spasticity developed asymmetrically in the arms with less severe changes in the legs.

All patients except the one in Case 4 had generally well-preserved pain, temperature, touch, and posterior column sensation. In Cases 1, 3, and 4, there was no deterioration in previous sphincter function. A feature of Cases 1, 2, and 4 was the development of a swollen tender meningomyelocele repair, often with burning dysesthesias radiating down both legs.

All five patients were initially evaluated by Pantopaque myelography. Accidental puncture of the ballooned cervical cord in Case 1 revealed extensive communicating hydromyelia. In Cases 3 and 5, markedly swollen cervicothoracic cords were demonstrated on myelography. In Case 2 there was swelling of the lumbar spinal cord with narrowing after a successful shunt procedure and clinical improvement. In Case 4 there was a complete myelographic block at the lumbar level.

Ventriculography with positive contrast in Case 3 demonstrated communication of the central canal with the ventricles. Extensive hydromyelia was demonstrated at autopsy. In Case 4, isotope ventriculography revealed filling of the central canal to a lumbar level. In Case 5, a communicating hydromyelia was demonstrated at the time of laminectomy.

Discussion

The development of spasticity and loss of function in meningomyelocele patients has previously been ascribed to stretching of the tethered cord,\textsuperscript{5,6,14,24} the development of gliosis with cavitation,\textsuperscript{20} or to compression by adhesions and lipomas.\textsuperscript{5,18,19} Spastic paraparesis may also be due to hydrocephalus with stretching of the cortical spinal fibers of the lower extremity to a greater extent than those of the upper extremity by the dilated lateral ventricles. However, the pathological association of hydromyelia with meningomyelocele has long been known. Keiller\textsuperscript{21} invariably found hydromyelia at autopsy of patients with meningomyelocele. Cameron\textsuperscript{7} found that 20 of 22 cases of meningomyelocele with a typical Arnold-Chiari malformation had associated hydromyelia. Communication of the central canal with the fourth ventricle has also been demonstrated at autopsy by injection of the meningomyelocele with positive contrast.\textsuperscript{2} Recognition of hydromyelia as a cause of progressive cord injury postnatally in meningomyelocele patients has been slow, however. Lassman, et al.,\textsuperscript{22} in 1968, reported an unshunted 7½-year-old meningomyelocele patient with a 6-year history of progressive spastic paraparesis. Laminectomy confirmed marked hydromyelia, but was not followed by clinical improvement.
Gardner and his coworkers, developing Morgagni's original observations, hypothesized that hydrodynamic forces arising from persistent fetal hydrocephalus acted down the central canal to rupture the cord and produce a meningomyelocele. If such hydrodynamic forces are operative, then persistence postnataally in unshunted patients could lead to progressive cord damage. We feel our cases confirm this by demonstrating that progressive neurological deficit in meningomyelocele patients can frequently be due to progressive communicating hydromyelia. Hydrocephalus was spontaneously compensated in these patients, possibly because of decompression down the central canal. These clinical observations accord well with recent experimental work. Eisenberg, et al., found by ventricular isotope and perfusion studies that hydrocephalus from fourth ventricular outflow obstruction in a cat was decompressed by means of the central canal.

We perform Pantopaque myelography by either cervical or lumbar routes routinely when we suspect hydromyelia. Significant hydromyelia is suggested by cord enlargement at either a cervical or a lumbar level. Reexamination of the dye column after successful shunting may reveal collapse of the cord as seen in Case 2, or opening of a myelographic block as was seen in Case 4.

Ventriculography or arteriography is carried out to establish the degree of hydrocephalus, which has generally been mild to moderate. Pantopaque introduced into the ventricles has demonstrated communicating hydromyelia. This has previously been recommended as a useful and safe procedure in newborn meningomyelocele patients. However, Bertrand feels that not all the cases of communicating hydromyelia will be demonstrated by positive contrast ventriculography. Such a procedure, consequently, is of questionable value and may be hazardous in older patients, as shown in Case 3. If it is used, we feel Pantopaque ventriculography should be followed soon by surgery. We have now tried radioisotope ventriculography with initial success.

Surgical treatment of progressive neurological deficits in meningomyelocele patients has generally involved exploration of the meningomyelocele and freeing of adhesions. The results have generally been unrewarding at this and other institutions. We believe a more rational approach is to treat the associated hydromyelia.

Laminectomy with cyst aspiration has long been used in syringomyelia with some success. However, in both Lassman's Case 6 and in our Case 5, this procedure failed to bring about improvement. Gardner has proposed decompression of the fourth ventricle with plugging of the obex, while others have felt decompression of the posterior fossa alone might be used in syringomyelia. The patient in our Case 1, treated by extensive decompression, continued to deteriorate postoperatively.

If ventricular pressure changes are the underlying pathogenesis of communicating hydromyelia, then ventricular drainage should decompress the central canal and stabilize cord pathology. Ventricular decompression by means of a shunt has now been used with some success in syringomyelia. Of our five cases of hydromyelia, three have now been treated by means of a ventriculoperitoneal shunt. Only Cases 1 and 2 have been followed for any period, and both patients have improved encouragingly. The patient in Case 1 had marked reduction of spasticity and some improvement of strength, while the one in Case 2 has recovered to his previous ambulatory status. Hydromyelia has been decompressed in Case 2 as demonstrated on reexamination of the dye column.

We feel that intracranial pressure, ventricular size, or head size may not be the only criteria in the future for the use of ventricular shunt systems. The prevention or treatment of communicating hydromyelia may also be considered a valid indication. Furthermore, the proposed use of head binding in meningomyelocele patients for the associated hydrocephalus may be contraindicated, since hydromyelia may be exacerbated in these cases.

References
Meningomyelocele and progressive hydromyelia


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