Syringomyelia following lumboureteral shunting for communicating hydrocephalus

Report of three cases

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Three patients are described in whom syringomyelia was identified long after the treatment of communicating hydrocephalus by a lumboureteral shunt. The reason for syrinx formation in these cases could not be determined. In two there was either autopsy-proven or presumed evidence for arachnoiditis, and in the third patient the symptoms of syringomyelia were acutely aggravated by temporary obstruction of the shunt. The development of a pressure drop from the intracranial compartment to the spinal compartment with crowding at the foramen magnum is also a suggested mechanism.

KEY WORDS • syringomyelia • hydrocephalus • lumboureteral shunt

PROGRESSIVE communicating hydrocephalus has been described in patients with compensated hydrocephalus and myelomeningocele. In this report, three patients are described in whom syringomelia was diagnosed (in one case by autopsy) following the treatment of communicating hydrocephalus by lumboureteral shunts. We have not previously recognized this complication despite wide experience with lumboperitoneal shunts and with lumboureteral shunts since their introduction in 1948 and their gradual discontinuance in the late 1960’s. Sterile and pyogenic meningitis have been serious problems in patients with lumboperitoneal shunts, and we have seen scoliosis, cauda equina syndromes, and arachnoiditis in patients with both lumboperitoneal and lumboureteral shunts involving Silastic or polyethylene tubes. Although we have not previously recognized syringomyelia in hydrocephalic patients with lumbar shunts, we feel that this indolent process should be conscientiously sought among these patients.

Case Reports

Case 1

This 23-year-old man was first seen in 1952, at 4 months of age, for evaluation of an enlarging head. Communicating hydrocephalus was diagnosed by ventriculography. After a left nephrectomy, a lumboureteral shunt with a polyethylene catheter was in-
Syringomyelia after lumboureteral shunt inserted at L-2. By 6 years of age he was noted to be slightly retarded; he had a mild scoliosis, walked with a stiff gait, and had bilateral ankle clonus and a left Babinski response. At 9 years of age the shunt became obstructed, causing headache, lethargy, and vomiting. The polyethylene tube had withdrawn from the ureter, and a new, longer tube was inserted. Examination at this time showed he had a stiff neck, mild weakness of dorsiflexion of his left foot, and a hyperactive left knee jerk.

At 11 years of age he had several episodes of headache interpreted as intermittent shunt malfunction, during which time air injection demonstrated communication between the lumbar subarachnoid space and the ventricles. At 12 years of age he was treated with intravenous antibiotics for a urinary tract infection due to *Streptococcus faecalis*. Meningitis was suspected and he had a pleocytosis of 13,600 white blood cells in the spinal fluid, which was sterile. Examination revealed a mild spastic paraparesis.

At 21 years of age he developed severe back and leg pains and progressive difficulty in walking. He had developed a severe kyphoscoliosis, had a stiff neck, left shoulder girdle weakness, weakness of plantar and dorsiflexors of both feet, and absence of both ankle and right knee jerks. Lumbar puncture was unsuccessful, but subarachnoid Pantopaque introduced via the lateral CI-2 approach traversed the spinal canal and passed through the shunt. The Pantopaque had a streaky appearance in the subarachnoid space, and the spinal cord could not be visualized. Also, ventricular filling could not be achieved with air injection by this approach. An arteriogram revealed ventricular enlargement and a ventricular peritoneal shunt was inserted. Over the next 4 months he had increasing back pain and leg weakness, as well as right arm spasticity and mild sensory loss over the left shoulder and lower right part of the body. A shunt reservoir tap revealed a pressure of zero, but cisternal and lumbar punctures were unsuccessful. The lumbar ureteral shunt was removed because of the unproved possibility of reflux from the ureter to the subarachnoid space, and he had marked relief from his pain and spasticity.

Two years later, in April, 1975, an anterior thoracic spinal fusion was performed without neurological complications. On the third postoperative day he was extubated and 18 hours later, while prone on a Stryker frame, he had a respiratory arrest from which he sustained severe brain injury and died 7 days later.

At autopsy there was severe acute necrosis and edema of the pons and necrosis and hemorrhage of the cerebellum, making it impossible to preserve the latter intact. The anatomical relationships of the cerebellum and brain stem at the foramen magnum were therefore unable to be determined; however, a large cystic cavity was present within the spinal cord, extending the entire length of the cord, lined partially by fibrillary astrocytes and in places incompletely by a single layer of low cuboidal ependymal cells. In portions of the cervical cord, the cystic cavity contained fragments of necrotic cerebellar cortex and subcortical white matter; however, the exact relationship between the cavity and the fourth ventricle could not be discerned. In the region of the cauda equina the leptomeninges were heavily overgrown with fibroblasts and included foci of calcification.

Case 2

This 27-year-old man was first seen in 1948, at 5 months of age, for evaluation of an enlarged head. Communicating hydrocephalus was diagnosed by air ventriculography and a right lumboureteral shunt was inserted with sacrifice of the right kidney. At 14 months of age he was unable to walk or stand; he had an inconstant equinus deformity of the right foot and bilateral Babinski reflexes. At 18 months he was hospitalized elsewhere for possible meningitis with a pleocytosis but sterile spinal fluid. When he was 6 years of age a stiff gait and developing scoliosis were noted. Between the ages of 9 and 15 years his scoliosis progressed despite bracing. At 26 years of age the thoracic scoliosis had progressed still further and a Harrington rod posterior fusion was performed from T-3 to L-2 with a benign postoperative course. Six months later he was readmitted to the hospital because of back pain at the level of his lumboureteral shunt.

Neurological examination revealed flattening of the left nasolabial fold, palatal droop on the left, mild tongue atrophy on the left, bilateral wasting of muscles in his arms and hands, spasticity of his legs, bilateral...
Babinski reflexes, and a suspended sensory impairment between C-2 and T-2. Air myelography demonstrated a normal cervical cord as well as patency of the shunt, but ventricular filling could not be achieved and a computerized tomography scan revealed hydrocephalus. The diagnosis of syringomyelia and syringobulbia was made on what was felt to be strong clinical evidence, even though the cervical cord appeared normal on air myelography. A ventriculoperitoneal shunt was inserted and the ureteral shunt removed. One year later his neurological examination was unchanged.

Case 3

This 15-year-old boy was first seen in May, 1960, at 2 months of age, for evaluation of an enlarging head. Communicating hydrocephalus was diagnosed by a ventriculogram, and following a left nephrectomy a lumboureteral shunt was inserted using polyethylene tubing. At 5 years of age he was evaluated for left flank pain for which no cause could be found. The only abnormality noted on examination was limitation of extension of the lumbar spine. At 9 years of age he developed a limp, and a mild scoliosis and neck stiffness were noted. The following year the scoliosis was again noted, as well as mild right calf atrophy.

At 15 years of age he noted the onset of weakness of his left hand. Examination revealed atrophy of the intrinsic muscles of the left hand, diminished reflexes in the left arm and a sensory loss to pinprick between the C-3 and T-1 dermatomes on the left. The patient was admitted to the hospital for air myelography to rule out syringomyelia. Lumbar and cervical punctures were attempted unsuccessfully, the latter yielding a few drops of blood-stained spinal fluid. After the study the urine was found to be bloody. About 15 hours later he developed acute respiratory distress requiring ventilatory assistance, and became quadriplegic with bilateral Babinski reflexes, papilledema, and retinal hemorrhages. A ventricular tap was performed and a pressure of over 600 mm H2O noted. After initial brisk drainage, however, constant ventricular drainage over the subsequent 14 hours yielded only 60 cc of CSF, indicating that his shunt, which had most likely been plugged by blood from the attempted diagnostic study, had probably begun working again. During the 48 hours following the establishment of ventricular drainage, he began to recover motor function, first in his legs. His right arm remained weak, particularly in the intrinsic hand muscles; weakness in his left hand had been the presenting complaint. At ventriculography he had mildly dilated ventricles and air was maneuvered into a dilated central canal of the cervical and upper thoracic cord (Fig. 1). A ventricular peritoneal shunt was inserted and recovery continued so that 3 months later the only neurological abnormalities noted were

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**FIG. 1.** Case 3. Air inserted into the lateral ventricle appeared in the central canal of the cervical spinal cord (single arrow). The posterior margin of the cord is demonstrated by the small arrows.
weakness of his right palmar interosseous muscles and a possible right Babinski reflex. One year later he was neurologically normal.

Discussion

Three cases are described in which symptomatic syringomyelia developed following lumboureteral shunting for communicating hydrocephalus. The diagnosis of syringomyelia was made by autopsy in Case 1, by strong clinical neurological findings in Case 2, and by demonstrating air within the syrinx in Case 3. Polyethylene catheters were used in all cases. All three patients developed scoliosis, two of them requiring spinal fusion. Severe arachnoiditis was found in the one patient who was autopsied (Case 1). This diagnosis was suggested in Case 3 because lumbar and cervical punctures were unsuccessful. In Case 2 there was failure of ventricular filling at pneumoencephalography but no other evidence of arachnoiditis. Some degree of ventricular dilation was present in each patient, however in none were the symptoms of spinal cord pathology preceded by more classical evidence of shunt malfunction. In one case temporary shunt obstruction resulted in severe spinal cord decompensation.

The causes of syrinx formation in our three patients cannot be definitely ascertained. It is possible that congenital factors or subclinical shunt obstruction were contributory; there was at least mild ventricular dilation in each case as evidence for the latter. In one patient severe arachnoiditis was unquestionably present and therefore could have been a definite contributing factor impairing the flow of CSF out of the fourth ventricle. Evidence for arachnoiditis was slight and indirect in the other two patients. If the development of arachnoiditis, either due to the presence of the subarachnoid polyethylene tubing or due to sterile or pyogenic meningitis, was a contributing factor to the syrinx formation, this would represent a clinical analog to experimental syringomyelia secondary to Kaolin-induced arachnoiditis.1,3,4,7,12 In these experiments it was presumed that arachnoiditis causes obstruction of the outlets of the fourth ventricle and, by such mechanisms as proposed by Gardner,6 or du Boulay, et al.,2 results in syrinx formation. Hall, et al.,7 noted that an association between syringomyelia and arachnoiditis was suggested by Joffroy and Archard9 in 1887.

Polyethylene shunt tubing in the lumbar subarachnoid space has been associated with severe arachnoiditis not only in our hospital, but also elsewhere, regardless of whether the distal end of the shunt was in the peritoneum6 or in the ureter.10 In a series of 285 patients with polyethylene catheter lumboperitoneal shunts, 72 patients had lordosis, kyphoscoliosis, significant neurological deficits, or, in five patients, paraplegia, all attributed to polyethylene-induced arachnoiditis.8 It is not reported that syringomyelia was recognized in any of these patients. In another series of 80 patients with polyethylene lumboperitoneal or lumboureteral shunts, 34 developed scoliosis, although specific neurological deficits were not described.10 In three of the cases, a severe fibrous arachnoiditis was demonstrated either at autopsy or at operation. In each of the two autopsied cases a syrinx was also demonstrated; however, the authors speculated only mildly that there may be a correlation between the scoliosis and the arachnoiditis or the syrinx. We have also observed scoliosis and cauda equina symptoms in patients with lumboperitoneal shunts that use Silastic tubing. In our three patients with syringomyelia, the relationship between arachnoiditis and syrinx formation remains uncertain.

A final possible explanation for the development of syringomyelia in patients treated by lumbar shunts is that the shunt in the lumbar space produces a pressure drop from the intracranial to the spinal compartments, resulting in a progressive crowding of cerebellar and brain-stem structures at the foramen magnum. Such an iatrogenic “Arnold Chiari malformation” would then impair outflow of CSF from the fourth ventricle, thus promoting the propulsion of fluid down the central canal of the spinal cord. Although this is a completely theoretical explanation for the development of syringomyelia in our cases, we have seen one patient with a lumbar shunt in whom serial pneumoencephalograms and arteriograms demonstrated a descent of the fourth ventricle and cerebellar tonsils, but without evidence, at least thus far, of syringomyelia.

In conclusion, syringomyelia appears to be a delayed risk in patients with communicating hydrocephalus treated by lumbar shunts.
The role of arachnoiditis or of the type of shunt tubing employed or the role of any other possible contributing factor in the development of syringomyelia in such patients remains to be determined. We feel that it is important to be alert to the possibility of an indolent syringomyelia in all patients with lumbar shunts for communicating hydrocephalus.

References


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