SYRINGO-ENCEPHALOMYELIA

DISCUSSION OF RELATED SYNDROMES AND PATHOLOGIC PROCESSES, WITH REPORT OF A CASE

HOMER D. KIRGIS, M.D., AND DEAN H. ECHOLS, M.D.

Section on Neurosurgery, Ochsner Clinic, and Department of Surgery, Tulane University School of Medicine, New Orleans, Louisiana

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The formation of cystic cavities within the brain and spinal cord may be secondary to a multiplicity of pathologic processes, including thromboses and emboli of the arteries, inflammatory and neoplastic reactions, trauma, and various other disorders that may interfere with the normal metabolic activity of some portion of the central nervous system. The syndromes that usually have been termed syringomyelia, syringobulbia, or syringopontia seem to present a special problem in the formation of cysts within the central nervous system. These terms have acquired certain connotations relative to the origin, location and progression of the cystic lesions. They have customarily been used to designate a syndrome suggestive of a slowly expanding, centrally located, intramedullary lesion. The pathologic process, at least in the majority of cases that have been examined in detail, has consisted largely of the cyst, with evidence of gliosis in the adjacent tissue. However, certain investigators\textsuperscript{7,17} have stressed the presence of areas of homogenization which they believe are sites of future cystic formation.

There is wide divergence of opinion regarding the mechanism of formation of the cyst and its fluid. Most investigators\textsuperscript{5,8,9,14,16} have considered the cyst to be the product of relatively benign glial proliferation and necrosis of the involved tissue. The process has generally been postulated to originate by atypical multiplication and differentiation of cells, probably spongiosblasts, in the vicinity of the deep portion of the posterior raphé of the spinal cord or the median raphé of the brain stem. Ependymal cells have been reported to form the lining of some syringomyelic cysts; others have apparently had no distinct epithelial wall. The presence of ependymal cells may, at least to some extent, be dependent upon the communication of the cyst with the central canal of the cord or with some remnant of the primitive neural canal. Regardless of the type of cells forming the wall of the cyst, the essential symptom-producing activity appears to be the gradual accumulation of fluid within the cyst with compression of neighboring tracts and nuclei. Although the original fluid of the cyst may be the result of necrotic tissue having undergone liquefaction, its volume probably is increased by transudation of fluid across the wall of the cyst or by the secretory action of the cells lining the cyst. One is impressed by the probable importance of the
latter process by the rapid re-accumulation of fluid in such cysts from which the fluid has been aspirated but in which no adequate, continued system of drainage has been instituted. The origin of syringomyelic cysts from what have been described as ruptured germinal epithelial cells has been suggested but the pathophysiologic process involved is not clear.

It has been pointed out that anatomically the central portion of the spinal cord, the area in which the great majority of these cysts seem to originate, is particularly vulnerable to vascular disturbances. This area is supplied by the terminal branches of the posterior and anterior spinal arteries and it has been postulated that generalized disease of the spinal arteries may result in greatest changes in this central area. Although some investigators have reported hyalinization and other pathologic changes in the adjacent vessels, these changes have not been consistently in evidence in this type of cystic disease.

Hassin concluded that syringomyelia and syringobulbia are manifestations of abiotrophy. At postmortem examination of the spinal cord of a patient with syringomyelia, he observed evidence of degeneration in the central area of the spinal cord at considerable distances from the cyst. These areas were described as zones of homogenization which represented a reaction of degeneration without evidence of previous glial proliferation.

A review of the literature reveals that processes, varying from cystic dilatation of the central canal of the spinal cord (which might be more correctly termed hydromyelia) to cystic degeneration within a glioma, have been discussed under the term syringomyelia. An insufficient number of cases that might be termed true syringomyelia and in which treatment consisted of establishing a permanent communication between the cavity of the cyst and the subarachnoid space has been followed to reach definite conclusions regarding the neoplastic significance of the proliferation of glial cells which has generally been reported to be present about the cyst. Purep, however, described 2 such cases of patients who were living 4 years and 3½ years, respectively. Each was still free of evidence of recurrence of compression by the cyst of adjacent neural structures. The same may be said in regard to reaching a final decision relative to the condition representing a progressive degenerative disease. Numerous patients have been observed to survive many years following acute manifestations of syringomyelia, finally succumbing to some intercurrent infection, which fact does not support the latter theory.

Most reports of postmortem examination of these patients indicate that cavitation appears to have begun in the region immediately posterior to the central canal and to have extended asymmetrically into the lateral and posterior funiculi and superiorly and inferiorly close to the base of the posterior columns of gray matter. Reports of the development and progression of neurologic defects in these patients support these observations and show that the lower cervical and cervicothoracic regions of the spinal cord are the most common sites of origin of the cysts. Multiple cysts have been
described\$ as well as a single cyst, which extended almost the complete length of the spinal cord.\$  
The preference of the syringomyelic process for the lower cervical or cervicothoracic region results in the common presenting complaint of diminution of normal sensation in the hands with weakness of the muscles of the hands and forearms and inability to make discrete movements with the fingers. The symptom complex most typical of syringomyelia includes a bilateral area of hypothermia and hypalgesia with reduced acuity of these sensations on only one side or more pronounced on one side below this level. The sensations of touch and pressure may be unaltered, and the term dissociation of sensations has been used to refer to this selective interruption of sensory paths. The acuity of proprioception and of the sensation of vibration may be greatly reduced. The sensations of touch and pressure survive more persistently, not only because of the greater distance of the anterior spinothalamic tract from the usual site of origin of the syringomyelic cysts, but because of the comparatively long ascending homolateral branches of the primary afferent neurons which mediate these sensations. Characteristically, motor manifestations of syringomyelia include spasticity and weakness of the muscles below the upper level of defective sensation. This may be confined to or considerably greater on the side opposite the more pronounced sensory loss. This neurologic picture, of course, is that of a deep lesion which has involved the fibers mediating the sensations of pain and temperature as they cross the mid-line and which has expanded asymmetrically to encroach upon the lateral and posterior funiculi. Invasion of the anterior columns of gray matter may be evidenced mainly by flaccid weakness or palsy of the intrinsic muscles of the hands or of the muscles of the forearms.

In addition to these sensory and motor manifestations of syringomyelia, further evidence of the disease may consist of so-called trophic disturbances in the periphery and mutilating lesions which might be expected in the absence of adequate pain and temperature sensibility.

If the lesion is confined to the lumbar region of the spinal cord, the motor deficiency may consist of a flaccid weakness of some of the muscles of the lower extremities with evidence of an upper motor neuron type of lesion affecting the muscles supplied by more inferior segments of the cord.

Syringobulbia and syringopontia may present a considerably more inconsistent picture. Usually there is evidence of damage to several of the nuclei of the brain stem. The most commonly affected include the vestibular nuclei, the nucleus of the spinal tract of the trigeminal nerve, and the visceral and somatic efferent nuclei of the medulla. The fiber tracts most often sufficiently involved at this level to form a significant part of the clinical picture include the medial lemniscus, the descending tract of the trigeminal nerve and the solitary tract. Unilateral damage to these tracts results in reduction in the normal acuity of exteroceptive and proprioceptive sensations on the contralateral side inferior to the head and similar reduction in sensation over the homolateral side of the head. Usually in syringobulbia
Syringo-encephalomyelia, there is reduction of these sensations bilaterally over the head, trunk and extremities. The common variation in the degree of this hypesthesia suggests the usual bilateral asymmetry of the lesion. Evidence of the pathologic process causing clinically recognizable dysfunction of the autonomic centers and the pyramidal and extrapyramidal systems is less frequently seen. There is, however, no complex of symptoms that can be said to be nearly so characteristic for syringobulbia and syringopontia as that which often is presented by syringomyelia. In general, the signs and symptoms of syringobulbia or syringopontia are those of any glioma of the brain stem except that they may signify greatest involvement of structures relatively close to the floor of the 4th ventricle, such as the vestibular nuclei, the nucleus ambiguus, and the nuclei of the 5th, 6th, 7th, and 12th cranial nerves and of the long ascending conduction pathways.

The therapeutic measures for cysts of the brain stem and spinal cord, which at this time are considered of value, are incision and drainage, and irradiation. The latter has been reported to give varying degrees of relief. Frazier and Rowe° emphasized the importance of remembering that irradiation affects only the gliosis, and consequently is of little value in the presence of cavitation. It is generally conceded that such patients demonstrate most improvement following exposure of the cyst with incision and drainage.4,5,6,10,12,13,15 It has been advocated that a foreign body be inserted into the cyst in such a way that the permanency of the communication between the cavity of the cyst and the subarachnoid space could be assured. Various substances such as dura mater, silk sutures, muscle, gutta percha and silver clips have been used. If the process is fundamentally benign and symptoms develop only because the continued secretory activity of the lining cells or some similar mechanism causes the slowly expanding cyst to exert undue pressure on adjacent structures, equalization of the fluid dynamics within the cyst and the subarachnoid space should effect a cure. It seems improbable that release of the contents of such a cyst into the subarachnoid space would ever overtax the cerebrospinal fluid absorptive mechanism. Obviously, the process of fluid accumulation within the cystic cavity is the major factor in precipitation of a severe phase of the disease. It is not evident that the proliferation of glial cells results in the formation of a tumor which alone is a significant factor in production of the various signs and symptoms characteristic of the disease.

The following case demonstrates the probable superiority of surgical treatment over irradiation therapy. It also proves the futility of merely establishing a communication between the cavity of the cyst and the subarachnoid space without taking measures to prevent its closure by the formation of scar tissue. Furthermore, it seems to represent a type of case for which the terms, syringomyelia, syringobulbia, syringopontia or syringo-encephalomyelia, might logically be reserved. This would seem consistent in view of the fact that the essential pathologic condition in this case has been adequately demonstrated to consist of formation of fluid within the
central nervous system with accumulation of the fluid in a cyst within the substance of the central nervous system. Involvement of the brain stem by the cyst without communication with the ventricular system seems to exclude the possibility that this case might be classed as an example of hydromyelia, that is, cystic dilatation of the central canal of the spinal cord. The accompanying neurologic disorder seemed dependent upon gradual expansion of the cyst as more and more fluid accumulated.

**REPORT OF CASE**

E. G. S., a white boy, aged 12 years, experienced a brief attack of dizziness in June 1946. Two months later a second attack occurred while diving and was immediately followed by persistent soreness of the muscles of the neck. During the next 3 days the vertigo, sometimes associated with vomiting, became severe. Attacks could be precipitated or intensified by turning the head or eyes to the left. Dysphagia and hoarseness soon developed. When admitted to the hospital on Sept. 5, 1946, he also was complaining of a dull, aching pain in the occipital region, neck and shoulder on the left.

*Examination* revealed severe dehydration, in ability to stand or walk because of generalized weakness and dizziness, horizontal nystagmus which was increased by lateral deviation of the eyes, especially to the left, paresis of the muscles of the pharynx, tongue, and larynx, and diminished acuity of proprioception in all extremities. Motor dysfunction was greater on the left in the case of each motor nucleus affected. There was no diminution of exteroceptive sensations in the distribution of either cranial or spinal nerves. Violent vertigo could be produced by slight degrees of active or passive rotation of the head. There was no papiledema.

The CSF pressure was 180 mm. of CSF. Cautiously applied jugular compression caused a slight rise of the fluid level without subsequent fall. The fluid was clear and the protein content was 325 mg./100 cc.

*1st Operation.* A ventriculogram, made on Sept. 9, 1946, revealed slight dilatation of the 3rd and lateral ventricles without displacement. On the same day, the posterior fossa of the skull was opened through a midline incision. Inspection of the floor of the 4th ventricle revealed an enlarged and cystic brain stem. The most pronounced thinning of the posterior wall of the cyst was at the medullol pontine junction in the region of the left vestibular nuclei. Slightly xanthochromic fluid was aspirated from the cyst at this point through a No. 26 gauge needle. A defect approximately 0.5 cm. in diameter was made in the posterior wall of the cyst in this area. By cautious exploration with a small flexible rubber catheter it was determined that the superior extent of the cyst was at the mesencephalic-pontine junction. The inferior pole of the cyst was not palpated although the catheter was directed downward to the middle-cervical level. The arch of the atlas and the laminae and posterior spinous processes of the 2nd and 3rd cervical vertebrae were removed. The incision in the dura was closed with interrupted sutures.

*Course.* Ability to swallow was regained within 48 hours, when tube feeding was discontinued. The vertigo disappeared and the nystagmus decreased. Phonation rapidly became normal and sensory function became almost intact. During the 2nd postoperative week roentgen-ray therapy directed at the cyst was begun. However, on the 20th postoperative day the patient complained of dizziness and within a week his condition had deteriorated to the preoperative level.
2nd Operation. On Oct. 4, 1946, the previously visualized portion of the brain and cord were reexamined and the remaining cervical segments of the spinal cord were exposed. It was obvious that the cyst had re-expanded. The scar tissue closing the aperture between the cyst and the 4th ventricle was easily identified. An incision was made in the cyst at the level of the 5th cervical segment. A catheter was passed upward in the cyst from this point well beyond the previous opening. It was passed downward slightly beyond the midthoracic level without encountering the lower end of the cyst. It was apparent that the cyst involved mainly the posterior funiculi of the spinal cord and extended from some point inferior to the midthoracic level, superiorly to the junction of the pons and mesencephalon (Fig. 1).

A drain, made by twisting several tantalum sutures together, was inserted into the cyst and brought out through another opening, which was made about 2 cm. inferior to the previously made opening. The ends of the drain were sutured to the deep surface of the dura and the dura was closed tightly.

Course. The patient’s 2nd convalescence was characterized by slow but steady improvement. Evidence of defective circulation of the CSF with inadequate absorption continued for several days. CSF was removed from the subarachnoid space in quantities of 30 to 50 cc. each day for a week. At the end of 2 weeks, the scalp over the craniotomy defect was pulsating normally and the symptoms began to clear more rapidly. The dynamics of the CSF as estimated by bilateral jugular compression became normal. It was evident that the intracystic fluid pressure had become equalized with that of the subarachnoid space undoubtedly as a result of the maintenance of the free transmission of fluid between the two spaces along the tantalum drain.

At the time of discharge from the hospital on the 24th postoperative day, the patient’s greatest complaint was stiffness and weakness of the posterior cervical muscles. Evidence of neuropathologic changes consisted mainly of slight nystagmus with extreme abduction of the eyes.

The patient re-entered school shortly after returning to his home, and in spite of having missed almost 2 months, soon had completed sufficient work to enable him to continue with the regularly scheduled assignments. Re-examination after 6 months revealed no evidence of neurologic defects and the only complaint was a sensation of weakness of moderate degree after holding the head erect for relatively long periods. Two years following the 2nd operation, the patient was living a nor-
mally active life with no complaints and the neurologic examination yielded no evidence of pathologic changes. Roentgenography revealed the tantalum drain to have the same relation to the adjacent bony structures as immediately postoperatively.

DISCUSSION

In the case reported, syringo-encephalomyelia, consisting of a cyst extending from the mesencephalic-pontine junction to some point below the midthoracic portion of the spinal cord, was manifested clinically mainly by evidence of involvement of nuclear groups of the medulla as well as the tracts mediating proprioceptive sensation. Inspection revealed the cyst to occupy mainly the posterior funiculi of the spinal cord and the region immediately anterior to the 4th ventricle in the brain stem. Decompression by suboccipital craniotomy and upper cervical laminectomy plus excision of a portion of the wall of the cyst gave pronounced but temporary relief. Recurrence of symptoms followed closure of the defect in the wall of the cyst by scar tissue with re-accumulation of fluid and expansion of the cyst. Roentgen-ray therapy did not prevent rapid reappearance of the syndrome. Clinical recovery to date (2 years postoperatively) followed exposure of the cervical segments of the spinal cord and insertion of a tantalum drain between the cyst and the spinal subarachnoid space. With the drain in place and the dura intact, the cyst has the status of an extra ventricle in the system of intracranial and intraspinal fluid formation and absorption. With this relationship established, the cyst cannot exert undue pressure on adjacent nuclei and fiber tracts nor interfere with the normal circulation of cerebrospinal fluid. The prognosis continues to appear favorable after 2 years.

SUMMARY

The terms, syringo-encephalomyelia, syringomyelia, syringobulbia, and syringopontia have usually been used in a restricted sense to refer to what appears to be a pathologic entity consisting essentially of a slowly expanding cyst originating in a paracentral site in the spinal cord or brain stem. Most investigators have considered the cystic formation to be a manifestation of a neoplastic reaction, a developmental defect, or a degenerative process. Regardless of the exact etiology, the significant pathophysiologic activity of the disease is the accumulation of fluid within the cyst and the resultant compression of adjacent neural structures. The clinical manifestations depend upon the exact location of the lesion. The syndrome most characteristic of syringomyelia is indicative of a deeply placed lesion in the cervicothoracic region, involving first fiber tracts crossing or adjacent to the midline and later extending peripherally, usually in the posterior or lateral funiculi. Commonly, there is evidence of destruction of the nuclei of the anterior column of grey matter. Such a cystic lesion in the brain stem most often is manifested by neurologic evidence of damage to the nuclei and conduction pathways close to the floor of the 4th ventricle. Although irradiation therapy has been advocated, it has become more and more apparent that the
only satisfactory method of treatment is incision followed by some additional maneuver to insure permanent communication between the cavity of the cyst and the subarachnoid space. A case of syringo-encephalomyelia illustrating the superiority of surgical over irradiation therapy as well as the importance of establishing a permanent communication between the cystic cavity and the subarachnoid space is reported.

REFERENCES


