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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 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 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 spongioblasts, 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
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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.

Hasson 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. Puusepp, 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