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Is Reversible Enlargement of the Spinal Cord a Presyrinx State?

Thomas H. Milhorat

Mechanisms of syrinx formation have long attracted interest and speculation. Until recently, most believed that syringomyelia was caused by a forceful diversion of CSF from the fourth ventricle into the central canal of the spinal cord attributed to either obstruction of the outlets of the fourth ventricle (1) or the development of a pressure gradient between the intracranial and intraspinal compartments (2). This classical hypothesis, which shaped operative treatment for half a century and led to innovations in surgical technique such as “plugging the obex” to prevent syrinx filling, was quickly challenged after the introduction of MR imaging. Clinicians were quick to note that only a minority of syrinxes communicate directly with the fourth ventricle, and that most are separated from it by a long segment of syrinx-free spinal cord. The need to understand better the mechanisms of syrinx formation better has resulted in a quickening pace of research on many fronts. In this issue of the *American Journal of Neuroradiology* (page 7), Fischbein et al describe a presyrinx state that has potentially important etiologic and therapeutic implications.

Syringomyelia can be classified into three general pathologic types: 1) central canal syrinxes that communicate with the fourth ventricle in association with hydrocephalus; 2) noncommunicating central canal syrinxes that occur with a wide variety of congenital and acquired disorders, including Chiari malformations, arachnoiditis, and extramedullary compressive lesions; and 3) parenchymal (extracanalicular) syrinxes that are found typically in the watershed area of the spinal cord and are associated with conditions that directly injure spinal cord tissue, such as trauma, infarction, and hemorrhage. Parenchymal syrinxes invariably produce neurologic deficits, whereas concentrically enlarged central canal syrinxes can be asymptomatic or produce only nonspecific neurologic findings unless the parenchyma of the spinal cord is dissected paracentrally. Precise clinicopathologic correlations can often be made on the basis of axial MR imaging with consecutive thin sections through the area of cavitation.

A question of critical importance is How do syrinxes fill? Ball and Dayan (3), and subsequently Aboulker (4), were the first to suggest that syrinxes fill from the spinal subarachnoid space and that CSF enters the central canal through the dorsal

roots or Virchow-Robin spaces. There is now convincing evidence that the spinal subarachnoid space is anatomically continuous with the central canal through a series of interconnecting, perivascular and interstitial spaces with a minimum diameter of 150 to 200 Å (5). Under normal conditions, the movement of interstitial fluid appears to occur by bulk flow from transmitted arterial pulsations and extends rostrally through the central canal to the fourth ventricle (6). Current evidence suggests that the formation of noncommunicating syrinxes depends upon an obstruction of the CSF pathways that exaggerates the spinal pulse wave and forces fluid under increased pressure into the interstitial spaces of the spinal cord.

No discussion of spinal cord cavitation can be complete without a consideration of central canal stenosis. Some degree of stenosis occurs in 70% to 80% of the general population, and is caused by a proliferation of subependymal gliovascular nodules that can lead to partial or complete obliteration of long segments of the central canal. Variations in the patency of the canal can affect the pathology of cavitation. Noncommunicating syrinxes, for example, are usually defined rostrally and caudally by canal stenosis, and the competence of the central canal almost certainly influences the ease of cavitation, the location and length of syrinxes, and the rarity of holocord enlargements in patients with adult-onset syringomyelia. It is assumed but not proved that extensive occlusion of the central canal can prohibit syrinx formation.

In the current article, Fischbein et al describe five patients with disorders known to produce syringomyelia who present with clinical evidence of myelopathy, no history of spinal cord trauma, and enlargement of the cervical spinal cord in the absence of cavitation. All patients had objective evidence of an alteration or obstruction of CSF flow, although definitive studies such as CT myelography and cine phase-contrast MR imaging were performed preoperatively in only two patients. The finding of chronic spinal cord enlargement with parenchymal T1 and T2 prolongation indicates an increased water content of the spinal cord from either interstitial edema or venous congestion and ischemia.

The authors advance a novel hypothesis that variations in the competence of the central canal can create a presyrinx state characterized by focal enlargement of the spinal cord when conditions are suitable for syrinx formation. Although there is obviously no way to clinically assess canal competence, the proposition that interstitial edema pre-

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cedes spinal cord cavitation is consistent with known data concerning the movement of CSF through the interstices of the spinal cord. Further support for a presyrinx state is provided by surgical evidence showing that correction of the underlying CSF obstruction can lead to resolution of cord swelling and neurologic improvement. The late evolution of syringomyelia in one patient in whom the patency of the CSF pathways could not be reestablished is of questionable significance because the operative technique included a myelotomy. The role of raised intramedullary pressure in the propagation of syrinxes is currently under investigation (7).

More experience will be required to understand the presyrinx state better. The disease process leading to syrinx formation appears to be a distinct clinical entity that warrants inclusion in the differential diagnosis of clinical myelopathy. Questions left unanswered are the prevalence and natural history of the disorder, and its usefulness in predicting syrinx formation. Because the presyrinx state is a potentially reversible myelopathy, it will be of obvious

interest to neurosurgeons. The goal of treatment, as with syringomyelia, should be to identify and correct the underlying CSF obstruction.

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