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Cord Cystic Cavities: Syringomyelia and Prominent Central Canal

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Syringomyelia is the term given to cystic cavities in the spinal cord, most of which are associated with congenital malformations of the craniocervical junction and represent dilation of the central canal of the cord. As such, syrinxes can be considered analogous to hydrocephalus. The exact etiology of syrinx formation remains a subject of debate, but there is ample evidence that they are the result of obstruction of the normal flow of cerebrospinal fluid between the intracranial and spinal compartments. The chances that a syrinx will progress over time are much greater when they are associated with a causative lesion (Chiari malformation, tumor, infection, and trauma), but asymptomatic central canal dilation may be a stable incidental finding. Although spinal cord neoplasms are a recognized etiology for syrinx formation, especially in adults, it is not always necessary to administer contrast when evaluating a syrinx for the first time with magnetic resonance imaging.
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Introduction

The term syringomyelia was first used to describe cystic cavities in the spinal cord in the late 19th century, although the lesion itself had been described in medical texts well before that time.[1,2] The name itself refers to the nymph of Greek mythology who was turned into hollow reeds in response to prayer; this is the origin for the term syringe as well. Some have suggested that syringomyelia be reserved for those cavities that are not lined by normal ependyma, with hydromyelia used for cavities that represent dilation of the central canal without the loss of ependymal cells. In a practical sense, this distinction is of little significance, as both variations can have a similar clinical and imaging appearance, and progression of central canal dilation can lead to the loss of ependymal lining.

A more substantial debate centers around the absolute or relative size that a dilated central canal must reach for it to be termed a syrinx. This was not an issue before the magnetic resonance (MR) imaging era, when the diagnosis was typically made in more severe and symptomatic cases. But with the development of high-resolution MR imaging of the spine, the range of the size of the "normal" central canal of the cord in vivo became apparent and the borderline between normal and pathologic became less obvious. A review of the anatomic, physiologic, and pathologic factors leading to enlargement of the central canal and development of cystic cavities in the spinal cord should help guide the assessment and management of this lesion.

Anatomical Considerations

The central canal of the spinal cord runs from the obex at the base of the fourth ventricle caudally to its termination in the conus medullaris. It is lined by ependymal cells, just as the ventricles are lined in the cranium. Like the ventricles, it serves as a conduit for cerebrospinal fluid (CSF). CSF has several functions—it provides buoyancy, and thus, some degree of protection to the brain and the spinal cord; it has a humoral function, transporting neurotransmitter and hormonal signals from one region of the central nervous system to another; and probably most importantly, it serves as the lymphatic system for the central nervous system, allowing for the regulation of interstitial fluid.[3] The flow of CSF throughout the ventricular system has been studied extensively, but the flow within the central canal of the cord is much less well understood. It is recognized that CSF must exit the ventricular system and circulate in the subarachnoid space to be resorbed, either through the cribiform plate or through arachnoid granulations actively transporting it into the dural sinuses.[3] Some have theorized that CSF is driven into the central canal through the

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obex by arterial pulsation; however, it is teleologically more sound to conclude that the central canal serves to gather interstitial fluid from the spinal cord and transmit it rostrally so that it can enter the subarachnoid space and be eventually resorbed, just as CSF exits the ventricular system in the brain. This rostral direction of CSF flow has been confirmed in animal models.[4]

Unlike the ventricles, which have 3 outlets for CSF to egress from the fourth ventricle, the central canal is an anatomical cul-de-sac, with the obex serving as the only macroscopic site of communication with the subarachnoid space. It is thus logical that lesions obstructing the central canal would result in dilation caudal to the level of obstruction; however, not all such obstructing lesions result in a more caudal syrinx cavity. Indeed, anatomical studies have demonstrated points of apparent obstruction in the central canal in a high percentage of autopsied adults, without associated syringomyelia.[5] This may be because there are microscopic channels that run from the subarachnoid space in the spinal canal to the central canal of the cord, similar to perivascular spaces in the brain.[6,7]Although the cause of the points of obstruction within the central canal is unknown, they may serve a protective function in limiting the cranial-caudal extension of syrinx cavities by forcing CSF out of the small collateral channels into the subarachnoid space. Thus, for obstruction of CSF flow in the central canal to cause syrinx formation, there must also either be insufficiency of these collateral channels or an additional obstruction to the CSF flow in the subarachnoid space (Fig. 1). Cosan et al[8] were able to demonstrate this pathophysiology in rats.

Just as all CSF-filled cavities in the brain are not due to dilation of the ventricular system, all cavities in the spinal cord are not a result of dilation of the central canal. Areas of injury to the cord can result in cavity formation, and these cavities can increase in size with time as interstitial fluid and CSF collect in them. Neoplasms may contain cystic foci filled with simple or complex fluid, and abscesses can form within the cord just as they do elsewhere. These traumatic, neoplastic, or inflammatory cavitary lesions may extend into or obstruct the central canal, resulting in cavities that are in part dilated ependyma-lined canal and in part cystic myelomalacia.

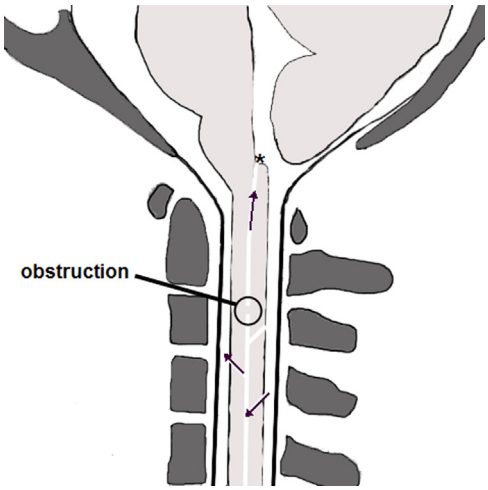

Figure 1 Schematic of the cranial-cervical junction and cervical cord showing key anatomical features contributing to syrinx development. The obex (*) is the only large channel for egress of CSF from the central canal of the cord, but multiple small channels (arrows) also connect the canal to the subarachnoid space along the length of the cord. Note the obstruction of the central canal, a nearly ubiquitous finding in the spinal cord that increases in frequency with age. (Color version of figure is available online.)

Conditions Associated With Syringomyelia

Congenital Malformations

Congenital malformations are the most common category of pathology associated with syringomyelia today. Syrinxes are the most significant complications associated with the Chiari 1 malformation but are also seen in the Chiari 2 malformation, Dandy-Walker malformation, and aqueductal stensosis, among others (Fig. 2).

Reviews of reports in the medical literature suggest an incidence of syringomyelia with the Chiari 1 malformation of more than 60%,[9] but there are clear biases toward complexity when reviewing only published cases. A review of spinal imaging in a large population of children in California found that 12% of the children diagnosed with Chiari 1 had a syrinx at presentation, and no new syrinxes developed over the course of the study.[10] This latter incidence is much more in keeping with our experience, although we have seen syrinxes develop over time in children with the Chiari 1 malformation (Fig. 3). The malformation results in varying degrees of obstruction of the obex and of the subarachnoid space at the craniocervical junction, thus satisfying the 2 anatomical criteria proposed earlier as necessary for syrinx development. The pistoning motion of the cerebellar tonsils with arterial pulsations results in repetitive pressure waves in the spinal subarachnoid space if the degree of obstruction caused by the malformation is sufficiently severe. This combination of factors serves to drive CSF into the central canal and trap it there. Once some degree of canal dilation results, the pressure on the walls of the dilated segment is accentuated, leading to a vicious cycle of syrinx enlargement.

Similar pathophysiology likely leads to the development of syrinxes in the Chiari 2 malformation. Several features of the Chiari 2 malformation might increase the likelihood of developing a syrinx, including the tethering of the cord at the site of myelomeningocele, hydrocephalus, and abnormalities of dural development or neuronal differentiation. Although one could argue that the open neural tube defect represents a de facto hydromyelia in all cases of myelomeningocele, the actual incidence of syringomyelia in children with the Chiari 2 malformation has not been well studied. Caldarelli et al[11] found syrinx cavities in 32 of 142 children (22.5%) with the Chiari 2 malformation in whom they reviewed spinal MR imaging. Our experience suggests a similar prevalence, with some cavities developing or enlarging over time.