Idiopathic syringomyelia

Syringomyelia not associated with any conditions.

With no overt etiology, surgical decision making and treatment can be challenging.

Pathophysiology

The underlying pathophysiology leading to syringomyelia is elusive with multiple flow-related theories constituting our current limited understanding of the disease process. Syringomyelia is associated with pathologies related to the disturbance of cerebral spinal fluid flow found in conditions such as Chiari I malformations, spinal malignancy, spinal cord tethering, trauma, or arachnoid adhesions.

Most hypotheses trying to explain the pathophysiology of idiopathic syringomyelia involve mechanisms whereby CSF is pumped against a pressure gradient, from the subarachnoid space into the cord parenchyma. On review, these theories have universally failed to explain the disease process. A few papers have suggested that the syrinx fluid may originate from the cord capillary bed itself. However, in these papers, the fluid is said to accumulate due to impaired fluid drainage out of the cord. Again, there is little evidence to substantiate this. This proffered hypothesis looks at the problem from the perspective that syringomyelia and normal pressure hydrocephalus are almost identical in their manifestations but only differ in their site of effect within the neuraxis. It is suggested that the primary trigger for syringomyelia is a reduction in the compliance of the veins draining the spinal cord. This reduces the efficiency of the pulse wave dampening, occurring within the cord parenchyma, increasing arteriolar and capillary pulse pressure. The increased capillary pulse pressure opens the blood-spinal cord barrier due to a direct effect upon the wall integrity and interstitial fluid accumulates due to an increased secretion rate. An increase in arteriolar pulse pressure increases the kinetic energy within the cord parenchyma and this disrupts the cytoarchitecture allowing the fluid to accumulate into small cystic regions in the cord. With time the cystic regions coalesce to form one large cavity which continues to increase in size due to the ongoing interstitial fluid secretion and the hyperdynamic cord vasculature [1)].

Diagnosis

Disorders of CSF dynamics such as syringomyelia and obstructive hydrocephalus can be caused by thin mobile obstructive lesions not visible on traditional MRI sequences. New imaging techniques with balanced steady-state free precession (bSSFP) and dynamic imaging with bSSFP cine allow visualization of these pulsatile structures within the CSF space. The authors present 2 cases involving pediatric patients-one who developed presumed idiopathic syringomyelia and one with presumed communicating hydrocephalus in association with Pfeiffer syndrome-who harbored thin dynamic obstructive lesions seen on bSSFP cine studies using 1.5-T MRI. In combination with traditional CSF cine studies and bSSFP, bSSFP cine sequence was able to detect dynamic membranous adhesions not seen on traditional MRI sequences. These previously undetectable lesions on traditional MRI sequences were the etiology of CSF obstruction, and tailored surgical approaches were performed to avoid shunting in both patients. These reports demonstrate the clinical utility for using these novel imaging tools for the detection of thin adhesions and dynamic lesions in the central nervous system. Balanced SSFP cine sequences can supplement conventional MR modalities to identify these otherwise poorly visualized lesions responsible for presumed communicating hydrocephalus or

idiopathic syringomyelia [2].

Treatment

Shunting procedures for the syringomyelia disease spectrum have been criticized due to the inconsistent long-term outcomes.

This is largely the result of small volume flow at a very low-pressure profile leading to occlusion or malfunction of the shunts.

Syringoperitoneal shunt.

Syringosubarachnoid shunt.

Syringopleural shunt

Iwatsuki et al., describe a new modified surgical approach to minimize the postoperative recurrence of a syrinx after surgery to treat syringomyelia associated with spinal adhesive arachnoiditis in two cases. Both patients presented with progressive gait disturbance without any remarkable history, and spinal magnetic resonance imaging revealed a syrinx and broad irregular disappearance of the subarachnoid space and/or deformity of the cord. They successfully performed a partial arachnoid dissection and syrinx-far distal subarachnoid shunt for both cases. [3].

Case series

Five patients, aged 22-50, presented with progressive neurologic symptoms associated with an idiopathic syrinx. All underwent decompressive laminectomy surgery with syringosubarachnoid shunting using the silastic wedge technique.

In five cases of idiopathic syringomyelia, clinical and radiographic follow up ranges from 3 to 36 months. Three patients have radiographic and clinical follow up greater than 24 months. All patients improved clinically and their symptoms have been stable [4].

1)
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4)
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