Holocord syringomyelia

Holocord syringomyelia, in which the syrinx involves most or all levels of the cord (typically from C2 to the lower thoracic or lumbar vertebrae, and often all the way to the conus medullaris) represents a unique subset of syringomyelia cases.

Although the central nervous system has a limited capacity for regeneration after acute brain and spinal cord injuries, it can reveal extensive morphological changes. Occasionally the formation of an extensive syrinx in the spinal cord can be observed causing no or only limited signs of functional impairment. This creates a unique condition to evaluate the mismatch between substantial morphological changes and functional outcomes.

Case series

2014

7 patients with holocord syringomyelia affecting the cervical cord following chronic traumatic thoracic/lumbar spinal cord injury (19 - 34 years after injury) or of non-traumatic origin were identified and anatomical syrinx dimensions (length, cross-sectional area) were determined using sagittal and axial MRI scans. Motor- and sensory-pathway integrity were evaluated using electrophysiological assessments (motor (MEP), sensory (dSSEP) and contact-heat (dCHEP) evoked potentials; nerve conduction studies (NCS)) and specifically compared to clinical measures of upper limb strength and grasping performance including 3-D motion analysis. Despite extensive anatomical changes of the cervical cord (on average 26% reduction of residual spinal cord area and intrusion of almost the entire cervical spinal cord) a clinically relevant impairment of upper limb motor function was absent while only subtle sensory deficits could be detected. dCHEPs revealed the highest sensitivity by disclosing impairments of spinothalamic pathways. Comparable to the brain extensive anatomical changes of the spinal cord can occur with only subtle functional impairment. Obviously, the time scale of very slowly emerging morphological alterations is essential to permit an enormous capacity for plasticity of the spinal cord [1)].

Case reports

A holocord syringomyelia due to Chiari Malformation Type 1.5 in a 12-year-old girl was serially imaged with 3 Tesla MRI over 4 years. The serial MRI showed a reduction in the size of the syrinx, without any surgical intervention or CM improvement, but rather due to spontaneous spinal cord tear. The tear was clearly demonstrated by evidence of flow signal across the tear between syrinx and subarachnoid space at the upper thoracic level. The tear showed spontaneous closure at follow-up. A medullary tear has been described in the adult population as one of the putative causes of spontaneous syringomyelia reduction, but its clear demonstration with modern high-resolution MRI has not been reported in the pediatric population. Moreover, this is the first reported case of syrinx reduction due to spontaneous fissuration in a pediatric patient [2)].

An 11-year-old male patient presented with progressive left hemiparesis and numbness on left half of the body for 4 years. Magnetic resonance imaging of the spine revealed peg-shaped herniation of tonsils 8 mm below the foramen magnum and holocord syringomyelia. No focal intraspinal mass was seen. Chiari I malformation with holocord syrinx was diagnosed. The patient underwent posterior fossa decompression with subpial resection of both tonsils with augmentation duraplasty. Post-operatively, patient improved clinically as well as radiologically [3].

1)
Awai L, Curt A. Preserved sensory-motor function despite large-scale morphological alterations in a series of patients with holocord syringomyelia. J Neurotrauma. 2014 Sep 15. [Epub ahead of print] PubMed PMID: 25219978.
2)
Staglianó S, Prodi E, Goeggel Simonetti B, Cianfoni A. Syrinx reduction due to spontaneous spinal cord tear: demonstration on 3 T MRI and review of the literature. BMJ Case Rep. 2021 Dec 21;14(12):e246235. doi: 10.1136/bcr-2021-246235. PMID: 34933894.
3)
Bansal S, Borkar SA, Mahapatra AK. A common case with an unusual association: Chiari I malformation with holocord syrinx. Asian J Neurosurg. 2017 Apr-Jun;12(2):241-243. doi: 10.4103/1793-5482.144169. PubMed PMID: 28484540; PubMed Central PMCID: PMC5409376.