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SPONTANEOUS RESOLUTION OF AN EXTENSIVE POSTTRAUMATIC SYRINGOMYELIA

The prevalence of symptomatic syringomyelia in patients with chronic spinal cord injury (SCI) is around 4.5%. It is an important cause of treatable neurologic deterioration, although surgical shunting of the syrinx or arachnoid adhesiolysis of tethered elements have unpredictable results. Classically, syrinx extension causes progressive neuropathic pain, often with dissociated sensory loss due to compression of the more central, spinothalamic projection neurons with relative preservation of dorsal column function. We report an unusual case of the total spontaneous resolution of an extensive cervicothoracic syrinx cavity in a patient with symptomatic posttraumatic syringomyelia (PTS) and describe the changes in contact heat-evoked potentials (CHEPs) associated with its development and disappearance.

Case report. A 32-year-old man with a chronic incomplete SCI (AIS C, sub T9) resulting from a motorcycle accident presented 9 years after the initial trauma with acute-onset neuropathic pain in the right shoulder blade and upper limb following an episode of heavy coughing. Cardiovascularly fit, he was a keen wheelchair sportsman. Extensive dissociated sensory loss was documented in dermatomes C6 to T10 on the right with impaired pinprick sensation but preserved light touch (figure 1A). Whole-spine MRI was in keeping with a mature thoracic syrinx cavity, with less well-delineated cord signal change extending cranially to segments C3/4 and a preexisting, posttraumatic cyst at T11 (figure 1B). Neurophysiologic assessment demonstrated impaired CHEPs at C6 and C8 bilaterally (figure 1C) with preserved dermatomal somatosensory evoked potentials. Inflammatory and ischemic etiologies were considered but deemed highly unlikely due to the relatively mild symptoms in the context of such extensive cord involvement. After 2 months of watchful waiting and with no improvement in symptoms, whole spine MRI was repeated. The syrinx cavity had consolidated in the cervical cord and CHEPs were abolished at C6 and C8 bilaterally. The patient was booked for elective surgical adhesiolysis. However, the evening before the procedure, he reported rapid symptomatic improvement over the course of several days. On MRI follow-up at 6 months, the cervicothoracic syrinx had completely resolved and CHEPs had improved, while the rostrocaudal dimension of the posttraumatic cyst had increased from 30 to 37 mm. The neuropathic pain was significantly improved and only subtle abnormalities of protopathic sensation remained in the right arm. Findings remained stable at 9 months (figure e-1 at Neurology.org).

Discussion. In the most widely accepted theory of PTS, the antecedent cause of syrinx formation is subarachnoid block due to arachnoid adhesions at or near the level of prior injury. Patients with PTS have reduced thecal compliance and higher pressures in the subarachnoid CSF at cardiac systole drives CSF into the cord through perivascular spaces. In vitro simulations of subarachnoid stenosis in PTS suggest that caudocranial syrinx extension results from ballooning of the rostral cavity during episodes of increased intracompartmental pressure, as occur on coughing. Such expansion likely proceeds relatively atraumatically along tissue planes and may account for the often subtle symptomology encountered, even in anatomically extensive PTS. The sudden and complete radiologic resolution of a syrinx, accompanied by subtotal clinical recovery, is almost without precedent. Interestingly, the only similar report also featured a posttraumatic cyst and the authors suggested that resolution resulted from a rupture of the syrinx cavity into the subarachnoid space. A similar rupture into the cyst may account for the observed increase in its size in this case. Our patient was involved in regular, strenuous sports, which he had to curtail as a result of syrinx-related pain. Perhaps a degree of subarachnoid block not quite sufficient for syrinx formation in a sedentary individual may result in PTS in the context of heavy exercise. As such, it may be reversible when exercise is reduced, although with a high risk of recurrence.

CHEPs are performed using a contact-thermodoe stimulator on a dermatome to elicit EEG responses. The thermal stimulus specifically activates Aδ nociceptors in the periphery and is propagated through the spinothalamic tract, which is specifically vulnerable in the case of an axially expanding, central syrinx, where horizontally crossing commissural fibers are more susceptible to damage than the longitudinal tracts of the dorsal columns.
Figure 1  Clinical, neuroimaging, and electrophysiologic findings in a spontaneously resolving, posttraumatic syrinx

(A) American Spinal Injury Association (ASIA) Impairment Scale (AIS) at 4 timepoints (clinical baseline 2 years postinjury, at index presentation, and 2 and 8 months later) in a 32-year-old patient with a chronic, incomplete (AIS C, neurologic level sub T9) spinal cord injury who presented with a symptomatic posttraumatic syrinx. (B) T2 MRIs of the same patient are presented from left to right from index presentation and 2 and 8 months later. No previous MRI was available. Between
noncommunicating with respect to the central canal, the path of expansion may meander somewhat eccentrically within the cord and this likely accounts for the sparing of spinothalamic function at T4 and T7 seen in this case, where axial images reveal the body of the syrinx to be more dorsally located (insets, figure 1B).

Spontaneous resolution in PTS is rare but possible and seems to be associated with sizeable posttraumatic cystic lesions, which may represent a therapeutic target in selected cases. CHEPs are ideally suited to the assessment of syringomyelia, in which a central syrinx tends to preferentially affect the small crossing fibers in the anterior commissure and may serve as an early marker of disease progression.

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