



Spinal Cord Injury Case No. 1, March 2005

Presenting Symptom(s):

Increased spasticity in a person with chronic spinal cord injury

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Appropriate Audience: Residents and practicing physicians

Learning Objectives: After completing this educational activity, participants will be able to: (1) identify factors that may affect spasticity in persons with chronic spinal cord injury; (2) formulate a differential diagnosis based on history and physical examination for changes in spasticity and neurologic condition in persons with chronic spinal cord injury; (3) identify diagnostic tests and treatment options for post-traumatic syringomyelia.

Level of Difficulty: Basic

History

The patient is a 57 year old man with a complete C7 tetraplegia secondary to diving injury 22 years ago. He presents for a new patient visit with complaints of increased spasticity over the last month. He has not otherwise been ill or had other symptoms. He lives at home with his wife, who is also his primary caregiver and a licensed vocational nurse.

- Prior to continuing, please develop a differential diagnosis and list each possible diagnosis in order of likelihood.
- Is there any additional information regarding the clinical history that might be helpful in clarifying your differential list or changing its order of priority?

Commentary I

When an individual who has had relatively stable spasticity presents with exacerbation of symptoms, potential sources of nociception should be considered and investigated. If present, management and treatment of the source of the noxious stimulation may be sufficient to decrease the spasticity rather than needing to initiate treatment specifically for the spasticity. A variety of secondary medical conditions or complications may exacerbate spasticity, including urinary tract infection, bladder calculi, pressure ulcers, ingrown toenails, hemorrhoids and bowel impaction. Other potential conditions include irritation from urethral catheters, fractures, syringomyelia, menstruation, deep venous thrombosis, appendicitis, cholecystitis, or other abdominal processes.



Additional history should assess any other changes in neurologic status (motor and sensory changes) and functional status which have accompanied the change in spasticity.

History, continued

He has managed his spasticity with 60 mg of baclofen per day, in divided doses, since his injury. He has not increased his dose, but notices more frequent and more severe spontaneous spasms, though his wife does not find him any more difficult to range. She does note that it is much more difficult to transfer and position him.

On review of systems, his bowel program is performed 3 times per week by his wife without problems. It has been going well for years without incontinences or more than an occasional bout of constipation, which they are able to resolve at home. He has had hemorrhoids for years, but there has been no change in them. He manages his bladder with a suprapubic catheter changed monthly. He has about 2 urinary tract infections per year, none requiring IV antibiotics. They have not noticed any change in sedimentation in his urine, nor has he passed any stones. He has not fallen recently, nor remembers any trauma. His wife checks his skin daily and has not noticed any areas of concern. He has not noticed any change in sensation, but he has begun to have difficulty wheeling, transferring and even lifting objects. He thinks his spasticity is the primary problem though. He has mild neuropathic pain, primarily below his level of lesion and only occasional neck pain with a headache. He does not take medications for these pain issues.

- If necessary, revise your differential diagnosis based on the additional clinical history.
- On which details of the physical examination should you focus at this point?

Commentary II

Although the history obtained from the patient and family appear to eliminate the more common potential secondary conditions or complications that can exacerbate spasticity, a thorough physical examination should be performed to rule out the presence of any of these conditions. A thorough and complete neurological examination is especially important in this situation given the lack of a clearly identifiable source of noxious stimulation that is exacerbating the patient's spasticity.

Physical Examination

The patient presents as a well-developed, well-nourished pleasant male who appears his stated age.

HEENT - scaling erythema over the eyebrow area and nasolabial folds

Heart and Lung - normal and chest expansion is as expected for his level

Abdomen - protuberant, but soft with normal bowel sounds; careful palpation of each quadrant does not provoke spasms or discomfort

Extremities - 3+ pitting, dependent edema in the lower extremities without erythema; no deformity or focal edema; upper extremities without edema or deformity other than expected atrophy

Skin - no breakdown and all red areas blanchable

- At this point, review your differential diagnosis and revise as appropriate.



Commentary III

The general examination appears to rule out the presence of the more common secondary conditions which frequently can exacerbate spasticity. There does not appear to be evidence of active pressure sores, hemorrhoids, bowel impaction or other acute abdominal process.

- *Are there additional observations on physical examination that might be helpful in narrowing your differential list?*

Physical Examination, continued

An additional part of the general examination that should not be overlooked is examination of the toenails as frequently, they can be a source of irritation that can exacerbate spasticity.

Also at this point, a complete and thorough neurological examination should be performed to determine if there has been any objective change in the patient's motor and sensory levels that might indicate a process within the central nervous system that is exacerbating his spasticity.

Toenails are thickened, consistent with dystrophic changes, but without areas of erythema, hypergranulation or purulence. Anterior/posterior squeezing of the toes does not provoke spasms.

Neurologic examination reveals the last normal sensory level to be C5 with 3 levels of partial preservation. Strength is 5/5 in biceps and 4+/5 in the wrist extensors and triceps bilaterally. Strength is otherwise absent below. Tone is 1+/4 in the lowers in the absence of a spasm, but 4/4 during a spasm. Spasms are noted with any superficial stimulation, and sometimes unprovoked. Reflexes are 2+ at the biceps and symmetric. During strength testing, the patient is surprised by his relative weakness. He reports that he previously had a trace of deep finger flexion on the right, which is now not elicitable.

- If necessary, revise your differential diagnosis based on the additional physical findings.

Commentary IV

Examination of the patient's toenails appears to eliminate ingrown toenails as the source of his increased spasticity. On neurological examination, however, the apparent decrease in motor strength raises concerns regarding changes within the spinal cord that might account for the increase in spasticity.

- *What diagnostic tests would you order at this time?*

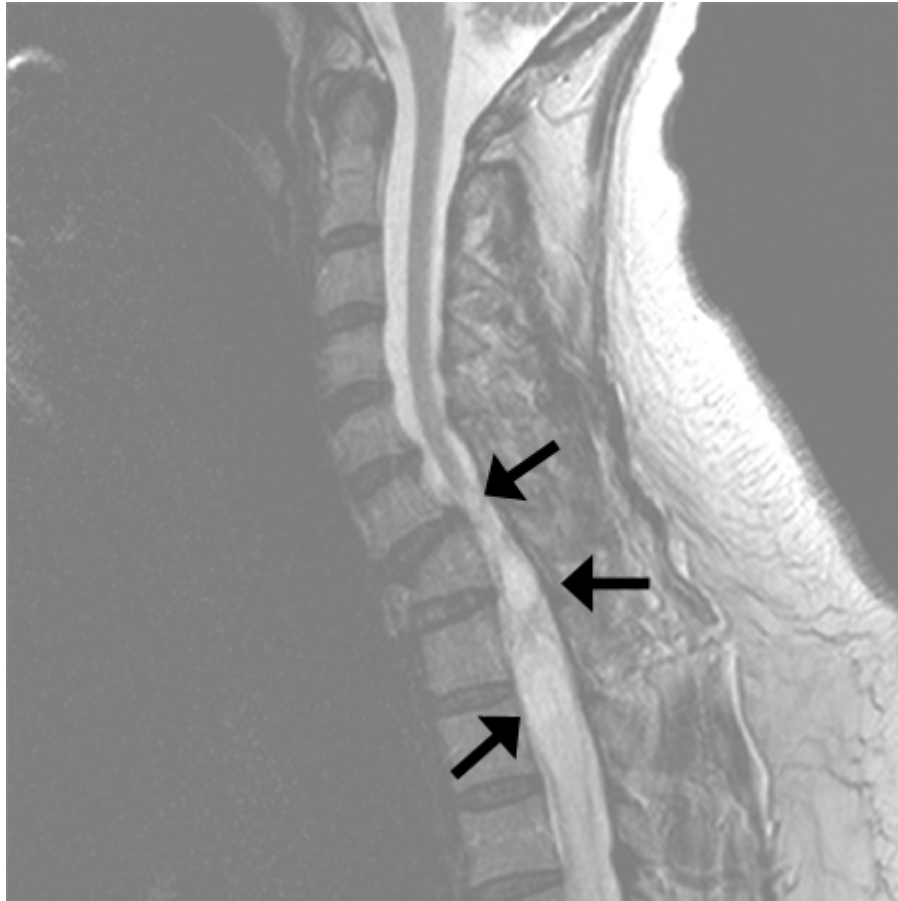
Commentary V

Given the clinical history and findings on physical and neurological examination, the possibility of pathologic changes within the spinal cord must be considered. A magnetic resonance imaging (MRI) scan of the cervical spinal cord may delineate any pathologic changes.

Test Results

- If necessary, augment or change the clinical impression based on the test results.

Figure 1. T2-weighted sagittal image revealing extensive cystic myelomalacia (arrows) within the lower portion of the cervical spinal cord



What additional diagnostic testing would you order?

CT myelogram, requested by the surgeon, revealed complete myelographic block at T3 from below and C6 from above.

Commentary VI

Magnetic resonance imaging serves as the gold standard for diagnosis of syringomyelia. T1 and T2-weighted spin echo sequences in both sagittal and axial planes are the preferred views for delineating the extent of the syrinx. Frequently, increased signal intensity from gliosis can be appreciated in the areas surrounding the syrinx on T1 and T2 weighted images. T2-weighted images can provide information on intrinsic cord disease and demyelination, both of which should be ruled out in making the diagnosis of syringomyelia. MRI CSF flow studies can also be very useful in determining the degree and extent of CSF block, delineating the mechanistic pathology.

In patients unable to undergo MRI evaluation, CT-myelogram is an invasive but informative diagnostic test. This modality can evaluate CSF flow, as well as length and breadth of cord expansion.



Commentary VII

Although useful in assessing patients with seemingly multiple potential causes of neurologic decline, electrophysiologic testing has limited value in both diagnosis and progression of syringomyelia. Nogues et al. have described increased spontaneous activity including fibrillations, positive sharp waves, neurogenic motor unit potentials, myokymic discharges, and synkinetic motor unit potentials with inspiration (respiratory synkinesis). Each of these is nonspecific so clinical correlation is necessary. Little and coauthors believe motor evoked potentials, calculated from the spine to the scalp, may provide additional diagnostic information. In general, electrodiagnostic testing provides its greatest utility in ruling out other entities such as radiculopathy or myelopathy.

- Considering all the data from the history, physical examination and laboratory studies, what is/are your final diagnostic impression(s)?

Final Diagnostic Impression

Post-traumatic syringomyelia with tethered spinal cord.

Commentary VIII

In several reports, pain and ascending sensory neurologic level were the most common complaints in persons who developed syringomyelia. According to the mechanism, early syrinx formation causes an increase in upper motor neuron signs and symptoms including increased spasticity and neuropathic pain, but can progress to lower motor neuron signs and symptoms including hyporeflexic bladder and loss of limb reflexes. Autonomic changes such as autonomic dysreflexia and hyperhidrosis can also occur. The clinical appearance is highly variable and can present as early as 2 months post injury or as distant as 34 years. One report observed onset of symptoms 39 months after initial spinal cord injury in complete (ASIA A) subjects and 101 months after injury in incomplete patients, but El Masri's series found no particular pattern between completeness of lesion and onset of syringomyelia

- What treatment would you now initiate for this patient?

Patient underwent duraplasty with lysis of adhesions. The syrinx was drained with a fine needle, but no shunt was placed. He was extubated within 24 hours easily. In the early post-operative period he had no spasticity, with a gradual recurrence over the next month. At 6 week follow up he felt well and had regained his prior biceps and wrist extension strength. He could also demonstrate trace triceps activity on the right. At 24 months post-procedure, he was well, at his baseline neurologic level with easily controllable spasticity at his baseline level of baclofen.

Commentary IX

Shunting of syringomyelic cysts may be attempted as a means of reducing the pressure within the fluid filled cavity. The transfer of cyst contents to the subarachnoid space yields satisfactory initial outcomes. However, failure of the shunt in the early postoperative period occurs with some frequency and is attributed to scarring of the subarachnoid space typical in patients with posttraumatic or postinflammatory cysts. Intraoperative sonography can be used to identify cystic cavities for selection of myelotomy site or shunt placement. Even with precise localization, additional complications in subarachnoid shunts arise, including collapse



of the cyst around the opening of a functioning shunt, glial ingrowth of shunt openings and cyst compartmentalization. These challenges have led to the advent of extrathecal approaches, specifically syringoperitoneal and syringopleural diversions. Yet in one series by several authors, a 50% complication rate was found among extrathecal shunts, with obstruction being the most common reason for failure. Other causes included spinal cord tethering, infection, low CSF pressure, and proximal dislocation.

In an attempt to reduce failure rates, Lee and colleagues have developed a 3-tiered protocol for treatment of those with syringomyelia. Among the most important considerations is whether or not spinal cord tethering has occurred, as this situation requires an initial untethering and lysis of adhesions prior to shunt placement. The group of subjects undergoing initial cord release was then subdivided according to whether or not the cyst persisted postoperatively. In all groups of patients, ultrasound imaging can be used to characterize the size and location of the cyst preoperatively, as well as confirm satisfactory post surgical decompression. Duraplasty was performed as the final step in all 3 groups. Results of Lee et al. revealed favorable outcomes in 73% of patients, with 20% remaining unchanged, but 7% became worse due to reformation of the cyst. The patients who developed recurrent cysts tended to demonstrate ventral tethering and long segments of tethered cord. Surgeons are now examining the value of duraplasty and lysis of adhesions as the primary management of post-traumatic syringomyelia, and are exploring the development of biophysical barriers that can be inserted between the cord and meninges to prevent retethering.

Final Discussion

Syringomyelia is defined as an expanding cystic cavity in the spinal cord. Although syringomyelia may occur secondary to hindbrain herniation, spinal cord tumor, or fibrosis following spinal meningitis, posttraumatic syringomyelia following spinal cord injury remains one of the more common causes of the disorder.

The pathology of post-traumatic syringomyelia begins with the development of cystic myelomalacia. The latter arises from direct spinal cord contusion, leading to inflammation and blood effusion. Subsequent liquefaction of a central area of the spinal cord occurs and, in a proportion of cases, results in a significant cavity. Syrinx formation transpires when CSF eventually gains access to the center of the cord. This step results from adhesions in the subarachnoid space or other causes of obstruction of CSF flow, such as narrowing of the spinal canal from deformity, osteoarthritis or disc herniation. The precise mechanism of how the fluid gains entry is not known. Ball and Dayan have proposed that the fluid tracks along perivascular spaces to gain entry to the cord; during transient elevations in intrathoracic or intraabdominal pressure, fluid can be forced upward and subsequently run down in a caudal direction, thereby continuing to propagate the size of the cavity and potentially expand further. Continued pressure within the cavity is necessary to maintain the size of the syrinx.

A syringomyelic cavity should be distinguished from that of a primary cyst occurring opposite a vertebral fracture site. Both pressure within the cavity and size of the syrinx are key components of differentiating a primary cyst from a syrinx. Because 51% of MR images can demonstrate primary cysts secondary to trauma, Williams has suggested that a cyst must extend more than two vertebral body lengths from the initial injury site, in a cranial and/or caudal direction to be considered a syrinx. In previous studies, mean lengths of a syrinx have varied from 3.6 to 14 vertebral levels. The diameter of cavities can reach 7 mm or larger. However, newer understanding of the mechanism of the formation of post-



traumatic syringomyelia suggests that any expanded cyst is likely under pressure, and therefore waiting until it reaches 2 vertebral levels is probably unwise.

Role of Physiatry

Few risk factors herald the latter appearance of syringomyelia. Sgouros et al. maintain that there is no association between severity of injury, particular neurologic level, age, or gender in the development of syringomyelia. Given the variable onset of symptoms, there appears to be no time at which the risk of developing a syrinx has passed. Therefore, close monitoring of a patient's pain, spasticity, and sensory level, as well as a heightened awareness of syringomyelia as a late cause of neurologic decline in a previously stable patient, becomes paramount in establishing the diagnosis. Physiatrists are perhaps in the best position to recognize these early warning signs.

Timely surgical intervention can prevent further neurologic progression, an overriding concern in a patient whose baseline function is already significantly compromised. Moreover, referral to a neurosurgeon well versed in a variety of surgical interventions and the need for continued physiatric involvement can provide a patient the best comprehensive approach to treatment of syringomyelia. Through a multidisciplinary approach, physiatrists will continue to serve a major role in returning the patient with syringomyelia to that individual's most favorable functional status.

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