Microcystic spinal cord degeneration causing posttraumatic myelopathy

Report of two cases

ROBERT L. MACDONALD, M.D., J. MAX FINDLAY, M.D., AND CHARLES H. TATOR, M.D., PH.D., F.R.C.S.(C)

Division of Neurosurgery and the Spinal Cord Injury Treatment, Research, and Prevention Centre, Toronto Western Hospital, University of Toronto, Toronto, Ontario, Canada

Two cases of progressive myelopathy occurring years after incomplete cervical spinal cord injury are presented. In both patients, the clinical features, as well as the "bull's-eye" appearance of the delayed computerized tomography (CT) myelography study and the circumscribed low density of the magnetic resonance image, were consistent with posttraumatic syringomyelia, but surgical exploration including intraoperative spinal sonography failed to reveal a syrinx. Although arachnoiditis was present in both patients, the striking abnormality found at surgery was the softened appearance and the microcystic degeneration of the cord. The microcystic spinal cord degeneration found in these cases represents a previously undescribed cause of late deterioration after spinal cord injury that may mimic the clinical, CT-myelographic, and magnetic resonance features of posttraumatic syringomyelia.

KEY WORDS · posttraumatic progressive myelopathy · syringomyelia · spinal cord injury · magnetic resonance imaging

POSTTRAUMATIC progressive myelopathy is an increasingly recognized clinical problem after spinal cord injury, with recent series indicating a prevalence of approximately 3% among spinal cord-injured patients.13 This delayed neurological deterioration after spinal cord injury represents an enormous threat to the already disabled patient, and it is imperative that the clinician recognize any potentially correctable cause. This disorder is usually attributed to posttraumatic syringomyelia,1,5 although other possible causes include arachnoiditis,3,19 spinal column instability with spinal cord compression,10 and spinal cord tethering.14 The correct identification of these various processes (in particular, posttraumatic syringomyelia) has been facilitated in recent years by the introduction of delayed computerized tomography (CT)-myelography and magnetic resonance (MR) imaging which have enabled appropriate therapeutic intervention, such as shunting of the syrinx in posttraumatic syringomyelia. This paper describes two patients with posttraumatic progressive myelopathy whose preoperative clinical, radiological, and MR evaluation was consistent with posttraumatic syringomyelia, but in whom operative exploration revealed arachnoiditis and a striking microcystic myelomalacia. The recognized causes and investigation of posttraumatic progressive myelopathy, as well as this newly described entity which we have termed "posttraumatic microcystic spinal cord degeneration," are discussed.

Case Reports

Case 1

This 29-year-old woman was involved in a motor-vehicle accident 3 years prior to referral, in which she suffered burst fractures of C-4 and C-5, a C3-4 dislocation with unilateral locked facets, and an incomplete spinal cord injury. She was treated with an anterior corpectomy of C-4 with decompression of the spinal cord, and fusion with iliac crest bone graft and metal plates from C-3 to C-5. She remained in a halo thoracic brace for 3 months. She improved neurologically for 1 year after the injury to the point where she was ambulatory but had a spastic right hemiparesis, diminished
Posttraumatic myelopathy

Fig. 1. Case 1. Computerized tomography scan at the C-3 level obtained 24 hours after iohexol myelography showing a hyperdense area (arrow) in the center of the cord.

pinprick sensation bilaterally below T-2, and a right posterior column sensory loss. She was hyperesthetic in the right upper extremity, and bowel and bladder function was clinically normal.

Two years prior to her present admission, the patient began to notice diminishing strength and increased spasticity in her right upper extremity, and severe burning pain in her right shoulder. The dysesthetic pain spread further in her right upper extremity and to both lower extremities, and sensory loss in her limbs increased. She had several episodes of bowel and bladder incontinence.

Examination. Examination on admission revealed wasting of the right deltoid, biceps, and triceps muscles, and a severe right lower-extremity spastic paresis. The left side had virtually normal muscle strength and tone. Sensory examination showed that pain and temperature sensation was decreased below C-4 on the right side, completely absent in the C-5 dermatome, and decreased below T-3 on the left. Touch was mildly impaired from T-3 down on the left, and proprioception and vibration sense were mildly impaired on the right.

The neurological deterioration and pain suggested posttraumatic syringomyelia. Plain radiographs showed evidence of a solid C3–5 fusion with a metal plate present. Cervical myelography with contrast material introduced through a lumbar puncture showed a patent subarachnoid space, but the cord was noted to be irregular from C-3 to C-5. Computerized tomography scanning confirmed an irregular cord outline at C4–5, and the CT study obtained 24 hours after myelography demonstrated a well-defined lesion in the cord at the C-3 level (Fig. 1). Magnetic resonance imaging was performed with a 0.15-tesla resistant magnet unit and an 8-in. surface coil with spin-echo pulse sequences. The echo time was 30 mseconds, the repetition time 1030 mseconds, and the slice thickness 1 cm. Sagittal images were interpreted as showing a syrinx extending from C-3 to C-5 on the right side of the spinal cord (Fig. 2).

Operation. Operative exploration with somatosensory evoked potential (SEP) monitoring was performed through a C4–5 laminectomy. The spinal cord appeared atrophied, and there was dense arachnoiditis over the right side. The right side of the cord was very abnormal in appearance with grayish discoloration and a reduction in overlying vessels. Intraoperative spinal sonography showed abnormal echogenicity in the cord, but failed to reveal an intramedullary cavity. A dorsal midline myelotomy was performed and under microscopy exploration extended anteriorly to the midportion of the cord. No syrinx was found, but the right side of the cord was soft and gelatinous in consistency and appeared to exude fluid from numerous microcystic cavities less than 1-mm in diameter. In the absence of a syrinx, the intended shunt placement was not performed. The myelotomy was approximately 3-mm long and remained open. The patient showed no neurological changes at 6 months postoperatively.

Case 2

This 39-year-old man suffered a burst fracture of C-5 in a diving accident 16 years prior to admission. Initially, he sustained an incomplete spinal cord injury with near quadriplegia, and was treated with a C4–6 decompressive laminectomy and halo thoracic bracing for 3 months. He recovered substantially and 5 years later was walking without leg stiffness and was active in sports with only mild residual impairment of fine motor control in all four limbs. However, during the 4 years...
FIG. 3. Case 2. Spin-echo magnetic resonance image, sagittal projection, showing an expanded cord (arrows) from C-4 to C-6 with central low-intensity signal compatible with posttraumatic syringomyelia.

prior to his present admission, he noted increasing stiffness and spasm in both lower extremities and worsening motor control in his upper extremities. He developed dysesthetic pain in the left interscapular area not aggravated by coughing or straining, and he began to suffer a painless burning sensation in his hands.

Examination. On admission, neurological examination revealed markedly increased tone in both lower extremities, and there was thenar and interosseous muscle wasting. Power was mildly decreased in all extremities in a pyramidal distribution. Reflexes in the upper extremities were normal, but in the lower extremities they were hyperactive with upgoing toes. Sensory examination showed decreased pinprick from C-6 to T-1 bilaterally with normal touch sensation. Position sense was mildly diminished in the toes, and vibration sense was decreased below the shoulder level bilaterally. The delay in neurological deterioration and the presence of a suspended, dissociated sensory loss raised the suspicion of posttraumatic syringomyelia.

Plain radiographs of the cervical spine showed evidence of a C4–6 laminectomy with old compression and deformity of the C-5 vertebral body, a fixed reversal of the normal lordotic curve at C4–5, and posterior displacement of the C-5 vertebral body. Cervical myelography performed after lumbar injection showed a patent subarachnoid space and no apparent arachnoiditis. Immediate and 24-hour delayed CT scanning revealed an irregular cord from C-3 to C-7 with an ill-defined central hyperdensity on the delayed CT scan. Magnetic resonance imaging of the cervical spine performed with the same unit and same parameters as in Case 1 showed an expanded cord from C-4 to C-6 with a central circumscribed low-density area diagnosed as a central syrinx cavity (Fig. 3).

Operation. The patient was operated on through a C-7 laminectomy, and intraoperative SEP monitoring was performed. The dura was opened from C-5 to C-7. The cord was relatively normal in size with arachnoiditis over its dorsal aspect bilaterally; however, the cord and the cord parenchyma appeared abnormal with gray-yellow discoloration. Intraoperative spinal sonography failed to reveal a discrete intramedullary cavity, but showed multiple ill-defined lucencies within the cord. Two dorsal midline myelotomies, each approximately 2 mm long, were made at the C-5 and C-6 levels with the use of the operative microscope. Tiny multiloculated cysts containing clear fluid and with numerous glial or fibrovascular trabeculae were found honeycombing the cord substance bilaterally at each site, but no cavity large enough to accept a shunt tube was found. Postoperatively, the patient awoke neurologically unchanged, and there has been no change in his condition during the 6-month follow-up period.

Discussion

The recognized causes of delayed neurological deterioration after spinal cord injury or “posttraumatic progressive myelopathy,” as it has more recently been termed, include posttraumatic syringomyelia, arachnoiditis, spinal column instability with spinal cord compression, and spinal cord tethering. Late deterioration can be a catastrophic development in an already disabled patient, and may reduce an ambulatory individual to a wheelchair, or render bedridden a paraplegic patient who has been competent in a wheelchair. In addition, the development of a pain syndrome can become equally incapacitating to the spinal cord-injured patient. Thus, it is essential for the physician caring for these patients to identify and correct, if possible, the cause of delayed-onset deterioration or pain.

The most common cause of posttraumatic progressive myelopathy is posttraumatic syringomyelia, with onset months to years after a spinal cord injury. The course is characterized by pain, followed by sensory loss and progressive weakness. Usually, the pain is a dull ache or burning felt in the neck, back, or arms, which is often initiated and then exacerbated by an exertional event or Valsalva maneuver. The sensory loss is usually asymmetric and dissociated. Although neither of our patients had pain that began or was exacerbated with straining, the clinical course was nevertheless consistent with posttraumatic syringomyelia. The radiological diagnosis of posttraumatic syringomyelia has been markedly improved by delayed CT myelography and more recently by MR imaging. With the advent of these techniques, gas and positive-contrast myelography have been employed less often for investigating posttraumatic progressive myelopathy as these methods rely largely upon demonstration of an enlarged or (rarely) a collapsing cord. It has been shown, however, that the size of the cord as determined by CT scanning or surgery often does not correlate with that suggested by myelography. In addition, syrinx cavities that have responded to shunting procedures have
Posttraumatic myelopathy

been detected in normal-sized and even atrophic spinal cords. Thus, these older diagnostic techniques do not accurately indicate cord size or the presence of a syrinx.

Central cord enhancement on CT scanning delayed at least 4 hours after myelography with a water-soluble contrast medium has been estimated to have a sensitivity of 91% and specificity of 87% for detecting syringomyelia of any etiology. Unfortunately, false-negative delayed CT studies for posttraumatic syringomyelia, as well as false-positive studies such as in our patients, have been described in detail in the literature. For example, Quencer, et al., failed to find syringes at surgery in two patients with posttraumatic progressive myelopathy who had exhibited well-defined areas of central cord hyperdensity on delayed CT myelography. In their cases, intraoperative spinal sonography demonstrated areas of abnormal echogenicity similar to those seen in our cases, but they did not open the dura or explore the spinal cord. They suggested but did not confirm that these areas consisted of "scar tissue and multiple vacuoles or microcysts." Our experience proves that not all centrally enhancing lesions on delayed CT myelography represent syringes, and that they may represent microcystic degeneration of the spinal cord.

Although MR imaging is a safer, more accurate, and painless way of detecting spinal cord cavities, this neuroimaging technique can also yield false-negative and false-positive studies for syringomyelia. For example, Pojunas, et al., reported the case of a false-positive diagnosis of syringomyelia with MR studies: the MR image showed a low-intensity lesion in the center of a minimally enlarged spinal cord consistent with syringomyelia, but surgical exploration and biopsy revealed viral myelomalacia. It seems probable, however, that with improved MR technology such as superconducting magnets, surface coils, and paramagnetic contrast agents, this technique will provide more precise preoperative diagnosis in the future.

Posttraumatic arachnoiditis is occasionally implicated as the cause of posttraumatic progressive myelopathy, although it remains a poorly defined entity. A satisfactory explanation for the associated myelopathy (when it occurs) has been lacking. However, it is noteworthy that arachnoiditis was present in both of our patients, and we believe that it may play an important role in the pathogenesis of the posttraumatic microcystic spinal cord degeneration which was found, as discussed below.

Ragnarsson, et al., recently described posttraumatic tethering of the spinal cord as a cause of posttraumatic progressive myelopathy. At surgery, the spinal cord was found to be enveloped in dense circumferential arachnoidal adhesions; after cord transection and release of the adhesions, the spinal cord retracted rostrally almost 1 cm. Marked postoperative clinical improvement was noted. We found no evidence of such tethering in our patients.

Delayed spinal instability with the development of fibrous or osseous extradural compression is another possible cause of posttraumatic progressive myelopathy, and should be assessed by routine radiological investigations including dynamic (flexion-extension) studies, as were performed in our patients. Patients suffering such spinal cord compression would benefit from surgical decompression and/or stabilization.

In our cases, the striking abnormality found at micr...
to be secondary to various mechanical factors such as stretching of the cord during flexion and extension with increases in the intracavitary pressure. In addition, there is the well-recognized association between arachnoiditis and syringomyelia. Thus, there may be a spectrum of posttraumatic spinal cord pathology from arachnoiditis to microcystic degeneration to syringomyelia (Fig. 4). Although this spectrum may not be applicable to all patients with posttraumatic syringomyelia, this is a useful theory, and we are following our two cases closely with MR imaging for any changes in spinal cord morphology.

Conclusions
When delayed neurological deterioration occurs after spinal cord injury, all potentially treatable conditions such as syringomyelia, spinal instability, and the recently described posttraumatic tethering of the spinal cord should be considered. If the imaging studies suggest posttraumatic syringomyelia, surgical exploration is indicated in order to perform a shunting procedure from the syrinx. Because microcystic cord degeneration can mimic posttraumatic syringomyelia, intraoperative spinal cord sonography should be employed; if this study fails to demonstrate a syrinx cavity, our experience would indicate that myelotomy is not warranted. At present, we can suggest no effective treatment for arachnoiditis or its associated microcystic spinal cord degeneration. It is unlikely that the myelotomies that we performed in our two cases will have any therapeutic value since the presence of fluid in the cord is more likely to be the result of the degeneration than its cause.

Acknowledgments
We thank our colleague Dr. Marty Simons of the Department of Radiology for performing and interpreting the intraoperative ultrasonography, and Miss Maria Vespa for preparing the manuscript.

References
Posttraumatic myelopathy


Manuscript received June 17, 1987.

Address reprint requests to: Charles H. Tator, M.D., Suite 4-034, Edith Cavell Wing, Toronto Western Hospital, 399 Bathurst Street, Toronto, Ontario M5T 2S8, Canada.