32 Subacute Progressive Ascending Myelopathy

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INTRODUCTION

Subacute progressive ascending myelopathy (SPAM) is an exceedingly rare and poorly understood complication of spinal cord injury. As the natural history of this process is quite peculiar, it is especially important for the imaging interpreter to be aware of this entity. Following a latent period a few weeks after spinal cord injury, patients usually present with an abrupt ascending neurologic deficit. The sudden onset of symptoms correlates with a characteristic and often dramatic extent of spinal cord edema extending more than four levels above the site of initial spinal cord injury. Remarkably, after a period of several months, near complete resolution of the ascending neurologic deficit and associated edema cephalad to the initial injury site is seen. Only a small focal area of myelomalacia close to the site of initial injury remains after resolution. Of the cases where SPAM reaches the brain stem, loss of vital functions may be fatal prior to resolution. There is no known effective treatment.

TEMPORAL EVOLUTION: OVERVIEW

As depicted in Fig. 32.1, the classic natural history of SPAM includes a latent period, ascending neurologic deterioration within a few weeks, and near complete resolution within a few months.

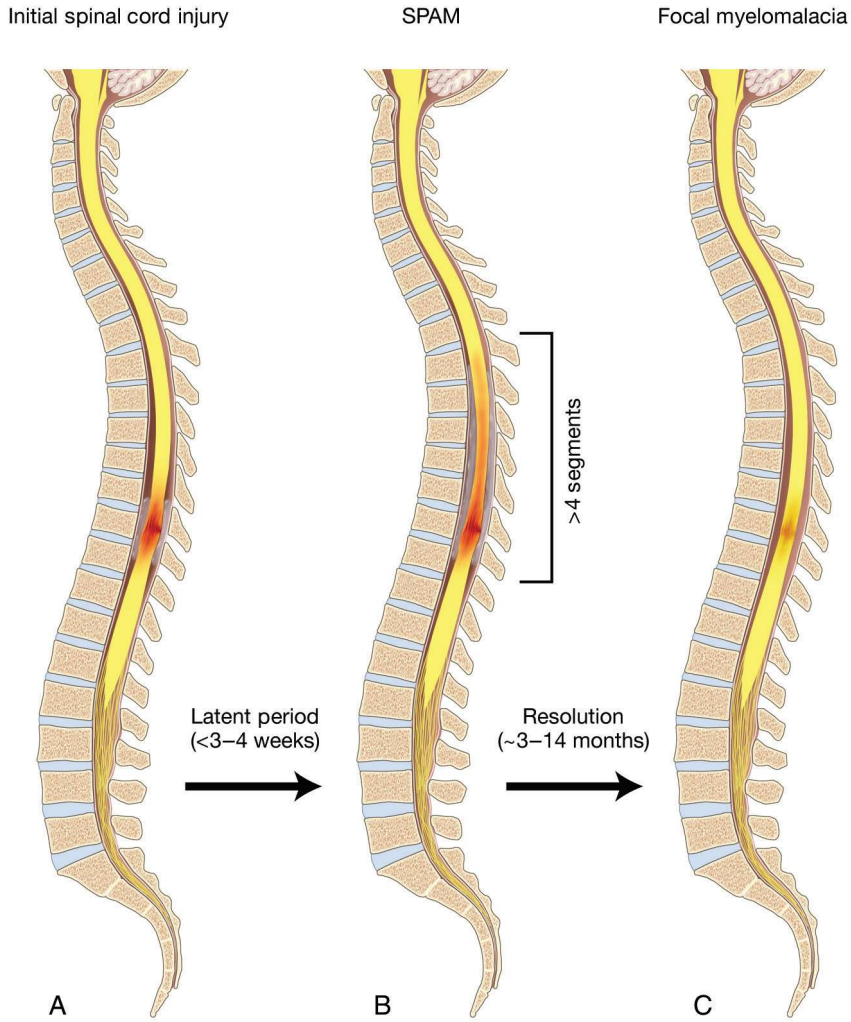

Figure 32.1. Subacute progressive ascending myelopathy *(SPAM)*. A known focal spinal cord insult is present prior to SPAM (A; *red oval*). Following a latent period, ascending neurologic deterioration is most commonly seen in the first 3 to 4 weeks following the initial injury. Imaging at the time of deterioration reveals prominent spinal cord edema extending at least four segments above the level of initial injury (B). These findings may progress over a period of days. Near complete recovery to a level close to but cephalad to the initial injury level usually takes several months. Resolution of edema with only a small focal area of residual myelomalacia extending slightly cephalad to the initial injury is generally evident (C).

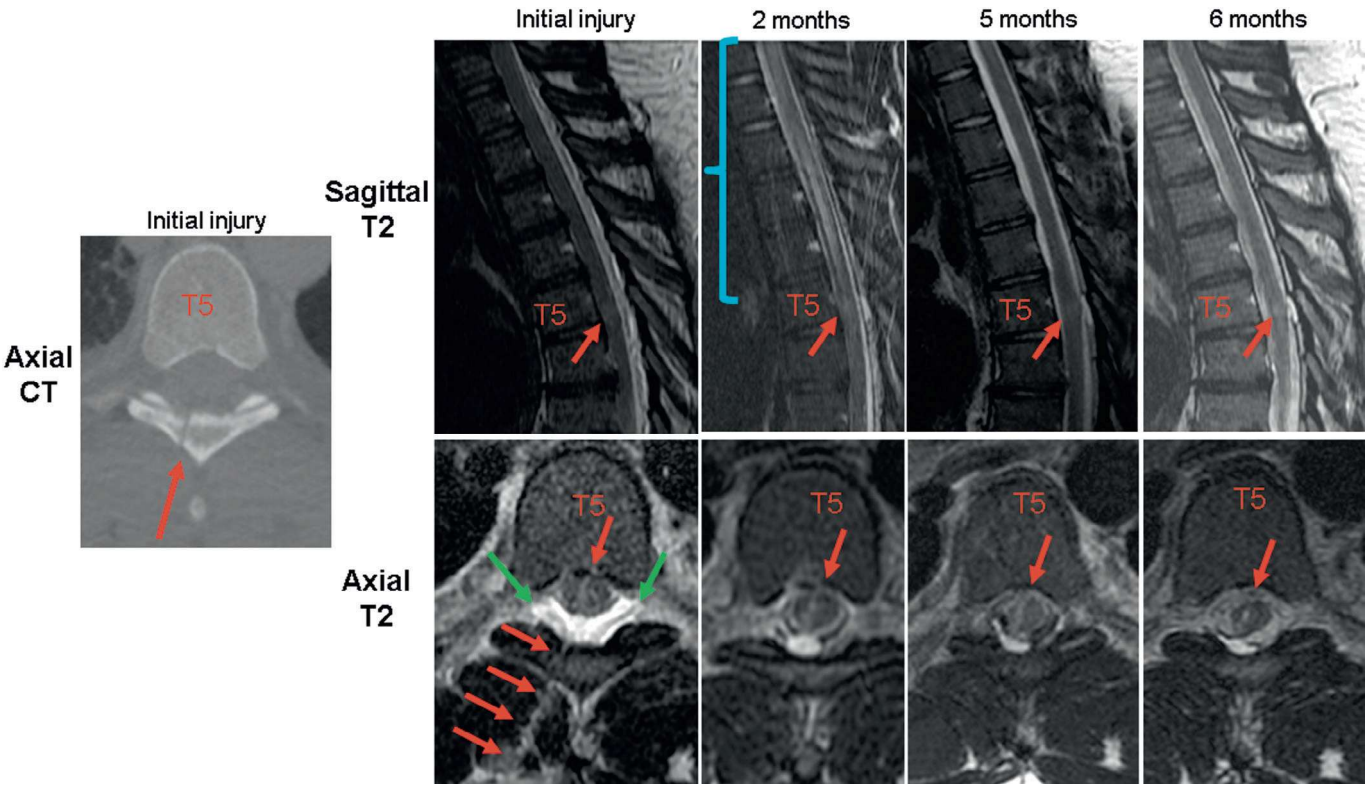

Figure 32.2. Subclinical subacute progressive ascending myelopathy in a young adult male status post T5 stab wound. Axial CT image at the level of T5 at the time of initial spinal cord injury demonstrates a linear stab wound defect along the right T5 lamina *(red arrow)*. Sagittal T2 and axial T2 images at the time of initial injury shows the trajectory of the stab wound (line of *red arrows*), an epidural hematoma *(green arrows)*, as well as focal linear defect through the central spinal cord (anterior *red arrow* on axial image). Two months after initial injury, the patient presented to the emergency room after several weeks of headaches and leg tingling; a repeat MRI demonstrated new spinal cord T2 hyperintensity extending more than 4 levels above the site of initial injury *(blue bracket)*. Five months after initial injury, T2 hyperintense signal above the level of injury has nearly completely resolved. Six months after initial injury, only a small focal area of myelomalacia minimally more prominent and cephalad to the area of initial injury remains *(red arrows)*.

A low-grade fever is present in some but not all patients. It is important to note that ranges of timing and extent of involvement have been reported in the literature. Initial deterioration is most commonly seen in the first 3 to 4 weeks but rarely up to 3 months following the initial injury (Fig. 32.2). All patients demonstrate abnormal nonenhancing T2 hyperintensity, likely representing edema within the central aspect of the cord, extending for at least four segments (and even up to 17 levels) above but not below the level of initial injury. Typical magnetic resonance imaging (MRI) features are mild cord expansion and hyperintense T2 signal with a medullary (central) distribution and a thin residual rim of peripheral cord. Neurologic deterioration and MRI changes can progress over a period of 2 to 12 days. In a small number of subclinical cases with only subtle neurologic deterioration, extensive MRI changes have nevertheless been reported. Near complete recovery to a level close to but cephalad to the initial injury level usually takes 3 to 14 months. Resolution of edema and near complete normalization of the spinal cord cephalad to the injury site is generally evident on imaging. Usually only a small focal area of myelomalacia, including the site of initial injury and a small segment immediately cephalad, can be seen. Although complete recovery does not occur, neurologic deficits are markedly decreased and approximate pre-SPAM levels.

SPAM is a poorly understood delayed complication of spinal cord injury and the etiology remains uncertain. Several hypotheses have been postulated, including secondary injury, venous thrombosis, congestive ischemia, infection, apoptosis, inflammatory or autoimmune reaction, fibrocartilaginous embolism, obstruction of the cerebrospinal fluid (CSF) pathways, and impaired venous drainage of the cord. However, no conclusive evidence is yet available. SPAM is distinct from acute secondary spinal injuries that may be caused by manipulation of an unprotected spinal cord as well as chronic secondary injuries related to syrinx formation or myelomalacia.

Depending on the site of initial spinal cord injury or the extent of superior involvement, the brain stem may be involved. Brain stem involvement is potentially fatal and a mortality of approximately 10% has been reported. As noted previously, there is no known effective treatment. Treatments that have been attempted include decompression, anticoagulation, and both steroid and osmolar therapy without documented benefit. Duroplasty, cordectomy, and cord untethering have been attempted in some cases with some patient-specific beneficial results.

DIFFERENTIAL DIAGNOSIS

Nonneoplastic causes of extensive spinal cord signal abnormality may appear similar to the extent of SPAM spinal cord involvement (Figs. 32.3–32.6). The clinical history, absence of recent focal traumatic spinal cord injury, the presence of enhancement, and/or other imaging findings are typically characteristic of other entities.

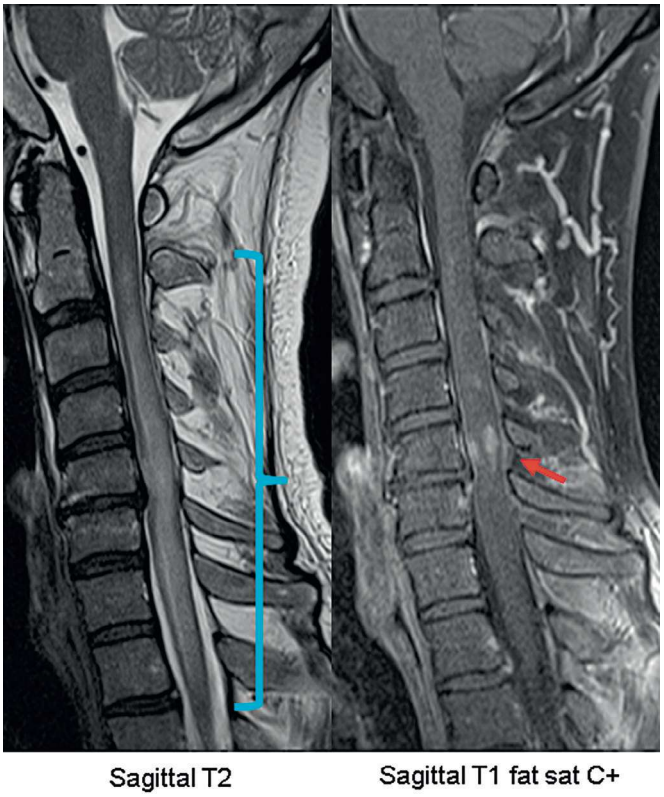

Figure 32.3. Transverse myelitis. Sagittal T2 and T1 fat-saturated postcontrast images in an adult male patient with transverse myelitis. Images demonstrate mild edema and extensive cervical spinal cord T2 hyperintense signal abnormality *(blue bracket)* associated with patchy spinal cord enhancement *(red arrow)*.

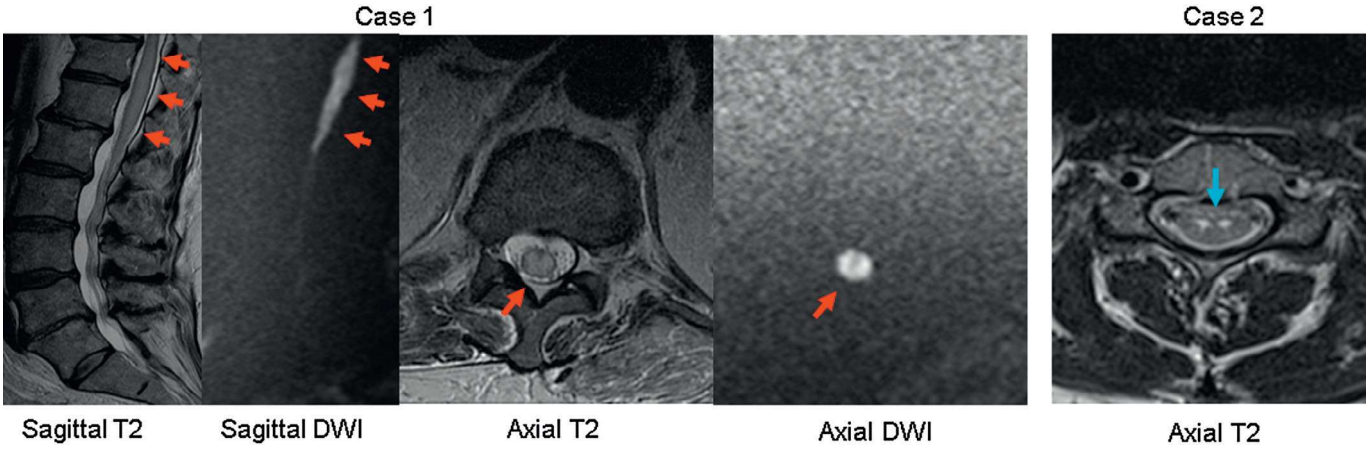

Figure 32.4. Spinal cord infarction. Sagittal and axial T2 and diffusion-weighted images *(DWI)* in a patient with acute spinal cord infarction (case 1) demonstrate a mildly expanded T2 hyperintense edematous conus with restricted diffusion *(red arrows)*. In a different patient (case 2), the classic "snake-eyes" appearance of a chronic spinal cord infarction is evident on an axial T2 image of the cervical spine *(blue arrow)*.

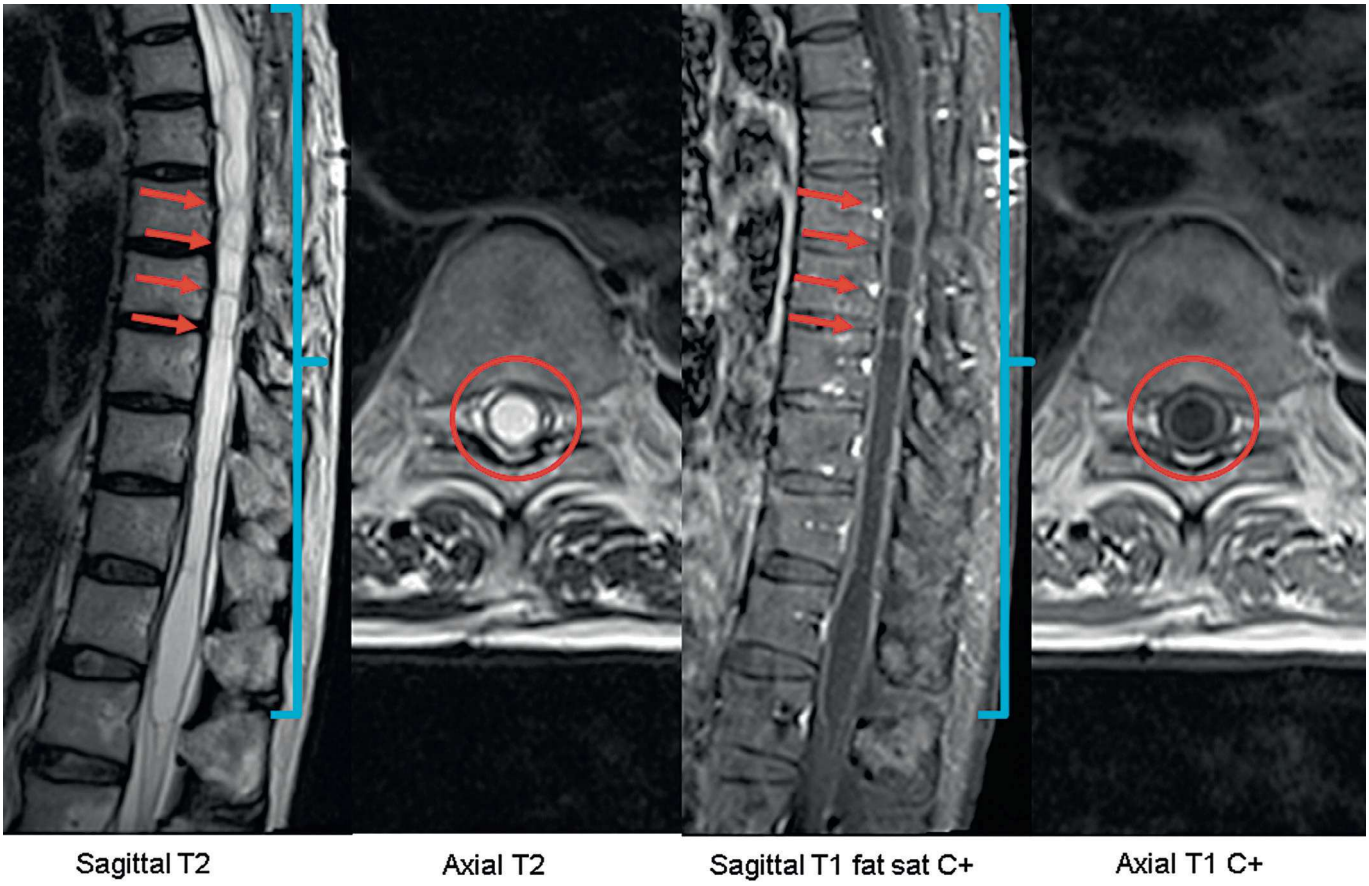

Figure 32.5. Hydrosyringomyelia. Sagittal T2 and sagittal T1 fat-saturated postcontrast images of the thoracic spinal cord demonstrate an extensive *(blue brackets)* nonenhancing, dilated, cystic-appearing intramedullary cavity with sacculated, beaded-appearing areas of narrowing/constriction *(red arrows)*. On axial images, cord expansion with only a thin rim of residual peripheral spinal cord tissue surrounding the dilated intramedullary cavity is evident *(red circles)*.

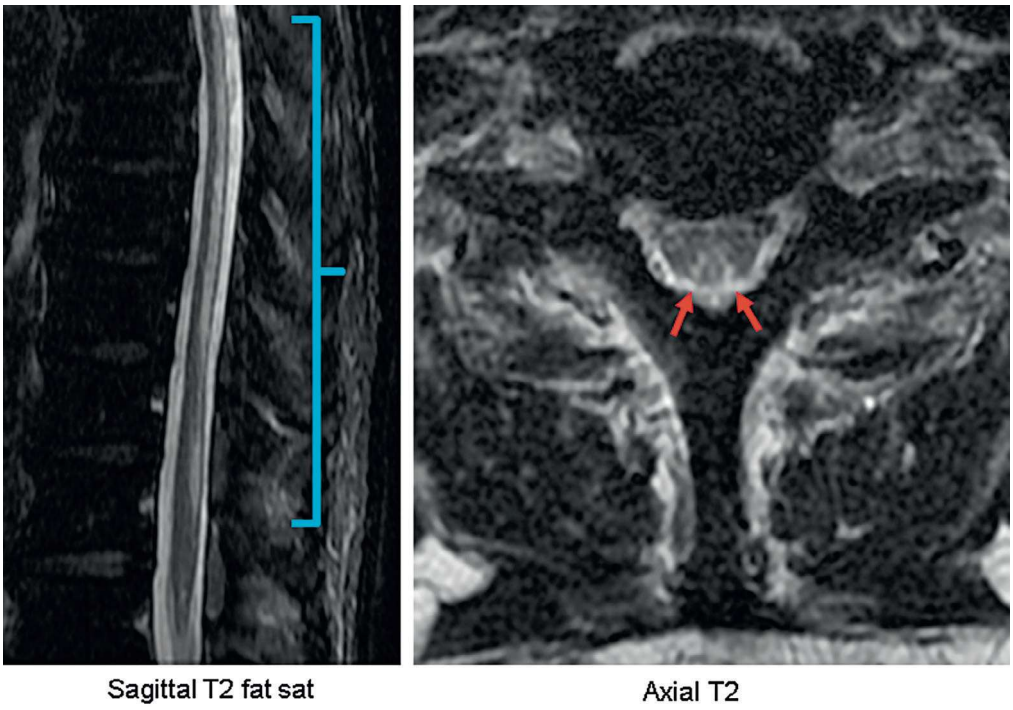

Figure 32.6. Subacute combined degeneration. A sagittal T2 fat-saturated image of the thoracic spinal cord demonstrates extensive *(blue bracket)* spinal cord T2 hyperintensity confined to the dorsal aspect of the cord. Axial T2 image confirms that the finding is isolated to the dorsal columns *(arrows)* in a patient with vitamin B12 deficiency.

SUGGESTED READINGS

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