Chapter 35
Myelopathy Due to Occult Trauma Mimicking Transverse Myelitis

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Case Presentation

A previously healthy 16-year-old girl presented over a 2-month period with numbness and tingling in her lower limbs that spread to her upper limbs with concomitant stiffness. Two weeks prior to presentation, she noticed increasing weakness in all four limbs, to the point of needing help for getting dressed and support for walking. She denied bladder or bowel problems or any systemic symptoms. During the previous year, she had experienced on and off right predominant shooting pain and tingling in the arms that lasted 30 min at a time. She had a positive family history of autoimmune disorders in two sisters and three second-degree relatives. Neurological examination at the time of presentation revealed upper limb predominant quadriparesis. She had increased tone throughout, and strength was 4/5 proximally and 3/5 distally in upper limbs and 4+/5 in lower limbs. Sensation was reduced multimodally in all four limbs without any clear level. Deep tendon reflexes (DTR) were 3+ throughout, and both

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Hoffmann's sign and extensor plantar responses were present bilaterally. Cognition and cranial nerves were intact. MRI of her neuraxis revealed intramedullary T2 signal change from C3 to C5 with patchy enhancement and degenerative changes in discs and vertebrae but no sign of compression (Fig. 35.1a–c). Brain MRI was normal. CSF analysis was normal (total protein of 33 mg/dL, glucose of 49 mg/dL, 1 lymphocyte, no oligoclonal bands, IgG index of 0.5, no bacteria on Gram staining, and negative cultures). Extensive workup included PCR for other herpesviruses and enterovirus in CSF, infectious serologies, and autoimmunity panel that returned negative except for positive antinuclear antibodies (ANA) at a 1:320 titer. She was first diagnosed with a myelitis of uncertain origin (infectious vs. inflammatory) and received high-dose steroids plus acyclovir IV but did not recover.

Two months after being discharged, she developed increasing weakness, and repeat MRI revealed persistence of the previously seen cord lesion and ongoing enhancement (Fig. 35.1d–f). HSV-1 serology was negative (IgG and IgM). Since the patient had a positive ANA titer, ongoing enhancement, clinical worsening, and a family history of autoimmune disease, an inflammatory transverse myelitis was suspected, and plasma exchange was performed for four cycles without clinical improvement.

Reexamination of imaging focused on disc and vertebral body changes raised additional consideration for the possibility of a past trauma. The patient had not had any major cervical trauma but had a motor tic involving her neck for the past 11 years with increasing frequency in the past months, up to several times per hour.

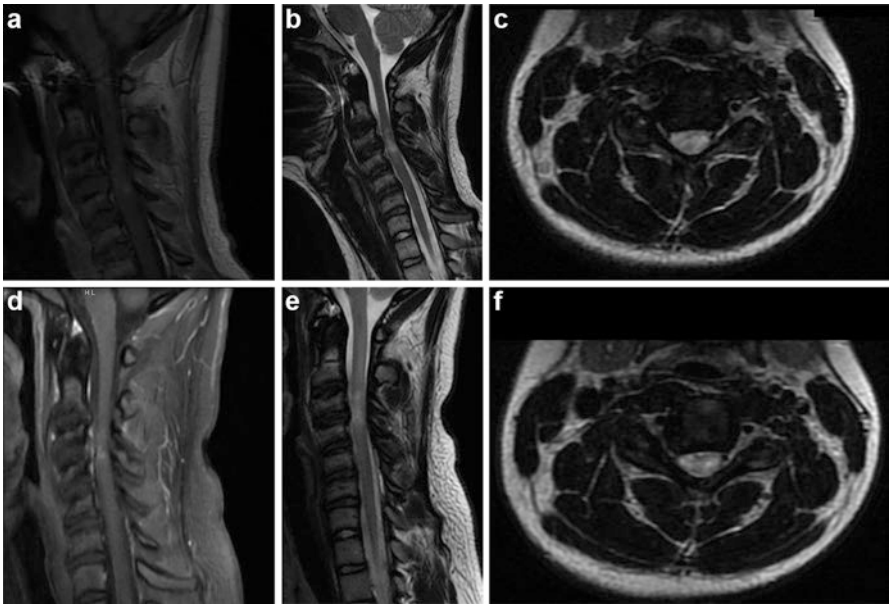

Fig. 35.1 MRI sagittal T1 post-contrast (*left*) and T2 sequences (*middle and right*) of cervical spine at presentation (**a–c**) and 5 months later (**d–f**). The T2 signal hyperintensity extending from C3 to C4 persists, while enhancement is slightly increased on follow-up. Note also mild disc protrusion at C3–C4, mild disc desiccation, and cervical canal narrowing, which are surprising findings for a 16-year-old girl

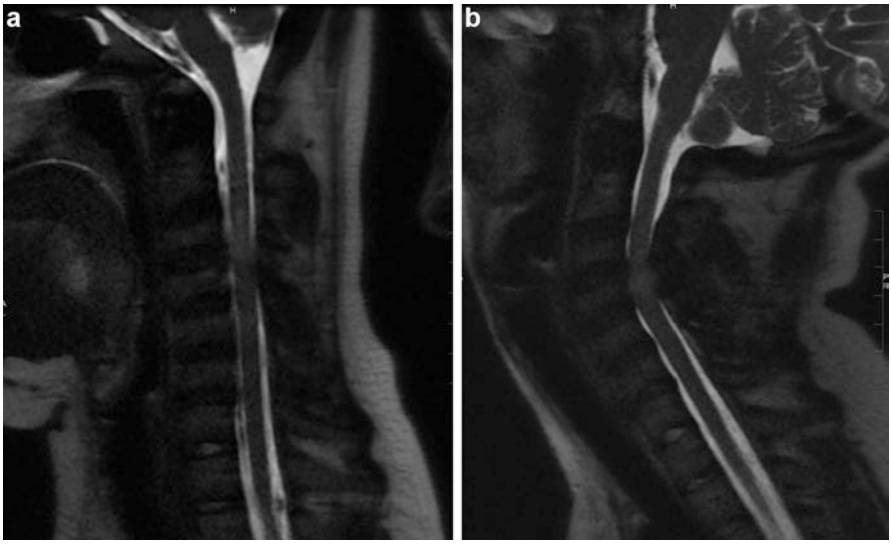

Fig. 35.2 MRI T2 sequence dynamic sagittal images of the spine demonstrate progressive spinal canal narrowing at C3–C4 in neck extension compared to neutral flexion position, related to combination of progressive disc bulge and ligamentum flavum buckling

The tic involved "cracking" her neck by pulling her head laterally to each side with the help of her hands. Dynamic plain radiographs of her neck did not show signs of spine instability. However, dynamic MRI demonstrated cervical spinal canal narrowing at the level of the lesion in neck extension due to disc protrusion and ligamentum flavum buckling (Fig. 35.2). The patient underwent multilevel laminoplasty, and upon clinical follow-up 8 months later, neurological deficits had stabilized and neuropathic pain had improved.

Clinical Questions

1. Where is the lesion based on her symptoms and physical exam?
2. What can cause an enhancing lesion in the spinal cord?
3. How can a spondylotic myelopathy present on MRI?
4. What may cause a spondylotic myelopathy in a teenager?

Discussion

1. Where is the lesion based on her symptoms and physical exam?

   The patient presented with a 10-week history of weakness and multimodal sensory loss involving both lower and upper limbs with increased DTR and plantar extensor responses. This presentation is consistent with cervical myelopathy.

A peripheral origin such of the Guillain-Barré spectrum can be ruled out because of very prominent upper motor neuron signs. Also, a higher than cervical lesion is unlikely due to sparing of cognitive function and cranial nerves. MRI of her neuraxis revealed a partially enhancing intramedullary lesion that spanned from C3 to C5 as well as some degenerative changes in discs and vertebra (see Fig. 35.1a–c)

2. What can cause an enhancing lesion in the spinal cord?

The differential diagnosis for an enhancing lesion in the spinal cord is broad and includes vascular, inflammatory, infectious, neoplastic, paraneoplastic, and compressive etiologies. Chronology of symptoms is crucial to narrow down the differential diagnosis. Myelopathies are acute when nadir of maximum disability is reached between 4 h and 21 days [1]. Hyperacute presentations (below 4 h) will usually have a vascular etiology. The differential diagnosis of acute myelopathy is discussed further in Chap. 32, "Neuromyelitis optica spectrum disease with transverse myelitis presentation."

Since presentation was consistent with a subacute process, causes for both acute and progressive myelopathies had to be considered. CSF analysis can help to further narrow down the differential diagnoses. Brain imaging can also help to detect accompanying abnormalities in inflammatory diseases (i.e., multiple sclerosis) and certain tumors (i.e., multifocal hemangioblastoma, multiple cavernomas).

- An infectious myelopathy was ruled out because of CSF parameters within normal range and negative PCR and serologies for common infectious agents. Transverse myelitis can be isolated (i.e., idiopathic) or part of an inflammatory disease. CSF can exhibit oligoclonal band (OCB) or increased IgG index but may be normal.

– Multiple sclerosis (MS) lesions in the spinal cord tend to be asymmetrical and short (<3 vertebral segments). Approximately, 90% of cases will have OCB in CSF at disease onset. Brain imaging will show typical lesions, although about 10% of isolated TM without brain abnormalities will convert to MS later in life.
– Neuromyelitis optica spectrum disorders (NMOSD) lesions tend to be symmetrical and longitudinally extensive (>3 vertebral segments). In the absence of other typical findings, serum positivity for AQP4-IgG is required for diagnosis. CSF is usually abnormal with elevated WBC.
– Serum autoimmunity panel (e.g., antinuclear antibodies) and other systemic findings are important to exclude rheumatological diseases or associated cancer when suspected.
– Persistence of enhancement beyond 2 months should question an inflammatory origin [2]. One exception to this rule is sarcoidosis, where enhancement may persist chronically. Meningeal enhancement and other systemic findings (i.e., hilar lymphadenopathy) may help with the diagnosis [3].

- Vascular malformation can present in many forms. Fluctuating symptoms due to vascular steal phenomenon are typical. MRI T2-weighted images may

show hyperintense intramedullary signal changes due to edema. Post-contrast T1 images may show enhancement and mimic transverse myelitis. CSF may show elevated protein without pleocytosis. The presence of abnormal vascular flow voids and serpentine enhancement along the surface of the cord are suggestive. Digital subtraction arteriography is the gold standard to confirm the diagnosis.

- Intramedullary tumors may be difficult to distinguish from transverse myelitis. The most frequent tumors in childhood include astrocytoma and ependymoma. A nearby syrinx may be present when the tumor obstructs the central canal. In contrast to inflammatory lesions, tumors will usually enlarge cord diameter. A lesion that continues to worsen beyond the acute time frame despite medical treatment should raise suspicion for a tumor.
- Compressive myelopathy is an uncommon cause for an enhancing lesion. See next question for further details.

3. How does a spondylotic myelopathy present on MRI?

   Spondylotic myelopathy presents with intramedullary hyperintense T2 signal that can be accompanied by gadolinium enhancement. In addition, cervical spondylosis is a common finding in the adult population and may coexist with other spinal pathology. Therefore, the combination of hyperintense T2 signal with post-contrast enhancement may first suggest transverse myelitis or intramedullary tumor [4]. Recognizing spondylotic myelopathy is important as it can be treated, while misdiagnosis may expose patients to hazardous treatments and procedures (i.e., biopsy). The lesion appears on T2 sequence as a nonspecific fusiform area that may extend longitudinally for a variable number of segments away from the site of compression. Gadolinium enhancement when present will show characteristic transverse band, often referred as "pancake like," just below the site of maximum stenosis. This band may be complete or partial. Interestingly, this pattern of enhancement may persist from several months to years despite successful decompression and clinical recovery [5]. However, our case presented with a more longitudinal and patchy enhancement pattern that may reflect dynamic factors contributing to the myelopathy.
4. What may cause a spondylotic myelopathy in a teenager?

   Children and teenagers are not expected to have degenerative spondylotic changes in their spine and, when present, should always raise suspicion for a secondary process. Our patient was discovered to have a motor tic, a potential cause for continuous, repetitive low-grade trauma to the spinal cord. The association between cervical tic and myelopathy has been previously reported. An association with other movement disorders such cervical dystonia has also been described [6]. In one case, the spinal cord lesion preceded spondylotic changes, suggesting that initial cord injury may arise from mechanical stretch or other dynamic factors [7]. In our case, dynamic MRI showed cervical canal narrowing on neck extension (Fig. 35.2) which may further explain the dynamic nature of the myelopathy.

Clinical Pearls

1. MRI for a suspected lesion to the cervical spine should also include brain imaging with and without contrast. Many etiologies causing a cervical lesion share concomitant brain abnormalities which may be clinically silent.
2. The presence of MRI enhancement or elevated protein in CSF does not necessarily indicate an inflammatory origin.
3. Clinical worsening beyond 3 weeks is unusual for an inflammatory lesion and should question alternative diagnosis. An exception to the rule is sarcoidosis.

Spondylotic myelopathy may cause intramedullary T2-bright changes and pancake-like gadolinium enhancement immediately caudal to the site of maximum stenosis.

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