05 Clinical Tests and Differential Diagnosis of Cervical Spondylotic Myelopathy

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Introduction

Cervical spondylotic myelopathy (CSM) is a disabling disease caused by a combination of mechanical compression and vascular compromise of the spinal cord. It is the most common cause of spinal dysfunction in older patients.[1] The onset is often insidious with long periods of episodic, stepwise progression and may present with different symptoms from one patient to another.[2] CSM is a clinical diagnosis that may involve broad-based gait disturbances first, associated with weakness of the legs, and then spasticity.[3] As spinal cord degeneration progresses, lower motor neuron findings in the upper extremities, such as loss of strength, atrophy, and difficulty in fine finger movements, may present.[3] Additional clinical findings may include: neck stiffness, shoulder pain, paresthesia in one or both arms or hands, radiculopathy, a positive Hoffman and/or Babinski sign, motor deficits, hyperreflexia, and bowel and bladder dysfunction.[3]

Physical examination findings are not always consistent with severity of disease in CSM; therefore, correlation to plain X-rays, MRI, and clinical symptoms is essential for a correct diagnosis. Anterior-posterior width reduction, cross-sectional evidence of cord compression, obliteration of the subarachnoid space, and signal intensity changes to the cord found on MR imaging are considered the most appropriate parameters for confirmation of a spinal cord compression myelopathy.[4] In some occasions when the diagnosis is still not clear, the use of other studies could help, such as diagnostic electrophysiology and cerebrospinal fluid (CSF) examination.

Clinical Tests

CSM is the most common cause of spinal cord dysfunction in the world. A meticulous physical examination of patients with cervical pathology can relatively make the distinction between radiculopathy or myelopathy easy. Routine physical examination of patients with cervical myelopathy should include special tests in addition to a thorough neurological examination. The relevant clinical examination tests are categorized in **Table 5.1**.

Table 5.1 Clinical examination tests used for cervical spondylotic myelopathy

Motor deficit	Sensory deficit	Upper motor neuron deficit	Provocative test	Gait and balance
Muscle weakness Finger scape test Grip and release	Proprioception Vibratory test Pinprick	Hoffman´s reflex Babinski reflex Sustained clonus Inverted radial reflex Scapulohumeral test Hyperreflexia	L´hermitte sign	Romberg test Heel and toe walk

Muscle Weakness

The causes of lack of muscle strength are many; however, in CSM, the most common cause is due to a peripheral muscular fatigue, which represents the inability of a specific muscle to work, secondary to a loss or dysfunction on any of the motor neurological pathways. This can be caused by osteophytes or prolapsed discs that compress either the spinal cord directly or the spinal nerves. It is important to distinguish weakness from fatigue or asthenia, which are separate conditions with different etiologies that can coexist with, or be confused for, weakness.

Technique

The patient stays in a sitting position and the doctor tests the level of strength in all muscle groups, examining from upper to lower extremities and quantifying the severity of muscle weakness, according to the Medical Research Council (MRC) criteria[5]:

- **Grade 0**: No contraction or muscle movement.
- **Grade 1**: Trace of contraction but no movement at the joint.
- **Grade 2**: Movement at the joint with gravity eliminated.
- **Grade 3**: Movement against gravity but not against added resistance.
- **Grade 4**: Movement against external resistance with less strength than usual.
- **Grade 5**: Normal strength.

This grading is not only an excellent tool to determine muscle weakness but also a great tool to evaluate progression and recovery after treatment.

The Wartenberg´s Sign or Finger Scape Sign

It was first described by Robert Wartenberg in 1939. Wartenberg's sign refers to the slightly greater abduction of the fifth digit, due to paralysis of the abducting palmar interosseous muscle and unopposed action of the radial innervated extensor muscles (digiti minimi, digitorum communis).[6]

From the physiological standpoint, this sign is easily understandable. Adduction of the little finger is performed by the interosseous and abduction by the hypothenar muscles. Both groups of the muscles are innervated by the ulnar nerve. However, in abduction of the little finger, the extensor digiti minimi and the branch to the little finger of extensor digitorum communis also play a definite part; both are innervated by radial nerve. If the muscles innervated by the ulnar nerve are weak, those innervated by the intact radial nerve predominate in strength and abduct the little finger. Thus, it is understandable why this abduction of the little finger is best seen when extensor digitorum communis comes into action and extends the fingers and the hand. In cases with combined palsy of ulnar nerve and radial nerve, this sign would not be present.

The finger scape sign is not only seen in ulnar nerve palsies but also in patients with syringomyelia and is an important examination test in the myelopathic hand together with grip and release.

Technique

The patient is placed with the wrist in a neutral position, and forearm fully pronated and instructed to perform full extension of all the fingers. Once digits are extended, patient is asked to fully abduct all fingers and then adduct all fingers. A positive sign is indicated with the observation of abduction, along with the inability to adduct the 5th digit when extended (**Fig. 5.1**).

Grip and Release Test

Grip and release test is used to evaluate hand function in myelopathic patients. Grip and release tests are part of a group of quantitative clinical tests together with 10-step test and the 30 meters walking test.[7] They are all common, in that they can demonstrate an improvement numerically, comparing

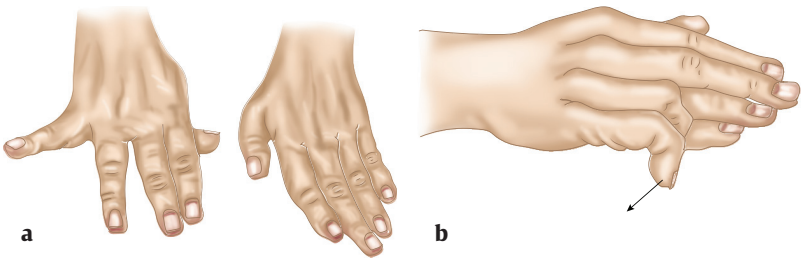

Fig. 5.1 Wartenberg scape test. 5th finger on abduction and flexion as the lesion does not allow the finger to adduct (radial nerve dominance).

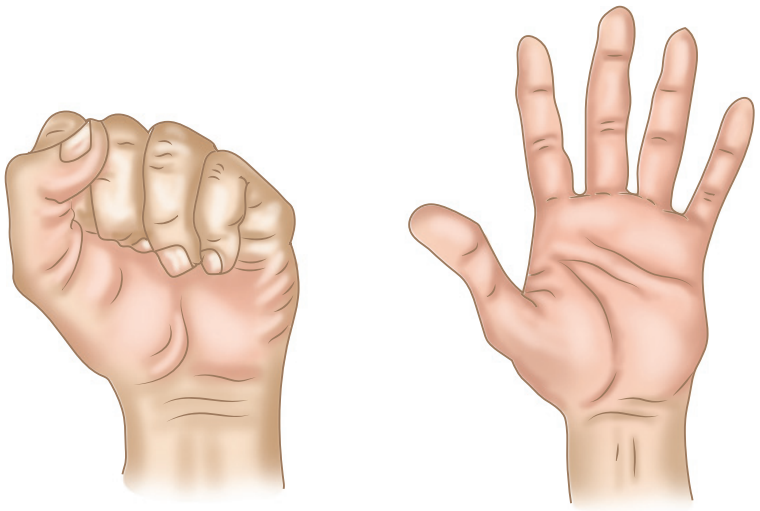

Fig. 5.2 Grip and release quantitative test. Opening and closing the hand. Normal value: > 20 in 10 seconds.

the performance before and after rehabilitation or surgery. This improvement normally coincides with better results in other clinical assessment subjective scales such as Japanese Orthopaedic Association (JOA), short form-36 (SF-36), neck disability index (NDI), and quality-adjusted life-years (QALY) and is reflection of a good management outcome.

It is believed that changes in grip aperture control are caused mainly by pyramidal tract damage in the spinal cord; however, recent studies confirm the importance of somatosensory inputs from the hand into grip aperture control.[8]

Technique

In this reflex, the patient is asked to form a fist and then extend the fingers, rapidly repeating the sequence. A normal response is more than 20 times in a 10-second period. Patients with cervical myelopathy (myelopathic hand) cannot achieve this goal (**Fig. 5.2**).

Proprioception

Proprioception, or kinesthesia, is the sense that lets us perceive the location, movement, and action of parts of the body. When we move, our brain senses the effort, force, and heaviness of our actions and positions and responds accordingly, for example, proprioception enables a person to close their eyes and touch their nose with their index finger. Proprioception includes a complex of sensations, including perception of joint position and movement, muscle force, and effort. In proprioception, the neuromuscular control is the efferent motor response to afferent (sensory) information, and it is a constant feedback loop that tells your brain what position you are in and what forces are acting upon your body at any given point in time.[9] Proprioception relies on the relationship between the body's central nervous system (CNS) and certain soft tissues, including muscles, tendons, and ligaments. Sensory

receptors in the muscles are called muscle spindles, which are long proteins encapsulated in sheaths that lay parallel to muscle fibers. These muscle spindles are stretched on muscle extension and is the degree of this stretch effort that is delivered through the spinal nerves to the nervous system, which facilitates a signal sent to the muscle to contract or relax.

Proprioception sensory output ascends through the dorsal columns in the spinal cord. Normal proprioception lets you move freely without giving your movements a second thought. Abnormal proprioception causes symptoms that can interfere with even the simplest activities and even can cause you to fall in severe cases.[10] In upper extremity injuries like in CSM, you may have difficulty reaching properly, and you may have problems with fine motor tasks that require precision of movement (myelopathic hand). This is caused by a loss in proprioception.

Technique

The patient is positioned lying on the bed. He or she is asked to relax and close his or her eyes. The physician then moves the big toe up, down, or keeps it in neutral, asking the patient to recognize which is the position of the big toe (**Fig. 5.3**). Proprioception can also be done in the upper extremity, using the fingers phalanx and asking the patient to recognize what is the position of the finger or phalanx.

Vibratory Sensation

Assessment of vibration sense is the best clinical test of the dorsal column pathway. Vibration sense is tested with a tuning fork (normally 128 Hz) placed on predefined bony prominences (with the eyes closed), and the patient is asked to report when the vibration starts and stops. Sensory receptors (mostly Pacinian and Meissner corpuscles) convert the vibration into a neural signal. The temporal resolution of the neural information transfer (action potentials) should be at least equal to the frequency of the vibration. With age, the vibratory response might be lost in the ankle and both feet. Vibratory sensation is often diminished in peripheral neuropathy and myelopathy but spared in disease confined to the cerebral cortex, orientating the problem to a spinal one.

Technique

The patient is asked to lay on the couch. The examiner explains the procedure until the patient obtains a comprehensive understanding. He or she is then asked to close their eyes and after a gentle strike to the tuning fork, the examiner applies it on top of the big toe (**Fig. 5.4**), asking the patient if he or she feels the vibration and when does it stop. It is important to perform this procedure in both feet and arms to detect any discordance between both sides.

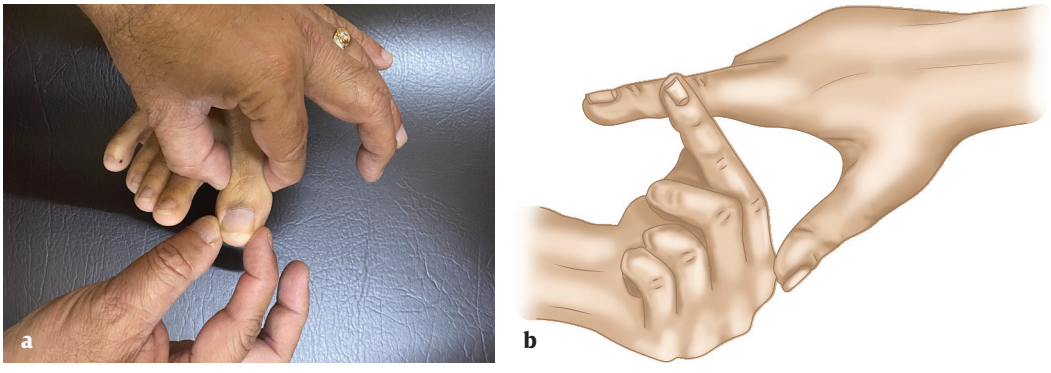

Fig. 5.3 Proprioception test in the phalanx of the big toe **(a)** and at the index finger **(b)**.

Fig. 5.4 Application of the tuning fork in the big toe in order to perform the vibration sense test.

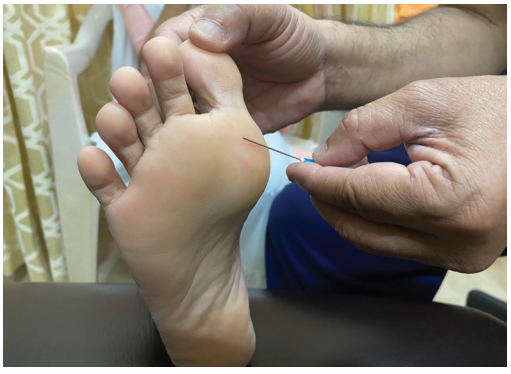

Fig. 5.5 Pinprick examination using a blunt needle or a pin, tapping in the skin and testing different areas of the body recognizing the dermatome involved.

Pinprick (PP) Test

The PP test is performed to test the numbness and pins and needles in a part of the body. The PP is a gross test that checks three variables: (1) the actual ability to feel a pinprick, (2) the ability to determine the difference between sharp and dull, and (3) the ability to locate the area where the pinprick has been tested on. Remember, pain and temperature sensory neurons entering the cord ascend ipsilaterally for two to three spinal segments in the dorsolateral tract of Lissauer before crossing just anterior to the central canal, in order to join the contralateral spinothalamic tract located in the lateral cord. Therefore, loss of pinprick or temperature sensation at a given level may indicate pathology two to three segments above the level detected on examination.

Technique

The patient is asked to lay on the couch. The examiner explains the procedure until the patient obtains a comprehensive understanding. Using a pin or a blunt needle and with the patient´s eyes closed, the examiner begins targeting different areas in accordance with the relevant dermatome to be examined (**Fig. 5.5**). The pin should never penetrate the dermis, as it would render an invalid test. In examining the sensibility, the patient is not only asked if he or she can feel but also where he or she can feel it, as feeling the sensation is as relevant as localizing it.

Babinski Reflex

The Babinski reflex (plantar reflex) was described by the neurologist Joseph Babinski in 1899 and is considered normal for infants below the age of 2.[11,12] This reflex tests the integrity of the cortical spinal tract (CST). The CST or pyramidal tract (upper motor neuron) is a descending tract travelling from the eloquent motor area of the brain throughout the spinal cord until it synapses with the alfa motor neuron (lower motor neuron), facilitating a normal motor response.[4,11,12]

A normal Babinski results after stimulating the lateral side of the sole (S1 myotome) and obtaining a plantar flexor response from the toes. Stimulation at the sole (nociceptive fibers S1) travel in an ascending path through the sciatic nerve until the S1 region of the spine, where synapse with the anterior horn cells produce a descending response, which travels through sciatic and tibial nerve until S1 motor fibers and induce the toe plantar flexion.

An abnormal or positive Babinski occurs when stimulating the lateral plantar side of the sole, which leads to dorsiflexion (extension) of the big toe, and on occasions, fanning

of the rest of the toes.[4] This is believed to be caused by overspreading of the S1 nociceptive response, reaching the L4 and L5 regions of the spine synapsing producing an overfiring of the anterior horn cells at that level, which descending cause and overwhelming extensor response through the peroneal nerve (extensor hallucis longus and extensor digitorum longus). An intact CST prevents this overspread.

A positive Babinski reflex is indicative of dysfunction of the CST, so not only does it reflect a problem in the cervical spinal cord but also all along the pyramidal tract from brain to lower thoracic spine.[4,11]

Technique

The patient has to be in supine position, relaxed with the legs resting on a couch. You need a pin or a blunt instrument, which has to be run up the lateral plantar side of the foot from the heel to the toes and across the metatarsal pads to the base of the big toe, watching for an extensor response (**Fig. 5.6**). Sharp instruments are not recommended.

There have been multiple variations to the Babinski sign, all eliciting same dorsiflexion response of the big toe. The most known include, Chaddock (stimulating under lateral malleolus), Gordon (squeezing calf), Oppenheim (applying pressure to the medial side of the tibia), and Throckmorton (hitting the metatarsophalangeal joint of the big toe).[12]

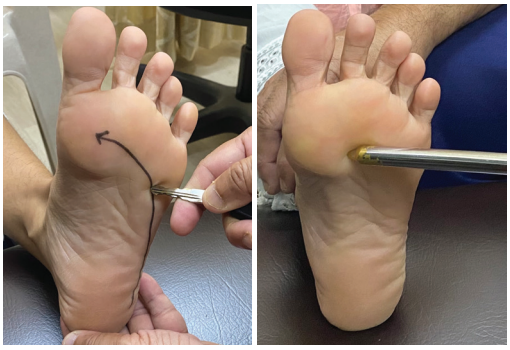

Fig. 5.6 Instructions in how to run the blunt instrument in the sole of the patient. Note the extensor response on the right.

Sustained Clonus

Clonus is a rhythmic, oscillating, repetitive stretch reflex; the cause of which is not totally known; however, it relates to lesions in upper motor neurons and therefore is generally accompanied by hyperreflexia. Like other signs of upper motor neuron syndrome clonus indicates some insult to the central rather than peripheral nervous system, so part of its utility as a clinical examination skill is in differentiating the two.[13]

The mechanism of this reflex is initiated by hyperexcitability produced in muscle stretch circuits when there is less tonic inhibition of motor neurons involved in the monosynaptic stretch reflex. This can occur when there is a lesion to descending motor nerves, predominantly the dorsal reticulospinal pathway, which can occur anywhere from the cortex to the spinal cord. The inhibitory dampening effect of these descending nerves on alpha and gamma motor neurons is removed, leading to a hyperexcitatory state in the muscle stretch reflex circuit.[13,14]

Technique

The patient is asked to relax with a passively flexed ankle to about 90 degrees and a passively flexed knee; this usually involves the examiner supporting the leg with the hand not performing clonus. Next, at the ankle, the examiner places their hand on the dorsum of the patient's forefoot and briskly dorsiflexes it, after which the former continues to maintain dorsiflexion pressure (**Fig. 5.7**). It is against this pressure that the clonus beats will be felt. Each beat will be felt as a plantar flexion followed by a relaxation. The initial beat is the longest, with decreasing duration of beats until the fourth beat, after which the beat frequency becomes equivalent from one to the next.[15] Count of three or over number of beats is considered pathological.

Hoffmann's Reflex

It was first described by the neurologist Johann Hoffmann.[16] This reflex is used to test

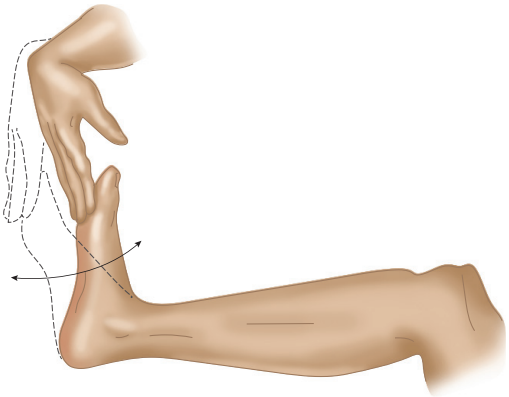

Fig. 5.7 Sustained clonus: In patients with cervical spondylotic myelopathy (CSM), after a brisk dorsiflexion on the foot, a sustained beat should be seen, confirming upper motorneurone damage.

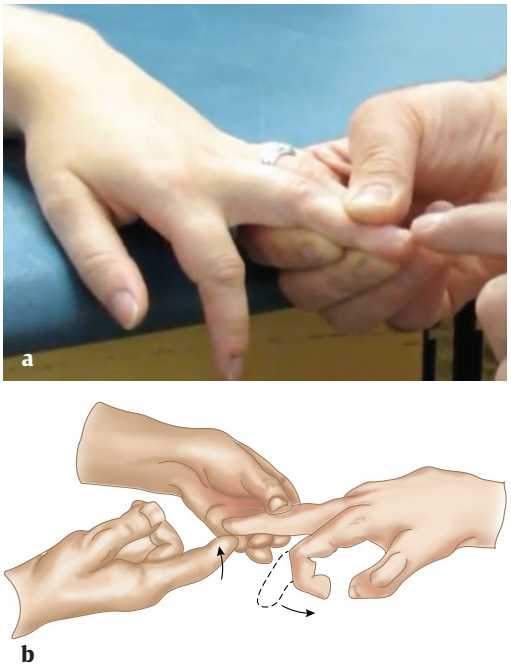

Fig. 5.8 On tapping the middle finger, the thumb and index fingers go on to flexion.

upper extremity reflexes and is considered a more reliable test when examining for suspected CSM compared to the Babinski reflex. Hoffmann's reflex has a different mechanism of reflex than Babinski, as it is a deep tendon reflex (spindle fiber) with a monosynaptic reflex pathway in Rexed lamina IX of the spinal cord, normally fully inhibited by descending input. Hoffman's reflex can be present on one side only, such as in Brown–Sequard syndrome in which only one-half of the spinal cord is affected.

Technique

The Hoffmann's reflex test itself involves loosely holding the middle finger and flicking the fingernail downward, allowing the middle finger to flick upward reflexively (**Fig. 5.8**). A positive response is seen when there is flexion and adduction of the thumb on the same hand, accompanied sometimes with flexion of the index or even all fingers.

Inverted Radial Reflex

The inverted supinator (brachioradialis) reflex is a sign that was introduced by Babinski in 1910.[11] The inverted radial reflex sign is commonly used in clinical practice to assess cervical myelopathy. It is also indicative of a spinal cord lesion at C5 or C6, for example, due to trauma, syringomyelia, or disc prolapse and occurs because a lower motor neuron lesion of C5 is combined with an upper motor neuron lesion affecting reflexes below C5. There are two components of this abnormal reflex on tapping the lower end of the radius:

- Absent biceps and exaggerated triceps reflexes.
- A hyperactive response of the finger flexor muscles, causing finger flexion.

Technique

The supinator reflex is tested by striking the lower end of the radius (styloid process) just above the wrist with a tendon hammer (**Fig. 5.9**). This normally causes contraction of the brachioradialis and hence flexion of the elbow. If the only response is finger flexion, then this reflex is said to be inverted.[17]

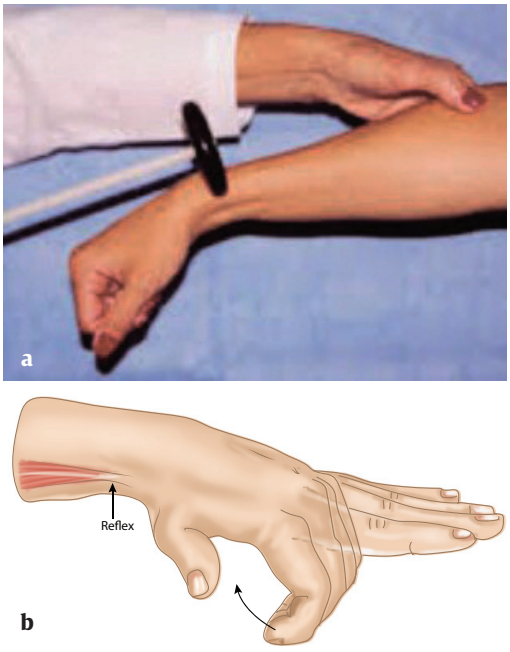

Fig. 5.9 Inverted brachial reflex. Tapping on the radius and unexpected finger flexion is obtained.

Scapulohumeral Reflex

The scapulohumeral reflex (SHR), which was first described by Shimizu in 1993, reflects damage in the upper cervical spine.[18] The reflex center of the SHR is clinically presumed to be located between the posterior arch of C1 and the caudal edge of the C3 body. A hyperactive SHR response was shown in 95% patients who exhibited neural compressive factors at the high cervical region.[18] Most of these cases had spastic symptoms and included several neurologic abnormalities (myelopathy, syrinx, EM). Hyperactive SHR provides useful information about dysfunctions of the upper motor neurons cranial to the C3 vertebral body level.

Technique

The SHR is elicited by tapping the tip of the spine of the scapula and acromion in a caudal direction. The SHR is classified as hyperactive only when an elevation of the scapula or an abduction of the humerus has been clearly defined after tapping at these points.

Hyperreflexia

Hyperreflexia is defined as overactive or overresponsive reflexes. Hyperactive deep tendon reflexes are a sign of upper motor neuron lesion and can be seen in normal but tense people. It can also be a consequence of loss of control ordinarily exerted by higher brain centers of lower neural pathways (disinhibition). Clonus is the higher representation of hyperreflexia. Diagnostic difficulty occurs when hyperreflexia and spasticity are the only findings. When hyperreflexia is found, it is wise to look for other features of upper motor neuron dysfunction such as positive Babinski sign and hypertonia, in order to determine a more clear diagnosis. Jaw clonus often indicates a lesion above the midpontine level.

Sensory nerve tracts eliciting the knee jerk reflex enter the spinal cord through L2, L3, and L4 spinal nerves. Synapse takes place between sensory and motor tracts in the spinal cord and immediately exits through the same spinal nerves. Hyperreflexia will occur when there is a lesion in the spinal cord which disrupts the inhibitory tract and originates in the brain and descends through the spinal cord, exiting via the spinal nerves of L2, L3, L4 to modulate the knee jerk reflex, or S1 to modulate the ankle jerk reflex.

The most common cause of hyperreflexia is spinal cord injury in which case the name changes onto autonomic hyperreflexia/dysreflexia. This condition results from chronic disruption of efferent impulses down the spinal cord, as seen in spinal trauma, neuromuscular diseases (amyotrophic lateral sclerosis [ALS]), or CSM. Autonomic hyperreflexia is uncommon if the level of the injury is below T5. Inciting stimuli such as bladder distention, bowel distention, or surgical stimulation can produce an exaggerated sympathetic response. This occurs because there is a

loss of normal inhibitory impulses from areas above the level of the lesion.

Technique for Deep Tendon Reflexes

The patient has to be sitting with the legs hanging and completely relaxed. The physician uses a patellar hammer to gently strike the patellar ligament, eliciting the reflex (**Fig. 5.10**). Proper technique of reflexes examination and experience play a major role in eliciting and categorizing deep tendon reflexes. An overactive response should be considered hyperreflexia.

L'hermitte Sign

Lhermitte phenomenon was described in 1916 by the French neurologist Jean Lhermitte.[19] It is described as an uncomfortable sensation, resembling an electric shock, provoked by active or passive neck flexion or when the physician pounds on the cervical spine while it is flexed. This trigger gives Lhermitte's sign its alternative name of barber's chair syndrome, as this movement is similar to when you move your head forward to let the barber cut the back of your hair. This sensation runs throughout the spinal cord, radiating from arms to legs, and is a result of a dysfunction of the posterior columns at the cervical level. It usually lasts a few seconds but can be very intense.

Technique

Position the patient in a sitting or a standing position. Ask the patient to try and touch the chest with their chin (flexion) or gently apply some force, resting your hand on the back of the head and bending the neck forward (**Fig. 5.11**). The electric shock sensation will appear and the patient will regain his or her normal neck position (neutral).

Fig. 5.10 Patellar reflex. With the leg resting, we tap the patellar ligament, obtaining a reflex.

Romberg Test

The Romberg test was first described in 1846 by the German neurologist Moritz H Romberg, which was originally described for the condition tabes dorsalis. The Romberg test is used for the clinical assessment of patients with imbalance or ataxia from sensory and motor disorders.[20]

Normal balance occurs when all acting forces (motor and sensory) are cancelled by each other, resulting in a stable balanced system. It is maintained through the sensory information from vestibular (VIII cranial nerve), somatosensory (dorsal columns of the spinal cord), and visual systems (II cranial nerve). A patient who has a problem with proprioception (somatosensory) can still maintain balance by compensating with vestibular function and vision. In the Romberg

Fig. 5.11 L´hermitte sign. A positive result is obtained when flexing the neck and observing an "electric-like" sensation running down the back to arms and legs.

test, the patient stands upright and is asked to close his eyes. A loss of balance is interpreted as a positive Romberg sign.[21]

Technique

The patient is kept in a standing position and legs are placed straight, with both feet touching each other and the head in a neutral position. A positive Romberg test results when the patient suddenly loses balance and has to be held to avoid a potential fall and/or injury when he or she is asked to close their eyes (**Fig. 5.12**).

Heel-to-Toe Walking

Heel-to-toe walking is another test to determine the integrity of the neurological pathways that control balance. The test result is considered positive when there is a demonstration of a loss of postural control.

Similar to Romberg's test, heel to toe positive test has been linked to all causes of proprioceptive deficits, including myelopathies of many causes, tabes dorsalis, and sensory neuropathies.

Technique

The patient is asked to walk in a straight line, touching the heel with the other´s foot toes (**Fig. 5.13**). They are asked to do this for 8 to 10 steps, and contrary to Romberg´s sign, the patient can keep their eyes open.

Nearly all of these tests are specific, versus sensitive, and are useful to rule in a suspected condition such as CSM. Furthermore, the tests, when used alone, may lead to a number of false negatives and, on rare occasions, false positives.

Differential Diagnosis

Only with a good history taking and physical examination can physicians derive appropriate differential diagnosis, which leads to proper imaging and other diagnostic tests that are cost-effective and minimally burdensome to patients. We can categorize the differential diagnosis broadly between compressive myelopathy, noncompressive myelopathy, and others (**Table 5.2**).

Compressive Myelopathies

Compression on to the spinal cord causes myelopathy. CSM with all its forms such as spondylosis, osteophyte formation, disc herniation, synovial cyst, ossification of posterior longitudinal ligament (OPLL), and subluxation, belong to this group of degenerative myelopathic diseases.

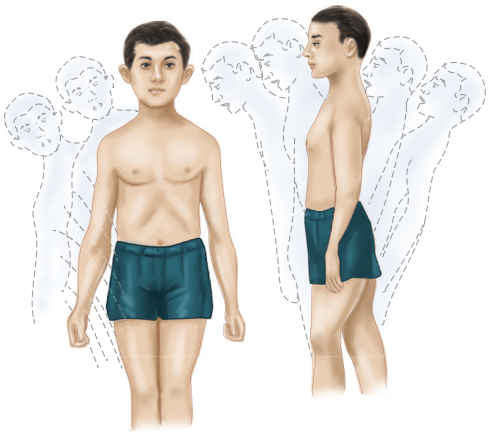

Fig. 5.12 Romberg test. On closing the eyes, the patient loses balance and has to widen the gait or advance a leg to avoid a fall.

Fig. 5.13 Heel-to-toe walk. Walking on a straight line, heel to toe shows the status of the balance.

Table 5.2 Types of myelopathies

Compressive myelopathy	Noncompressive myelopathy	Others
Degenerative	Spinal cord infarct	AVM (vascular malformations)
Congenital malformation	Inflammatory/autoimmune	Dural fistula
Traumatic injury	MS	Retrovirus myelopathy (HIV)
Spinal tumors	Rheumatoid arthritis	Syringomyelia
Epidural hematoma	Acute infective myelitis	Subacute degeneration (vitamin B deficiency)
Hematomyelia	Post-infection myelitis	Tabes dorsalis
Epidural abscess	Lupus/Sjögren	Familial spastic paraplegia
	Sarcoidosis	Adrenomyeloneuropathy
	Radiation myelopathy	
	Discitis/osteomyelitis	
	ALS	
	Neuromyelitis optica	

Abbreviations: ALS, amyotrophic lateral sclerosis; AVM, arteriovenous malformations; HIV, human immunodeficiency virus; MS, multiple sclerosis.

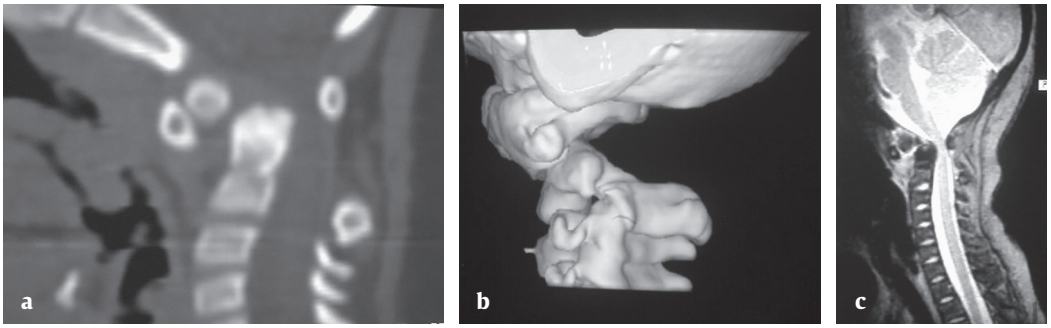

Fig. 5.14 Os odontoideum. **(a)**: CT showing bony invasion of the cervical canal; **(b)**: CT-3D spinal malalignment associated with os odontoideum; **(c)**: MRI confirming severe compression to the spinal cord.

Spinal congenital malformations such as defects of the anterior or posterior arches of C1, occipital assimilation of the atlas, basilar invagination or impression, os odontoideum (**Fig. 5.14**), Klippel–Feil syndrome (**Fig. 5.15**), and cervical spine congenital stenosis can cause myelopathy by reducing the vertebral canal area and compressing the spinal cord. In congenital cervical stenosis, the average canal diameter, which produces the neurological symptoms, ranges from 7 to 9 mm[21]. In other congenital anomalies of the craniocervical junction such as Down´s syndrome, achondroplasia, and odontoid and atlas hypoplasia, the compression is caused by spinal malalignment associated with or without ligament laxity, which causes the area to be hypermobile, damaging the spinal cord on repetitive flexion and extension movements during normal living.[22] Chiari malformation, which is also a congenital disease, causes myelopathic symptoms on account of spinal cord compression caused by tonsillar descent, which occupies the vertebral canal, pressing the spinal cord posteriorly.

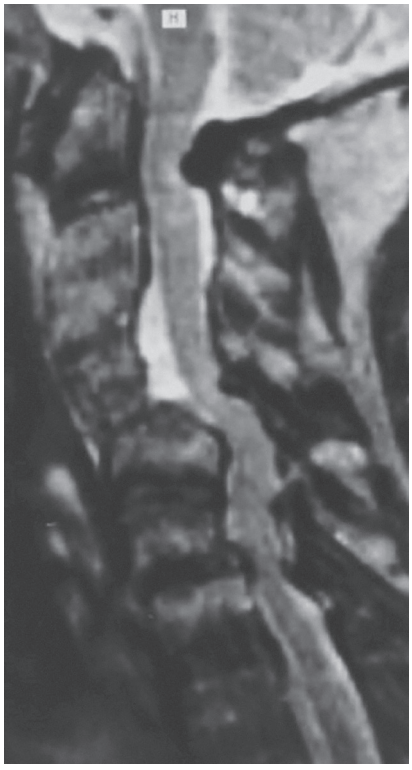

Fig. 5.15 MRI of a Klippel–Feil case. Spinal deformity; vertical translocation and staircase deformity, compressing the spinal cord on different levels.

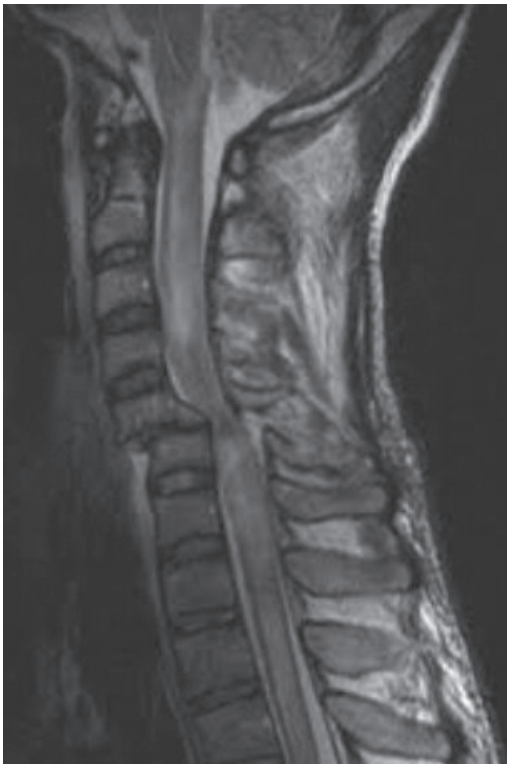

Fig. 5.16 A 27-year-old man cervical MRI demonstrating an unstable fracture dislocation on C5/6 with severe cord compression and an incomplete tetraparesis.

Trauma to the cervical spine also causes myelopathy by compression of the bony fragments that had invaded the vertebral canal, compressing the spinal cord. Lesion to the spinal cord can be either a complete lesion (no function below the level of the injury) or an incomplete one (some useful motor function below the level of injury). In the event that no neurological deficit has occurred, classification of the cervical fracture is mandatory to determine its stability. On a stable fracture, a collar will suffice, while on an unstable fracture (**Fig. 5.16**), the management will include decompression, realignment, and stabilization.

A spinal tumor is an abnormal mass of tissue within or surrounding the spinal cord and/or spinal column. The tumors can be benign or malignant and can be classified according to their relation with the spine: extradural, intradural-extramedullary, and intradural intramedullary tumors.[23] They cause compression to the spinal cord by the presence of its mass, which competes with the spinal cord to space in the vertebral canal. The most common spinal tumors causing myelopathy are metastasis (lung, breast, and prostate), followed by meningiomas (**Fig. 5.17**) and schwannomas (intradural-extramedullary spinal tumors). Primary tumors of the bone and intramedullary tumors are infrequent.[24] On intramedullary tumors (**Fig. 5.18**), the presence of an intramedullary syrinx is common, which can extend to several levels, causing additional myelopathic symptoms and signs.

Cervical epidural hematoma (CEH) is a collection of blood between the dura mater and the bone, which can cause cord compression

and myelopathy. CEH was first described by Jackson.[25] Hematomas can occur, following a cervical traumatic injury (venous), spontaneously in case of a clotting disorder (**Fig. 5.19**) or as a result of a postsurgical complication after a cervical laminectomy or an incomplete tumor removal.[26] Hematomyelia is the term used when the hemorrhage occurs inside the spinal cord. This hemorrhage is always due to a vascular anomaly or bleeding from a hemangioblastoma.

If the hematoma is confirmed by appropriate imaging and is causing a neurological deficit, the hematoma needs to be removed urgently; trying at the same time, to solve its cause.

Cervical epidural abscess (CEA) is a rare condition that leads to devastating neurological deficits and may be fatal. The classical triad of symptoms includes back pain, fever, and neurological deterioration. The infection often begins in the bone (osteomyelitis) and spreads inside the vertebral canal, causing myelopathy through two different mechanisms: direct compression on the spinal cord and due to a chemical reaction induced by pus and infectious pathogens (bacteria), which cause inflammation with local ischemia in the spinal cord, significantly worsening the symptomatology.[27,28] Abnormal blood inflammatory markers with fever and neurological deterioration would lead to a high suspicion of CEA. MRI with and without contrast and fat suppression is the imaging of choice with high sensitivity and specificity (**Fig. 5.20**).

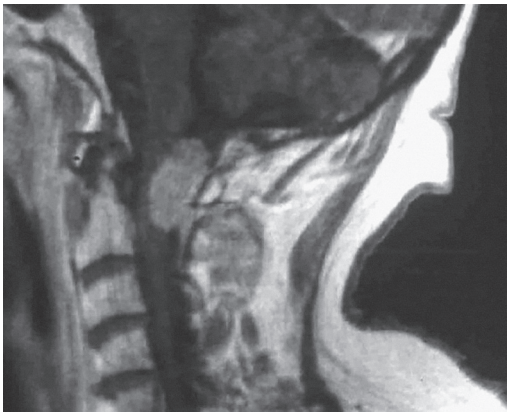

Fig. 5.17 MRI showing a meningioma at the C1–2 complex, compressing the spinal cord posteriorly.

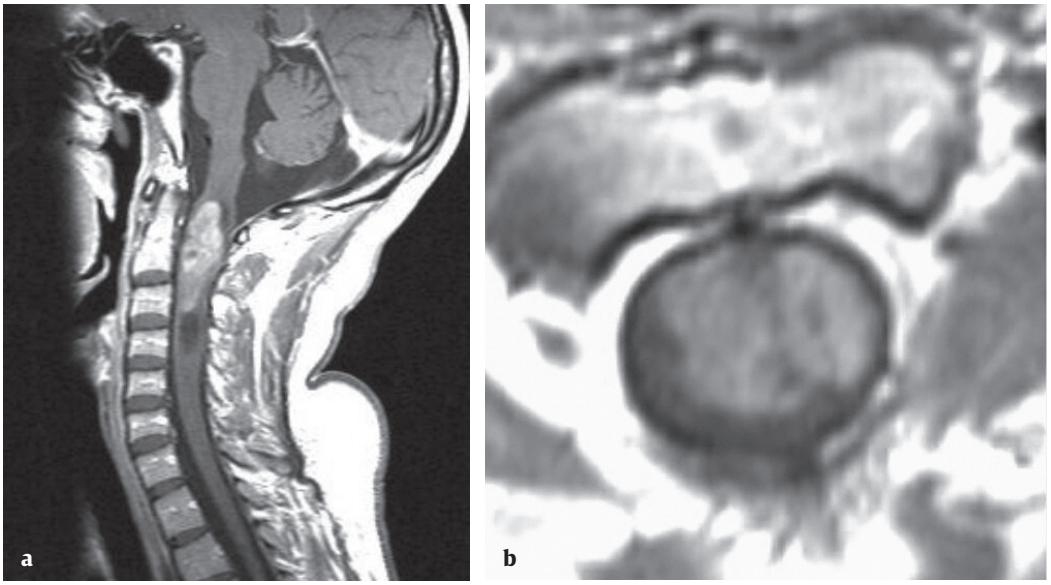

Fig. 5.18 MRI of an intramedullary tumor (ependymoma), causing severe cord compression and myelopathy.

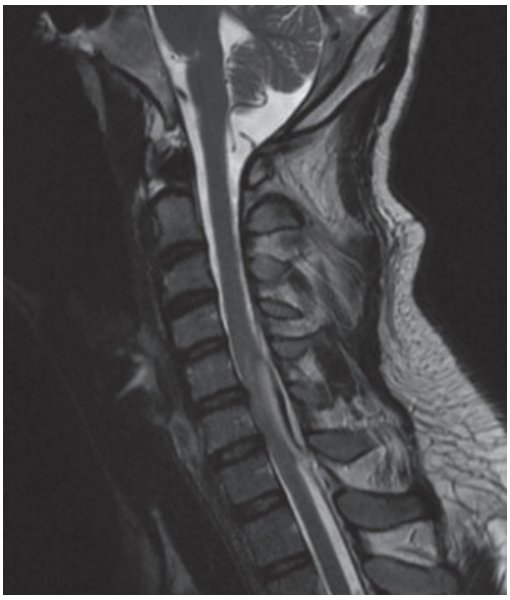

Fig. 5.19 MRI showing a spinal epidural hematoma (EDH) which is most common, as in this case, due to spontaneous venous bleeding, often in the setting of coagulopathy or over-anticoagulation.

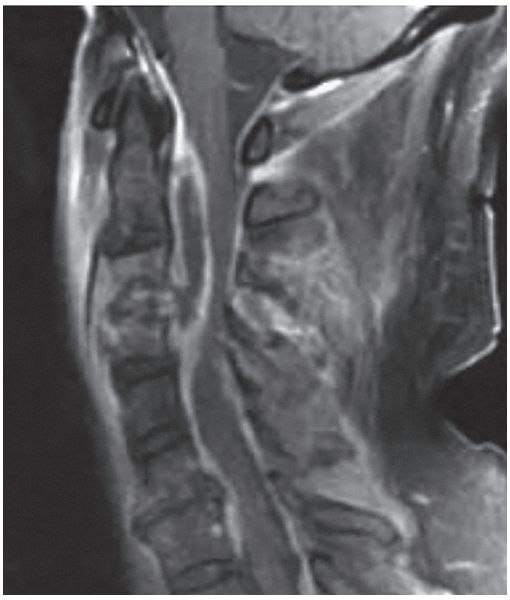

Fig. 5.20 MRI demonstrating anterior cervical epidural abscess following C3/4 discitis and osteomyelitis in C3 and C4, causing severe cord compression and tetraparesis.

Once the diagnosis is confirmed, urgent evacuation of the abscess is mandatory.

Noncompressive Myelopathies

Spinal cord dysfunction can result from noncompressive myelopathy that could be broadly grouped into inflammatory and non-inflammatory causes. The inflammatory group includes transverse myelitis, infections, and demyelinating and vasculitic diseases.[29] The noninflammatory groups comprise vascular, toxic and physical agents as well as degenerative, metabolic and inherited myelopathies.[30] Also, noncompressive myelopathies are clinically characterized by patterns of selective involvement of different anatomical structures of the spinal cord, and these patterns help the etiological diagnosis. Some of the classical syndromes with their most common causes are: Complete spinal cord syndrome (e.g., transverse myelitis), Brown–Sequard syndrome (e.g., multiple sclerosis), anterior spinal cord syndrome (e.g., anterior spinal artery infarct), posterolateral cord syndrome (e.g., vitamin B12 deficiency), central cord syndrome (e.g., neuromyelitis optica), and posterior syndrome (e.g., posterior spinal artery infarct and tractopathies or ALS).

Noncompressive myelopathy affects people across all age groups with a clear preponderance on patients in their middle age. Diagnosis of these conditions can be difficult, and it can be elusive in up to 1/3rd of the patients. Diagnostic tools to help to distinguish these syndromes include imaging (bone scan, MRI, fluorodeoxyglucose-positron emission tomography [FDG-PET]), CSF evaluation, blood cultures, and electromyography (EMG) studies.[31,32]

Transverse myelitis (TM) is an inflammation of the spinal cord, a major part of the CNS. The spinal cord carries nerve signals to and from the brain through nerves that extend from each side of the spinal cord and connect to nerves elsewhere in the body.

The term myelitis refers to inflammation of the spinal cord; transverse refers to the pattern of changes in sensation—there is often a band-like sensation across the trunk of the body, with sensory changes below.[29]

Causes of TM include infections, immune system disorders, and other disorders that may damage or destroy myelin, the fatty white insulating substance that covers nerve cell fibers. Inflammation within the spinal cord interrupts communications between nerve fibers in the spinal cord and the rest of the body, affecting sensation and nerve signaling below the injury. Symptoms include pain, sensory problems, weakness in the legs and possibly the arms, and bladder and bowel problems. The symptoms may develop suddenly (over a period of hours) or over days or weeks. Diagnosis of TM includes MRI (**Fig. 5.21**), blood tests looking for autoantibodies (antiaquaporin-4, antimyelin oligodendrocyte), and a host of antibodies associated with cancer (paraneoplastic antibodies), which may be found in people with TM and lumbar puncture, wherein CSF contains more protein than usual and an increased number of white blood cells among some people. A lumbar puncture also helps out to rule other infectious diseases.[32] Initial cases might render normal results through investigations; in such cases, repeating the MRI and CSF in 5 to 7 days is recommended. Treatment includes steroids, plasmapheresis, and immunoglobulins. Recovery can be long.

Spinal cord infarction usually results from ischemia and originates in an extravertebral artery. The first symptom of spinal cord infarction is usually sudden pain in the neck or the back, with tightness radiating circumferentially, followed within minutes by segmental bilateral flaccid weakness and sensory loss. Pain and temperature sensation are disproportionately impaired. Position and vibration sensation, conducted by the posterior columns, and often light touch are relatively spared.

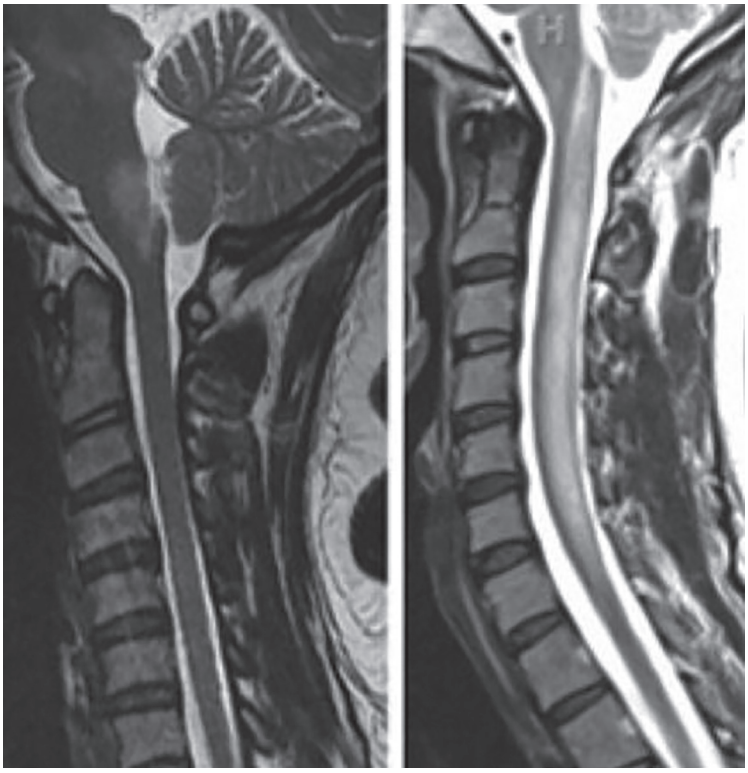

Fig. 5.21 MRI showing a case of transverse myelitis; please note the severe swelling of the spinal cord, affecting the entire cervical spine.

The main blood supply to the posterior third of the spinal cord is by two posterior spinal arteries; for the anterior 2/3 of the spinal cord, the anterior spinal artery is the only supply. The anterior spinal artery has only a few feeders from the cervical region and one large feeder in the lower thoracic region (artery of Adamkiewicz). All feeders originate from the aorta. Injury to an extravertebral feeder artery or the aorta (e.g., due to atherosclerosis, dissection, or clamping during surgery) causes infarction more commonly than intrinsic disorders of spinal arteries. Thrombosis is an uncommon cause, and polyarteritis nodosa is a rare cause.

Diagnosis of spinal cord infarction is conducted by good history taking and confirmed with an MRI (**Fig. 5.22**). If MRI is unavailable, CT myelography can be done.

The causes of Vitamin B12 and folate deficiency in author's patients were a combination of strict vegetarian diet, malnutrition, alcohol consumption, and phenytoin therapy. Vitamin B12 deficiency can also lead to symptoms similar to CSM, including sensory and motor deficiencies and gait ataxia. Deep tendon reflexes are usually absent or severely diminished, whereas pathological reflexes (Babinski sign) are present. Usually, these neurological findings are present with symptoms of dementia and/or other psychiatric symptoms. Patients with a history of pernicious anemia or gastrointestinal abnormalities who present with symptoms of gait ataxia and motor or sensory deficits should have a high index of suspicion for Vitamin B12 deficiency.[33] The patients afflicted with hepatic myelopathy had decompensated alcoholic liver disease with encephalopathy. The cause of spinal cord dysfunction could be multifactorial (toxic effect of adulterated alcohol, nutritional deficiency, nitrogenous breakdown products bypassing the liver through shunt, and venous hypertensive myelopathy).[34]

Spinal cord sarcoidosis usually affects the cervicodorsal cord. MRI of the spinal cord may be normal or show linear signal abnormality on T2-weighted imaging associated with patchy gadolinium enhancement, cord swelling with T2 hyperintensity without enhancement, subpial enhancement, or thickening with enhancement of the cauda equina. Isolated spinal cord sarcoidosis is extremely rare.[31] Serum angiotensin-converting enzyme (ACE) level is raised in only 50% of the patients. CSF ACE is less sensitive but relatively specific (94–95%) for CNS sarcoidosis.[35]

Multiple sclerosis (MS) is an autoimmune disease in which the body´s immune system damages the myelin coating around the nerve fibers in the CNS and the nerve fibers themselves, interfering with the transmission

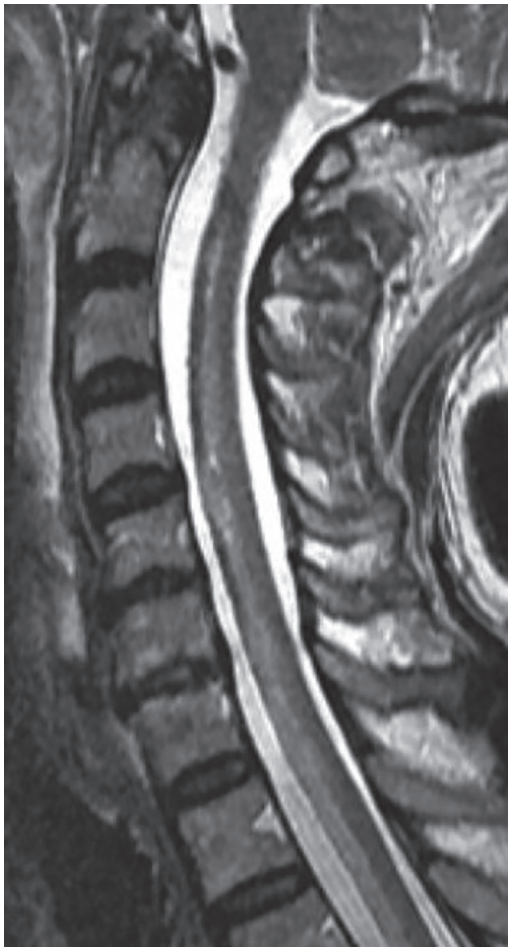

Fig. 5.22 Spinal cord infarction. Anterior spinal cord syndrome. Stroke on the anterior spinal artery territory causing flaccid tetraparesis.

of the nerve signals between the brain, spinal cord, and the rest of the body. MS is most commonly diagnosed in females aged between 20 to 40, but may occur at any age and among both genders, and contrary to ALS, often progresses slowly over many years (about 25 years). Patients with MS display relapsing symptoms involving the white matter tracts and, similarly to CSM, may present with L'hermitte phenomena, and motor, sensory, and bladder/bowel dysfunction.[33] Often, they show other symptoms that could help to distinguish CSM (visual defects, cognitive problems, epileptic fits).[33]

The diagnosis of MS usually involves a neurologist who will take the medical history, do blood tests, conduct tests to measure electrical activity in the brain and other areas, carry out an MRI (**Fig. 5.23**), and perform an analysis of CSF.

ALS, also known as motor neurone disease (MND) or Lou Gehrig's disease, is a neurodegenerative disease that causes death through gradual deterioration of neurons controlling voluntary muscles. Motor neurons are nerve cells that extend from the brain to the spinal cord and to muscles throughout the body. In ALS, both the upper motor neurons and the lower motor neurons degenerate or die and stop sending messages to the muscles. Unable to function, the muscles gradually weaken, start to twitch (fasciculations), and waste away (atrophy). Eventually, the brain loses its ability to initiate and control voluntary movements. Most people with ALS die from respiratory failure, usually within 3 to 5 years from when the symptoms first appear. However, about 10 percent of people with ALS survive for 10 or more years.

Electrophysiology tests are best to differentiate between ALS and CSM. Measuring motor-evoked potentials (MEP) from the trapezius muscle was found to be abnormal in all patients with ALS but normal in patients with CSM, whereas abnormal MEP´s were found in all limbs in both CSM and ALS.[36] Furthermore, EMG resulted abnormal for both upper and lower limbs in patients with ALS-like symptoms, compared to abnormal EMG findings in only the upper limbs

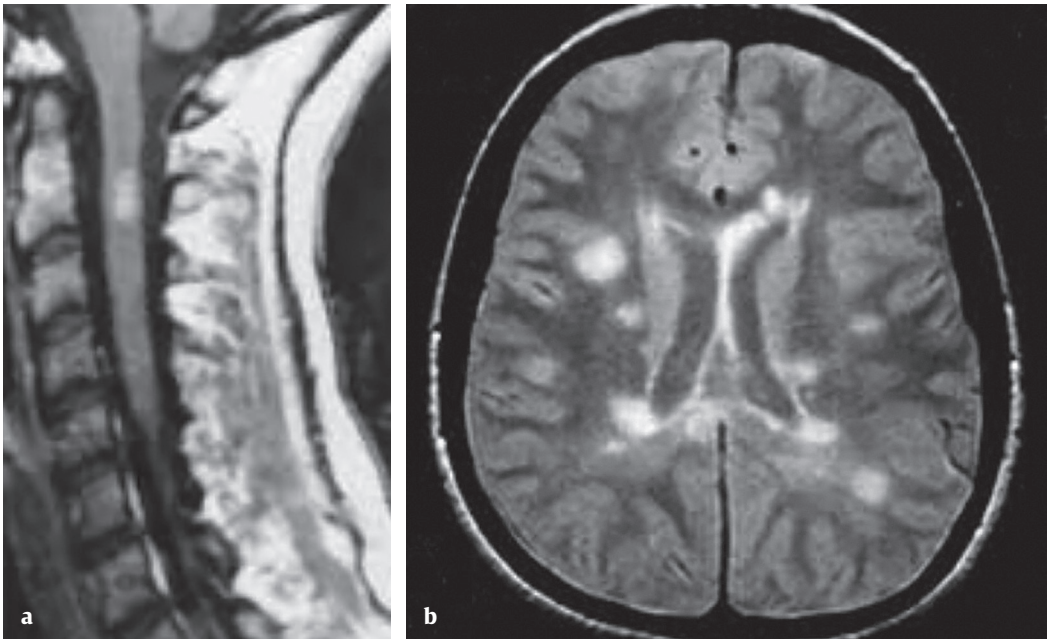

Fig. 5.23 Cervical and brain MRI showing some features of multiple sclerosis (MS). Typical MS plaque in the cervical spinal cord **(a)**. Normally, it is accompanied with multiple plaques in the brain **(b)**.

on the CSM group.[37] Another study recorded responses of masseter muscles to transcranial magnetic stimulation and found that the activation of corticobulbar descending fibers was absent or delayed in the majority of patients with ALS but normal in all patients with CSM, providing a means of distinguishing the two conditions.[38] CSF analysis can also help in the diagnosis of ALS.[32] The best biomarkers for distinguishing ALS from other neurodegenerative diseases and healthy subjects are erythropoietin (decreased),[39] hepatocyte growth factor (upregulated), monocytic chemotactic protein (increased),[40] neurofilament light and heavy subunits (upregulated), and cystatin C and transthyretin (reduced).[41]

Neuromyelitis optica (NMO) is an autoimmune, inflammatory, and demyelinating disorder of the CNS, with a predilection for the optic nerves and spinal cord often resulting in permanent blindness and/or paralysis and is characterized by the presence of serum aquaporin-4 immunoglobulin G antibodies (AQP4-IgG).[42] The pathological features of NMO include perivascular deposition of immunoglobulin and activated complement, loss of astrocytic AQP4, inflammatory infiltration with granulocyte and macrophage accumulation, and demyelination with axon loss. Immunosuppression and plasma exchange are the mainstays of therapy for NMO optic neuritis.

Syringomyelia refers to a disorder in which abnormal fluid-filled cavities or cysts form in the spinal cord.[43,44] This syrinx can get bigger and elongate over time, damaging the spinal cord and compressing and injuring the nerve fibers that carry information to the brain and from the brain to the rest of the body. The symptoms begin earlier than those of CSM, but, like CSM, its onset is insidious and progresses irregularly.[44] Symptoms include sensory loss, areflexic weakness and atrophy in the upper limb, leg spasticity, bladder and bowel dysfunction, and Horner syndrome.[44] The best diagnostic tool is an MRI (**Fig. 5.24**).

There are two major forms of syringomyelia:

Congenital syringomyelia (communicating syringomyelia): In most cases, syringomyelia is caused by a Chiari malformation, which may allow a syrinx to develop, mostly in the cervical region. Straining or coughing can force CSF into the ventricles, causing the person to develop headache or even lose consciousness (so-called cough syncope).

Acquired syringomyelia (noncommunicating syringomyelia): Causes of acquired syringomyelia include spinal cord injury, meningitis, arachnoiditis after intradural surgical operations, tethered cord syndrome, a spinal cord tumor, and bleeding into the cord (hemorrhage).

Vascular pathologies that have to be on the differential diagnosis of CSM include dural arteriovenous fistulas (DAVFs) and arterial venous malformations (AVMs).

Fistula means abnormal connection between two structures that are normally

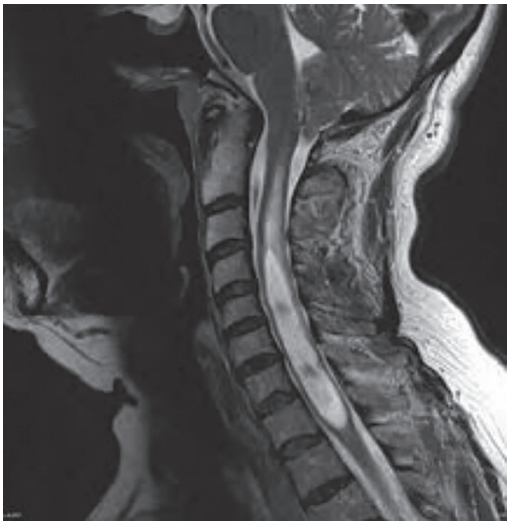

Fig. 5.24 MRI showing syringomyelia and Chiari malformation. Syrinx is a fluid-like cavity caused by the reopening of the congenital central canal, which is filled by cerebrospinal fluid (CSF) and expands the spinal cord, compressing the neuropathways and causing myelopathic symptoms.

not connected. An arteriovenous fistula is therefore an abnormal connection between an artery and a vein. When a fistula forms, blood from an artery under high pressure and flow goes directly into a vein, which is a low pressure and low flow structure. Even though the dural fistula is usually not directly on or within the spinal cord, it nevertheless causes dysfunction by interfering with normal spinal cord circulation, eventually producing severe and sometimes irreversible problems.[45] Spinal DAVFs are the spinal vascular malformations that are encountered most often, and they are usually encountered in the lower thoracic region. Cervical spine DAVFs are exceedingly rare and may be difficult to differentiate from radicular AVMs, epidural arteriovenous shunts, or perimedullary AVFs. Treatment includes embolization, surgical disconnection, or combined.[46,47]

Spinal AVMs **(Fig. 5.25)** are abnormal collections of blood vessels in the spinal canal that have a direct connection between the arterial system and the venous system without intervening capillaries.[48] Most of the AVMs will produce progressive neurological symptoms over months to years, especially back pain associated with progressive sensory loss and lower extremity weakness. They can be classified as: DAVF, the most common type; intradural AVMs, which are located between the spinal cord and the dura; and intramedullary AVMs, which are those located within the spinal cord. Diagnosis is made with MRI and spinal angiography, and treatment, as in DAVF, can be achieved through embolization, surgical excision, or a combination of both. Radiosurgery has also been used for some forms of AVM with promising results.

Conclusion

Physical examination findings are not always consistent with severity of disease in CSM, therefore, familiarizing with the clinical tests as an adjuvant to patient examination and correlating those findings with appropriate imaging are essential for arriving at the correct diagnosis. An MRI result is crucial in helping to distinguish between compressive and noncompressive myelopathic pathologies. In some cases where these imaging studies are still equivocal, use of other studies should be considered including electrodiagnostic studies as well as CSF examination.

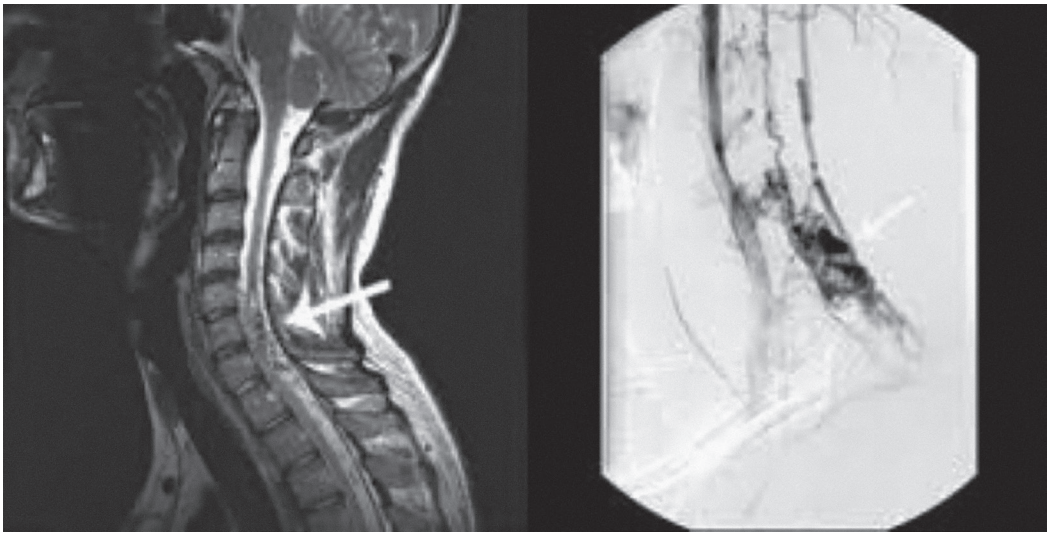

Fig. 5.25 Cervical AVM. Left: MRI demonstrating (arrow) a spinal arteriovenous malformation (AVM) with its classical "bag of worms" appearance. Right: Spinal angiography demonstrating the anatomical characteristics of the AVM.

Take-home Points

- Clinical tests are important tools to compliment a good anamnesis and neurological examination.
- The clinical tests mainly help to differentiate between CSM and radiculopathy.
- Clinical tests are cheap and easily done in the physician consulting rooms.
- MRI is an invaluable tool in the diagnosis of CSM together with plain X-rays (anteroposterior, lateral, and dynamic).
- Patients with bilateral sensory complaints in the hands should be presumed to have cervical cord pathology and should have cervical MR imaging, even if the EMG/NCS suggests bilateral carpal tunnel syndrome.
- Differential diagnosis in CSM can sometimes be extremely different, as many related pathologies can be present at the same time.
- ALS is distinct from CSM in the presence of cranial nerve involvement (CN XI) and the absence of pain or sensory changes.
- Electrodiagnostic studies demonstrate distinct characteristics in ALS and CSM.
- MS and NMO can be distinguished from CSM in the presence of visual symptomatology as well as the patient demographic is much younger.
- A good clinical examination together with a thoughtful investigation selection and good cooperation with our neurologist will help us to diagnose virtually all cases with myelopathy, providing patients with a professional and speedy solution to their problems.

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