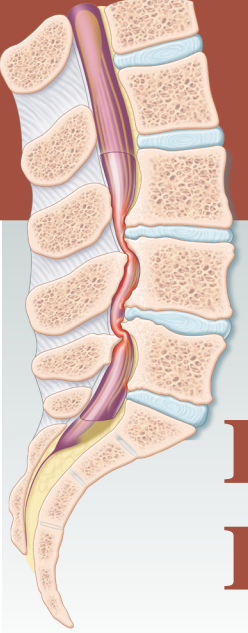

Primary Care Approach to Degenerative Cervical Myelopathy

ABSTRACT

Degenerative cervical myelopathy is an umbrella term describing all degenerative conditions that present with cervical myelopathy due to compression of the spinal cord. The role of primary care physicians (PCPs) in early identification is vital as delayed diagnosis can lead to irreversible neurological impairment. Patients often present with subtle neurological deficits associated with neck or upper extremity pain. Screening for upper motor neuron signs, gait disturbances, fine motor abnormalities and bowel bladder symptoms is critical. Currently, surgical decompression is the treatment of choice but with future advancements in non-operative treatments, PCPs are expected to play a larger role in treatment plans.

KEYWORDS: degenerative cervical myelopathy, primary care, cervical spondylotic myelopathy, degenerative disc disease

CME Pre-test Quiz

Introduction

Degenerative cervical myelopathy (DCM) is an umbrella term that encompasses a variety of degenerative pathologies characterized by compression of the cervical spinal cord resulting in progressive injury to neural tissue (Figure 1).[1] DCM is one of the most common causes of non-traumatic spinal cord impairment in adults worldwide.[2,3] The management of DCM has advanced in the last decade and a structured approach to patient care is expected to enhance outcomes.[4,5]

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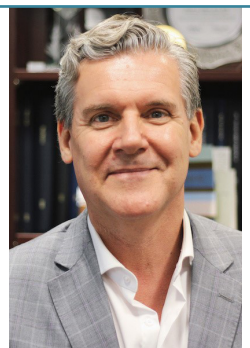

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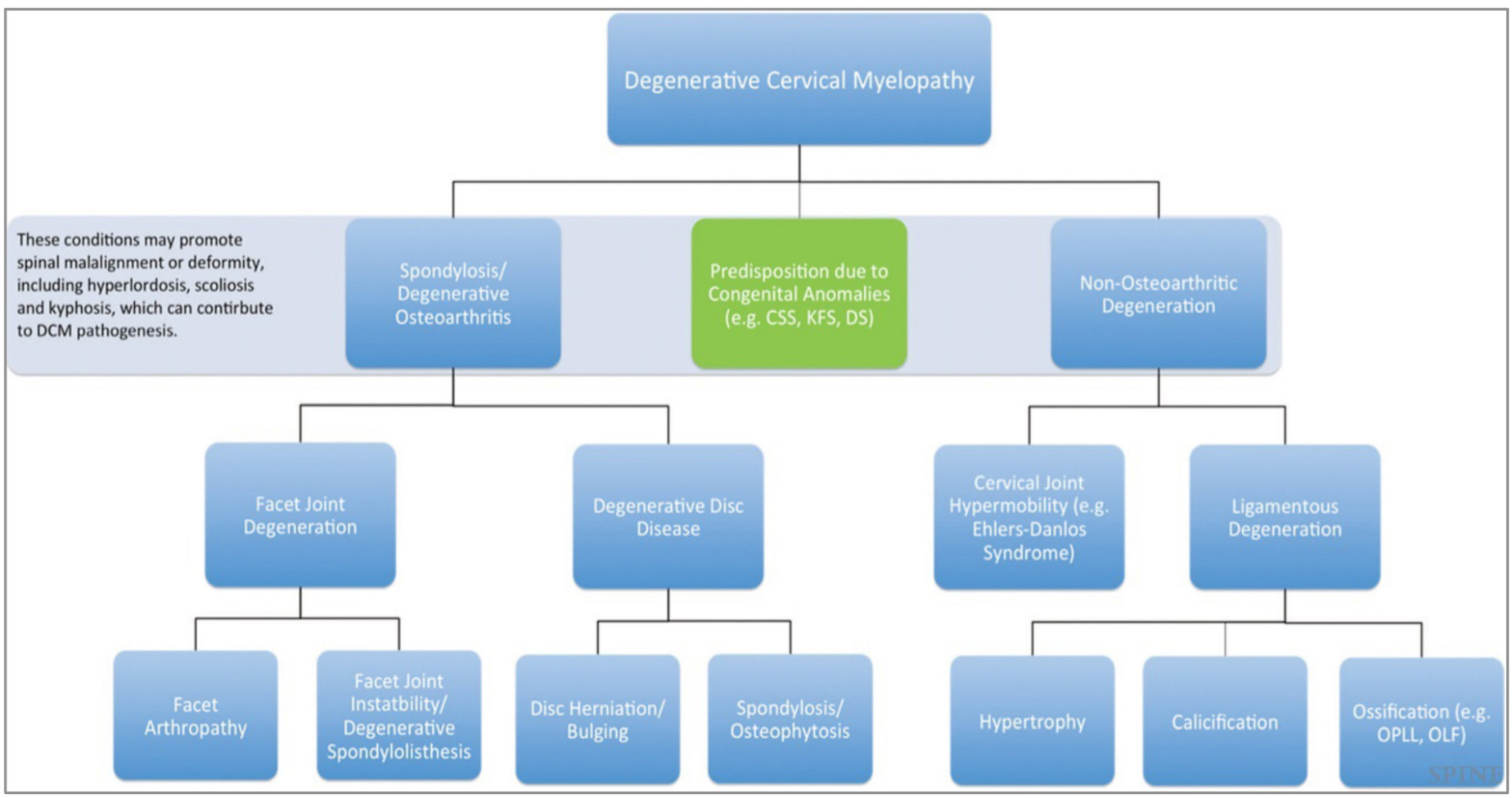
Figure 1: Breakdown of Conditions Classified under Degenerative Cervical Myelopathy



CSS: congenital spinal stenosis. KFS: Klippel-Feil syndrome. DS: Down Syndrome

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One of the most important aspects in the care of DCM is early diagnosis as delayed treatment can result in irreversible neurological impairment. This highlights the essential role of primary care providers (PCPs) who are often the first to encounter a patient with DCM. Unfortunately, a delay in referral for surgical treatment is partly due to a late diagnosis of DCM in the primary care setting.[6,7] There is a critical need to maintain a high index of suspicion for the recognition of DCM among primary care practitioners. Although the necessity for urgent diagnosis is very much less, degenerative cervical myelopathy, like cauda equina syndrome, can result in permanent neurological impairment and should have the same level of recognition. In this article, we aim to provide a summary of the appropriate diagnosis and management of DCM in primary care.

Diagnosis

The diagnosis of DCM requires clinical signs and symptoms of myelopathy coupled with radiological evidence of cervical cord compression. Patients presenting with neck and upper limb pain associated with difficulty walking should be assessed for DCM. Clinical screening means inquiring about focal neurologi-

cal deficits, gait abnormalities, fine motor control problems as well as bowel and bladder incontinence. While it is uncommon, degenerative cervical myelopathy can present without neck or arm pain. Ideally, DCM should be suspected on clinical presentation and confirmed on imaging as opposed to the reverse.[8] The early clinical signs of DCM are often subtle. Physical exam findings can include hyperreflexia, clonus, muscle wasting, focal motor deficits, sensory deficits, gait ataxia, rigidity, the Spurling test, and Hoffman's and Babinski's signs. Table 1 gives a summary of the signs and symptoms of DCM. The modified Japanese Orthopedic Association (mJOA) scale is commonly utilized as a measure of severity in DCM research (Figure 2).[9] The mJOA scale is a

Table 1: Signs and symptoms of Degenerative Cervical Myelopathy (DCM)

Clinical signs of DCM on history (Estimated Prevalence)	
Upper limb pain (80%)	Often the presenting complaint
Neck pain (60%)	Often the presenting complaint with a broad differential diagnosis.
Focal neurological signs (62%)	This includes weakness or any sensory dysfunction. Unlike radiculopathy, a specific dermatome or myotome cannot always be identified.
Sphincter dysfunction (17%)	Bowel and bladder incontinence. This is often a late finding.
Gait disturbances (67%)	Patients present with instability. Screening questions could inquire whether patients require use of handrail when going up the stairs.
Problems with hand dexterity (62%)	Fine motor control problems can help narrow the differential diagnosis. Screening questions can inquire about ability to button up a shirt, open a jar or use kitchen utensils.
Physical Exam findings	
Hoffman signs	Flexion of index finger or thumb on flicking of the nails of the ring or middle finger.
Spurling test	Pain in upper extremities with axial compression of neck in extension
Babinski Sign	Extensions and fanning of the great toe in response to stroking of the lateral sole of the foot.
Clonus	Repeated muscular spasms: sustained clonus is defined as three or more beats.
Spasticity	Velocity dependent increase in muscle tone with hyperexcitability of the stretch reflex
Gait disturbance	Difficulties with tandem gait
Upper extremity range of motion	DCM can often present with a decrease in neck ROM
Cranial nerves	DCM should not result in any cranial nerve abnormalities. This test should be done as part of the differential diagnosis.

Figure 2: The modified Japanese Orthopedic Association (mJOA)

Category	Score	Description
Upper Extremity Motor Subscore (/5)	0	Unable to move hands
	1	Unable to eat with a spoon but able to move hands
	2	Unable to button a shirt but able to eat with a spoon
	3	Able to button a shirt with great difficulty
	4	Able to button a shirt with mild difficult OR other mild fine motor dysfunction (marked handwriting change, frequent dropping of objects, difficult clasping jewelry, etc.)
	5	Normal hand coordination
Lower Extremity Subscore (/7)	0	Complete loss of movement and sensation
	1	Complete loss of movement, some sensation present
	2	Inability to walk but some movement
	3	Able to walk on flat ground with walking aid
	4	Able to walk without walking aid, but must hold a handrail on stairs
	5	Moderate to severe walking imbalance but able to perform stairs without handrail
	6	Mild imbalance when standing OR walking
	7	Normal walking
Upper Extremity Sensory Subscore (/3)	0	Complete loss of hand sensation
	1	Severe loss of hand sensation OR pain
	2	Mild loss of hand sensation
	3	Normal hand sensation
Urinary Function Subscore (/3)	0	Inability to urinate voluntarily (requiring catheterization)
	1	Frequent urinary incontinence (more than once per month)
	2	Urinary urgency OR occasional stress incontinence (less than once per month)
	3	Normal urinary function

The mJOA is an 18 point score of functional disability specific to cervical myelopathy, including upper extremity motor subscore, lower extremity subscore, upper extremity sensory subscore, and sphincter function. The descriptions of each score are modified slightly from Benzel et al. (1991).[2]

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clinically administered tool that evaluates four functional domains: upper limb motor, lower limb motor, upper limb sensory, and sphincter. The domains of the mJOA scale can act as a guide for the clinical assessment of patients with suspected DCM.

One of the challenges in the diagnosis of DCM is its broad differential diagnosis. This can include demyelinating/inflammatory conditions such as multiple sclerosis or transverse myelitis, cervical radiculopathy, peripheral nerve pathologies such as diabetic neuropathy and carpal tunnel, amyotrophic lateral sclerosis (ALS), or structural brain pathologies such as stroke or tumors. In particular, DCM should always be considered as the underlying diagnosis for patients presenting with symptoms of bilateral hand numbness, pain or motor dysfunction. If DCM is suspected, the appropriate next step is to acquire neural imaging. If diagnostic uncertainties remain after the imaging, an urgent referral to a neurologist is appropriate. A detailed neurological exam and history coupled with nerve conduction studies or electrophysiology tests can resolve diagnostic uncertainties. Advancements in ancillary testing and personalized medicine should lead to enhanced clinical tools for the diagnosis of DCM.[4]

Imaging evidence of spinal cord compression is required in the diagnosis and management. Patients with a high level of suspicion for DCM require an MRI. For those that have contraindications to an MRI, a CT myelogram can be obtained. Cord edema characterized by T2 hyperintensity within the cord, while suggestive of DCM, is not a requirement.[10] The degree of compression on a static MRI is not necessarily correlated with disease severity and any degenerative deformation of the cord can produce dynamic spinal cord compression, which may not be visible in a standard static MRI.[11] Emerging techniques that assess the microstructural anatomy of the cord (such as T2*-weighted imaging) can provide further clarity.[12] CT and/or dynamic X-ray imaging are necessary for operative planning.

Treatment

The treatment decision for DCM is largely dependent on the severity of the disease. The most recent clinical practice guidelines from the AO Spine group recommend surgery for those with severe (mJOA <12) and moderate (mJOA 12-14) disease.[13] The main surgical goal is to halt progression, however many patients with moderate or severe symptoms will also improve to some degree. The efficacy of surgical decompression for DCM is well established in the literature.[9,14] Given our lack of understanding of the natural history of those with mild DCM (mJOA 15-17) without progression, the current guidelines recognize clinical equipoise between surgery

and a structured non-operative approach to management. The recommendations favor an operative intervention for mild patients who exhibit any signs of clinical progression.

Structured non-operative treatments for DCM can be offered for all patients as an adjuvant to surgical decompression or as the main treatment in mild cases under surveillance. Various approaches to non-operative treatments for DCM are documented in the literature, with neck therapy to relieve pain and restore function being the most common.[15] However, there is no consensus as to the best therapy and different modalities such as posture training, balance training, core stability, or electrotherapy are all recommended. The choice of non-operative care will usually be dependent on patient preference and available local resources at each center.[16–19] Referral to an experienced physiotherapist or physiatrist is recommended. Although not currently accepted treatments, non-operative medical approaches centered on emerging neuroprotective and neuroregenerative therapies such as riluzole,[20] mesenchymal stem cells,[21] and protein injections,[22,23] are expected to play a larger role in the future management of patients with DCM. This expansion of medical treatment should expand the role of primary care.

Surveillance and post-operative care

After the diagnosis of DCM, perhaps the most important role for the primary care clinician is the surveillance of patients for neurological worsening. Patients should be fully educated and examined at regular intervals. This can either be to detect disease progression in mild DCM patients who are being monitored or to detect recurrence of the symptoms in previously operated patients. With neurological progression, patients can become candidates for surgical decompression. Referral to the spinal surgeon is required. It is estimated that, over a 3-7 year time span from the time of diagnosis, 20-60% of patients will deteriorate.[24–26] Patients who have already had decompression need to be monitored for further degeneration at adjacent cervical motion segments. Early disease progression can be subtle and we recommend a detailed neurological exam with a focused functional history. Ideally, PCPs' clinical assessment for neurological progression should be done routinely in patients with known DCM. Serial MRI scans in isolation, without clinical assessment, should not be used. With expected advancements of multiparametric quantitative MRIs, serial MRIs could potentially be

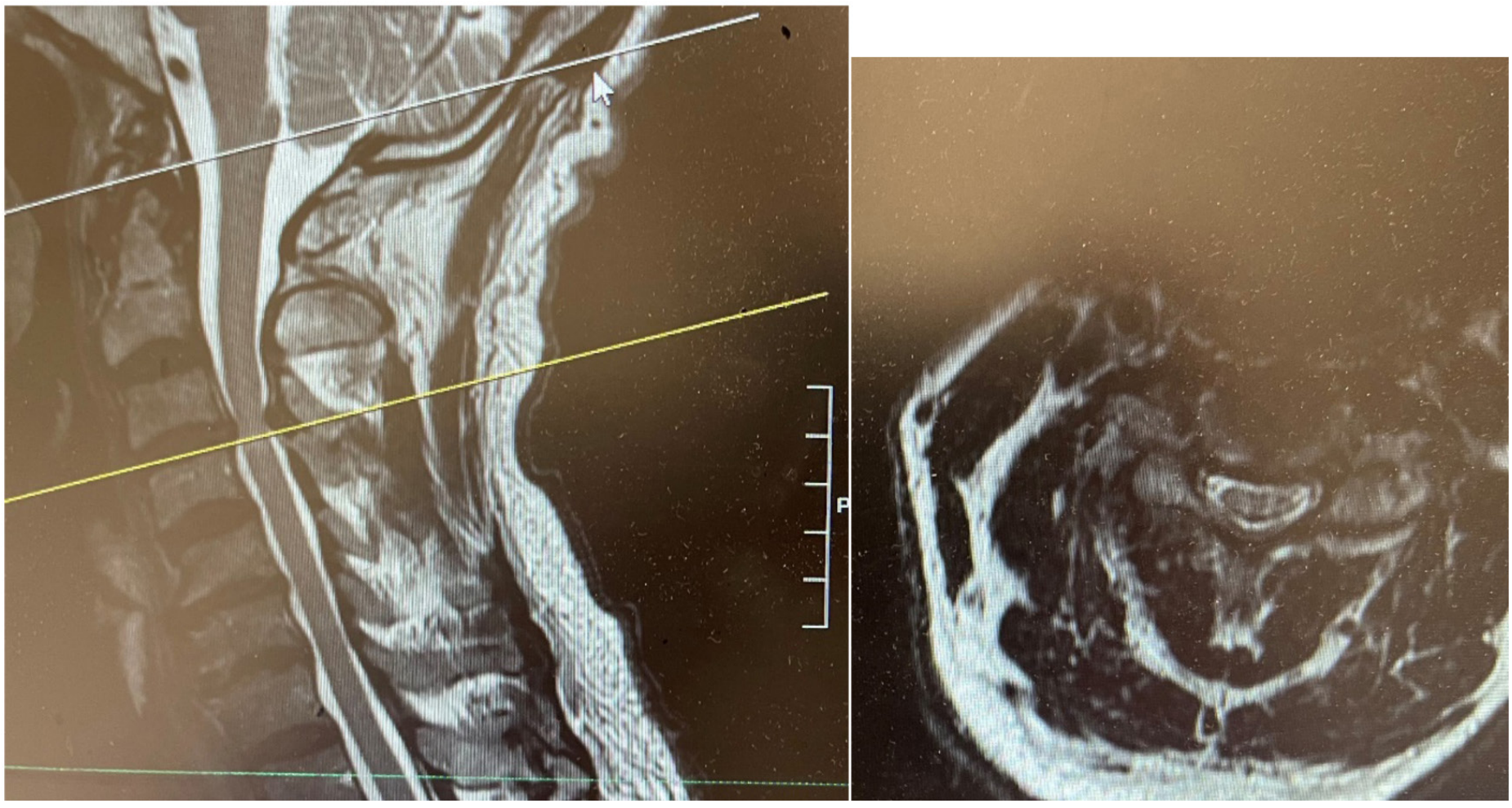
adopted to confirm neurological worsening.[27]

In addition to surveillance, physiotherapy and analgesia are often part of the routine for post-operative care of DCM patients. The efficacy of post-operative physiotherapy is a topic of active research with several registered clinical trials.[28] Currently, it is recommended to employ active physiotherapy as part of post-operative care.[5] The goal of physiotherapy is to improve flexibility, range of motion, fine mother control and gait. Strengthening of the deep cervical muscles aids posture. Pain management of DCM patients can often be challenging, particularly given multiple comorbidities and other degenerative pathologies. Referral to a pain specialist may be warranted in the long-term management. Optimal analgesia should be pre-emptive, multi-modal, and opiate sparing.[29–31]

Typical Case

56-year-old right handed accountant presenting with a 6-month history of neck pain. He has tried physiotherapy once a week as well as ibuprofen with some alleviation of his pain. On further questioning he reveals that over the last three months he has noticed numbness in both hands. He also reports that he has difficulty buttoning up his shirt but is able to use utensils for eating. He does not report any recent falls but indicates that he requires holding on to the hand rail when going up the stairs. He does not report any urinary or bowel symptoms. He does not report any leg or back pain. He does not report

Figure 3:



SUMMARY OF KEY POINTS

PCPs play a vital role in the management of DCM as a delayed diagnosis can lead to irreversible neurological impairment

A heightened level of awareness with a comprehensive history and a focused physical examination are essential.

With advancements in biomarkers and emerging neuroprotective and regenerative agents, we can expect an increased role in the primary care medical management of DCM patients soon.

The approach to DCM management is multidisciplinary and generally will involve PCPs, spinal surgeon, physiotherapist, pain specialist, and neurologist.

any recent weight loss. He has no history of trauma. No recent infections or history of fever or chills. He is otherwise well and is on levothyroxine for hypothyroidism.

On physical examination he does have numbness as well as altered sensation to light touch in the hand bilaterally in no specific dermatomal distribution. He has negative Hoffman reflex. He is hyper reflexia at C5 and C6 bilaterally. He has a downgoing plantar reflex with normal reflexes in the patellar and Achilles tendon. He has 5/5 power in all the major muscle groups of the upper and lower extremity. On gait assessment he is noted to have difficulty with tandem gait. He has normal cranial nerve examination. Cerebellar function is normal.

Given concerns for degenerative cervical myelopathy he was sent for urgent outpatient MRI of the cervical spine. The MRI (Figure 3) reveals central broad based disc protrusion at C3-4 with effacement of the cervical spinal cord and T2 hype-intensities within the spinal cord at that level.

An urgent referral was sent to a spinal surgeon. Patient was seen and confirmed to have moderate

CLINICAL PEARLS

Patients with query bilateral carpal tunnel syndrome should be assessed for DCM.

Patients with moderate to severe DCM or unequivocal progression of mild DCM require surgical treatment while there exists clinical equipoise between structured non-operative therapies and surgical decompression for mild non-progressive cases of DCM.

Clinically monitor patients with mild DCM frequently and carefully for subtle signs of neurological progression

CME

Post-test Quiz

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DCM (mJOA 13). Given the natural history of his condition he was offered surgical decompression via an anterior discectomy and fusion (ACDF) at C3- C4 level. The indications quoted for the surgery were to halt disease progression. Risks of surgery quoted were remote chance of significant neurological impairment or death. Other risks such as infection, CSF leak, adjacent segment disease, swallowing difficulties and changes in voice were also explained. The patient agreed to proceed and was booked for a 1-level ACDF with pre-operative CT scan for operative planning.

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