Cervical Spondylotic Myelopathy, Be Vigilant of Miscellaneous Presentations: A Case Series

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Abstract

Background: Cervical spondylotic myelopathy (CSM) is the result of prolonged compression of the central spinal canal. Clinical and radiological manifestations of CSM could be misleading and physicians need to be vigilant while evaluating patients with neurologic complaints otherwise delay in diagnosis and management would happen that could finally lead to a poor outcome. The aim of this study was to report our series of patients with CSM to reiterate that CSM could have diverse manifestations, and it can be only identified if the physicians are aware and do a thorough investigation. The progressive nature of CSM makes its early diagnosis and subsequent appropriate management vital to avoid further complications.

Methods: A total of 10 patients who had diagnosed with CSM and undergone surgical treatment at our institute evaluated according to the pre-operative and post-operative Nurrick classification and also pre-operative history and physical examination findings and elecrodiagnostic examination.

Conclusions: Because of progressive nature CSM and wide range of clinical manifestation, high index of suspicion, early diagnosis and early treatment prior to permanent spinal cord injury is recommended.

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Introduction

Cervical spondylotic myelopathy (CSM) is the result of prolonged compression of central spinal canal. Symptoms of CSM including gait disturbances, difficulty in walking, loss of fine motor function, sensory deficit in extremities and bowel and bladder dysfunction (1-3). Myelopathy is the most common cause of spinal cord dysfunction in patients over 55 (1,3). Symptoms of CSM usually presents in the 6th decade of life, which can be different on an individual basis and may be quite subtle and stable for a long time (4-8). The classical findings in the physical examination are upper motor neuron abnormalities like Hoffman and Babinski sign in addition to interosseous muscles atrophy and myelopathy symptoms such as small finger scrape test and positive grasp-release test in the hand. Some patients may report the Lhermitt's sign, i.e., the "electric shock"-like sensation in the extremities and trunk generated by neck flexion (4-8).

However, there are many patients with CSM, who are asymptomatic and have normal physical examination. On the other hand, patients with CSM could be presented with subtle symptoms in areas other than neck (4). In addition, though the natural history of CSM usually follow the stepwise progression over the years, catastrophic quadriparesis can be developed within hours in 5% of patients with CSM (7). Therefore, proper diagnosis and treatment of CSM are required to prevent the further disastrous complications.

The aim of this study was to report our series of patients with CSM to reiterate that CSM could have diverse manifestations, and it can be only identified if the physicians are aware and do a thorough investigation. The progressive nature of CSM makes its early diagnosis and subsequent appropriate management vital to avoid further complications.

Materials and Methods

From April 2013 to June 2015, we studied retrospectively 10 patients who had diagnosed with CSM and undergone surgical treatment at our institute. These patients had been undergoing mistreatment due to misdiagnosis when they referred to our clinic. We evaluated them according to the pre-operative and post-operative Nurrick classification and also pre-operative history and physical examination findings and elecrodiagnostic examination. Details of all patients have been demonstrated in table 1.

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Table 1. Characteristics of our patients with CSM

Case	Age	Sex	Comorbidity	Early clinical presentation	EMG-NCV	Misdiagnosis	MRI finding	Physical finding before operation	Nurrick
1	33	Male	Non	Neck pain-both non-radicular pain	normal	Axial pain	Sever stenosis C4-C5 + signal change	Motor weakness mp = 3-4/5 in upper ext. Hofmann	1
2	75	Male	IHD	Arm pain, gait instability, paresthesia in upper extremity, motor weakness, interossei muscles atrophy	Chronic radiculopathy C5, C6, C7	Shoulder rotator calf tear, CTS	Multilevel stenosis C3-C7	Myelopayhy hand symptoms, weakness of upper and lower ext., Hofmann, Babinski	4
3	48	Male	IHD, DM, psoriasis	Gait disturbance, progressive upper extremity weakness	Mild C5, C6 radiculopathy	Symptoms related to psoriasis and aging	Sever stenosis at C3-C4, signal change	Unstable gaite, urine incontinence, myelopathy hand, pathologiv reflexes Hofmann	5
4	59	Female	Non	Subtle weakness in hand	C5, C6, mild radiculopathy without active denervation	CTS	Central canal stenosis, C5-C6, C6-C7, signal change	Motor weakness 4/5 in upper extremity, Hofmann	2
5	65	Male	Non	Gait disturbance	Multiple root radiculopathy	CVA, psychsomatic disorder	Diffuse cervical stenosis C4-C7	Motor weakness, pathologic reflex, gait instability	4
6	60	Male	Non	Gait disturbance, hand clumsiness	Chronic C6, C7 radiculopathy	Age related disease, CTS	Diffuse cervical stenosis C4-C6	Motor weakness, pathologic reflexes, gait instability	4
7	57	Female	Non	Occationally neck and arm pain	Mild C6, C7 radiculopathy	Axial pain	Diffuse cervical stenosis C4-C7, cervical kyphosis	Subtle Motor weakness 4/5, pathologic Hofmann	1
8	74	Male	Non	Gait disturbance	Multiple root involvement	OA of knee, age related disease	Cervical stenosis C4-C6, cervical kyphosis	Motor weakness in upper and lower ext., Hofmann, flat plantar reflex	3
9	58	Male	DM	Weakness and paresthesia of hands	C5, C6, C7 sensory polyneuropathy	Diabetic poly neuropathy	Cervical stenosis C4-C6, OPLL	Weakness of hands, interossei muscles atrophy, gait disturbance, Hofmann, flat plantar reflex	3
10	84	Male	RA	Gait instability, hand clumsiness	C5, C6, C7, mild radiculopathy	RA related disorders	Cervical kyphosis, vertebral subluxation, cervical stenosis signal change	Myelopathy hand, Hofmann reflex, motor weakness	4

IHD: Ischemic heart disease, DM: Diabetes mellitus, RA: Rheumatoid arthritis, EMG-NCV: Electromyogram-Nerve Conduction Velocity, CTS: Carpal tunnel syndrome, CVA: Cerebral vascular accident, OA: Osteoarthritis, MRI: Magnetic resonance imaging, OPLL: Ossified posterior longitudinal ligament, CSM: Cervical spondylotic myelopathy

Discussion

CSM is the most common cause of cervical myelopathy and was described by Clark and Robinson (5). Clinical and radiological manifestations of CSM could be misleading and physicians need to be vigilant while evaluating patients with neurologic complaints otherwise delay in diagnosis and management would happen that could finally lead to a poor outcome (1,3,4,6,7,9,10). Furthermore, it is possible that patient was considered to have CSM while he has other pathology as a cause of his neurologic symptoms. Dvorak et al. reported four patients with neurologic findings who admitted in spine unit for consideration for surgery, but after careful neurologic workup the second diagnosis has been made. They believed that multiple sclerosis, syringomyelia, Guillain-Barre syndrome and amyotrophic lateral sclerosis should be considered in the differential diagnosis of CSM. They concluded that if the clinical presentation cannot explicated by radiologic findings, further investigation including cerebrospinal fluid analysis, neurophysiologic studies, and further imaging technique should be applied to exclude systemic or psychogenic disorders (11).

In our series of patients, carpal tunnel syndrome (CTS) (3 of 10 cases) was the most common misdiagnosis that other physicians made and led to the delay in treatment of CSM. All of these patients had been undergone operation before the diagnosis of cervical myelopathy was confirmed. Similarly, Epstein et al. (12) reported six patients which for them operation for bilateral carpal tunnel syndrome was performed and after that due to recognition of radiculomyelopathy symptoms cervical myelopathy was diagnosed and decompressive laminectomy was done. They also stated that because of the resemblance of the symptom of CTS and cervical myelopathy, appropriate clinical neurologic examination, electro-diagnostic and imaging study in patients with presumed diagnosis of CTS, will recognize the correct diagnosis and prevent mistreatment (12). The second most common misdiagnosis of our series was the age-related disease, such as osteoarthritis of hip, knee, and hands, (3 of 10 patients) that their symptoms had been related incorrectly to the aging process by the treating physicians. Therefore, it is very important to have high index of suspicion in especially older patients who present with gait disturbance and difficulty in hands functions (9).

For one of our patients cerebral vascular accident (CVA) had been diagnosed, because he had presented with gait disturbance in the onset of disease. He admitted in the neurologic ward and underwent workup for CVA without any consideration of cervical myelopathy and spinal cord evaluation. For differentiation between CVA and cervical myelopathy, "jaw jerk test" is helpful. Closing of the mouth, upward jerking of the lower mandible, while tapping of the lower mandible at a downward angle with the mouth held slightly open, constitutes a positive test. Positive test represent that the pathology is in the brain (4,7,8,13,14).

Other misdiagnosis in our series were rheumatoid arthritis (RA) related disease, diabetic polyneuropathy and psychosomatic disorder in patients with previous history of RA, diabetic mellitus and depression, respectively.

In terms of symptoms and signs of myelopathy, according to the Ferguson (7) classification, the clinical presentation depends on the location of cord compression in the cervical spine, for this reason the clinical manifestation is different among the patients. In our cases, case no. 8, presented at the beginning of disease with mild gait disorder and insecure gait during walking and he explained that during walking he was feeling instability during the stance phase that this is the result of posterior column compression and proprioceptive dysfunction.

The most common symptoms of our patients were motor weakness in the upper extremity, hand clumsiness and gait disturbance which sometimes explained by the patients' family. Likewise, the most common findings in physical examination in order of decrease were positive Hofmann sign, myelopathy hand symptoms, motor weakness especially in upper extremity and pathologic reflexes in upper and lower extremities such as Babinski sign, hyperreflexia and clonus. Hence, the comprehensive history taking and physical examination have a great value in approaching the patients with neurologic symptoms and gait disturbance.

One of the most important finding in physical examination is Hoffman's sign. Because sometimes it is the only positive finding in patients (case 4 and 7) and should be considered in all patients suspected for neurologic disorders. Denno et al. (15). Introduced dynamic Hoffman sign which "was checked with the head in neutral (static) and during multiple active full flexion to extension as tolerated by the patient (dynamic)," and believed that this test should be added to neurologic examinations to help making early diagnosis of CSM.

Sometimes due to coexistence of severe foraminal stenosis or peripheral polyneuropathy with cervical myelopathy the signs of upper motor neuron symptoms such as Babinski sign and hyperreflexia may be masked and patients can present with diminished or absent reflexes. Because peripheral nerves must be functioning well in order to transmit the hyperreflexia of myelopathy. Hence, the physician should be aware of concomitant foraminal and central cervical stenosis, or cervical stenosis accompany with lumbar foraminal stenosis which is called "Tandem stenosis," in interpreting the symptoms in patients complaining of

neurologic symptoms (4-8,13,15,16).

For our patients after comprehensive history taking and complete neurologic examination, magnetic resonance imaging of the cervical spine was requested to confirm the diagnosis and also rule out the other diagnosis mimicking cervical myelopathy.

We choose surgical approach according to the number of stenosis level, location of compression on spinal cord, cervical alignment, and present or absence of instability in cervical spine. We choose anterior approach in patients with maximum three levels of stenosis and in patients with cervical kyphosis which was not correctable in dynamic view. In patients with more than three levels stenosis or compression of cord from behind posterior approach was selected.

After a short period of time, 6 months after operation, improvement was more significant in patients who had lower grade of Nurrick and in patients who was younger and were operated in less than 6 moths of onset of disease. Because delay in diagnosis result in prolong cord ischemia and permanent cord injury and as a result worst neurologic function representing by higher grade of Nurrick classification. It seems that surgical treatment is effective in all patients suffering from cervical myelopathy, and there was not any significant difference in outcome and also complications between patient underwent anterior approach, and patients underwent posterior approach both anterior and posterior approach, although the further study and clinical trial are needed to conclude which approach is better.

Conclusion

Because of verity of symptoms in patients with cervical myelopathy and also different clinical presentation, full neurologic examination, and comprehensive history taking should be performed. It should be emphasized that just as the pain is not a predictor of cervical myelopathy, complete normal physical examination does not rule out the diagnosis of myelopathy (4-8,13,15,16). Motor and sensory examination may be completely normal or may subtle changes. Searching the pathologic reflexes should be done in patients, but in approximately 20% of patients with cervical myelopathy pathologic reflexes may not be demonstrated (4-8,13,15,16).

Because of progressive nature of cervical myelopathy, it is very important to have high index of suspicious in patients with more subtle or milder degree of myelopathy and do comprehensive history taking and complete neurologic examination. And also with usage of proper imaging studies and elect physiologic tests diagnosis should be confirmed to prevent misdiagnosis. Because of progressive nature of this entity, early diagnosis, and early treatment prior to permanent spinal cord injury is recommended (4-8,13,15,16).

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