Dropped Head Syndrome Proceeding Cervical Spondylotic Myelopathy: Case Report

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ABSTRACT

Abstract: The dropped head syndrome is a rare feature of a variety of neuromuscular diseases characterized by forward flexion of the head due to severe weakness of the extensor muscles of the neck. A patient with the dropped head syndrome or head ptosis is unable to lift the chin off the chest and look forward. There are only isolated cases of cervical spondylosis in whom the dropped head syndrome proceeded months to years before appearance of cervical myelopathy.

Herein, we will present a 72-year-old female who was admitted because of gradual, flexion deformity of the neck over the last 18 months. Flexion deformity of the neck was of a degree that the chin was almost near the chest although it was quite correctable manually and with the use of a collar. She also experienced progressive weakness of all four limbs in the last six months. The imaging disclosed multilevel degenerative cervical spondylosis, myelomalacia of the cord as well as kyphosis. The degree of kyphosis could not be correlated with the degree of degenerative changes of the vertebral bodies. Both conditions improved with multilevel laminectomy and internal fixation from C2 to C7.

KEY WORDS: Amyotrophic lateral sclerosis, Cervical spondylotic myelopathy, Dropped head, Stabilization

INTRODUCTION

The dropped head syndrome (DHS) is characterized with weakness of the extensors or increased tone of the flexor muscles of the neck. Clinically, it is typified by the patient's inability to keep the neck in a neutral position and finally this progresses to chin on chest deformity. Twenty various conditions associated with DHS have been recognized (1-11,12-21,23,24). The dropped head syndrome attributed to cervical spondylopathy has been rarely reported and since Kawaguchi et al in 2004 who described this association for the first time only two more cases have been published so far (10,14,22). It is postulated that cervical spondylosis may cause preferential denervation and weakness of the posterior cervical muscles presumably by compromising the microcirculation of the anterior horn cells innervating a few corresponding myotomes of the cervical cord.

Herein we present a new case of dropped head syndrome in an old female in whom clinical features of cervical spondylotic myelopathy were manifested about a year after appearance of dropped head syndrome. Both conditions improved with decompressive cervical laminectomy and screw–rod instrumentation.

CASE REPORT

A 72-year-old woman was admitted because of progressive numbness and weakness of the upper and lower limbs of six months duration. Initially, at the beginning of the discomfort, despite of numbness of the limbs, she was able to walk without clumsiness, and her grip strength remained acceptable. Later, after three months, her difficulty in walking being started as unsteady gait became apparent. In the last 4 weeks walking was so disturbed that she could only walk with the aid of a walker. Past history revealed that she had been unable to hold her head in upright position and could not prevent her head from dropping on her chest over the last 18 months diagnosed to be dropped head syndrome. The head ptosis
could be corrected with use of a hard collar that was quite important for having a meal and forward vision. Physical examination revealed flexion deformity of the neck with the chin on the chest. However, this deformity could be corrected easily with passive extension. Neurological examination revealed spastic quadripareisis with bilateral extensor planter response. MJOA score was around 10.

Lateral plain radiographs in neutral position revealed chin to chest deformity and degenerative destructive changes of the vertebas and mild slip of C4 on C5. The flexion deformity was not in proportion to the degree of degenerative changes (Figure 1).

Reconstructed CT disclosed degenerative changes with kyphosis (Figure 2).

MRI in supine position revealed cervical spondylotic changes and myelopathy at C4-C5 level. The aforementioned deformity was partially corrected in supine position (Figure 3A, B).

She underwent decompressive laminectomy of C3 to C6. Then pedicle screw of C2 and C7 as well as lateral mass crewes of the intervening vertebas were inserted.

Later, after assembling the rods, the nuts were tightened with the head in extension (Figure 4A,B).

The patient tolerated the procedure very well and was discharged with the head in the upright position although she was advised to use a Philadelphia collar for the next 8 weeks.

Three months after surgery, dramatic improvement of kyphosis continued and the patient was able to look straightforward. Neurologically, her MJOA score increased to 13 and she was very satisfied with her recovery. A plain lateral radiograph confirmed the correction of kyphosis (Figure 5).

**DISCUSSION**

Dropped head syndrome is a chin on chest deformity that is either due to severe weakness of the extensors or increased tone of flexors of the neck and this deformity should be correctable by passive neck extension.

The weakness or atrophy of the cervical posterior extensor muscles can be a symptom of various neuromuscular diseases including amyotrophic lateral sclerosis, myasthenia gravis, congenital myopathies, polymyositis, chronic demyelinating polyneuropathy, fascio-scalpo-
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Figure 3: A) T1-weighted MRI reveals spondylotic changes. Note partial correction of kyphosis in supine position despite using a head-rest under the head. B) T2-weighted MRI showing myelomalacia at the C4-C5 region.

Figure 4: A) Intraoperative view. The screws could be inserted only in extreme flexion. B) Intraoperative view. After assembling the rods, nuts are tightened in extension with correction of deformity.
humeral dystrophy, Nemaline dystrophy, carnitine deficiency syringomyelia, camptocormia, amyloidosis-isolated neck extensor myopathy and spinal muscular atrophy (1-11,12-21,23,24).

However, there are only a few isolated case reports of dropped head syndrome proceeding cervical spondylotic myelopathy (10,14,22).

Patients with this syndrome are unable to lift their chin off the chest wall and have difficulty in looking forward. They also have difficulty in walking, talking and eating and usually use their hands to support the chin. Constant flexion of the neck may result in various degree of neck pain (1-25).

Association of cervical spondylotic changes as a causative factor of this disabling syndrome has been described in a few occasions. In this association, the dropped head syndrome usually occurs a few months to years before appearance of cervical spondylotic myelopathy (10,14,22).

This condition is thought to result from spinal cord microcirculatory disturbances and secondary anterior horn cell degeneration due to ischemia caused by spondylotic changes of the cervical spine.

At the beginning of the disease, the preferential involvement of anterior horn cells innervating the paraspinal muscles results in weakness limited to cervical extensor muscles that causes head ptosis or dropped head syndrome. Later with progression of the disease and subsequent affection of the cord, the clinical features of myelopathy appear.

This means that in a patient with cervical spondylosis, the dropped head syndrome might be present months to years before the appearance of myelopathic changes. Actually, preexisting destruction of cervical vertebrae and silent myelopathy might progress gradually to a symptomatic cervical spondylotic myelopathy secondary to repeated minor traumas resulting from mechanical stress and minor instability.

Electrodiagnostic testing of the patients with dropped-head syndrome due to cervical spondylosis might show active denervation limited to the cervical paraspinal muscle (9). Biopsy of cervical paraspinal muscles is usually not conclusive.

Amyotrophic lateral sclerosis (ALS) is one of the first disorders that should be considered in the differential diagnosis of patients suffering from both cervical spondylosis and the dropped head syndrome. Actually, ALS might present with the dropped head syndrome a few months before appearance of the characteristic features of the disease (3). This is also due to the isolated affection of the anterior horn cells that innervate the paraspinal muscles. However, sooner or later, the disabling picture of ALS will become evident where electrodiagnostic features of this disease in the muscles of the shoulder and hands become apparent. This rare form of ALS is of great importance, in particular if we accept that coexistence of cervical spondylosis and ALS is not infrequent.

Isolated extensor myopathy is another entity that should be considered in the differential diagnosis of the dropped head syndrome. However this benign kind of myopathy is not necessarily associated with cervical spondylotic changes and can be easily diagnosed with neck muscle biopsy that might show myopathic changes with variability of fiber size, internalized nuclei and fiber splitting (8,9,10,14,22,23).

Considering the treatment, it should be noted that the dropped head syndrome not only causes significant disability but is also a source of social embarrassment.
Therefore, the patient can be advised to wear a collar till the correct diagnosis can be made.

Generally, treatment of dropped head syndrome is considered specific to the underlying disease. Patients with DHS of known etiology might respond to an appropriate treatment, such as Parkinson with levo dopa, dystonia with isolated botox injection, polymyositis with long-term corticosteroids and syringomyelia with surgery (4,7,15,20,21).

In pathologies with poor prognosis such as amyotrophic lateral sclerosis, a collar might be the only solution but in benign pathologies, corrective surgical strategy with the application of various internal instruments are recommended (2,14,16,21,22, 25).

Once DHS is associated with cervical spondylotic changes or cervical spondylotic myelopathy, decompressive laminectomy followed by stabilization with screw rod construct is advised. Decompressive surgeries such as laminoplasty eventually may not result in an acceptable outcome (9).

However, combination laminectomy and stabilization has been reported to be associated with good outcome (2,14,16,25). We also recommend this formulation and believe that with application of this strategy both pathologies will eventually improve.

The length of the instrumentation depends the judgment of the surgeon and the severity and the extent of kyphosis. In complicated kyphosis associated with sagittal imbalance, occipitocervical, cervicothoracic and occipito-cervico-thoracic instrumentation have been advocated by different surgeons. However, according to Riew, if the plumb line dropped from the basion falls anterior to the manubrium, the corrective surgery should include upper thoracic spine. Otherwise instrumentation from C2 to C7 will suffice (20,21,23).

With early diagnosis, and before appearance of myelopathy, corrective surgery will result in excellent outcome. However, with delay, both in diagnosis and surgery, myelopathic changes might remain irreversible.

In conclusion, cervical spondylotic should be considered a rare cause of dropped head syndrome. In such instances appropriate surgical intervention probably will avert the upcoming cervical myelopathy. Once a surgeon is faced with both conditions in a patient, a favorable response and a good outcome can be expected if appropriate decompression and corrective stabilization are performed without delay.

REFERENCES

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