Cervical myelopathy caused by dropped head syndrome

Case report and review of the literature

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✓ The authors present a rare case of cervical myelopathy caused by dropped head syndrome. This 68-year-old woman presented with her head hanging forward. After 1 month, she was admitted to the medical service because of head drop progression. Examination of biopsy specimens from her cervical paraspinal muscles showed nonspecific myopathic features without inflammation, and isolated neck extensor myopathy was diagnosed. The patient’s condition did not respond to the administration of corticosteroids. During follow up as an outpatient, the patient’s head drop continued to gradually progress. At 1 year after onset, she developed bilateral weakness of the upper and lower extremities, clumsiness of the hands, and gait disturbance. A radiograph of the cervical spine obtained in a standing position showed a pronounced kyphotic deformity and instability at the level of C4–5. Magnetic resonance imaging demonstrated spinal cord compression at C-3 and C-4. The patient underwent a C3–4 laminectomy and occipitocervicothoracic fixation. Gait and hand coordination gradually improved, and she was able to walk with no support 1 month postoperatively. Surgical fixation was beneficial in this patient with dropped head syndrome, myelopathy, and cervical instability.

KEY WORDS • dropped head syndrome • cervical myelopathy • surgical fixation • isolated neck extensor myopathy

In dropped head syndrome, the patient’s head hangs forward when the body is in the erect or sitting position. Although commonly considered a benign condition by itself, dropped head syndrome limits activities of daily living because the patient can only look downward. The dropped head is primarily due to excessive tension in the anterior cervical muscles associated with multiple systemic atrophy, or in other cases from weakness or atrophy of the posterior cervical muscles. Dropped head syndrome may be caused by one of several neuromuscular disorders. The patient ordinarily complains of head drop and neck pain to some degree. Myelopathic symptoms are rare. In this case report, we describe the development of a kyphotic deformity and its potential for surgical correction. This patient had cervical myelopathy caused by dropped head syndrome. The patient underwent a C3–4 laminectomy and occipitocervicothoracic fixation for a C3–4 cord compression and C4–5 instability. Symptoms improved dramatically after surgery.

Abbreviations used in this paper: MMT = manual muscle test; MR = magnetic resonance.

Case Report

History. This 68-year-old woman first presented with her head hanging forward. After 1 month, she was admitted to our hospital because of the worsening of symptoms. She showed no neurological deficits. A radiograph of her cervical spine showed a kyphotic deformity and spondylotic change in the neutral position, with exacerbation of the kyphotic deformity in flexion (Fig. 1). Gadolinium-enhanced MR imaging demonstrated increased signal intensity within the paraspinal muscles (Fig. 2). Intramuscular electromyograms of the cervical paraspinal muscles and the proximal and distal limb muscles were normal. Biopsy specimens obtained from the cervical paraspinal muscles revealed variations in fiber size, increased connective tissue, and central nuclei. These specimens showed nonspecific myopathic features without inflammation. Laboratory tests, including muscle biopsy procedures, did not establish a specific diagnosis; therefore isolated neck extensor myopathy was diagnosed in the patient. Corticosteroids were administered with no response. During outpatient follow up, the head drop gradually progressed; 1 year after onset, the patient was readmitted to our medical ser-
vice because bilateral weakness had developed in the upper and lower extremities. One month after admission, progression of the patient’s bilateral upper and lower extremity weakness resulted in difficulty getting out of bed. The patient was transferred to the neurosurgery department.

**Examination.** Motor strength was reduced in both the upper extremities (MMT Grade 3/5) and lower extremities (MMT Grade 4/5). Hyperreflexia was present in the upper and lower extremities. Hoffmann and Babinski signs were present bilaterally. Proprioception and light touch sensation were reduced in all four limbs. The patient could not walk and displayed impaired hand movement coordination.

A radiograph of the patient’s cervical spine in a standing position revealed a severe kyphotic deformity, instability at the level of C4–5, and a radiolucent increase with osteoporotic change (Fig. 3). Magnetic resonance imaging showed spinal cord compression at the C3–4 level (Fig. 4). Myelopathic symptoms did not improve with the use of a neck collar for cervical immobilization. Therefore, the patient underwent cervical fixation with a halo brace to treat spinal myelopathy caused by cervical instability. Two weeks after initiating halo brace fixation, the myelopathic symptoms improved. One month after initiating brace fixation, we proceeded with surgical fixation in an effort to decrease the patient’s upper and lower extremity weakness.

**Operation and Postoperative Course.** The patient was positioned prone on bolsters, with her head supported by three-pin fixation after reduction of cervical instability. Using a midline posterior skin incision, the occipital bone, spinous processes, and laminae from C-1 to T-2 were identified. A C3–4 laminectomy was then performed. Cervical fixation from the occipital bone to T-2 was accomplished with an occipitocervicothoracic rod system (Medtronic Sofamor Danek). The rods were fixed in place with a hook system at the occipital bone, sublaminal wiring with radiolucent cable (Nesplon Cable System, Alfresa, Inc.) was placed on the cervical spine, and pediculotransverse process hooks were placed at the level of T-1

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**Fig. 1.** Lateral radiographs of the patient’s cervical spine on initial presentation showing kyphotic change in the neutral position *(left)* and in flexion *(right).*

**Fig. 2.** Axial T₁-weighted MR image with Gd enhancement of the cervical spine demonstrating increased signal intensity within the paraspinal muscles.
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and T-2 (Fig. 5). The fusion substrate used the spinous processes and laminae of C-3 and C-4.

Postoperatively, the patient’s neck was immobilized with a collar for 1 month. Gait and hand coordination improved gradually. One month postoperatively the patient experienced improvements in extremity strength and could walk without support. Although the patient experienced slight neck pain, the signs and symptoms of dropped head syndrome and myelopathy had not recurred 1 year later.

Discussion

Initial conservative therapy for this patient’s dropped head syndrome in an outpatient setting did not prevent worsening of the head drop. Ultimately myelopathy developed as a result of cervical instability. Cervical spine derangements were a consequence of the dropped head associated with weakness of the posterior cervical paraspinal muscles. Significant neck extensor muscle weakness contributed to the development of cervical segmental instability and possibly an accelerated degenerative change. A flexible cervical kyphotic deformity can occasionally result in myelopathy.

To our knowledge, there is no previous report of cervical myelopathy secondary to dropped head syndrome. Amin et al.1 reported vertebral body fractures of T-3 and T-10 in patients with myelopathy associated with dropped head syndrome. These patients presented with spastic paraparesis. Both the head drop and paraparesis were improved by C-2 to T-11 fixation. In another study, Kawaguchi et al.10 reported on an 80-year-old woman with dropped head syndrome associated with cervical spondylotic myelopathy. A laminoplasty from C-2 to C-6 was performed. One year after the operation, she could maintain her cervical spine in a neutral position easily. Those authors concluded that spondylotic compression led to weakening of the cervical extensor muscles.

Dropped head syndrome is caused by various neuro-muscular disorders including amyotrophic lateral sclerosis,3,11 myasthenia gravis,4,12 polymyositis,12 chronic inflammatory demyelinating polyneuropathy with spinal muscular atrophy,5 nemaline myopathy,8 inclusion body myositis,6 parkinsonism,2,13 and Parkinson disease.7 Authors of a recent study found that dropped head syndrome in Parkinson disease is not rare in Japan, with a 6% incidence observed in Parkinson patients.7 Katz et al.9 reported dropped head syndrome in four patients, each showing a nonprogressive myopathy characterized by severe neck extensor weakness, and proposed the designation “isolated neck extensor myopathy.” Our patient experienced a head drop caused by weakness of her posterior cervical paraspinal muscles. A lack of other specific neuromuscular disorders and nonspecific myopathic features in cervical paraspinal biopsy specimens resulted in a diagnosis of isolated neck extensor myopathy.

Generally, treatment of dropped head syndrome is considered specific to the underlying disease. Although a head drop may improve upon treatment of the underlying disease,9 improvement of dropped head with direct treatment is rare. In the four cases of isolated neck extensor myopathy reported by Katz et al.,9 short-term progression did not occur, presumably because worsening was gradual and treatment had little effect. Common treatment of isolated neck extensor myopathy includes corticosteroids, azathioprine,14 fixation of the neck, neck rest,4 and nonsteroidal antiinflammatory drugs. However, no specific treatment has been established for dropped head syndrome.

The patient in our case did not benefit from neck rest or corticosteroids. Operative cervical fixation was performed after cervical instability caused myelopathy. We decided to perform a posterior spinal fixation because the dropped head resulted from weakness or atrophy of the cervical posterior muscles. The patient underwent a C3–4 laminectomy and occipitocervicothoracic fixation because of C-3 and C-4 cord compression caused by C4–5 instability. Cervical pedicle screw fixation and short fusion with sublaminar wiring were considered but were excluded because of poor bone strength associated with osteoporotic changes. Instead, a long fusion by occipitocervicothoracic fixation was performed and the head drop and myelopathy both improved. We believe that fixation could benefit other patients with dropped head syndrome, cervical instabil-

Fig. 3. Preoperative lateral radiograph of the cervical spine showing spondylotic change and severe kyphotic deformity. Note the instability at the C4–5 level.

Fig. 4. Sagittal (left) and axial (right) T2-weighted MR images demonstrating spinal cord compression at the C3–4 and C4–5 levels.
ity, and consequent myelopathy. This myelopathy can respond favorably to surgical correction of the deformity as well as fusion.

References


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Fig. 5. Postoperative anteroposterior (left) and lateral (right) radiographs of the cervical spine showing fixation extending from the occipital bone to T-2.