Isolated neck extensor myopathy causing a dropped head: a case report

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ABSTRACT

This report is of a 71-year-old woman who presented with a dropped head and difficulty in extending her neck. She was diagnosed with isolated neck extensor myopathy. Conservative treatment (use of a cervical collar and muscular strength training) temporarily improved her symptoms. However, destruction of cervical vertebrae and myelopathy progressed secondary to repeated microfractures from mechanical stress. The patient underwent 2-stage combined anterior and posterior decompression and fusion using autografts. At one-year follow-up, numbness of the bilateral upper limbs had resolved, and bone union was achieved. The patient was able to look straight ahead and was very satisfied with the outcome.

Key words: kyphosis; neck pain; spinal cord compression

INTRODUCTION

Paroxysmal dropped head associated with dizziness1 or drooping eyelid and masticatory motor dysfunction2 have been reported. This pathology can be associated with muscular or central nervous system diseases or cervical diseases.3,4 This report is of a 71-year-old woman with isolated neck extensor myopathy who presented with a dropped head.

CASE REPORT

In August 2005, a 71-year-old woman presented with a dropped head and difficulty in extending her neck. She had had an uncomfortable feeling in her neck since June 2005. On examination, no sensory disturbance or deteriorated muscular strength was observed, and the deep tendon reflex was normal. The result of the Spurling test was negative. Cranial neurological signs were negative. Lateral radiographs showed a 47º kyphosis between the C2 and C7 vertebrae (Fig. 1). Magnetic resonance imaging showed no obvious
compression of the spinal cord and nerve root (Fig. 2). She was treated with oral non-steroidal anti-inflammatory drugs, a cervical collar, and muscle strength training for the neck. By October 2005, the neck pain was relieved and the kyphosis improved (Fig. 1). In March 2006, the neck pain recurred without any underlying disease, and her dropped head worsened. Muscle biopsy of the trapezius showed diffuse, non-uniform muscle fibres and growth of interstitium, with no muscular inflammation (Fig. 3). This indicated a myogenic atrophy, rather than neurogenic deterioration. Needle electromyography showed neither neurogenic nor myogenic deterioration. Blood tests showed no abnormalities and a normal level of thyroid hormone. Tensilon test and serum acetylcholine receptor antibody test were both negative. Neurological diseases (amyotrophic lateral sclerosis, myasthenia gravis, and Parkinson’s disease) were excluded from the diagnosis. Based on the muscular biopsy findings, she was diagnosed with isolated neck extensor myopathy.

In June 2006, destruction of the C4 to C7 vertebrae worsened, despite no change in subjective symptoms. In November 2007, numbness developed in the bilateral upper limbs, but she was able to work without clumsiness, and her grip strength remained at 24.5 kg (right) and 20.0 kg (left). Further kyphosis was noted, even in the extended position, with deterioration of motion. Magnetic resonance imaging showed a compressed spinal cord corresponding to the destroyed vertebrae (Fig. 4). In June 2009, she underwent anterior decompression and fusion of C4 to C7 and then posterior fusion of C3 to C7 using autologous fibula and iliac grafts. Histopathological examination of the vertebral body and disk was not performed. At one-year follow-up, numbness of the bilateral upper limbs had resolved, and bone union was achieved. The kyphosis was improved to 20° (Fig. 5). The patient was able to look straight ahead and was very satisfied with the outcome.

**DISCUSSION**

The symptom of a dropped head has been associated with hypothyroidism, severe neuromuscular

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**Figure 1** Lateral radiographs showing (a) a 47° kyphosis between C2 and C7, and (b) improvement to 10° after 4 months of conservative treatment.

**Figure 2** Initial magnetic resonance images showing no compression of the spinal cord and nerve root.

**Figure 3** Needle biopsy of the trapezius reveals diffuse, non-uniform muscular fibres and growth of the interstitium, with no muscular inflammation (H&E, x200).
Dropped head is considered to be a pathognomonic symptom occurring in patients with multiple system atrophy (also known as disproportionate antecollis), which is a systematic neurodegenerative disease of unknown aetiology involving the cerebellum, nigrostriatal corpus, and autonomic nervous system.\textsuperscript{7} Multiple system atrophy includes olivopontocerebellar atrophy, striatogniral degeneration, and Shy-Drager syndrome.\textsuperscript{7}

Dropped head is also considered to be a non-inflammatory myopathy limited to the cervical extensor muscle and not associated with a particular disease.\textsuperscript{8} The condition is also known as isolated neck extensor myopathy.\textsuperscript{9} Thus, this symptom has been associated with muscular and central nervous system diseases, as well as cervical spondylosis and myelopathy. Dropped head may be caused by excessive tension of the anterior cervical muscle or declined strength of the posterior cervical muscle. The former is associated with multiple system atrophy, whereas the latter includes amyotrophic lateral sclerosis, myasthenia gravis, and polymyositis.

In our patient, there was no manifestation of neurological pathology, and needle electromyography showed no abnormalities. Her serum creatine kinase level was normal and the Tensilon test result was negative. She was suspected to have a Charcot spine because of progressive bone destruction and mild neck pain. However, this diagnosis was ruled out, as the patient had no history of tabes dorsalis, syringomyelia, diabetic polyneuropathy, hereditary sensory neuropathy, or spinal cord injury. Muscle biopsy of the trapezius showed diffuse non-uniform muscle fibres and growth of the interstitium. This was considered to be a non-grouping atrophy, rather than neurogenic deterioration. Thus, the diagnosis of isolated neck extensor myopathy was made, despite inconsistency with the needle electromyography findings. The prognosis is relatively good.

Our patient underwent anterior decompression and fusion of C4 to C7 and then posterior fusion of C3 to C7 using autologous fibula and iliac grafts for destruction of the cervical vertebrae secondary to repeated microfractures under mechanical stress. Although C2 is important for cervical stability, fixation of the vertebrae above C3 may result in limited rotation.\textsuperscript{10} Thus, fixation of vertebrae up to C3 may be acceptable, even though the kyphosis cannot be corrected completely.

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