Dramatic Response of Dropped Head Sign to Treatment with Steroid in Parkinson’s Disease: Report of Three Cases

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Abstract

Neck dystonia is the most common cause of dropped head sign in parkinsonism. Isolated neck extensor myopathy, which is a rare condition, can also cause dropped head sign in parkinsonism, but no improvement has been achieved with immunosuppressive therapy. We report three cases of treatable neck extensor myopathy causing dropped head sign in patients with Parkinson’s disease. Needle electromyography and magnetic resonance imaging suggested a restrictive active myopathy affecting neck extensor muscles. All cases responded dramatically to steroid therapy. Routine needle electromyography should be performed to explore treatable myopathy in Parkinson’s disease.

Key words: dropped head sign, neck extensor myopathy, needle electromyography, Parkinson’s disease, prednisolone


Introduction

Dropped head sign (DHS) due to neck extensor muscle weakness may be present in a variety of neuromuscular disorders, including amyotrophic lateral sclerosis (1-4), myasthenia gravis (1, 2, 4), chronic inflammatory demyelinating polyneuropathy (5), polymyositis (1, 4) and inclusion body myositis (1, 6), and in metabolic myopathy, such as hypothyroid myopathy (7), mitochondrial myopathy (8) and carnitine deficiency (1, 2, 9). DHS is also a part of the clinical signs of parkinsonism such as multiple system atrophy (MSA) (10-12), diffuse Lewy body disease (12), and Parkinson’s disease (PD) (12-14). Although the etiology of DHS in parkinsonism has not yet been elucidated, it is known that this symptom can accompany two different conditions, dystonic hyperflexion of the neck or neck extensor muscle weakness. Dystonic hyperflexion of the neck was first described in MSA by Quinn (10) in 1989 and was termed “disproportionate antecollis”. This is the most common cause of DHS in parkinsonism (10, 14). The latter condition, neck extensor muscle weakness, characterized by severe weakness of the neck extensors due to a restricted non-inflammatory myopathy was termed “dropped head syndrome” (15) or “isolated neck extensor myopathy (INEM)” (1). To our knowledge, there are only 3 reports in the literature which discuss cases of INEM with parkinsonism (16-18). No improvement with steroid or levodopa therapy was described in those cases. We report three cases of treatable and dramatic steroid-responsive neck extensor myopathy causing DHS in PD.

Case Report

Patient 1

A 77-year-old woman with a past history of Sjögren’s syndrome had a 5-year history of slowly progressive walking difficulties and postural instability. She walked slowly, taking short steps, and had decreased arm swing predominantly on the right side. There was moderate cogwheel rigidity, bradykinesia, and resting tremor. Levodopa therapy at 300 mg/day improved her walking difficulties. 

123I metaiodobenzylguanidine (MIBG) myocardial scintigraphy for differentiating PD from MSA exhibited low uptake. She noticed dropped head characterized by moderate weakness of
the extensor muscles of the neck 3 months prior to admission. At the time of admission, neck extension was grade 3 on a modified Medical Research Council (MRC) scale (Fig. 1A). Otherwise muscle strength was normal. Abnormal results of laboratory tests included mildly elevated creatine kinase (CK) of 268 IU/L (normal, <189 IU/L) and positive test results of anti-nuclear antibody and anti-SSA/Ro antibody. Anti-SSB/Ro antibody and anti-acetylcholine receptor (anti-AChR) antibody were negative. Erythrocyte sedimentation rate (ESR), serum immunoglobulins and thyroid function were normal. Needle electromyography (EMG) revealed profuse fibrillation potentials and positive sharp waves (PSWs) in the right C6 and C7 paraspinal muscles. Many obvious small-amplitude short-duration motor unit potentials (SASD MUPs) and rapid recruitment with pathological interference was noted in those muscles (Fig. 2A, B). EMG of the right deltoid and biceps muscles was normal. Magnetic resonance imaging (MRI) of the cervical spine showed no spondylosis and showed high signal intensity in the bilateral splenius capitis muscles on short tau inversion recovery (STIR) images (Fig. 3). An increase in the dose of levodopa to 400 mg/day did not result in any improvement of the DHS. Immediately after the start of prednisolone treatment at 20 mg/day, dramatic improvement in the strength of her neck was seen, and she was able to raise her head completely 2 weeks later (Fig. 1B). Her neck condition has been stable for 3 months, and prednisolone was tapered and discontinued.
Patient 2

A 74-year-old woman presented with walking difficulties and stiffness in the body for approximately 10 years. These problems became progressively worse and there were difficulties in rising from a chair without support or turning around 180 degrees. On physical examination, there was moderate cogwheel rigidity, bradykinesia and resting tremor predominantly on the left side. Initially, a small dosage of levodopa had been effective for controlling the symptoms. However, after several years, increase in the dosage of levodopa up to 500 mg/day and administration of pramipexole at 1.5 mg/day were required for effective treatment. She also developed extreme neck extensor weakness over a 2-month period and was unable to lift her chin off her chest wall (Fig. 1C). Neck extension was grade 2- on an MRC scale, but all other muscles had normal strength. CK was normal, but myoglobin was mildly elevated (126 ng/mL; normal, <65 ng/mL). ESR, serum immunoglobulins and thyroid function were normal. Anti-AChR antibody was negative. EMG findings were typical SASD MUPs and rapid recruitment with profuse fibrillation potentials and PSWs in the right C5 and C6 paraspinal muscles. EMG of the left deltoid was normal. MRI of the cervical spine showed mild spondylotic changes but no root or cord compression. STIR images in the neck extensor muscles did not show abnormal signal intensity although EMG findings in these muscles were indicative of active myopathy. After starting prednisolone administration at 20 mg/day, gradual improvement in the strength of her neck was seen, and she recovered almost completely after 2 months (Fig. 1F).

Patient 3

A 69-year-old woman with pigmentary degeneration of the retina complained of progressive walking difficulties and postural instability for 5 years. There was moderate cogwheel rigidity, bradykinesia and resting tremor predominantly on the left side. MIBG myocardial scintigraphy for diagnosing PD exhibited low uptake. She was treated with 100 mg/day of levodopa, 0.5 mg/day of pramipexole and 7.5 mg/day of selegiline, and obvious improvement in walking difficulties was observed. Extreme neck extensor weakness developed over a 2-month period and she was unable to raise her head against gravity (Fig. 1E). It thereby became difficult for her to look forward when walking. Neck extension was grade 2 on an MRC scale. All other muscles had normal strength. CK was normal, but myoglobin was mildly elevated (157.5 ng/mL). ESR, serum immunoglobulins and thyroid function were normal. Anti-AChR antibody was negative. EMG revealed typical SASD MUPs and rapid recruitment with profuse fibrillation potentials and PSWs in the left C6 and C7 paraspinal muscles. EMG of the left deltoid was normal. MRI of the cervical spine showed mild spondylotic changes but no root or cord compression. STIR images in the neck extensor muscles did not show abnormal signal intensity although EMG findings in these muscles were indicative of active myopathy. After starting prednisolone administration at 20 mg/day, gradual improvement in the strength of her neck was seen, and she recovered almost completely after 2 months (Fig. 1F).

Discussion

The remarkable clinical aspect of our patients was the occurrence of severe neck weakness without involvement of other muscle groups during the course of PD. DHS due to neck weakness developed over a period of 2 to 3 months. The onset was around 70 years of age or older. Electro-physiological abnormalities localized to the cervical paraspinal muscles were indicative of active myopathy. A small dosage of steroid showed a dramatic effect on neck weakness within 2 months, whereas the general symptoms of parkinsonism remained unchanged during the treatment on steroid.

Although it has been considered that “disproportionate antecollis” due to neck dystonia is the most common cause of DHS in parkinsonism (10, 14), the present cases did not seem to be due to dystonia. In all 3 patients described herein, there was no sign of a “psychic pillow”, which is a term used to describe an uncontrollable neck dystonia (19), because, when lying down, the neck would go back to a normal position immediately. Neck dystonia has been reported to be much more frequent in MSA than in PD (10-12), whereas, in the present cases, the diagnosis of PD was made on the basis of existence of asymmetrical motor impairment and a good levodopa response in parkinsonism. In addition, in patients 1 and 3, MIBG scintigraphy, which helps to differentiate PD from MSA, showed reduced uptake of the isotope indicative of PD (20).

DHS characterized by isolated neck extensor weakness was initially termed “dropped head syndrome” by Suarez and Kelly (15) in 1992. Subsequently, Katz et al (1) suggested the name “INEM”. INEM cases have occasionally been described over the past decade (16-18, 21). Patients with INEM have rapid onset and less progressive myopathy

Figure 3. Axial STIR images in patient 1 showing hyperintensity of the bilateral splenius capitis muscles (arrows).
of neck extensor muscles. The onset is usually around 60 years of age or older. Muscle biopsies in previously reported INEM cases have shown a nonspecific myopathic change without inflammatory infiltrates. Although the cause of INEM remains unclear, it has been suggested that mechanical stretching causes injury of neck extensor muscles (1, 21). Overloading to the neck extensor muscles is an aggravating factor to induce further neck drop in the case of kyphotic postural changes or loss of tissue elasticity associated with aging. Equally, pronounced neck rigidity or dystonia in parkinsonism may place increasing workloads on the cervical paraspinal musculature that leave some individuals susceptible to injury. To our knowledge, there have been only 3 reports (9 cases) of INEM in parkinsonism to date (16-18). Okamiya et al (16) reported an 84-year-old man having INEM caused by vascular parkinsonism. Askmak et al (17) reported that 1.5% (7 of 459 patients) of patients with parkinsonism were found to have INEM. All 7 patients with INEM were older than 60 years of age (up to 91 years of age) and made a diagnosis of MSA probable (17). Lava and Factor (18) reported a 70-year-old woman who developed INEM during the course of PD. The clinical features in the present patients are similar to those of INEM. However, we could not diagnose INEM for two reasons. First, in all 3 patients, muscle biopsy findings were not available because we could not obtain the patient’s consent. Second, unlike our cases, there have been no cases previously reported of INEM successfully treated with steroid therapy. The dramatic response to steroid therapy suggests that the etiology of the present cases should be different from INEM initially reported by Suarez and Kelly.

Although the term “INEM” has been used for idiopathic restricted non-inflammatory myopathy of the neck extensors, the underlying basis for INEM is still uncertain. Goh et al (22) suggested that DHS caused by a myopathic origin is a syndrome of mixed etiology and that there is a spectrum of pathological processes ranging from non-inflammatory to pronounced inflammatory myopathy. So far, only 7 reports (9 cases) of DHS due to a restricted myositis have appeared in the literature (22-28). Gaeta et al (26) used the term “inflammatory INEM” for this condition. Recently, Gdynia et al (28) described histopathological data from 19 muscle biopsies in patients with PD and concomitant dropped head or bent spine syndrome. Three patients demonstrated an inflammatory myopathic change and other patients demonstrated histological characteristics of a necrotizing myopathy or a myopathic change with mitochondrial abnormalities. Most of the cases that had shown inflammatory infiltrates in affected muscles on biopsy responded well to steroids (22, 24, 26-28) and good disease control was achieved with immunosuppressive therapy such as azathioprine (24, 27) and intravenous immunoglobulin (25). The etiology of the present cases was unclear, but the clinical features seemed to be an immune-mediate mechanism such as an inflammatory type of myopathy because of dramatic response to steroid therapy.

The most notable feature in our cases was that steroid-responsive neck extensor myopathy occurred in PD. When one is faced with a patient with DHS in PD, EMG of the neck extensor muscles should be considered in order to explore myopathic change. Moreover, steroid therapy can be tried to confirm steroid-responsive neck extensor myopathy, which is a potentially treatable disease.

The authors state that they have no Conflict of Interest (COI).

References


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