ABSTRACT: Weakness of neck extension causing a dropped head may result from many neuromuscular disorders. One etiology is isolated neck extensor myopathy. A similar focal myopathy of the lower axial muscles may cause the bent spine syndrome, which manifests as flexion of the trunk and inability to stand upright. Combination of both dropped head and bent spine myopathies is uncommon. Inflammation is usually not pronounced in these conditions and response to immunosuppressive treatment is rare. We present an 81-year-old man who developed progressive weakness of neck and trunk extension over several months, with a prominent inflammatory process in the thoracic paraspinal muscles, which responded dramatically to treatment with intravenous immunoglobulin (IVIg). This case, together with other rare reports, suggests that the presence of inflammation in the biopsy of an affected muscle may predict treatment response.

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RESPONSE OF THE DROPPED HEAD/BENT SPINE SYNDROME TO TREATMENT WITH INTRAVENOUS IMMUNOGLOBULIN

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The dropped head syndrome due to weakness primarily localized to the neck extensors may be caused by a variety of neuromuscular disorders, including motor neuron disease, myasthenia gravis, chronic inflammatory demyelinating polyneuropathy, and inflammatory, dystrophic, and metabolic myopathies. The "bent spine syndrome" is characterized by pronounced flexion of the trunk, with limited extension, is reduced when supine,[9] and may be associated with head drop.[6] Both conditions can be caused by axial myopathy. The occurrence of both dropped head and bent spine together has been reported infrequently.[6] In several series, bent spine has been attributed to a primary disorder of the paraspinal muscles, with fatty degeneration, increased connective tissue, and usually little or no inflammation, except in one case.[2,5] Response of myopathic neck extensor weakness to treatment with corticosteroids typically has been disappointing.[4,6,10] We report a case of a man with the dropped head/bent spine syndrome due to an inflammatory axial myopathy who responded dramatically to intravenous immunoglobulin (IVIg).

CASE REPORT

An 81-year-old man with hypertension, coronary artery disease, and atrial fibrillation developed progressive slumping forward of his torso beginning about 1 year prior to presentation. Six months later, he began having increasing difficulty in keeping his head elevated, although his head drop improved when he was lying supine. These symptoms caused him difficulty in walking and climbing stairs. He denied any rashes, muscle pain, or cramps. He did not have any difficulty in speaking or swallowing, nor were there any sensory symptoms. On examination, he had marked forward head drop, with 4− strength on the Medical Research Council scale. He had a prominent scoliosis, concave to the right, with weakness of trunk extension. Neck flexion was normal, as was the remainder of muscle strength in the arms and legs. He had no atrophy or fasciculations, and muscle tone was normal. Vibratory sensation was mildly diminished in the hands and feet, but sensory testing was otherwise normal. Coordination was normal. Due to the head drop and stooped posture, he

Abbreviations: EMG, electromyography; IVIg, intravenous immunoglobulin

Key words: axial myopathy; bent spine; dropped head; inflammatory myopathy; intravenous immunoglobulin (IVIg)

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walked with very slow, unsteady steps. Deep tendon reflexes were brisk and symmetric in the arms and at the knees, and were mildly reduced at the ankles. Plantar responses were flexor.

Blood chemistry panel, including sodium, potassium, chloride, carbon dioxide, urea nitrogen, creatinine, and glucose, and a complete blood count were essentially normal. Serum creatine kinase was mildly elevated at 361 IU/L (normal range, 0–175 IU/L). Serum thyroid-stimulating hormone, vitamin B_{12}, and aldolase, and serum protein electrophoresis were normal. Rapid plasma reagin was nonreactive. Acetylcholine receptor antibodies were negative. Radiographs of the thoracic and lumbar spine revealed a 35° levoscoliosis from T-11 through L-3 with associated pelvic tilt.

Electromyography (EMG) and nerve conduction studies of the right arm and leg were normal. Fibrillations and positive sharp waves were noted in the thoracic paraspinal muscles, with normal-appearing motor unit potentials. EMG of the right lumbar paraspinals also revealed fibrillations and positive sharp waves. A biopsy of a thoracic paraspinal muscle revealed multiple foci of endomysial and perivascular chronic inflammatory cells (Figs. 1 and 2). There was prominent fiber size variation attributable predominantly to polygonal atrophy and endomysial fibrosis, with necrotic and regenerating fibers. Electron microscopy was not performed.

Because of his age and coexisting medical conditions, it was decided to treat the patient with IVIg rather than steroids. He was started on IVIg at 2 g/kg over 5 days once a month. Improvement in neck and spine extensor strength occurred after the first course. After the second course of IVIg, his head drop had resolved and he was able to sit and stand up fully erect. Neck and back extension strength were normal. This benefit was sustained with the same regimen of IVIg (2 g/kg monthly) over the 12 months of follow-up and weakness has remained confined to the spine. A reduction of the dose to 1 g/kg every 4 weeks resulted in increased weakness. Questioning of his treatment by his health insurer led to the addition of prednisone, 60 mg daily, but the deterioration continued to the point where he needed hospitalization. An intense course of IVIg at 2 g/kg every 2 weeks for six cycles returned his spine strength to the point before the deterioration.

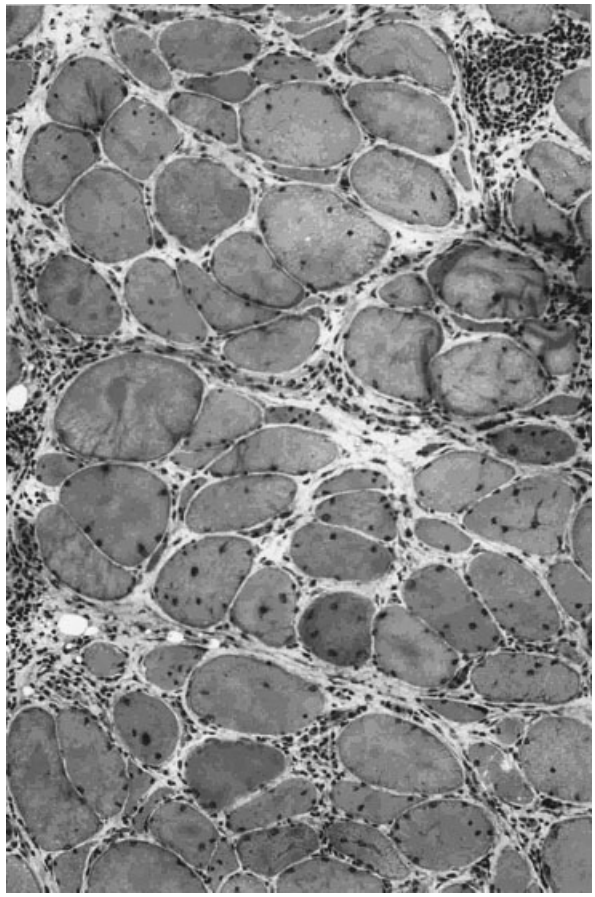

FIGURE 1. Thoracic paraspinal muscle biopsy. Hematoxylin–eosin stain on formalin-fixed paraffin-embedded tissue. Extensive rounded or polygonal myofiber size variation is associated with endomysial fibrosis and chronic inflammation. ×200.

DISCUSSION

It has been proposed that dropped head and bent spine syndrome, together or separately, may be due to an entity known as axial myopathy.[6,7,11] Our patient is similar to those previously described, although he had the apparently uncommon combination of both dropped head and bent spine. Muscle biopsies in previously reported cases of dropped head or bent spine have usually shown nonspecific myopathic features, including fiber splitting, fiber size variation, moth-eaten fibers, type 2 atrophy, and internal nuclei.[4,6,10] However, there have been reports of biopsies with some inflammatory cells present,[3,6,7,9] and our patient's muscle biopsy showed a prominent inflammatory cell infiltrate. The most striking aspect of this case is our patient's dramatic response to IVIg treatment, with no apparent benefit from additional steroids. In most cases in which steroids were tried there was no benefit.[4,6,10] In one such case, prednisone, plasmapheresis, and azathioprine were used, with some improvement in limb strength but not neck extension.[4] One explanation proposed for the lack of response to steroids

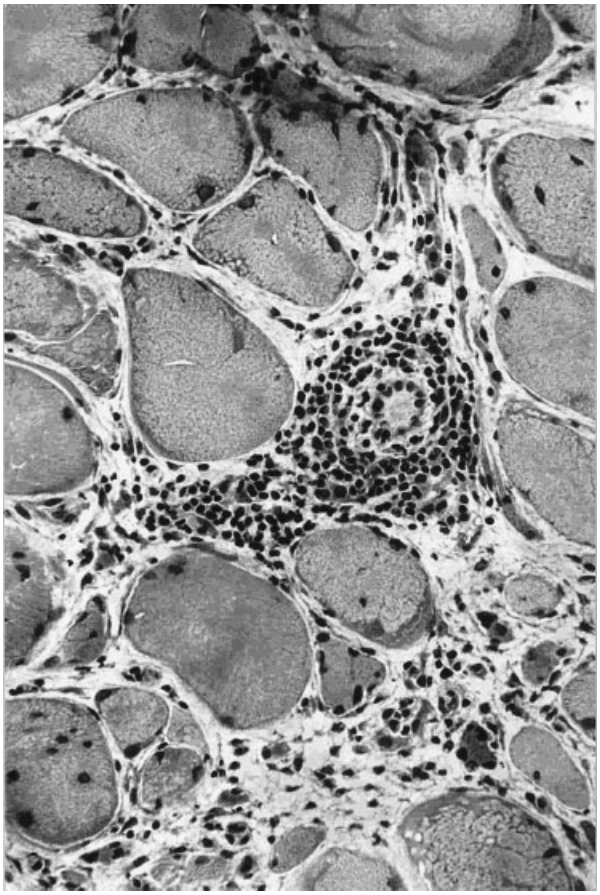

FIGURE 2. Thoracic paraspinal muscle biopsy. Hematoxylin–eosin stain on fresh-frozen tissue. There is perivascular endomysial chronic inflammation. Internal nuclei are increased and intermyofibrillary networks are often smudged or blurred. ×400.

is that the dropped head position produces mechanical stretch injury to neck extensor muscles over time, with associated loss of tissue elasticity.[4]

It is noteworthy that the only other case of isolated myopathic dropped head syndrome that responded to treatment also had prominent inflammatory infiltrates in affected muscles[1]; this patient responded to methylprednisolone, followed by oral prednisone. A case of "dropped head plus syndrome," with neck extensor and arm and leg weakness, responded to the combination of prednisone and azathioprine.[8] Biopsy of the trapezius in that case revealed regenerating fibers, some small angular fibers, and scattered foci of inflammatory cells, which did not invade muscle fibers.[8] Although it seems intuitive to biopsy the affected paraspinal muscles in cases of isolated axial weakness, this has not always occurred.[10] Biopsies of muscles other than the affected paraspinals, such as the deltoid or biceps, may not be particularly revealing.[10]

To our knowledge, there have been no cases previously reported of dropped head/bent spine syndrome successfully treated with IVIg. Additionally, the only cases of isolated dropped head or bent spine syndrome that have responded to immunosuppressive treatment have had inflammatory infiltrates on biopsy of an affected muscle, an observation that may help predict response to treatment and prognosis. Further cases will be necessary to determine this.

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