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Chiropractic management of a U.S. Veteran with myofascial pain and concurrent distal bimelic amyotrophy (Hirayama disease): a case report

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Background: *Distal bimelic amyotrophy (DBMA) also known as Hirayama disease, is a rare, self-limiting motor neuron disease manifesting as atrophy of C7-T1 innervated muscles. We present a case report describing the chiropractic management of neck and thoracic pain in a patient with known DBMA.*

Case presentation: *A 30 year-old black male U.S. veteran with DBMA presented with myofascial pain of the neck, shoulder, and back. A trial of chiropractic care was undertaken involving spinal manipulation of the thoracic spine and cervicothoracic region, manual and instrument-assisted soft tissue mobilization, and*

Prise en charge chiropratique d'un vétéran américain souffrant de douleurs myofasciales et d'une myélopathie cervicale basse (maladie d'Hirayama): rapport de cas

Contexte : *La myélopathie cervicale basse, également connue sous le nom de maladie d'Hirayama, est une maladie rare et spontanément résolutive du motoneurone qui se manifeste par une atrophie des muscles innervés C7-T1. Nous présentons un rapport de cas décrivant la prise en charge chiropratique de douleurs cervicales et thoraciques chez un patient atteint d'une maladie d'Hirayama connue.*

Présentation du cas : *Un vétéran américain noir de 30 ans, atteint de myélopathie cervicale basse, s'est présenté avec des douleurs myofasciales au cou, aux épaules et au dos. Un essai de soins chiropratiques a été entrepris comprenant des manipulations vertébrales de la colonne thoracique et de la région cervicothoracique, des mobilisations manuelles et instrumentales des tissus mous, et la prescription d'exercices à domicile.*

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home exercise prescription. The patient reported modest improvement in pain intensity and did not experience any adverse events.

Summary: *This case presents the first documentation of chiropractic services in musculoskeletal pain management of a patient with concurrent DBMA. At this time there is no guidance in the existing body of literature for the safety and effectiveness of manual therapy in this population.*

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KEY WORDS: chiropractic; bimelic amyotrophy; hirayama disease; myelopathy; manual therapy

Le patient a fait état d'une amélioration modeste de l'intensité de la douleur et n'a pas ressenti d'effets indésirables.

Résumé : *Ce cas présente la première documentation des services chiropratiques dans la gestion de la douleur musculo-squelettique d'un patient souffrant d'une myélopathie cervicale basse. À l'heure actuelle, il n'existe pas d'orientation dans la littérature existante sur la sécurité et l'efficacité de la thérapie manuelle dans cette population.*

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MOTS CLÉS : chiropratique ; myélopathie cervicale basse ; maladie d'Hirayama ; thérapie manuelle

Introduction

Distal bimelic amyotrophy (DBMA) is a phenotypic variant of monomelic amyotrophy, also known as Hirayama disease. It is a motor neuron disease first described by Hirayama *et al*.[1,2] in 1959. Other proposed terms for DBMA, depending on the location of extremity involvement, include brachial monomelic amyotrophy, crural monomelic amyotrophy, or proximal bimelic amyotrphy.[2,3] Early descriptions of DBMA only recognized upper extremity involvement, with the most identifiable characteristic being sensory-sparing, motor amyotrophy commonly limited to weakness in the C7 to T1 myotomes.[4] Clinically presenting as an insidious onset of weakness and atrophy in the unilateral or bilateral distal upper extremities, DBMA has been described in the literature as self-limiting, with muscle atrophy plateauing around five years after onset.[5] Although DBMA is considered self-limiting, the extent of atrophy acquired is permanent[6], and atrophy-related fatigue is the most common long-standing symptom[7].

Monomelic amyotrophy is reported to range from 8% to 29% of all motor neuron diseases (e.g. amyotrophic lateral sclerosis, Madras motor neuron disease).[8–10] The dominant limb is affected more commonly, as reflected in a 3:1 right-sided predominance.[11] It is seven times more prominent in males and more commonly reported in Asian countries.[6] The initial diagnosis classically occurs during the adolescent and young adult years.[6] DBMA is a rare condition and the prevalence is unknown.[12] The rarity of cases has limited investigation into familial, or other potentially causal, relationships which may predispose individuals to developing DBMA.[13] Although the exact etiology is unknown, it has been theorized that DBMA presentation is the result of growth discrepancies between the vertebral column and spinal canal contents.[7,14] Individuals with a long vertebral column and short thecal sac may be at higher risk of repetitive cord compression, or venous congestion[13], as a result of the anterior thecal sac being displaced into the posterior surfaces of the vertebral bodies during cervical flexion[14,15]. Table 1 presents common and less commonly observed features of distal bimelic amyotrophy.

Table 1.
Distal bimelic amyotrophy characteristic features.[8]

Common	Less common
Young age of onset Sporadic occurrence Male preponderance Weakness and atrophy affecting intrinsic muscles of hand and forearm Commonly confined to single limb	Cold paresis Hyperhydrosis with abnormal sympathetic skin response Bilateral

Unlike presentations of cervical myelopathy or amyotrophic lateral sclerosis (ALS), the lower extremity is typically spared in DBMA. The differential diagnosis for DBMA includes peripheral neuropathy (e.g. pronator

syndrome), spinal cord pathologies (e.g. syringomyelia), and multisystem genetic disorders (e.g. myotonic dystrophy).[16] Diagnosis of DBMA may be suspected through cervical MRI and is confirmed through repeat MRI with the patient positioned into cervical flexion.[14,15] The addition of a cervical flexion MRI demonstrates the anterior displacement of the thecal sac along with a high signal intensity crescent-shaped lesion in the posterior epidural space, most easily seen with T2 weighted sequences.[17] With this displacement, compression into the posterior element of the vertebral bodies occurs at the anterior horn of the spinal cord and ventral roots, resulting in the hallmark motor deficits with sensory sparing.[14,16]

Patients with DBMA are commonly advised to avoid cervical flexion due to the increased pressure placed on the anterior spinal cord, putting it at risk for further injury, and they often describe electric-like pain/sensations in the upper extremities when performing cervical flexion movement.[18,19] Surgical management may be considered to reduce the risk of continued cord compression.[13] Physical and occupational therapy is reported to improve and maintain adaptations to living with muscle atrophy.[18,20] To the authors' knowledge, there are no prior reports in the literature detailing the role of chiropractic care in the management of musculoskeletal complaints in patients with DBMA.

Case presentation

This case report was approved by the VA Puget Sound Privacy Officer. The patient provided consent for publication, and we followed CARE guidelines of reporting for case reports.[21] A 30 year-old black, right-hand dominant male was referred by a Veterans Health Administration (VHA) physical medicine and rehabilitation physician to a VHA chiropractic clinic for neck pain and upper back tension. Ten years prior to his chiropractic presentation, he began experiencing neck pain and hand weakness that limited his ability to complete his service responsibilities in the U.S. Army. His symptoms progressed over two years and he was medically discharged from military service and initiated evaluation with VHA. Neurosurgery consulted and deemed him to not to be a suitable candidate for surgical intervention due to the stabilization of symptoms, and recommended conservative management. At the time of his presentation to the chiropractic clinic, 10 years after initial presentation, his treatment team consisted of a physical medicine & rehabilitation physician, psychologists, and occupational therapists.

On presentation, he described muscular neck and upper back pain. Pain intensity was rated four out of 10 on a numeric pain rating scale (NPRS). Function and disability were rated 14 out of 50 (28%) with the Neck Disability Index, correlating with a "mild disability" score.[22] He reported bilateral upper extremity weakness, atrophy, and muscle fatigue which worsened with aerobic activity and cold weather, with the left upper extremity weakness more prominent. This often resulted in clumsiness of his hands (dropping items, general difficulty with hand dexterity) throughout the day. He reported managing his symptoms with cool showers and self-massage of the neck and upper back musculature. Prior to the COVID-19 pandemic, the patient participated in long-boarding and exercising at a mix-martial arts gym. His review of systems was unremarkable. Upon review of the electronic health records, a cervical MRI from six years prior demonstrated mild atrophy of the anterior cord at levels C5 through C7 without abnormality of the cord signal, findings consistent with DBMA (Figures 1 and 2). A cervical MRI with flexion views was not available in the records. The patient's past medical history was not significant for any other relevant comorbidities. At the time of presentation to the VA chiropractic service the patient was not taking any prescription medications.

Diffuse atrophy of the upper extremities distal to the elbow was noted. Neurological examination revealed right arm pronator drift, a single beat of clonus in the left ankle, and bilateral strength deficits at the C8 and T1 myotomes. Romberg's test, heel and toe walk were performed without difficulty, Hoffman's reflex was absent bilaterally, cranial nerves were grossly intact, heel-to-shin test was unremarkable, muscle stretch reflexes of the upper and lower extremities were all 2+ bilaterally, and sensation to light touch was normal in all extremities. The patient had previously been instructed to avoid cervical flexion movements, but did so at the examination without being prompted and no range of motion deficits or pain of the cervical spine was observed. Axial compression, cervical distraction, shoulder depression, and upper limb tension tests were all unremarkable. Joint movement restrictions were noted in the thoracic spine with hypertonicity of the upper trapezius muscles. The patient was diagnosed with

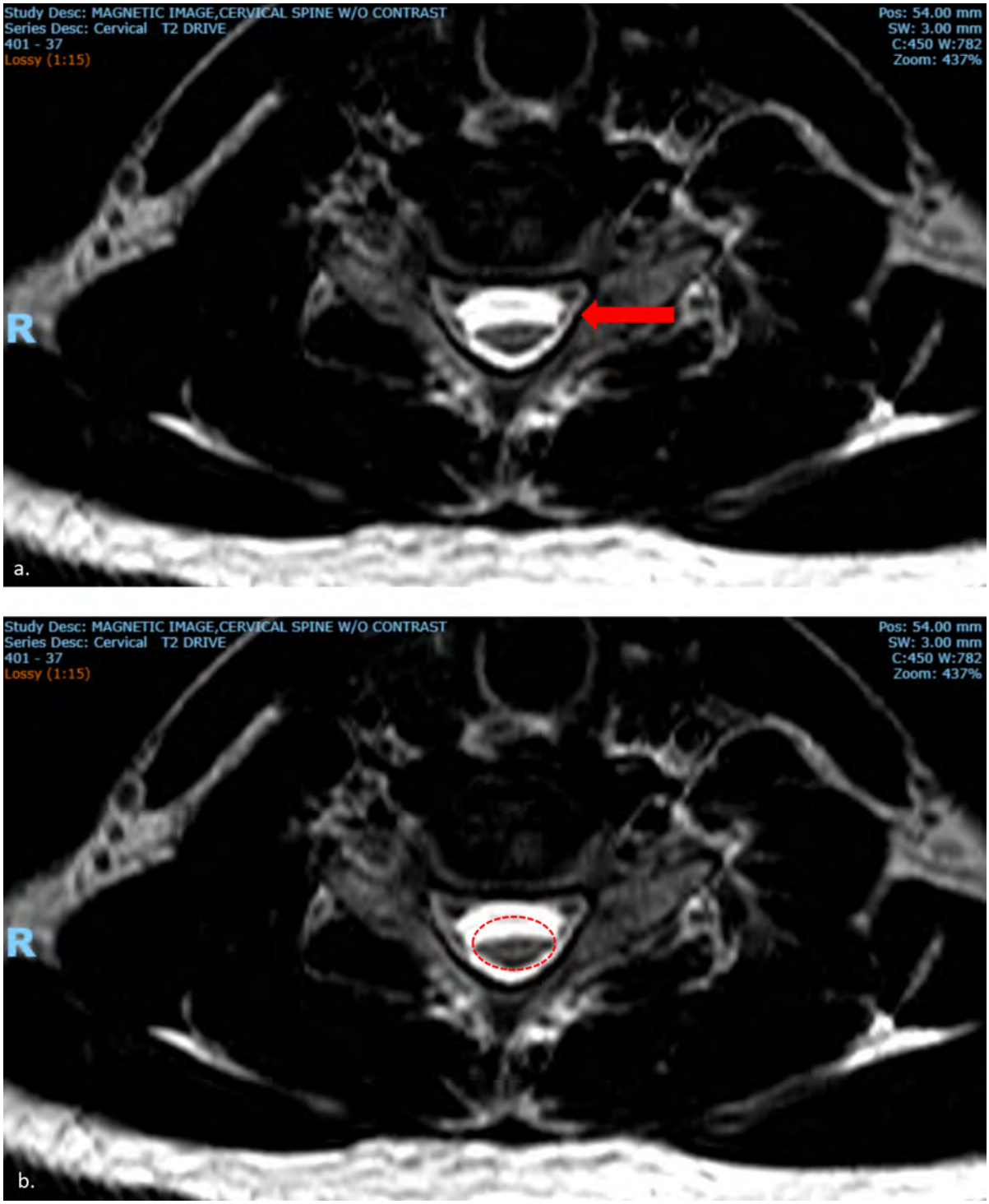

Figure 1.
a.T2 weighted axial cervical MRI demonstrating mild anterior cord atrophy at C6-C7 (red arrow). b. Red dotted line approximates normal cord volume at this spinal level.

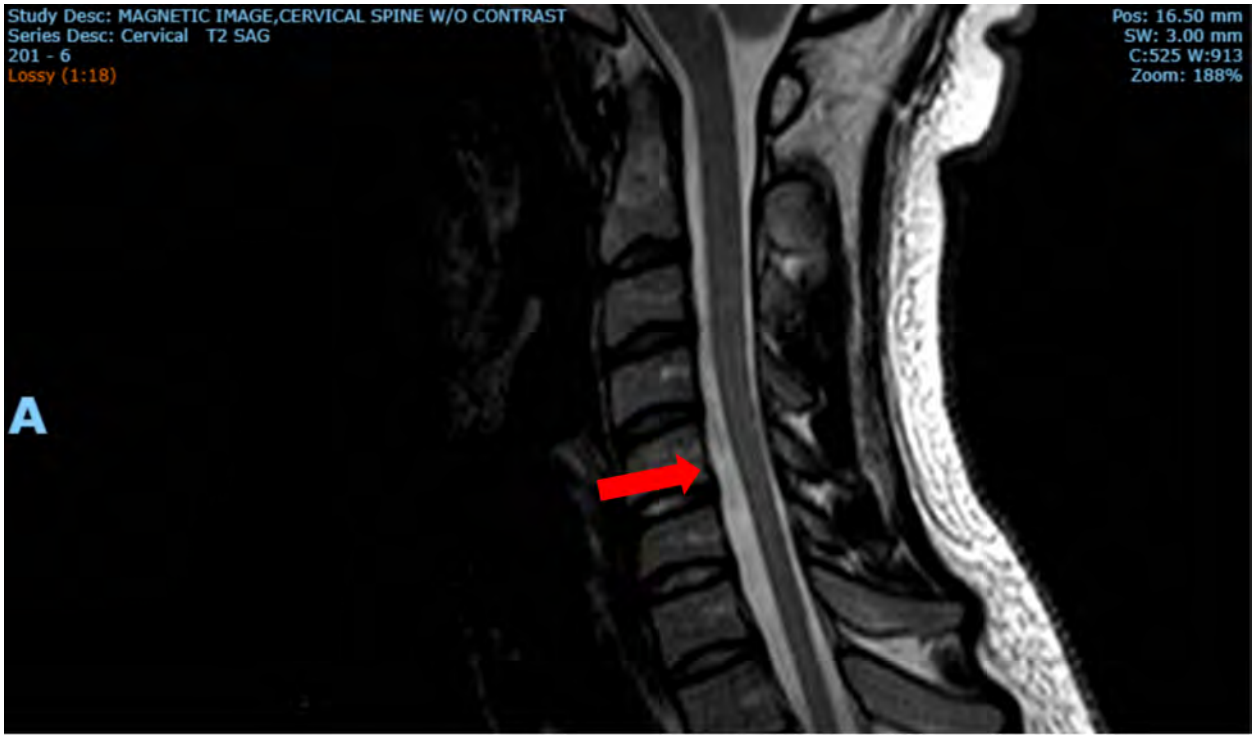

Figure 2.
T2 weighted sagittal cervical MRI demonstrating mild atrophy of the anterior cord at the C5, C6, and C7 levels (red arrow). No abnormal cord signal noted. The remainder of the visualized spinal cord was normal in course, caliber, and signal.

myofascial upper thoracic pain complicated by concurrent DBMA.

A trial of manual therapy was initiated with high-velocity, low-amplitude (thrust) supine thoracic spine manipulation, manual myofascial release, and post-isometric relaxation to the upper trapezius bilaterally. At each session, following manual therapy, the patient participated in supervised exercise and was provided home exercise instructions to perform four bodyweight rehabilitation exercises (Y, T, W, L) for five repetitions each[23] (Figure 3). Each movement was then repeated with elastic resistance bands (e.g., TheraBand) added.

At a follow-up session six weeks later, the NPRS was rated 0 out of 10, but he noted two out of 10 "tension" in the upper thoracic and cervicothoracic region. He described increased general activity and engagement in the home exercise program. Over a subsequent four visits in a three month period, seated cervicothoracic long axis manipulation (Figure 4) and instrument-assisted soft tissue mobilization were additionally incorporated into the treatment plan. Soft tissue techniques were applied to the suboccipital, cervical paraspinals, levator scapulae, rhomboids, and periscapular muscles as indicated. Care was taken to maintain neutral cervical positioning and no active or passive movements were attempted that involved cervical flexion.

At four months following the initial presentation an updated NDI was scored 11 out of 50 (22%), and he reported benefit in general mobility following manual therapies, and quality of life as a result of encouragement from the treatment team to remain active. He was seen for seven visits over five months. After his final visit he was referred to a Whole Health[24] introductory class intended to inform veterans about additional opportunities offered within the

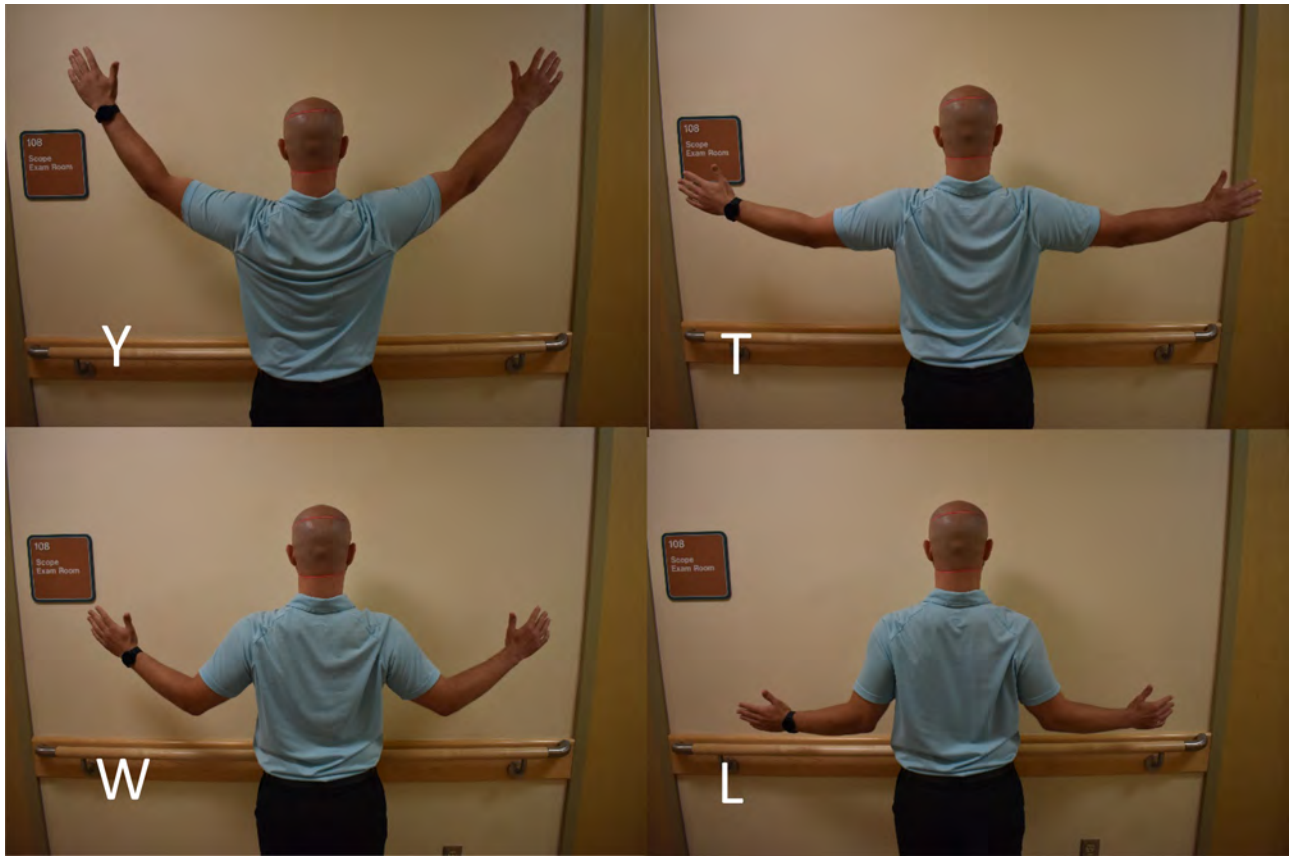

Figure 3.
Depiction of Y,T,W,L movements. The patient is not pictured.

VHA to engage in self-directed active care strategies (e.g. tai chi, yoga). Over the course of care, no adverse events occurred. The patient was lost to follow-up.

Discussion

This case report is the first documented account of the chiropractic management of cervico-thoracic region myofascial pain in a patient with previously diagnosed distal bimelic amyotrophy. Due to the relatively low intensity of the patient's pain experience, it was difficult to determine the impact of chiropractic care on his complaints; however, there was no exacerbation or progression of DBMA and no adverse events were reported. Current literature has focused on the recognition and diagnosis of the DBMA disease entity and very little exists on the symptomatic management of this disorder.[25]

Surgical management options for DBMA diagnosis include posterior cervical duraplasty or anterior cervical fusion.[13] Cervical duraplasty is typically performed via a two-step surgical procedure: laminectomy followed by expansile duraplasty (an opening is created in the dural sac and covered using grafted fascia).[26] This procedure aims to create more space around the spinal cord during flexion-based movements. Anterior cervical fusion, with the aim of restricting cervical flexion, through the addition of instrumentation to immobilize segments of the spinal column is another option in patients with progressive symptoms. A recent study compared the outcomes of surgical (duraplasty) to conservative management (i.e. cervical collar) revealing better patient outcomes with surgery.[26] A clinical practice guideline on diagnosis and treatment of Hirayama disease recommended offering both options with the individual patient assessed for risk and benefit of each.[13] Our patient was referred for neuro-

Figure 4.
Depiction of seated cervicothoracic spinal manipulation with the cervical spine maintained in a neutral position. An upward force is generated by the provider as part of this manipulation maneuver. The patient is not pictured.

surgical evaluation to discuss surgical intervention which was deemed not to be appropriate. There is a paucity of literature to guide clinicians in the nonsurgical management of pain conditions in patients with DBMA.

As the patient presented 10 years after the initial disease presentation and without recent progression of signs or symptoms, we considered the patient to be neurologically stable and thus proceeded with a trial of care. At each session, we assessed for appropriateness of high velocity, low-amplitude thoracic manipulation by applying pre-loading positioning consistent with the given spinal manipulation technique and sought patient feedback on tolerance. With the potential risk of injury to the spinal cord from cervical range of motion in the presence of DBMA, cervical spine manipulation was considered to be contraindicated and was not performed. Spinal manipulation was initiated in only the thoracic spine, with addition of the cervicothoracic junction after established tolerance without adverse event. In accordance with guidelines[13] and prior patient education, cervical flexion was avoided during all manual treatment and rehabilitative exercise instruction. The patient was able to tolerate the exercise as well, although he struggled to sustain strength improvements due to fatigue of the upper extremities with repetitions. The Y, T, W, L exercises prescribed have been demonstrated to induce moderate to high activation of the periscapular and lower trapezius muscles.[23]

The score change of the NDI did not achieve the minimal clinically important difference (MCID) of a five-point (10%) decrease.[22] However, the NPRS score did decrease by four points (meeting MCID) and subjective improvements were noted by the patient. Treatment focused on providing pain relief and improving the mobility of the neck, shoulder, and back musculature. The authors hypothesize that atrophy of distal upper extremity muscles may have led to increased stress on the spinal and proximal muscle groups. Further research into this population may prove difficult given its rarity. The majority of current evidence exists from individual cases and expert consensus guidelines and provides no insight on manual therapy in the long-term management of this condition.[13] Future case reports and cohorts detailing conservative management by chiropractors, physical therapists, occupational therapists, and other manual therapy providers may help to guide nonpharmacologic management. The authors theorize that introducing manual therapy and other conservative interventions following the plateau of disease progression could positively impact patient symptom management and quality of life.

Limitations

There is a paucity of evidence for conservative management in the setting of stable DBMA. Patient safety and provider confidence with treating rare disorders should be considered before beginning any trial of care. Bimelic amyotrophy is a rare condition and the findings of our report of a single case may not be generalizable. While our patient did not report any adverse events, this does not mean that adverse events are unlikely to occur with manual therapy approaches being utilized in patients with DBMA, and trials of care should be undertaken with caution.

Summary

Bimelic amyotrophy is a rare and complicated condition with the potential for serious adverse events (e.g., spinal cord trauma). This case demonstrates conservative

management of concurrent myofascial pain in a patient with known DBMA without experiencing any reported adverse events. Co-management with physical medicine & rehabilitation, or other specialty physicians, is appropriate in the presence of stable bimelic amyotrophy.

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Authors contributions

KM, MP, SP, and CD all cared for the patient. KM, MP, and CD drafted, and all authors critically revised and approved the final manuscript.

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