

Treatment of a patient with posterior cortical atrophy (PCA) with chiropractic manipulation and Dynamic Neuromuscular Stabilization (DNS): A case report

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Objective: Posterior cortical atrophy (PCA) is a rare progressive neurodegenerative syndrome which unusual symptoms include deficits of balance, bodily orientation, chronic pain syndrome and dysfunctional motor patterns. Current research provides minimal guidance on support, education and recommended evidence-based patient care. This case reports the utilization of chiropractic spinal manipulation, dynamic

Objectif : L'atrophie corticale postérieure (ACP) est un syndrome neurodégénératif évolutif ayant des symptômes inhabituels, notamment un manque d'équilibre, un trouble d'orientation du corps, des douleurs chroniques et une organisation motrice dysfonctionnelle. Les recherches actuelles offrent peu de renseignements à propos du soutien, de l'éducation et des soins basés sur des données probantes recommandés aux patients. Cette étude de cas analyse l'utilisation de manipulations chiropratiques vertébrales, de stabilisations

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Disclaimers: This paper was written as part of the Research Honors program at Palmer College of Chiropractic, Davenport IA. This original article received the Research Honors Award presented by the Palmer Center for Chiropractic Research, in June 2013.

Principal investigator and corresponding author: Vinicius T. Francio, DC, MS

Sources of support: None

Patient provide informed consent.

Conflicts of Interest: The authors declare no conflicts of interest.

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neuromuscular stabilization (DNS), and other adjunctive procedures along with medical treatment of PCA.

Clinical features: A 54-year-old male presented to a chiropractic clinic with non-specific back pain associated with visual disturbances, slight memory loss, and inappropriate cognitive motor control. After physical examination, brain MRI and PET scan, the diagnosis of PCA was recognized.

Intervention and Outcome: Chiropractic spinal manipulation and dynamic neuromuscular stabilization were utilized as adjunctive care to conservative pharmacological treatment of PCA. Outcome measurements showed a 60% improvement in the patient's perception of health with restored functional neuromuscular pattern, improvements in locomotion, posture, pain control, mood, tolerance to activities of daily living (ADLs) and overall satisfactory progress in quality of life. Yet, no changes on memory loss progression, visual space orientation, and speech were observed.

Conclusion: PCA is a progressive and debilitating condition. Because of poor awareness of PCA by physicians, patients usually receive incomplete care. Additional efforts must be centered on the musculoskeletal features of PCA, aiming enhancement in quality of life and functional improvements (FI). Adjunctive rehabilitative treatment is considered essential for individuals with cognitive and motor disturbances, and manual medicine procedures may be considered a viable option.

(JCCA 2015; 59(1):37-45)

KEY WORDS: chiropractic, spinal manipulation, neurodegenerative disease, physical and rehabilitation medicine

neuromusculaires dynamiques et d'autres procédures d'appoint connexes au traitement médical de l'ACP.

Caractéristiques cliniques : Un homme de 54 ans s'est présenté à une clinique de chiropratique avec des douleurs lombaires non précisées, des troubles de la vue, de légères pertes de mémoire et un contrôle cognitif moteur inadéquat. Après un examen clinique, une IRM du cerveau et une tomographie par émission de positons, l'ACP a été diagnostiquée.

Intervention et résultats : On a utilisé des manipulations chiropratiques vertébrales et des exercices de stabilisation neuromusculaire dynamique à titre de soins d'appoint au traitement pharmacologique conservateur de l'ACP. Selon l'évaluation des résultats, le patient voit une amélioration de sa santé de l'ordre de 60 %, soit le retour des fonctions neuromusculaires, l'amélioration de la locomotion, la posture et l'humeur, une diminution de la douleur, la capacité à réaliser les activités de la vie quotidienne, et une satisfaction d'ensemble de la progression de la qualité de vie. Toutefois, on n'a pas observé d'amélioration sur le plan de la progression de la perte de la mémoire, l'orientation visuelle dans l'espace et la parole.

Conclusion : L'ACP est un trouble progressif et débilitant. Souvent, les patients souffrant d'ACP ne reçoivent pas tous les soins dont ils ont besoin, car les médecins connaissent mal ou ne connaissent pas l'ACP. On doit investir plus d'efforts dans les caractéristiques musculosquelettiques de l'ACP dans le but d'améliorer la qualité de vie et la fonctionnalité. Le traitement d'appoint de réadaptation est considéré comme essentiel aux patients atteints de troubles cognitif et moteur, et il convient de considérer les procédures de médecine manuelle comme étant une option viable.

(JCCA 2015; 59(1):37-45)

MOTS CLÉS : chiropratique, manipulation vertébrale, maladie neurodégénérative, médecine physique et de réadaptation

Introduction

Posterior cortical atrophy (PCA) is a rare progressive neurodegenerative syndrome characterized by deficits in higher-order visual processing, in which memory, judgment and insight is preserved.¹ This disorder may be considered a variant of Alzheimer disease, but can also result from corticobasal degeneration, Creutzfeldt-Jakob, dementia with Lewy bodies and/or subcortical gliosis. Typically, the age of onset for PCA is 50-65 years old. However, prevalence and incidence are unknown. Furthermore, etiology often remains uncertain until postmortem examination.² Clinical features of PCA constitute a wide variety of signs and symptoms; nevertheless, the most frequent include hemianagnosia (deficit in awareness of one side of space), optic ataxia (lack of coordination between visual inputs and hand movements, resulting in inability to reach and grab objects), visual agnosia (an impairment in recognition of visually presented objects), alexia (difficulty to understand written words), acalculia (difficulty with simple mathematical tasks), and agraphia (loss in the ability to communicate through writing).³ Functions related to the parietal, occipital, and occipitotemporal regions of the brain may be affected, and may include left-right disorientation, language skills, and space perception deficits.¹⁻³ Some unusual symptoms are potentially linked to different brain arrangements, such as visuovestibular and pontinomedullary reticular formation interactions, evoking a wide range of phenomena, including disturbance of balance, bodily orientation, chronic pain syndrome, decomposition of motion and dysfunctional motor patterns.³⁻⁴ Early identification and increased awareness is key for appropriate management of PCA, since the condition is debilitating and rapidly progressive in its early years. Nevertheless, current research provides minimal guidance on support, education and recommended evidence-based patient care.⁵ Benson and colleagues¹ recognized the clinical presentation of PCA for more than 2 decades and yet, compared to other conditions, it is essentially overlooked by researchers and clinicians.

Therefore, the aim of this study is to describe the case of a 54-year-old male with PCA who sought chiropractic care, as well as how the condition was co-managed along with conservative pharmacological care.

Case report

A 54-year-old male sought care for musculoskeletal pain

associated with stiffness, and tightness in his neck and back. The symptoms began 6 months earlier. About that time, he noticed he was bumping into things, forgetting familiar faces, and was having difficulty reaching out to pick up and grasp objects. Activities such as tying his shoes and balancing his cheque book were becoming progressively more difficult. His symptoms were primarily affecting activities of daily living (ADLs), such as the ability to drive, perform household chores, dress, and his leisure activities. Family members also reported fluctuations with mood and behavior. Previous medical history was unremarkable, except for a triple bypass surgery in the past.

Upon physical examination performed by the chiropractic physician, satisfactory mental status was accomplished via the mini-mental status test. However, decreased nasal field, and peripheral vision, mainly in the left eye, was noted. Along with reported blurred vision during eye examination. Glabellar reflex (tap sign) produced persistent response only in his right eye, and finger snap activation in the auditory canal activated bilateral blink response on his right ear, and only right blink response (unilateral) on the left ear. Eye movements were conjugated, but he was unable to follow the examiner with increased speed and specific smooth pursuits, eyes opposite and prosaccade eye exercises were conducted. During the neurological exam, strong concentration and reinforcement was required due to difficulty regarding right-left discrepancy. Presence of hyper-reflexive knee jerks (+3) bilaterally, grossly intact plantar reflex, and sensory examination was found. Manual muscle testing indicated very mild left upper extremity flexor angulation and lower extremity extensor angulation. Bilateral increase pronator drift and right-sided wrist cogwheel rigidity was noted. Hoffman's sign was positive bilaterally. Other pathological reflexes were negative. Inability to perform finger-to-nose on the left, and difficulty with graphognosia, barognosia and stereognosia testing were noted. Spinal exam revealed overall loss of flexibility and range of motion in all planes, with anterior head carriage, and hyperkyphotic antalgic posture. Diffuse superficial tenderness to palpation in the back with spinal core stabilizers weakness and joint hypomobility was found. Functional motor pattern analysis revealed mild akathisia and deconditioning syndrome.

Spinal radiographs showed multilevel cervicothoracic

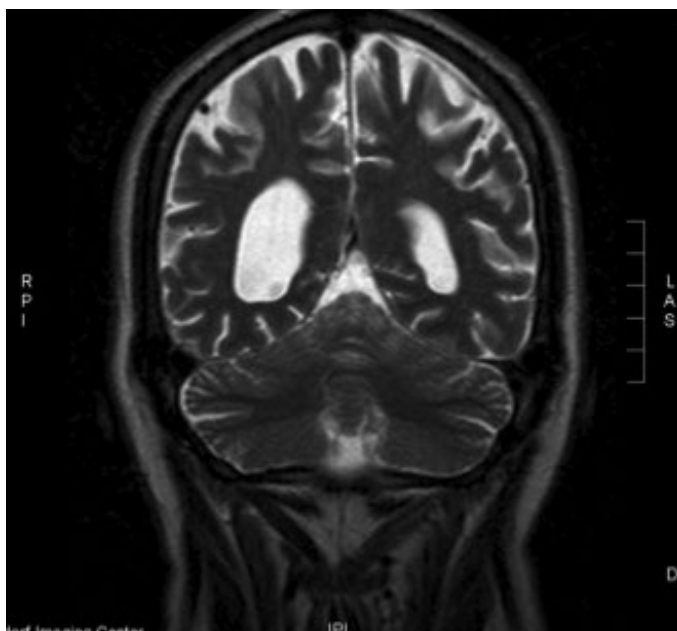


Figure 1.

Coronal plane; Brain magnetic resonance imaging (MRI) revealing enlarged ventricles

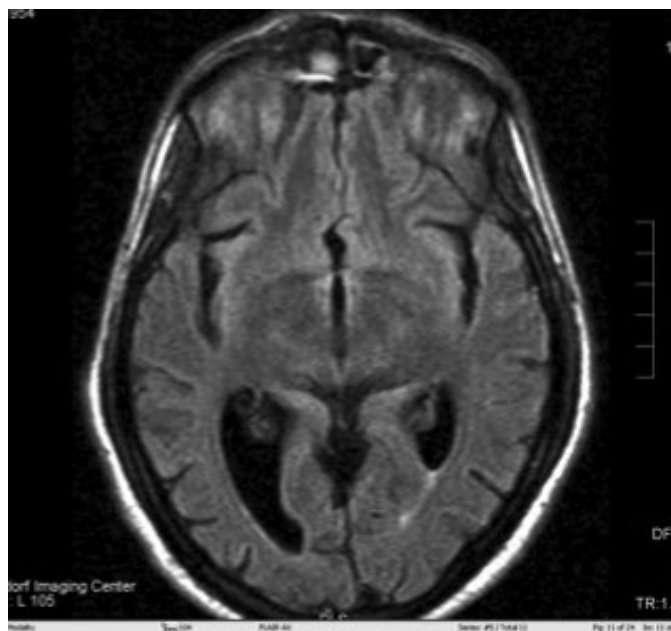


Figure 2.

Transverse plane; Brain magnetic resonance imaging (MRI) revealing enlarged ventricles

spine degenerative disc/joint disease, reduction of lordotic curve and moderate anterior head carriage. The neurologist ordered first a magnetic resonance image (MRI) of the brain (see Figure 1 and 2), which revealed mild ventricle enlargement due to ex vacuo dilatation secondary to the diffuse, primarily parieto-occipital cortical atrophy. In addition, a positron emission tomography (PET) study was performed, which indicated lack of radiotracer uptake in the parieto-occipital cortices bilaterally, more pronounced and with a larger volume of involvement on the right (see Figure 3). This study was performed to determine the areas of the brain that were not functioning properly. The clinical diagnosis of posterior cortical atrophy (PCA) was accomplished after a thorough investigation, with corroborative findings from the neurologist consult. Communication between the neurologist and the chiropractic physician regarding exchange of records, treatment and outcomes occurred several times. The neurologist recommended daily dosage of Donepezil, a common medication prescribed for Dementia-like symptoms, which is standard of care for PCA, and regular follow-up regarding the condition's prognosis. The patient decided to adhere to the suggestion, and also pursue chiropractic

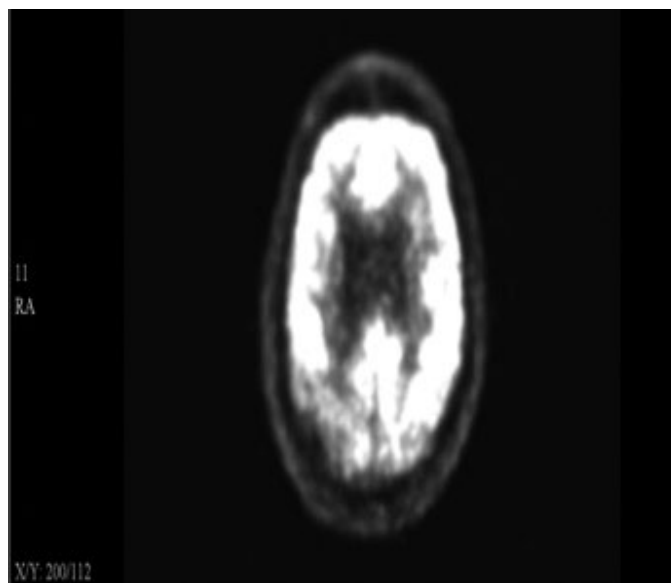


Figure 3.

Brain positron emission tomography (PET) scan with FGD radiotracer. Brain positron emission tomography (PET) scan revealing lack of radiotracer uptake in the parieto-occipital cortices bilaterally, more pronounced and with a larger volume of involvement on the right.

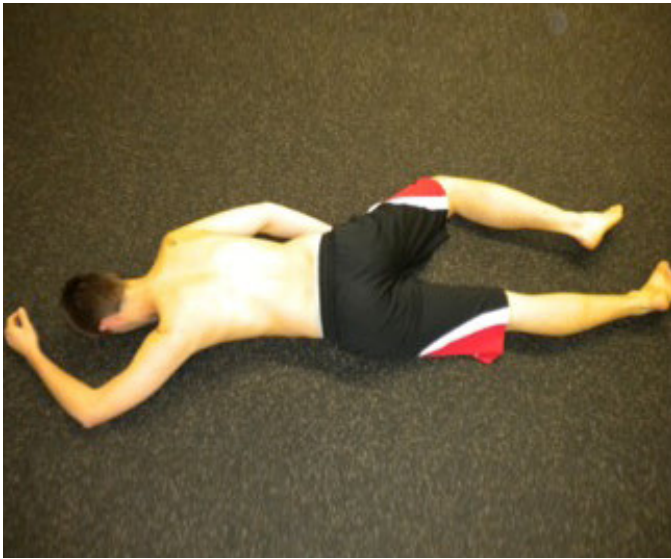


Figure 4.

Dynamic Neuromuscular Stabilization (DNS) procedure utilizing crawl position 1 with treatment points. This is not the actual patient, rather only a model illustration about the procedure sequence.



Figure 5.

Dynamic Neuromuscular Stabilization (DNS) procedure utilizing crawl position 2 with treatment points. This is not the actual patient, rather only a model illustration about the procedure sequence.

manipulative therapy and musculoskeletal rehabilitation hoping to enhance his neuromuscular function.

Our treatment consisted of manual medicine and rehabilitation procedures focused on the neuromuscular features of PCA, in a 42-week period initially, and then another 13-week period. Frequency and duration of treatments were variable throughout these periods. At times, visits were twice a week, others once every two weeks due to conflicting schedule. Treatment visits consisted of the use of vibratory stimuli therapy over the distal extremities (to enhance vibratory and proprioceptive input to the CNS), with the use of a 128hz tuning fork pre and post high-velocity low-amplitude (HVLA) spinal manipulation. Proprioceptive retraining exercises standing on balance training ball performing finger-nose exercise and catching various size objects was also incorporated. DNS therapy was performed in the supine and prone position, with 2 or 3 doctors asking the patient to perform specific motor pattern motions, average approximately 12 minutes (see Figure 4, 5 and 6). At-home therapeutic exercises for thoracic extension, and hamstrings/adductor stretch-hold exercises were encouraged.

Progress evaluation focused on functional improve-

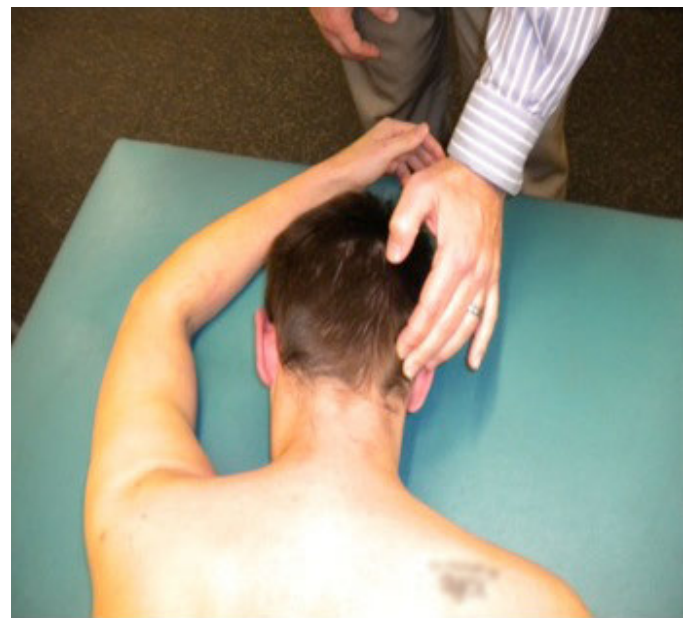


Figure 6.

Dynamic Neuromuscular Stabilization (DNS) procedure utilizing crawl position 3 with treatment points. This is not the actual patient, rather only a model illustration about the procedure sequence.

ments (FI) involving activities such as the ability to drive, dress, perform household chores, climb stairs, play golf, motor behavior, and overall global health status. The response to the DNS therapy, and proprioceptive exercises was used every visit to monitor progress. In a 10-month period, the Health Status Questionnaire (HSQ) demonstrated a 38% functional improvement. Within a 13-month period, HSQ and Back Bournemouth Questionnaire (BBQ) demonstrated a 60% improvement, which is extremely significant regarding musculoskeletal pain and FI. These two outcome measurements are a short, self-reported questionnaire that measures objective improvements with different dimensions in patients with musculoskeletal pain, aiming pain and disability, but also takes the affective and cognitive aspects of pain into account. They are well known for their validity, reliability and responsiveness. Likewise, functional impairments previous noted, such as inability to play golf, self-care (dressing), climbing stairs and motor control were enhanced. Better control of involuntary motion, right and left discrimination, and refined motor skills for reaching objects were observed. No progression in memory loss, space orientation, and speech disturbances was observed.

Significant functional improvement of neuromusculoskeletal control and pain was accomplished. Altered movement patterns between agonists and antagonists secondary to dysfunctional sensory inputs were restored with improvement in the synergistic function between different muscle groups and fasciae. In this case, the integration of chiropractic care with DNS resulted in a number of remarkable musculoskeletal restorations, with improved mechanics and tolerance to activities of daily living. The patient was encouraged to continue with conservative care, in addition to daily routine of exercise, and active rehabilitative program.

Discussion

PCA is a progressive, debilitating, condition characterized by difficulties in visuospatial tasks, writing, and motor control.⁶ The loss of cognitive motor function related to PCA, and the presence of prominently asymmetric limb apraxia, illustrates an uncommon feature of this syndrome.⁷ Musculoskeletal examination in most cases is unremarkable; however, severe progressive visual and cognitive findings are common. According to Kas and colleagues⁸, the visual symptoms are perhaps more

likely to be detected than other deficits due to its greater impairment level, nevertheless, neuromuscular dysfunctions are equally as important to address. Because of poor awareness of PCA by physicians, patients usually receive incomplete care. Adjunctive rehabilitative treatment is considered fundamental for individuals with cognitive and motor disturbances. Special efforts must be centered on the neuromuscular features of PCA, and appropriate management with manual medicine, rehabilitation, exercises and cognitive therapy is recommended as an option for care, aiming to enhance quality of life, and improve neuromusculoskeletal functional.⁹ According to Kolar¹⁰, influenced by the work of Vojta, Dynamic Neuromuscular Stabilization may be considered an adjunctive therapy in such cases to help enhance specific functions of the musculoskeletal system. Such an approach was chosen by the clinician as the most suitable therapy to facilitate passive movement patterns in this case, while enhancing primitive subcortical kinesiological patterns to stimulate appropriate neuromuscular function.

Dynamic neuromuscular stabilization studies suggest that our motor behavior is predetermined. During motor development, characteristic muscle synergies are stored in the brain as a matrix, responsible for posture, locomotion, and movement patterns.¹⁰ The highest level of integration occurs at the cerebral cortex, which allows us the ability to develop new skills, refines, and predicts specific movements. The mechanism, which the nervous system responds to external stimuli and adjusts future responses based on previous outcomes, is defined as 'functional neural plasticity,' and its understanding is fundamental in the field of neuromusculoskeletal rehabilitation.¹⁰ The function of each part of the nervous system is dependent on the central integrative state of each hemisphere, which is determined to a large extent by the awareness of afferent stimulation from the periphery. This fact supports the concept of functional hemisphericity, which involves asymmetric cortical modulation of various forms of input that can result in decreased postural tones, spinal stiffness, pelvic floor weakness, and inhibition of the intrinsic stabilizer spinal muscles.¹⁰⁻¹¹ The pontomedullary reticular formation receives nearly 90% of the output dedicated to the neuraxis and cortex modulation, ipsilaterally. The impact that an asymmetrical cortical output has clinically in the neuromusculoskeletal system is evident with symptoms such as, decrease in muscle tone ipsilaterally,

ipsilateral pain syndrome, generalized spinal stiffness and deconditioning syndrome.^{11,12,13}

As evidenced by the present case, this clinical presentation of PCA was associated with musculoskeletal dysfunctions and asymmetric cortical modulation. The patient's presentation with ipsilateral flexor angulation of upper limb, extensor angulation of lower limb, spinal stiffness, deconditioning, and imbalance motor patterns supports the application of manual medicine and rehabilitation as adjunctive therapeutic procedures. SMT and DNS were utilized focusing on movement pattern analysis. According to Vacek¹⁴ if the central nervous system (CNS) is not functioning normally, the postural program will change and kinesiological analysis of postural activity and re-actions can be used to evaluate the CNS. Also, mobilization and manipulation of spinal joints and fascia may influence global movement patterns allowing higher levels of cortical function, stimulating appropriate cortex output through balanced peripheral afferent stimuli to the PMRF.¹¹ Other therapeutic procedures included inhibitory stretching protocols to the facilitated muscles as described in the upper and lower cross syndrome, as well as facilitating exercises to increased activity in the inhibited muscles.¹³ Balance, vibration therapy over the extremities and proprioception exercises using solid ground and 2-way wobble boards were used with eyes open and closed, to stimulate appropriate cortical awareness of proprioceptive and cerebral pathways.^{14,15}

According to several studies^{11,13,16,17}, a number of pathways may explain the potential effects of manual medicine procedures on the neuraxis, and therefore theoretically corroborate the positive neuromuscular improvements in this case. These include, but are not limited to, exciting spinoreticular pathways and dorsal column pathways to the PMRF, modulation of vestibulosympathetic pathways and vestibulocerebellar activation of the nucleus tractus solitarius, dorsal motor nucleus of vagus, and nucleus ambiguus. Yet, spinal manipulation and afferent peripheral stimulation may result in brain hemisphere influence via descending excitation of PMRF pathways, inhibitory control of IML cell column, which may alter central integration of the brain stem and hypothalamus via spinoreticular and spinothalamic afferent direct connections.^{11 14-19} These manual medicine procedures tend to facilitate global stability and enhance musculoskeletal functional patterns addressing a number of anatomical

structures that help maintain appropriate neuromuscular stimulation. Mechanoreceptors, proprioceptors, Golgi tendon organs, muscle spindles, and her sensory organ are all directly influenced by mobilization and directive manual therapy approach.²⁰⁻²³ Ultimately, extensive sensory input into the CNS contributes to the neuromuscular adaptation demanded by the cortex needs in its process of reorganization. Hence, manual medicine procedures play a fundamental role in adding sensory stimulation to peripheral receptors, known to influence motor response patterns, which directly affect posture, balance, locomotion, and musculoskeletal function.²⁴⁻²⁶

The manual medicine procedures used in this case improved the neuromuscular function and quality of life of this patient. Yet, no changes in memory, visual spatial or writing ability were seen. Furthermore, the results of this case report, and likewise cases^{27,28}, where similar procedures were utilized, supports the value of such procedures in the co-management of neurological conditions associated with musculoskeletal dysfunctions. Chiropractic, DNS, and adjunctive therapies can help manage neuromuscular compensations and altered movement patterns, secondary to improper spinal mechanics, and dysfunctional sensory inputs between muscle groups focusing in enhancing structural mobility, postural integrity and enhance neuromuscular function.^{29,30} Our clinical approach for the management of musculoskeletal dysfunction, such as in this case, emphasizes the utilization of manual stimulation of specific zones of the body by properly mobilizing/manipulating joints to evoke predetermined efferent motor patterns by the central nervous system, facilitating sensory input to enhance cortex assimilation, and appropriate re-organization or adaptation.

Limitations

This report has the limitations of all case reports that represent the experience of a single patient. Therefore, findings may not be generalizable to other patients.

Conclusion

As evidenced by current literature and substantiated by the present case, PCA is a progressive neurological disorder associated with higher cortical dysfunctions, and impaired neuromusculoskeletal ability. Medical research provides minimal guidance regarding the appropriate management of PCA. Yet, multidisciplinary care is con-

sidered essential for successful results. Chiropractic, DNS and other procedures were utilized in this case. There was no progression in memory loss, speech or visuospatial orientation noted, though significant musculoskeletal improvements were achieved, as endorsed by an improvement in 60%, according to the HSQ, and BBQ outcome assessment tools, especially in quality of life, and musculoskeletal function. Our goal with this study is to increase multidisciplinary management awareness, and include chiropractic care in future research considering its contributory use for functional enhancement in the co-management of neurological disorders with musculoskeletal dysfunctions.

Acknowledgments

We would like to thank Dana Lawrence, DC, MMedED, MA for his comments and review of this article.

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