

REMISSION OF HEADACHE AND NECK PAIN FOLLOWING CHIROPRACTIC MANIPULATIVE TREATMENT IN A PATIENT WITH NEUROFIBROMATOSIS

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ABSTRACT

Objective: To describe the case of a 21-year-old male with a 10-year history of neurofibromatosis type 1 (NF1) who complained of nuchal pain (2 on a 0-10 point numeric pain rating scale, NRS) and right frontal headache (8 on NRS) for six months.

Clinical features: The painful symptoms were associated with a great impact on the sleep and routine tasks during the month preceding the first presentation to the practitioner. The patient scored an 84% on the Neck Disability Index (NDI). Spinal palpation revealed restricted range of motion at C2/3, C4/5, C5/6, T1/2, T3/4 and T7/8 vertebral segments. Radiographic evaluation of the cervical spine revealed reduced cervical lordosis with Cobb angle 3°.

Intervention and outcomes: Chiropractic treatment aimed to reduce pain, muscle hypertonicity and restore spinal mobility. Treatment consisted of thermal ultrasound therapy, cervical manipulation with a high-velocity, low-amplitude force, and skin to skin contact manual adjustment. As the result of 12-month treatment, the patient experienced a resolution of neck pain and headache. He reported a reduced headache score from 8 to 0 on a scale of 10. His NDI reduced from 84 % to 8%, and cervical lordosis was restored from 3° to 20° as measured by the Cobb angle between the inferior endplate of C2 to the inferior endplate of C7.

Conclusion: This case report details the long-term recovery of a patient with NF1 from severe headaches and neck pain following chiropractic treatment. It provides evidence and highlights the value of chiropractic intervention in improving neuromuscular functions and resolving cervicogenic headache in a patient with NF1, especially when the problems cannot be effectively solved by pharmacological or other conservative means. Chiropractic manipulative therapy may be a viable option as conservative management of musculoskeletal dysfunction resulting from NF1.

Keywords: chiropractic manipulation therapy, headache, neck pain, neurofibromatosis

Introduction

Neurofibromatosis (NF) is a group of three genetic disorders that cause tumors to develop anywhere in the nervous system. The three types are neurofibromatosis type 1 (NF1), neurofibromatosis type 2 (NF2), and schwannomatosis [1]. NF1 is the most prevalent of this group and characterized by benign tumor growth on nerve sheaths called neurofibromas and cutaneous spotting called café-au-lait spots and multiple tumors starting from the central and peripheral nervous system [2]. Although the

function of the mutated, naturally occurring protein neurofibromin is not completely understood, it appears to regulate Ras proteins that promote cell division and growth. The presentation and complications of NF1 are highly variable, even within immediate family members [3]. NF2 is characterized by the development of benign, slow-growing tumors in both ears (acoustic neuromas), which can cause hearing loss. Schwannomatosis is rare disorder that can cause schwannomas to grow along the cranial, spinal and peripheral nerves [2].

Around 50% of NF1 patients develop significant musculoskeletal manifestation, such as scoliosis, congenital pseudarthrosis, bone cysts, cortical bone thinning, and subperiosteal bone hyperplasia [2]. Decreased serum vitamin D levels have been noted in patients with NF1. It is possible that vitamin D and neurofibromin could interact at the level of cell proliferation and reducing the pro-apoptotic and antiproliferative effects of vitamin D in patients with NF1, promoting the tendency to form tumors [4]. The musculoskeletal complaints could be associated with decreased activated vitamin D levels, increased parathyroid hormone levels, and increased markers of bone breakdown. Due to NF1 influencing multiple systems, patients are likely to benefit most from a multidisciplinary treatment strategy [2].

The standard of care for the management and follow-up of patients with NF consists of an annual physical examination and the treatment of symptomatic manifestations [5]. Physiotherapy focusing on muscle stretching and strengthening regimens is beneficial in patients with NF1 [6]. Patients with NF1 may seek chiropractic care if they experience musculoskeletal pain, as well as clinical neurologic deficits, as a result of decreased spinal range of motion and muscular strength caused by the presence of neurofibromas [4,7]. The purpose of this case report is to present the results of a five-year conservative management for neck pain, cervicogenic headache and spinal deformity in a patient with NF1. Chiropractic intervention may be a viable option for addressing musculoskeletal pain and cervicogenic headache resulting from NF1.

Case report

A 25-year-old salesman with type I neurofibromatosis (NF1) diagnosed by a dermatologist 10 years ago complained of pain on the right side of the head and neck for six months. The symptoms worsened in the cold temperature, and aggravated by mental stress. He had no history of trauma or other neurological disorders. The patient was also concerned about his painless bumps (neurofibromas) on the skin that caused him distress.

The patient's self-reported peak neck pain intensity was 2/10 on a 0-10 point numeric pain rating scale (NRS) and 8/10 for the frontal headache. He described as feeling like a pulsating squeezing around the right forehead, lasting about an hour and 2-3 times per day that disturbed sleep and daily activities over the preceding month. The patient had a prior negative brain MRI and neurological assessment of his headache. He had received a coincidental diagnosis of attention deficit disorder and learning disability by a clinical psychologist. He was given a diagnosis of chronic headache by his primary care physician. Non-steroid anti-inflammatory drugs, anticonvulsants, narcotics, physical therapy, acupuncture, and psychotherapy had been used to manage his headache but did not provide adequate relief and were discontinued. His therapeutic reliance was analgesics alone.

The patient presented with a restricted neck range of motion and loss of cervical lordosis. Upon inspection, several flat, light brown spots (café-au-lait spots) were located over the back skin (Figure 1). Multiple soft, bluish nodular lesions (cutaneous neurofibromas) appeared on back and upper extremities bilaterally. Physical examination yielded tenderness and hypertonicity of bilateral cervical extensor muscles from C3-7 and restricted range of motion to 40° at cervical extension (normal reference >60°) and 50° at rotations (normal reference >80°). Spinal palpation revealed intersegmental dysfunction at C2/3, C4/5, C5/6, T1/2, T3/4 and T7/8 vertebral segments. The assessments of eye movement, cerebellar, neurological, orthopedic, and walking functions were normal. His Neck Disability Index (NDI) was rated at 84%. Total NDI score ranges from 0 to 50 with the score converted to a percentage, higher scores indicating greater disability. Radiographic evaluation of the cervical spine revealed reduced cervical lordosis with Cobb angle 3° (Figure 2). Based on his clinical findings and patient history, he was diagnosed as having a cervical syndrome (ICD-10 code M53.82) plus secondary headache (ICD-10 code was G44.88: Chronic headache attributed to other head and/or neck trauma or injury). The International Classification of Headache Disorders 3rd edition [8] code 11.2.1: Cervicogenic headache.

Treatment and follow-Up

Chiropractic treatment aimed to reduce pain, muscle hypertonicity and restore spinal mobility. The patient began a regular schedule of chiropractic treatment consisting of thermal ultrasound therapy, cervical manipulation with a high-velocity, low-amplitude force, and skin to skin contact manual adjustment treated daily for the first 6 days for pain relief. He reported a reduced pain score of his headache from 8/10 to 5/10 on the NRS. Treatment session was then applied 3-times-weekly for additional 4 weeks. The patient described a decrease of frequency and duration of his headache, improvement of sleep quality, and reduction of reliance on analgesics. The second phase of treatment performed twice-weekly for another 8 weeks. After 13-week treatment session, the patient reported relief from neck pain, and improved strength and mobility of the neck. The NDI score returned to 8%.

The patient was treated monthly following the initial 13 weeks of therapy. In this phase of care management was supplemented with the prescription of a home based posture correction exercise regimen. Upper trapezius stretches, scapular retraction, and cervical retraction neck exercise were prescribed in 10 repetitions, and 3 sets a day; however, patient only followed the prescription in the first week and was uncompliant due to a

busy work schedule. The patient remained asymptomatic for 12 months. The patient then took a break from treatment for three months. With the cessation of maintenance care, the headache quickly reappeared but resolved immediately after treatments (2 times a week for 2 weeks). Radiographic re-evaluation performed in the third year showed a change in cervical lordosis from Cobb angle 3° to 10°. After exacerbation of the symptoms with the three-month cessation of care, the patient chose to return to maintenance care at one monthly interval to remain symptoms free.

Maintenance care was continued into the fifth year with no return of presenting signs and symptoms whilst maintaining normal activities of daily living. His re-evaluation radiograph demonstrated a cervical lordosis Cobb angle of 20° (Figure 2). The cutaneous neurofibromas (yellow arrow) had increased in both size (largest from 1cm to 2cm) and number (55 to 70)

Discussion

Musculoskeletal complaints are the most common orthopaedic manifestation of NF1 [2] and is considered to be caused by osteomalacia, intraspinal neurofibromas that erode and infiltrate bone, and endocrine dysfunction [9]. Rib penciling, vertebral scalloping, dumbbell lesions, and dural ectasia are all examples of dystrophic alterations in NF1. Less often occurring is dystrophic alterations including: transverse process spindling,

spinal wedging and rotation, foraminal enlargement, wider interpediculate distance, and pedicle dysplasia [10]. None of these changes were noted in this case.

Scoliosis is the most common orthopaedic finding in NF1 patients and often develops around the age of 10, possibly be associated with osteopenia and subsequent dysplastic bones [11]. Individuals with NF1 will require routine assessment with MRI or CT scans to determine the deformity accurately [9], and those with musculoskeletal dysfunctions should be appropriately co-managed with a primary care provider or neurologist. The management in this case brings into question whether chiropractic manipulative therapy can have a role in the treatment of neck pain and spinal deformity in patients with NF1. Evidence on the effectiveness of chiropractic for spinal pain in NF1 is limited. The chiropractic Thompson-drop technique (mechanically assisted manipulation) has been described to be effective in a NF1 patient with low-back pain [12] and the chiropractic cervical flexion-distraction technique has been shown to be effective and safe for addressing a patient's cervical joint restrictions [6]. With regard to this patient, low amplitude high velocity thrust (HVLA) manipulations with prescribed home exercise programs were found to improve musculoskeletal dysfunction provided there are no gross osseous contraindications. The mechanism of the effect provided by manipulation provided in this study may possibly due to a variety of possible effects including: correction of spinal dysfunction, breaking-up joint adhesions, mobilizing restricted tissues, facilitation of trunk muscle strength, and possibly releasing the nerve entrapped by limited movement. The treatment effects may eventually facilitate the normalization of muscle strength and improved functional mobility that had previously been lost [13,14].—The positive changes observed after chiropractic manipulative therapy may be unrelated to treatment specificity but rather is a manifestation of a systemic effect of functional changes related to the biomechanical interdependence of musculoskeletal structures [15]. Additionally, we have previously hypothesized in a case report that autonomic dysfunction may correlate with tension type headaches and may also be relevant in the improvements noted in this case [16].

It is noteworthy in this case that the patient utilized chiropractic manipulative therapy as a conservative management of his symptoms in response to previous failed medical and allied health attempts to control his pain and dysfunction and to avoid medications. It is important that manipulation of the spine should be applied with great caution in patients with NF. It is possible for tumors growing on and near the spine may lead to an overall weakening of the spine which may be associated with adverse outcomes [17]. To mitigate this possible eventuation prior to therapy, a screening radiograph is warranted prior to the application of high velocity spinal manipulative therapy.

The primary limitation of this case report is that the etiology of the neck pain and headache was unknown and the application of chiropractic treatment was based on clinical possibilities with the long-term consequences of the chiropractic treatment beneficial in this particular case. Furthermore, radiography has inherent limitations such as pitfalls in relation to radiographic technique and patient positioning. As the clinical report is not research we cannot conclude that the treatment caused the effects, only

that they were associated with them and that this may have happened for reasons not noted by the practitioner or the patient.

Conclusion

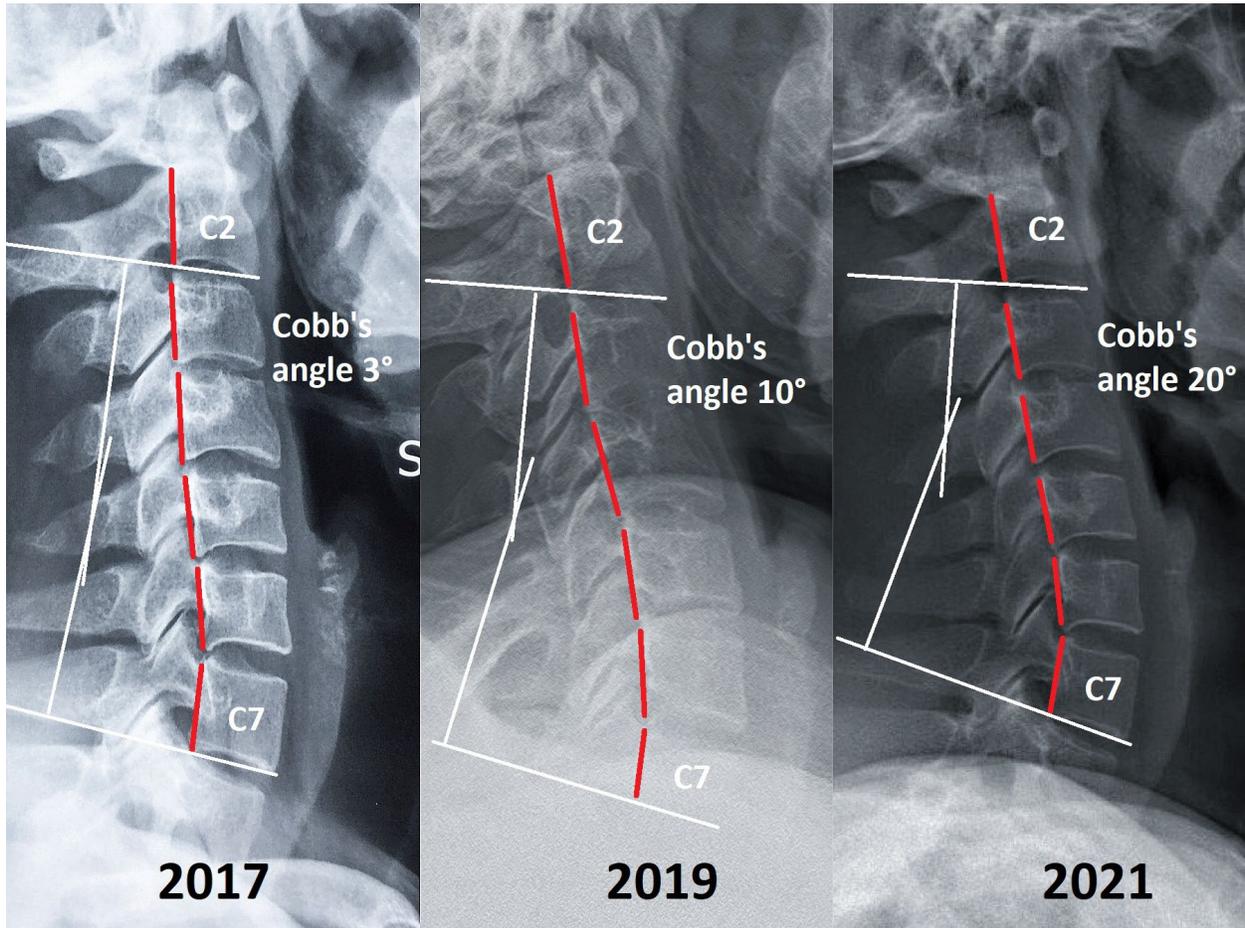
This case report details the long-term recovery of a patient with NF1 from severe headaches and neck pain following chiropractic treatment. Musculoskeletal symptoms occur in NF-1 patients just as they do in the general population. The reasons for neuromuscular dysfunction vary, and the appropriate treatment is determined by the etiology, scope and severity of the symptoms. The goal of normalizing spine curvature in this context may help to alleviate cervicogenic headache and musculoskeletal complaints associated with cervical misalignment.

Legends

Figure 1: *Multiple flat brownish spots (café-au-lait spots) and raised brownish nodules (neurofibromas) are identified on the back of the 25-year old patient with neurofibromatosis. Compare photos between 2017 and 2021, the cutaneous neurofibromas (yellow arrows) had increased in both size and magnitude.*



Figure 2: *Sagittal cervical radiographs. (2017) Initial radiograph showed reduced lordosis with 3° Cobb angle. (2019) At the 3rd year follow-up visit, the cervical curvature improved as compared with the initial radiograph with Cobb angle 10°. (2021) At the fifth year follow-up visit, the cervical curvature was restored to Cobb angle 20°.*



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