Effects of Cervical Deep Muscle Strengthening in a Neck Pain: A Patient with Klippel-Feil Syndrome

YOUNGSOOK BAE, PT

Department of Physical Therapy, College of Health Science, Gachon University: Yeonsu-dong, Yeonsu-gu, Incheon City, Republic of Korea

Abstract. [Purpose] This study aimed to identify the effects of cervical deep muscle strengthening (CDS) on neck pain in a patient with Klippel-Feil syndrome (KFS). [Subjects and Methods] The subject was a 39-year-old woman with neck pain and KFS that included incomplete block vertebrae in the C2–3 segments and block vertebrae in the C6–7 segments. The subject performed a rehabilitation program including cervical strengthening exercise (level 1) and CDS exercise (level 2) for 6 weeks. Neck pain intensity was measured using a visual analog scale (VAS) and the pressure pain threshold (PPT). All measurements were obtained before and after the CDS exercise program. [Results] The VAS and PPT measurements decreased; range of motion in the cervical joint increased. [Conclusion] CDS exercises were effective interventions for reducing neck pain in a patient with Klippel-Feil syndrome.

Key words: Cervical deep muscle strengthening, Neck pain, Klippel-Feil syndrome

INTRODUCTION

Klippel-Feil syndrome (KFS) was first identified in 1912 by French neurologists Maurice Klippel and André Feil. KFS is defined as congenital fusion of two or more cervical vertebrae and is believed to result from segmentation failure along the embryo’s developing axis during the 3rd to 8th gestational weeks. The incidence of this rare disease is 1 in 42,000, and nearly 65% of KFS cases occur in women. Common signs and symptoms of KFS include a short neck, limited range of motion (ROM) in the cervical spine, muscular weakness, atrophy, and neurological sensory loss. In addition, spinal scoliosis (60%) is the most common associated anomaly in patients with KFS, followed by spina bifida, high scapulae, urinary tract problems (15–20%), congenital heart defects, and cardiovascular anomalies (15–25%).

One particular consequence of KFS is a decrease in ROM when the vertebrae are fused, possibly restricting cervical motions of rotation, flexion, and extension; this is the most common clinical finding in KFS patients. Moreover, the upper cervical spine adjacent to the fused vertebrae may lead to hypermobility and instability, which is a predisposing factor that can induce neurological problems; the lower cervical spine close to the fused vertebrae may suffer disc degeneration. Clinically, instability resulting from spinal hypermobility is a condition under which the ability of the spine to maintain physiologic loads has been lost and deformity or pain may be triggered. The deep muscles around the spine maintain its stability, and dysfunction of such muscles may cause pain. Therefore, spinal instability in patients with KFS may not only trigger cervical pain but also aggravate deformity and as a result may bring about neurological problems.

As noted earlier, KFS causes hypermobility of the vertebrae that are not fused and the resulting neck pain necessitating intervention to preserve the stability of the vertebrae. Accordingly, the purpose of this study was to apply cervical deep muscle strengthening (CDS) exercise that can stabilize the cervical spine in a KFS patient and evaluate the subsequent decreases in neck pain.

SUBJECTS AND METHODS

A 39-year-old woman had been experiencing pain in the posterior midline of the neck for a few months but had not received specific treatment for the pain. She had no other systemic diseases and neurologic signs. On radiological examination, the cervical spine was observed to resemble a forward-straightened cervical column. With respect to specific vertebral segments, the examination revealed an arthritic change in C1–C2, complete block vertebrae in the C2–3 segments, right eccentric foraminal body hypertrophy and disc degeneration in the C5–6 segments, and incomplete block vertebrae in the C6–7 segments (Fig. 1).

The severity of pain in the neck, which was her main symptom, was measured using the visual analog scale (VAS). The VAS is a tool for measuring musculoskeletal pain with excellent reliability and validity. It is usually a horizontal line, 100 mm in length, that divides pain into grades of 0 to 100 ranging from 0 = "no pain" to 100 = "unbearable pain", and subjects are asked to indicate the pain level that they currently experience.
felt by using this scale. When the C2–C3 segments are incompletely blocked, excessive mechanical stress is applied to C1 and C2, thereby triggering excessive motion and instability of the atlantoaxial junction\textsuperscript{10}. Considering that pain may be induced by excessive stress on the capital extensor muscle as a result of excessive stress on the C1–C2 segments, the patient’s pressure pain threshold (PPT) was measured. The PPT is a valid and highly reliable tool in clinical studies of pain related to musculoskeletal pain problems\textsuperscript{11}. After vertically placing a pressure algometer on the belly of the patient’s capital extensor muscle, the tester gradually applied pressure to the belly at the pressure level of 0.2 lb and then held this pressure level for 5 seconds. At the moment when pain began, the tester paused and used the indicated pressure.

In addition, pain is a factor that may affect restriction of ROM\textsuperscript{8}; therefore, changes in cervical ROM were also measured. Cervical ROM was measured with a goniometer during flexion, extension, and bilateral rotation in a sitting position. For measurement of flexion and extension, the axis of the goniometer was placed with the external auditory meatus and proximal arm vertically aligned with the ground and the distal arm aligned with the base of the nares, and then measurement were taken. For measurement of rotation, the axis of the goniometer was placed with the cranial aspect of the head and the proximal arm aligned in parallel with the acromion and the distal arm aligned with the tip of the nose\textsuperscript{12}.

CDS exercise was initiated with light positive exercise, and resistance was gradually increased (by classifying the intensity into level 1 and level 2). The exercise methods were as follows. At level 1, strengthening exercise consisted of lifting the head from the supine or prone position and performing cervical flexion, extension, and lateral rotation ROM exercise in combination with cervical rotation in the supine position. To prevent any stress that might result from excessive exercise, the patient was told to carry out the cervical exercise as much as she wanted for each session. These movements were held for 5 seconds each. The patient performed 3 sets of 10 repetitions of this strengthening exercise daily for 2 weeks.

At level 2, the cervical deep muscle exercise consisted of tucking the chin down towards the chest in the supine position without lifting the head off the floor. Then, the patient lay face down with her hands supporting her forehead, took away her hands, and stayed in the prone position. A therapist placed her hand on the side of the subject’s head and provided gentle resistance, as if she was bending the subject’s head to one side. The patient also performed resistance exercise against cervical flexion, extension, lateral bending, and rotation in a sitting position using a Theraband. This exercise consisted of first lifting her neck up from the prone position and then lifting her neck up from the supine position. All movements were held for 5 seconds each, and the patient completed 3 sets of 10 repetitions of strengthening exercise daily for 4 weeks. To prevent exercise-related stress, resistance was gradually increased from the minimum amount of resistance imposed for the cervical exercise. The cervical stretching exercise was followed by strengthening exercise to complete the level 2 exercise.

The subject understood the purpose of this study and agreed to participate in the research; she signed an informed consent form approved by the Institutional Review Board before participating in the study.

\section*{RESULTS}

The VAS was 58 mm, and the PPT was 1.28 kg/cm\textsuperscript{2} for neck pain at baseline. The VAS was reduced to 31 mm, and the PPT increased 1.37 kg/cm\textsuperscript{2} after the exercise program. Prior to the exercise, cervical ROM was 29° during flexion, 39° during extension, 58° during left rotation, and 50° during right rotation. After the exercise, cervical ROM increased slightly increased to 32° during flexion, 46° during extension, 62° during left rotation, and 55° during right rotation. It was verified that CDS exercise had positive effects on neck pain and cervical ROM in a KFS patient (Table 1).

\begin{table}[h]
\centering
\begin{tabular}{|l|c|c|}
\hline
 & Before & After \\
\hline
VAS (mm) & 58 & 31 \\
PPT (kg/cm\textsuperscript{2}) & 1.28 & 1.37 \\
Flexion & 29 & 32 \\
Cervical Extension & 39 & 46 \\
ROM & Left rotation & 58 & 62 \\
 & Right rotation & 50 & 55 \\
\hline
\end{tabular}
\caption{Comparison of VAS, PPT, and cervical ROM between before and after the intervention}
\end{table}

\section*{DISCUSSION}

KFS is defined as congenital fusion of two or more cervical vertebrae, and myelopathy or radiculopathy and neck pain have been reported as its clinical symptoms\textsuperscript{6}. The fused vertebrae grow asymmetrically, restricting cervical ROM and bringing about deformity such as scoliosis\textsuperscript{3}. Restricted cervical ROM is the cause of cervical vertebrae...
Cervical vertebrae adjacent to the fused vertebrae exhibit hypermobility, leading to excessive mechanical stress. Hypermobility in the vertebrae can lead to induction of intervertebral disc damage, or degeneration may be induced during cervical active motion. The subject in this study exhibited an arthritic change in the C1–C2 segments and disc degeneration in the C5–C6 segments, with hypermobility resulting from the fusion of the C2–C3 and C6–C7 segments. Hypermobility may increase load on the cervical facet joint and trigger pain during active ROM. Instability due to segmental hypermobility reduces the ability to stabilize nerve muscles for movement control, triggering inappropriate control of vertebra motions and restricting pain and joint movements.

Cervical deep muscles play a key role in stabilizing and protecting the cervical spine, whose dysfunction may lead to pain in the head and neck. The cervical extensor muscle is a muscle that needs exercise for muscle dysfunction management in patients with neck pain and cervical deep muscle strength and endurance training may be useful in the management of symptoms related to mechanical impairment of the neck. Deep muscles reduce increased stress on bone-ligament assistive structures and enhance spinal stability, decreasing pain. Deep muscles have a lot of nociceptive receptors around the joints. Abnormally applied excessive stress on the muscles may be a cause of pain; pain may also be triggered by deep muscle dysfunction.

In the present study, the VAS and PPT decreased after cervical deep muscle strengthening exercise. Cervical deep muscle strengthening improved the stability of the hypermobile cervical spine and helped to decrease mechanical stress applied around the spine, thereby alleviating pain.

In addition, KFS may restrict cervical rotation, flexion, and extension ROM. Cervical ROM in normal adults is 40±12° during flexion, 50±14° during extension, 72±7° during right rotation, and 73±6° during left rotation. However, in this subject, Cervical ROM was restricted. After CDS exercise, the subject's ROM increased slightly, although it was restricted compared with normal ROM. Moreover, pain acts as a factor that restricts ROM, and therefore, cervical ROM is considered to have increased as cervical stability improved; pain accordingly decreased as well.

In conclusion, CDS exercise had positive effects in a patient with KFS and neck pain who had no neurological symptoms. Most KFS patients select surgery as a therapeutic intervention against hypermobility of the spine that may worsen neurological problems. Nonetheless, hypermobility of the cervical spine in KFS patients may not be related to risks that increase their symptoms or neurological signs. Thus, CDS exercise is regarded as an appropriate intervention method for spinal stability in patients with KFS who present with neck pain and no neurological signs. This study had some limitations, as it was conducted with only one subject. Further studies are therefore needed to confirm the effects of the aforementioned exercises on neck pain in patients with KFS.

REFERENCES

1) Klippel M, Feil A: Un cas d’absence des vertebres cervicales. Avec cage thoracique remontant jusqu’a la base du crane (cage thoracique cervicale). Nouv Iconog Salpetriere, 1912; 25: 223–250.
2) Thomsen MN, Schneider U, Weber M, et al.: Scoliosis and congenital anomalies associated with Klippel-Feil syndrome types I–III. Spine, 1997; 22: 396–401. [Medline] [CrossRef]
3) Tracy MR, Dormans JP, Kasumi K. Klippel-Feil syndrome: clinical features and current understanding of etiology. Clin Orthop Relat Res, 2004; (424): 183–190. [Medline] [CrossRef]
4) Farid IS, Omar OA, Inslker SR: Multiple anesthetic challenges in a patient with Klippel-Feil Syndrome undergoing cardiac surgery. J Cardiothorac Vasc Anesth, 2003; 17: 502–505. [Medline] [CrossRef]
5) Henninger RN, Jones ET: Developmental orthopaedics. II: The spine, trauma and infection. Dev Med Child Neurol, 1982; 24: 202-218. [Medline] [CrossRef]
6) Kisner C, Colby LA: Therapeutic Exercise, 5th ed. Foundation and Techniques. Philadelphia: F.A. Davis Company, 2007, pp 450–479.
7) White AA, Panjabi MM: Clinical Biomechanics of the Spine, 2nd ed. 1990, Philadelphia: JB Lippincott.
8) Norkin CC, White DJ: Measurement of Joint Motion: A Guide to Goniometry, 4th ed. Philadelphia: F.A. Davis Company. 2009.
9) Pizzutillo PD, Woods M, Nicholson L, et al.: Risk factors in Klippel-Feil syndrome. Spine, 1994, 19: 2100–2116. [Medline] [CrossRef]
10) Shen FH, Samartzi D, Herman J, et al.: Radiographic assessment of segmental motion at the atlantoaxial junction in the Klippel-Feil patient. Spine, 2006; 31: 165–169. [Medline] [CrossRef]
11) Ohrbuch R, Gale EN: Pressure pain thresholds, clinical assessment, and differential diagnosis: reliability and validity in patients with myogenic pain. Pain, 1989; 39: 157–169. [Medline] [CrossRef]
12) Norkin CC, White DJ: Measurement of Joint Motion: A Guide to Goniometry, 4th ed. Philadelphia: F.A. Davis Company. 2009.
13) Aprill C, Bogduk N: The prevalence of cervical zygapophysial joint pain. A first approximation. Spine, 1992; 17: 744–747. [Medline] [CrossRef]
14) Kirkaldy-Willis WH, Burton CV: Managing Low Back Pain, 3rd ed. New York: Churchill Livingstone, 1992.
15) Chung SH, Her HG, Ko TS, et al.: Effects of exercise on deep cervical flexors in patients with chronic neck pain. J Phys Ther Sci, 2012; 24: 629–632. [CrossRef]
16) Schomacher J, Falla D: Function and structure of the deep cervical extensor muscles in patients with neck pain. Man Ther, 2013; 18: 360–366. [Medline] [CrossRef]
17) Iqbal ZA, Rajan R, Khan SA, et al.: Effect of deep cervical flexor muscles training using pressure biofeedback on pain and disability of school teachers with neck pain. J Phys Ther Sci, 2013; 25: 657–661. [Medline] [CrossRef]