

Large C4/5 Spondylotic Disc Bulge resulting in Spinal Stenosis and Myelomalacia in a Klippel-Feil Patient: Knowing When Not to Treat Conservatively

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Abstract:

Objective: The purpose of this report is to describe the rationale behind not conservatively treating a patient presenting with cervical radiculopathy.

Clinical Features: A 39 year-old male was referred to a private chiropractic practice for a second opinion, more specifically from a certified Cox® practitioner. The patient presented with pain and limited motion in his neck with pain and numbness radiating down both arms and left leg. Diagnostic imaging revealed a patient with Klippel-Feil syndrome and a large spondylotic disc bulge at C4/5 compressing the cord and causing myelomalacia.

Outcome: After examining this patient and reviewing the diagnostic imaging, it was decided that cervical flexion-distraction manipulation would not be performed and that the patient should proceed with the surgical intervention recommended by his neurosurgeon and considered by his chiropractor.

Conclusion: Although this patient was referred by another chiropractor to evaluate the possibility of treating his condition with cervical Cox® flexion-distraction decompression manipulation, it was decided that surgery would be the most advantageous for this patient.

Introduction:

Klippel-Feil syndrome may include a plethora of congenital musculoskeletal anomalies and is generally defined by two or more non-segmented levels of the cervical and upper thoracic spine. The fused vertebrae are known to cause hypermobility and increased tensile stress on the adjacent discs predisposing them to disc degeneration, bulges and herniations. This syndrome may include a variety of other clinical findings such as the appearance of a short neck or low hairline, hearing loss, and facial nerve palsy and asymmetry. Additional congenital anomalies may be present such as Sprengel's deformity, scoliosis, and genito-urinary tract malformations. Chiropractic management of these patients would be a consideration for treating the affected adjacent disc and not the anomalous segment.

Case report:

HISTORY & BACKGROUND

This 39-year-old man was referred by his chiropractor for an opinion from a certified Cox® practitioner regarding whether or not cervical flexion-distraction manipulation would be a reasonable alternative to the decompression with fusion surgery recommended by the patient's neurosurgeon.

Subjective complaints included several months duration of pain and stiffness in his neck with numbness in all of his fingers bilaterally. The numbness was more pronounced distally and on the right with the middle three fingers primarily affected. He also reported a more recent onset of achy pain radiating down his left leg from the inner thigh to his foot with numbness in his left foot. He had consulted his general physician who performed lab work to rule out diabetes or other causes of peripheral neuropathy. No etiology for his complaints was determined with laboratory studies and the recommendation was for this patient to increase his exercise and lose weight. His lower extremity symptoms continued to worsen even though he had undergone physical therapy for his lumbar spine.

This patient reported that his left foot occasionally 'jumps' or moves on its own. His wife relayed that his foot frequently shakes at night. He reported no weakness in his legs however did report a loss of fine motor control in the fingers which affects his activities of daily living. He stated he makes frequent errors while typing since he has difficulty feeling the buttons on his computer keyboard. Upon further questioning, he relayed that when attempting to complete the paperwork before this examination, the pen felt like it was slipping out of his hands, possibly because he did not have the strength to hold it firmly. He is noticing progressive loss of grip strength and increase in numbness in his arms and left foot.

IMAGING





Sequential sagittal MRI images demonstrating Large C4/5 disc-osteophyte complex, spinal stenosis, and myelomalacia

Imaging brought in by the patient included an MRI of his lumbar spine, bilateral hips and sacroiliac joints. These were reported as normal with the exception of minor degenerative changes at the L4/5 level with no disc herniations or stenosis noted. These imaging procedures were ordered by his chiropractor due to the symptoms radiating down his left leg.

The cervical spine MRI brought in by the patient included T1 weighted, T2 weighted and stir sagittal images, as well as T2 weighted coronal and gradient echo axial images through the C2/3-C7/T1 levels. C2/3 was reported as unremarkable. C3/4 had a rudimentary disc and congenital fusion with the spinous processes being separate.

The C4/5 level had a disc decreased in its T2 signal intensity but normal in stature. There was a large circumferential disc bulge with adjacent endplate spur formation anteriorly and posterolaterally. No focal disc herniation was reported; however the disc osteophyte complex produced moderate-to-marked central canal stenosis with compression and displacement of the adjacent spinal cord. Bilateral uncovertebral joint degeneration produced moderate-to-marked bilateral foraminal stenosis with probable impingement on the exiting C5 nerve roots. There was a focal region of abnormal increased signal within the spinal cord parenchyma at this level. The spinous processes at that level had an abnormal appearance with no distinct cortex seen, as well as a generalized increased signal in that area.

A congenital non-segmentation anomaly with a rudimentary disc and fused spinous processes was noted at C5/6. The disc was decreased in its T2 signal intensity with normal stature and a circumferential disc bulge. Uncovertebral arthrosis was present bilaterally with narrowing of the intervertebral foramina; however no mechanical compression of the spinal cord or nerve roots was demonstrated. The C7/T1 segment was unremarkable. The sagittal curve was straightened with a levoscoliosis in the mid-to-lower cervical region and a dextroscoliosis of the mid-to-upper cervical region.

The radiologist's impressions concluded that this large C4/5 spondylotic disc bulge produced moderate-to-severe central canal and intervertebral foraminal stenosis which compressed and displaced the cord. The region of abnormal increased signal intensity on the T2 weighted images in the spinal cord parenchyma was consistent with myelomalacia. Klippel-Feil syndrome with an abbreviated segmentation defect at C3/4 and a congenital block vertebra at C5/6 was noted with mild degenerative disc disease and bulging at C6/7.

This patient was evaluated by a neurosurgeon the day before this consultation. The surgeon stressed the importance of undergoing surgery within the next two weeks stating that with his current condition even a mild car accident could be catastrophic. The patient and the patient's referring chiropractor were seeking a second opinion to see if there was any possibility that surgery could be prevented, specifically with the flexion-distraction manipulation procedure.

The primary goal of the examination was to determine the level of upper motor neuron pathology in this patient. The examination in regards to orthopedic testing and ranges of motion of the cervical spine were limited so as not to aggravate this patient's condition.

Visual inspection revealed significant facial asymmetry, appearance of a short neck and a severely deformed and hypoplastic right external ear. He was previously informed that his ear and facial deformities were due to the fact that his mother fell down a flight of stairs when he was in utero which caused his fist to impact and damage his facial nerve and external ear. Upon

further questioning, the patient reported a history of some urinary tract issues, although no details were recalled.

The neurological examination revealed general hypoesthesia to pinwheel examination at the C6, C7 and C8 dermatomes bilaterally, most marked at C6 on the right. Positive Tromner's finger flick test was positive bilaterally indicative of an upper motor neuron lesion. His upper extremity strength revealed weakness of the biceps muscles bilaterally, graded a 4, and weakness of opposition of the thumb and 5th finger on the right.

Examination of the lumbar spine was essentially normal and included negative nerve root stretch tests, negative Valsalva for low back or leg pain, negative orthopedic testing and normal strength. However, his patellar reflexes were brisk bilaterally. His right ankle reflex elicited two beats of clonus and his left ankle reflex elicited five-to-six beats of clonus. After performing this reflex examination, he reported that when he hooks his feet under the couch to perform abdominal exercises, his left foot repeatedly twitches or jumps, similar to the clonus just elicited.

After the examination and review of the MRI films and reports, it was determined that surgical intervention as recommended by the neurosurgeon would be the best course of action for this patient.

Conclusion:

Although this patient was referred for the possibility of receiving Cox flexion-distraction decompression manipulation to treat a large C4/5 disc bulge resulting in radiculopathy, it was determined that surgical intervention was the best method of care. The rationale included the severity of his condition, the progressive neurological deficits in the upper and lower extremities including pathological reflexes, and the presence of Myelomalacia. This patient was surprised to hear that he had Klippel-Feil syndrome, having never been informed prior to this consultation, especially since the diagnosis helped explain a variety of physical issues throughout his life. After six months time, the patient was called in hopes of determining his post-surgical status. He informed us that he had recently returned to work; however, no other details were given at that time.

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