Anomalous rib presenting as cervical myelopathy: a previously unreported variant of Klippel-Feil syndrome

Case report

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A patient is reported with an anomalous rib that caused compression of the cervical spinal cord and presented with cervical myelopathy. This appears to be the first reported instance of this particular anomaly. The clinicoanatomical aspects of this case are discussed.

KEY WORDS • Klippel-Feil syndrome • rib anomaly • cervical spine • myelopathy

As originally described in 1912, the Klippel-Feil syndrome consisted of a clinical triad of limitation of neck movement, a short neck, and a low posterior hairline. Current parlance allows the designation to indicate the congenital fusion of two or more cervical vertebrae. The classic clinical triad is noted in less than 50% of cases. Both skeletal and visceral congenital anomalies frequently accompany this syndrome and can be considerable threats to these patients. In this report, we present a previously undocumented skeletal anomaly of the cervical spine associated with cervical myelopathy.

Case Report

This 27-year-old black man presented with weakness and loss of muscle mass on the left side of the body. He first noted the left-sided asymmetry approximately 5 years prior to admission, when the left arm and leg seemed smaller than the right. Gradually his condition progressed to the point that he could no longer participate in sports. When he attempted to run, the left side became stiff and he lost his balance. When he turned his head to the extreme left a tingling sensation would spread down his left arm. This symptom was relieved when his neck returned to a neutral position. He had no radicular pain and denied sensory loss, seizures, or other cerebral, cerebellar, or brain-stem symptoms. There was no history of trauma at birth, head injury, meningitis, or familial neurological disease. The patient denied the use of alcohol, tobacco, or street drugs.

Examination. Physical examination revealed a well-nourished black man with normal physiological appearance. There was obvious asymmetry in muscle bulk, with the left arm and leg much smaller than the right. The neck was supple, there were no midline abnormalities, and the spine was straight. Mental status and cranial nerve examination were unremarkable. Muscle tone was spastic in the left extremities and normal in the right. Although bulk was decreased in all muscle groups in the left upper and lower extremities, power was normal in all muscles except the left triceps (4/5), extensor carpi radialis (4/5), extensor carpi ulnaris (4+/5), opponens pollicis (4+/5), flexor digitorum (4+/5), ilopsoas (4+/5), and quadriceps (4+/5). Sensory examination was normal for light touch, pinprick, vibration, and position sense. Graphesthesis, stereognosia, and double simultaneous stimulation responses were normal. Reflexes were 3+ on the right and 4+ on the left, with Hoffmann’s sign present on the left. There was sustained clonus on the left and unsustained clonus on the right. Cross adductor reflexes were positive bilaterally. He was clumsy on the left side with rapid alternating movements, but finger-to-nose and heel-to-shin testing were normal. Gait was spastic and Romberg’s sign was negative. As the patient increased his speed of gait he tended to drag the left foot.

Plain x-ray films and tomograms of the cervical spine demonstrated a segmentation defect at the base of the dens and fusion of the vertebral bodies from C-5 through C-7. Both lesions were stable in flexion and...
extension studies. Myelography revealed a severe stenosis at the mid-cervical level with spinal cord compression predominantly from the dorsal aspect of the canal (Fig. 1). Note was made of a right-sided ventrolateral soft-tissue mass thought to be consistent with intervertebral disc herniation or an osteophyte. Computerized tomography demonstrated a dorsal element passing directly through a defect in the C-5 lamina and causing compression of the left side of the spinal cord. Hemiatrophy of the left side of the spinal cord was also noted below the C-5 level. Magnetic resonance (MR) imaging demonstrated the dorsal compressive element.

Serum B12, folate, vitamin E, erythrocyte sedimentation rate, anti-nuclear antibody level, rheumatoid factor, serum protein electrophoresis, venereal disease, and thyroid function tests were unremarkable. Electrodiagnostic analysis was also within normal limits.

Based on the clinical and radiological examinations, it was concluded that this patient had a myelopathy caused by the cervical lesion. Because the cord was compressed from the left posterior aspect of the canal, it was decided that decompression would best be effected through a posterior approach.

Operation. At surgery, bilateral exposure of C4–7 was obtained. An anomalous rib was noted within 3 cm of the skin, coursing down toward the lamina of C-5 (Fig. 2 upper). The rib continued parallel to the spinous process of C-5 and traveled through a defect in the lamina of C-5 (Fig. 3). After removal of the C-5 lamina, compression of the spinal dura was apparent (Fig. 2 lower). The rib did not arise from a bone process, and was detached from its origin in the soft tissues at the base of the neck posterior to the T-1 lamina. Pathological analysis disclosed that the specimen was consistent with rib. The postoperative course was uneventful.

Postoperative Course. Three months after surgery, MR imaging showed that the dorsal cord compression had been relieved; however, a bright intensity signal remained within the spinal cord (Fig. 4). The patient had noted a subjective decrease in spasticity but weakness and hyperreflexia had not changed.

Discussion

Although the classic triad of Klippel-Feil syndrome consists of a short neck, a low posterior hairline, and limitation of neck movement, fewer than 50% of patients have these symptoms. As currently applied, the designation "Klippel-Feil anomaly" refers to the congenital fusion of at least two cervical vertebrae.7 The reported incidence of congenital fusion of the cervical vertebrae is as high as 0.5% on spinal roentgenograms.5 The deformity may result from an insult to the developing fetus which is believed to take place at approximately the 25th day of gestation.1 Although unproven, another theory (proposed by Gardner and Collis) postulates that overdistention of the neural tube may be involved in the pathogenesis of the Klippel-Feil syndrome as well as syringomyelia, diastematomyelia, myelomeningocele, and the Arnold-Chiari syndrome.5 Many malformations have been described in association with the Klippel-Feil syndrome; among the more...
common are scoliosis, Sprengel's deformity, hemivertebrae, and spina bifida. Less common but significant disorders include those of the urogenital and cardiovascular systems. Cervical rib abnormalities are present in 10% to 15% of cases. Some patients with Klippel-Feil syndrome will be at risk for development of neurological deficits based on their respective cervical spinal abnormalities. Nagib, et al., reviewed a series of 21 patients with Klippel-Feil syndrome in an effort to predict which patients might be at risk for neurological deterioration. They concluded that three risk groups can be recognized: unstable fusion patterns; craniocervical abnormalities; and cervical spinal stenosis. In their series, 43% of patients required decompression and stabilization for neurological deficits.

To the best of our knowledge, the congenital abnormality described above represents a previously unreported anomaly of the cervical spine associated with the Klippel-Feil syndrome and therefore denotes a unique cause of cervical myelopathy. The radiographic abnormalities consisted of congenital fusion of the vertebral bodies from C-5 to C-7, a segmentation defect at the base of the dens, and an anomalous rib originating in the paraspinal muscles dorsally and not articulating with either C-7 or T-1. The rib variant appeared to have the attributes of a normal rib with a head, neck, and shaft. Based on this arrangement it would appear that the proximal part of the rib was located in the paraspinal muscles. The distal aspect of the rib passed through a cleft in the lamina of C-5 and extended just beneath the lamina of C-4 where it caused severe spinal cord compression and resulting atrophy of the left side of the spinal cord.

Care had to be taken throughout the initial surgical exposure because of the excessive mobility of the rib which was noted to slide easily in and out of the laminar defect. We believe that, even though the rib did not have a bony origin, it was seated deep enough in the musculature that movements of the head and neck would cause recurrent compression of the spinal cord by the distal end of the rib. After excision of the rib, an obvious indentation remained in the dura. We did not perform posterior fusion in view of the fact that the limited laminectomy from C-4 to C-6 did not compromise the facet joints; however, the potential for delayed instability may exist and this was thoroughly discussed with the patient. At follow-up evaluation 4 months after surgery there was no evidence of spinal instability.

References

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