Anomaly of the axis causing cervical myelopathy

Case report

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Although the craniovertebral junction is one of the most common sites at which anomalies develop, spina bifida occulta of the axis (C-2) associated with cervical myelopathy is extremely rare. The authors present the case of a 46-year-old man who developed progressive tetraparesis caused by a cervical canal stenosis at the level of the axis. The spinal cord was compressed by an invaginated bifid lamina of the axis. The patient made a remarkable recovery after undergoing decompressive laminectomy of C-3 and removal of the bifid posterior arch of the axis.

KEY WORDS • anomaly • axis • craniovertebral junction • myelopathy • spina bifida

Developmental stenosis of the spinal canal is a recognized cause of cervical myelopathy; however, localized stenosis of the spinal canal due to an anomaly of the axis (C-2) is rare. We report the case of a patient who developed an anomaly of the axis with its lamina invaginating the cervical spinal canal and discuss the embryological development and the causes of this condition.

Case Report

History. This 46-year-old man was admitted to our hospital with a 6-month history of numbness of both hands and stiffness of all four extremities. During the 2 months prior to admission, he had begun to notice a slowly progressive gait disturbance. He had no history of neck or head injury.

Examination. Physical examination on admission revealed no cutaneous hallmark around the neck. There was no fasciculation or evidence of muscle atrophy. Diminution of all sensory modalities, including touch and pain sensations and vibratory appreciation, was noted. Deep tendon reflexes were hyperactive in the upper and lower extremities. Hoffmann’s sign was present bilaterally. The patient suffered spasms and needed the assistance of a cane to walk.

Plain x-ray films of the cervical spine revealed the absence of the spinous process of the axis and an enlarged spinous process of C-3 (Fig. 1 upper left). Magnetic resonance imaging revealed a marked spinal cord compression at the C2–3 level (Fig. 1 upper right). Computerized tomography (CT) myelography demonstrated a bifid lamina of the axis that invaginated the spinal canal and compression of the spinal cord (Fig. 1 lower left). Three-dimensional CT better delineated this anomaly; the bifid lamina of the axis was surrounded by the large spinous process of C-3 (Fig. 1 lower right).

Operation. The patient underwent a C-3 laminectomy and removal of the bifid lamina of the axis. During surgery, we observed the short rotator muscles such as rectus capitis posterior major muscle inserting into the enlarged spinous process of C-3. The bifid lamina and atrophic ligamentum flavum were observed as well. Anteroposteriorly the dural sac was markedly pinched by the bifid lamina of the axis.

Postoperative Course. Two weeks postoperatively, the symptoms had disappeared, and the patient regained normal neurological status. He returned to his occupation as a carpenter within 2 months after surgery.

Discussion

Developmental anomalies in the region of the axis have been reported as follows: irregular atlantoaxial segmentation; dens dysplasias including ossiculum terminale persistence, os odontoideum, and hypoplasia/aplasia; and segmentation failure of C-2 and C-3. With the exception of fusion anomalies, most congenital anomalies of the axis are confined to the odontoid process. Posterior arch defects of the axis are thought to be rare, and their clinical significance is not known.

The designation “spina bifida occulta” is a radiological one referring to a defect of the spinous process and/or asso-
associated posterior neural arch. Spina bifida occulta results when the halves of the vertebral arch fail to fuse. The development of the vertebra takes place in three essential stages: precartilage, chondrification, and ossifications. In the precartilage stage, the cells of the sclerotome migrate in three directions: ventromedially to surround the notochord, dorsally to cover and to form the neural arch, and ventrolaterally to form the costal process. The chondrification of the posterior arch begins at the pedicles during the 6th week of embryogenesis and ends in the midline during the 4th month. This cartilaginous arch ossifies by 3 to 4 years of age. Therefore, the posterior elements defect of the axis may either be caused by failure of the extension process of the chondrification centers in the posterior arch or by failure of the ossification process.

In our search of the literature we found only five cases in which there was myelopathy due to canal stenosis at the level of the axis (Table 1). Benitah, et al., reported on a 78-year-old man in whom cervical myelopathy due to cord compression was demonstrated to be caused by large osteophytes at the level of the axis. This case was thought to be acquired, but in addition to our case, the remaining four cases are considered congenital anomalies. Benitah, et al., described another case of a 41-year-old man in whom cervical myelopathy with a hypertrophied posterior arch of the axis had developed. Prakash, et al., have reported a similar case in which a hypertrophied posterior arch of the axis was demonstrated. The cause of the hypertrophy might have been a premature fusion of the cartilaginous synchondrosis or abnormal ossification in the posterior arch. In contrast, Koyama, et al., and Kawano, et al., have described cases similar to ours in which myelopathy was caused by rare anomalies of the axis. These authors reported that cervical myelopathy in their patients was caused by anomalous bony structures that invaginated the spinal canal. The cause of this invagination might be due to failure of the chondrification process because failure of the two chondrification centers on one side to fuse into a single ossification center.
which is associated with failure of posterior fusion, may result in dysplastic bony structures in the spinal canal. In our patient the spinous process of C-3 was also enlarged. This enlargement is probably caused by compensation for the defect of the spinous process of the axis, into which the short rotator muscles insert themselves. For this compensation to occur, the cause of the anomaly in this case would have occurred in the earlier stage of embryogenesis such as the chondrification stage.

Our patient and four of those reported in the literature2–4,7 harbored congenital anomalies that did not manifest clinically until the fourth and fifth decades of life. In contrast, Morizono, et al.,6 have reported the case of a 20-year-old man with spina bifida of the axis associated with enlarged posterior elements of C-3. Barber-Riley1 has also described a case with a hypoplastic lamina of the axis and an enlarged spinous process of C-3 in an 18-year-old woman. Although the radiological findings in these patients was very similar to ours, the patients were young, asymptomatic, and their anomalies were found incidentally. Therefore, the chronic myelopathic symptoms in the other reported cases likely resulted from complications that arose during the aging process (such as spondylolytic change, ligament hypertrophy, venous stasis with cord edema, or injury due to hyperextension) that caused progressive narrowing of the cervical spinal canal and spinal cord compression in a congenitally small canal.

All of the six patients underwent a surgical decompressive procedure and made favorable outcomes (Table 1). Therefore, decompressive surgery including removal of the anomalous bony structures or laminectomy resulted in improved symptoms and clinical findings in all six cases. We think this is an effective treatment for symptomatic patients with this condition.

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123