Chiari I malformation and spinal cord injury: Cause for concern in contact athletes?

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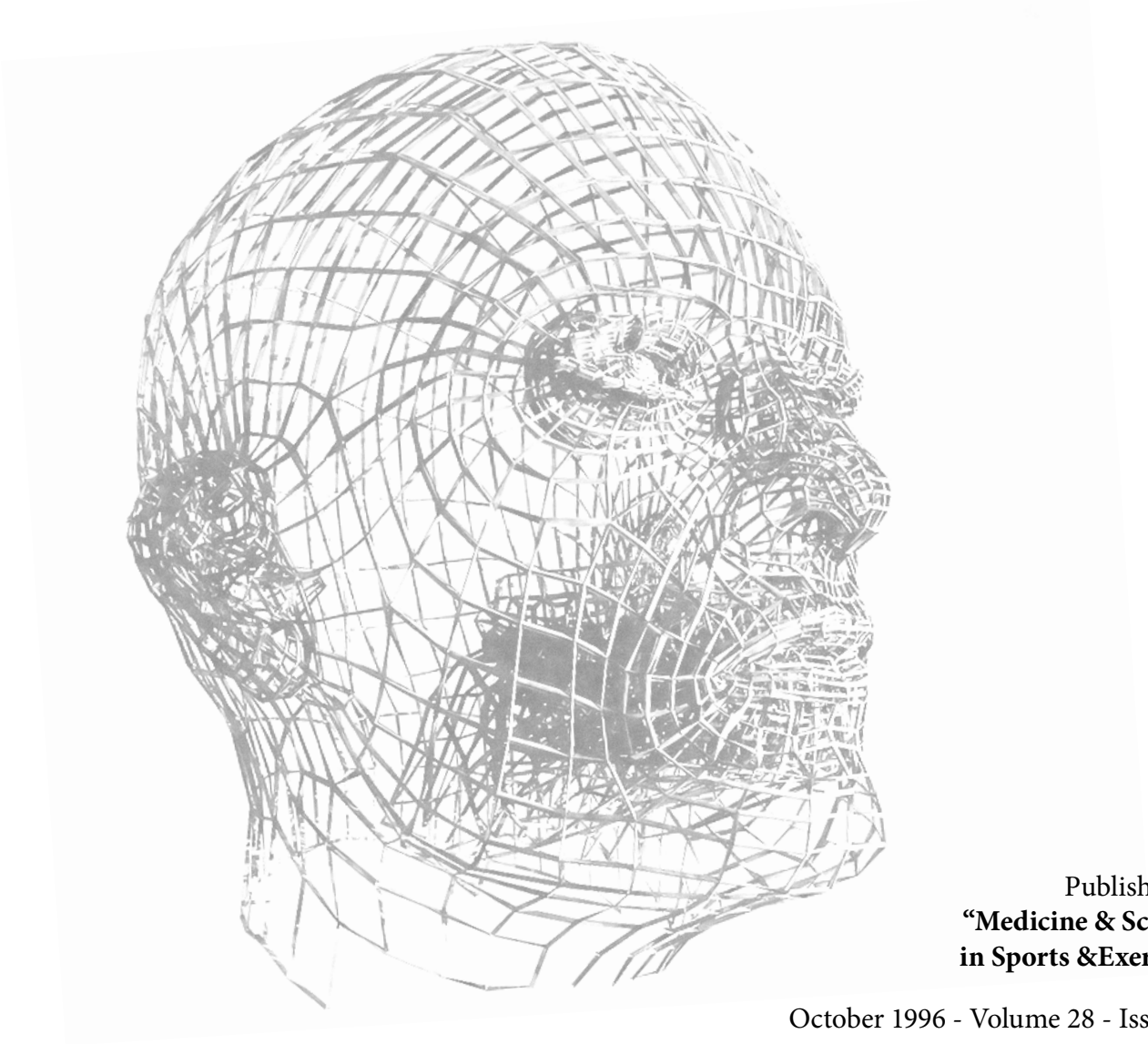

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Chiari I malformation and spinal cord injury: cause for concern in contact athletes?

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We present a case of transient quadriparesis in an 8-yr-old football player with a normal cervical spinal canal but Chiari I malformation of the hindbrain. Chiari I consists of herniation of the cerebellar tonsils through the foramen magnum, which reduces space available for the spinal cord. Magnetic resonance imaging (MRI) of the brain or cervical spine can identify asymptomatic Chiari I, whereas previously myelography was necessary. The incidence and natural history of Chiari I malformation is reviewed, with special reference to the question of return-to-play in young athletes.

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CASE REPORT

A previously healthy 8-yr-old male football player suffered an axial blow to the head and flexed neck during a tackle. He did not lose consciousness but experienced tingling in both anterior thighs. His arms felt normal. He felt short of breath for less than a minute. When examined on the field, he had normal motor and sensory findings in all extremities. He did not have unusual neck pain or restriction of motion. He was able to stand up and walk off the field with assistance.

Subsequent neurologic examination was normal. The paresthesia in the thighs resolved after 24 h. Plain radiographs of the spine demonstrated mild dysplasia of the C1 ring. Flexion-extension lateral radiographs demonstrated normal alignment. A magnetic resonance imaging (MRI) study demonstrated an Arnold-Chiari type I malformation, with extension of the cerebellar tonsils 16 mm inferior to the foramen magnum to the level of the base of the odontoid process (Fig. 1). The subarachnoid space was significantly diminished at the foramen magnum and C1 levels. The anterior aspect of the inferior medulla was indented by the dens. The remainder of the MRI was normal.

The patient was advised not to return to contact sports. After consultation with several orthopedic and neurosurgical spine specialists, the patient and his family decided to proceed with decompression of the foramen magnum. The patient has recovered from surgery uneventfully, with full resuspension of the cerebellar tonsils.

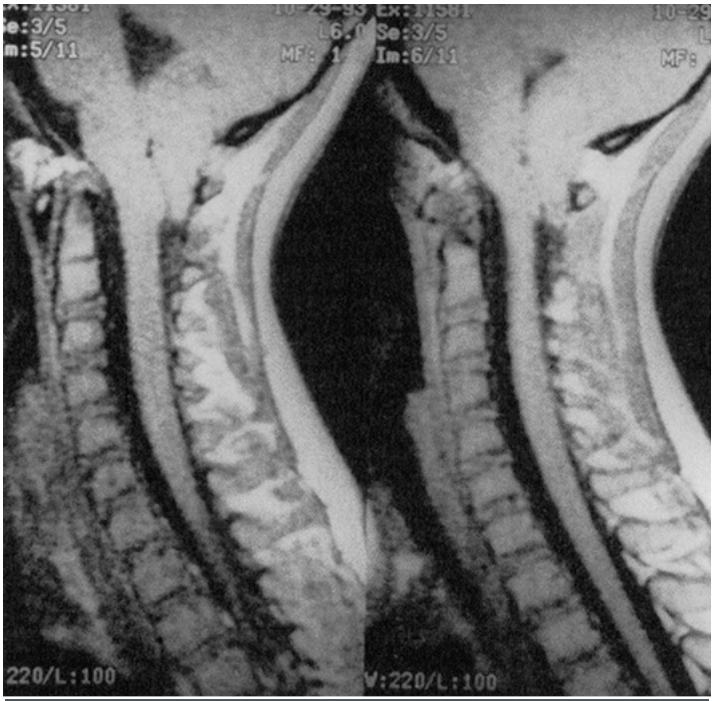

Figure 1 – A sagittal T1-weighted image of the cervico-occipital junction shows crowding of the spinal cord by the cerebellar tonsils. The medulla and upper spinal cord occupy the anterior half of the foramen magnum. The anterior aspect of the medulla appears to be indented by the odontoid process of C2. The cerebellar tonsils occupy the posterior half of the foramen magnum and extend 1.6cm below the foramen to level of the ring of C1. The subarachnoid space (black) is diminished by crowding of the medulla and cerebellar tonsils between the foramen and C1.

DISCUSSION

Significant spinal stenosis is usually considered a medical contraindication to participation in collision sports[2]. After an episode of transient paralysis, an athlete's neck should be stabilized for transport to the hospital. The athlete should undergo x-ray and MRI evaluation of the spine to rule out fracture, instability, or intervertebral disc herniation. In the absence of these lesions, some athletes may have a "functional" stenosis of the cervical canal, which predisposes them to recurrent spinal cord injury.

The radiologic definition of functional spinal stenosis is problematic. The width ratio of spinal canal to vertebral body was proposed as a screening tool for subaxial cervical spinal stenosis by Torg et al.[12, 15]. Subsequent studies showed the ratio to be abnormal in 25-49% of asymptomatic college and professional football players[8, 10]. The ratio may retain prognostic value when used as part of the definition of "spear tackler's spine," which also requires straightening of cervical lordosis, post-traumatic changes in the cervical spine, and previous speartackling behavior[16].

Because of normal variation in vertebral body, spinal canal, and spinal cord size among individuals, Cantu[2] has proposed that obliteration of the subarachnoid space on MRI should be primary radiologic evidence of functional spinal stenosis. This criterion was met in 6 of 11 cases of sports-related quadriplegia that were evaluated by MRI and registered at the National Center for Catastrophic Sports Injury Research between 1989 and 1991, but it has not yet been tested in asymptomatic athletes. Because this criterion does not depend on measurements of the vertebral body, it may be useful in the upper as well as in the lower cervical spine.

The Chiari I malformation is a congenital extension of the cerebellar tonsils into the upper cervical spinal canal, which may constitute functional spinal stenosis by the Cantu definition. Chiari[3] described autopsy findings of four malformations of the cerebellum in 1891, the least severe of which was termed type I. Prior to the advent of MRI, the lesion was diagnosed by clinical exam and cervical myelography in patients who became symptomatic as adults. The use of MRI to assess head and neck problems in athletes, however, will identify these malformations in younger, relative-

ly asymptomatic individuals who wish to participate in contact sports.

A previously published review of 12,226 consecutive head and cervical spine MRIs at a referral medical center identified 68 cases of Chiari I, establishing a reasonable upper estimate of its prevalence at 0.56%[5]. Forty-two patients (62%) were female. Forty-seven patients (69%) had symptoms referable to the malformation. The mean age of symptom onset was 30.6 yr (range 3-65 yr) and was unrelated to the type of symptoms. Twenty-five patients (37%) had central cord signs, with numbness, weakness, or pain involving the upper extremities, most commonly at the C4-C6 levels. Fourteen (21%) had brainstem dysfunction such as horizontal or vertical nystagmus, ataxia, dysarthria, dysphagia, lower cranial nerve palsies, or sleep apnea. Eight patients (12%) had headache or neck pain, without localizing neurologic findings. The symptoms and age of onset in this study are consistent with other reports.

Radiographic findings in this group of 68 Chiari I patients included skeletal or cerebral anomalies in 16 cases (24%), usually atlanto-occipital assimilation, platybasia, or basilar invagination. The MRI identified syringomyelia in 27 cases (40%), most of which were in the central cord symptom group. The mean tonsillar descent was 7.3 mm (range 5-12 mm) in the asymptomatic group, 8.6 mm (range 5-20 mm) in the headache symptom group, 10.1 mm (range 3-20 mm) in the central cord symptom group, and 12.3 mm (range 4-24 mm) in the brainstem symptom group (ANOVA $F = 3.32$, $P = 0.026$).

Chiari I with syringomyelia is well described as a predisposition to spinal cord injury. For example, Frogameni et al.[7] reported a 20-yr-old male football player who experienced 10 min of left-sided hemiparesis after weightlifting. MRI demonstrated Chiari I with a C3-C7 syrinx. No treatment was undertaken, but the patient was advised to forgo sports. Olivero and Dinh[11] describes a 28-yr-old woman who hit her head in a motor vehicle accident and developed bilateral upper extremity weakness over the subsequent 24 h. MRI revealed Chiari I and a large syringomyelia extending from C2 to C7. Her arm weakness resolved over 2 months with supportive care, and a follow-up MRI at that time showed almost complete resolution of the syrinx. Scully et al.[13] reported a case of dysarthria, hemihypesthesia, and nystagmus occurring a week after chiropractic neck manipulation in a 17-yr-old female with MRI evidence of Chiari I and a syrinx from C2 to the conus medullaris. The patient underwent a cervical laminectomy with shunting of the syrinx to the subarachnoid space. She had resolution of most symptoms except nystagmus.

At least six cases of isolated Chiari I (without syrinx) with cord injury have been reported. Mampalam et al.[9] describes a 13-yr-old female pedestrian who was hit by a car and suffered transient upper extremity weakness, lower cranial nerve palsies, and cerebellar dysfunction. MRI showed only a Chiari I malformation. Her symptoms incompletely resolved after a month of supportive care and rehabilitation. Vlcek and Ito[17] describes a 2-yr-old boy who fell onto the apex of his head while in a position of neck flexion and suffered lower extremity paraparesis and urinary retention. Again, the MRI demonstrated only Chiari I. The symptoms resolved after cervical laminectomy and suboccipital decompression. Tomaszek et al.[14] describes a 3-yr-old boy who fell downstairs and had progressive lethargy, confusion, and fatal apnea over 24 h. Autopsy showed mild brain swelling and Chiari I.

Erlich et al.[6] describes a 2-yr-old girl who fell onto the back of her head and suffered bilateral upper extremity paralysis, eponymically termed Bell's cruciate palsy. CT showed fracture of the left lateral mass of C1. MRI showed Chiari I and edema or contusion of the medulla. Symptoms incompletely resolved over 5 months of rehabilitation. Bondurant and Oro[1] describes a 2-yr-old boy who fell from the porch onto his chin and chest. Over 15 min, he developed quadriparesis, confusion, urinary retention, and respiratory arrest. He was treated with mechanical ventilation and antibiotics for pneumonia; the neck was stabilized with a hard collar. He had incomplete motor recovery (legs better than arms) and cognitive recovery three months after injury. Dong[4] describes a patient who developed torticollis after endotracheal anesthesia and was subsequently shown to have Chiari I on MRI.

Despite such reports, Chiari I has not been described in cases of spinal cord injury submitted to the National Football Head and Neck Injury Registry or the National Center for Catastrophic Sports Injury Research (J. Torg and R. C. Cantu, personal communication, 1994). However, since both Chiari I and sports-related spinal cord injury are rare, even a few cases like ours could indicate a predisposition of patients with Chiari I to spinal cord injury.

We disqualified this individual from play because of his single episode of quadriparesis, the Chiari I malformation with an unusually long tonsillar herniation, and the obliteration of the subarachnoid space at the level of the medulla. The decision for surgery, made separately, was based on these factors and the perceived risk of subsequent development of syringomyelia. Decisions

about return-to-play and surgery must be made on a case-by-case basis, and medical opinions may differ.

Several conclusions can be drawn. Chiari I malformation with or without syringomyelia may be a cause of spinal cord injury without radiographic abnormality. MRI should be considered in evaluation of athletes with transient quadriparesis. Patients with Chiari I with syringomyelia, obliteration of the subarachnoid space, or indentation of the anterior medulla should probably not play contact sports and should have neurosurgical evaluation. Further studies will be necessary to define the proper management of athletes with asymptomatic Chiari I malformation identified by MRI.

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