Chiari Malformation in a Collegiate Volleyball Player

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Brain Anatomy

The brain consists of the cerebrum, the diencephalon (which contains the thalamus, hypothalamus, tuber cinereum, and infundibulum), the brainstem (which includes the midbrain, pons and medulla oblongata), and the cerebellum. The cerebrum and the cerebellum form the bulk of the brain (Figure 1). Cerebellar tonsils are two pegs of tissue that project from the inferior surface of the cerebellum at the base of the brain. The inferior vermis is a bundle of tissues connecting the two halves of the cerebellum. These two structures can also protrude downward into the foramen magnum and are significant in the diagnosis of Chiari Malformation Type I.

Chiari Malformation (CM), also known as Arnold-Chiari Malformation, is a congenital hindbrain deterioration characterized by downward displacement of the cerebellar tonsils into the spinal canal. CM is not uncommon itself, although it typically manifests itself in conjunction with other problems. Some researchers believe that it occurs as a result of fetal developmental malformations in the posterior container of the brain.

CM is subdivided into four categories according to degree of displacement and severity of the symptoms (Table 1). Chiari Malformation Type I (CM-I) is characterized by a 3-5 mm herniation of the cerebellar tonsils below the foramen magnum. In CM-I, neurological symptoms are not typically found until adolescence or adult life. The symptoms of CM-I include headache that is mainly occipital, cerebellar ataxia, progressive quadriplegia, and down-beating nystagmus. The headache is a rather specific, and usually protracted, suboccipital-occipital headache of variable quality and duration. The headaches are aggravated by Valsalva’s maneuver, exertion, cough, or postural changes. The headaches are typically relieved by occipital-suboccipital craniectomy to remove excessive pressure. Both migraine and tension-type headaches occur with the same frequency that they occur in the general population. Other typical complaints include visual and neuro-opthalmologic abnormalities, lower cranial nerve symptoms, and spinal cord disturbances, such as those presented in Table 2.

Concussions are the most common type of head injury seen in all sports. Studies have shown that 60%
Table 1 Diagnostic Characteristics of Chiari Malformation

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<th>Category</th>
<th>Diagnostic Characteristics</th>
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| Type I   | • Downward displacement of the cerebellar tonsils out the inferior opening of the skull into the spinal canal.  
          • No other structures of the brain are displaced.  
          • Common association of fluid accumulation within the interior of the spinal cord. |
| Type II  | • Greater displacement of brain structures into the spinal canal.  
          • Displacement of the inferior vermis, fourth ventricle, choroid plexus, and medulla. |
| Type III | • Includes myelodysplasia with spina bifida (division or defect in the posterior portion of the bony spine) with an associated herniation of a fluid sac into the overlying defect in tissue closure. |
| Type IV  | • Very rare  
          • Characterized by hypoplasia (incomplete development) or aplasia (lack of development) of a portion of the cerebellum.  
          • Not considered a hindbrain herniation. |

Table 2 Secondary symptoms of Chiari Malformation

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|                      | • Vision changes  
                      | • Dizziness  
                      | • Autonomic symptoms (orthostatic intolerance)  
                      | • Muscle weakness  
                      | • Unsteady gait  
                      | • Numbness and tingling in the extremities  
                      | • Chronic fatigue  
                      | • Sleep apnea  
                      | • Tinnitus (ringing, buzzing or watery sounds in the ears)  
                      | • Hearing loss  
                      | • Impaired ability to coordinate movement  
                      | • Episodes of acute pain in and around the eyes |
|                      | These symptoms are exacerbated by exertion, leaning the head backward, or coughing. |

of concussions cases seen in Emergency Departments return for follow-up with post-concussion symptoms. The same post-concussion symptoms can mimic CM-I. Thus, CM-I is often misdiagnosed as Post-Concussion Syndrome.

Diagnostic Techniques for CM-I

The most reliable test for CM-I is Magnetic Resonance Imaging (MRI). With the advent of MRI, physicians have acquired the ability to diagnose CM earlier.

Treatment Options

Surgical intervention is an option for all grades if symptomatic CM and/or herniation of more than 5 mm. Several surgical procedures for CM include posterior fossa reconstruction, craniovertebral decompression, and craniectomies. The surgery of choice depends on the severity of the anomaly and its associated symptoms, along with physician preference.

The purpose of this case report is to describe a common brain anomaly that may be asymptomatic, thus having the potential be undiagnosed in an athletic setting. Head injuries to a person with CM-I may result in more prominent or longer-lasting symptoms and neurologic deficits. There is a significant amount of published literature on CM in children, but there is little published information on CM after a traumatic injury. This anomaly was a chance occurrence, but it highlights the importance of referring an athlete to a physician when he or she has vague symptoms after a closed head injury that does not resolve in a timely manner.