CASE REPORT

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Death Following Minor Head Trauma in Two Adult Individuals with the Chiari I Deformity

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ABSTRACT: Reports of sudden death associated with minor head trauma in adults with the Chiari I malformation are rare. We describe two such cases, occurring in a 71-year-old individual and in a 22-year-old individual. In both cases, postmortem examination revealed prominent cerebellar tonsillar herniation and, in one, associated tonsillar sclerosis and hydrocephalus. Evidence of trauma was minimal in both, and was not sufficient to explain these fatalities. We describe the neuropathologic findings and discuss the differential diagnostic considerations in these cases of sudden death. The implications regarding manner of death are also emphasized.

KEYWORDS: forensic science, Chiari I malformation, sudden death, minor head trauma

In 1891, Chiari described three types of malformations of the posterior fossa structures associated with congenital hydrocephalus (1). The type II malformation presents in infants and consists of caudal displacement of the posterior cerebellar vermis into or through the foramen magnum, with associated brain stem displacement and usually also a spinal dysraphic lesion. In contrast, a type I malformation consists of downward displacement of the cerebellar tonsils into or through the foramen magnum, usually with sclerosis of the tonsillar folia and often with associated hydrocephalus. Central defects (e.g., syringomyelia) may be present in the cervical cord. The type I configuration is often referred to as ''chronic tonsillar herniation,'' and, depending on the presence or absence of associated abnormalities, as the adult type of Arnold-Chiari malformation (2).

The Chiari type I malformation frequently produces symptoms or signs of a chronic nature. These symptoms and signs have been categorized in various clinical complexes such as paroxysmal intracranial hypertension, central cord disturbance, cerebellar dysfunction, spasticity, and bulbar palsy (3). Age at presentation is variable, but is often adulthood. Surgical decompression of the posterior fossa is frequently beneficial (3,4). Case reports and series have documented sudden death in children or adolescents who were found at autopsy (2,5) or by radiography (6) to have a Chiari I malformation. The latter individuals usually do not have prior symptoms suggestive of the presence of the underlying malformation. Such deaths are usually, though not always (6), associated with temporally related minor head trauma. Sudden deaths in adults associated with the Chiari I malformation are much more rare; the literature documents only one report of a 25-year-old individual with this posterior fossa anomaly who expired after a minor blow (during an altercation) to the head (7). Two additional cases of adults dying unexpectedly, and in whom the Chiari I malformation may have contributed to their deaths, are described here. These cases are discussed in the context of other plausible causes of death, and in relation to determination of the manner of death.

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Case Histories and Postmortem Findings

Case 1—A 71-year-old white male embarked on a morning exercise excursion and was later discovered prostrate on the ground adjacent to his bicycle. Events immediately surrounding his demise were not witnessed. No past medical or surgical history was available. At autopsy, external examination did not reveal evidence of trauma or violence to the body. However, examination of the head revealed a single small superficial puncture wound in the right parietal scalp. Internal examination was limited to the head and brain. Focal subgaleal hemorrhage was present under the puncture wound, but no skull injury, subdural hemorrhage, or epidural hemorrhage were present. Examination of the brain revealed no subarachnoid hemorrhage and no contusions. The cerebellar tonsils were markedly prominent and descended bilaterally, with molding around the medulla (Fig. 1). On the cut surface the tonsillar folia appeared sclerotic where these structures impinged on the edge of the foramen magnum (Fig. 2). Microscopic examination confirmed the presence of focal atrophy of the tonsils; sections demonstrated leptomeningeal fibrosis, thinning of the underlying cerebellar molecular layer, loss of Purkinje cells and thinning of the granular cell layer (Fig. 3). Coronal slices of the cerebrum revealed striking hydrocephalus without gyral flattening, sulcal widening, parenchymal edema, or transtentorial or subfalcine herniation (Fig. 4). No contusion or other evidence of traumatic injury was present in the cerebrum, cerebellum or brain stem.

FIG. 1—*Case 1, anterior view of the cerebellum with the brain stem removed. Prominent cerebellar tonsils.*

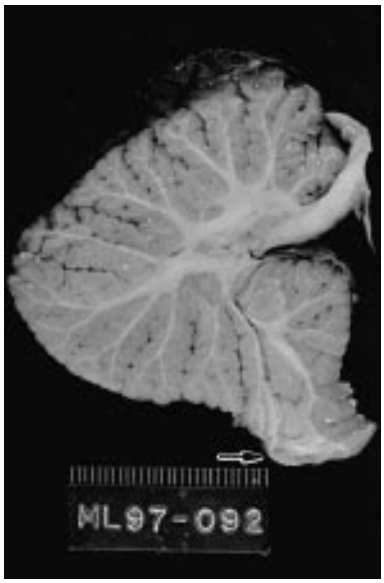

FIG. 2—*Case 1, parasagittal section of the cerebellum. Focal sclerosis of the tonsillar folia is evident (arrow).*

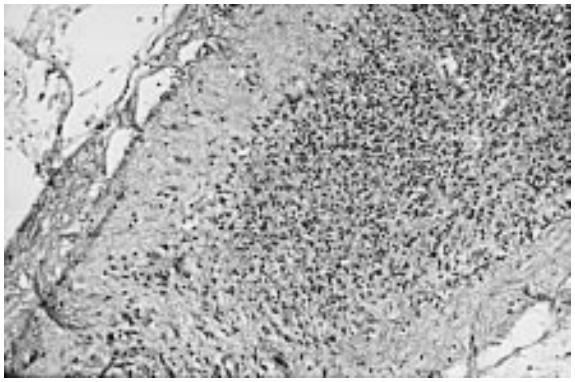

FIG. 3—*Microscopic section of a sclerotic tonsillar folium, Case 1. Depleted Purkinje cells, thinned molecular and granular cell layers, and leptomeningeal fibroglial scar (hematoxylin-eosin/luxol fast blue; original magnification ×12.5).*

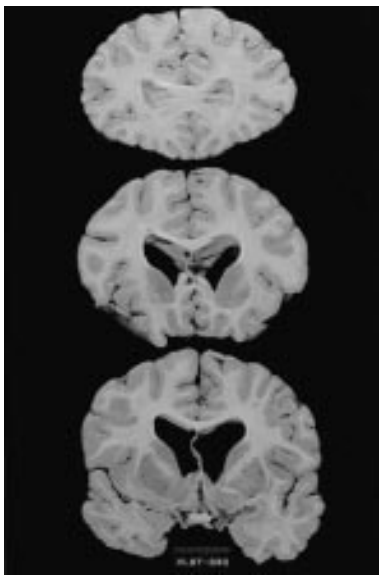

FIG. 4—*Case 1, hydrocephalus. No transtentorial herniation.*

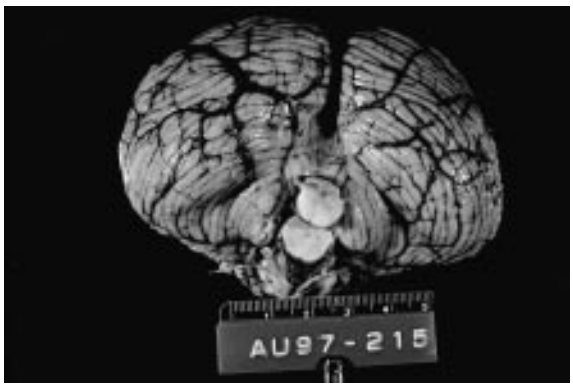

FIG. 5—*Case 2, inferior view of the cerebellum with brain stem attached (medulla is partially transversely sectioned. Prominent cerebellar tonsils with marked groove corresponding to the edge of the foramen magnum.*

Case 2—A 22-year-old previously healthy black male was playing basketball with some friends when he suddenly lost consciousness, fell to the ground, and sustained minor trauma to the occipital region of his head. Following this, his pupils were dilated and his blood pressure was unobtainable. Despite resuscitative attempts, he was pronounced dead approximately one hour after the fall. A postmortem drug screen was negative for alcohol or common drugs of abuse. At autopsy, no traumatic lesions were present on the extremities, trunk, or abdomen. Examination of the head revealed a superficial laceration of the occipital scalp. The skull was not fractured and no subgaleal or subdural hemorrhage was present in the occipital area. Examination of the brain revealed very mild and focal diffuse subarachnoid hemorrhage of the inferior frontal lobes, spanning the gyri rectus bilaterally, consistent with a recent contrecoup type injury. No contusion of the occipital lobes was evident. The cerebellar tonsils were prominent and descended caudally bilaterally, with molding around the medulla, but the tonsils were not sclerotic on gross or microscopic examination (Fig. 5). Serial coronal slices of the cerebrum did not reveal hydrocephalus or other diffuse or focal abnormalities. In particular, the gyri were

not flattened, the sulci were not compressed, and there was no other evidence of cerebral edema. No gross or microscopic abnormalities of the brain stem or spinal cord were present. Examination of the other internal organs revealed a floppy mitral valve and microscopic evidence of acute subendocardial ischemia.

Discussion

The cases described here are unusual in that they represent examples of the rarely reported association of adult sudden death and an underlying Chiari I malformation. In each case, associated minor head trauma was present. In case 1, the events immediately surrounding the death were not witnessed, and it is therefore unclear whether the minor trauma (small parietal scalp puncture wound assumed to have been incurred upon the subject's fall from the bicycle) was antecedent or subsequent to his death. If the latter is the case, it is possible that neck hyperextension, or other minor trauma, was at play. However, as mentioned above, deaths in children with this posterior fossa abnormality have been reported without antecedent trauma (6). In the second example, witnesses suggested that the individual lost consciousness and fell as a result. An occipital head impact was documented and frontal contrecoup injuries were present at autopsy. Again, loss of consciousness may have been precipitated by the posterior fossa anomaly because of neck hyperextension or other positional insult, or without a direct traumatic event. The mechanism of death is probably the same whether direct minor head trauma occurs or whether the insult is purely positional, as in neck hyperextension. We hypothesize that sudden pressure of the bony edge of the foramen magnum against the displaced cerebellar tonsils produces immediate compression of the underlying respiratory centers or cardiac centers within the medulla. This sudden compression may therefore impair vital centers (causing respiratory arrest or a cardiac arrhythmia) without producing acute anatomic changes.

An important point illustrated by these examples is the difficulty in assigning the exact cause of sudden death in cases where several potential explanations exist. In case 1, head and brain examination were performed expressly to rule out the presence of a direct major traumatic cranial insult because of the external finding of a puncture wound. Inspection of the fixed brain revealed the cerebellar abnormality. The presence of chronic tonsillar herniation suggests a possible cause of death, but is not conclusive. An acute cardiac event (arrhythmia, ischemia/infarct) may also be suggested to explain his death. However, examination of the heart in such cases is unlikely to reveal anatomic evidence of acute myocardial infarct after such a short survival and would not demonstrate anatomic evidence of a terminal arrhythmia. In case 2, a floppy mitral valve was noted at autopsy. The valvular abnormality may have been contributory to this patient's demise (sudden death probably due to arrhythmia is known to occur in a small fraction of patients with this valvular abnormality (8,9)), or may have been merely an incidental finding. Studies of floppy mitral valves from patients dying from sudden death versus those dying of other causes document only minor population differences in valvular dimensions; no clear-cut differences separate these two entities in an individual patient (10). Subendocardial myocytolysis was present, but this is a nonspecific indicator of terminal ischemia and does not explain death. An alternative sequence is that the valvular abnormality may have led to palpitations, sudden loss of consciousness, minor head trauma (potentially survivable in a normal individual), and ultimately death because of the posterior fossa abnormality.

These cases also demonstrate some potential difficulties in determining the manner of death in some cases of sudden death. Sudden death related to the presence of a Chiari I malformation in children is not characterized by acute brain swelling, acute brain infarct, or other anatomic evidence of acute brain injury. Similarly, sudden death related to mitral valve prolapse, or very early myocardial infarct, for example, is not accompanied by anatomic evidence of an acute cardiac lesion. Indeed, one accepted requisite for attribution of death to a floppy mitral valve is that it is the sole anatomic finding (for example, Ref *9*). The question is of more than mere academic interest. In case 1, for example, if the Chiari I malformation were the underlying cause of death, the death was probably precipitated by a fall from the bicycle—manner of death: accident. However, if his demise were cardiac in nature, and antecedent to his fall, the manner of death is natural. Case 2 is similar in that if this individual's demise was cardiac in nature, the manner of death is natural. However, if death were due to the Chiari I malformation, it may have been precipitated by neck hyperextension, or by his fall (since accounts of witnesses may not be entirely accurate concerning such a quick, unexpected event)—manner of death: accident.

In conclusion, we report two cases in which the presence of a Chiari I anomaly may have precipitated sudden death in two adult individuals. Sudden death is more frequently described in children with this anomaly, but we feel that routine close examination of the posterior fossa structures in adults dying suddenly and unexpectedly may reveal a higher than previously expected incidence of this association. More experience with similar cases may lead to a better understanding of the mechanism of death in such cases, and may allow for more definitive determination of the cause of death in these persons. Finally, the presence of this anomaly may, in many instances, influence the assignment of manner of death, as detailed above.

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