

Unexpected Brain Death in a 20-year-old Man with Chiari Type I Malformation after Trauma: Case Report and Review of Literature

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Abstract The Chiari I malformation is a congenital anomaly of unknown origin consisting of caudal displacement of the cerebellar tonsils protruding through the foramen magnum, whereas the brainstem and fourth ventricle remain in the cranial vault. We present a case of a sudden brain death of a 20-year-old adult man with trauma who subsequently was found to have a previously undiagnosed Chiari I malformation.

Keywords: Chiari I malformation, brain death, trauma

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1. Introduction

Chiari malformation (CM) is a congenital hindbrain anomaly caused by displacement of the brainstem and cerebellum from the foramen magnum into the cervical spinal canal. CM can sometimes be acquired, and the cause is unknown, but many theories have been suggested for the etiology such as hindbrain dysgenesis and developmental pause theory, small posterior fossa/hindbrain overgrowth and neuroectodermal-mesodermal spatial dyssynchrony. However, none of these theories can explain all CM types and it is believed to occur through their combinations [1-7].

In Chiari malformation type 1 (CM-1), cerebellar tonsils are symmetrically herniated more than 5 mm below foramen magnum as a result of the combination of several theories suggested for the etiology [1,2,3,5,6,8]. CM-1 is the simplest form of Chiari malformation and called 'Adult type CM' since it occurs more in the third and fourth decades. CM-1 may be congenital or acquired [5,7].

The present paper aimed to present a case of CM-1 detected in a patient who was under follow-up in the ICU after trauma. Brain death occurred afterward, which has not been previously reported in the literature.

2. Case

A 20-year-old male patient was brought to the emergency service because a heavy object fell on him. He was conscious, oriented, and cooperative upon admission (GCS=15); his blood pressure was 110/60 mmHg, his

heart rate was 104/min and his respiratory rate was 16/min. There was an open fracture in the right tibia in the FM, and no pathology was found in his routine laboratory findings. After initial intervention, he was taken to the ICU with a diagnosis of multitrauma.

The craniocervical CT revealed cerebellar tonsils with extension over the inferior to the foramen magnum (approximately 6.5 mm) (Figure 1, Figure 2, Figure 3), and a cystic lesion that could be consistent with the appearance of syrinx cavity in lower cervical and thoracic region medulla spinalis. CT findings were consistent with CM-1. It was ascertained from the medical history through his family that his father had CM-1 and died unexpectedly at the age of 45.



Figure 1. Cranial CT scan of patient showing 6.5 mm tonsillar herniation consistent with Chiari 1 malformation



Figure 2. Sagittal section of cranial CT scan of patient



Figure 3. Axial section of cranial CT scan of patient

The thoraco-abdominal and pelvic CT revealed a fracture of the twelfth thoracic vertebra, segmental fracture in left femoral diaphysis, and segmental fracture in bilateral ramus superior and inferior to the pelvis.

The open fracture of the right tibia was splinted and antibiotic treatment was initiated. The patient was closely followed hemodynamically and was administered four units of erythrocyte suspension in the emergency service and four units in the ICU. A cystofix was placed for urinary retention caused by the pelvic trauma.

The patient had bleeding from the open fracture in the tibia and was surgically treated under general anesthesia in order to place a tibial and pelvic external fixator. There were no hemodynamic problems during the operation. The patient was followed-up as intubated postoperatively, but he did not recover consciousness, although he did not receive any sedation, and a control cranial CT was performed. Cerebral edema was detected and anti-edema treatment was initiated. The brain stem reflexes could not be obtained on day 3 of the follow-up, and it was determined that he did not have spontaneous ventilation.

Doppler USG and SPECT, which were performed upon the absence of significant activity in EEG, showed no cerebral flow (Figure 4). The apnea test was positive and the patient was diagnosed with brain death. The proposal for organ donation was not accepted by the family and the patient died.

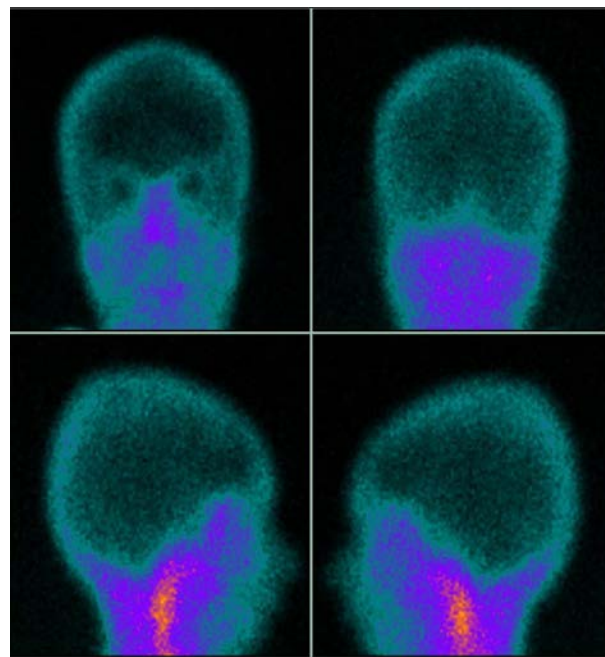


Figure 4. Static (anterior, lateral) scans and axial sections of SPECT scan of patient showing no blood flow in brain

3. Discussion

The prevalence of symptomatic CM-1 is approximately 1 in 1000 in general population. Although it was shown in the autopsies early on, it has been observed to be more common than estimated with the use of MRI. Patients are asymptomatic and diagnosed incidentally, but may also present a wide spectrum of various clinical and radiological symptoms. The pathogenesis of Chiari malformation has not been precisely understood. The increased number of reported cases, and its pathogenesis and clinics have presented the need for more understanding [1,2,3,6].

A great majority of the cases is congenital; however, there may be CM-1 cases acquired secondary to CSF hypotension (iatrogenic or spontaneous) or increased intracranial pressure. CM-1 is mostly seen in the third and fourth decades of life and called adult type CM [1,3,5]. In the present case, the male patient was asymptomatic until the age of 20 and CM-1 was incidentally detected in cranial CT performed in the emergency service after trauma.

In CM-1, cerebellar tonsils are symmetrically herniated more than 5 mm below foramen magnum [5,9,10,11]. Patients are symptomatic if the herniation of cerebellar tonsils is > 12 mm, and 30% of cases with herniation between 5 and 10 mm and all of the cases with herniation < 5 mm are asymptomatic [5]. In the present case, cerebellar tonsils were herniated approximately 6.5 mm downwards in CT measurements and the patient remained asymptomatic until the age of 20.

MRI delivers the final diagnosis in CMs. Malformation diagnosis with MRI has facilitated the detection of the extent of cerebellar herniation and the syringomyelia that can accompany it [3,5,8]. In the present case, a CT was performed upon admission to the emergency service, and an MR could not be performed in the subsequent process due to external fixators. Therefore, syringomyelia could not be clearly detected. A previous study indicated that CM could be diagnosed via CT or MRI, but some cases could be missed with CT [12].

The assumption for CM is that the posterior fossa is very small, causing tonsillar herniation and resulting in obstruction of cerebrospinal fluid (CSF) flow at intracranial and spinal subarachnoid distance at the foramen magnum level. CM-1 is reported to involve hydrocephalus at a rate of 7% [4]. Hydrocephalus was not detected in the present case.

The clinical findings of CM, according to Milhorat, are classified as suboccipital headache, ocular disorders, otoneurological disorders, brainstem-lower cranial nerves-cerebellum involvement, and spinal cord disorders. In the presence of syrinx, there may be weakness of the legs and occasionally accompanying scoliosis. At times, these symptoms may occur after a trauma or an acute physical activity [2,3,5,8]. In the present case, the asymptomatic patient was diagnosed incidentally after trauma.

One study, which investigated 364 patients diagnosed with CM-1, found common findings as syringomyelia in 65%, scoliosis in 42% and bacillary invagination in 12% of cases. A positive family history was reported for CM or syringomyelia in 12% of the patients. Twenty-one families were shown to have genetic transmission consistent with autosomal dominant or recessive inheritance [3]. In the present case, the medical history ascertained through his family revealed that his father was symptomatic with the diagnosis of CM-1 and died unexpectedly at the age of 45 despite treatment.

As is the case with all neurosurgical patients, general medical treatment should be considered as the first step of treatment in CM, if possible. If CM does not involve syringomyelia and it is asymptomatic, there are no surgical indications. If syringomyelia is present and it is asymptomatic, then the surgical indication should be discussed. In general, surgery is performed in patients with progressive neurological symptoms. Asymptomatic patients should be followed-up [2,8]. One previous study emphasized that the clinical findings of increased intracranial pressure should be considered during the preoperative evaluation and a multidisciplinary approach should be adopted in pregnant patients with CM-1 [13]. Another study showed cranial constriction, cranial compression, spinal cord union, intracranial hypertension, and intraspinal hypotension to be associated with CM etiology [6]. In the present case, the clinical presentation might have been caused by the intraoperative increased intracranial pressure. It was reported that a 29-year-old patient diagnosed with CM-1, who was administered a narcotic analgesic for pain management, had cardiopulmonary arrest due to hypercarbia-related increased intracranial pressure and decompensation of the cardiopulmonary center in the brainstem [10]. In the present case, the patient had no problems related to the partial CO₂ levels during the follow-up.

CM-1-related sudden neurological deficits and sudden deaths are defined in several ways. Deaths usually tend to be associated with chronically increased intracranial pressure in the posterior fossa [10,12,14,15,16]. It was reported that intracranial pressure might increase and result in increased herniation of cerebellar tonsils after a minor head trauma in a CM-1 patient with headache and obstructive hydrocephalus. Again, the same study reported that the cerebellum became more sensitive to tonsillar ectopia in an 11-year-old CM-1 patient with cerebellar contusion after head trauma [16]. The literature reports for unexpected 'sudden death' CM-1 are usually related to respiratory arrest, trauma, surgery/general anesthesia, and alcohol use; however, the role of such factors should be investigated. There are reports for CM-related sudden death after trauma in the literature [14]; however, no report has been found for CM-related brain death. We believe that the present case is the first case with CM-1 accompanied by brain death.

In conclusion, we believe that treatments should be planned to reduce intracranial pressure; proper analgesia and sedation should be provided, and surgical traumas should be minimized once the anomaly is detected since there is low tolerance to increased intracranial pressure in such rare cases.

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