Delayed hydrocephalus associated with traumatic atlanto-occipital dislocation: Case report and literature review

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Abstract

Background: Traumatic atlanto-occipital dislocation (AOD) is a rare but often fatal injury. Consequently, long-term data regarding surviving patients have been limited. In particular, the occurrence of hydrocephalus is not well-documented.

Case Description: A 33-year-old male sustained AOD as a consequence of a motor vehicle collision. Although he did well initially after an occipitocervical fusion, 1 month after his operation, he exhibited signs of increased intracranial pressure (bilateral abducens nerve palsies, worsening headaches, and fatigue). He was found to have hydrocephalus, which was responsive to shunting.

Conclusion: Manifestations of hydrocephalus after AOD can be variable, ranging from interval ventricular dilatation to pseudomeningoceles and syringomyelia. In addition, the timing of presentation can be acute, requiring emergent external ventricular drainage, or delayed, requiring ongoing vigilance. Consequently, as more patients survive this once thought to be fatal injury, caution for hydrocephalus is stressed.

Key Words: Atlanto-occipital dislocation, hydrocephalus, occipitocervical fusion

BACKGROUND

Traumatic atlanto-occipital dislocation (AOD) is a rare but often fatal injury. Consequently, long-term data regarding surviving patients have been limited. In particular, the occurrence of hydrocephalus is not well-documented. We report a patient with delayed hydrocephalus and review the literature.

CASE PRESENTATION

A 33-year-old male presented after a high-velocity motor vehicle collision. On presentation, his Glasgow Coma Scale (GCS) was 3T, with reactive pupils but no corneal/cough/gag reflexes. Imaging demonstrated AOD [Figure 1], with associated intraventricular hemorrhage (IVH) and subarachnoid hemorrhage (SAH) [Figure 2]. An external ventricular drain (EVD) was placed for low GCS and progressive dilatation of the ventricles. Shortly thereafter, he was able to follow simple commands with the right extremities. He underwent occiput to C4 fixation/fusion. His EVD was weaned within 7 days. He continued to progress neurologically. At 1 month, he exhibited bilateral abducens nerve palsies, worsening headaches, and fatigue. A computed tomography (CT) of the head showed interval enlargement of his ventricles.
He underwent a ventriculoperitoneal shunt placement. At discharge, 2 months after presentation, he was doing well, ambulating with stand-by assist/walker.

**DISCUSSION**

AOD involves significant instability of the craniocervical junction. The tectorial membrane and alar ligaments are the most commonly injured ligaments. Most frequent mechanisms of injury are motor vehicle accidents and pedestrian vehicle accidents. Patients often die in the field; autopsy studies have noted that AOD was present in 6–20% of the patients who sustained fatal traffic accidents.\(^1\,\,^2\,\,^5\,\,^11\) For those who remain alive upon arrival to a hospital, AOD constitutes 1% of cervical spine injuries.\(^1\,\,^2\) Pediatric patients, due to large head-to-body ratio, flatter atlanto-occipital articulation, and weaker ligaments, have a higher potential for AOD compared to adult patients.\(^8\,\,^10\,\,^14\)

Neurological presentation can be variable, ranging from no deficits to severe deficits (from injuries to cranial nerves, brain stem, and spinal cord), ventilator dependence, and death.\(^10\) Moreover, due to the inherent high-energy impact typically involved with AOD, the presence of polytrauma is frequent.\(^11\) The abducens nerve and hypoglossal nerves are commonly injured due to abrupt distraction forces.\(^1\) With improved medical care and advanced diagnostic imaging, survival has now become more common. Consequently, only small case series and case reports exist regarding long-term outcomes from AOD.\(^1\)

Manifestations of hydrocephalus after AOD include pseudomeningoceles, syringomyelia, and interval ventricular dilatation. Table 1 summarizes the available literature. The majority of the patients have been pediatric patients. Five have exhibited retropharyngeal pseudomeningoceles and 2 have exhibited cervical syrinx. Moreover, most of the cases have exhibited delayed hydrocephalus, diagnosed up to 3.5 months after the initial injury. On the other hand, acute obstructive hydrocephalus can occur, which can be secondary to a retroclival hematoma, requiring emergent ventricular drainage. Klimo et al.\(^9\) noted that 4 of 14 pediatric patients with AOD and 5 of 16 pediatric patients with AOD and atlantoaxial fracture-dissociation exhibited hydrocephalus, without describing the details of their presentation.

Though some series report a high rate of hydrocephalus (up to 30%) associated with AOD,\(^9\) other series report no occurrence of hydrocephalus.\(^11\) Overall, the actual number of documented cases remains low because these series had small population sizes and overall survival data have been limited. Mechanism for ventricular dilatation may be related to scarring along the basal subarachnoid cisterns, arachnoid villi, or fourth ventricular outlets secondary to intracranial hemorrhage.\(^11\) Pseudomeningoceles can occur through a dural defect sustained from the initial trauma; with distorted cerebrospinal fluid (CSF) dynamics, CSF drainage may persist through the defect. Post-traumatic syringomyelia, also thought to be secondary to distorted CSF dynamics, has been known to be a late complication.
after spinal trauma.\(^{[7,11]}\) It remains unclear if surgical fixation increases the potential for hydrocephalus. Several surgical procedures have been employed, including dural defect repair and shunting (ventriculoperitoneal and lumboperitoneal).

Our patient exhibited bilateral abducens nerve palsies associated with ventricular dilatation. This presentation is consistent with a delayed hydrocephalus. Though injury to the abducens nerve is commonly associated with AOD, the time delay to this symptom argues against its association with AOD. Our patient also had scattered IVH and SAH, requiring a temporary EVD. This likely increased his risk for delayed hydrocephalus.

**CONCLUSION**

Manifestations of hydrocephalus after AOD can be variable, ranging from interval ventricular dilatation to pseudomeningoceles and syringomyelia. In addition, the timing of presentation can be acute, requiring emergent EVD, or delayed, requiring ongoing vigilance. Consequently, as more patients survive this once thought to be fatal injury, caution for hydrocephalus is stressed.

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**Conflicts of interest**

There are no conflicts of interest.

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**Table 1: Review of Literature**

| Literature       | Year | Age | Gender | Mechanism       | Time of diagnosis of hydrocephalus | Surgical fixation prior to diagnosis? | Ventricular dilatation | Syringomyelia | Retropharyngeal pseudomeningocele | Treatment                              |
|------------------|------|-----|--------|-----------------|-----------------------------------|-------------------------------------|------------------------|--------------|-------------------------------|---------------------------------------|
| Cognetti[11]     | 2006 | 19  | Male   | MVA            | 3 weeks from injury               | Yes                                 | x                      | x            | x                            | Lumboperitoneal shunt                  |
| Collalto[11]     | 1986 | 3   | Male   | Pedestrian vs car | 12 weeks from injury             | No                                  | x                      | x            | No                           | VP shunt                               |
| Gutierrez[11]    | 2008 | 29  | Female | MVA | 3 weeks from injury | No                                  | x                      | x            | No                           | VP shunt                               |
| Naso[11]         | 1997 | 26  | Male   | MVA            | 3.5 months from injury            | No                                  | x                      | x            | No                           | VP shunt                               |
| Reed[11]         | 2005 | 9   | Male   | Pedestrian vs car | 4 weeks from injury              | Yes                                 | x                      | x            | No                           | Foramen magnum/posterior fossa decompression and duraplasty |
| Vera[11]         | 2007 | 5   | Female | MVA | Promptly after admission | No                                  | x                      | x            | No                           | EVD, died shortly afterwards          |
| Williams[11]     | 1995 | 3   | Male   | MVA            | Promptly after admission         | No                                  | x                      | x            | No                           | VP shunt                               |

MVA: Motor vehicle collision, VP: Ventriculoperitoneal. Klimo et al[11] also reported a small series that noted patients who exhibited hydrocephalus without further details.