Traumatic Atlanto-occipital Dislocation in Children Followed by Hydrocephalus – A Case Report and Literature Review

Deslocamento atlanto-occipital em crianças seguido por hidrocefalia – Relato de caso e revisão da literatura

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Abstract

Traumatic atlanto-occipital dislocation (TAOD) are uncommon injuries associated with high immediate mortality rate and occurs more than twice in children than adults, due to biomechanical properties and immaturity of children's cervical spine. We report a pediatric patient with TAOD, who underwent occipitocervical stabilization and also developed a late hydrocephalus requiring a shunt procedure. A six-year-old boy was admitted to the emergency department after a car accident with refractory cervical pain. A cervical computed tomography (CT) scan showed an anterior C1–C2 level hematoma, and a dynamic CT scan demonstrated an increasing basion-dens interval on extension. Cervical magnetic resonance imaging (MRI) showed discontinuity of the tectorial membrane and diffused hyperintense signal on the left alar ligament. These findings were attributed to TAOD, and an occipitocervical fusion was performed. The pain and neurological status improved after surgery, but after 3 months he returned with persistent vomiting, headache, and a CT scan showing hydrocephalus. Then, a ventriculoperitoneal shunt was performed, improving the symptoms. One year after the injury, the patient remained asymptomatic, and a later radiography demonstrated satisfactory bone fusion. In conclusion, the decision-making process regarding treatment should consider several clinical and radiographic findings. Occipitocervical fusion is the treatment of choice, while hydrocephalus is not an unusual complication in children.

Keywords

► atlanto-occipital dislocation
► pediatric spine
► trauma
► hydrocephalus

Resumo

O deslocamento atlanto-occipital (DAO) é uma lesão incomum associada a uma alta taxa de mortalidade imediata que ocorre duas vezes mais em crianças do que em adultos, fato relacionado às propriedades biomecânicas e à imaturidade da coluna cervical pediátrica. Relatamos o caso de um paciente pediátrico com DAO traumático...
Introduction

Traumatic atlanto-occipital dislocation (TAOD) is a rare injury that is associated with a high mortality rate, since it is frequently related to cardiorespiratory arrest at the trauma scene and severe neurological impairment due to injury of the cervicomedullary junction.\textsuperscript{1-3} Traumatic atlanto-occipital dislocation occurs more than twice in children than in adults. This is due to biomechanical properties and the immaturity of the pediatric cervical spine, such as smaller occipital condyles, and a horizontal atlanto-occipital joint, which is less resistant to shear forces.\textsuperscript{4,5} Some case series reported that a certain number of children who survive the initial trauma have a positive outcome despite presenting neurological deficits.\textsuperscript{6,7}

Because of a wide range of clinical presentations, from a neurologically intact patient to one with cardiac arrest at the emergency department, as well as the association with other traumatic brain injuries, the diagnosis of pediatric TAOD can be delayed or even missed entirely, risking irreversible traumatic brain injuries. A dor e o status neurológico melhoraram após a cirurgia, mas 3 meses após, o paciente evoluiu com vômitos persistentes, cefaleia e TC de crânio evidenciando hidrocefalia. Em seguida, foi realizada uma derivação ventriculoperitoneal, com melhora dos sintomas. Um ano após, o paciente permaneceu assintomático, e a radiografia demonstrou fusão óssea satisfatória. Em conclusão, o processo de tomada de decisão quanto ao tratamento deve levar em consideração diversos achados clínicos e radiográficos. A fixação occipitocervical é o tratamento de escolha, enquanto a hidrocefalia não é uma complicação incomum em crianças.

In the present study, we report a pediatric patient with TAOD who underwent occipitocervical stabilization and also developed a late hydrocephalus requiring a shunt procedure. The diagnosis of TAOD is discussed in detail.

Case Report

A six-year-old boy was admitted to the pediatric emergency department following a car accident, where he was on the backseat wearing a conventional seat belt, and the driver ran off the road and crashed. On admission, he was reporting abdominal pain and had undergone two episodes of emesis. He was otherwise stable, wearing a cervical collar, notifying neck pain. Neurological examination showed evident pyramidal signs, with mild left side weakness. An initial cranial CT scan revealed traumatic subarachnoid hemorrhage, and a cervical CT scan showed an anterior C1—C2 level hematoma (\textsuperscript{\textbullet}Fig. 1). We performed all the following radiological measurements to diagnose TAOD using a CT scan: Wholey dens-basion interval (DBI),\textsuperscript{10} Powers’ ratio,\textsuperscript{11} Harris’ basion-axis interval (BAI),\textsuperscript{12} and Sun’s interspinous ratio,\textsuperscript{13} but all results had normal values (\textsuperscript{\textbullet}Fig. 1). Additionally, the abdominal CT scan showed a mesenteric rupture, treated non-operatively. He was then admitted to the intensive care unit with cervical immobilization. Despite the algescic management, the cervical pain was refractory, leading us to perform a dynamic (flexion and extension) cervical CT and a MRI.

On the dynamic CT scan, the vertebral bodies and facet joints remained aligned, except for the increasing distance between the basion and the odontoid process on extension (\textsuperscript{\textbullet}Fig. 2A-B). The cervical MRI demonstrated a transfixing rupture between the posterior arches of C1 and C2, an anterior arch subluxation with discontinuity of the anterior longitudinal ligament on the medium third of the dens and on the tectorial membrane. The diffuse hyperintense signal on the nuchal ligaments and on the left alar ligament was attributed to a distension/partial...
lesion (► Fig. 3A-B). The transverseligament was unimpaired as on method (►Fig. 3C). Additionally, an epidural hematoma of 1.9 mL was attached to the left anterolateral spine canal. The entirety of the findings allowed us to diagnose the TAOD, despite the near normal CT findings. Based on the exams and clinical presentation, a multidisciplinary case discussion was held, and a decision was made to perform an occipitocervical fusion (OCF).

While under general anesthesia, the patient was prone positioned, the neck was kept neutral using a Mayfield head holder, and the shoulders were retracted caudally. We performed a posterior median incision from the inion to C3. A subperiosteal dissection exposed the squamous part of the occipital bone, the posterior tubercle of C1, and the spinous process of C2, and then the lamina, and the inferior articular process of C2 and the C2–C3 joint. The posterior arch of the C1 was exposed further with some bleeding from the vertebral plexus, controlled by hemostatic agents and bipolar cauterization. We found and drained the epidural hematoma, secondary to the traumatic avulsion of the right C1 nerve root, with a dural injury and a high debit cerebrospinal fluid (CSF) leakage, fixed with a fat graft and fibrin.

Fig. 1 Admission cervical computed tomography (CT) scan. (A) Slightly Left–right asymmetry between the atlas and vertebrae axis in the coronal plane. (B) Sagittal CT scans showing a normal Powers ratio measurement (0.79; normal ≤ 1), but an epidural collection in the upper cervical canal. (C) Normal Harris’ basion-axis interval (3.11). The normal distance is between 12 and 0 mm in children. (D) Normal Sun’si ratio (1.16). The interspinous ratio is indicative of atlanto-occipital dislocation (AOD) by a C1–C2/C2–C3 ratio of more than 2.5.
Fig. 2 (A and B) Dynamic computed tomography (CT) scan showing the Wholey’s dens-basion interval. Note an increasing dens-basion interval on extension, in this case. (C and D) Conventional CT scan with placement of the measurement points (arrowheads) on the coronal (A) and sagittal (B) planes for calculation of the occipital condyle–C1 interval (CCI) in this case. A value of 4.05 mm was obtained (normal is < 4 mm).

Fig. 3 Cervical magnetic resonance imaging (MRI). (A) Evident hypointense collection on anterior spine canal, causing spinal cord edema, compatible with subacute hemorrhage. There is diffuse hyperintensity on nuchal ligaments. (B) On coronal plane, a left-right asymmetry and also a left alar hyperintensity are evident. (C) The transverse ligament was preserved as on method.
An occipital plate was fixed with four screws of $4.5 \times 6 \text{ mm}$ (2) and $4.5 \times 8 \text{ mm}$ (2), centered over the thickest portion of the occipital bone. Guided by fluoroscopy, bilateral polyaxial screws were positioned on the C2 pars ($3.5 \times 14$ and $3.5 \times 12 \text{ mm}$) and unilateral ones on the left C1 lateral mass ($3.5 \times 26 \text{ mm}$). The system was fixed with 2 adjusted bars, and positioned with an additional cross-link (Fig. 4A-B). Bone graft was extracted from the iliac crest and placed between the occipital bone and C2. There was no significant intercurrence during surgery. A follow-up CT further confirmed the adequate screw positioning.

Thereafter, the patient was transferred to an intensive care unit with significant improvement of the pain, being able to sit on day 1, and having no further symptoms. After hospital discharge, he underwent the follow-up process at the outpatient unit, remaining pain-free. Three months after surgery, however, he presented repetitive nighttime vomiting and headache with no signs of fever. Following a CT scan, a communicating hydrocephalus was diagnosed (Fig. 5). We collected a CSF sample and ruled out meningitis. Then, a frontal medium pressure ventriculoperitoneal shunt was performed, with radiological resolution of hydrocephalus and clinical improvement. One year after the injury, the patient remains asymptomatic, and later radiography showed satisfactory bone fusion (Fig. 4C-D).

Discussion

Traumatic atlanto-occipital dislocations are uncommon injuries associated with high immediate mortality rates. However, the likelihood of a pediatric patient surviving this almost invariably lethal injury clearly improved with time, due to
advances in emergency resuscitation and prehospital care, as well as in the quality of radiological imaging.\textsuperscript{14,15} Children who survive initial injury may have a favorable outcome with early diagnosis and treatment, before irreversible damage occurs in the cervicomedullary junction.\textsuperscript{16}

High-energy trauma is usually required to cause TAOD, typically in the form of sudden acceleration-deceleration forces on the head.\textsuperscript{17} The mechanism of injury most often reported is an automobile accident,\textsuperscript{18} although accident in which a pedestrian is struck by a motor vehicle is also a common cause in children. The high-energy mechanism of injury with TAOD frequently results in further additional injuries, especially traumatic brain injury, which may hamper the diagnosis of TAOD.\textsuperscript{19} Our patient had traumatic subarachnoid hemorrhage and blunt abdominal trauma in addition to TAOD.

**Anatomical Background**

The craniocephalic junction is the most mobile part of the spine, and stability is provided mainly by the ligaments.\textsuperscript{3} The transverse ligament secures anteriorly the odontoid process against the anterior arch of the C1, while the alar ligament attaches the dens to the anterolateral part of the foramen magnum.\textsuperscript{20} The tectorial membrane is the continuation of the posterior longitudinal ligament and connects the axis with the clivus.\textsuperscript{3,20}

Children younger than 10 years of age are particularly predisposed to TAOD because of the larger head-to-body ratio, smaller and flatter atlantooccipital joints and more flexible and weaker ligaments.\textsuperscript{21} Previous studies showed that rupture of the alar ligaments and the tectorial membrane are sufficient to result in TAOD, since the remaining ligaments that attach the upper cervical spine to the occiput are insufficient to maintain adequate stability, and these abnormalities were also noted in this case.\textsuperscript{22–24} There are usually no fractures associated with this injury, although in older children or adolescents, stronger ligaments can result in avulsion fractures at the ligamentous attachment of the skull base.

**Imaging/Diagnosis**

Although several radiographic methods to detect TAOD have been described, such as those proposed by Power, Harris, Wholey, and Sun,\textsuperscript{10–13,25} none have been proven adequate as a single diagnostic criterion.\textsuperscript{18} Also, there have been reports of significant variances from the previously accepted normal values on plain radiographs compared with the CT scans for most of these methods.\textsuperscript{26} High-quality MRI is valuable for detecting ligamentous injury and careful surgeon-supervised flexion-extension CT scan (or even a CT scan performed under cranial traction) can also be informative.

In 1979, Powers et al.\textsuperscript{11} described the Powers-ratio as a tool for the evaluation of TAOD, diagnosed by a ratio of more than 1. However, this method is only sensitive for the diagnosis of anterior TAOD. A vertical dissociation injury could result in a normal value, like in this case, and consequently go undiagnosed.

Harris et al.\textsuperscript{12} established a reliable and accepted method to diagnose TAOD on lateral cervical radiographs, the BAI. In all 50 children (100%) with no occipitocervical abnormalities, the BAI was less than 12 mm, which is considered as the upper limit of normal.\textsuperscript{12} However, this test alone has been found to have a sensitivity of 50%.\textsuperscript{16–27}

The DBI was originally described by Wholey et al.,\textsuperscript{10} and the commonly accepted cutoff on plain radiograph is 12 mm. However, normal values on CT scans were significantly different from the accepted ranges of normal on plain radiographs.\textsuperscript{26,28} Considering the pediatric population, Bertozzi et al.\textsuperscript{28} showed that the DBI was shorter than 10.5 mm in 97.5% of patients.

Pang et al. describe both the normal anatomy and radiographic findings suggestive of TAOD,\textsuperscript{15,16} but it has been discussed that these parameters cannot be applied to all age groups.\textsuperscript{5} They showed that the normal occiput–C1 joint in children has an extremely narrow joint gap (condyle–C1 interval or CCI).\textsuperscript{15} With a cutoff value of 4 mm, the CCI criterion had the highest diagnostic sensitivity and specificity for TAOD among all other radiographic criteria in their study.\textsuperscript{16} In this case, the combined CCI value (average of both the sagittal and coronal CCIs) was 4.05 and represents the only abnormal radiographic standard test, proving this test as an important tool for TAOD diagnosis in children with less evident dislocations (\textsuperscript{–Fig. 2C-D}).

**Treatment and Complications**

Occipitocervical fusion in the pediatric population has been a challenging surgery even for experienced spine surgeons. Posterior spinal fusion is the pillar treatment for TAOD, and the use of screws has now become more common in young children, in whom other techniques, such as rib grafting with wiring followed by a halo vest, are used.\textsuperscript{29–34} In a systematic review in which 285 patients underwent OCF, Hwang et al.\textsuperscript{35} found that both screw and wiring groups had very high fusion rates (99% and 95%, respectively, \( p = 0.08 \)); however, wiring was associated with a higher complication rate.

Pediatric OCF carries serious risks, and the complication rates reported in the literature range from 7.5 to 26%.\textsuperscript{36} Short-term complications include vertebral artery injury, blood loss, neurologic deterioration, dural tear, and CSF leak.\textsuperscript{36} There are also long-term risks, including hardware-related complications, infection, pseudoarthrosis, and deformity.\textsuperscript{36}
In our case, the patient developed hydrocephalus, which is a complication has also been described in previous case reports of TAOD. In a series of 14 patients, the most common postoperative complication was hydrocephalus, and the authors hypothesize that it occurs as a result of posthemorrhagic scarring within the basal cisterns or outlets of the 4th ventricle. In our case, the CSF leak associated with the epidural hematoma may have played a role in the development of a CSF disturb, as well as the root injury.

Conclusion

In conclusion, TAOD is an uncommon and challenging subject in the pediatric population. Surgical stabilization is life-saving in cases of TAOD, while missing an unstable injury could have catastrophic consequences. The diagnosis criterion is not unique, generally requiring multimodal image, especially in less evident dislocations. Fixation of the occipitocervical junction with screws have a higher fusion rate and should be considered as the treatment of choice when feasible. Finally, hydrocephalus is not an unusual complication in children, and attention is necessary, with close clinical and radiological follow-up.

Conflict of Interests

The authors have no conflict of interests to declare.

References

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