Case Report
Tumor/Infection
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LOCKED-IN SYNDROME AFTER TRAUMATIC ATLANTO-AXIAL SUBLUXATION: A CASE REPORT

SÍNDROME DO ENCARCERAMENTO DECORRENTE DE SUBLUXAÇÃO ATLANTOAXIAL TRAUMÁTICA: RELATO DE CASO

SÍNDROME DE ENCLAUSTRAMIENTO POR SUBLUXACIÓN ATLANTOAXOIDEA TRAUMÁTICA: INFORME DE CASO

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ABSTRACT

Traumatic atlanto-axial subluxation is a rare and underdiagnosed condition due to its high rate – reported to be between 60 and 80% – of early mortality. Its diagnosis takes into account the trauma mechanism, precise analyses of the imaging tests and the clinical presentation of the patient. This article describes a rare presentation of atlanto-axial subluxation associated with craniocervical dislocation as a case of locked-in syndrome. ***Level of evidence V; Retrospective observational study – Case report.***

Keywords: Locked-in syndrome; Atlanto-axial joint; Craniocervical injuries.

RESUMO

Subluxação atlantoaxial traumática é uma condição de diagnóstico raro e subestimado, devido a sua alta taxa – descrita entre 60% e 80% – de mortalidade precoce. Seu diagnóstico leva em conta o mecanismo do trauma, as análises precisas dos exames de imagem e a apresentação clínica do paciente. Este artigo relata um quadro raro de subluxação atlantoaxial associada à luxação craniocervical como um caso de síndrome do encarceramento. ***Nível de evidência V; Estudo observacional retrospectivo – Relato de caso.***

Descritores: *Síndrome do encarceramento; Articulação atlantoaxial; Traumatismos craniocervicais.*

RESUMEN

La subluxación atlantoaxoidea traumática es una afección rara y subdiagnosticada debido a su alta tasa - descrita entre el 60 y el 80%- de mortalidad temprana. Su diagnóstico tiene en cuenta el mecanismo del traumatismo, los análisis de imagen precisos y la presentación clínica del paciente. Este artículo informa de una rara condición de subluxación atloaxoidea asociada a una dislocación craniocervical como un caso de síndrome de enclaustramiento. ***Nivel de evidencia V; Estudio observacional retrospectivo – Informe de caso.***

Descriptores: *Síndrome de enclaustramiento; Articulación atlantoaxoidea; Traumatismos craneocervicales.*

INTRODUCTION

Injuries to the craniocervical junction account for 10 to 30% of the cases of cervical spine trauma.[1] Craniocervical dislocation and atlanto-axial dislocation are conditions that have high rates of morbidity and mortality, reaching mortality rates above 33% in hospital care alone, a significant number that certainly increases when considering cases with fatal outcomes that occur outside the hospital, such as traffic accidents.[2] Its initial presentation can be difficult to diagnose, and its consequences can lead to severe neurological damage.[3] In this case report, we present the unusual outcome of a patient who developed locked-in syndrome as a result of a rare craniocervical dislocation associated with traumatic atlanto-axial subluxation.

METHODS

The clinical data from the retrospective observational study was acquired through a review of the electronic medical records. The Term of Commitment for the Use of Data was signed by the authors, ensuring the confidentiality and secrecy of the data.

CASE REPORT

A 40-year-old woman pedestrian was hit by a car and subsequently hit by a pole. She was found at the scene with ECG 3 and blood pressure levels of 80 x 50 mmHg, and extra-hospital orotracheal intubation and volume expansion were performed. On arrival at the emergency service, she presented with isochoric and non-reactive pupils and no movement of the extremities. Computed tomography (CT) of the cervical spine was initially evaluated by the radiologist and there were no changes described in her report. However, after evaluation by the orthopedic team specialized in spine surgery, signs of craniocervical dissociation and atlanto-axial subluxation were seen. Harris' Rule of 12 indicated an alteration, the basion-dens interval being equivalent to 18.3 mm and the basion--axial interval to 15.8 mm (Figure 1). Anterior bilateral atlanto-axial dislocation is characteristic of Fielding and Hawkins classification type III.[4] The calculation of Power's ratio yielded a value of 0.87, considered normal (Figure 2).[5] In the coronal CT sections of the cervical spine, the noticeable increase in the occipito-C1 distance indicates vertical altanto-occipital dislocation, characteristic of type II

Study conducted at the Hospital Regional Homero de Miranda Gomes, Orthopedics and Traumatology Service, São José, SC, Brazil.
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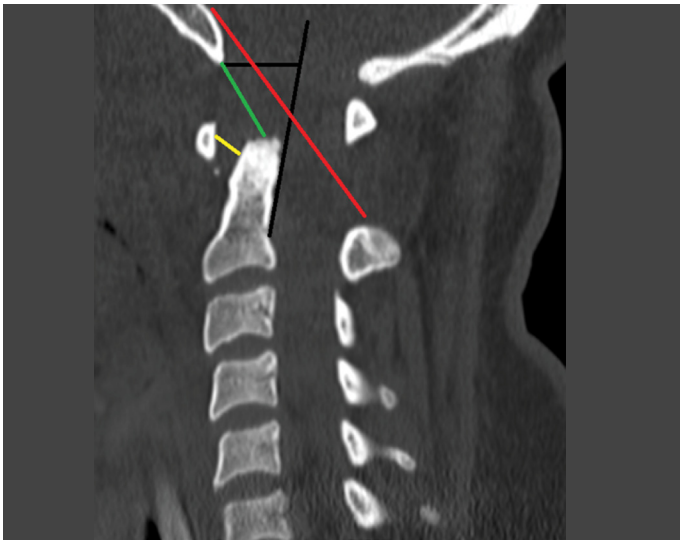

Figure 1. CT of the cervical spine, sagittal section with measurement lines. Sagittal section of the CT of the cervical spine showing the Wackenheim line (in red) posterior to the top of the odontoid, suggesting atlantocervical subluxation. The measurement of the anterior atlantodental interval (in yellow) corresponds to 5.5 mm; the basion-dens interval (BDI, green) is equivalent to 18.3 mm. The basion-axis interval (BAI, black) has a value of 15.8 mm.

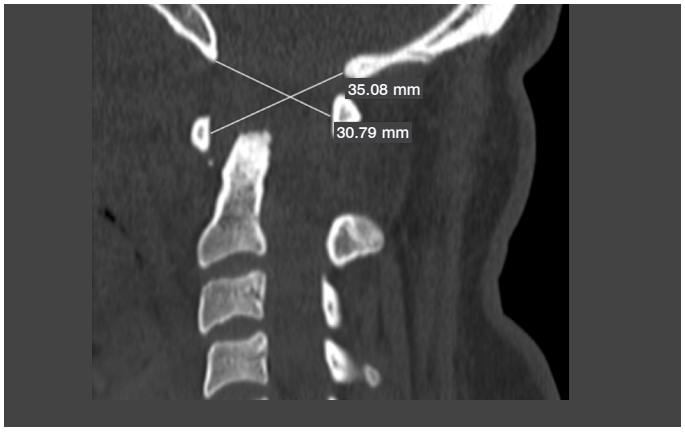

Figure 2. CT of the cervical spine, sagittal section. Measurements for the calculation of the Power's ratio, yielding a value of 0.87.

dislocation according to the classification of Traynelis et al.[6] Figure 3 shows the increase in the articular spaces, characterizing dislocation. The CT of the skull did not show lesions or hemorrhagic signs in the cerebral parenchyma. On the third day of hospitalization in the intensive care unit, the patient spontaneously opened her eyes, moved them slightly to be able to fix her gaze on the examiner, and succeeded in blinking her eyes. Due to the improvement in her condition, the patient was able to be transferred to another institution for an MRI of the skull and the cervical spine, which showed, respectively, signs of diffuse axonal injury, mainly in the region anterior to the pons next to the bulbopontine transition on the left, and signs of craniocervical dislocation and anterior subluxation of C1 over C2 (Figure 4). In the third week of hospitalization, after stabilization and clinical release, the patient was taken to surgery for occipitocervical arthrodesis. During the postoperative period, she was able to define the location of the pain and answer dichotomous questions by blinking her eyes (using a prolonged blink for "yes"). She died on the 40th day of hospitalization due to cardiorespiratory arrest resulting from nosocomial pneumonia due to bronchoaspiration.

DISCUSSION

Traumatic craniocervical and atlanto-axial dislocations can be caused by high-energy traumas that generate forces of distraction,

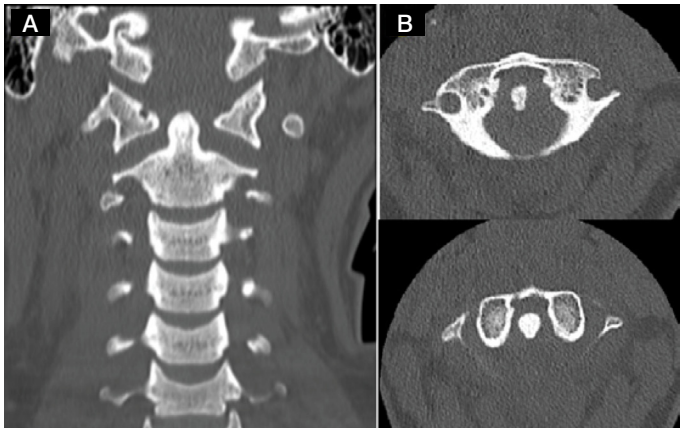

Figure 3. CT of the cervical spine. Coronal (A) and axial (B) sections showing an increase of the articular spaces with loss of joint congruence.

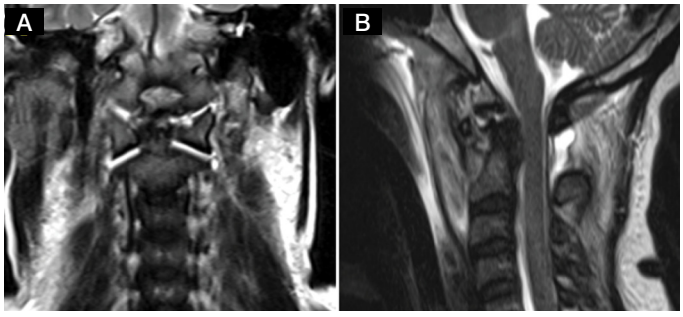

Figure 4. MRI of the craniocervical transition. T2-weighted sagittal (A) and coronal (B) sections. Visible increase of the articular space between the anterior neural arch of C1 and the odontoid process, suggesting anterior subluxation of C1, as well as an increase in the distance between the corresponding spinous processes, with the presence of fluid collection.

rotation, or flexion-extension. They have a high out-of-hospital mortality rate. In atlanto-axial dislocations, the most recurrent symptoms are weakness or paresthesia of the extremities, affecting up to 70% of patients, and posterior cervical pain or a decreased range of cervical motion, affecting up to 50%.[7] Diagnosis starts with the investigation of the entire spine. The computed tomography trauma series, including imaging of the craniocervical transition, associated with a detailed analysis using the standard anatomical reference measurements described in the literature, are essential for diagnosis and timely management of these injuries.[8,9] It is not uncommon for the diagnosis of craniocervical or atlanto-axial dislocation to be delayed or not realized, as a high index of suspicion and a protocol for the methodological visualization of radiological images are essential. Magnetic resonance imaging of the cervical spine can help in the diagnosis of high cervical region ligament injuries, possibly showing rupture of the alar, apical, or transverse ligament of the atlas. Evidence of free fluid in the joint and of ischemic signs in the nerve tissue may support the suspicion and diagnosis of injuries. Vertical dissociations, which can be caused by strong distracting forces when the cervical spine is bent, can damage multiple structures, such as the tectorial membrane, the longitudinal ligaments, facet joints, and even the cervical musculature, which are liable to increase the C1-C2 distance as seen in our patient's images. In a case cohort study by Meyer et al.,[10] all the distraction injuries in a series of three cases had vertebral artery lesions.

Craniocervical dissociations are generally caused by high-energy blunt trauma and were classified by Traynelis et al. into 3 types: Type I, anterior displacement of the occiput in relation to the atlas; Type II, dislocation of the occiput in relation to the atlas; and Type III, posterior dislocation of the occiput in relation to the atlas.[6] In 1977, Fielding classified atlanto-occipital dislocation into four categories. This classification is based on imaging to evaluate the direction

of the atlanto-axial displacement in relation to the central axis. In Fielding's classification, Type I is a rotatory dislocation without anterior displacement of the atlas. The odontoid acts as a pivot and the transverse and alar ligaments are intact. This is the most commonly occurring type within the normal range of rotation of the joint. Type II is a rotatory dislocation with anterior displacement of the atlas of from 3 to 5 mm, in which one of the joint masses acts as a pivot. It is associated with a deficiency in the transverse ligament. Type III is a rotatory dislocation with anterior displacement of the atlas greater than 5 mm. This degree of displacement implies a deficiency in the transverse and alar ligaments. Type IV is a rotatory dislocation with posterior displacement of the atlas. This type is rare and severe.[4]

The locked-in syndrome is characterized by preserved cognitive ability, the ability to move the eyelids, and vertical eye movement, associated with paralysis of all other voluntary muscles. It occurs due to an injury of the white matter along the pons resulting from factors such as infarction, bleeding, or trauma. Despite advances in patient management, the mortality rate is still high, as are the psychosocial complications of the disease. Some studies suggest a 10-year survival of up to 80% and the ability to resume social life at home with early rehabilitation care.

To our knowledge, after a review of the PUBMED, SciELO, and Scopus databases, only one case of craniocervical dissociation associated with atlanto-occipital subluxation and two cases of locked-in syndrome following traumatic cervical dislocation were described.[11–13] Desai et al. described the possibility of shear injury due to cervical hyperextension, resulting in subluxation and posterior axonal injury in the spinal cord. Brain contusions are less uncommon causes of locked-in syndrome, however due to circuitous path and high possibility of injury to the vertebral arteries in cases of craniocervical dislocation, insufficient blood flow to the pons can occur, causing the locked-in syndrome.[14]

CONCLUSION

Craniocervical injury must be strongly suspected in high-energy traumas and the urgent and adequate evaluation of imaging studies is essential. In this way, more timely and accurate diagnoses can benefit the patient.

All authors declare no potential conflict of interest related to this article.

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