Atlanto-occipital fusion and its neurological complications: a case report

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Abstract

During routine activities in the Laboratory of Human Anatomy at the University of Santa Cruz do Sul – Brazil, an atlanto-occipital fusion was observed in a Caucasian cadaver skull. The skull used in our study had complete fusion of the occipital bone with the atlas vertebra, except in traffic areas of the vertebral arteries. Some important neurological disorders seem to be related with atlanto-occipital fusion. The presence of other anatomical variations was not verified. Thus, the present study shall be important for health sciences and those who keep some interest in pathologies associated with brain.

Keywords: atlanto-occipital fusion, vertebra, neurological disorders, anatomy.

1 Introduction

The atlanto-occipital joint on each side is that located between the superior articular facet of the lateral mass of atlas and the occipital condyle corresponding. These two bones are also connected by atlanto-occipital membrane and back, extending from the anterior and posterior arches of the atlas, respectively, to anterior and posterior edges of the foramen magnum. The facet of the occipital condyle is usually reniform or hourglass-shaped, occasionally split (GARDNER, GRAY and O'RAHILLY, 1988). The atlanto-occipital joint is synovial type and has joint capsule thin and loose. The main movement of this joint is of bending, with a slight lateral tilt and rotation of the head (MOORE, DALLEY and AGUR, 2011).

Atlanto-occipital fusion of the atlas is an important congenital malformation of the craniovertebral region because of the proximity to the spinomedullary region (AL-MOTABAGANI and SURENDRA, 2006). Furthermore, the atlanto-occipital fusion is that not all cases can be easily distinguished from the Arnold-Chiari malformation (KASSIM, LATIFF, DAS et al., 2010).

The morphological aspects of atlanto-occipital fusion in humans as well as their neurological complications are described in this study.

2 Case report

In a Caucasian cadaver skull, with approximately 40 years old, belonging to the didactical collection of the Laboratory of Human Anatomy from University of Santa Cruz do Sul - Brazil, was observed an atlanto-occipital fusion during routine activities (Figure 1). All measurements performed in our study were taken with a digital pachymeter from Digimess®. The skull used in our study had complete fusion of the occipital bone with the atlas vertebra, except in traffic areas of the vertebral arteries. The diameters of the channels formed by the passage of the vertebral arteries were of 0.25 and 0.31 mm on the right and left, respectively. The posterior arch of the atlas vertebra showed bifid format (see Figure 1). Thus was formed a left and right posterior arch length of 0.91 and 1.10 mm, respectively. Another important aspect was the absence of foramen of the transverse process on both sides of the atlas vertebra (see expanded field of Figure 1). We did not observe any apparent deformity of the skull examined.

3 Discussion

According to Al-Motabagani and Surendra (2006), atlanto-occipital fusion was first described by Rokitansky in 1844 and demonstrated roentgenographically by Schuller in 1911. The same authors also reported that the incidence of atlanto-occipital fusion varies from 0.5-1.0% in Caucasians. Furthermore, Murlimanju, Prabhu, Paul et al. (2010) in a recent study showed that the occipital bone may fail in some centers of ossification.

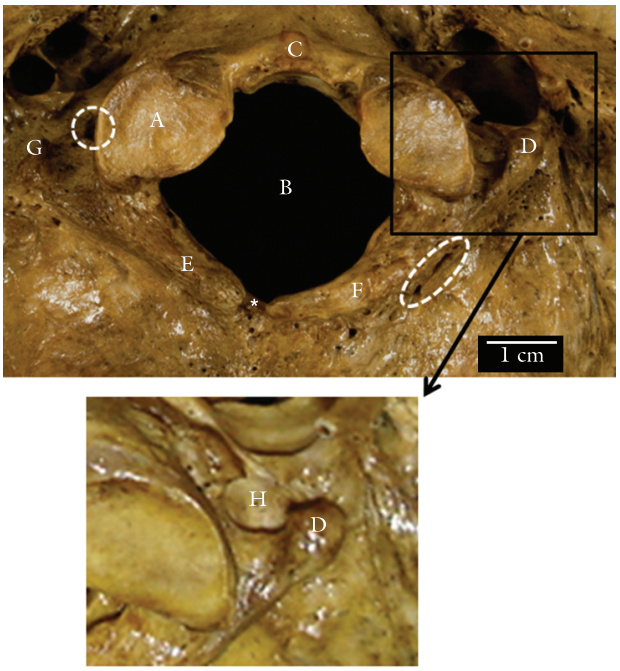

Figure 1. Inferior view of the skull. a) inferior articular facets, b) foramen magnum, c) anterior tubercle, d) left transverse process, e) right posterior arch, f) left posterior arch, g) right transverse process. *Bifid posterior arch. The dotted circles correspond to the channel formed by the entry of the vertebral arteries. Note at the expanded field that the transverse process foramen appears as a notch (H) and not a true foramen.

Kassim, Latiff, Das et al. (2010) commented that not all cases of atlanto-occipital fusion can be easily distinguished as Arnold-Chiari malformation as the pathophysiology of both are similar. According to Moore and Persaud (2004), the Arnold-Chiari syndrome is the most common congenital anomalies involving the brain. This syndrome is defined as a structural defect of the central nervous system in which portions of the cerebellum are located below the foramen magnum. The anomaly results in a type of hydrocephalus in which it interferes with the absorption of cerebrospinal fluid, consequently the entire ventricular system becomes strained. The frequency of appearance of Arnold-Chiari malformation is one in 1.000 births and is commonly associated with spina bifida with meningomyelocele, spina bifida with myeloschisis and hydrocephalus.

Graham, Davis, Gouvernayre et al. (2011) described four kinds of Arnold-Chiari malformation. Of the four types of Arnold-Chiari malformation, emergency physicians are most likely to encounter Type I. Arnold-Chiari I malformation is typically caused by congenital anomalies of the posterior skull fossa. In addition, this kind of malformation may be acquired later in life secondary to the development of brain or spinal cord processes. Regardless of cause, Arnold-Chiari malformation may result in downward pressure on the cerebellum or brainstem, with subsequent blockade of cerebrospinal fluid flow. This can lead to a range of clinical findings including cephalalgia, vertigo, weakness, numbness, visual disturbance, ataxia, or deficiencies in coordination. The most common variety may remain asymptomatic throughout life, and is frequently discovered initially as an incidental anatomic abnormality seen on cerebral imaging. Congenital Arnold-Chiari I malformation is most frequently asymptomatic, but may be encountered in the emergency department setting due to an exacerbation of subacute or chronic Arnold-Chiari related symptoms.

Kagawa, Jinnai, Matsumoto et al. (2006) reported a case of Arnold-Chiari I malformation accompanied by assimilation of the atlas, Klippel-Feil syndrome, and syringomyelia. X-ray images and CT scans demonstrated assimilation of the atlas to the occipital bone, C2 and C3 fusion, abnormal passage of the vertebral arteries, and an anomalous bony mass on the right lateral mass of the atlas protruding into the spinal column. The odontoid process was also deviated to the left. Magnetic resonance images demonstrated bilateral descent of the cerebellar tonsils and syringomyelia extending from C6 to T8.

In a case report of the an atlanto-occipital fusion in a Japanese Brown calf, Morimoto, Tsuda and Miyamoto (2001) discussed that the atlanto-occiptital fusion is a developmental disorder due to abnormal segmentation and development of the caudal occipital and cranial cervical sclerotomes.

Abumi, Avadhani, Manu et al. (2010) described that the occipitocervical fusion is formed primarily in instability due to congenital and post-traumatic deformities as well as infections, tumors and inflammatory conditions at the craniocervical junction.

El Abd, Rosenberg, Gomba et al. (2008) studied the lateral atlanto-axial joint as a source of headache in congenital atlanto-occipital fusion. They found that a 47 year old woman presented with severe right sided neck pain and headache, predominantly in the right occipital region, for 3 years. The patient's examination was unremarkable except for reduced neck motion and prominent right occipital tenderness.

Al-Motabagani and Surendra (2006) performed a study with a sample of 109 adult skulls of Asian origin examined for evidence of atlanto-occipital fusion. They found that only one specimen that had this type of anomaly, that was totally synostosed atlas with the occipital bone with multiple bony defects.

Recently, Kassim, Latiff, Das et al. (2010) mentioned that it should be noted that the vertebral artery is an important blood vessel related superiorly to the atlas vertebra and any compression of the artery may compromise the blood flow to the brain.

Wang, Wang, Liu et al. (2009) researched the anomalous vertebral artery in craniovertebral junction with atlanto-occipital fusion. They classified four different pathways of the vertebral artery at the cranio vertebral junction with atlanto-occipital fusion were found: Type I, wherein the vertebral artery enters the spinal canal below the C1 posterior arch, and the course of the vertebral artery is below the atlanto-occipital fusion C1 lateral mass; Type II, the vertebral artery enters the spinal canal below the C1 posterior arch, and the course of the vertebral artery is on the posterior surface of the atlanto-occipital fusion C1 lateral mass, or makes a curve on it; Type III, wherein the vertebral artery ascends externally laterally after leaving the axis transverse foramen, enters an osseous foramen created between the atlas and occipital bone, then into the cranium; and Type IV, in which the vertebral artery is absent.

In domestic mammals, anomalies of the cranio vertebral joint are referred to as occipitoatlantoaxial malformations. Such malformations involve different degrees ofsymmetrical or asymmetrical, unilateral or bilateral fusionof the atlas to the skull (SEVA, GÓMEZ, PALLARÉS et al., 2008). The differential diagnosis of congenital malformation includes fetal infection with a teratogenic virus [Akabane disease virus, Cache Valley virus, Pestivirus and Orbivirus], ingestion of certain toxic plants [*Astragalus* spp., *Oxytropis*, *Conium maculatum*, *Nicotiana glauca*, *Lupinus formosus* and *Veratrum californicum*] and vitamin D imbalance [high levels of vitamin D have led to marked ankylosis and scoliosis of thecervical vertebrae along with exostosis of the atlantooccipital and atlantoaxial joints] (SCHMIDT, FORSYTHE, COWGILL et al., 1993).

According to Al-Motabagani and Surendra (2006) the congenital malformations at the cervicooccipital region are, in general, of considerable consequence because of their proximity to the spin medullary, region with the possibility of neurological compression syndrome. The occipitalization of the atlas, in particular, can produce a wide range of neurological signs and symptoms, which vary from a transitory headache to a full-blown neurological syndrome.

We undertook this study with the aim of providing a more accurate report about the atlanto-occipital fusion, because of its interesting relationships with neuropathologies in adjacent structures. Finally, this study is useful for academics, clinicians and surgeons who handle and have special interest in anatomical structures discussed in this work.

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Received August 31, 2011
Accepted June 5, 2012