Anatomical changes in occipitalization: is there an increased risk during the standard posterior approach?

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Abstract

Purpose The purpose of this study was to examine the anatomic changes in a case of occipitalization of the atlas.

Methods Occipitalization of the atlas was found accidentally in a 64-year-old male cadaver. Anatomic dissection was carried out to examine the posterior aspect of the upper cervical region and craniocervical junction. The occipitalized atlas was then harvested and macerated to study the bony anomaly.

Results In this case of occipitalization, fusion was observed at both lateral masses and at the left posterior hemiarch of the atlas. We found the following soft tissue changes: the rectus capitis posterior minor muscle was lacking on the left side and was atrophic on the right, the obliquus capitis superior muscle was present on both sides showing moderate atrophy and fatty changes. The posterior atlanto-axial membrane was thinner and asymmetric, had a free edge on the right side. Lateral to this edge the dura was lying free. We believe that these changes of the posterior atlanto-axial membrane together with the increased distance between the fused posterior arch of the atlas and the lamina of the axis could cause the observed “dura bulge”

at this level. The vertebral artery was entering the skull through a canal on the left side.

Conclusions In our case, occipitalization considerably changed the anatomy of the upper cervical spine and craniocervical junction. Special care must be taken when using the posterior approach to avoid neurovascular injury in cases with occipitalization.

Keywords Occipitalization of the atlas · Craniovertebral junction · Developmental anomaly · Upper cervical spine

Introduction

Occipitalization of the atlas is a developmental anomaly in which part or the complete atlas is fused to the occiput. Developmental background of occipitalization is a failure of segmentation between the fourth occipital sclerotome (proatlas) and the first cervical sclerotome during embryonic development [1]. Occipitalization is the most common anomaly of the craniocervical junction with a prevalence of 0.08–2.76 % in the population [2]. It could be associated with atlanto-axial instability or basilar invagination resulting in compression of the medullary cord. Symptoms may vary from deformity with a cosmetic concern, through pain, to varying degrees of neurological dysfunction. Surgery, i.e. occipito-cervical fusion with or without decompression, may be indicated in case of severe symptoms, especially in case of neurologic deterioration [3]. As several components apart from the skeleton of the proatlas may be involved in the malformation, any surgical intervention carries an increased risk of injuring the dura, neural or vascular structures. Special attention must be paid to map out the course of the vertebral artery before surgery, therefore a preoperative angiogram is strongly recommended. Because

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of the rare prevalence of occipitalization of the atlas, the anatomical changes associated with this condition have not yet been fully explored although were already mentioned by Morgagni in 1762 [4]. We report an anatomic study on a case with occipitalization of the atlas where we focused our attention on the associated soft tissue aberrations and correlated this to bony anomaly. To our knowledge, this is the first anatomical study carried out on a fresh cadaver, where the anatomy of the soft tissues and also the bony anatomy of the macerated specimen could be studied.

Materials and methods

Occipitalization of the atlas was incidentally found in a cadaver of a 64-year-old man during autopsy. An anatomic dissection was carried out using surgical loupe magnification to examine the structures at the posterior aspect of the craniocervical junction and upper cervical spine. A detailed photo-documentation was carried out. After dissection, the occiput with the atlas was harvested and macerated to allow a meticulous study of the bony relations. Pictures were made showing the bony anomaly. All measurements were done using a caliper and rounded off to the nearest millimeter or degree. The findings were also compared with some of our clinical cases.

Before his death, the 64-year-old male patient was diagnosed and treated at a neurology department with supranuclear palsy and multiple cerebellar infarcts. His secondary diagnoses consisted of high blood pressure, atherosclerosis, chronic pancreatitis, pancreatogenic diabetes, polyneuropathia diabetica and dementia. According to the patient's anamnesis, he had suffered from gait ataxia in the last 5 years. Cervical MRI was not carried out to rule out cervical myelopathy.

The bony specimen is owned by the Museum of the 2nd Department of Pathology at the Semmelweis University in Budapest.

Results

Soft tissue dissection

The anatomic dissection was done on the posterior aspect of the upper cervical spine. To perform this, we removed the skin and prepared the subsequent layers of the posterior cervical musculature. The superior layers of this musculature showed no deviation from normal in size, expansion or symmetry of the muscle groups. At the level of the suboccipital neck muscles, we found the following variations: the rectus capitis posterior minor muscle was missing on the left side (it was replaced by a matrix of connective

and fatty tissue) and was atrophic on the right side (it originated below the inferior nuchal line and was attached to a tubercle formed on the tip of the right semiarch). The rectus capitis posterior major muscle showed no changes. The obliquus capitis superior muscle was present on both sides but showed moderate atrophy and fatty changes while the obliquus capitis inferior muscles were showing hypertrophy when compared with normal subjects.

We removed the above-mentioned muscles to examine the posterior aspect of the upper cervical spine (Fig. 1). On the left side, we found a rudimentary posterior hemiarch of the atlas fused to the occiput. Posterior ramus of the C1 nerve was passing dorsal between occiput and the hemiarch. The hemiarch on the right side was not fused to the occiput. Its posterior tubercle approached but did not reach the posterior midline. It served as an attachment for the right m. rectus capitis minor. The two hemiarches were bridged by fibrous connective tissue, which macroscopically did not contain cartilage. The posterior atlanto-axial membrane seemed to be thinner and asymmetric. It was present on the left side, but on the right side it formed a free edge running obliquely from the right hemiarch of the atlas to almost the middle of the lamina of the axis. Lateral to this free edge, the dura was lying free.

The course of the vertebral artery was normal on both sides, running cranially after exiting the C2 transverse process to enter the C1 transverse process. The vertebral artery was covered with a layer of connective tissue above the C1 hemiarches. On the left side, it entered the skull through a tunnel formed by the occiput and the fused hemiarch of the atlas (Fig. 3b), whereas on the right side it

Fig. 1 Posterior aspect of the upper cervical spine is demonstrated from dorsal. The tips of the two hemiarches of the atlas are connected by a bridge of connective tissue (*). Between the hemiarches of the atlas and the lamina of the axis spans the posterior atlanto-axial membrane forming a free edge on the right side (arrow). The C2 nerve root, its ganglion, ventral and dorsal ramuses and the vertebral artery (arrowheads) show no deviation from normal. The obliquus capitis superior muscle was preserved on both sides

had a normal course running on top of the hemiarch in the groove of the vertebral artery entering the skull the usual way.

Bony anatomy after maceration

The macerated bone specimen showed the following bony alterations. The atlas showed an asymmetric fusion to the occiput. The midsagittal axis of the atlas was shifted to the right about 5 mm according to the midsagittal axis of the occiput determined by the basion-opisthion axis. The atlas was slightly rotated to the right (about 2° – 3°) (Fig. 2c). The superior articular processes of the atlas were forming a solid mass with the occipital condyles on both sides. Articular gap was not visible. The anterior arch of the atlas was well developed and had a marked anterior tubercle. The superior aspect of the anterior arch together with the anterior margin of the foramen magnum formed an irregular anterior atlanto-occipital foramen, limited by the fusion masses on the sides. The diameter of this opening was about 2–4 mm vertically and it was 18 mm horizontally (Fig. 2b). The articular facet for the odontoid was pronounced, and showed slight degenerative changes (osteophyte). The transverse processes were well developed, bearing a foramen transversarium for the vertebral artery on both sides. The posterior arch of the atlas was not formed; instead, there were two hemiarches (Fig. 2a). The left hemiarch was completely fused to the margin of the

foramen magnum between the canal for the vertebral artery and the tip of the hemiarch (Fig. 3b). This canal where the left vertebral artery entered the skull was formed by the posterior arch of the atlas caudally, the occiput cranially and the two fusion masses, the fused mass of the atlanto-occipital articulation ventrally and the margin of the fused mass of the left hemiarch dorsally. The canal was 16-mm long with a diameter of 4 mm. The tip of the fused left hemiarch approached the midline to 3 mm defined by the basion-opisthion axis. The right hemiarch of the atlas was not fused to the occiput, although bony spurs protruding towards the hemiarch were visible on the margin of the foramen magnum (Fig. 3a). The distance between the arch and the margin of the occiput was only 2 mm. The right hemiarch had a sulcus for the vertebral artery, which entered the skull the usual way. The tip of the right hemiarch approximated the midline to 5 mm. The distance between the tips of the two hemiarches was 8 mm. The hypoglossal canals were normal in position and appearance on both sides. The foramen magnum was distorted, oval shaped and narrowed by the left fused hemiarch (Fig. 2d).

Discussion

Realdo Colombo (1516–1559), a student of Vesalius was the first who mentioned the occipitalization of the atlas. The earliest discussion of the bony anomaly and the

Fig. 2 Photographs demonstrating the occipitalized atlas vertebra from different aspects. Macerated bone specimen. **a** posterior, **b** anterior, **c** caudal view, demonstrating the occipitalized atlas and

the foramen magnum, **d** “internal view”, demonstrating the foramen magnum from cranial

Fig. 3 Pictures **a** and **b** illustrate the *right* and *left* side of the bony specimen. On the *right* side (picture **a**), the vertebral artery entered the skull the usual way. Picture **b** demonstrates the canal formed between the occiput and atlas through which the vertebral artery enters the skull

accompanying changes of the muscles date back to 1762 by Morgagni. There were numerous theories explaining the formation of this deformity. In 1862, Luschka believed the occipitalization of the atlas to be similar to the sacralisation of the 5th lumbar vertebra. Schiffner in 1878 considered intrauterine forces to be responsible for the “ankylosis of the upper cervical articulation”. In the following years, anatomists and pathologists showed growing interest in this condition. The next hundred years it was disputed if occipitalization of the atlas was a sign of phylogenetic “future form” or only an occasional variant. [4]. An anatomic study was done by Ingelmark in 1947 and a clinical-radiologic study by McRae and Barnum in 1953. The latter authors used the term “occipitalization of the atlas” first [5, 6].

Today, we know that occipitalization of the atlas is caused by failure of segmentation between the fourth occipital sclerotome (proatlas) and the first cervical sclerotome during embryonic development [1]. The deformity represents the most common anomaly of the cranio-vertebral junction. Its reported frequency shows great deviations.

It is 0.14 % according to Macalister from 1892 while Stiemens in 1929 found it to be 0.25 % (he examined 56,000 human skulls from a collection) [5]. Later Menezes [7] also found the incidence of occipitalization to be 0.25 % examining 5,300 patients. Occipitalization occurs mostly in patients with some sort of a congenital syndrome (Klippel-Feil, 22q11.2 deletion or Goldenhar syndrome). However, patients with occipitalization—without any congenital syndrome—show similar clinical characteristics to Klippel-Feil syndrome, i.e. low posterior hairline, short neck, torticollis and restricted neck movements. Associated congenital abnormalities are present in 20 % of the cases. These include anomalies of the jaw, incomplete cleft of the nasal cartilage, cleft palate, congenital deformities of the external ear, cervical ribs, hypospadias and urinary tract anomalies. Most patients with occipitalization of the atlas probably remain asymptomatic and undetected. For those that develop symptoms the onset is usually in the third or fourth decade. Common complaints are neck pain, stiffness and varying degree of myelopathy [3].

Numerous authors reported surgical techniques to treat atlanto-axial dislocation in association with occipitalization of the atlas. Jain [8] reported 50 cases in 1993, he formed an “artificial atlas” by drilling a groove on the occiput and used wire fixation between the occiput and the C2 lamina. Atul Goel [9] reported eight cases in 2004; he used posterior C1–C2 fusion using a plate and screw construct. According to Brockmeyer, surgical intervention is indicated in cases of atlanto-axial subluxation in case of myelopathy, neural compression, C1–C2 subluxation with an anterior atlanto-dental interval greater than 6 mm and progressive C1–C2 subluxation. If neural compression can be reduced with traction or positioning, posterior C1–C2 fusion is proposed. In cases of nonreducible compression, occipito-cervical fusion is carried out after adequate decompression with transoral odontoidectomy (in case of ventral compression) or posterior decompression (in case of posterior compression) [10].

During the preparation, we made the following observations that may help surgeons to avoid intraoperative complications in a similar case.

In our case of occipitalization, atrophy of the rectus capitis posterior minor muscle was observed on the right side, while it was replaced by a matrix of connective and fatty tissue on the left side. In this case, there would be less muscle bulk covering the posterior surface of the atlanto-occipital region leaving the interval between the lamina of the atlas and axis vulnerable during the subperiosteal dissection.

During the anatomic dissection, the fused arch of the atlas was found about 1.5-cm deeper than the level of the C2 spinous process. In our case, the right hemiarch had a tubercle, which could erroneously be identified as the

posterior tubercle of the atlas, except that it was not lying in the midline. In this case, the right hemiarch did not reach the midline, between the tips of the hemiarches was a band of connective tissue, which could be confused with the nuchal ligament. Care should be taken not to penetrate between the fused lamina of the atlas and the lamina of the axis either with finger or with an instrument while trying to find the posterior tubercle.

Due to the fusion of the atlas to the occiput, the distance between the lamina of the atlas and axis is increased. Between the two laminae, the dura is covered only by the posterior atlanto-axial membrane. In this case, the posterior atlanto-axial membrane was asymmetrical. Because of this asymmetry, the dura was lying free on the right side. We searched our database of CT-Myelograms and MRIs of the upper cervical spine and found that in cases with occipitalization, there is sometimes a considerable “dura bulge” between the posterior arch of the fused atlas and the lamina of the axis. This was best visible on mid-sagittal images. (Small amount of “dura bulge” is also present in normal cases between C1–C2.) We found this increased expansion also on the axial pictures. This “dura bulge” could lead to unwanted opening of the dura during the muscle dissection from C1 or C2. We believe that the reason for this “dura bulge” is the increased distance between the two laminae and also the thin- or missing-posterior atlanto-axial membrane. Similar dura bulge is visible on postoperative MRIs of the cervical spine after laminectomy.

In our case, the vertebral arteries had a normal course on both sides, exiting the C2 transverse foramina and running through the C1 transverse foramina. According to Shengling Wang et al., anomalies of the vertebral artery occur more often in cases with occipitalization. In his 36 cases of occipitalization, he found the vertebral artery to enter the spinal canal below the occipitalized posterior arch in 33.3 % of the cases without running through the transverse foramen of the atlas [2]. This variant is analogous to the “C2 segmental type of vertebral artery” described by Tokuda [11].

Although in our case the hypoglossal canals showed no aberrations, changes in shape and size of the hypoglossal canal were described by Gordon Harrower in 1923. He described nine human skulls from a collection of the Anatomical Museum of Glasgow University showing congenital anomalies, occipitalization or an occipital

vertebra. Among these he found five anomalous hypoglossal canals, most of them showing minor variations. Only one of these cases showed marked deformity of the hypoglossal canal in a skull with an occipital vertebra [12].

Conclusion

Occipitalization of the atlas considerably changes the anatomy of the upper cervical spine and craniocervical junction. Preoperative diagnostic using CT angiography is of paramount importance to detect these changes and avoid neurovascular injury during surgery.

Conflict of interest None.

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