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THE ATLANTO-AXIAL SYNOSTOSIS — A CASE OF VERTEBRAL ANOMALY

Abstract: *The atlanto-axial synostosis — a case of vertebral anomaly*
The subject of this analysis are the first two cervical vertebrae (the atlas and the axis), fused together as consequence of a pathological process. A detailed analysis of the specimen revealed a synostosis which existed between the lateral facets of the atlanto-axial joint. Hence, a fusion between the anterior arch of the atlas and the dens of the axis, and an incomplete ossification of the yellow ligaments was observed. The dimensions of the fused vertebrae, except for the length of the C2 vertebral foramen, remain within the normal range of variation for an adult male. Morphological appearance of the specimen allowed to exclude the congenital nature of the synostosis. Therefore we attribute a post-traumatic etiology to the studied anomaly.

Key words: Atlanto-axial fusion, Cervical vertebra, Facet joints

INTRODUCTION

The atlanto-axial joint is the articulation which provides mobility between the first (atlas) and the second (axis) cervical vertebra [1, 2]. Formation of the synostosis within the atlanto-axial joint would mainly inhibit rotatory movements of the head. The other vertebral joints can hardly compensated its function [3, 4]. Synostosis within the vertebral column may occur on any level. However, synostosis between the C1 and C2 seems to be an uncommon case and is rarely described in literature.

The objective of this study was to present the morphological appearance of the atlanto-axial synostosis, and consider the clinical implications of its etiopathology.

MATERIAL AND METHOD

The osseous specimen which shows fusion between the first two cervical vertebrae (C1 — atlas and C2 — axis) was the subject of this morphological analysis. The vertebrae were in a very good condition, and did not suffer from postmortem damage. This permitted to perform a precise evaluation of their morphology, and to describe the anatomy of the fused atlanto-axial joint. The specimen is housed in the Department of Anatomy, Jagiellonian University Medical College. Unfortunately, we are not able to attribute any ethnic origin to the investigated sample. There are also no convincing data which would allow to determine the gender and age at death of the individual who was afflicted with the atlanto-axial synostoses. Only by comparing the diameters of the fused vertebrae to literature data, we can hypothesize that the specimens comes from a male [5]. We deem that the fused vertebra derives from an individual of about 60 years of age. This is because of the visible notches on the medial border of the joint surfaces, which, with age, are prone to atrophy [6]. During anatomical evaluation, we paid particular attention to the size of the articular facets, their mutual orientation, and angle that they made with the sagittal plane. The block of fused vertebrae was subjected to conventional measurements, which definitions are presented in Table 1. Moreover, two additional measurements characterizing morphology of the atlanto-axial synostosis were performed:

— lateral rotation C1–C2 — measured as a lateral displacement of the posterior tubercle of C1 towards the apex of the C2 spinous process;
— length of the callus between the anterior surface of the dens and the posterior surface of the C1 arch.

Parameters, which were measured on the atlas and the axis, were defined in Table 1. Their values were compared to the normative literature dimensions of the atlas and axis [7]. The linear measurements were taken using a digital caliper (accuracy 0.1 mm). The photographic documentation was obtained with a digital camera (Canon EOS 5D) equipped with the Tamron Macro 1 : 1 lens.

Table 1

Definitions of measurements performed on the atlas and the axis

Measurement	Atlas (C1)	Axis (C2)
Length of the vertebra	Distance between the anterior tubercle and the posterior tubercle, respectively on the anterior and posterior arch.	Distance between the antrior-most point of the vertebral body and the end of the spinous process.
Width of the vertebra	Distance between both tips of the transverse process.	
Length of the vertebral foramen	Distance between the posterior surface of the anterior arch and the anterior surface of the posterior arch.	Distance between the posterior surface of the vertebral body and the anterior surface of the arch.
Width of the vertebral foramen	Maximum transverse diameter within the foramen.	

Measurement	Atlas (C1)	Axis (C2)
Length of the superior articular facets of the C1	Distance between the most posterior point of the articular facet of the fovea dentis and the most posterior point of the fovea dentis.	
Width of the superior articular facets of the C1	Distance between the most medial point of the articular facet of the fovea dentis and the most lateral point of the fovea dentis.	
Height of the dens	Distance from the apex of the dens to the horizontal plane, which passed along the superior edge of the superior articular facets.	
Width of the dens	Maximum transverse diameter of the dens.	

RESULTS

Visual inspection of the vertebral specimen revealed total fusion of the atlanto-axial joint. A complete synostosis was visible between the lateral facets of the atlas and axis, and between the dens of the axis and the anterior arch of the atlas (Fig. 1, Fig. 2). Distortion of the atlas towards the axis, and right side inclination in respect to the horizontal plane are remarkable characters of the specimen. This abnormal position caused also displacement of the transverse processes of both vertebrae. Due to this fact the foramina in the transverse processes do not precisely coincide with each other. The size and shape of the transverse foramina are both normal in the atlas and the axis. Nevertheless, the normal passage of the vertebral vessels was altered within the atlanto-axial segment of the spine.

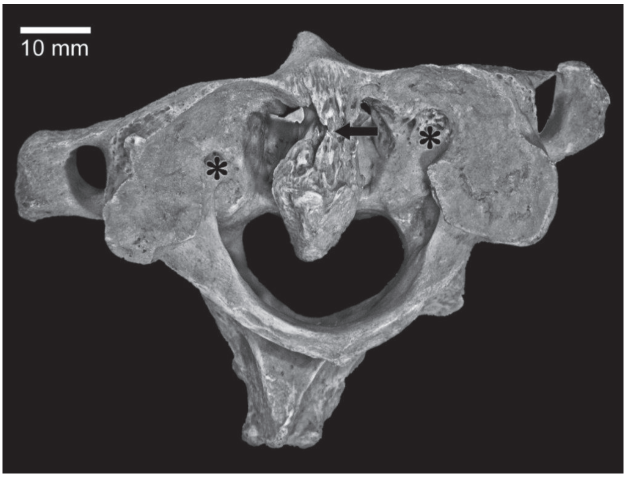

Fig. 1. Superior view of the atlas and axis which is abnormally rotated. Visible bilateral notches located medially on the superior articular facets (indicated by asterisks). Visible fusion of the dens (C2) with the anterior arch of the C1 (indicated by arrow)

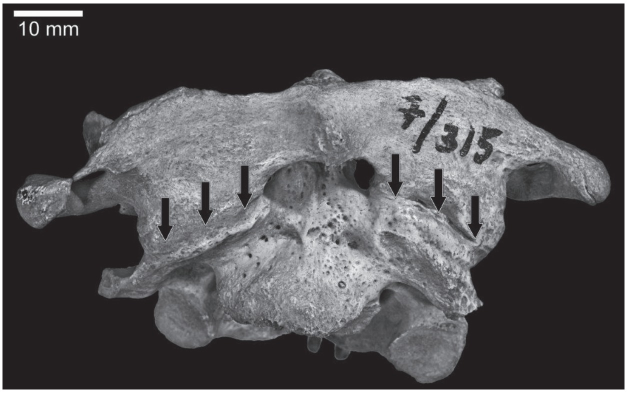

Fig. 2. Anterior view of the atlas and axis with visible synosthosis between the lateral facets (indicated by arrows)

Overall, anatomy of each individual vertebrae remained basically normal. Most of the parameters of the atlas and axis did not exceed the normal range of variation. Only the length of the vertebral foramen of the axis seemed to be smaller than normal (Table 2). Hence, some parts of the atlas and axis show defects. The lower part of the body of the axis is obliquely deformed and protruded. This was proably dictated by the loss of mobility within the atlanto-axial joint, which was compensated by abnormal motion of the rest of the cervical spine.

Table 2

Measurements of the fused atlas and axis with range of normal variation [8]

Measurement	Value [mm]	Min–Max [mm]
Length of the atlas	48.1	42–54
Length of the axis	52.0	46–64
Width of the atlas	83.8	70–95
Width of the axis	54.5	49–64
Width of the vertebral foramen (C1)	31.0	27–35
Width of the vertebral foramen (C2)	22.5	21–28
Length of the vertebral foramen (C1)	33.8	28–40
Length of the vertebral foramen (C2)	15.1	18–27
Length of the superior facet (C1)	31.8 (R), 33.8 (L)	17–31
Width of the superior facet (C1)	17.4 (R), 16.8 (L)	10–16
Height of the dens	15.3	14–22
Width of the dens	12.9	9–13

R — right side, L — left side

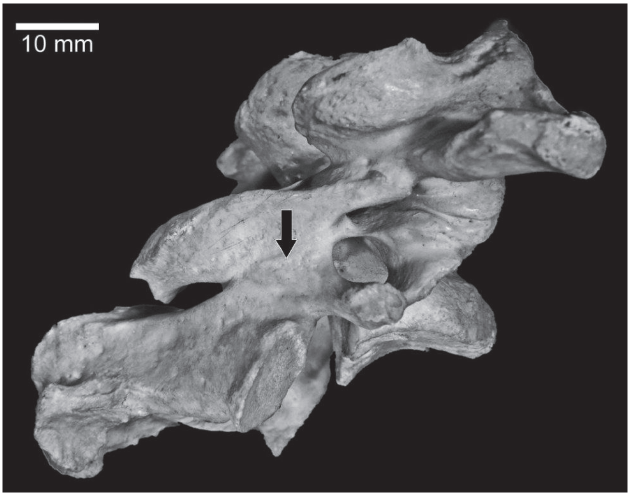

Fig. 3. Lateral view of the atlas and axis. Note the ossified yellow ligament (indicated by arrow)

Particular attention should be paid to the shape and size of the superior articular facets of the C1 vertebra. These facets are flat plates with an uneven, rough surface and numerous bone tissue losses visible on the medial aspect of the left and right articular surface. It should be also noted that the transverse and the sagittal diameters of the superior articular facets of the atlas slightly exceed upper normative value.

The other important feature of the specimen is a partial fusion between the right side of the posterior arch of C1 and the arch of C2. This is most likely the ossified yellow ligament, which extends on the distance of 13.7 mm (Fig. 3).

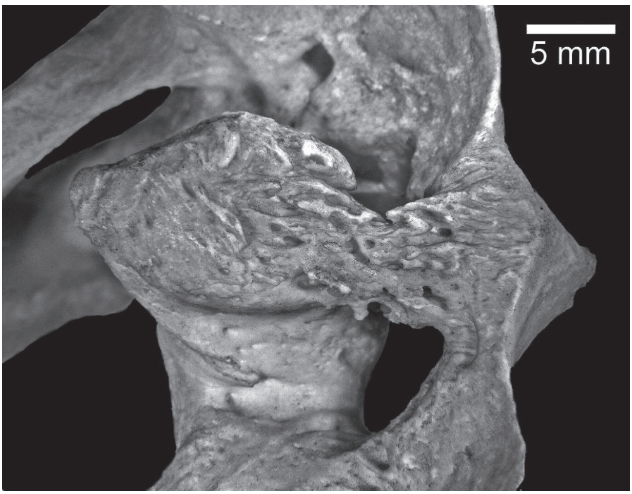

Fig. 4. Close-up view of the synostosis between the dens and the anterior arch of the atlas

Synostosis between the dens of the C2 and the anterior arch of the C1 shows characteristic jagged form which comprises the median atlanto-axial joint. This manifests as a conglomerate of thick bony trabecules which vary in length between 5.0–12.0 mm (Fig. 4). The posterior surface of the dens is remarkably thickened. This may be caused by calcification and ossification of the transverse ligament of the dens.

DISCUSSION

Presented specimen of the atlanto-axial synostosis is a fine example of cervical spine malformation, which is rarely reported in the literature. Only few similar cases termed as the atlanto-axial fusion or C1–C2 block vertebra were recognized in children and adults [9–13]. Cave [9] described three types of the atlanto-axial fusion:

1. Fusion of the separated dens with the ventral atlantal arch.
2. Complete (bilateral) fusion of the atlas and the axis, with or without assimilation of the first vertebra by the second.
3. Incomplete (unilateral) fusion, one-half of the atlas retaining its independence, with or without some degree of assimilation.

According to this classification, presented fusion of the first two cervical vertebra should be included in the second group, because of total ossification that occurred between both lateral and medial atlanto-axial joints.

The specimen at our disposal did not represent a reliable basis to draw conclusions about the time, mechanism and reason of atlanto-axial synostosis formation. Certainly, bilateral obliteration of the atlanto-axial joints results in the absolute immobility of that joint. Extensive fusion between the atlas and the axis significantly influenced the vertebral biomechanics of the neck. The primary effect of the atlanto-axial synostosis was that head rotation, within the atlanto-axial joint, was seriously limited. Rotation of the head occurs almost entirely at the joint between the atlas and the axis. The vertebral column itself contributes only slightly to head rotation.

That lack of mobility was probably associated with heterotropic ossification, which occurred within the articular capsules of the intervertebral joints, the atlanto-dental joint, and the partially afflicted right yellow ligament. The whole process of complete pathological ossification may result from heavy trauma of the craniovertebral region. Therefore, we presume that fusion of the atlanto-axial joint was of acquired rather than congenital nature. A similar case was reported by Holck who analyzed a skeleton with traumatic whiplash neck injury [14]. That individual had broken the dens of the axis, and fused it with the arch of the atlas.

An injury-type etiology of the atlanto-axial synostosis can be supported by the rotatory subluxation of the atlas and axis. Dislocation of these vertebrae from normal position was the effect of an extreme load or force directed from above and from the right. In consequence, it caused an injury of the osseous and ligamentous structures of the cervical portion of the vertebral column. This led to the formation of a peri- and intraarticular hematoma. Next, the hematoma might have calcified, promoting heterotopic ossification within the atlanto-axial complex.

It is believed that trauma activates local factors such as bone morphogenic proteins or systemic factors such as the E2 prostaglandin, or both. These factors could induce bone-forming mesenchymal cells to differentiate to osteoblasts and stimulate bone formation. Another possible mechanism that may trigger heterotopic ossification in the case of trauma, is the disruption of joint proprioception after neurologic damage. This changes the relationship between the different periarticular tissues [15]. Even local micro trauma and decreased blood flow can facilitate the formation of new bone in connective tissues surrounding the joints.

Fusion between cervical vertebrae can be regarded as a congenital anomaly — asymptomatic or with manifested clinical features (e.g.: myelopathy, pain in the head, neck, shoulders). Hence, it can be associated with diseases such as the Klippel-Feil syndrome. In this syndrome several vertebrae are incorporated into one osseous block of grossly abnormal appearance, accompanied by clinical symptoms [16]. In our case, morphological features of the atlas and axis are not definitely altered. Both the vertebral body (C2) and the arches (C1, C2) have all processes properly developed. They were also of proper size and shape. Only the superior articular facets of the atlas showed altered morphology. They were flattened, with an uneven rough surface, and bone tissue defects. This could be an effect of a secondary lesion, which is typical for degenerative joint diseases [17].

A remarkable feature of the studied atlanto-axial block is thickening of the posterior surface of the dens. This happened probably due to ossification of the transverse ligament. A similar condition can be found in chondrocalcinosis, when the transverse ligament of the atlanto-axial joint calcifies just behind the dens. In chondrocalcinosis, deposits of pyrophosphate dihydrate crystals, which are found within the articular facets, cause inflammation. This pathological process may induce osteogenesis in soft tissues [18].

Total fusion of the neighboring vertebrae occurs also during the advanced phase of ankylosing spondylitis. One of the characteristic traits of this autoimmune disease is new bone formation. Osteoproliferative processes occur often at previously inflamed areas such as syndesmophytes, calcified entheses, and ligaments [19].

In our case the process of new bone formation occurred vividly between the dens and the anterior arch of the atlas. Also, small osteophytes originating from the inferior-anterior aspect of the dens could be an effect of an osteoproliferative process. Hence, ossified parts of the yellow ligament and articular capsules support the idea of heterotopic bone formation.

We are not able to establish what kind of neurological symptoms accompanied the male who was the donor of the studied specimen. Nevertheless, it should also be stressed that permanent displacement between the atlas and the axis could have an influence on the normal course of the vertebral vessels. These vessels might have been twisted during head movement, and subjected to compressions. This probably diminished blood flow in these vessels. Hemodynamic changes in the vertebral artery could cause occipital head and neck pain, vertigo, focal neurological deficits, and cerebral ischemic infarction [20].

Concluding, we speculate that formation of the atlanto-axial synostosis, in this particular case, was the effect of trauma of unknown origin. Surface deformation of the atlanto-axial joint seems to suggest a mechanical rather than an inflammatory etiology. This is also supported by the fact that metabolic or autoimmune diseases are not limited to a single vertebra, but usually encompass larger segments of the vertebral column. In consequence, the morphology of the entire vertebral column or at least its part is considerably altered.

CONFLICT OF INTEREST STATEMENT

None declared.

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