Occipitalization of atlas with other associated anomalies of skull

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SUMMARY

Congenital fusion of the atlas with the occipital bone is the common denominator of a galaxy of skull defects. Some skull anomalies may result in sudden unexpected death.

Seventy dry skulls were obtained from the Department of Anatomy. Of these, 2 skulls showed occipitalization. These were photographed and radiographed. Several measurements, including cranial index, dimensions of the foramen magnum, basal angle and basal impression, were taken. Both skulls showed near by complete fusion of the atlas with the occipital bone. A continuation of the confluence of the sinuses only to the left transverse sinus was observed in both skulls.

One skull was asymmetrical in shape. The foramina transversaria of atlas in this specimen were bilaterally reduced to admit only the diameter of a safety pin. The thickness of the wall of the other skull was asymmetrical on the two sides. It also showed a shelf-like bony triangular projection (dural band/ spur), which was attached to the upper margin of the groove for the transverse sinus on the right side. The cranial indices of both the skulls were greater than 80 (brachycranial/brachycephalic). Basal angles were within the normal range (120°-140°).

These variations are discussed in light of developmental aspects and their clinical significance.

Key words: Atlas – Foramina transversaria – Occipitalization – Basal angle

INTRODUCTION

Occipitalization of the atlas is defined as congenital bony fusion of the atlas vertebra to the base of the occipital bone of the skull. The incidence of atlanto-occipital fusion ranges from 0.14% to 0.75% of the population, both sexes being equally affected (Lanier, 1939a, b; Al-Motabagani and Surendra, 2006).

Occipitalization of the atlas can produce a wide range of neurological signs and symptoms, which vary from transitory headache to a full blown neurological syndrome.

Cervical cord compression is due to the soft tissue and bony abnormalities associated with occipitalization of the atlas. This leads to weakness and ataxia of the lower extremities. Numbness and pain in the upper extremities is a prominent complaint. Occipital headache may be present.

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Objective findings chiefly comprise hyperreflexia, positive Babinski and Hoffmann reflexes, weakness and other long-tract signs in both the upper and lower extremities. Ataxia may be present to a marked degree, and nystagmus is frequent. Sensory findings are less frequent.

Several examples of bony distortion of the foramen magnum were reported by early anatomists. There are two principal types of congenital anomalies: the first is *atlanto-occipital fusion or occipitalization* / assimilation of atlas, first described by Rokitansky in 1844 and demonstrated roentgenographically by Schüller in 1911, and second is the manifestation of *occipital vertebra*, described by Meckel in 1815 and again by Kallman in 1905. Several other authors have also reported this bony abnormality and the neurovascular complications associated with occipitalization of the atlas (Gladstone and Erichsen, 1915; Green, 1930; Lanier, 1939a; Mac Rae and Barnum, 1953; Misra, 1954; Shehata, 1964; Kalka et al., 1989; Al-Motabagani and Surendra, 2006).

Occipitalization of the atlas is an important congenital malformation of the craniovertebral region because of its proximity to the spinomedullary region, with the possibility of a neurological compression syndrome. It is one of the most common skeletal abnormalities of the upper cervical spine. According to Rowe and Yochum (1987), occipitalization represents the majority of «blocked» cephalic vertebra encountered in the spine.

To obtain a diagnosis of morphological assimilation of the atlas, it is necessary to demonstrate some degree of bony union between the skull and the atlas (Mac Rae and Barnum, 1954). Such assimilation usually involves the anterior arch of the atlas, the lateral masses, or the entire atlas. Partial fusion is more common. Probable criteria based on the position of the hypoglossal and suboccipital nerves and the vertebral artery are the most reliable for differentiating occipitalization of the atlas from manifestations of an occipital vertebra (third occipital condyles, a paracondyler process, a transverse basioccipital fissure, basilar process, bipartite atlantal facets).

Barge demonstrated that the atlanto-occipital joint is intersegmental and not segmental (Mac Rae and Barnum, 1953).

The aim of the present study was to determine the prevalence of atlanto-occipital fusions and to describe the skull changes associated with such an anomaly, along with a brief embryological explanation. Knowledge of this rare anatomical variation in the craniovertebral juncture is important for radiologists, neurologists and neurosurgeons.

Material and Methods

Seventy human adult skulls from the northwest region of India were studied for evidence of occipitalization of the atlas vertebra. Each specimen was examined carefully for any associated anatomical variations/anomalies. In the event of atlas assimilation being detected, the degree of fusion between the first cervical vertebra and occipital bone was examined. Skulls were photographed and X-ray films were also obtained in a lateral view. A vernier caliper was used for linear measurements. The following details were noted:

– Maximum cranial breadth: the maximum breadth taken at a right angle to the mid sagittal plane.
– Maximum cranial length: the linear distance between the glabella and opisthocranion.
– Cranial index = $\frac{\text{Maximum cranial breadth x 100}}{\text{Maximum cranial length}}$
– Antero-posterior diameter of the foramen magnum: the linear distance between the basion and opisthion.
– Antero-posterior length behind the odontoid process: the linear distance between the opisthion and midpoint taken on an imaginary line drawn between the tubercles to which the transverse ligament of the atlas is attached.
– Breadth of foramen magnum: the greatest transverse breadth of the foramen magnum.
– Antero-posterior length of the vertebral foramen of the atlas vertebra: the maximum antero-posterior distance in the midline of the vertebral foramen.
– Maximum breadth of the vertebral foramen of the atlas vertebra: the linear distance between two points of the vertebral foramen of the atlas vertebra on the most laterally placed margins.
– Length at the upper border of the dural spur: the linear distance between the apex and a point at the upper end of the base of the spur.
– Length at the lower border of the dural spur: the linear distance between the apex and a point at the lower end of the base of the spur.

– Length of the base of the dural spur: the linear distance between the upper and lower ends of the base of the dural spur.
– Basal angle (on X-ray film): measured between lines drawn from the tuberculum sellae to the nasofrontal suture and to the anterior lip of the foramen magnum on the X-ray film.
– Basilar impression: a transverse line drawn between the groove for the posterior belly of the digastric muscle of one side to the same point on the other side. The position of this line in respect of the atlanto-occipital joint or sqamous part of the occipital bone was noted. If the line lies at or below these structures, the basilar impression is present.
– Cranial capacity: cranial capacity was measured using sand: all holes were plugged with plasticine and the skull was filled with sand. The sand was then measured volumetrically.

Results

Of seventy skulls, only two exhibited the anomaly of occipitalization of the atlas vertebra. Both skulls were sexed subjectively using morphological features given in standard text books of anatomy, anthropometry and forensic science (Al-Motabagani and Surendra, 2006; Singh and Bhasin, 1989; Standring et al., 2005), showing the skulls probably belonged to approximately 40-45-year old males.

Skull-I measurements:

Maximum cranial length – 16.09 cm
Maximum cranial breadth – 13.33cm
Cranial index – 82.84% (brachycranial)
Cranial capacity – 1370 c.c. (mesocephalic)
Basal angle – 122° (normal 120°-140°)
Basal impression – present
Foramen Magnum:
Antero-posterior length – 2.85 cm
Transverse width – 2.50cm
Atlas (vertebral foramen):
Antero-posterior length – 3.05 cm
Antero-posterior length behind the odontoid process – 1.7 cm
Transverse width – 2.70cm
Dural spur:
Length at upper border – 2.1cm
Length at lower border – 1.9cm
Length at base – 1.5cm

Features

The bilateral presence of a carotido-clinoid foramen was noted (Fig. 1). The thickness of parietal wall was asymmetrical on both sides. Thickness was greater on the right side as compared to the left side. The hypophyseal fossa was shallow.

The groove for the left sigmoid sinus was larger and spacious as compared to the right side. The confluence of sinus was continuous with the left transverse sinus. The groove for the occipital sinus split at the posterior aspect of the foramen magnum and joined the groove for the sigmoid sinuses of both sides.

A bony triangular shelf-like projection, whose base was attached between the right parietal and sqamous part of the right temporal bone was seen. This is the dural spur present at the upper margin of the attachment of the tentorium cerebelli (Fig. 1).

Occipitalization of the Atlas

– Atlas:

The atlas vertebra was almost completely fused with the occipital bone at the base of the skull, except at the transverse processes on both sides.

Anterior arch: This part of the atlas was fused with the anterior margin of the foramen magnum, leaving a single anterior midline (0.4 cm) opening between the anterior tubercle and the basiocciput (Fig. 2). A small circular facet of approximately 6-7 mm in diameter, located on the posterior surface of the anterior arch in the midline for articulation with the odontoid process of the axis vertebra was seen. No part of the axis vertebra was fused with the atlas.

Posterior arch: Almost the whole of the posterior arch was synostosed with the posterior rim of the foramen magnum. A few small foramina and cleft-like openings were observed along the line of fusion. The posterior tubercle had lost its typical appearance. On both the sides, the groove for the vertebral artery on the upper surface of the posterior arch was converted into a bony canal. The canals for the vertebral arteries were equal in diameter on the two sides (Fig. 2). This part of the posterior arch was thinned out. On the left side, there was a small foramen close to the posterior arch, most probably serving for the dorsal ramus of the suboccipital nerve.

Lateral masses: The lateral masses were completely fused with the condylar parts of the occipital bone, as well as being symmetri-

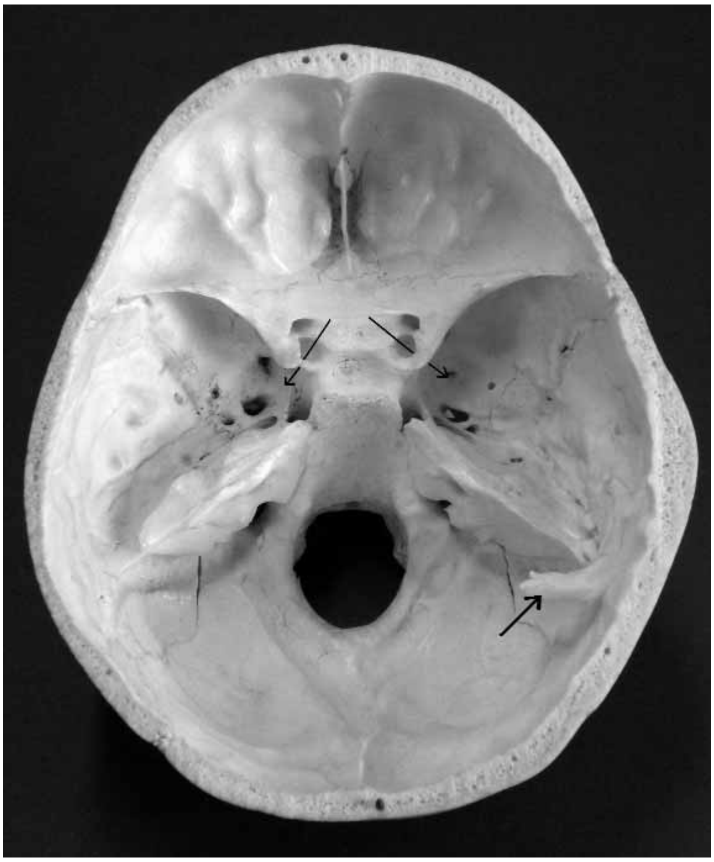

Fig. 1. Skull I: Interior of skull. Thin arrows: bilateral presence of carotido-clinoid foramen. Thick arrow: dural spur.

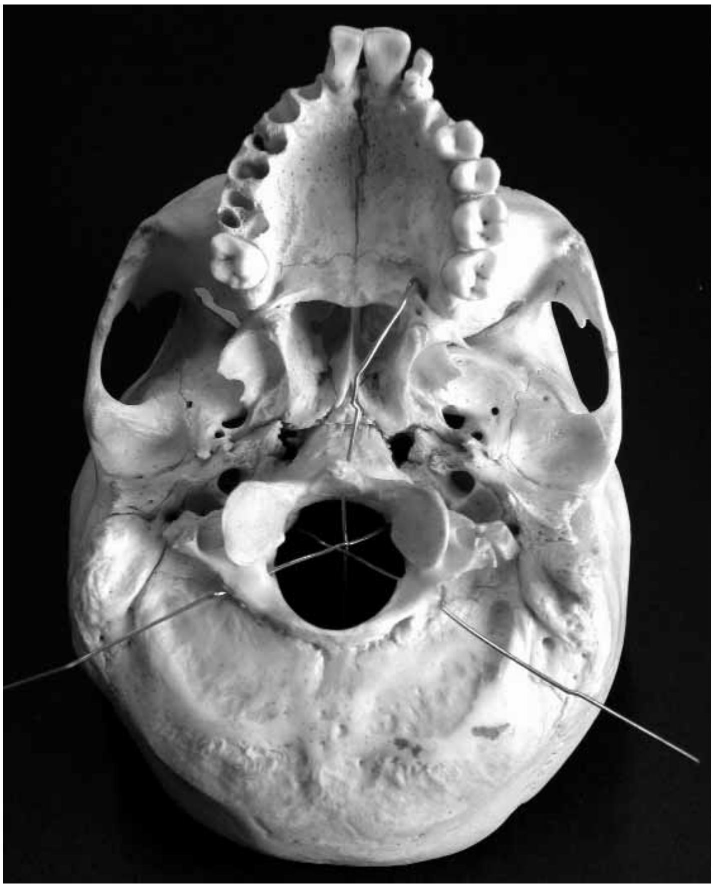

Fig. 2. Skull I: Inferior surface of the skull. Anterior midline probe: shows a single anterior midline opening between anterior tubercle of atlas and the basiocciput. Two posterior probes on either side: Show the canals for the vertebral arteries.

cal on the two sides. Plain X-rays revealed no evidence of a joint cavity (Fig. 3).

On both the sides, the inferior articular facets were almost oval in shape, with a narrow anterior extension. The facets were relatively smooth and flat, directed anteromedially along the long axis and downwards and medially along the transverse axis.

Transverse processes: On both sides, the transverse processes were directed downwards and laterally. On the left side, the costotransverse bar was missing, causing an incomplete foramen transversarium. The transverse processes were not fused with the occipital bone or paracondylar processes. The right foramen transversarium was larger than the left one.

– Occipital bone:

The margin of the foramen magnum was completely fused with the synostosed atlas vertebra. Bilateral paracondylar processes were observed.

Basilar part: The length of the clivus was 3.6 cm (normal: 4.05 cm). The pharyngeal tubercle was prominent and fused with the right margin of the anterior tubercle of the anterior arch of the atlas vertebra.

Condylar parts: The occipital condyles were completely covered by the lateral masses of the fused atlas. Hypoglossal canals were normal. Condylar fossae and posterior condylar canals were not seen. The jugular processes were normal. The left jugular notch was wider and deeper as compared to the right one.

Squamous part: This was normal.

Foramen magnum: The shape was nearly normal. A slight encroachment of the foramen magnum by the anterior arch as well as by the fused condylar parts with the lateral masses of the atlas vertebra was seen. No incidence of a third occipital condyle on the anterior margin of the foramen magnum was noticed.

– Cranial base:

The basal angle was 120° (Fig. 3) and within the normal range (120°-140°). A basilar invagination was present.

Skull II measurements:

Maximum length – 17.12cm
Maximum breadth – 13.89cm

Cranial index – 81.13% (Brachycranial)
Cranial capacity – 1390 c.c. (Mesocephalic)
Basal angle – 136° (normal 120°-140°)
Basal impression – not present
Foramen magnum:
  Antero-posterior length – 2.90 cm
  Antero-posterior length behind the odontoid process – 1.7 cm
  Transverse width – 2.15 cm
  Distorted in shape
Atlas:
  Antero-posterior length – 3.05 cm
  Transverse width – 2.30 cm
  Vertebral canal distorted in shape

Features

The skull was asymmetrical in shape. The right temporal wall was pushed inside as compared to the left side. The left parietal eminence was prominent and thicker than the right one. The hypophyseal fossa was deep and the dorsum sellae was observed as a thin vertical chip of bone. The clivus was vertical. The groove for left sigmoid sinus was deeper and wider as compared to right side. A continuation of the confluence of sinuses to the left transverse sinus was noticed. The shape of the foramen magnum was distorted.

Occipitalization of the Atlas

– Atlas:

The atlas vertebra was almost completely fused with the occipital bone at the base of the skull, except at the transverse processes on both sides.

Anterior arch: The anterior arch was much higher than the posterior arch. This part was fused with the anterior margin of the foramen magnum, leaving a single midline opening (0.3 cm) between the anterior tubercle and the basiocciput. This opening entered the vertebral canal. No facet was observed on the posterior surface of the anterior arch of the atlas vertebra for the articulation of the odontoid process of the axis vertebra. The left inferior articular facet's anterior narrow portion extended anteriorly over the posterior surface of the anterior arch of the atlas vertebra. The right portion of the anterior arch was longer than the left one (Fig. 4). No part of the axis vertebra was fused with the atlas vertebra.

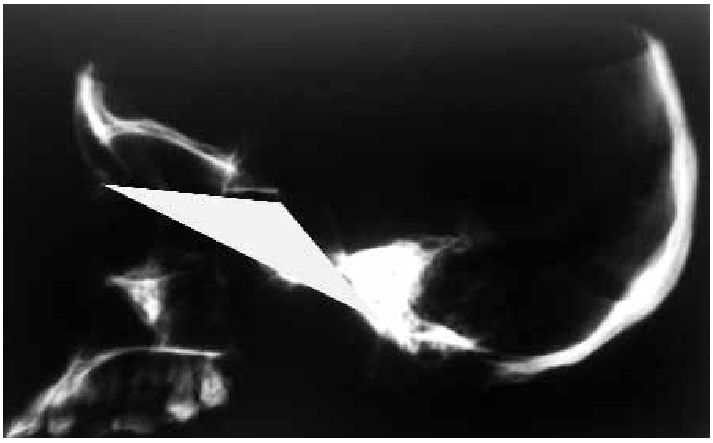

Fig. 3. Skull I: Plain X-ray. No evidence of a joint cavity and the basal angle is within the normal range.

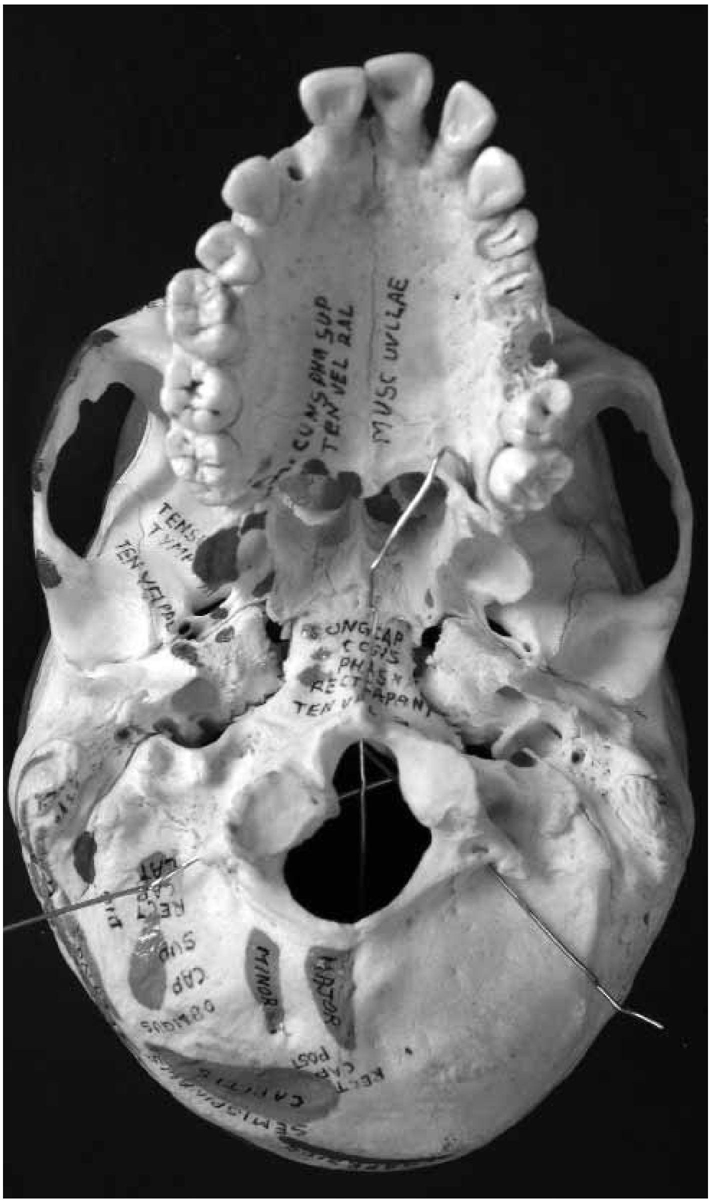

Fig. 4. Skull II: Inferior surface of the skull. Probes: as in skull-I. Right portion of the anterior arch is longer than the left. Both articular processes are oblique. Left transverse process is larger than the right one, and a single mass can be seen lateral to the foramen transversarium.

Posterior arch: Almost the entire posterior arch was united with the posterior rim of the foramen magnum. A few small foramina were observed along the line of fusion. The left portion of the posterior arch was longer than the right one. The posterior tubercle of the posterior arch had lost its typical appearance. At the inferior margin, the posterior arch in the midline showed a notch. On both sides, the grooves for the vertebral arteries did not show any curve, because the foramina transversaria were located immediately below the gap between the upper surface of the transverse process and the lower margin of the foramen magnum. On both sides, foramina for the suboccipital nerve (dorsal ramus) were present, but on right side it was incomplete, both leading into the canal for the vertebral arteries.

Lateral masses: The bony masses representing the fused occipital condyles and lateral masses were asymmetrical. The height of the fused right lateral mass with the occipital condyle was more prominent than on the left side. Plain X-ray revealed no evidence of a joint cavity.

Right inferior articular facet and process:

Position - slightly distal and posteriorly placed to the left one
Shape - oval
Size - smaller than the left one
Axis - anteromedially (nearly transverse)
Surface - concave along its longituidinal axis, slightly concave at the right angle to its axis.

Projecting into the vertebral canal of atlas/foramen magnum.

Two vascular foramina were present on postero-medial surface.

Left inferior articular facet and process:

Position - slightly proximal and anteriorly placed to the right one
Shape - oval with an indentation at its posterior margin
Size - larger than the right one
Axis - antero-medial with slightly upward tilt (longitudinal)
Surface - the lateral margin was lower than the medial margin. The facet itself faced towards the right vertebral canal/foramen magnum. The anterior end of the facet reached the anterior midline.

Transverse: Exhibited an upward slope from the lateral to the medial side.

– Axis:

The vascular foramen was present on the medial surface of the articular process. Both articular processes were present obliquely. The left one was placed more anteriorly than the right one (Fig. 4).

Transverse process: The bilateral transverse processes were smaller, directed posterolaterally, and were not fused with the occipital bone or paracondylar processes. The left transverse process was larger than the right one, and a single mass (anterior tubercle of the transverse process) was noticed lateral to the foramen transversarium (Fig. 4). On the right side, anterior and posterior tubercles, as well as the costotransverse bar were noticed. The foramina transversaria on both sides were small in size, with a diameter of only a safety pin. The right foramen transversarium was larger than the left one. No signs of fusion between the atlas and axis were evident.

– Occipital bone:

The margin of the foramen transversarium was completely covered by the fused atlas vertebra. Bilateral paracondylar processes were observed.

Basilar part: A longitudinal ridge ran from the upper margin of the gap between the anterior arch of the atlas and the anterior margin of the foramen magnum. The length of the clivus was reduced to 3.8 cm (normal - 4.05 cm).

Condylar parts: The occipital condyles were completely covered by the lateral masses of the fused atlas. The hypoglossal canal of the right side had a large circular opening as compared to the left hypoglossal canal. The left canal was smaller, flattened and oval shaped, and a bony ridge/spicule divided the canal into upper and lower parts, representing the fused intervertebral foramina of the occipital sclerotomes (Fig. 5).

The canal for the suboccipital nerve (dorsal ramus) was present bilaterally and it opened into the canal for the vertebral artery. The left suboccipital canal was incomplete. No condylar fossae or posterior condylar canals were seen on either side. Jugular processes were seen, but on left side it was raised superiorly to form a fossa. The jugular notch was wide and shallow on the left side as compared to the narrower and deeper notch on the right side.

Bilateral paracondylar or paramastoid processes were observed.

Squamous part: Most of the anatomical features of the external and internal surfaces of the occipital squama were distorted due to asymmetrical changes in the bone.

Foramen magnum: This opening had irregular margins and shape. Anteriorly, the margin of the foramen magnum was found to be overlapped by the inferior articular facets of the fused atlas, much more so on the right side. Posteriorly, the rim of the foramen magnum was encroached by the fused posterior arch due to the oblique union of the atlas (Fig. 4). Because of this invasion by a portion of the occipitalized atlas, the functional size of the foramen magnum was reduced. No incidence of a third occipital condyle on the anterior margin of the foramen magnum was noticed.

– Cranial base:

The basal angle was 136° and within the normal range (120°-140°). Basilar invagination was not present (Fig. 6).

Discussion

Occipitalization of the atlas is commonly associated with concomitant neurovascular and skeletal anomalies at the craniovertebral junction, which may produce a variety of symptoms. Absolute immobility of an occipitalized atlanto-occipital joint results in compensatory hypermobility between the atlas and axis. Approximately 50% of patients with atlanto-occipital fusion develop atlanto-axial instability (Warner, 2003) and concurrent subluxation (Turek, 1984). In the present study, skull I had inferior articular facets like those observed in a normal unfused atlanto occipital joint and the facet for the articulation of the odontoid process was also present. This type of skull can develop atlanto axial instability or subluxation. Skull II showed asymmetrical inferior articular facets and the direction of these facets helped the axis vertebra to retain it in a position to prevent subluxation. The left lateral mass of the fused atlas had the appearance of a socket holding the superior articular facet of the axis.

Fig.5. Skull II: Posterior and part of the middle cranial fossae. The left hypoglossal canal is divided by a bony ridge/spicule.

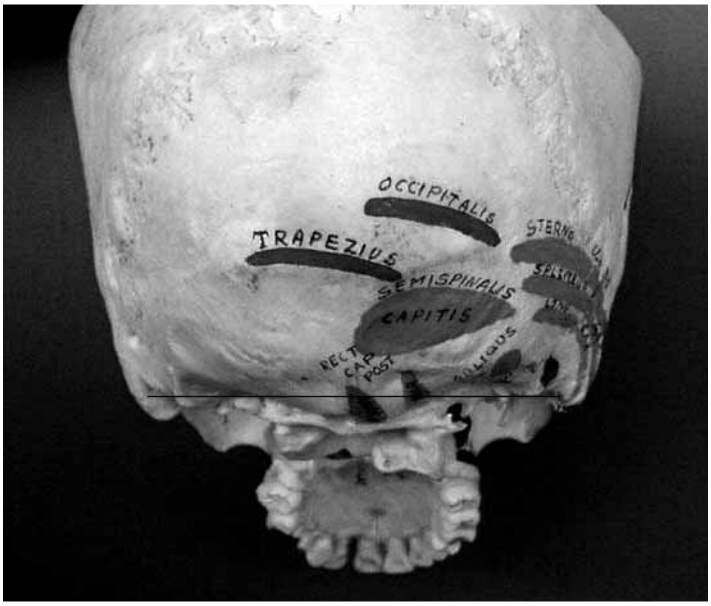

Fig. 6. Skull II: Posterior view of the skull. Basilar invagination is not present as indicated by a transverse line lying above the atlanto-occipital joint.

A congenital anomaly of the cranio-vertebral junction, assimilation of the atlas with the occipital bone, is generally associated with bony torticollis (McRae and Barnum, 1953; Bharucha and Dastur, 1964; Tachdjian, 1990). The muscular markings, especially in the region of squamous part of the occipital bone, were well marked. The thickness of the wall of the skull was also asymmetrical. These are some of the features which might cause bony torticollis. Vertebral artery anomalies with atlanto-occipital fusion were observed by Bernini et al. (1969). In the present study, skull I showed a slight difference in the size of the foramina transversaria. In skull II, the foramina transversaria were very small. These findings indicate that the vertebral arteries were of unequal size and that they may have had an aberrant course passing between the atlas and the axis. A possible explanation for this anomaly is the embryological basis of the development of the vertebral artery. An error may occur in the formation of the first cervical intersegmental artery and its branches and in establishing its post costal anastomosis with the second cervical intersegmental artery.

In skull II, the left hypoglossal canal was divided by a bony spicule into two portions, representing the fused intervertebral foramina of the occipital sclerotomes. This finding was also reported by Al-Motabagani and Surendra (2006).

Constriction of the foramen magnum leads to spinal cord compression, which always occurs when the saggital spinal canal diameter behind the odontoid process is less than or equal to 14 mm. It is possible when the above diameter is between 15 to 17 mm but almost never when it is > 18 mm. In the present study, both cases had a diameter of 17 mm, and hence spinal cord compression may have been present. Primary basilar invagination of congenital type, shown by the interdigastric groove line, indicates the cranial displacement of the odontoid process into the foramen magnum. In the present study, the basilar invagination was seen in skull I, which was not associated with platybasia. This is in agreement with previous findings that in the presence of basilar invagination, the basal angle may be normal or increased (Rowe and Yochum, 1987; Tachdjian, 1990).

Variations and malformations of the craniovertebral region can be seen during the embryonic period long before ossification. Lanier (1939a, b) and Frame (1960) attributed this to cranial shifting of the intersegmental boundaries during the regional grouping of vertebra. Hadley (1956) described it as being due to non-segmentation or a failure of segmentation rather than fusion. Harcourt and Mitchell (1990) suggested a lack of segmentation and separation at the caudalmost occipital sclerotome. Tachdjian (1990) mentioned that the dense caudal part of the fourth occipital somite apparently unites with the cephalic part of the subjacent somite only in cases of atlanto-occipital fusion. Chandraraj and Briggs (1992) explained it as being due to an interruption in the differentiation of the occipito-cervical somites early in the embryonic period. Thus, the available embryological data regarding the occipitalization of the atlas are confusing and inadequate. The cervico-occipital junction develops between the last occipital and first sclerotomic segments.

The basiocciput is formed by the cranial part of the fourth occipital sclerotome with the remaining three occipital sclerotomes cranial to it and the medial part of the caudal portion of the fourth sclerotome. Occipital condyles are formed from the lateral part of the caudal portion of the fourth sclerotome. The tip of the dens and part of the occipital condyles are formed by the cranial part of the first cervical sclerotome, which appears to fuse with the supraadjacent caudal part of the fourth occipital sclerotome.

The caudal portion of the first cervical sclerotome mainly gives rise to the neural arch of the atlas (posterior arch) and the hypochordal bow becomes the anterior arch. The caudal portion of first cervical sclerotome also helps to form the upper part of the dens. The middle part of the dens, which represents the centrum of the atlas, is derived from the cranial part of the second cervical sclerotome, which fuses with the caudal part of the first cervical sclerotome. The dense caudal part of the second cervical sclerotome forms the neural arch of the axis and a rudimentary disc between the atlas and axis.

The occipitalization of the atlas seen in the present study is most likely due to a failure in differentiation of the fused caudal and cranial segments of the fourth occipital and first cervical sclerotomes, respectively, and a lack of segmentation and separation between the cranial and caudal parts of the first cervical sclerotome.

Skull I also showed some very important bony variations. It is well known that the anterior and middle clinoid processes are linked by a carotido-clinoid ligament. However, sometimes it ossifies to form a carotido-clinoid foramen. In our study, skull-I had bilateral carotido-clinoid foramina. The presence of this foramen is rare.

The clinical importance of this rare finding is that while doing the carotid arteriogram, it may confuse the radiologists, so Physicians should be aware of this uncommon anomaly (Patnaik et al., 2003).

A dural spur was also observed in the same skull and was formed as a sequel to ossification of the duramater (tentorium cerebelli). During acceleration and deceleration accidents, it can cause severe damage to the brain, leading to sudden death.

In conclusion it may be stated that occipitalization of the atlas reduces the foramen magnum diameter, leading to neurological complications due to compression of the spinal cord.

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