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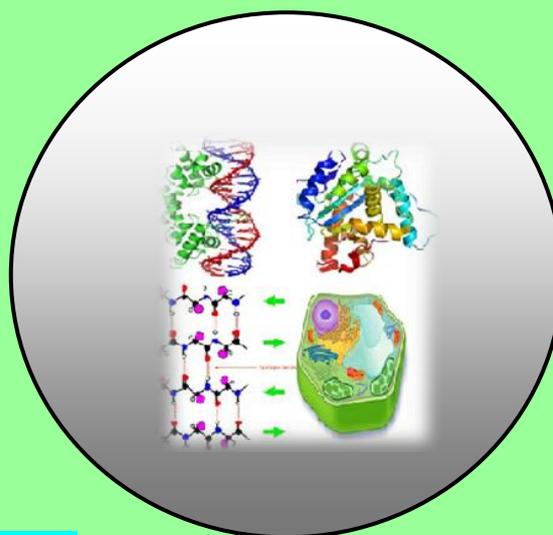
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[http:// www.sasjournals.com](http://www.sasjournals.com)[http:// www.jbcr.in](http://www.jbcr.in)jbiolchemres@gmail.cominfo@jbcr.in**CASE REPORT**

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Atlanto-Occipital Synostosis: Embryological Basis

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Uttar Pradesh, India*Department of Anaesthesia, Sanjay Gandhi Post Graduate Institute of Medical Sciences,
Lucknow-226014, Uttar Pradesh, India**ABSTRACT**

Atlantooccipital synostosis is a rare anomaly. It is also known as assimilation or occipitalization of atlas. It is defined as a congenital fusion of atlas vertebra with the base of occipital bone; it may be complete or partial. This anomaly results due to failure in segmentation and separation of last occipital sclerotome and first cervical sclerotome during development of foetus.

During routine demonstration classes of undergraduate medical students of King George's Medical University, Lucknow, Uttar Pradesh, India, we noticed a cranium which was fused with the first cervical vertebra. The anterior and posterior arches of atlas were completely fused with occipital bone and the superior articular facets of atlas were completely fused with respective condylar facets of occipital bone.

This type of anomaly may alter the dimensions of foramen magnum and may compress the spinal cord and vertebral artery. Therefore, improved knowledge of the fusion of atlas with the occipital bone is important for neurosurgeons, orthopedicians, anaesthetist, radiologist, physiotherapist and anatomist, as it shows multiple variations and combinations.

Keywords: Atlas, Occipital Bone, Synostosis, Craniovertebral Junction, Variation and Sclerotome.

INTRODUCTION

The occipital bone forms the back and base of cranium and its inferior surface possess two occipital condyles. The first cervical vertebra supports the skull, so also called atlas (after the

mythical greek God who supported the globe) (Basmajian and Slonecker, 1997). Atlas vertebra is ring shaped, without a body having an anterior arch, posterior arch and two lateral masses. Its superior articular facets articulate with the occipital condyles to form atlantooccipital joint which is a synovial joint of ellipsoid variety having movements of flexion, extension and slight lateral rotation (Standring et al. 2008). There are various anatomical abnormalities of this region of which atlantooccipital-synostosis is most common (Lang et al. 1995). It is defined as congenital fusion of atlas to the base of the occiput. The other terms for the same is occipitalization of atlas, assimilation of atlas, occipitocervical synostosis and atlantooccipital fusion. It was first described by Rokitsansky in 1884 and first radiological report was published in 1911 by Suchuller. Its incidence is 1 in 109 adult skulls of Asian population (Motabagani and Surendra 2006). The fusion may be complete or incomplete.

CASE REPORT

During routine demonstration classes of undergraduate medical students of King George's Medical University, Lucknow, Uttar Pradesh, India, we noticed a cranium which was fused with the first cervical vertebra. The bone was studied carefully and photograph was taken from the base of the cranium to see the fusion properly. Measurement of foramen magnum and inferior articular facets was done by vernier calliper. There was complete fusion of anterior and posterior arches of atlas with the anterior and posterior margins of foramen magnum. Superior articular facets of atlas were completely fused with occipital condyles. Just behind the fused lateral masses, a canal was observed bilaterally between atlas and occipital bone which most probably gave passage to vertebral artery (Fig. 1). Both transverse processes appeared to be normal. On left side, it was free from the base of the cranium but on right side, a spicule of bone connected the costotransverse bar with jugular process due to which a foramen was created that most probably gave exit to first cervical nerve (Fig. 2). The maximum antero-posterior diameter of foramen magnum was 27 mm and transverse diameter 25 mm. There was absence of articular facet for dens on the anterior arch of atlas. The right and left inferior articular facets of atlas were irregular in shape and the right facet was divided into two parts i.e. horizontal part and vertical part (Fig. 3). The horizontal part was present at the normal position of inferior articular facet while the vertical part was protruding into foramen magnum making an angle of 90° with horizontal part. The antero-posterior and transverse diameters of inferior articular facets of both the sides were measured with vernier calliper (Table 1).

As the articular facet for dens was not present on anterior arch, it seems that the dens articulated with atlas on vertical part of right inferior articular facet (Figure 3). The external occipital crest was underdeveloped while the external occipital protuberance was more marked.

DISCUSSION

During development, the ventro-medial portion of somite forms sclerotome which surrounds the notochord and develops into vertebral body. The dorsal portion surrounds the neural tube and develops into posterior vertebral arch. Then the caudal half of each sclerotome combine with the cranial half of the sclerotome below it.

The cranial half of first cervical sclerotome combine with the caudal half of last occipital sclerotome to form the base of skull. While the caudal half of first cervical sclerotome combine with the cranial half of second cervical sclerotome to form the first cervical vertebra. The pattern continues in this way to form rest of the vertebra. The dens of second cervical vertebra is formed by the body of first cervical vertebra, thus the first cervical vertebra doesn't have body(Sadler 2007).



Figure 1. Photograph showing bilateral canal between atlas and occipital for passage of vertebral artery (red wire).



Figure 2. Photograph showing a foramen (yellow wire) formed between costotransverse bar (blue arrow) and jugular process on right side for exit of first cervical nerve.

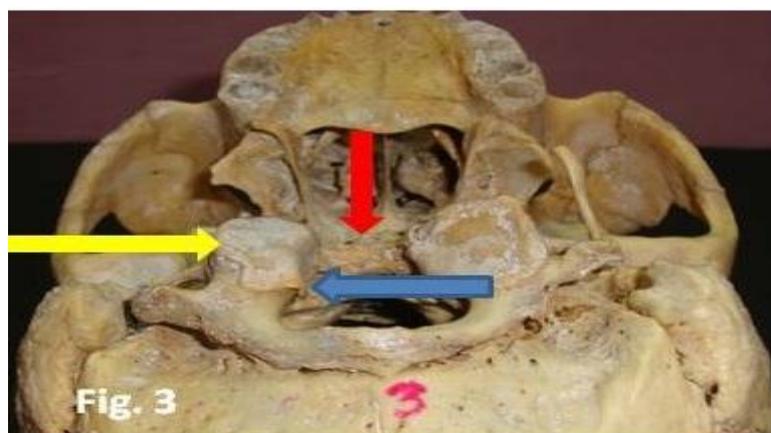


Figure 3. Photograph showing horizontal (yellow arrow) and vertical (blue arrow) parts of right inferior articular facet and absence of articular facet for dens (red arrow) on anterior arch of atlas.

Table 1. Measurements of inferior articular facet of atlas.

Diameter	Left inferior articular facet	Right inferior articular facet (Horizontal part)	Right inferior articular facet (Vertical part)
Antero-posterior	21 mm	18 mm	13 mm
Transverse	19 mm	11 mm	5 mm

The present case most probably arises as a result of failure of segmentation and separation of most caudal occipital sclerotome and first cervical sclerotome during initial weeks of foetal life (Yochum and Rowe 1987). This variation may often be unnoticed, but incidentally found during radiological and operative procedure or during autopsy (Jadhav et al. 2012). The onset of clinical symptoms can be sudden or may be precipitated by minor trauma and sudden death has also been reported (Hensinger 1986). It may produce narrowing of foramen magnum and spinal canal which may compress spinal cord or brain stem. The standard dimensions of foramen magnum ranges between 28-38 mm for sagittal and 25-40 mm for transverse diameter (Motabagani and Surendra 2006) and the spinal cord compression occurs when sagittal diameter is less than 18 mm (Greenberg 1968). It can produce signs and symptoms which vary from simple headache to full blown neurological syndrome i.e. occipital headache, neck pain, numbness, abnormal head posture and convulsion. The vertebral artery and first cervical nerve are related with superior aspect of atlas, so there may be chances of their compression which can cause compromise blood flow to the brain leading to dizziness, seizure, syncope and neurological symptoms (Jayanti et al. 2003). If cranial nerve is involved, there may be tinnitus and visual disturbance. Clinically, wide variety of other signs and symptoms may be associated like restricted or absent movement, ataxia, muscular spasticity and torticollis. The atlantooccipital synostosis may be classified into 4 types based on the zone of atlas that is fused with occiput (Gholve et al. 2007).

Zone -1 = Fusion of anterior arch

Zone -2 = Fusion of lateral masses

Zone -3 = Fusion of posterior arch

Zone -4 = Combination of zones

According to this classification, the present case can be categorised into Zone 4. Some associated malformations may commonly occur with atlantooccipital synostosis like fusion of C1 and C2, absence or malformation of transverse ligament, hyperplasia or aplasia of dens and anomalies of vertebral artery. The findings of the present case suggest that patient might have left torticollis with restricted neck movement and absent or malformed transverse ligament.

CONCLUSION

This anomaly should be checked in time especially by neurosurgeons and orthopaedic surgeons because this can cause severe neurological symptoms, convulsion, seizure, severe neck pain and even sudden death. As the transverse process is very important landmark for surgeons, the knowledge of fusion may be important for head and neck surgeries. It may cause failure of cisternal puncture, therefore also important for anaesthetist.

Physiotherapist dealing with neck pain and radiologist dealing with abnormalities of cervical spine, must also be aware of this condition. Because of the serious consequences of this type of osseous anomaly there is a need of thorough clinical assessment and evaluation of every patient.

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