

Occipitalization of Atlas: A case report.

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Abstract:

Occipitalization of atlas is an osseous anomaly of the cranio-vertebral junction which occurs at the base of the skull in the region of the foramen magnum due to failure of segmentation and separation of the most caudal occipital sclerotome and the first cervical sclerotome during the first few weeks of foetal life. Atlas is the first cervical vertebra and the atlanto-occipital joint belongs to ellipsoid variety of synovial joints. It does not have a body like other vertebrae. Rarely, it gets occipitalized, where the lateral masses of the atlas vertebra fuse with the condyles of the occipital bone. One skull showing Occipitalization of Atlas was observed in the Department of Anatomy, B.J. Medical College, Ahmedabad, Gujarat. The knowledge of such a fusion may be of importance for radiologists, anaesthesiologists, orthopaedic and neurosurgeons because skeletal abnormalities at the cranio-cervical junction may result in sudden unexpected death. It can result in dysphagia, dysarthria or torticollis because of compression of cranial nerves. They should be aware that such an anomaly may exist without any typical symptomatic presentation, and thus, serious consequences of upper cervical spinal manipulative therapy may arise when a complete and adequate clinical assessment is missed.

Key words: Atlanto-Occipital joint, Atlas, Occipital bone, Occipitalization.

Introduction:

Atlas is the first cervical vertebra. It is ring like structure, consisting of anterior and posterior arches including two lateral masses along with the transverse processes. Two lateral masses bear a kidney shaped superior articular facets which articulate with occipital condyles to form atlanto-occipital joint of ellipsoid variety. Inferior articular facets of these lateral masses are almost circular and flat. It is oriented more obliquely to the transverse plane than the superior articular facet and faces more medially. Inferior articular facets of atlas form lateral atlanto-axial plane joint¹. The anterior arch which forms anterior 2/5 of ring is slightly convex anteriorly and bears anterior tubercle. The anterior longitudinal ligament and longus colli muscles are attached to anterior tubercle. Its upper border provides attachment to the anterior atlanto-occipital membrane which often gets ossified. The posterior surface of the anterior arch bears concave circular facet for dens¹. This condition was initially described by Rokitansky in 1844 and was demonstrated radiologically by Schuller in 1911². According to Yochum and Rowe⁸ occipitalization

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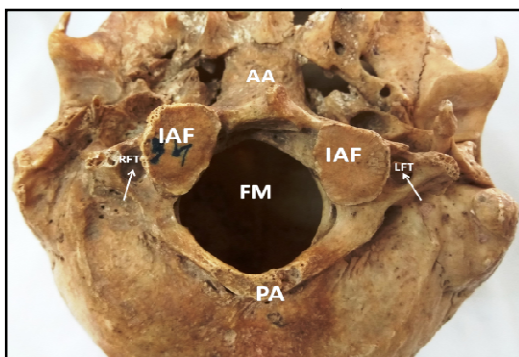


represents the most cephalic ‘blocked’ vertebra encountered in the spine. Atlanto-occipital fusion can be complete or partial, unilateral or bilateral and bony or fibrous. Its incidence is reported from 0.14 to 3.63% according to different authors in the whole population²⁻⁷, with equal repartition in both sexes and it is considered the most common anomaly of the cranio-cervical junction²⁻⁴. More often congenital, atlas assimilation may be associated with atlanto-axial instability. Atlas assimilation may be asymptomatic otherwise its onset is normally characterized by progressive appearance of neurologic symptoms in the third or fourth decade due to brain stem compression or vertebro-basilar insufficiency⁴⁻⁶.

Case Report:

Partial and asymmetric occipitalization of atlas was found in the Department of Anatomy, B.J. Medical College, Ahmedabad, Gujarat. Image 1 showing anterior arch, posterior arch, inferior articulating facet, right foramen transversarium, left foramen transversarium and foramen magnum. Anterior arch of atlas was partially fused with anterior margin of foramen magnum leaving a slit like gap measuring 0.9 mm transversely between it and the basilar part of occipital bone (Image 2). Superior articular facets of atlas were completely fused with condylar facets of occipital bone. Tip of the right transverse process was fused with lateral part of the occipital bone leaving a gap measuring 10mm in diameter. Both the foramen transversarium was normal in dimension. Right posterior arch was complete and it was fused with the right part of the posterior margin of foramen magnum except for a small foramen on its superior surface close to the transverse process, most probably for the transmission of the right vertebral artery (Image 3). Left posterior arch was also complete but not fused with left part of posterior margin of foramen magnum and leaving a gap measuring 10mm for transmission of left vertebral artery (Image 4). Another 15mm gap was noted in between the posterior arch tip and the basiocciput (Image 5). Dimensions of Left Inferior Articulating Facet were 16mm in length and 15mm in width, Right Inferior Articulating Facet was 19mm in length and 14mm in width. Foramen magnum was 35mm in sagittal diameter and 30mm in transverse diameter. An articular facet measuring 14mm in width and 7mm in length was found on the posterior surface of anterior arch in the midline for articulation with the odontoid process of axis vertebra.

**Image 1: Occipitalization of Atlas
(Inferior View)**



**Image 2: Anterior arch of atlas
partially fused with
Anterior margin of foramen magnum**



AA- anterior arch, PA- posterior arch, IAF- inferior articulating facet,
RFT- right foramen transversarium, LFT- left foramen transversarium, FM- foramen magnum.

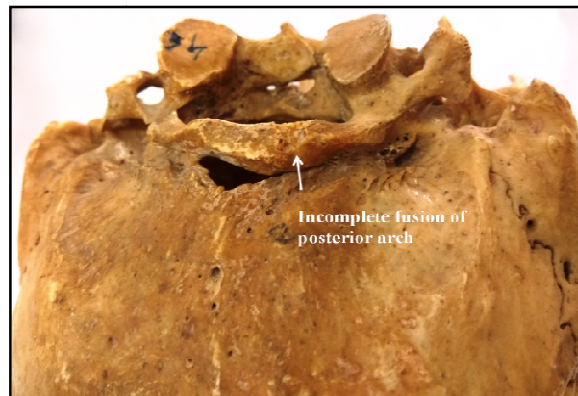
**Image 3: Atlanto-occipital joint
(Right Side)**



**Image 4: Atlanto-occipital joint
(Left Side)**



Image 5: Atlanto-occipital joint (Posterior view)



Discussion:

By the beginning of the fourth week cells forming the ventral and medial wall of the somite lose their compact organisation to become polymorphous and shift their position to surround the notochord by a protein product gene, Somic hedgehog (shh) released by the notochord and the floor plate of neural tube which induce the ventromedial portion of the somite to become sclerotome, forming loosely woven tissue the mesenchyme⁹. The induced sclerotome express the transcription factor PAX1 which initiates the cascade of cartilage and bone forming genes for vertebra formation. They surround the spinal cord and the notochord to form the vertebrae⁹.

In atlas assimilation, the spinal canal may be anteriorly compressed by fibrosis in the retro-odontoid space or posteriorly by a partially assimilated posterior arch, and so on¹⁰. According to Greenberg, spinal cord compression always occurs when the sagittal spinal canal diameter behind the odontoid process is ≤ 14 mm¹⁵. Spinal cord compression occurs when the sagittal cord diameter is between 15 and 17 mm¹⁴. The incidence has reported as 1.4-2.5per 1000 children, affecting both males and females equally¹⁶.

According to Kang Rae Kimn et al patient can have numbness of all his fingertips in his left hand, hyperesthesia, posterior neck pain, and left shoulder pain, Lhermitte's sign and a positive Hoffmann's sign¹⁰. Song SH et al studied recurrent posterior circulation infarction caused by anomalous occipital bony process in a young patient with cranio-cervical anomalies¹¹. Kotil K et al reported recurrent posterior circulation infarction caused by

anomalous occipital bony process in a young patient with anomalous¹². Gergely Bodon et al studied that a patient with atlanto occipital assimilation was diagnosed and treated at a neurology department with supranuclear palsy and multiple cerebellar infarcts, high blood pressure, atherosclerosis, chronic pancreatitis, pancreatogenic diabetes, polyneuropathia diabetica and dementia¹³. Patients with atlanto-occipital fusion usually have low hair lines, torticollis, short neck and restricted neck movement. Half of the patients have associated atlanto-axial instability. Kyphosis and scoliosis are associated with this deformity. Other congenital anomalies seen in 20 % of atlanto- occipital fusion are anomalies of jaw, incomplete cleft of nasal cartilage, cleft palate, external ear deformities, cervical rib and urinary tract anomalies¹⁶.

Conclusion:

Occipitalization of atlas remains asymptomatic for first 2 decades of life. Future scope of this study lies in the field of orthopaedics, kinesiology and pain management as these manifestations may lead to chronic pain and movements at the atlanto-occipital joint may be restricted partially or completely which may indirectly lead to disfigurement of the facial and neck region. Also this knowledge of such fusion may be of importance for radiologists, anaesthesiologists, and neurosurgeons because skeletal abnormalities at the cranio-cervical junction may result in sudden unexpected death.

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