

## CASE REPORT

# OCCIPITALIZATION OF ATLAS

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### ABSTRACT

Occipitalization of atlas is an osseous anomaly of the craniovertebral junction which occurs at the base of the skull in the region of the foramen magnum. The knowledge of such a fusion is important because skeletal abnormalities at the craniocervical junction may result in sudden death. During bone cleaning procedure and routine undergraduate osteology teaching, three skulls with Occipitalization of atlas were encountered in the department of Anatomy at MMIMSR, Mullana, India. In one skull, both anterior and posterior arch were completely fused with occipital bone while the transverse process on the right side was not fused whereas left transverse process was fused with occipital bone. Both anterior and posterior arch were completely fused whereas transverse process on both sides were not fused in other skull. In another skull, partial and asymmetrical Occipitalization of atlas vertebra with occipital bone was found with bifid posterior arch of atlas at the level of posterior tubercle. Anterior arch was completely fused with basilar part of occipital bone but both the transverse processes were not fused. Reduced diameter of foramen magnum due to the atlanto-occipital fusion might cause neurological complications due to compression of spinal cord or medulla oblongata, vertebral vessels, 1st cervical nerve, thus, knowledge of occipitalization of the atlas may be of substantial importance to orthopaedicians, neurosurgeons, physicians and radiologists dealing with abnormalities of the cervical spine.

**KEY WORDS:** Occipitalization, Atlas, Assimilation, Occipital bone, Atlanto-occipital fusion.

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### Access this Article online

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DOI: 10.16965/ijar.2014.549

**Web site:** International Journal of Anatomy and Research  
ISSN 2321-4287  
[www.ijmhr.org/ijar.htm](http://www.ijmhr.org/ijar.htm)

Received: 29 Nov 2014

Peer Review: 29 Nov 2014 Published (O):31 Dec 2014

Accepted: 13 Dec 2014 Published (P):31 Dec 2014

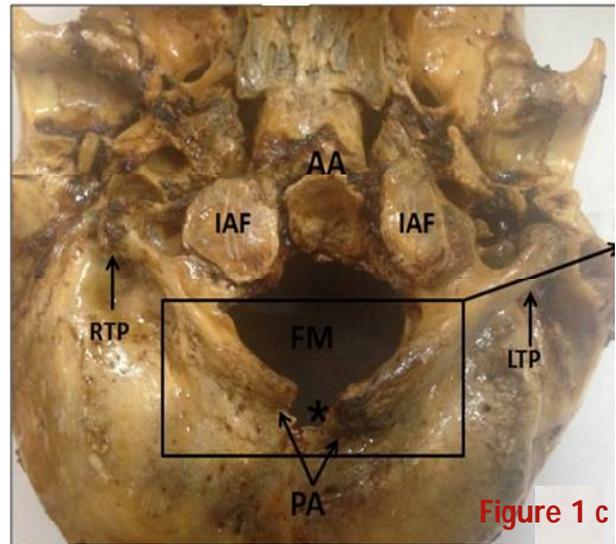
### INTRODUCTION

Occipitalization of the atlas or occipito cervical synostosis is fusion of the atlas with the base of occiput. It is one of the most common skeletal abnormalities at the craniovertebral junction [1]. The Atlas vertebra partially or completely fuses with the occipital bone. Complete fusion is more common while multiple variations of partial Occipitalization have been reported and may involve any aspect of atlanto-occipital articulation. This condition is first, described by Rokitansky in 1844 and Schuller in 1911 demonstrated this anomaly roentgenographically [2].

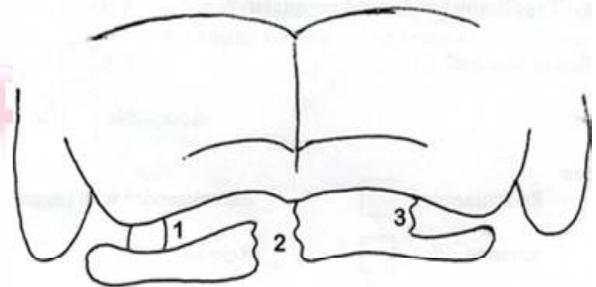
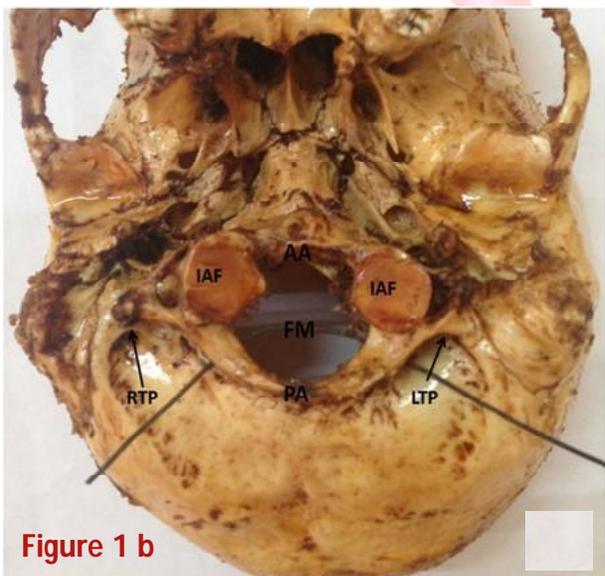
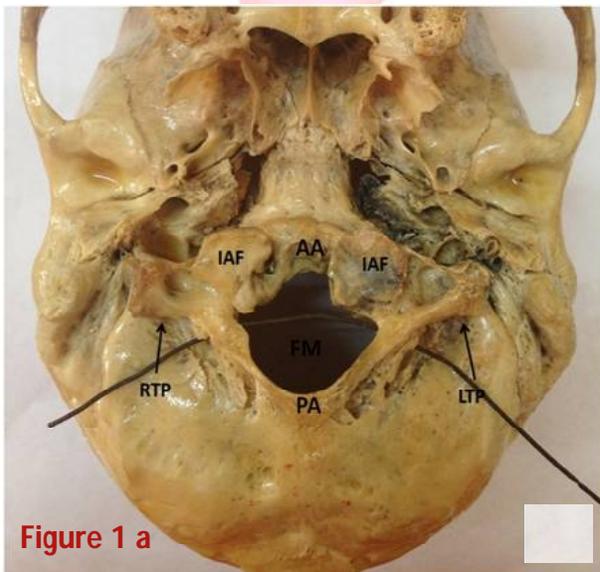
Atlantal Occipitalization may be congenital or acquired. Because of its proximity to the spinomedullary region, congenital malformation of the cervico-occipital region is of considerable consequence as it can produce wide range of symptoms of neurovascular and skeletal conditions [3]. Neurological signs and symptoms associated with Occipitalization vary from transitory headache to neurological syndrome. Occipitalization also results in cervical cord compression due to soft tissue and bony abnormalities which can lead to weakness and ataxia of the lower extremities while numbness and

pain in the upper extremities. Apart from this vertebral artery compression or even its total occlusion in the bony canal leading to dizziness, seizures, mental deterioration and syncope has also been reported [4]. However, an assimilation of atlas is found incidentally when other diseases are being ruled out as it is mostly asymptomatic. Thus, due to its multiple variations knowledge of such anomaly is essential for orthopedic, surgeon, anesthetist, and clinician. We encountered three skulls with occipitalization of atlas during bone cleaning procedure and during routine dissection.

**Fig. 1:** Occipitalization of Atlas. (a & b) showing complete fusion of anterior arch and posterior arch of atlas vertebra with occipital bone. (c) Illustrates partial and asymmetrical assimilation of atlas with occipital bone with bifid posterior arch. Anterior arch was completely fused with basilar part of occipital bone. Probe showing foramen on the superior surface of posterior arch.



AA- Anterior Arch of the Atlas, IAF- Inferior articular facets of the atlas, FM- Foramen Magnum, PA- Posterior Arch of the Atlas, RTP- Right Transverse Process, LTP- Left Transverse Process, \* Bifid Posterior Arch.



**CASE 1 (Figure: 1a)**

This skull showed completely fused atlas vertebra with the occipital bone at the base of the skull, except at the transverse processes on both sides. The anterior arch was fused with the basilar part of the occipital bone leaving a gap in the synostosed part. The whole of the posterior arch was synostosed with the posterior rim of foramen magnum. The hypoglossal canals were present. The two superior facets of atlas had inseparably fused with the occipital condyles. The right and the left transverse process of the atlas each containing complete transverse foramen were not fused with the occipital bone. The left foramen transversarium was larger than the right one. The skull showed foramen on the superior surface of posterior arch on both sides. The inferior articular facet on the right side (17X17mm) appeared to be larger than left (14X16mm) and facets were smooth and flat. The anterior aspect of the foramen magnum was minimally decreased by the right and left lateral mass of the atlas.

**CASE 2 (Figure: 1b)**

Atlas vertebra was completely fused with the occipital bone at the base of the skull, except the right transverse process. In this skull both the anterior and the posterior arches were completely fused with the occipital bone but there was small non synostosed part between anterior arch and basilar part of occipital bone. Hypoglossal canals were normal on both sides. Both left and right superior articular facets were fused with occipital condyles. The skull showed foramen on the superior surface of posterior arch on both sides. Correspondingly, there was an incomplete fusion of the transverse process of the atlas in the right side. The transverse processes were having complete foramen transversarium. The right articular process (17X14mm) was larger than left (15X15mm). Both right and left lateral mass protruded into foramen magnum making it smaller in shape.

**CASE 3 (Figure: 1c)**

Partial and asymmetrical occipitalization of atlas vertebra with occipital bone was found. The skull showed midline defect in posterior arch of atlas because of failure of fusion of right and left halves of posterior arch. Distance from posterior margin of foramen magnum to inferior margin of left posterior arch is more(11mm) whereas right posterior arch is completely fused and distance from posterior margin of foramen magnum to inferior margin of fused atlas is 7mm. Anterior arch was completely fused with basilar part of occipital bone. Both the transverse process were not fused. There was incomplete foramen transversarium on left side. The skull showed foramen on the superior surface of posterior arch only on right side. Hypoglossal canal were normal on both sides. Both condylar facets were completely fused with superior articular facet of atlas. Inferior articular facet of right side(16X21mm) was circular in shape whereas on left side(21X10mm) was oval. Lateral mass of right side was protruding into foramen magnum.

**DISCUSSION**

Occipitalization of atlas is an osseous abnormality occurring in the base of skull in the

region of foramen magnum. In the development of basilar occiput and atlas, the rostral half of the first cervical sclerotome combines with the caudal half of the last occipital sclerotome to form the base of the skull. While the caudal half of the first cervical sclerotome combines with the rostral half of second cervical sclerotome to form 1st cervical vertebra and odontoid process. In small number of cases the disruption of this merging process may result in atlanto-occipital fusion [5]. Prevalance of occipitalization in Asian countries has been found to be varying from 0.32%-2.86%.

Affected individuals may have the following physical features: low hairline, torticollis, restricted neck movements, and an abnormally short neck [6]. Absolute immobility of an occipitalized atlanto occipital joint results in compensatory hyper mobility of atlas on the axis due to exerting stress on occipitodontoid ligament with over stretching [7].

The sagittal diameter of the foramen magnum is an important parameter in spinal cord compression. The atlanto-occipital fusion may reduce the lumen of the foramen magnum and lead to neurological complications due to compression of the spinal cord. The standard dimensions for foramen magnum range between 28-38mm for the sagittal diameter and between 25-40 mm for the transverse diameter [2]. In present case, lateral mass has been seen protruding into foramen magnum making it irregular in shape but the sagittal and transverse diameters have remained within normal range. The protruding lateral mass in foramen magnum causes reduced diameter which may compress vertebral artery influencing the blood flow to the brain resulting in syncope, seizures, dizziness, vertigo and neurological symptoms which may not develop until second decade of life due to a gradual increasing degree of ligamentous laxity and instability with aging [1]. It may also result in compression of 1<sup>st</sup> cervical nerve affecting the sub occipitalis muscle which are postural muscles, thus, giving rise to an abnormal posture of head and an unsteady gait [8].

Although asymptomatic in early age, but neurological symptoms usually occur in third and fourth decades and vary depending on the area of spinal cord impingement. There may be

anterior compression of medulla oblongata leading to long tract signs such as dysfunction to the lateral corticospinal tract, may be present in the upper and lower extremities in the form of hyper reflexia, spasticity, Hoffman's sign, and Babinski's sign. The impingement of posterior column due to presence of dural bands, posterior column signs and symptoms [Paresthesiae, numbness, impairment of 2 point discrimination and vibration and conscious proprioception impairment] predominate [9]. The headaches associated with this condition are characterized as dull and aching, and are located over the posterior two thirds of the skull. The headaches may be precipitated by coughing or by neck movements. Cranial nerve findings associated with occipitalization of the atlas include tinnitus, visual disturbances, auditory disturbances, lower cranial nerve palsies leading to dysphagia, dysarthria and nystagmus [6].

Occipitocervical synostosis is associated with other skeletal malformations such as Spina bifida of atlas, basilar invagination, cervical stenosis, Klippel-Feil syndrome (fusion of the second and third cervical vertebrae) and Arnold Chiari I malformation [5]. An important aspect of atlantooccipital fusion is that the neurological symptoms and signs cannot be distinguished from those of the Arnold Chiari malformation as the pathophysiology of both is essentially the same [10].

In 3<sup>rd</sup> skull, there was incomplete assimilation leading to difference between posterior margin of foramen magnum and inferior margin of posterior arch of atlas on right and left side which could be the possible cause for right and left asymmetry in length of vertebral column, hence, causing clinical problems. Hence, it is of importance, is that skilled diagnostician should be aware that such an anomaly may exist without any typical clinical presentation. Restriction or absence of movement in atlantooccipital assimilation may be the first sign to attract the attention of surgeons regarding assimilation. The knowledge of assimilation may be of

importance to orthopedic surgeons dealing with the pathologies of upper cervical spine. It may cause anesthetic problems like failure of cisternal puncture. This condition may also be of importance to physiotherapist dealing with the neck pain and radiologist dealing with abnormalities of cervical spine [10]. Transverse process is very important landmark for head and neck surgeons, when it is inclined and fused to occipital bone, there may be difficulty in reaching various structures and may led to asymmetry in structure and shape of apertures for the vessels and nerves around the foramen magnum. Therefore, the knowledge of such anomaly is essential.

**Conflicts of Interests: None**

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### How to cite this article:

Sween Walia , Bhawani Shankar Modi, Nidhi Puri, VV Gopichand Patnaik. OCCIPITALIZATION OF ATLAS. *Int J Anat Res* 2014;2(4):781-784. DOI: 10.16965/ijar.2014.549