

Occipitalization of the Atlas - A Case Report

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Abstract: *Occipitalization is a congenital synostosis of the atlas to the occiput, which is a result of failure of segmentation and separation of the most caudal occipital sclerotome and the first cervical sclerotome during the first few weeks of fetal life. Occipitalization of atlas is associated with abnormalities as a result of narrowing of the foramen magnum, compressing the spinal cord or the brain stem. However, this anatomical variation may often go unnoticed. One of the human skulls of Indian origin collected from Department of Anatomy, DM – wayanad Institute of Medical Sciences, Wayanad Kerala, India, showing occipitalization of Atlas. Although atlanto-occipital fusion is a congenital condition, many patients do not develop the symptoms until the second decade of life. This may be due to a gradual increasing degree of ligamentous laxity and instability with aging.*

Keywords: Atlas, Foramen magnum, Occipitalization, Skull, Variation

1. Introduction

Craniovertebral abnormalities have been recorded for many years in morphological and clinicoradiological studies. Occipitalization is a congenital synostosis of the atlas to the occiput, which is a result of failure of segmentation and separation of the most caudal occipital sclerotome and the first cervical sclerotome during the first few weeks of fetal life[1]. Occipitalization of the atlas or atlanto- occipital fusion is one of the most common osseous anomalies of the craniovertebral junction. According to Yochum and Rowe[2] occipitalization represents the most cephalic 'blocked' vertebra encountered in the spine.

There may be varying degrees of bony fusion between atlas and occiput; complete and partial assimilation have been described[3,4]. In a majority of cases, assimilation occurs between the anterior arch of the atlas and the anterior rim of the foramen magnum and is associated with other skeletal malformations. Occipitalization of atlas is associated with abnormalities as a result of narrowing of the foramen magnum, compressing the spinal cord or the brain stem[5,6]. However, this anatomical variation may often go unnoticed.

2. Case Report

One of the human skull of Indian origin collected from Department of Anatomy, DM – wayanad Institute of Medical Sciences, Wayanad Kerala, India, showing occipitalization of Atlas was observed the lateral and both anterior and posterior arches were completely fused with the occipital bone. In addition the transverse processes of the atlas were also fused to the occipital bone (Fig-1,2,3).



Figure 1: Anterior view



Figure 2: Posterior view



Figure 3: Lateral view

3. Discussion

Fusion, either partial or complete, of the atlas with the occipital bone may occur in about 1% of cases so-called atlas assimilation [7]. The ventral portion of the sclerotome surrounds the notochord and provides the form, which develops into the vertebral body. The dorsal portion

surrounds the neural tube and provides the form, which develops into the posterior vertebral arch. The caudal half of each sclerotome combines with the rostral half of the sclerotome below it. 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 the second cervical sclerotome to form the first cervical vertebra, the pattern continues in this fashion to form the other vertebrae[8,9]. In a small number of cases, the disruption of this merging process may result in atlanto-occipital assimilation. This condition may be partial or complete, as was the cases here. The complete fusion of the atlas is more common than the incomplete [10].

Neurological symptoms associated with occipitalization of the atlas are attributed to the laxity of the transverse ligament about the odontoid process caused by repeated flexion and extension of the neck leading to the compression of the spinal cord or actual indentation of the medulla oblongata. Although atlanto-occipital fusion is a congenital condition, many patients do not develop the symptoms until the second decade of life. This may be due to a gradual increasing degree of ligamentous laxity and instability with aging.

Hensinger RN [11] reported that the onset of clinical symptoms can be sudden and precipitated by relatively minor trauma, the most common course is a progressive but sudden onset or instant death. Lopez et al[12] reported that three patients with atlanto-occipital fusion have had cervical pain and two patients had tonic or clonic convulsions. Iwata et al[13] reported a case of atlanto-occipital fusion with unusual neurological symptoms. The neurological symptoms and signs of atlanto-occipital fusion can not be distinguished from those of the Arnold Chiari malformation as the pathophysiology of both is essentially the same. Fusion between atlas and occiput occurs anteriorly between the arch and rim of the foramen with some segment of the posterior arch of C1 present in some instances. This fragment can frequently constrict the spinal canal causing intermittent symptoms depending on the position of the head[14].

4. Conclusion

Occipitalization of the atlas may cause both orthopedic problems and occasionally induce neurological effects. This happens particularly when the lumen of the foramen magnum is constricted by the abnormally ossified parts of the atlas. Therefore, improved knowledge on the fusion of the atlas with the occipital bone is important for medical practice. The future scope of the study lies in the field of orthopedics, 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. Further regional studies should be done to find out the environmental and racial factors which may lead to these complications.

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