



## CASE STUDY

### OCCIPTOCERVICAL SYNOSTOSIS AND ITS CLINICAL SIGNIFICANCE –A CASE REPORT

\*Dr. Varalakshmi K. L., Dr. Padmavathi G., Dr. Roopashree Ramakrishna and Dr. Suman Tiwari

Department of Anatomy, MVJ Medical College and Research Hospital, Bangalore, India

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#### ABSTRACT

Craniovertebral junction shows the presence of various anatomical variations and abnormalities. The present case report describes a case of fusion of superior articular facets of atlas vertebra with the occipital condyles, observed during the routine osteology classes for undergraduate medical students of MVJ Medical College and Research Hospital, Bangalore. Such type of complete or partial fusion of atlas vertebra with occipital bone is known as occipitocervical synostosis. Occipitalisation of atlas is associated with various neurological signs and symptoms ranging from mild headache to sudden unexpected death due to the narrowing of foramen magnum compressing the spinal cord or brain stem.

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#### INTRODUCTION

Occipitocervical synostosis is a congenital malformation characterized by a partial or complete union between the atlas vertebra and occipital bone of the Skull. It also is known as “occipitalisation of the atlas” or “assimilation of the atlas” (Ranade *et al.*, 2007). The fusion ranges from a total incorporation of the atlas into the occipital bone to a bony or fibrous band uniting one small area of the atlas to the occiput. (Paramesh *et al.*, 2013) It is a most common osseous anomaly in the cervicooccipital region and is of great clinical importance. Occipitalisation occurs in 1 in 109 adult skulls of Asian origin. It leads to various neurological signs and symptoms due to the narrowing of foramen magnum and compression of brain stem, cervical part of spinal cord and vertebral artery as these structures pass through foramen magnum. (Sani *et al.*, 2009) Hence the detailed knowledge of occipitocervical synostosis is important for neurosurgeons, neurologists and radiologists to prevent and avoid possible complications and achieve best operative results as well as diagnostic interventions.

#### Case report

During the routine osteology demonstration class for 1st MBBS students of MVJ Medical College and Research Hospital, Bangalore, we noticed a skull with occipitalisation of atlas vertebra. The bone was thoroughly studied, photograph was taken (Figure 1) and following observations were made.

Superior articular facets of atlas were completely fused with the occipital condyles. Anterior arch of atlas was partially fused with basiocciput leaving a gap between the two bones. The posterior arch of atlas showed complete non fusion with occipital bone and to certain extent it has encroached on the foramen magnum dividing the foramen magnum into two compartments. The fusion was not exactly in the midline, slightly inclined to the right side. Transverse processes were normally developed and free from the base of the skull. Foramen transversarium was normal on both the sides. The hypoglossal canal was in its normal position.

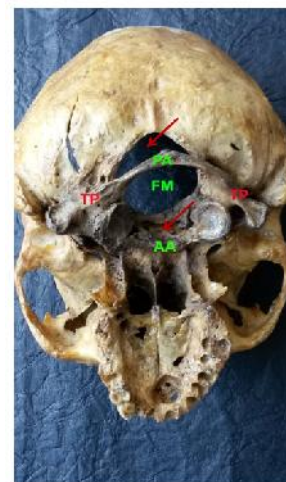


Fig.1. Photograph of the base of the skull showing the fusion of superior articular facets of atlas vertebra with occipital condyles. An Arrow mark indicates partial fusion between the anterior arch and non-fusion of posterior arch with the occipital bone

\*Corresponding author: Dr. Varalakshmi K. L. Department of Anatomy, MVJ Medical College and Research Hospital, Bangalore, India.

AA-anterior arch of atlas,  
 PA-posterior arch of atlas,  
 FM-foramen magnum,  
 TP-transverse process of atlas

## DISCUSSION

Occipito cervical synostosis is a congenital condition caused due to the faulty development during the 1<sup>st</sup> week of fetal life. It occurs due to the failure of segmentation and separation of the caudal occipital and 1<sup>st</sup> cervical sclerotome. (Sangeeta *et al.*, 2012). Multiple variations of partial fusion of atlas with occipital bone have been reported and many involve some aspect of atlanto-occipital articulation. (Khamanarong *et al.*, 2013). The study on assimilation of atlas was first described by Rokintansky in 1844 and later by Schuller in 1911 on roentgenographically. (Al –Motabagani and Surendra). Surekha. Yadhav *et al.* conducted study on 150 adult skulls and observed a single case of assimilation of atlas where the superior articular facets of atlas were fused with occipital condyles. The anterior arch had incompletely fused and posterior arch and right transverse process were completely fused with occipital bone. (Jadhav *et al.*, 2012). Jayanthi *et al.* have observed two cases of occipitalisation associated with spina bifida atlas in one (Jayanthi *et al.*, 2003). Tun and his colleagues reported a case in which CT images revealed a fusion between the posterior arch of atlas and occipital bone. It also showed the presence of hypertrophy of occipital condyles with reduction in the transverse diameter of foramen magnum (Tun *et al.*, 2004). According to Mc Raes and Barnon the patients of occipitocervical synostosis may have the following clinical features: spasticity, hyperreflexia, muscular weakness, wasting due to the lesion of pyramidal tract. Vertigo, syncope, seizures, unsteady gait due to the compression of vertebral artery compromising the blood flow to brain. The cranial nerve lesions results in tinnitus, visual disturbances, dysphagia and dysarthria (Mc Rae and Barnon 1953). The signs and symptoms of occipitocervical synostosis is similar to Arnold chiari malformation as pathophysiology of both are identical. (Nayak *et al.*, 2005) Even though atlanto occipital fusion is a congenital condition, signs and symptoms usually occurs during the second decade of life. The onset of symptoms can be sudden and precipitated by minor trauma. (Hensinger 1986) Greenberg *et al.* concluded that the spinal cord compression occurs when the sagittal spinal canal diameter behind the odontoid process is less than or equal to 14 mm. Cord compression is possible when the sagittal canal diameter is between 15 and 17 mm and it is almost never happens at a distance of 18 mm or more (Saheb *et al.*, 2010). In the present case the posterior arch of atlas covers the foramen magnum and divides it into two parts. This will not only compress the structures but also changes the sequence of passage of structures through the osseous and neurovascular compartments of foramen magnum.

## Conclusion

Improved knowledge of occipito cervical synostosis is important for clinicians, surgeons, radiologists as it reduces the dimensions of foramen magnum and leads to neurological complications due to the compression of important neurovascular structures. The head and neck surgeons should be aware that occipito cervical synostosis may exist without much clinical symptoms and may leads to sudden unexpected death.

## REFERENCES

- Al –Motabagani MA, Surendra M. Total occipitalization of the atlas vertebra. *Anatom Scien Inter* 2006; 81:173-180.
- Hensinger RN. Osseous anomalies of cranio vertebral junction. *Spine* 1986; 11:323-333.
- Jadhav SD *et al.* Assimilation of atlas in Indian dry skulls. *JKIMSU* 2012; 1(1):102-106.
- Jayanthi V, Kulkarni R, Kulkarni R N. Atlanto –occipital fusion: Report of two cases. *J.Anat.Soc. India* 2003; 52(1): 71-73.
- Khamanarong K *et al.* Occipitalization of the atlas:Its incidence and clinical implications. *Acta Medica Academica* 2013; 42(1):41-45.
- Mc Rae DL, Barnon AS. Occipitalisation of the atlas. *American Journal Roentgenology* 1953; 70:23-45.
- Nayak S, Vollala VR, Raghunathan D. Total fusion of atlas with occipital bone: A case report. *Neuroanatomy* 2005; 4:39-40.
- Paramesh RM *et al.* An osteological study of occipitocervical synostosis: Its embryological and clinical significance. *Journal of Clinical and Diagnostic Research* 2013; 7(9):1835-1837.
- Ranade AV, Rai R, Prabhu LV, Kumaran M, Pai MM. Atlas assimilation: A case report. *Neuroanatomy*. 2007;(6):32-3.
- Saheb HS *et al.* Occipitalization of atlas:a case report.*J Biomed Sci and Res* 2010; 2(2):73-75.
- Sangeeta RJ, Ila SM, Jitendra RK. An unusual case of unilateral atlanto occipital assimilation with skullasymmetry. *National Journal of Medical research* 2012; 2(20):238-240.
- Sani V, Singh R, Bandopadhyay M, Tripathy S, Shamal S. Occipitalization of the atlas: its occurrence and embryological basis. *IJAV* 2009; 2:65-68.
- Tun K, Okutan O, Kaptanoglu E, Gok B, Solaroglu I, Beskonakli E. Inverted hypertrophy of occipital condyles associated with atlantooccipital fusion and basilar invagination: A case report. *Neuroanatomy* 2004; 3:43-45.

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