OCCIPITOCERVICAL SYNOSTOSIS AND ITS CLINICAL SIGNIFICANCE – A CASE REPORT

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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.

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 passes 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.

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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 1st week of fetal life. It occurs due to the failure of segmentation and separation of the caudal occipital and 1st 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. Yadav 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 Barnon1953). 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**