Cervical Vertebra Synostosis (C2-C3) - A Case Report

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Received June 22, 2014; Revised July 02; Accepted July 07, 2014

Abstract Skeletal abnormalities of cervical region or in craniocervical region are of interest to the anatomists, orthopaedicians, neurologists, neurosurgeons and even orthodontists. These abnormalities may result in severe neck pain, decreased neck mobility, muscular weakness and sensory deficits of both upper limbs and sudden unexpected death. During the routine osteology teaching, it was observed that the axis vertebra is fused with the third cervical vertebra. It was observed that body, laminae and pedicles of C2 and C3 were completely fused on both anterior and posterior aspects. The features of these fused cervical vertebrae were analysed and the specimen was photographed from different aspects. This is a condition of block vertebra which has embryological importance and clinical implications.

Keywords: blocked vertebra, fusion, cervical vertebrae

Cite This Article: Yogesh Yadav, Preeti Goswami, and Veena Bharihoke, "Cervical Vertebra Synostosis (C2-C3) - A Case Report." Cervical Vertebra Synostosis (C2-C3) - A Case Report, vol. 2, no. 6 (2014): 120-122. doi: 10.12691/ajmcr-2-6-3.

1. Introduction

Cervical vertebrae are seven in number. C3-6 are typical whereas C1 (atlas), C2 (axis) and C7 (cervicae prominens) are atypical.C2 vertebrae is different from other by the presence of Dens (odontoid process), which projects cranially from the superior surface of the body. The axis acts as an axle for rotation of atlas and head around the dens [1]. The C3 vertebra is typical with the features similar to other cervical vertebrae. Congenital anomalies at craniovertebral or cervical region are common [2] Among these congenital anomalies the important ones are the fused cervical vertebrae (FCV).Congenital fusion of axis with the third cervical vertebrae limits the movements between these bones and because of this the third vertebrae was given the name as "vertebrae critica" by Cave [3]. As sacrum is a well known example of block vertebrae which is formed by the union of 5 sacral vertebrae, if we follow same principle for C2-C3 fused vertebrae, it can be given the name of cervical sacrum. This fusion of C2 with C3 may be associated with neurological signs and symptoms. Severe neck pain and sudden unexpected death may occur due to these abnormalities [4].

2. Case Report

During the osteology demonstration of cervical vertebrae it was observed that the axis was fused with 3rd cervical vertebra. The features of this fused cervical vertebrae was analyzed and the specimen was photographed from different aspects. Body, laminae,

spines, pedicles and transverse processes of C2 and C3 were completely fused on both anterior and posterior aspects. Foramen transversarium were present on both the vertebrae (Figure 1 - Figure 3).



Figure 1. anterior view



Figure 2. lateral view



Figure 3. posterior view

3. Discussion

Fused cervical vertebrae (FCV) have clinical and embryological importance. The anomalies of cervical region are of interest to anatomists, orthopedists, neurologists, neurosurgeons and even orthodontists. In FCV two vertebrae appear both structurally and functionally as one. This fusion may be congenital or acquired [5]. The orthodontist may be the first person to detect cervical spine abnormalities as they are asymptomatic until adolescence or young adulthood. Early diagnosis is based on incidental radiographic findings.

It is important to identify the cause of FCV – whether it is congenital or acquired or pathologic condition. Acquired FCV is generally associated with diseases like tuberculosis, other infections, juvenile rheumatoid arthritis and trauma. Clinical symptoms may vary from asymptomatic to myelopathy, limitation of the neck movement, or the muscular weakness, atrophy and neurological sensory loss or associated with Klippel-Feil syndrome [5]. Congenital FCV is one of primary malformations of chorda dorsalis [6,7].

All these abnormalities may lead to clinical signs and symptoms which are: Shortening of spine in the cervical region; The trapezei are unduly prominent laterally and give a webbed appearance; Limited neck motion; Osseous malformation (scoliosis, kyphosis, torticollis); Signs of peripheral nerve irritation such as pain, burning sensations and cramps; Signs of nerve compression such as hypoaesthesia/anaesthesia, weakness/paralysis, fibrillations and reduced deep reflexes [4].

These are thought to be due to defects which take place during development of occipital and cervical somites [8]. A combination of environmental and genetic factors mainly during the third week after conception is thought to be the main causative factor for this anomaly [9]. According to Farman and Escobar, the radiographic appearance of congenital anomalies of the vertebral bodies may be due to defects in fusion or normal segmentation occipitalization of the atlas, odontoid and atlas malformations, spina bifida and abnormal ossifications. There is increased incidence of osteophyte formation in the adjacent levels in cervical fusion. Up to 70% of occipitalizations have an accompanying C2–C3 fusion with instability at the C1–C2 articulation [10].

Early diagnosis of these anomalies will be helpful in documenting the change due to an injury, aging, or

progression of a degenerative process and also motivates the patients to change their lifestyles to lead a normal life. Surgical intervention for block vertebra carries a higher risk of morbidity and mortality. On the contrary, cervical vertebral fusions are commonly done following traumatic, degenerative and inflammatory dislocations of upper and lower cervical spine [9]. The awareness and monitorization of complications like anesthetic risk during intubation with neck extension needs to be considered. Appropriate counseling on the management of known risk factors, including therapeutic options that can precipitate complication should be encouraged (like avoiding undue trauma, extension and rotational maneuvers which may place the spinal cord and vertebral artery at risk) [10].

3.1. Embryological Significance

Somites are formed from para-axial mesoderm that lies on each side of neural tube. The somites are divided into three parts: Ventromedial sclerotome; Intermediate myotome; and Lateral dermatome. The vertebral column is formed from the sclerotome of the somites. The body, posterior arch and transverse process of C2 vertebra is derived from second cervical sclerotome, tip of dens is derived from cranial half of 1st cervical sclerotome [11]. Normal segmentation of the sclerotomes is important for the development of a normal vertebral column. But in certain cases due to decreased local blood supply during the third to eight week i.e. embryonic period results in abnormal segmentation and formation of congenitally fused vertebrae or block vertebrae. Vertebral fusion anomalies are likely to be associated with disturbance of Pax-1 gene expression in the developing vertebral column [12]. Congenital fusion of vertebrae is most commonly seen in the cervical region although it may involve other segments of vertebral column also. Sacrum itself is an example of block vertebrae [13]. The incidence of CFCV of C2-C3 is around 0.4% to 0.7% with no sex predilection [10] The location of block vertebra in order of frequency are cervical spine (C2-3, C5-6), lumbar spine (L4-5) and thoracic spine (any section) [14].

Block vertebra results from embryological failure of normal spinal segmentation due to decrease in local blood supply during the third to eighth week of fetal development. The commonly encountered anomaly is block vertebrae [15].

According to recent report long-standing congenital or acquired fusion of upper cervical vertebrae may lead to stretching and laxity of the ligaments between the occiput and the atlas, resulting in excessive motion and brainstem or cord compression [16]. Cervical vertebrae fusions are sometimes done following traumatic, degenerative and inflammatory dislocations of upper and lower cervical vertebrae [9]. The presence of block vertebra results in a greater biomechanical stress in the adjoining segments leading to premature degenerative changes at adjoining motion segments. The other common consequences are vertebral disc tear, rupture of the transverse ligament, fracture of the odontoid process, and spondylosis [10]. Embryological vascular disorders like subclavian artery supply disruption sequence have been hypothesized to result in Klippel-Feil syndrome [10].

The radiologic appearance of congenital anomalies may be due to defects in fusion or normal segmentation, occipitalization of the atlas, odontoid and atlas malformations, spina bifida and abnormal ossifications. In about 70% of occipitalizations, we see an associated C2-C3 fusion with instability at the C1-C2 articulation [10]. If these anomalies are diagnosed early, they will help us in finding the change due to an injury, ageing or progression of a degenerative process and also motivates the patient to change their lifestyles to lead a normal life. These anomalies like block vertebrae cannot be treated surgically as they carry a higher mortality and morbidity.

3.2. Clinical Significance

While doing endotracheal intubation, extension of the neck is done. So in persons with block vertebrae in cervical region we have to take care to prevent hyperextension as it can precipitate disc prolapsed. If cisternal puncture or lumbar puncture is to be done, we should look for possibility of block vertebrae in cervical and lumbar regions respectively.

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