Indian Journal of Medical Case Reports ISSN: 2319–3832(Online)
An Open Access, Online International Journal Available at http://www.cibtech.org/jcr.htm
2018 Vol.7 (2) April-June, pp. 1-5/Budhiraja et al.
Case Report

BLOCK VERTEBRAE IN LOWER CERVICAL SPINE- A RARE CADAVERIC FINDING

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ABSTRACT
Variations of vertebral column are frequently reported. One of the variation or anomaly of vertebral column is fusion of the two or more vertebrae, known as block vertebrae. This may be congenital or acquired, complete or incomplete. Here we present a rare fusion at the level of C5 & C6. Fusion of the Upper Cervical vertebrae has been easily accessible in the previous literature. However, fusion of vertebrae in lower cervical spine has not been frequently reported. Thus it is important to report such cases as the patient may present with muscular or neurovascular symptoms in the form of muscle weakness, limited neck movements, root irritation & spinal cord compression. Early diagnosis is helpful to improve the quality of life.

Keywords*: Cervical, Vertebrae , Fusion, Spine, Klippel- Feil Syndrome*

INTRODUCTION
The cervical spine is shaped primarily to impart flexibility, which is gained at the expense of stability (Maclister, 1869). Variations in the apophyseal articulations of any of the cervical vertebrae or fusion between them will produce irregular motion & at times indefinite instability of the joint involved (Overton and Grossman, 1952). Fusion of the cervical vertebrae is a condition in which two vertebrae appear not only structurally as one but also function as one (Dunsker *et al.,* 1980). This fusion may be congenital or acquired (Graaf, 1982; Resnick, 1992). Fusion may be complete or incomplete, involving the body of vertebra alone. In pathological conditions like Diffuse Idiopathic Skeletal Hyperostosis (DISH) or Ankylosing Spondylosis (AS), there will be ossification of paraspinal ligaments in addition to the fusion of body of vertebrae. There are many instances of fusion of the occipital bone with atlas & of atlas with axis i.e in the upper cervical spine. However, the fusion of vertebrae in lower cervical spine is rare according to previous literature. This anomaly may be asymptomatic or may be associated with syndromes such as Klippel- Feil (Graaf, 1982; Schlitt *et al.,* 1989; Nagashima *et al.,* 2001), limited neck movements (Bharucha & Dastur, 1964), muscle weakness, atrophy & neurological sensory loss (Kameyama *et al.,* 1993). Thus, early diagnosis & treatment of this anomaly for preventing any serious damage such as osteoarthritis is important.

CASE
On examining the vertebral column of a 60 year old male cadaver in the Department of Anatomy, Kalpana Chawla GMC, Karnal, fusion between 5^{th} & 6^{th} cervical vertebrae was observed and photographed. Vertebrae were fused incompletely. Morphology of the fused vertebrae was as follows: (See Figures 1, 2, 3).

1. The undersurface of body of C5 was fused with the upper surface of C6.
2. Superior articular processes, Transverse processes & pedicles of both vertebrae were not fused on either side.
3. Inferior articular process of C5 was fused with the Superior articular process of C6 on right side whereas on left side, these were normal.
4. Laminae were fused with each other.

5. Spinous processes were not fused. The spinous process of C5 was in midline, however the spinous process of C6 was deflected to left side.

Rest of the vertebrae of this cadaver were found to be normal.

Figure 1: Anterior View of Fused Cervical Vertebrae

Figure 2: Posterior View of Fused Cervical Vertebrae

Figure 3: Lateral View of Fused Cervical Vertebrae

DISCUSSION

Fusion of two vertebrae also called as Block vertebra has been reported by Barclay Smith (1910), Cave (1938), Gray *et al.,* (1964), Gunderson *et al.,* 1967) & others. The fusion may be congenital or acquired (Graff, 1982, Resnick, 1992). This anomaly may be asymptomatic; however, it may also appear with manifestations of serious clinical features such as myelopathy or may be associated with syndromes such as Klippel-feil (Graff, 1982; Schlitt *et al.,* 1989; Nagashima *et al.,* 2001), limitation of neck movement (Bharucha & Dastur,1964) or the muscular weakness, atrophy & neurological sensory loss (Kameyama *et al.,* 1993). Klippel & Feil (1912) described a syndrome with congenital fusion of cervical vertebrae. In 1919, Feil classified this syndrome into 3 types based on the extent & the type of cervical fusion (Fielding *et al.,* 1986).

Type 1: Massive fusion of many cervical & upper thoracic vertebrae into bony blocks

Type 2: Fusion of only 1 or 2 interspaces, usually C2-C3 or C5-C6, but there can be intrafamiliar variability.

Type 3: Both cervical fusion & lower thoracic or lumbar fusion, often associated with multiple organ anomalies & subsequent neurological compromise.

Raas-Rothschild *et al.,* (1988) suggested a type 4 with an association of sacral agenesis & cervical fusion.

Thus, our fused vertebral specimen fall into Type 2 of above classification as rest of the vertebral column was found to be normal.

Ontogeny

In man, the development of vertebra occurs in three stages viz precartilage, chondrification & ossification. The neural arches proceed directly backward to a variable extent to form the pedicle, articular process & the lamina. The less dense tissue that separates the sclerotome segments develops into the intevertebral discs (William *et al.,* 2005).

The segmented pattern of human spine is established during embryogenesis when the somites, the epithelial spheres that contain the precursors of the vertebrae, are rhythmically added to the developing posterior part of the embryo. In humans, somite formation begins around the 3rd weak postfertilization & continues until a total of around 52 somites are formed. These somites divide into dermomyotomes dorsally, which give rise to muscles & dermis and sclerotomes ventrally which form the spine. Scrélotome further subdivides into an anterior & posterior compartment. This subdivision plays a key role in the definitive pattern of vertebrae that form when the posterior part of one somite fuses to the anterior part of the consecutive somite during a process termed as resegmentation. During the fourth week of embryonic development, the sclerotomes shift their position to surround the spinal cord & notochord. As the sclerotomes develops, it condenses further eventually developing into the vertebral body & the neural arches proceed directly backwards to a variable extent to form the pedicle articular process & the lamina (Romer *et al.,* 1977; Galis, 1999).

A fusion between the different vertebrae may be because of failure of the normal segmentation of cervical somites during 3rd to 8th week of gestation (Galis, 1999).

Genetics

Pedigree analysis suggests a human genetic locus for fusion of vertebrae especially in Klippel-Feil Syndrome. Fusion between C5 & C6 has been attributed to autosomal recessive inheritance. A dominant interference of Hox-1.1 transgene has been blamed for the craniofacial abnormalities. This interference with developmental programmes occur around Day 9 of gestation, that is the time of neural crest migration & somite differentiation (Kessel *et al.,* 1990).

Phylogeny

Evolutionary changes in the development of man have been accompanied by number of skeletal changes in the form of shortening of spinal column, broadening & flattening of all the spinal segments & inclusion of last lumbar vertebra into sacrum (Willis, 1929). Gupta (1966) studied fusion of cervical vertebrae in different species of rodents & found fusion of C2-C3 vertebrae in majority of *Dipodomys spectabilis spectabilis* & *Dipodomys spectabilis baileyi* (Gupta, 2009).

Clinical significance

Any deviation in the morphological characteristics of cervical spine can have catastrophic consequences. Mobility of cervical region is greater compared to other levels of vertebral column. Thus, fusion of cervical vertebrae affects day to day activity of patient because of the restricted neck movements. Absence of motion segment can cause stress in the free articulations above & below the block segment, resulting in premature degenerative spondylosis & arthrosis (Yadav, 2014). Patient may present with neurological symptoms due to root irritation & spinal cord compression. Major neurological sequelae include death or quadriplegia following minor trauma in high risk patients (Samartzis *et al.,* 2006). Further, fused cervical segments may be associated with developmental anomalies like stunted growth, non union of two halves of vertebral arch, low hair line, short neck etc (Nayak, 1931).

Conclusion

Fusion of cervical vertebrae may be associated with serious clinical manifestations such as such as myelopathy, limited neck movements or neurological symptoms. Thus, early diagnosis with the help of imaging techniques such as X-Ray, MRI may be helpful to prolong the normal life & to plan appropriate corrective surgical measures. Knowledge about the rare fusion of vertebrae is essential for clinical anatomists, radiologist & forensic medicine experts.

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