Fusion of axis with third cervical vertebra: a case report

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Abstract: *Introduction*: Fusion of one or more contiguous vertebral segments is usually the result of embryological failure of normal spinal segmentation. It may be associated with syndromes such as Klippel-Feil. Fused cervical vertebrae (FCV) may also be acquired or pathologic. FCV is generally associated with disease like tuberculosis, other infections, juvenile rheumatoid arthritis and trauma. The commonest site of involvement is C2-C3. In condition of fusion the two vertebrae appear not only structurally as one but also function as one. This anomaly may be asymptomatic; however, it may also manifest in the form of serious clinical features such as myelopathy, limitation of the neck movement, muscular weakness, atrophy or neurological sensory loss. *Case report*: We observed the fusion of axis with 3rd cervical vertebra. Body, laminae and spines of C2 and C3 were completely fused on both anterior and posterior aspects, whereas the pedicles and transverse processes were not fused. Foramen transversarium was present on both the vertebrae bilaterally. *Conclusion*: This variation is noteworthy to neurosurgeons and radiologists in studying computed tomography (CT) and magnetic resonance imaging (MRI) scans.

Key words: Axis, 3rd cervical vertebra, fusion, block vertebra, foramen transversarium, variation

Introduction

Cervical vertebrae are seven in number of which C3-6 are typical, sharing similar features whereas C1 (atlas), C2 (axis), and C7 (vertebrae prominence) are atypical having different characteristic features. (1) Vertebrae and intervertebral discs are one of the main manifestations of body segmentation or metamerism. (2) Developmental and ossification process of C2 vertebra is the most complex among all the vertebrae. (3)Fusion of one or more contiguous vertebral segments results from the embryological failure of normal spinal segmentation. The incidence is 0.4% to 0.7% with no sex predilection. (4) Recent studies have documented associations between fusion of the cervical vertebral column and craniofacial morphology,

including head posture in patients with severe skeletal malocclusions. This finding is expected to have importance for diagnostics and elucidation of etiology and thereby for optimal treatment. (5) Skeletal abnormalities of cervical region or in cranio-cervical region are of interest to the anatomists, neurosurgeons radiologists and even orthodontists. (6)

Block vertebrae, formed after fusion of adjacent vertebrae is a condition which has embryological importance and clinical implications. Since the fusion of cervical vertebrae 2nd with 3rd has a clinical importance, we need to emphasize the importance of multidisciplinary approach to help establish the precise occurrence of this congenital anomaly for preventing any serious damage such as osteoarthritis by early diagnosis and treatment.

Case report

During routine survey of bones in the Department of Anatomy, King George's Medical University, Lucknow, Uttar Pradesh, India, it was observed that the axis was fused with 3rd cervical vertebra. Features of this block vertebra were analyzed and the specimen was photographed from different aspects. Body, laminae and spines of C2 and C3 were completely fused on both anterior and posterior aspects. A ridge was noted on the dorsal surface of fused arches. Joints between the inferior articular facet of C2 and superior articular facet of C3 displayed synostosis bilaterally (Figures 1 and 2). Diameters of foramen transversaria were measured from supero-lateral aspect of C2 and inferior aspect of C3. The diameters of foramen transversarium of C2 on right and left side were 5.5 mm and 5.0 mm respectively while of C3 vertebra was 6.0 mm on right side and 6.5 mm on left side. The pedicles and transverse processes were not fused (Figure 3).

Figure 1 - Photograph showing complete fusion of body of C2 and C3 from anterior aspect

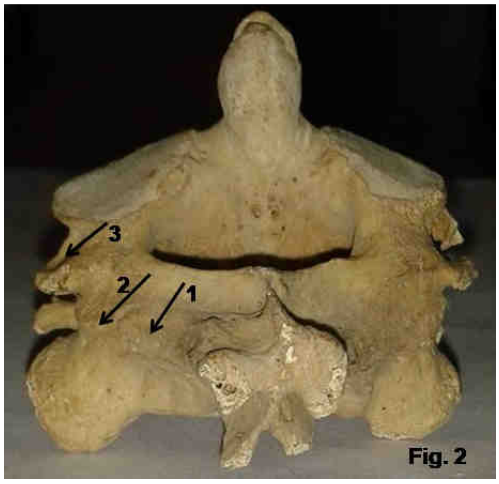

Figure 2 - Photograph showing complete fusion of laminae and spines of C2 and C3 from posterior aspect, presence of ridge on dorsal surface of fused arches (arrow 1), bilateralsynostosis between inferior articular facet of C2 and superior articular facet of C3 (arrow 2), foramen transversarium is seen in C2 vertebra superolaterally (arrow 3).

Figure 3 - Photograph showing non-fusion of pedicles and transverse processes of C2 and C3 in left lateral aspect

Discussion

In condition of the fusion of cervical vertebrae, two vertebrae appear not only structurally as one but also function as one. (7) It is important to identify the cause of FCV i.e. congenital, acquired or pathologic. Embryologically, C2-C3 fusion was explained as improper separation of adjacent somites or their associated mesenchyme. (8) Also it was found to be due to non-segmentation of primitive sclerotome.(9) In a condition known as chorda dorsalis, congenital fused cervical vertebra is one of the primary malformations believed to be due to defects during the development of the occipital and cervical somites. Between 3rd to 8th weeks of embryonic life, cartilaginous framework of a vertebra is formed from paraxial mesoderm. (10) Congenital fusion of two or more cervical vertebrae as seen in Klippel-Feil syndrome was believed to result from faulty segmentation along the embryos developing axis during weeks 3-8 of gestation. (11) Cause of this anomaly is often a combination of environment and genetics which occurs during the third week post-conception. It has been described as an autosomal dominant condition. Mutation in Pax gene and notch signaling pathway and a chromosomal inversion inv (8) (q22.2q23.3) causes familial Klippel-Feil syndrome. (9) This syndrome was also hypothesized to result from embryological subclavian artery supply disruption sequence. (12) It was suggested that decreased local blood supply during third to eighth week of development may be a causative factor4. Absence of the joints between articular facets in the fused vertebrae suggests failure of normal development and differentiation of vertebrae (i.e. fusion at the pre-cartilaginous stage of vertebral development). Independent pedicle, transverse process and ridge on dorsal surface of fused arches suggest normal initial development followed by fusion. Acquired FCV is generally associated with diseases like tuberculosis, other infections, juvenile rheumatoid arthritis and trauma. Cause of these abnormalities may be multifactorial which also includes the role of certain drugs like thalidomide, lovastatin and progestin/estrogen on the developing fetus.

The present case is very much similar to some previous studies in which the body, laminae and pedicles of C2 and C3 were completely fused on both anterior and posterior aspects. (1, 6) But in our case the pedicles were not fused while spines of the two vertebrae were fused on both anterior and posterior aspects. This anomaly may be

asymptomatic, may appear with manifestations of serious clinical features such as myelopathy, muscular weakness, atrophy and neurological sensory loss or may be associated with syndromes such as Klippel-Feil in the form of limitation of neck movement. (7) The orthodontist may be the first person to detect cervical spine abnormalities as they are asymptomatic until adolescence or young adulthood and early diagnosis is based on incidental radiographic findings. Symptoms vary according to the extent of pathology and may result in severe neck pain, decreased neck mobility, muscular weakness or sensory deficits of both upper limbs and sudden unexpected death. (6) Normally aligned congenital synostosis of C2-3 is rarely associated with a junctional problem, whereas a kyphoticsynostosis is associated with a caudal junctional problem.(13) Persons with Klippel-Feil syndrome and cervical stenosis may be at increased risk for spinal cord injury after minor trauma as a result of hypermobility of the various cervical segments. Cervical spondylosis, disc herniation and secondary degenerative changes are more at levels adjacent to fused vertebra. There is increased incidence of osteophyte formation in the adjacent levels in cervical fusion. Neurological signs and symptoms are variable, depending on the degree of pathology. Another syndrome associated with this anomaly is Wildervanck syndrome, characterized by Klippel–Feil (KF) deformity of the cervical spine in association with abducens palsy with retracted bulbi and hearing loss. The main clinical consideration of this deformity is a difficult airway due to the short, thick and immobile neck secondary to fused cervical segments, which compromises bag and mask ventilation if airway management is required. (14) Persons with Klippel-Feil syndrome may be at high risk for developing a transient neurolgoic deficit due to cervical spinal cord injury following minor trauma, as these subjects are particularly prone to hyperextension trauma.

Conclusion

Fusion of axis and third cervical vertebrae is a rare and unusual finding which is clinically significant. This variation is noteworthy to neurosurgeons and radiologists in studying computed tomography (CT) and magnetic resonance imaging (MRI) scans. It is of equal significance to the anaesthetists doing endotracheal intubation as in persons with block vertebra in cervical region hyperextension may precipitate disc prolapse. 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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References

1.Wazir S, Mahajan A. Fusion of axis with third cervical vertebrae- a case report. Indian Journal of Fundamental and Applied Life Sciences. 2011; 1(4): 164-166.

2.Standring S. Gray's Anatomy- The Anatomical Basis of Clinical Practice. 40thed. Churchill Livingstone; 2008.p 720-721.

3.Pate D. Normal Radiographic Differences between the Adult and Pediatric Cervical Spine.Dynamic Chiropractic.2012; 30(4).

4.Solan S. Fused Axis Vertebrae With A Third Cervical Hemivertebrae - A rare case report.IOSR Journal of Dental and Medical Sciences. 2013; 10(4): 83-85.

5.Sonnesen L.Associations between the cervical vertebral column and craniofacial morphology.International Journal of Dentistry.2010; 2010.

6.Yadav Y, Goswami P, Bharihoke V. Cervical Vertebra Synostosis (C2-C3) - A Case Report, American Journal of Medical Case Reports. 2014; 2(6): 120-122.

7.Erdil H, Yildiz N, Cimen M. Congenital Fusion of Cervical Vertebrae and Its Clinical Significance.J Anat. Soc. India. 2003; 52(2): 125-127.

8.Kaplan KM, Spivak JM, Bendo JA. Embryology of the spine and associated congenital abnormalities: review article. The Spine Journal.2005; 5: 564-576.

9.Kulkarni V, Ramesh BR. A spectrum of vertebral synostosis. International Journal of Basic and Applied Medical Sciences. 2012; 2(2): 71-77.

10.Moore KL, Persaud TVN, Torchia MG. The developing human: clinically oriented embryology in skeletal system. 9th ed. Elsevier.New Delhi; 2013.p348-350.

11.Vaidyanathan S, Hughes PL, Soni BM, Singh G, Sett P. Klippel-Feil syndrome – the risk of cervical spinal cord injury: A case report. BMC FamPract.2002; 3: 6.

12.Tiwari A, Chandra N, Naresh M, Pandey A, Tiwari K. Congenital Abnormal Cervical Vertebrae–A Case Report.J Anat. Soc. India. 2002; 51(1): 68-69.

13.Moon MS, Kim SS, Lee BJ, Moon JL, Lin JF, Moon YW. Radiographic assessment of congenital C2-3 synostosis. Journal of Orthopaedic Surgery. 2010; 18(2): 143-7.

14.Schisler T, Huttunen H, Tang R,Vaghadia H. Ultrasound-assisted spinal anaesthesia in a patient with Wildervanck syndrome and congenital abnormalities of the lumbar spine. British Journal of Anaesthesia.2012; 109(2): 290-291.