Congenital Fusion Of Cervical Vertebrae and Its Clinical Significance

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Abstract. — The present study is based on clinical analysis of 5 cases with common complaints. Plain x-ray, and MRI revealed congenital fusion of cervical vertebrae at various levels. Although there was no other associated malformations, syndrome or trauma, all the patients presented with clinical disturbances and limited anatomical movements in the region of interest. Since it has a clinical and diagnostic significance, this anomaly needs to be investigated in terms of occurrence pattern, associated anomalies and clinical manifestations.

Key words: Cervical Vertebrae, Fusion, Block, Congenital

Introduction :

For many years anomalies of cervical region were of interest mainly to anatomists (Mc Rae and Barnum, 1953). Congenital anomalies are common in the vertebral column (Romanes 1981). In condition of fusion of the cervical vertebrae (FCV), two vertebrae appear not only structurally as one but also function as one (Dunsker et al, 1980). This fusion may be congenital (CFCV) or acquired (Resnick, 1992; Graaf, 1982). 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 (Graaf, 1982; Schlitt et al, 1989; Nagashima et al, 2001), limitation of the neck movement (Bharucha and Dastur, 1964), or the muscular weakness, atrophy and neurological sensory loss (Kameyama et al, 1993).

Since this anomaly has a clinical importance, with reliable evaluation by x-ray or Magnetic Resonance Imaging (MRI), propose of this study is to emphasise importance of the multidisciplinary approach to help establish the precise occurrence of this congenital anomaly for preventing any serious damage such as osteoarthritis by early treatment and diagnosis. It is discussed by comparing with other related articles.

Material and Methods :

In this study, patients admitted to the Department of Physical Medicine and Rehabilitation of Social Security Hospital (SSK), Sivas, Turkey with the common complaints were examined. The plain x-ray and MRI showed CFCV. There were observed degenerative changes in nonsegmented cervical regions both on x-ray and MRI. Vertabral heights were normal & antero-posterior (AP) diameters were decreased. However, there were no abnormal appearance at neural foramina.

Cases : Case 1 : A 47 year-old female with CFCV at C 5-6 (fig. 1), Case 2 : A 50-year-old female with CFCV between the C1-2 and C 6-7 (fig. 2), Case 3 : A 45-year-old female white CFCV at the levels C 4-5 and 6-7 (fig. 3), Case 4-A 63-year-old female with CFCV between C4-5-6 (fig. 4). Case 5 : A 49-year-old female with CFCV observed on MRI at C 6-7.(fig. 5a, 5b).

Observations:

Common Complaints of Cases: In all cases range of the motions of the neck and head like extension, flexion and lateral flexion were decreased. Although it seemed in case 2, as if there was minor intersegmental disc on upper segment FCV yet after the clinical examination and history of the patient we excluded the acquired pattern, it was believed to be a CFCV. In addition, all patients suffered from pain in the head, neck, shoulders and both upper limbs for many years. Muscular weakness of both upper limbs were observed. There was complaint of exhaustion, besides hypoesthesia of both upper limbs, paraesthesia of arms and hands. Their history was elaborated especially to rule out any associated malformations or syndrome. Neurological examinations, and deep tendon reflexes were found normal.

Discussion :

Congenital FCV is one of primary malformations of chorda dorsalis (Meschan, 1973; Besnich & Niwayama, 1985, Sutton 1993), believed to be due
to defects which take place during the development of the occipital and cervical somites. (Dunsker et al., 1980; Sadler 1990; Chandaraj and Briggs, 1992). Cause of this anomaly is often a combination of environment and genetics which occurs during the 3rd week postconception (Bethany and Mette, 2000). Although radiologic appearance of FCV has a characteristic feature, its precise diagnosis is complex, particularly, among young cases (Graaf, 1982). It is because ossification of the vertebral body is not complete till adolescence and cartilage which has not ossified, may appear like a normal disc area (Gray et al., 1964).

Since our cases were not young, it was easy to diagnose FCV without any confusing appearance on x-ray due to ossification process of the vertebra (Gray et al., 1964; Graaf, 1982). Cimen, (2000) reported FCV at C1-4 in cadaver skeleton and discussed possible syndromes and diseases.

In vivo, it is easy to diagnose the existence of the FCV by plan x-ray, but important point is to be able to distinguish between a pathologic condition and whether it is CFCV or acquired (Graaf, 1982, Ericson et al., 1984). If it is acquired FCV, it is generally associated with serious diseases such as tuberculosis, and other infections, juvenile rheumatoid arthritis and trauma (Gray et al., 1964; Graaf, 1982; Resnick, 1992). Although FCV may be silent; in advanced age it causes degenerative changes in nonsegmented cervical regions, and secondarily it leads to development of hypermobility and degenerative arthritis above and below of the fused cervical region (Mc Rae and Barnum, 1953; Yin et al., 1989), and may be associated with serious disease (Graaf, 1982; Schlitt et al, 1989), or Klippel-Feil syndrome as reproted by Nagashima et al (2001). The most important differential diagnosis in CFCV is decreased AP diameter of the vertebra, and individual measurements of the two vertebrae’s bodies height is equal to the two fused vertebrae’s height including the intervertebral disc. There are calcifications and atrophic appearance in intervertebral disc (Meschan, 1973; Besnich & Niwayama, 1985; Dahwerts; 1991; Sutton, 1993) on x-ray or MRI. History of the patient and physical examination is enough to make diagnostic differentiation between the acquied and CFCV (Gray et al. 1964). In consonance with the above mentioned literature the distance between the fused vertebrae’s height were the same in our cases and AP diameters of the vertebra were decreased (Meschan, 1973; Besnich et al, 1985; Dahwerts, 1991; Sutton 1993). In addition there was no tuberculosi, or any other infection, juvenile rheumatoid arthritis, trauma and rheumatoid spondylitis diseases in their history (Gray et al, 1964; Graaf, 1982; Resnick 1992). There were observed degenerative changes in nonsegmented cervical regions (Mc Rae and Barnum, 1953; Yin et al, 1989), but not associated with serious diseases or Klippel-Feil syndrome (Graaf, 1982, Nagashima et al, 2001), so we excluded the acquired pattern and believed that they were CFCV.

Patients suffered from progressive muscular weakness and sensory loss of both upper limbs. They have been suggested conservative therapy including immobilization of the neck position which will improve muscular weakness of both upper limbs, although in serious cases such as cervical myelopathy, it is inescapable to perform surgical treatment (Graaf, 1982, Yin et al, 1989). Yin et al (1989) suggested that the best cure is the conservative treatment for those with symptoms caused by radiculopathy. They analysed 87 cases with FCV, at C2-3 and the C3-4, showing no abnormality, or any other malformations. Our cases also showed no other abnormality or malformations, except the muscular weakness of both upper limbs, neck pain, paraesthesia and minor but intermittent head and neck pain. We suggested conservative therapy to improve muscular weakness of both upper limbs. The patients who do not respond to any conservative treatment may be advised cranectomy or C1-2 laminectomy. However, surgery has lots of risk factors. Although some patients were cured following the posterior decompression, most of them had same symptoms, while the others lost their lives during or after surgery (Mc Rae and Barnum, 1953; Bharucha and Dastur, 1964). Therefore we assert that, unless symptoms are unbearable, the conservative treatment should be done and repeated, because of the risk associated which surgery. Also we emphasise the importance of early diagnosis and treatment beside the multidisciplinary approach and systematical investigation.
References:

Fig. 1 Case 1: A 47-year-old female who had congenital FCV at C5-6.

Fig. 2 Case 2: A 50-year-old female who had congenital FCV between the C1-2 and C6-7.

Fig. 3 Case 3: A 45-year-old female who had congenital FCV between the C4-5/C6-7.
Fig. 4. Case 4: A 63-year-old female who had fusion between C4-5-6.

Fig. 5 a. Case 5: A 49-year-old female who had congenital FCV C6-7 on MRI, sagittal section.

Fig. 5 b. Case 5: A 49-year-old female who had congenital FCV C6-7 on MRI, coronal section.