Magnetic Resonance Imaging and Multidetector Computed Tomography Evaluation of Craniovertebral Junction Abnormalities

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ABSTRACT

BACKGROUND
The craniovertebral junction is a complex articulation between occiput, atlas, axis and supporting ligaments enclosing the soft tissue structures of cervicomedullary junction which includes medulla, spinal cord and lower cranial nerves. The incidence of different types of CVJ anomalies varies with demographic environment & ill-defined genetic factors. CVJ anomalies are more frequently found in Indian subcontinent than anywhere else in the world. Even in India, these anomalies are more frequently documented from Bihar, Uttar Pradesh, Rajasthan and Gujarat. The reason for this geographical clustering is more speculative. The CVJ anomalies can be either due to bony or soft tissue anomalies. They are common in all age groups and almost equal in both sex groups. The anomalies can be due to congenital or acquired causes. There has been a renewed interest in the normal anatomy & pathological lesions of CVJ anomalies with dynamic x-rays, computed tomography (CT) and magnetic resonance imaging (MRI). The clinical features are often delayed up to 2nd or 3rd decade, since they are subtle and often missed. Various congenital anomalies and acquired disease processes can affect the craniovertebral junction. They often cause diagnostic dilemmas. Only few studies have been conducted in this regard. This study is an attempt to define importance of precise diagnosis for pre-treatment evaluation and systematic classification of CVJ abnormalities with MRI and multi-detector computed tomography (MDCT).

METHODS
We conducted this cross-sectional descriptive study with 55 patients, who had been referred to us for CT / MRI from Department of Neurology. 3 Tesla MRI (GE Healthcare) and 16 slice MDCT (Philips) were used in this study.

RESULTS
In our study, congenital anomalies were the most common type of CVJ abnormality followed by degenerative changes and trauma. MRI proved to be better at detecting soft tissue abnormalities and assessing spinal cord compression, although CT was very much accurate at demonstrating bony lesions with short scan times and ability to reconstruct in three orthogonal planes.

CONCLUSIONS
CT and MRI cannot be compared in imaging the craniovertebral junction and should be complementary to each other.

KEYWORDS
Craniovertebral Junction, MRI, MDCT
**BACKGROUND**

CVJ is a complex region extending from a line drawn between internal occipital protuberance and mid-point of distance from the dorsum sellae to the anterior margin of foramen magnum up to C2—C3 interspace level. It encloses occipital bone, clivus, foramen magnum and upper cervical vertebrae (axis and atlas), their articulation and connecting ligaments and soft tissue structures of cervico-medullary junction which includes mainly medulla, cervical cord, cerebellum and lower cranial nerves. There are several ligaments connecting the bones, however, primary stabilising ligaments\(^2,3\) are the alar ligaments and transverse atlantal band\(^3\) of the cruciform ligament.

The CVJ abnormalities can lead to medullary-cervical cord compression,\(^4\) cranial nerve or spinal nerve compression, vertebral artery compression and obstructive hydrocephalus. These may present with neck pain, limb weakness. CVJ abnormalities can be a result of congenital anomalies or acquired disease processes (e.g. trauma, infection, inflammatory processes etc.).

Congenital CVJ anomalies include segmentation anomalies\(^5,6\) of atlas (assimilation), occiput, axis; atlas arch defects (rachischisis)\(^7,8\) anomalies of odontoid peg including osodontoidenum,\(^9,10\) ossiculumterminale,\(^11\) hypoplasia.\(^12\) Acquired abnormalities are trauma traumatic fracture\(^13,14,15,16,17,18\) of atlas, axis body or odontoid; inflammatory conditions viz. rheumatoid arthritis, ankylosing spondylitis; infections like tuberculosis,\(^19,20,21,22,23\) which is usually spread from elsewhere in body. Degenerative changes are very common, especially in Klippel-Feil syndrome\(^24,25\) where excessive movement occurs at CVJ to compensate for the rest of the non-mobile fused cervical vertebrae. The incidence of different types of abnormalities varies with demographic environment and has ill-defined genetic factors.\(^26\) The Indian subcontinent with very high population density of varying socio-economic strata, high incidence of infectious diseases and lack of healthcare awareness among population shows wide spectrum of CVJ abnormalities, in their late stages. These different types of abnormalities with complex pathological bony anatomy need precise diagnosis radiologically for individual management decision tailored for that particular case. This study was conducted for aetio logically categorising abnormalities, making precise diagnosis for pre-treatment evaluation and ruling out mimickers.

Plain radiographs of CVJ show overlap of many soft tissue structures. Due to anatomic complexities of the CVJ and high frequency of cranio-vertebral trauma with muscle spasm, plain radiographs pose limitations in accurate diagnosis.\(^27,28\) In this study we will be considering cross-sectional imaging with MRI and MDCT, as MDCT is the ideal modality for evaluation of complex osseous anatomy associated with CVJ abnormalities while MRI, with its multiplanar capabilities and high soft tissue contrast resolution has become the mainstay in radiological evaluation of CVJ.\(^27,28,29,30\) The craniometry of CVJ\(^1\) uses a series of lines, planes and angles to define the normal anatomic relationships of the CVJ namely Chamberlain line, McGregor line, McRae line, Wackenheim's or clivus line, height index of Klaus, Welcher basal angle, clivus canal angle, atlanto-occipital joint axis angle (Schmidt angle), Fischgold's digastic line and Fischgold's bimastoid line.

Also, CT,\(^29\) with its sagittal and coronal reconstruction confirms the diagnosis and helps to know the occipitalisation of the atlas, hypoplastic posterior arch of atlas. Enhancing soft tissue pannus at CVJ in cases with pharyngeal abscesses and lytic lesions in vertebrae in cases of tuberculosis were precisely diagnosed with CT in previous studies. Hence knowing the underlying cause of the abnormality helps in better prognostication and treatment of the patient's condition. Lastly and most importantly both modalities will be used to assess the atlanto-axial instability that may lead to spinal cord compression, which undoubtedly is the main concern for which clinicians refer them for imaging and depending on which further management is planned.

With this background, this study was performed to outline normal anatomy, normal variants of CVJ as seen in CT scan and MRI, systematically classify frequently detected CVJ abnormalities in local population and establish the role of MRI and CT for pre-treatment evaluation of CVJ abnormalities.

**METHODS**

This was an institution based cross-sectional observational study. Patients of either sex or age clinically suspected to have CVJ abnormality and evidence of same on dynamic x-ray were included. Study setting of MRI (3T) section was done at I.P.G.M.E & R and SSKM Hospital. CT Scan (16 slice) was done at Bangur Institute of Neurosciences. Informed consent form was obtained from the patients. This study was done in the Department of Radiodiagnosis, I.P.G.M.E & R and SSKM Hospital, Kolkata from January 2019 to August 2020. Control was not required as it was a prospective type of descriptive study. Study variables included age, sex, MRI and MDCT findings, dynamic x-ray findings. Demographic data such as age, sex, height, weight, religion etc. was collected. 55 patients were included in the study.

**Exclusion Criteria**
- Severely ill patients needing cardio-pulmonary support.
- Patients of acute trauma.
- Patients with claustrophobia.
- Patients with contraindications to MRI.
- Post-operative patients.

**Clinical History Recorded**
- Symptoms and duration
- For non-adult population - birth history and development history.
- For adult population - history of trauma, tuberculosis or any other diseases known to affect CVJ.
RESULTS

We conducted this cross-sectional observational study to understand the anatomy and various pathologies of the cranio-vertebral junction and assess their prevalence among the general population. Our institution is a tertiary care government hospital and cases which were suspected to have CVJ abnormalities were referred to our department for imaging by MDCT or MRI. In our study period we assessed many patients with such abnormalities and 55 of them, who met all the criteria for inclusion were selected for the study. As this was a simple random sampling, it should reflect the status of the majority of the population especially of West Bengal.

| Total Overall Cases | S5 | Percentage (%) |
|---------------------|----|----------------|
| Congenital anomalies| 46 | 83.63 |
| Trauma              | 9  | 16.36 |
| Infection           | 1  | 1.81  |
| Inflammation        | 2  | 3.63  |
| Neoplastic          | 2  | 3.63  |
| Degenerative changes| 26 | 47.27 |

Table 1. Relative Proportion of Different Types of CVJ Abnormalities Encountered in Our Study

| AAIS IN                      |  |
|------------------------------|--|
| OC hypoplasia                | 3 |
| Assimilation                 | 10|
| Rachischisis                 | 10|
| Os odontoideum               | 15|
| Os terminale                 | 3 |
| Dens hypoplasia              | 1 |
| Retroverted dens             | 4 |

Table 2. Different Types of Congenital Abnormalities Encountered in This Study with Presence or Absence of Atlantoaxial Instability

83.63 % patients had some sort of congenital abnormality making it the most common category of abnormality. Next most common category was degenerative changes (47.27 %) followed by trauma (16.36 %). Inflammation (3.6 %), bone tumour and tumour like lesions (3.6 %) and infection (1.81 %) being uncommon pathologies overall.

Among the patients having congenital abnormalities, os odontoideum (33 %) and atlanto-occipital assimilation (22 %) as a part of segmentation anomalies and different types of rachischisis (20 %) were the most common ones. Among the patients with os odontoideum, 93.3 % had instability, 30 % of atlanto-occipital assimilation had instability and 100 % (1 case only) hypoplastic dens had instability. Rest of the anomalies were rather stable.

Among patients with trauma to CVJ, type 2 odontoid fracture was most common (67 %), followed by atlas fracture, axis fracture and transverse atlantal ligament (TAL) rupture each accounting for approximately 11 %. About 67 % of patients with segmentation anomalies had degenerative changes at CVJ and out of them 54.5 % had atlantoaxial instability, making degenerative change the most common cause of atlantoaxial instability in segmentation anomalies.

Among the patients with atlantoaxial instability, os odontoideum (41 %), degeneration (32 %) and trauma (21 %)
were more common ones. Inflammatory disease (rheumatoid arthritis) was the cause for instability in 6 %.

Rheumatoid arthritis, as shown above, has caused instability in 100 % of cases. But due to a smaller number of overall patients of rheumatoid arthritis (RA), the contribution to the instability percentage is significantly low.

Following the Steel's rule of thirds, only 44.12 % of patients with severe instability had caused cord compression. This is because of the larger canal diameter at CVJ.

Two patients had associated Chiari type 2 malformation and one patient with Down Syndrome had Dandy Walker variant.

**DISCUSSION**

In the study conducted by Rajshree et al.,27 in Mumbai, congenital anomalies were the most common CVJ abnormality, similar to our study. However, trauma was found to be more common than degenerative changes, unlike our findings. This can be a regional variation. Their findings were compatible with the findings of the study conducted by Bhagwati.

In the study conducted by Mohan Kumar et al.,28 on acquired CVJ abnormalities, among the trauma patients, type 2 dens fracture was the most common followed by fracture of atlas. Significant male predominance was noted among the trauma patients. Rheumatoid arthritis and TB were next most common pathologies. Our findings were similar to this study if we take out the degenerative changes secondary to segmentation anomalies.

According to Roy Riascos et al.,29 CT should be the first imaging modality of choice when trauma to neck is suspected, rather than MRI. According to them, sensitivity and specificity of MRI to detect ligamentous injury in patients without CT evidence of instability is debatable and MRI should be used in trauma patients only when neurological status cannot be fully evaluated even after 48 hours of injury including those with normal initial CT. Our observations were correlated with this study.

According to Ramen Talukdar et al.,30 MRI should be the imaging modality of choice as its superior soft tissue resolution provides better understanding of topographic relationships between structures and also allowing to eliminate beam hardening artefacts observed in CT, especially in pre-operative assessment. CT is good only for bony abnormalities including anomalies and fractures. While we agree with their findings, role of CT cannot be negated completely as mentioned above.

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