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Spectrum of non-traumatic craniovertebral junction disorders. Diagnosis and demonstration with magnetic resonance imaging and multidetector computed tomography

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ABSTRACT

Introduction: Craniovertebral junction is the "zone of transition" between the skull and cervical spine so its detailed discussion is lacking in so many standard textbooks addressing these regions. These anomalies are especially common in the Indian subcontinent. Accurate diagnosis, probable aetiology and pre-treatment evaluation considerably affect the prognosis of patients. The aim of this study was to classify various Craniovertebral junction disorders according to their aetiology and to correlate the imaging findings with the clinical profile of the patients and histopathology whenever possible.

Methods: This prospective observational study was done in our department over the period of one year from August 2016 to July 2017. 57 North Indian patients from all age groups and both sex, who had imaging features suggesting craniovertebral junction pathology were selected for the study.

Results: Our study group comprised 34 males and 23 females with a male to female ratio of 1.47: 1. The most common age group was 11-20 years and the most common aetiology being developmental followed by infective, autoimmune and inflammatory disorders.

Conclusions: CVJ abnormalities are a group of treatable neurological disorders, and are approached with much caution by clinicians. Thus, it is crucial that radiologists should be able to make a precise diagnosis, categorize them into etiological groups, and give precise anatomical information on MDCT and/or MRI, as this information ultimately helps determine the management of such abnormalities and their prognosis.

INTRODUCTION

The term craniovertebral junction (CVJ) refers to a cone shaped area, the upper limit of which is a line joining the internal occipital protuberance to the spheno- occipital synchondrosis& the lower limit is inferior margin of the body of axis.[15] It is a transition site between

mobile cranium and relatively rigid spinal column. Disorders of the CVJ present a dilemmatic problem due to presence of neurologically vital structures in the vicinity, potentially unstable articulation and grave prognosis, if gone untreated and undetected.

Our study is an attempt to systematically classify different CVJ abnormalities according to their etiopathogenesis and to describe the importance of accurate diagnosis and precise anatomical information for pretreatment evaluation with Multidetector computed tomography (MDCT) and/or magnetic resonance imaging (MRI).

The main objectives of our study were:

1. To evaluate the patients with non traumaticcraniovertebral junction disorders by multiplanar and dynamic imaging utilizing MRI and/ or Multidetector CT.
2. To classify various craniovertebral junction disorders according to their etiology.
3. To correlate the imaging findings with clinical profile of the patients.
4. To correlate radiological diagnosis with diagnosis after medication /surgery / biopsy whenever undertaken.

METHODS

This prospective study was done in Department of Radiodiagnosis & Imaging, Ram Manohar Lohia Institute of Medical sciences, Lucknow, over the period of one year from August 2018 to July 2019.The Permission from Ethics Committee of our institute was taken. Written informed consent for the study was obtained from each patient prior to the examination.

North Indian Patients from all age groups and both sex, who underwent MRI and / or CT for clinical suspicion of non-traumaticCranio-vertebral junction disorder were evaluated. Out of these, 57 patients who had imaging features suggesting cranio-vertebral junction pathology were selected for the study. Contrast enhanced imaging was restricted only to those with suspicion of infective, inflammatory or neoplastic etiology. Dynamic scanning in neutral, active flexion and extension posture was done in all cases except in those with atlanto-axial instability. Only exclusion criterion was presence of at least one absolute contraindication for MRI.

MR imaging was performed with a 1.5 T whole body MR imaging system (SIGNA EXCITE GEMSOW) and the following sequences were obtained:

1. Sagittal -T1-weighted spin echo (T1W SE), T2-weighted turbo spin echo (T2W TSE) and T2-weighted fat suppressed.
2. Axial- T1-weighted spin echo (T1W SE) & T2-weighted turbo spin echo (T2W TSE).
3. Coronal - T2-weighted turbo spin echo (T2W TSE).
4. T2W TSE in flexion and extension whenever required.
5. Contrast enhanced T1W fat saturated (FS) images in axial, coronal and neutral sagittal planes.

Scanning parameters used for T1W SE sequence were a repetition time (TR) of 600 ms and an echo time (TE) of 9.7 ms; for T2W TSE sequence TR of 4,000 ms and TE of 100 ms; for T2W fat suppressed sequence TR of 3,300 ms, TE of 110 ms. These sequences were done with slice thickness of 4 mm and spacing of 0.5 mm.

CT examination was done on a Multislice spiral CT scanner (BRILLIANCE CT, Phillips medical system, Nederland, B.V.5684 PC Best). After acquisition of thin axial images, multiplanar reconstruction into sagittal and coronal planes was obtained.

Craniometric measurements used for radiologic assessment of CVJ abnormalities include Chamberlain's line, McGregor line, McRae line, Wackenheimclivus line, Fishgold digastric line, Fishgoldbimastoid line, Welcher basal angle, and atlantooccipital joint axis angle.

Detailed history of the patients was taken with respect to age, sex, socio-economic status, clinical profile & correlated with the spectrum of cranio-vertebral junction finding on MRI and Multislice CT. These patients were followed up to look for clinical outcome after conservative or surgical treatment.

Statistical Methods: Statistical analysis was done using ratio and percentages for age and gender distribution. Percentage distribution was calculated for each disease affecting the CVJ. Pearson's chi square test was used to establish the association between two different anomalies.

RESULTS

Our study group comprised of 34 males and 23 females with a male to female ratio of 1.47: 1. Maximum number of patients (21) was in the age group between 11-20 years, which included 8 females (34.78% of all females) and 13 males (38.24% of all males). This was followed by 21-30years age

group (12 patients) comprising of 3 females (13.04% of all females) and 9 males (26.47% of all males).
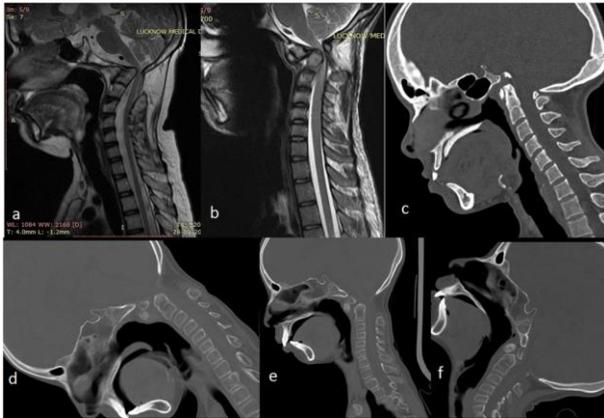
Figure 1. a) Sagittal T2W MRI shows Platybasia with flattened clivus, blocked C2- C3 complex with atlanto-occipital assimilation, AAD and BI causing compression over cervico-medullary junction **b)** Sagittal T2W MR shows Osodontoideum with AAD causing compressive myelopathy. **c)** Sagittal NCCT shows shallow posterior fossa with hyoplasticclivus related to basioccipital hypoplasia. A bony arch is noted at the tip of clivus which is continuous laterally with occipital condyles consistent with prebasioccipital arch. **d, e, f)** Sagittal dynamic NCCT shows mobile type atlantoaxial dislocation with ossiculumterminale causing cord compression.

Developmental cause of atlanto-axial diseases most commonly presented in 11-20 year age group (18 patients-40.91%), followed by 21-30 year age group (12 patients-27.27%). None of the patient of 51-60 years presented with a developmental cause, while one of these (2.27%) presented in 61-70 years age group. Patients with acquired causes of atlanto-axial diseases most commonly presented in 41-50 years age group (4 patients-30.77%) followed by 3 patients each (23.08%) in 11-20 and 51-60 years age group and 2 patients (15.38%) in 31-40 year age group [Table 1].
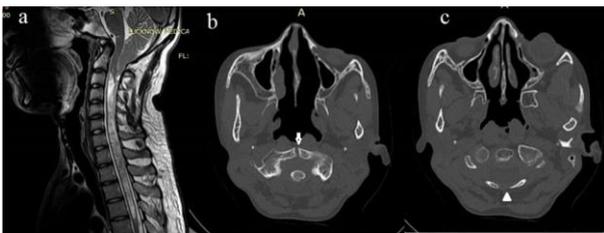
Figure 2. a) Sagittal T2W MRI shows small posterior fossa with hypoplasticclivus and cerebellar tonsillar herniation causing crowding at foramen magnum with compression over proximal cervical cord causing syringomyelia in cervico-dorsal cord. **b)** Axial non contrast bone window image shows anterior and posterior rachischisis with atlanto-axial dislocation and hypoplastic posterior arch of atlas.
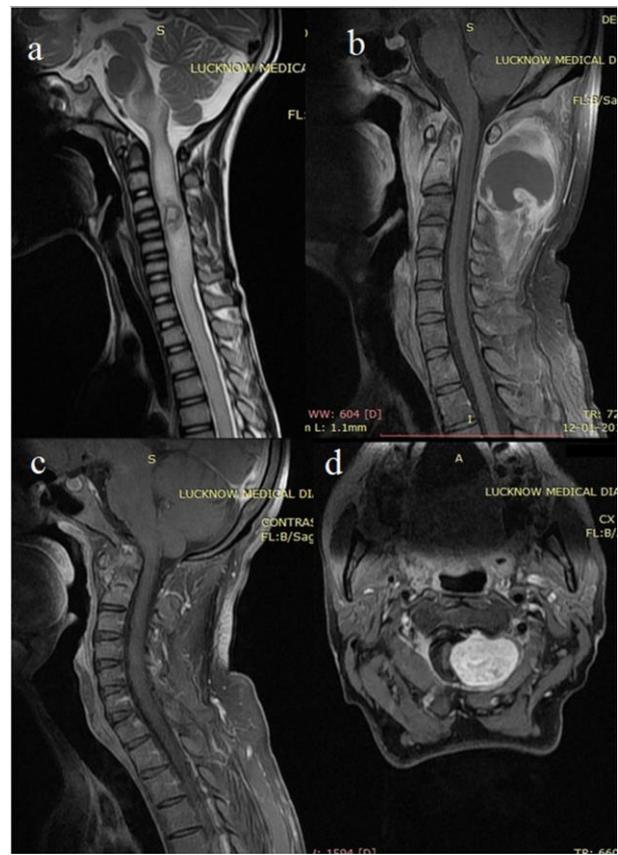
Figure 3. a) Sagittal T2W MR Image shows inflammatory granuloma in cervical cord with marked edema involving cord and brainstem. T1W Fat sat contrast enhanced **b)** sagittal image showing contrast enhancement in atlas and axis vertebra with erosion of dens. Enhancing granulation tissue is noted adjacent to atlanto-axial joint with a large abscess in paraspinal soft tissue. **c)** Sagittal image showing erosion of dens with mild heterogenous contrast enhancement in anterior atlas arch and axis vertebra. Enhancing soft tissue s/o pannus is noted adjacent to atlanto-axial joint with increased atlantoaxial interval. **d)** axial image shows a well defineddural based lesion with strong homogenous contrast enhancement at C1- C2 level on left side causing right lateral displacement and compression of cord. Radiological diagnosis of meningioma was made.

Most common presenting symptom was neck pain seen in 33 patients (57.89%), followed by weakness of both upper and lower limb in 24 patients (42.11%) and neck stiffness in 23 patients (40.35%). These were followed by fever, hemiparesis, sensory symptoms, paraparesis, headache and upper limb weakness. However, weakness of limbs (motor

symptoms) was overall the most common clinical feature present in 35 patients (61%).
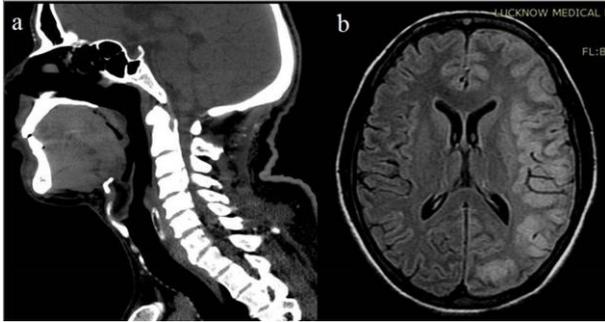
Figure 4. a) Sagittal CT shows thickened and ossified posterior longitudinal ligament causing spinal canal narrowing. **b)** Axial T2 FLAIR shows hyperintense signals in left cerebral hemisphere involving cortex and subcortical white matter. Diagnosis of Acute disseminated encephalomyelitis was made.

Age	Developmental	Percentage (%)	Acquired	Percentage (%)
0-10	4	09.09%	1	07.69%
11-20	18	40.91%	3	23.08%
21-30	12	27.27%	0	00.00%
31-40	4	09.09%	2	15.38%
41-50	5	11.36%	4	30.77%
51-60	0	00.00%	3	23.08%
61-70	1	02.27%	0	00.00%
TOTAL	**44**	**100.00%**	**13**	**100.00%**

Table 1. Age wise distribution of developmental and acquired abnormalities of cranio-vertebral junction.

Abnormality	No. of cases	Percentage (%)
Clivus segmentation	6	10.53%
Basilar invagination	31	54.39%
Condylar hypoplasia	1	01.75%
Platybasia	1	01.75%
Occipitalization	21	36.84%
Hypoplasia/aplasia of atlas arches	3	5.26%
Atlantoaxial fusion	0	00.00%
Fixed atlantoaxial instability	31	54.39%
Mobile atlantoaxial instability	4	7.01%
Cord compression with myelopathic changes	30	52.63%
Os- odontoideum	8	14.02%
Os- terminale	1	01.75%
Syrinx	4	07.02%
Tuberculosis	6	10.53%
Spondyloarthropathy	1	01.75%
Neoplasms	1	01.75%
Chiari malformation	7	12.51%
Inflammatory granuloma with myelitis	1	01.75%

C2/c3 block vertebra	5	08.77%
Thickened calcified pll	1	01.75%
Adem	2	03.51%

Table 2. Spectrum of cranio-vertebral junction diseases

Most common disorder was developmental anomalies (46 patients, 80%) followed by infective (7patients, 12.2%), autoimmune (2 patients, 3.5%), inflammatory (1 patient, 1.7%) and neoplastic (1 patient, 1.7%) etiology [Table 2]. The developmental anomalies were co-existing and were found in various combinations. Overall, Atlanto-axial instability was the most common abnormality found in 35 patients (fixed in 54.39% + mobile in 7.02%= 61.4% total) followed by Basilar invagination (31 patients, 54.39%) and Atlanto-occipital assimilation (21 patients, 36.84%) whether partial or complete (figure 1a). Osodontoideum was noted in 8 patients (14.04%) (figure1b) and Chiari malformation in 7 (12.5%) (figure 2a). Others being, C2/C3 block vertebra in 5 cases (08.77%) (figure 1a). Syrinx was found in 4 patients (07.02%) each (figure 2a) and hypoplasia/aplasia/rachisis of atlas arches in 3(5.26%) (figure 2b,c), clivus hypoplasia (figure 1a, 1c,2a) and tuberculosis found in 6 patients (10.53%) each (figure 3b).

ADEM were found in 2 patients each (03.51%) (figure 4b), while condylar hypoplasia, platybasia (figure 1), os-terminale (figure 1 d, e, f), spondyloarthropathy, neoplasm, inflammatory granuloma (figure 3) and thickened calcified posterior longitudinal ligament were found in only 1 patient each (1.75%) (figure 4 a).

Cord compression with myelopathic changes were observed in abovementioned abnormalities in 30 patients (52.63%) and were most frequently associated with atlanto-axial instability and basilar invagination (figure 1).

Out of 21 patients with Occipitalization, basilar invagination was seen in 19 patients (90.48%) with a p value of <.001 indicating a significant association between the two.

Basilar invagination also had an important association with atlantoaxial instability as Out of 31 patients presenting with basilar invagination, 26 also had atlantoaxial instability with a p value of <.001.

Out of 23 patients who were operated, 22 completely recovered, while partial recovery and post operative worsening was noted in one patient. Rests of the patients were managed conservatively.

One of the patients expired while one patient who presented with isolated syrinx was kept on monitoring.

DISCUSSION

Craniovertebral junction (CVJ) is a collective term for occipital bone, clivus, atlas, axis, supporting ligaments and underlying part of neural axis (medulla, spinal cord and lower cranial nerves)[2]. Any process congenital, developmental or acquired, which affects these structures, can give rise to abnormalities of CVJ. CVJ has intricate relationship with the major neurovascular structures which can lead to compression over cervico-medullary junction, lower cranial or spinal nerve and vertebral artery [10]. Due to its complex anatomy and relationships, CVJ disorders are difficult to treat, so, their proper classification, accurate diagnosis and precise anatomical information is of utmost importance as shown in our study.

Our study group of 57 patients, revealed male to female ratio of 1.47:1 which correlated well with the study done by Rajshree U. Dhadve et al & N.J.M. Mwang'ombe et al which also showed male predilection and a ratio of 1.6:1 and 2:1 respectively.[6,13] In our study, the most common age of presentation was 11-20 years followed by 21-30 years (21 patients, 37%) as compared to the studies by N.J.M. Mwangómbe et al and De Barrorswhich had a peak in third decade.[13,5] Most common presenting complaint in our study was neck pain followed by limb weakness and neck stiffness which was in accordance with most of the previous studies.[6,14]

However. Mwang'ombe stated that most common symptom was weakness of limbs followed by sensory disturbances, headache and neck pain.[13] The imaging features of cranio-vertebral junction in first decade are confusing as ossification is completed only after 12 years of age[11].

These composite disorders need accurate diagnosis and precise anatomical information by experienced radiologists so that decision for management can be tailored on individual basis. Due to complex anatomy of CVJ and overlap of many soft tissue structures, plain radiographs create limitations in diagnosis and anatomical delineation.[3] So, we used cross-sectional imaging (MRI and CT) for its evaluation. Due to its capability of volume rendering and multiplanar reformations, MDCT is "state of the art" imaging technique for evaluation of complex osseous anatomy of CVJ. MRI, due to its high soft tissue contrast resolution and multi-planar capabilities, has turned into the mainstay in radiological evaluation of this region [18]. Quantitative assessment of the CVJ uses a series of lines and angles, as discussed in material and method, to describe various anatomic relationships. [14]

In our study, developmental anomalies were the most frequent etiological group, followed by infective, autoimmune, inflammatory and neoplastic which correlated well with the studies done by Bhagwati et al [1] and Kale.[10] In our study, Atlantoaxial dislocation was most common etiology seen in 61.4% cases which was comparable with results produced by J.S. Chopra et al who stated that congenital AAD accounted for 51.5–68% of all CVJ anomalies.[3] Atlantoaxial instability was followed by basilar invagination and occipitalization which correlated well with study by Rajshree U. Dhadve.[6] Compressive myelopathic changes were seen in 52.63% cases which was in accordance with studies by Ramen Talukdar.[17] Os-odontoideum was found in 14.02% which was in correspondence with previous studies.[4] Os-terminale was seen in 1.75% of the cases

Among acquired disorders, tuberculosis was most common entity seen in 6 patients (10.5%) which may be explained by high prevalence of the disease in Indian subcontinent and correlated well with study done by Ramen Talukdar.[17] Out of these 6 cases, 4 patients showed erosions and destruction of dens and 2 of anterior atlas arch. Atlanto-axial dislocation was noted in 5 patients and basilar invagination in two. Prevertebral and paravertebral granulation tissue was seen in 5 cases and frank abscess formation in 2 cases. 3 patients showed extension of granulation tissue/ abscess in anterior epidural space and compressive myelopathy was seen in 2 patients. 2 cases showed involvement of the skull base in form of marrow edema and post contrast enhancement in clivus. Involvement of occipital bone was not seen in any of these patients. These findings were in accordance with study by Rajshree U. Dhadve.[6]

CVJ involvement in Rheumatoid arthritis was noted in one patient who showed erosions of dens and associated fibrous pannus resulting in atlanto-axial dislocation.

In our series of 59 patients, four had Syringomyelia and two had Chiari I malformation. The later entity was associated with platybasia, flattened clivus, blocked vertebra and atlantoaxial dislocation in 100 % of the cases. Scoliosis and platybasia were seen in two patients each which was in accordance with previous studies.[17] One patient had neoplastic etiology of CVJ, an extra-axial dural based meningioma. Elderly patients (between 50-70 years), who presented with complains of neck pain, headache and walking difficulty, developmental cause was seen in only one patient and acquired causes in 3 patients. Bony and ligamentous degeneration explained the above complaints in one patient as imaging revealed ossified posterior longitudinal ligament.

Our study showed basilar invagination in 90% of the cases who had atlanto-occipital assimilation indicating strong association between these two entities. This association was first noted by by Grawitz[9] and has since been reported frequently. [2,12,16,19,7] Many articles have regarded atlanto-occipital assimilation as a characteristic feature of basilar invagination.

Similar association was also noted between atlanto-axial instability and basilar invagination. Previous studies state micro-trauma due to instability related repeated cord injuries to be the defining factor in the entire pathophysiology of basilar invagination. [8]

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