

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

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Abstract

Background: Craniovertebral junction (CVJ) abnormalities constitute an important group of treatable neurological disorders with diagnostic dilemma. Their precise diagnosis, identification of probable etiology, and pretreatment evaluation significantly affects prognosis and quality of life of patients. **Aims:** The study was to classify various craniovertebral junction disorders according to their etiology and to define the importance of precise diagnosis for pretreatment evaluation with multidetector computed tomography (MDCT) and magnetic resonance imaging (MRI). **Materials and Methods:** This is a prospective observational study of 62 patients referred to our department between October 2012 and September 2014. All patients suspected to have a craniovertebral junction disorder were included in the study, from all age groups and both genders. Detailed clinical history was taken. Radiographs of cervical spine were collected if available. All patients were subjected to MDCT and/or MRI. **Results:** In our study of 62 patients; 39 were males and 23 were females, with male to female ratio of 1.6:1. Most common age group was 2nd-3rd decade (19 patients, 30.64%). Developmental anomalies (33 patients, 53.22%) were the most common etiology group followed by traumatic (10 patients, 16.12%), degenerative (eight patients, 12.90%), infective (four patients, 6.45%), inflammatory and neoplastic (three patients each, 4.8%), and no cause found in one patient. **Conclusions:** CVJ abnormalities constitute an important group of treatable neurological disorders, especially in certain ethnic groups and are approached with much caution by clinicians. Thus, it is essential that radiologists should be able to make a precise diagnosis of craniovertebral junction abnormalities, classify them into etiological group, and rule out important mimickers on MDCT and/or MRI, as this information ultimately helps determine the management of such abnormalities, prognosis, and quality of life of patients.

Keywords: Abnormalities, craniovertebral junction, magnetic resonance imaging, multidetector computed tomography

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Introduction

Craniovertebral junction (CVJ) abnormalities constitute an important group of treatable neurological disorders, especially in certain ethnic groups like Indian subcontinent. These are one of the major causes of spinal cord, vascular and nerve compression, and

hydrocephalus. Hence, in every patient presenting with these features, CVJ abnormalities should be excluded.^[1]

CVJ is a complex region that incorporates the occiput as well as the C1 and C2 vertebrae, while the stability and flexibility to CVJ is provided by the bony as well as

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ligamentous nature of atlantooccipital and atlantoaxial joints. Variety of abnormalities can affect the CVJ with their diagnostic dilemmas and few studies have been conducted in this regard.^[1]

This study is an effort to systematically classify various CVJ abnormalities according to their etiological group and to define the importance of precise diagnosis for pretreatment evaluation with Multidetector computed tomography (MDCT) and/or magnetic resonance imaging (MRI).

Materials and Methods

This study presents the data of 62 patients referred to our department between October 2012 and September 2014. Written informed consent for the study was obtained from each patient prior to the examination. All patients clinically suspected to have a CVJ disorder were included in the study, from all age groups and both genders. Exclusion criteria: None. Detailed clinical history was taken. Radiographs of cervical spine were collected if available. The abnormalities involving CVJ had been grouped as developmental or congenital, traumatic, degenerative, infective, inflammatory, neoplastic, and unknown. All patients were subjected to MDCT and/or MRI and contrast was given if required in patients with normal serum creatinine (normal up to 1.3 mg%).

The CT study was performed on Siemens Somatom 64 slice CT machine. CT was performed in neutral, flexion, and extension positions; except in situations where patient was not in a condition to perform flexion and extension and in cases of trauma. After thin axial images were obtained, these were isotropically reconstructed into coronal and sagittal planes. Contrast was administered in suspected cases of infection and inflammation. After contrast, images were acquired in neutral position only.

MRI was done on 3T Philips machine and the following sequences were obtained:

1. T1-weighted spin echo (T1W SE) and T2-weighted turbo spin echo (T2W TSE) in the sagittal and axial planes.
2. T1W SE and short tau inversion recovery (STIR) in coronal plane.
3. T2W TSE in flexion and extension whenever required.
4. Post gadolinium administration T1W fat saturated (FS) images were obtained in all three planes.

Typical parameters used for T1W SE sequence were a repetition time (TR) of 598 ms and an echo time (TE) of 27 ms; for T2W TSE sequence were TR of 4,100 ms and TE of 100 ms; for STIR sequence were TR of 2,300 ms, TE of

60 ms, and inversion time (TI) of 150 ms. These sequences were done with slice thickness of 3 mm with an inter-slice gap of 0.3 mm. Subsequently, in cases where contrast was required, gadolinium enhanced (administered in a dose of 0.1 mmol/kg) T1W FS sequences were obtained in all three planes.

Craniometric measurements used in radiologic assessment of CVJ abnormalities include Chamberlain's line, McGregor line, McRae line, Wackenheim clivus line, Fishgold digastric line, Fishgold bimaoid line, Welcher basal angle, and atlantooccipital joint axis angle.

Statistical analysis

Statistical analysis was done using ratio and percentages for gender distribution and percentage distribution of each disease affecting the CVJ.

Results

In our study of 62 patients, 39 were males and 23 were females with male to female ratio of 1.6:1. Most common age group was 21-40 years (19 patients, 30.64%) followed by older age of more than 60 years (17 patients, 27.41%). The most common presenting complaint in these group of patients was neck pain (26 patients, 41.93%) followed by limb weakness (17 patients, 27.41%); with others being headache, tingling numbness, restriction of neck movements, ataxia, giddiness, and scoliosis. Developmental anomalies were the most common etiology group followed by traumatic, degenerative, infective, inflammatory, and neoplastic, and no cause found in one patient in our study [Table 1].

Among the developmental anomalies, congenital atlantoaxial dislocation (AAD; 56.45%) [Figure 1a and b], basilar invagination (37.10%) [Figure 2], and occipitalization of atlas (20%) were the common radiological features and often exist with each other in varying combinations. Other developmental anomalies were os odontoideum (nine patients, 14.51%), Chiari malformation (eight patients, 12.90%) [Figure 3], platybasia (five patients, 8.06%),

Table 1: Distribution of patients according to etiological group

Etiological group	No. of patients	Percentage (%)
Developmental	33	53.22
Traumatic	10	16.12
Degenerative	8	12.90
Infective	4	6.45
Inflammatory	3	4.8
Neoplastic	3	4.8
No cause	1	1.6
Total	62	100

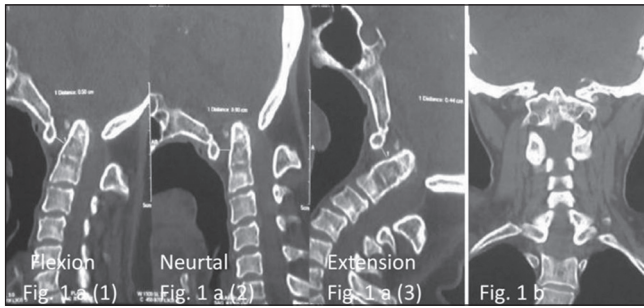


Figure 1: CT CVJ region bone window images. (a) Sagittal reconstruction showing reduced anterior atlantodental interval on extension. (b) Coronal reconstruction showing proper alignment of lateral atlantoaxial joints. This is suggestive of reducible atlantoaxial instability. 1 = flexion, 2 = neutral, 3 = extension of neck



Figure 2: CT CVJ region bone window image sagittal reconstruction showing tip of the odontoid process 'invaginated' into the foramen magnum and was above the Chamberlain line (longer line) and McRae's line (shorter line) in basilar invagination. CT = Computed tomography, CVJ = craniovertebral junction

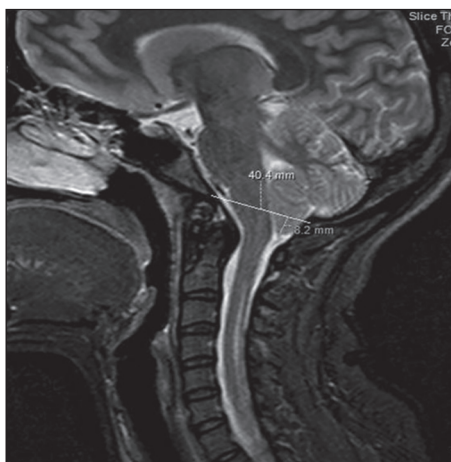


Figure 3: MRI CVJ region STIR midsagittal image showing syringomyelia and peg-like tonsils 8.2 mm below the foramen magnum. MRI = Magnetic resonance imaging, STIR = short tau inversion recovery

Klippel-Feil syndrome, and unassimilated rudimentary atlas (one patient each, 1.61 %).

Out of the 10 patients in traumatic group, five patients had type 2 fracture of the dens [Figure 4], two had type 3 fracture of the dens, one patient had tear drop fracture of anteroinferior aspect of the axis body, and one each had bilateral atlantooccipital dislocation and type 3 atlantoaxial rotatory dislocation.

Degenerative changes affecting the CVJ commonly mimic CVJ anomalies. Amongst the infective etiologies, tuberculosis (TB) [Figure 5a and b] was seen in four patients. Inflammatory pathology like rheumatoid arthritis (RA) [Figure 6 a and b] was seen in three patients. The neoplasms detected were, one meningioma [Figure 7], another was chordoma of axis which was also involving C3 vertebra, and a nerve sheath tumor was found at C2 vertebral level. Only two patients had a syndromic association, one was Klippel-Feil syndrome and other was Down's syndrome.

Discussion

The craniovertebral junction is a complex region formed by the occipital condyles, atlas (C1), axis (C2) vertebrae, and their articulations.^[2] Any process which can affect these structures can give rise to abnormalities of CVJ. These processes can be congenital, developmental, or acquired. CVJ has intricate relationship with the major neurovascular structures which can lead to medullary-cervical cord compression, cranial or spinal nerve compressions, vertebral artery compression, and obstructive hydrocephalus.^[3] Therefore, treatment of various types of CVJ abnormalities poses many challenges which are critically affected by the precise diagnosis of the abnormality as shown in our series by classifying the abnormalities into etiological group with their precise diagnosis.

The clinical manifestations are often delayed into the 2nd and 3rd decade because they are usually subtle and easily missed in children unless looked for specifically.^[4-6] In our study of 62 patients, male to female ratio was 1.6:1 (39 males and 23 females) and the most common age group was 21-40 years (19 patients, 30.64%), which correlated well with the study by Jawalkar *et al.*,^[7] and Sankhe and Kumar.^[1] In our study, most common presenting symptom was neck pain, followed by limb weakness, which correlated well with most other studies.^[1] But delayed symptom reporting is another hurdle in early diagnosis. Moreover, the radiological picture is confusing as the ossification of the bones is completed only by 9 years of age.^[8] The incidence of different types of abnormalities varies with the demographic environment and has ill-defined genetic factors.^[9-12] The Indian subcontinent with very high population density of varying socioeconomic strata, high incidence of infectious diseases, and lack of healthcare awareness



Figure 4: CT CVJ region bone window coronal reconstruction showing type II dens fracture

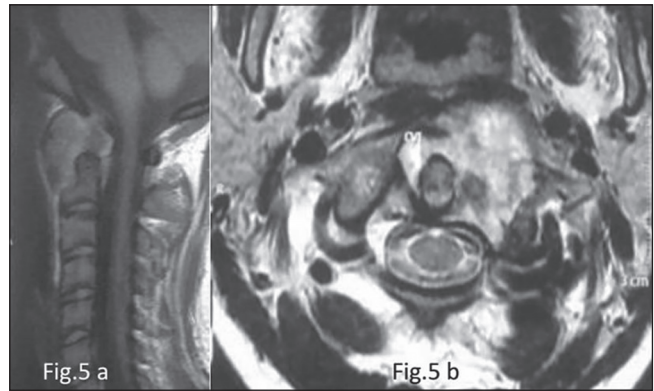


Figure 5: MRI CVJ region; (a) Midsagittal T1W and (b) axial T2W images. Hyperintense soft tissue in the pre-dental and left paravertebral space extending through C1-C2 foramen with erosions of dens and left lateral anterior arch of C2, which was to be of tuberculous origin. T1W = T1-weighted

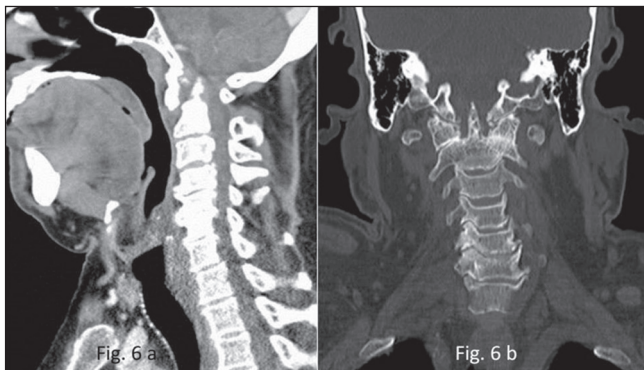


Figure 6: CT CVJ region in patient with known RA since 7 years. (a) Postcontrast soft tissue window midsagittal reconstruction showing enhancing soft tissue pannus surrounding the dens. (b) Precontrast bone window coronal reconstruction showing erosion and thinning of the dens suggestive of RA. RA = Rheumatoid arthritis

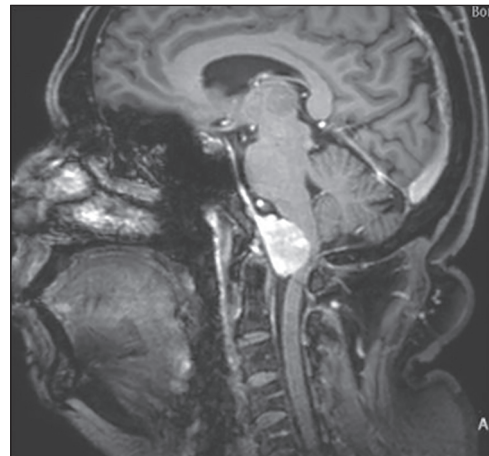


Figure 7: MRI CVJ region postcontrast T1W midsagittal image showing strongly enhancing anterior dural-based CVJ mass lesion with dural tail sign suggestive of meningioma

among the population shows wide spectrum of CVJ abnormalities and in its late stages. These different types of abnormalities with a complex pathological bony anatomy need precise diagnosis by experienced radiologists for individual management decision tailored for that particular case, which is why this study was conducted for categorizing the abnormalities etiologically, making precise diagnosis for pretreatment 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 craniocervical trauma with muscle spasm, plain radiographs pose limitations in accurate diagnosis. In our series, the cross-sectional imaging used was MDCT and MRI, as MDCT is ideal modality for evaluation of complex osseous anatomy associated with CVJ abnormalities while MRI, with its multi-planar capabilities and high soft tissue contrast resolution, has become the mainstay in radiological evaluation of the CVJ.^[13] The craniometry of the CVJ uses

a series of lines, planes, and angles to define the normal anatomic relationships of the CVJ namely Chamberlain's line, McGregor line, McRae line, Wackenheim clivus line, height index of Klaus, Welcher basal angle, clivus canal angle, atlantooccipital joint axis angle (Schmidt angle), Fishgold digastric line, and Fishgold bimaistoid line.^[1] Also CT with its sagittal and coronal reconstruction confirms the diagnosis and helps precisely to know the occipitalization of atlas, hypoplastic posterior arch of atlas, and C1-C2 instability. Enhancing soft tissue pannus at CVJ in cases of RA, retropharyngeal abscess, and lytic lesions in the vertebrae in cases of tuberculosis were precisely diagnosed with CT. Hence, knowing the underlying cause of the abnormality helps in better prognostication and treatment of the patient's condition.

In our study, developmental anomalies were the most common etiology group, followed by traumatic, degenerative, infective, inflammatory, and neoplastic,

and no cause found in one patient [Table 1]. This distribution of etiology was comparable with the study done by Bhagwati.^[14] In this study out of 159 cases of CVJ anomaly, developmental anomalies were the most common affecting 147 patients, neoplasm was found in 10 patients. Infective and traumatic cause was seen in one patient each. Another study of 189 cases by Kale and colleagues^[3] showed developmental cause as the most common followed by traumatic and tuberculosis in decreasing order. In clinicoradiological practice, congenital AAD accounted for 51.5-68% of all CVJ anomalies,^[15] which correlated with our study. In our study, os odontoideum was found in nine patients with male to female ratio of 2:1 (6 males:3 females) which correlated with study done by Dai *et al.*,^[16] and Spiering and Braakman,^[17] where male to female ratio was almost 3:1. Traumatic atlantooccipital dislocation is uncommon and often fatal. In our series, one patient presented with bilateral atlantooccipital dislocation and left lateral atlantoaxial subluxation after road traffic accident. One patient had traumatic type III atlantoaxial rotatory dislocation, which is very uncommon in adult patient. Till date, a total of 16 patients have been reported in the literature.^[18]

In our four patients of CVJ TB, involvement of the skull base was not seen. Loculated abscesses involving the prevertebral and paravertebral region were seen in two patients. Osteolytic destruction of the anterior arch of atlas was seen in three patients and in another three; erosions were noted in the dens. Basilar invagination was seen in one case and atlantoaxial dislocation was seen in two patients. In one patient, active pulmonary tuberculosis was present, while in other three patients there was past history of tuberculosis for which they had completed treatment. Chest radiograph in two of three patients showed sequelae of old tuberculosis. In our three patients of RA, the history of RA was long standing, more than 5 years in all patients. All the three patients had erosion of the odontoid process and enhancing soft tissue pannus along the dens. Atlantoaxial dislocation was found in all of them. Basilar invagination was found in two patients.

In our series of 62 patients, eight patients had Chiari I malformation. Syringomyelia was found in five patients, scoliosis in two, and platybasia in two. "Peg-like" tonsillar herniation of more than 8 mm was seen in all eight patients. One patient of Chiari I had multiple osseous abnormalities such as AAD, platybasia, fusion of C2 and C3 vertebrae, and hemivertebra at C4. These results were comparable in order of frequency with the study carried out in Ayub Teaching Hospital, Abbottabad between July 2008 and July 2010 with a prospective cohort of 60 symptomatic patients.^[19] Three patients had neoplasm affecting the CVJ. These were

meningioma which was anterior dural based, another was chordoma of the C2 body and spinous process and also involving the C3 vertebral body, and one C2 nerve sheath tumor causing AAD. In three patients in the age group of 75-81 years, who presented with complains of neck pain, walking difficulty, and imbalance; we found AAD and basilar invagination. Thus, these patients with developmental anomalies presented very late in life. Age-related degeneration of ligaments and bones caused the patients to present with above symptoms.

Conclusion

CVJ abnormalities constitute an important group of treatable neurological disorders, especially in certain ethnic groups, and are approached with much caution by clinicians. Thus, it is essential that radiologists should be able to make a precise diagnosis of CVJ abnormalities, classify them into the appropriate etiological group, and rule out important mimickers on MDCT and/or MRI; as this information ultimately helps determine the management of such abnormalities, prognosis, and quality of life of patients.

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Conflicts of interest

There are no conflicts of interest.

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