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EVALUATION OF CRANIOVERTEBRAL JUNCTION ABNORMALITIES WITH THE HELP OF MAGNETIC RESONANCE IMAGING AND MULTIDETECTOR COMPUTED TOMOGRAPHY

Radiodiagnosis

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ABSTRACT

Craniovertebral junction (CVJ) abnormalities constitute an important group of treatable neurological disorders caused by various etiologies like congenital, developmental, infective, inflammatory, neoplastic and traumatic. Their precise diagnosis, identification of probable etiology, and pretreatment evaluation significantly affects prognosis and quality of life of patients. MRI has more recently come into play to assess the ligaments, brainstem and spinal cord at the CVJ. This prospective study was conducted on 50 patients to classify various CVJ disorders according to their etiology and to define the importance of precise diagnosis for pretreatment evaluation using MDCT and MRI. Male to female ratio was 1.9:1 (33 males and 17 females) and the most common age group was >50 years. Multiplanar MRI was of great value in identifying the CVJ abnormalities precisely except for trauma where CT was accurate. Commonest CVJ abnormality was developmental anomaly 62% (n=31) followed by trauma 18% (n=9), arthritis 8% (n=4), infective 6% (n=3) and neoplastic 6% (n=3). Developmental anomaly was basilar invagination 45.1% (n=31) out of which BI-II (64.2%) was more common than BI-I (35.7%). Commonest finding on CT and/or MRI was cord compression followed by atlantoaxial dislocation in patients with anomaly.

KEYWORDS

Craniovertebral Junction, Basilar Invagination, MRI, Multidetector Computed Tomography.

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 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).

AIMS AND OBJECTIVES

- To study the utility of magnetic resonance imaging and multidetector computed tomography to evaluate the craniovertebral junction abnormalities
- To study most common developmental and acquired CVJ abnormalities.
- To arrange frequently detected CVJ pathologic imaging findings.

MATERIALS AND METHODS

This study presents the data of 50 patients referred to the department of Radiodiagnosis at Geetanjali Medical College and Hospital between Jan 2017 to June 2018. 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. Detailed clinical history was taken. The abnormalities involving CVJ had been grouped as developmental or congenital, traumatic, degenerative, infective, inflammatory and neoplastic. All patients were subjected to MDCT and/or MRI and contrast MRI was done if required in patients with normal serum creatinine (normal up to 1.3 mg%).

All trauma patients underwent CT to avoid unnecessary delay in scanning whereas MRI was done only in suspected cases of spinal cord trauma and spinal canal stenosis. MRI contrast study was performed in all suspected cases of infective, neoplastic and inflammatory etiology. CT and/or MRI scans with neck flexion and extension were performed whenever needed except in case of uncooperative patient.

INCLUSION CRITERIA:

1. Patients referred to department of radio-diagnosis clinically suspected to have a CVJ abnormality.
2. Patients with previous history or diagnosed with CVJ abnormality.
3. Patients were included irrespective of age or sex.

EXCLUSION CRITERIA:

1. Patients with ferromagnetic implants, pacemakers and aneurysm clips were excluded from MRI.
2. Patients with unstable vital parameters especially in the setting of trauma.
3. Pregnant and lactating patients were excluded from MDCT.
4. Patients with history of previous allergic reaction to MR contrast media and serum creatinine > 1.3 mg/dl were excluded from contrast study.
5. Claustrophobia.

Technique of CT and MRI examination:

The MDCT study was performed on 64 slices multidetector row CT scanner SOMATOM Sensation (SIEMENS). Thin axial images were obtained and reconstructed into coronal and sagittal planes.
MRI was done on MAGNETOM AVANTO (SIEMENS) 1.5 T 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, clivus canal angle, McRae line, Wackenheim clivus line, Welcher basal angle, Boogard's angle and atlantooccipital joint axis angle.

BI was divided into those associated with craniocervical instability in which odontoid process invaginated inside foramen magnum (Type I Basilar Invagination) and those not associated with instability but with flattened basecranium (Type II Basilar invagination)[2,3]

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 50 patients, male to female ratio was 1.9:1 (33 males and 17 females) and the most common age group was >50 years (12 patients, 46%) followed by 41–50 years (11 patients, 22%). The most common presenting complaint was neck pain (44 patients, 88%) followed by vertigo (35 patients, 70%); with others being headache, tingling numbness, restriction of neck movements, dizziness, fever, torticollis and short neck. Developmental anomaly (62%) was the commonest etiological group in CVJ abnormalities followed by trauma (18%), inflammatory (8%), infective (6%) and neoplastic (6%) [Table 1].

Table 1 : Spectrum of CVJ abnormalities (n=50)

CVJ Abnormalities	No. of Patients	%
Developmental anomalies	31	62
Trauma	9	18
Inflammatory	4	8
Infective	3	6
Neoplastic	3	6

Among the developmental anomalies, commonest was basilar invagination (14 patients, 45.16%) and commonest CT and/or MRI finding was CMJ compression (61.29%) followed by congenital atlantoaxial dislocation (48.38%). Other radiological findings were basilar invagination (45.16%), atlanto-occipital assimilation (32.25%), Arnold chiari I malformation (19.35%), etc (Fig.1) and often existed with each other in varying combinations. Other developmental anomalies were Arnold chiari I malformation (6 patients, 19.36%) [Fig. 1], os odontoideum (4 patients, 12.90%), C1 posterior arch rachischisis (4 patients, 12.90%), platybasia (4 patients, 12.90%), Arnold chiari II malformation (2 patients, 6.46%), etc.

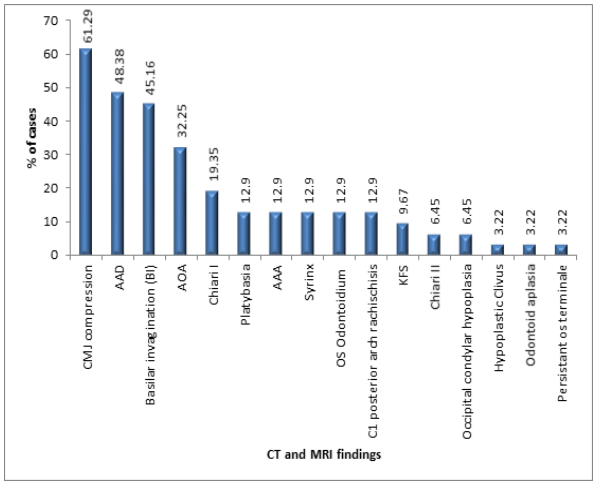

Fig 1: Distribution of CT and/or MR findings in developmental anomalies (n=31)

The incidence of developmental anomaly was found more common in the age group 11-20 years and males were more commonly affected than females (58.06% male and 41.94% female). Male to female ratio was 3:1 (3 males: 1 female) in cases of os odontoideum and 4:3 (8 males: 6 females) in cases of basilar invagination. Most common combination of developmental anomalies found was 25.80% for BI + AOA followed by 12.90% for BI+AAD and AOA+AAD, 9.67% for BI+AOA+AAD, KFS+BI, OO + AAD, and 6.45% for BI+ condylar hypoplasia, platybasia + AAA, platybasia + AOA and AOA + condylar hypoplasia.

Maximum CVJ injuries were fracture of odontoid process 66.66%, followed by fracture of atlas 33.33% and fracture of axis 11.11% (Fig 2). Commonest radiological finding among CVJ injury patients was fracture of odontoid process 66.66%, followed by fracture of atlas 33.33% and fracture of axis 11.11%. Cord compression and cord oedema were also found in 33.33% cases, whereas AAD was found in 22.22%. Commonest odontoid fracture found was type II (66.67%) whereas both type I and type III were found in 16.67% cases only.

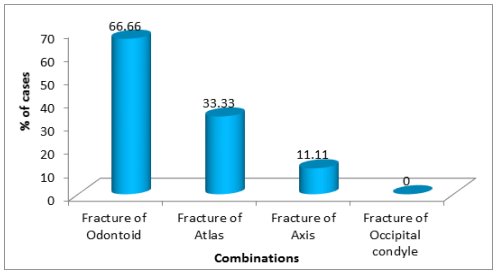

Fig.2: Incidence of various CVJ injuries (n=9)

CVJ arthritis was found in elderly patients only. 75% were above age of 60 and only one 1 cases was below 60 years. Out of total 4 cases of CVJ arthritis, one case was of psoriatic arthritis. Ligament thickening and pannus were found in all the patients (100%), whereas bone erosion and congenital comparison (75%) and AAS 25% (Fig 3).

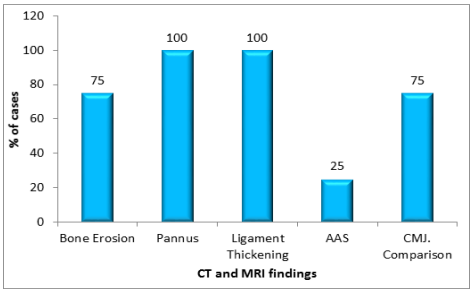

Fig.3: CT & MRI findings of CVJ arthritis (n=4)

Os odontoideum was found maximum in 0-20 years of age group (50%) and one (25%) was found in 41-60 years of age group and in 21-40 years each. Male to female ratio was 3:1. CT and MR conducted for os odontoideum showed 75% cases of AAD and cord compression each and only one case of BI (25%). Atlanto axial dislocation was the commonest associated finding in patients with os odontoideum with cord compression.

BI was found in 6 cases (42.86%) of age group 41-60 years and 0-20 years each. 2 patients (14.29%) were found in the age group of 21-40 years. Male to female ratio was 4:3 (8 males: 6 females). BI type II (64.2%) was more common than BI type I (35.7%). Platybasia was seen in combination with BI-II in 14.28%. KFS was seen in combination with BI-II in 21.42%, Anterior AOA was seen in combination with BI-I in 28%, Posterior AOA was seen in combination with BI-II in 21.42% and complete AOA was seen in combination with BI-II in 7.14%. No patient of BI-I showed posterior AOA, complete AOA, platybasia, KFS and OO. Out of 64.28% patients of BI-II with CMJ compression, 21.42% showed compressive myelopathy, whereas all patients of BI-I with CMJ compression showed compressive myelopathy. Among 4 patients of platybasia, BI-II was seen in 50% cases, whereas no BI-I was seen in 50% of cases (Fig 4).

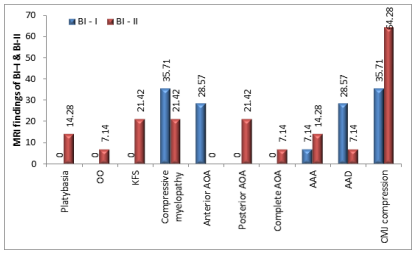

Fig.4: CT and/or MRI findings of BI–I & BI-II

Thus our study showed that platybasia and Klippel-Feil syndrome were more commonly seen with BI type II patients where as compressive myelopathy and atlanto-axial dislocation were more commonly seen with BI-I patients. Anterior atlanto occipital assimilation was more commonly seen with BI-I and posterior/total atlanto occipital assimilation was more commonly seen with BI-II. Platybasia and Klippel-Feil syndrome were more commonly seen with BI type II patients where as compressive myelopathy and atlanto-axial dislocation were more commonly seen with BI-I patients.

Arnold chairi malformation was most commonly found in the age group of 41-60 years (50%), whereas 37.50% were found in below 20 years of age group. Among 8 cases, commonest was chiari I (75%) followed by chiari II (25%).

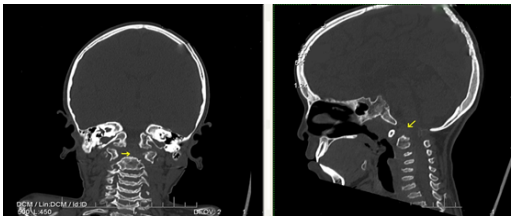

Figure 5 (26): Case of odontoid aplasia

There is absence of odontoid process on the body of C2 vertebra [yellow arrow fig (5a and 5b)].These imaging findings are suggestive of odontoid aplasia.

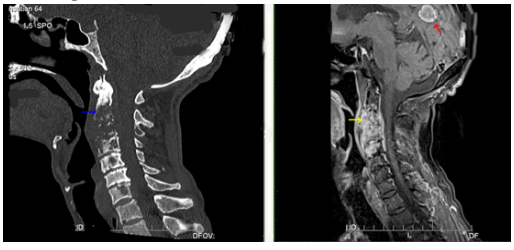

Figure 6: Case of metastasis from ca lung

Figure (6a) mid sagittal CT and (6b) post contrast T1 mid sagittal MRI images

65 year old man on chemotherapy for ca lung (biopsy proven) showing multiple bony destructive lytic lesions in C2,C3,C4,C6 vertebrae on CT scan [blue arrow fig.6(a)] with enhancement on post contrast MRI [yellow arrow fig.6(b)] and enhancing lesion in cerebrum [red arrow fig.6(b)]. These imaging findings are suggestive of vertebral and cerebral metastasis from ca lung.

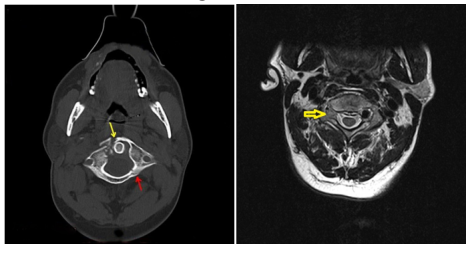

Figure 7: Case of Jefferson fracture

Figure 7(a) axial CT and 7(b) axial T2 MRI images

[Fig 7(a)and 7(b)] of 24 year old male showing fractures involving anterior and posterior arches of C1 vertebra [yellow and red arrows fig.7(a)] with loss of normal flow void in right vertebral artery [yellow arrow fig.7(b)] suggestive of vascular injury

Figure 8: Case of os odontoideum

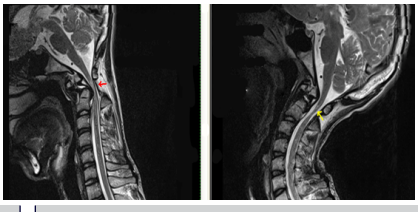

Figure 8(a) T2 mid sagittal flexion and 8(b) T2 mid sagittal extension images

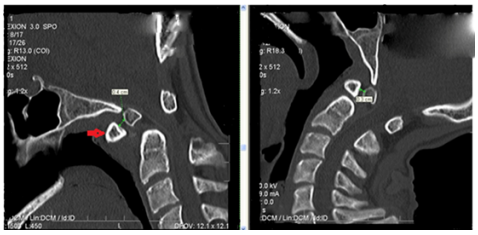

Figure 8 (C) mid sagittal flexion and 8(d) mid sagittal extension CT images

T1W MRI in a 20 year old patient with quadriparesis showing rounded bony fragment, lying above and anterior to the base of dens. Dens is hypoplastic, smooth and well corticated and anterior arch is hypertrophied [red arrow fig 8©] and rounded differentiating the condition from fracture. There is increased distance between anterior arch of atlas and rounded bony fragment on flexion extension CT [green line fig 8(c and d)]. MR also revealed marked ligament thickening, spinal canal narrowing on neutral and neck flexion with cord compression and myelomalacic changes [red arrow fig 8(c)]. These imaging findings are suggestive of os odontoideum with AAD.

Figure 9: Case of CVJ tuberculosis.

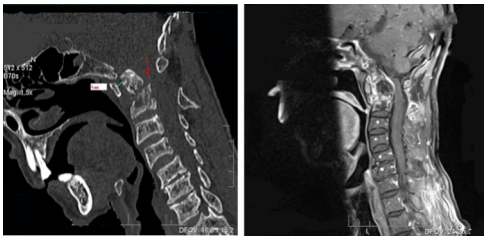

Figure 9(a) flexion mid sagittal CT Figure 9(b) post contrast T1 mid sagittal images

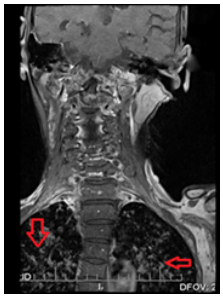

Figure 9 (C) coronal T2 images

71 year old male presented with bilateral limb weakness, hemoptysis and cough with sputum showing multiple bony lytic lesions and type II odontoid fracture [red arrow fig 9(a)] with displaced fracture fragment on neck flexion causing spinal canal narrowing, atlanto axial dislocation[blue line fig 9(a)], prevertebral abscess, granulomatous tissue and soft tissue thickening around odontoid process on T2 w image [fig 9(c)] showing post contrast enhancement [fig 9(b)] and multiple military opacities in both lungs [red arrow fig 9(c)]. These imaging findings are suggestive of tuberculosis of CVJ with pathological fracture of dens.

Figure 10: Case of type II basilar invagination

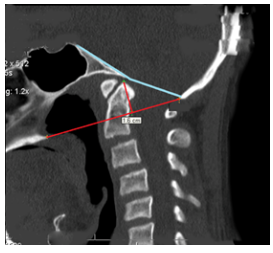

Figure 10 mid sagittal CT images of CT scan

18 year old male showing violation of Chamberlain's line [red line fig 10], acute clivus canal angle, normal Welcher's basal angle and increased Boogard's angle [blue line fig 10]. These imaging findings are suggestive of type II basilar invagination.

Figure 11: Case of platybasia

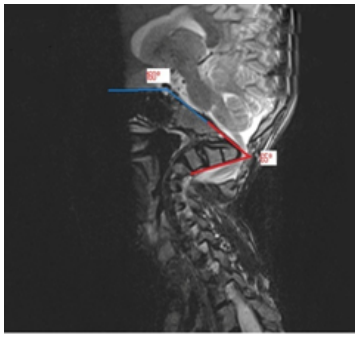

Figure 11(a) T2 mid sagittal images

18 year old male showing acute clivus canal angle [red line fig 11(a)], increased Welcher's basal angle [blue arrow fig 11(a)]. These imaging findings are suggestive of platybasia.

DISCUSSION

The craniovertebral junction is a complex region formed by the occipital condyles, atlas (C1), axis (C2) vertebrae, and their articulations.[4] 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. 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.[5] In our study of 50 patients, male to female ratio was 1.9:1 (33 males and 17 females) and the most common age group was 11–20 years (10 patients, 20%).

In our study, most common presenting symptom was neck pain 88% followed by vertigo 70% and restriction of neck movements 64% which correlated well with most other studies[6] 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. The incidence of different types of abnormalities varies with the demographic environment and has ill-defined genetic factors.[7] The Indian subcontinent with very high population density of varying socioeconomic strata, high incidence of infectious diseases, and lack of healthcare awareness 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. 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, McRae line, Wackenheim clivus line, Welcher basal angle, clivus canal angle, atlantooccipital joint axis angle (Schmidt angle), Boogard's angle.[6] 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 (62%) were the most common etiology group, followed by traumatic (18%), arthritis (8%), infective (6%) and neoplastic (6%). [Table-1]. This distribution of etiology was comparable with the study done by Bhagwati.[8] 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 et al[9] showed developmental cause as the most common followed by traumatic and tuberculosis in decreasing order.

In clinico-radiological practice, congenital AAD accounted for 51.5–68% of all CVJ anomalies,[10] which did not correlated with our study. In our study, commonest developmental anomaly was basilar invagination found in 45.1% of patients and commonest CT and/or MRI finding was CMJ compression (61.29%). In our study, commonest presenting complain was neck pain (88%) followed by restricted neck movements (64%). In our study, os odontoideum was found in four patients with male to female ratio of 3:1 (3 males:1 female) which correlated with study done by Dai *et al*,[5] where male to female ratio was almost 3:1.

In our study, commonest CVJ trauma seen was odontoid fracture (66.6%) followed by fracture of atlas (33.3%). Commonest odontoid fracture seen was of Type II (66.6%). Traumatic atlantooccipital dislocation is uncommon and often fatal. In our study, one case was of bilateral atlantooccipital dislocation and left lateral atlantoaxial subluxation after road traffic accident and one case was of Jefferson's fracture with atlanto axial dislocation.

There was one patient of CVJ tuberculosis in our study. Involvement of the skull base was not seen. Loculated abscesses involving the prevertebral and paravertebral region, osteolytic destruction of the anterior arch of atlas and odontoid process with type II pathological fracture of dens and bony erosions were noted.

Among four patients of CVJ arthritis in our study, the history of RA was long standing, more than 5 years in all patients. All the four patients had pannus and ligament thickening, three patients had erosion of the odontoid process and one patient had atlantoaxial joint sclerosis. There was one case of psoriatic arthritis with erosion of odontoid process in our study in correlation with few studies like Lassoued et a1[11] showed low incidence of odontoid erosions (12%) in their series of 56 patients with psoriatic arthritis, makes it a unique case in our study.

In our series of 50 patients, six (19.3%) patients had Chiari I malformation. Syrinx was found in four (12.9%) patients, scoliosis in two, and platybasia in four (12.9%). “Peg-like” tonsillar herniation of more than 8 mm was seen in all eight patients. These results were comparable in order of frequency with the study carried out in Abbottabad between 2008-2010 with a prospective cohort of 60 symptomatic patients.[12] In our study one patient had craniosynostosis (absent sagittal suture) in association with Arnold Chiari I malformation.

In our series of 50 patients, three patients had neoplasm affecting the CVJ. Two patients had metastasis from carcinoma of lung and breast involving C1 and C2 vertebra with post-contrast enhancement. One patient had compression fracture of C3 vertebral body. Third patient had CML of the clivus with enlarged enhancing cervical lymph nodes.

In our study, two patients around the age of 50 years presented with complain 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. In our study, patients with BI type II (64.2%)

were more common than BI type I (35.7%) in agreement with studies by Botelho et al.[2]

The importance of atlas assimilation in Basilar invagination: In our study, in the type I BI group, 28.57% patients presented with anterior arch assimilation (AAA), and 21.47% patients with type II BI presented with posterior arch assimilation, either in isolation or associated with anterior arch assimilation which is in agreement with study by Ferreira et al[13] and Botelho et al[14], in the type I BI group, all patients presented with anterior arch assimilation (AAA) and 63% of the patients with type II BI presented with posterior arch assimilation, either in isolation or associated with anterior arch assimilation. In the control group, no patients had atlas assimilation. It was suggested that anterior atlas assimilation leads to type I BI. Posterior atlas assimilation more frequently leads to type II BI. At least in Type I BI, AAA is strongly associated with craniocervical instability and craniocervical junction kyphosis.

In our study, 50% patients of BI type II had platybasia and not a single patient of BI type I had platybasia in agreement with studies by Botelho et al[14]. Platybasia was suggested to be more prevalent in type II BI than in type I BI.

Thus, the results of our study were in common agreement with other studies[1,15]; describing CVJ abnormalities as an important group of treatable neurological disorders and in disagreement with study of Chopra[10] describing atlanto-axial dislocation as commonest CVJ anomaly. Our study was also in common agreement with Botelho et al[14], Jânio et al[3], etc; describing types and associations of basillar invagination.

CONCLUSION

In our study of north western region of India, developmental anomaly appears to be the commonest CVJ abnormality. Most prevalent CVJ anomaly is basilar invagination out of which BI-II is more common than BI-I. Platybasia and Klippel-Feil syndrome are more commonly seen with BI-II. Basilar invagination with atlanto-occipital assimilation is the commonest combination among CVJ anomalies. Type II odontoid fracture is the commonest CVJ injury.

MRI is better for demonstration of ligaments and cervicomedulary junction distortion in all patients, where as MDCT is better for demonstration of osseous abnormality. The angular craniometry differs among different types of craniovertebral junction anomalies. Many CVJ abnormalities are not isolated. Therefore, a detailed examination of the different bone and soft-tissue structures is essential for diagnosis of CVJ abnormalities, management, prognosis and quality of life of patients.

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