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Imaging In Craniovertebral Junction (CVJ) Abnormalities

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Abstract:*The craniovertebral junction (CVJ) consists of complex anatomy of osseous, ligamentous and neurovascular structures including multiple lower cranial nerves. CVJ may be congenital, developmental or due to malformation secondary to any acquired disease process. These anomalies can lead to neural and vascular compromise, obstructive hydrocephalus and cerebrospinal fluid dynamics. Aims and Objectives: To outline normal anatomy, normal variants, to arrange frequently detected CVJ abnormalities and emphasize clinical implications to improve our radiological report. A prospective study of 46 cases of CVJ abnormalities using 16 slice Philips MX16 CT machine and 1.5T SIEMENS Tim Avanto MRI machine, 28 cases (61 %) was congenital anomalies, 10 (22 %) trauma, 4 (8 %) tuberculosis,4 (9 %) of rheumatic arthritis. Males were more common than females (3: 1). There was maximum incidence of cervicomedullary junction compression and atlantoaxial dislocation followed by Chiari I with syrinx.Due to advances in computed tomography and magnetic resonance imaging and ability to image multiplanar sequences the complex anatomy is well understood. Due to limited published articles on CVJ, our study is aimed to study Indian scenario of craniovertebral junction abnormalities.*
Keywords: *Congenital, craniovertebral junction, magnetic resonance imaging, tuberculosis, trauma*

I. Introduction

The craniovertebral junction (or craniocervical) (CVJ) consists of occiput (posterior skull base), foramen magnum, clivus, atlas, axis, ligaments of atlantooccipital and atlantoaxial articulations.It encloses the soft tissue structures of the cervicomedullary junction (medulla, spinal cord, and lower cranial nerves). CVJ may be congenital, developmental or due to malformation secondary to any acquired disease process. These anomalies can lead to neural and vascular compromise, obstructive hydrocephalus and cerebrospinal fluid dynamics. An understanding of development of craniovertebral junction is essential for the recognition of pathological abnormalities.

This article is focused on studying craniovertebral junction complex anatomy, normal variants, congenital and acquired abnormalities. Due to advances in computed tomography and magnetic resonance imaging, our ability to image multiplanar sequences in the complex CVJ anatomy is well understood. Due to limited published articles on CVJ, our study is aimed to study Indian scenario of craniovertebral junction abnormalities.

II. Aims And Objectives

- To outline normal anatomy of the craniovertebral junction (CVJ)
- To study most common developmental and acquired CVJ abnormalities
- To arrange frequently detected CVJ pathologic imaging findings
- To emphasize clinical implications to improve our radiological report

III. Materials And Methods

A prospective study of 46 patients referred from outpatient and emergency department of Gauhati Medical College and Hospital during July 2012 to October 2013 were studied. Computed tomography (CT) scan was performed on a 16 slice Philips MX16 CT machine and magnetic resonance imaging (MRI) on 1.5T SIEMENS Tim Avanto machine. Multiplanar sequences using T1, T2, GRE, MR myelogram were used. Final diagnosis was made after MRI findings with clinical correlation and in some patients confirmed on surgery.

Inclusion criteria: Patients with clinical suspicion of CVJ abnormalities evaluated by CT and Magnetic Resonance Imaging.

Exclusion criteria: Claustrophobia, Cardiac implants, unstable patients

IV. Results And Observations

This is a prospective study of 46 cases of CVJ abnormalities, 28 cases (61 %) congenital anomalies, 10 (22 %) trauma, 4 (8 %) tuberculosis,4 (9 %) of rheumatic arthritis. Males were more common than females (3:1). There was maximum incidence of cervicomedullary junction compression and atlantoaxial dislocation followed by Chiari I with syrinx.The combination of OA+AAD was seen in 20% patients.

Combination of BI+OA was seen in 14.2% and BI +OA+AAD was seen in 7.1% patients and meant a localized congenital affection affecting both atlas and basiocciput.

V. Charts, Tables, Images

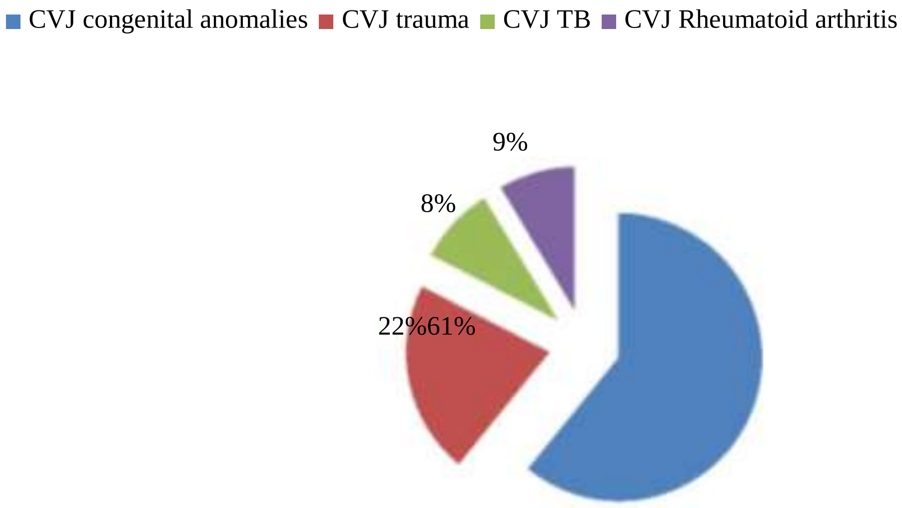


CHART 1: Various CVJ abnormalities (Cases / percentage wise distribution)

Age group (years)	No of Patient			Percentage
	Male	Female	Total	
0-10	6	2	8	2 8.57%
11-20	5	1	6	21.4%
21-30	5	2	7	25%
31-40	2	1	3	10.71%
41-50	1	3	4	14.2%
>50	19	9	28	100%

Table 1: AGE distribution of CVJ anomalies

Sex	No. Of Patients	Percentage
Males	19	67.85%
Females	09	32.1%

Table 2: Sex Distribution of CVJ Anomalies

MRI findings	Number	Percentage
Basilar invagination (BI)	6	21.4%
Platybasia	2	7.1%
Occipitalisation of atlas (OA)	8	28.57%
Atlanto-occipital dislocation	1	3.52%
Atlantoaxial Dislocation (AAD)	13	46.4%
Hypoplastic clivus	1	3.52%
Chiari I (Cerebellar herniation)	12	42.8%
Chiari II	02	7.1%
Chiari III	1	3.52%
CMJ compression	19	67.85%
Syrinx	9	32.1%
KlippelFeil syndrome	01	3.5%
Odontoid aplasia	1	3.5%
Odontoid dysplasia	1	3.8%
Os odontoideum	5	19.2%
Retroflexed Odontoid	2	7.1%
Hypoplasia of Atlas	6	3.8%
Hypoplasia of condyles	1	3.8%

Table 3: Distribution of MR Findings in CVJ Anomalies

Combination	Number	Percentage
OA+AAD	05	17%
BI+OA	04	14.2%
0A+Hypoplasia of atlas	03	10.71%
AAD+OD	02	7.1%
BI+OA+AAD	02	7.1%
PB+OA	01	3.5%

Table 4: Combinations of CVJ Anomalies

MRI findings	Number	Percentage
Odontoid Fractures	9	90%
Fracture of Atlas	2	20%
Fracture of Axis	1	10%
Occipital Condyle Fracture	0	0%

Table 5: Incidence of Various CVJ injuries

MRI findings	Number	Percentage
Odontoid Fractures	9	90%
Fracture of atlas	2	20%
Fracture of axis	2	20%
Atlanto-occipital dislocation	1	10%
Atlantoaxial Dislocation (AAD)	1	10%
Cord compression	2	20%
Cord edema	3	30%
Absent Flow void in vertebral artery	01	10%

Table 6: MRI findings in injuries of CVJ

Odontoid Fractures	Number	Percentage
Type I	2	22.2%
Type II	5	55.55%
Type III	2	22.2%

Table 07: Incidence of different types of odontoid Fractures

Age group	No	Percentage
0-20	1	25%
21-40	1	25%
41-60	1	25%
>60	1	25%

Table 08: Age distribution of CVJ tuberculosis

CT Findings	No	Percentage
Bone erosion	2	50%
Soft tissue component	2	50%
AAD	2	50%
Epidural component	1	25%
Contrast Enhancement	2	50%

Table 09: CT findings in CVJ tuberculosis

MR Findings	No	Percentage
Bone erosion	2	50%
Soft tissue component	2	50%
AAD	2	50%
Epidural component	1	25%
Transverse ligament breach	1	25%
Transverse ligament thickening	1	25%
Contrast enhancement	3	75%
CMJ compression	2	25%
Cord edema	1	25%

Table 10: MR findings in CVJ tuberculosis

Age Group	No	Percentage
0-20	0	0%
21-40	0	0%
41-60	1	25%
>60	3	75%

Table 11: Age distribution of CVJ Rheumatoid Arthritis

MR Findings	No	Percentage
Bone Erosion	3	75%
Pannus	4	100%
Ligament Thickening	3	75%
AAS	2	50%
CMJ Compression	3	75%

Table 12: MR findings of CVJ Rheumatoid Arthritis

.

Imaging Findings	LEE et al	Signoret F, et al	S Basu et al	**Present study**
Odontoid fractures	32.5%	49%	46%	90%
Fracture of Atlas	47.5	23%	28%	20%
Fracture of Axis	32.5%	37%	14%	10%

Table 13: Analysis of CVJ injuries- Comparative study

Imaging Findings	Lee et al	Dickman et al	Present study
C1+Odontoid Fractures	32.5%	37.5%	20%
C1+Miscelleaneous C2 #	15%	28%	0
C1+hangmans #	10%	12%	0

Table 14: Combination of CVJ injuries – Comparative study

MR Findings	Krishnan A et al	Present study
Dens involvement	62%	50%
Prevertebral soft tissue	75%	50%
Epidural component	86%	25%
CMJ compression	41%	75%

Table: 15: Analysis of MR Findings in CVJ tuberculosis

MR findings	BUNDSCHUH ET AL	Present study
Bone Erosion	100%	100%
Pannus formation	100%	100%
Atlantoaxial subluxation	53%	50%
CMJ Compression	26%	75%

Table 16: Analysis of MRI findings in CVJ Rheumatoid Arthritis

Table: 17 Classification of Craniovertebral Junction Anomalies

I Congenital

- Malformation of the occipital bone
  - ✓ Malformations of occipital bone.
  - ✓ Clivus segmentation
  - ✓ Remnants around foramen magnum
  - ✓ Atlas variants
  - ✓ Dens segmentation anomalies
  - ✓ Basilar invagination
  - ✓ Condylar hypoplasia
  - ✓ Assimilation of the Atlas
- Malformations of the atlas.
  - ✓ Assimilation of the atlas
  - ✓ Atlantoaxial fusion
  - ✓ Aplasia of atlas arch
- Malformations of the axis.
  - ✓ Irregular atlantoaxial segmentation
  - ✓ Dens dysplasias
  - ✓ Ossiculum terminals persistent
  - ✓ Os odontoideum
  - ✓ Hypoplasia/Aplasia
- Segmentation failure of C2/C3

II Acquired

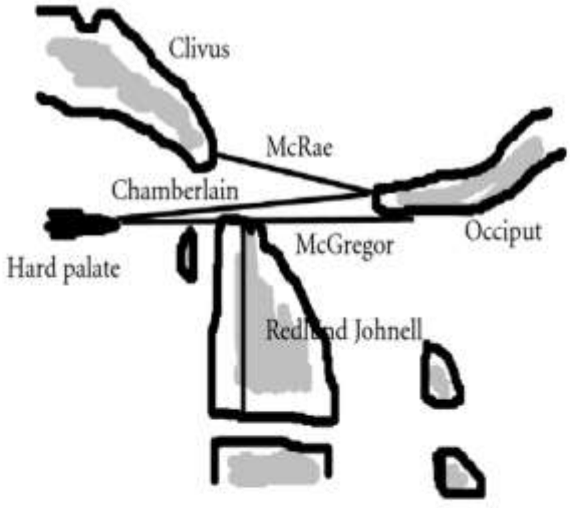
(A) Abnormalities at foramen magnum.

- ✓ Secondary basilar invaginations (Paget's disease, rheumatoid arthritis)
- ✓ Foraminal stenosis (e.g., achondroplasia)

(B) Atlantoaxial instability.

- ✓ Errors of metabolism
- ✓ Down's syndrome
- ✓ Infections (Grisel's syndrome)
- ✓ Inflammatory (RA)
- ✓ Traumatic
- ✓ Tumors- neurofibromatosis
- ✓ Miscellaneous- Syringomyelia, fetal warfarin syndrome, contradi's syndrome

NORMAL LANDMARKS FOR CVJ CRANIOMETRY



Chamberlain's line: Line extend between posterior pole of the hard palate and opisthion. Tip of dens commonly lies below or just tangent to the line or may normally project several mm above this line.

McGregor line: Line between the posterior pole of the hard palate to the lowest portion of the occipito-squamosal surface. Tip of the dens should be <5mm above this line.

McRae's line: Line between the basion and opisthion, tip of the dens should be below this line, if above this line s/o basilar invagination.

Wackenheim Clivus baseline (basilar line): It is the line along the clivus which is tangential to posterior aspect of the dens. Line should fall tangential to posterior aspect of the dens; if not s/o basilar invagination.

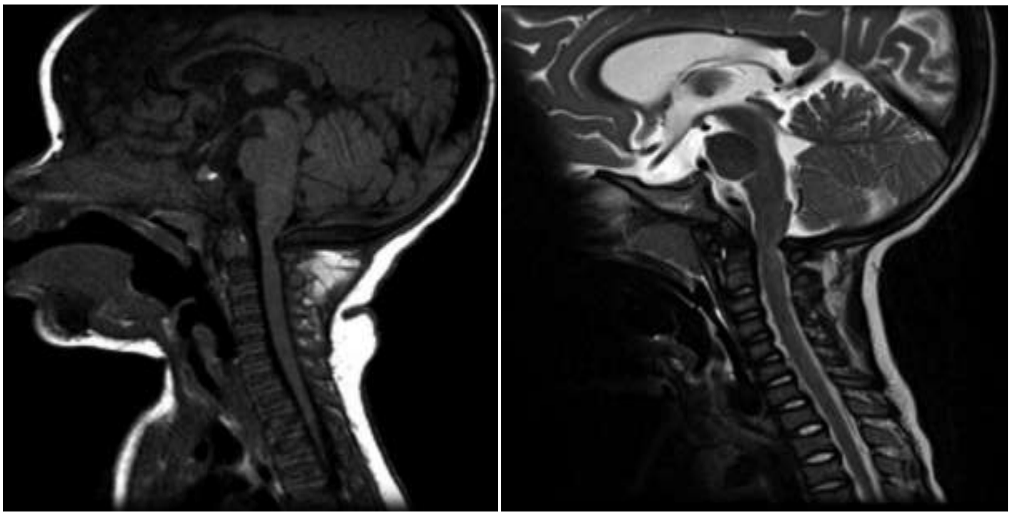
Clivus canal angle: Angle formed at the intersection of the Wackenheim clivus baseline with a line constructed along the posterior surface of the axis body and dens. Clivus canal angle should range between 150-180 degree.

Welcher basal angle – Angle formed by intersection of the nasion-tuberculum line and tuberculum-basion line. It should be less than 140 degree.

Atlanto-occipital joint axis angle (Schmidt-Fisher angle) - Angle formed by line drawn parallel to both atlanto-occipital joints which typically intersects at the center of the dens when condyles are symmetric.
Average angle is 125 degree (124-127 degree) and becomes obtuse in condylar hypoplasia.

IMAGES OFDEVELOPMENTAL AND ACQUIRED CVJ ABNORMALITIES

Figure 1: Achondroplasia. A. Skull AP view shows a relatively large cranial vault with small skull base and prominent forehead with depressed nasal bridge
B. Sag T2 (right) and C. T1(Left) weighted MR images showing foramen magnum stenosis causing compression of cervicomedullary junction



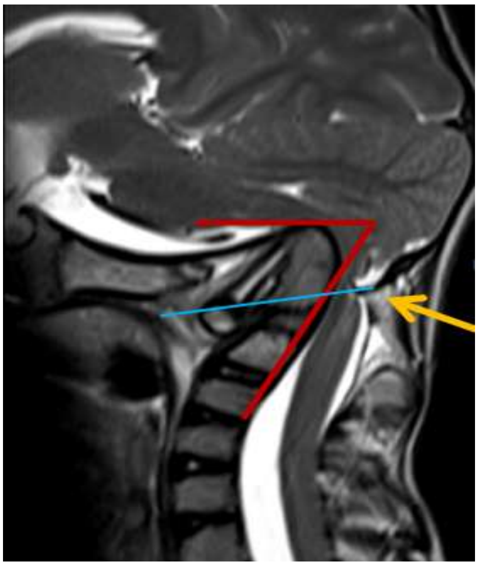


Figure 2: Platybasia. 32 year old gentleman with decreased clivus canal angle (red line), violation of Chamberlain's line (blue line) acute angulation, compression of cervicomedullary junction (yellow arrow).

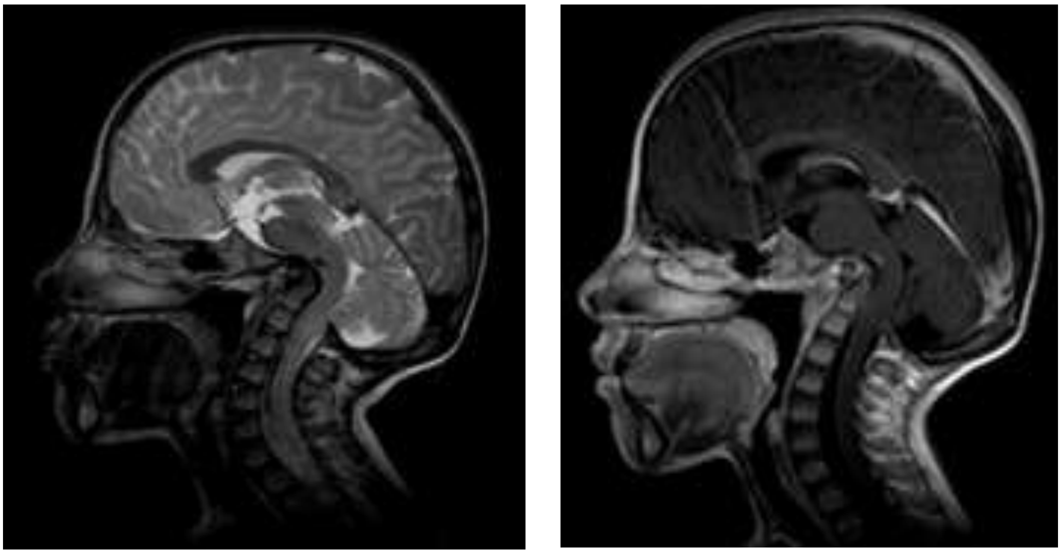


Fig 3: Osteogenesis imperfecta: Sagittal T1(Right) and T2(Left) weighted MR images showing basilar invagination with odontoid process in foramen magnum, platyspondyly, cervical cord compression.

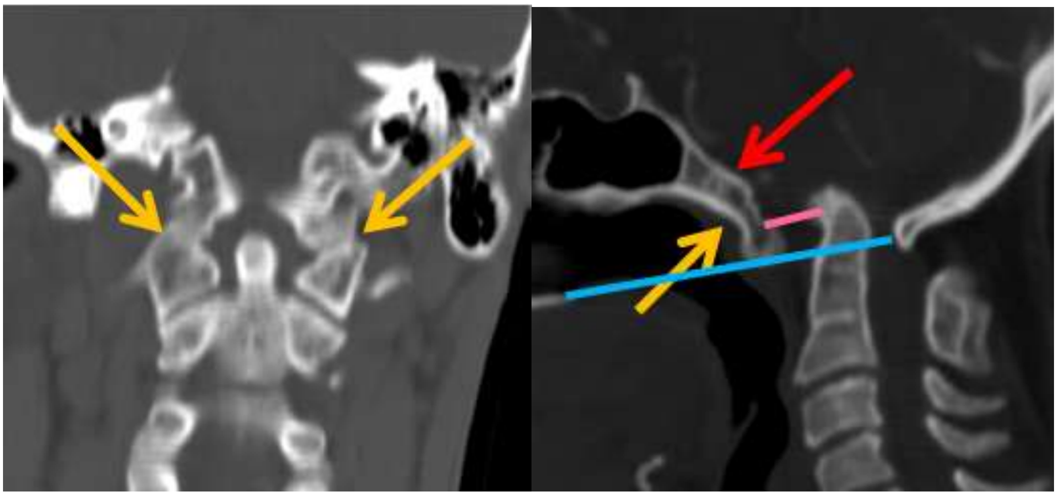


Figure 4: Atlanto-occipital assimilation. A. CT coronal section showing complete atlanto-occipital assimilation on right side and incomplete atlanto-occipital assimilation on left side. B. CT sagittal section showing complete atlanto-occipital assimilation, short Clivus, violation of Chamberlain's line-basilar invagination and atlantoaxial dislocation.

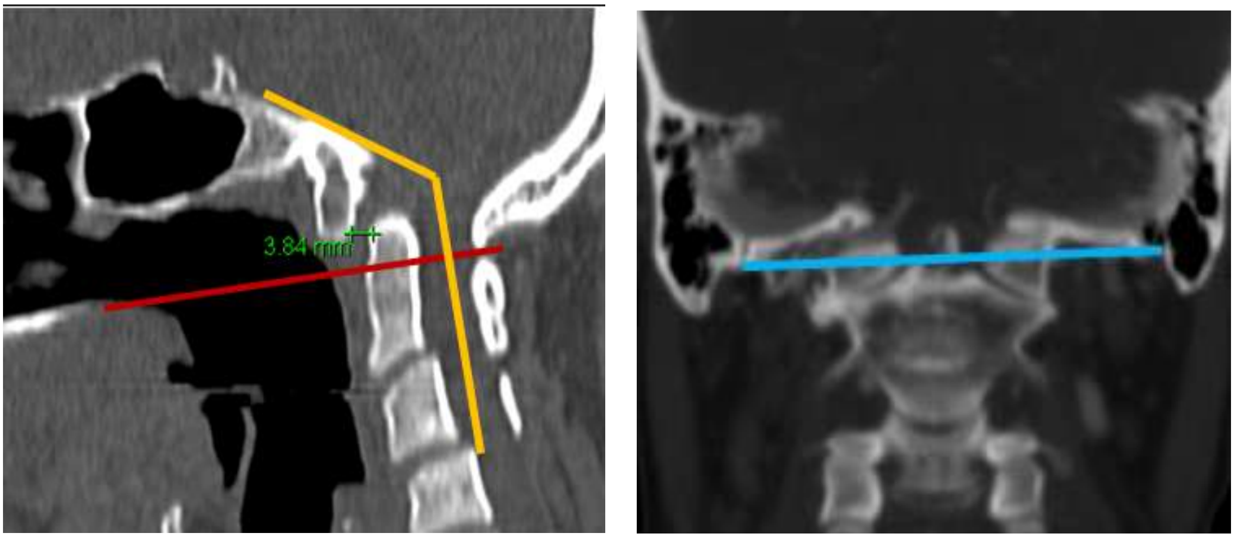


Figure 5: Basilar invagination.
24 year old gentleman with violation of Chamberlain's line (red line) and digastric line (red line), atlantoaxial dislocation (atlantodens interval-3.8mm)

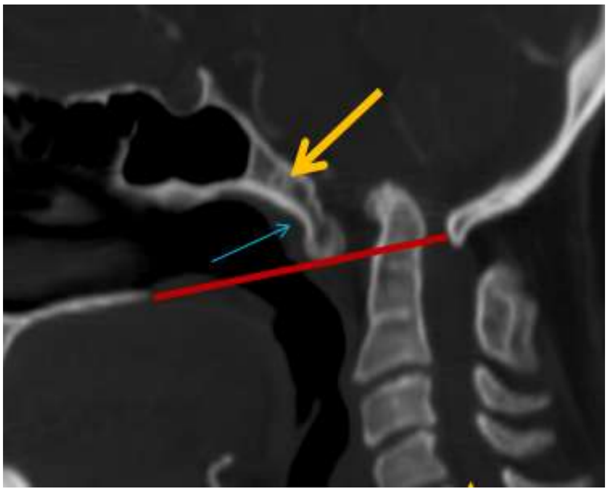


Figure 06: Basiocciput hypoplasia. CT sagittal section showing short clivus (yellow arrow), atlantooccipital assimilation (blue arrow) and violation of Chamberlain's line (red line).

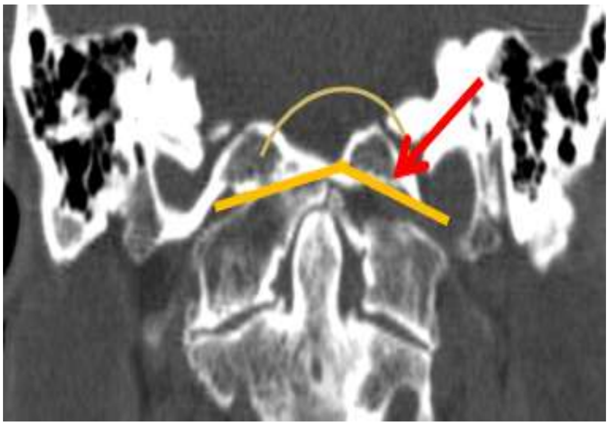


Figure 07: Flattened condyles. CT coronal section showing flattened occipital condyles (red arrow) and widening of atlanto-occipital joint axis angle (yellow line).

Figure 08: Condylus tertius. CT coronal section showing remnant ossification center at distal end of clivus (yellow arrow)

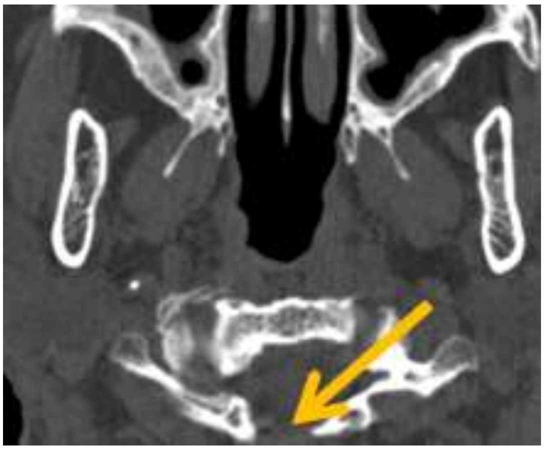


Figure 09: Posterior atlas arch rachischisis. CT axial section showing posterior atlas arch rachischisis.

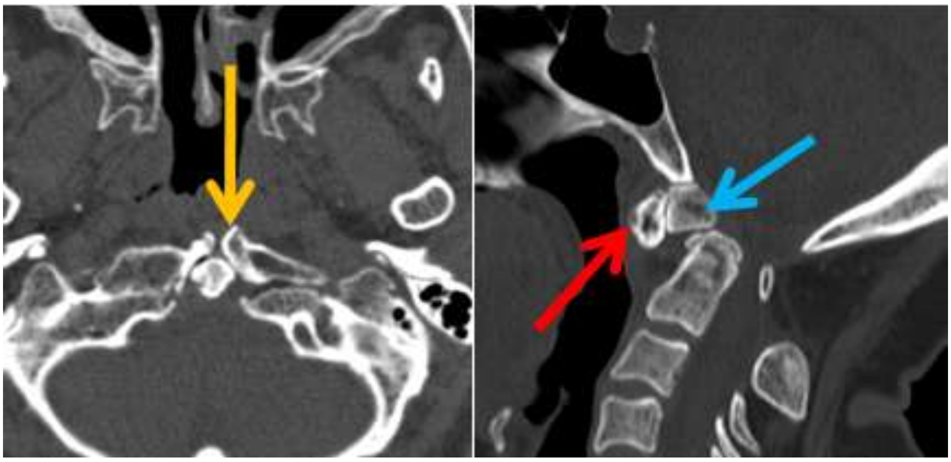


Figure 10: CT axial section showing partial anterior arch rachischisis (yellow arrow) and os odontoideum (blue arrow), hypertrophic anterior arch (red arrow); corticated margins.

Figure 11: T1W MRI sagittal section showing os odontoideum

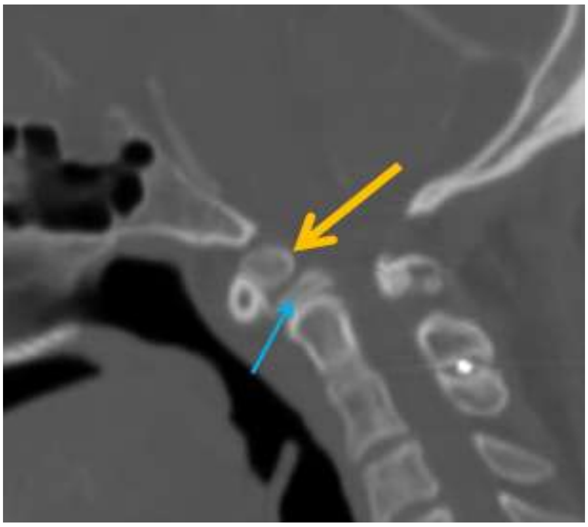


Figure 12: Ossiculum terminale. CT sagittal section showing os odontoideum (blue arrow) with ossiculum terminale (yellow arrow).

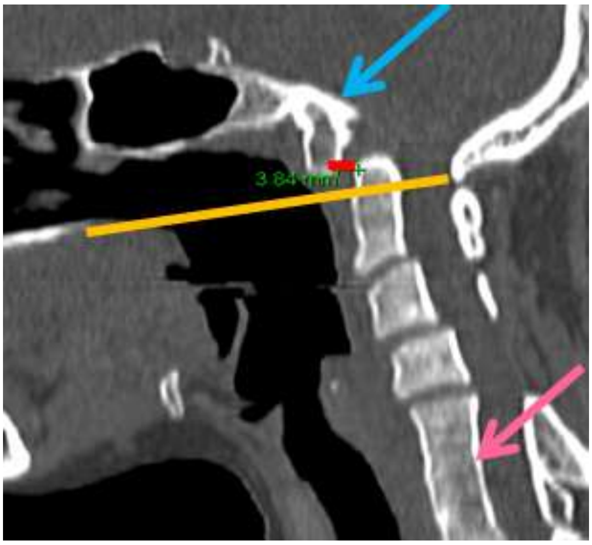


Figure 13: KlippelFeil syndrome. CT sagittal section showing violation of Chamberlain's line (yellow line), atlantooccipital fusion (blue arrow), atlantodens interval of 3.9mm (red line), fused C5-C8 (pink arrow).

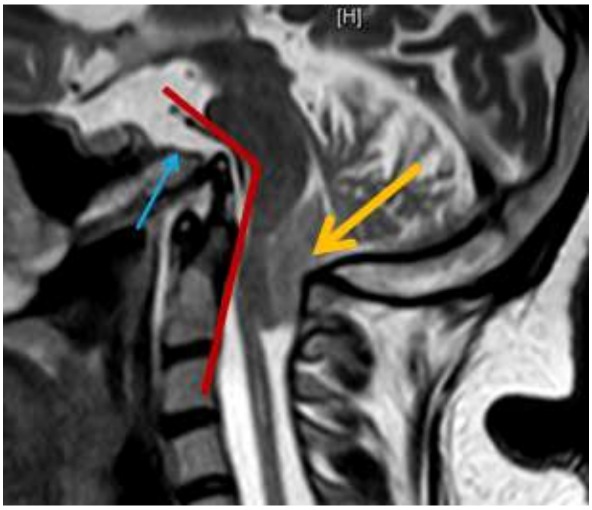


Figure 14: Chiari malformation. 16 year old lady with herniated tonsils (yellow arrow), acute clivocanal angle (red line), short clivus (blue arrow) and cervical cord compression.

Figure 15: Spontaneous Atlantoaxial dislocation.
38 year old lady with increased atlantodens interval.

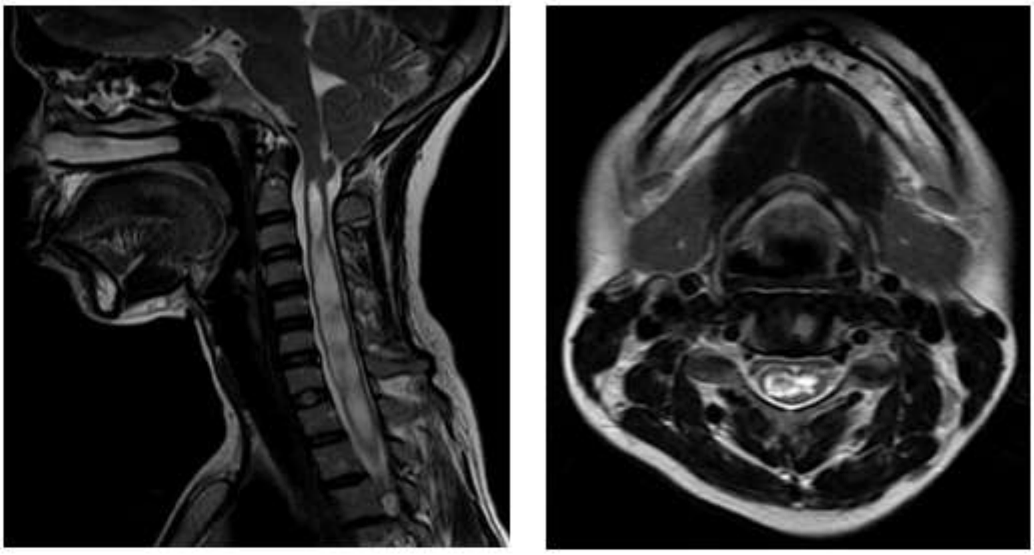


Figure 16: Syringohydromyelia. 32 year old history of numbness and tingling sensation.

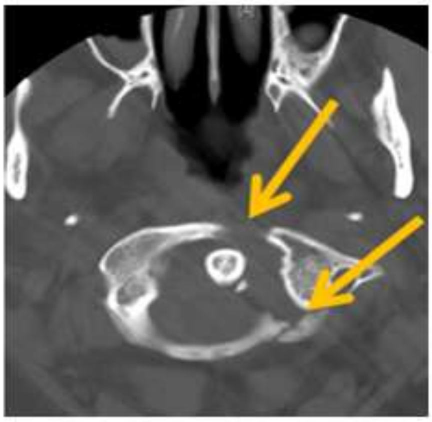


Figure 17: Jefferson's fracture. CT axial section demonstrates displaced fracture of anterior and posterior arches (yellow arrow).

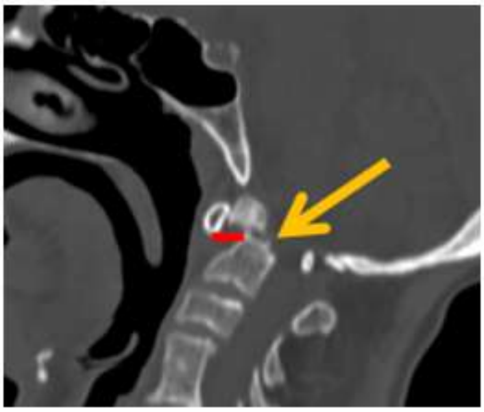


Figure 18: Trauma. 20 year old man with type 2 dens fracture (irregular margins (yellow arrow) and atlantoaxial dislocation (red line).

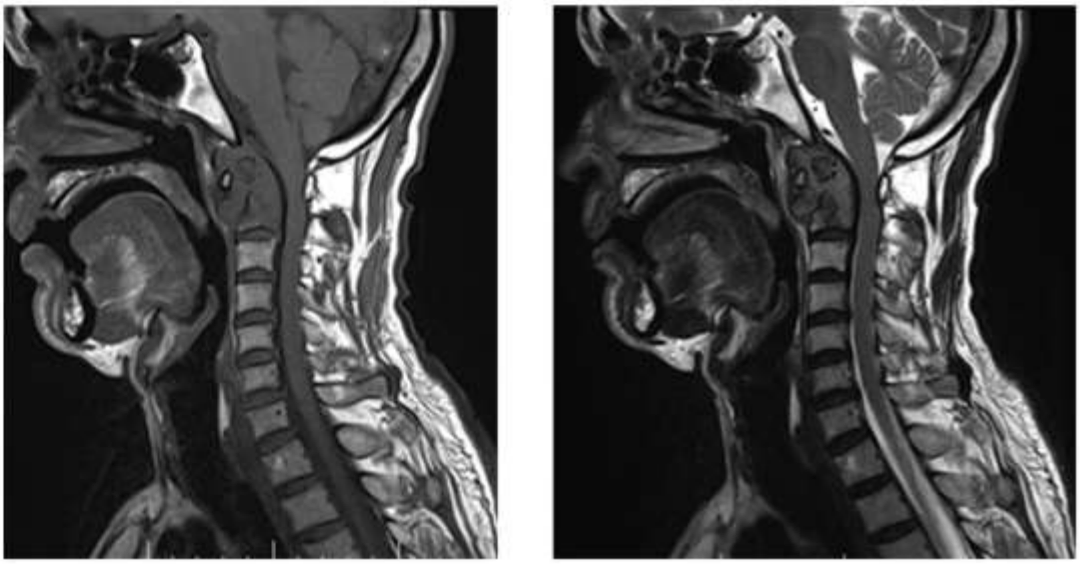


Figure 19: CVJ tuberculosis. 40 year old history of bilateral limb weakness. Sagittal T1 and T2 weighted images showing destruction of odontoid process of atlas and anterior arch of C1 with prevertebral abscess, secondary spinal canal stenosis.

Figure 20: Rheumatoid arthritis. 47 year old lady with basilar impression, sclerosis of atlantoaxial joint (yellow arrow) and atlantoaxial dislocation (red line).

VI. Discussion

Craniovertebral junction (CVJ) extends from a line drawn between the internal occipital protuberance and the midpoint of the distance from the dorsum sellae to the anterior margin of the foramen magnum to the C2-3 interspace level. It encloses the occipital bone, Clivus, foramen magnum and upper cervical vertebrae that is axis and atlas, their articulation and connecting ligaments and soft tissue structures of the cervico medullary junction, which includes mainly medulla, cervical cord, cerebellum and lower cranial nerves. The articulation between the atlas and the occipital bone consists of a pair of condyloid joints. The ligaments connecting the bones are articular capsules, anterior atlantooccipital membrane, posterior atlantooccipital membrane, lateral ligaments and synovial membranes[1, 2]. Various ligaments connecting the axis with the occipital bone are membrana tectoria (occipitoaxial ligament), alar ligaments and apical odontoid ligament. The ligaments connecting atlas and axis are articular capsules, anterior atlantoaxial ligament, posterior atlantoaxial ligament, synovial membranes and transverse ligament of the atlas[3].

Development of the human spine starts in the triploblastic stage of embryo and ends in the third decade of life[4].The occipito-atlanto-axial region has a complex development background. The mesodermal somites, 42 in number appear at the fourth week of fetal life. Each somite differentiates into an outer dermatome, inner myotome and a medial sclerotomes and then cluster around the previously formed notochord[5].

Congenital anomalies of the craniovertebral junction exists as singularly or more than one anomaly and involving both osseous and neural structures[6]. An insult to both may occur between the fourth and seventh week of intrauterine life and may result in a combination of anomalies. A careful look at the embryology and development of the CVJ makes it obvious that anomalies in this region would consist of failures of segmentation, failures of fusion of different components of each bone or hypoplasia and ankylosis[6].

Dysplasia of the occipital segments may flatten the Clivus and platybasia. When the basiocciput and rim of foramen magnum are underdeveloped, the odontoid and arch of atlas may grow normally to overhang along the sides. Odontoid and arch of atlas invaginate resulting in basilar invagination. The proatlas may develop into separate vertebrae and form occipital vertebra. The third occipital condyle represents midline hyperplasia of the proatlas hypochordal bow. At birth the odontoid base is separated from the body of the axis by a segment of cartilage. This synchondrosis persists till the age of eight. The whole of the odontoid process or a part of it may remain separate from the body of the axis (os odontoideum)[7]. The terminal segment of the dens develops from the proatlas and fuses with odontoid process at the age of 12. In case it does not fuse, it persists as "os terminale" Two or more vertebrae that are normally separate may fuse to give rise to condition like Klippel-Feil syndrome and occipitalisation of atlas[7].

Physiology & biomechanics[2, 3]: The cervical spine is the most mobile portion of the spine. The occipito-atlantoaxial complex serves as a transition zone between the vertebral structures and the skull. The unique anatomical configuration of the craniovertebral junction creates distinct biomechanical behavior that differs from other spinal joints. The complex allows necessary range of motion and also supports the head. Spinal movements are characterized by two distinct types of motion: rotations (angular motions) and translations (linear motions). Coupling refers to the simultaneous motions that occur secondary to a main motion.

Congenital CVJ anomalies: Genetics and Etiology- Homeobox or Hox genesthat regulate differentiation processes of the axial and appendicular skeletons, segmentation of the craniocaudal axis by activation and repression of DNA sequences that encode the transcription factors and proteins affecting the order and direction of development of the axial skeleton. Mutations of the homeobox genes may be responsible for congenital anomalies[8, 9, 10]. A familial Klippel-Feil syndrome gene locus[11] on the long arm of chromosome 8. Bavinck and Weaver[12] believed that disruption of the blood supply of the vertebral vessels and their branches during development might be responsible for the vertebral defects. Teratogenic events, such as the maternal consumption of ethanol during pregnancy have also been suggested to cause vertebral anomalies[13]. Congenital anomalies includes condylus tertius, condylar hypoplasia, basiocciput hypoplasia, atlantooccipital assimilation (occipitalisation of atlas), Atlanto- axial dislocation, Atlantoaxial rotatory subluxation, hypoplasia, split, posterior arch abnormalities of atlas, axis anomalies include persistent Ossiculum Terminale, odontoid aplasia, Os Odontoideum, Arnold chiari malformations I to IV, KlippelFeil syndrome.

Acquired CVJ: Tuberculosis usually occurs secondary to TB elsewhere in the body such as pulmonary TB, cervical/mediastinal lymph nodes or other sites. The infection spreads in retrograde direction by lymphatic route to reach the synovial lining of the occipito-atlanto-axial joints. The disease then spreads to ligaments causing ligamentous destruction and instability. Subsequently it extends to the surrounding bone causing destruction and collapse. It usually takes 2 months to 2 years to produce symptoms which are mainly, features of cervico-medullary compression, cranial nerve deficits, atlanto-axial instability and abscess formation[14,15,16,17].

Rheumatoid arthritis [18, 19]: Although the etiology is unknown, the disease process is thought to be initiated by an autoimmune response to an antigen expressed on synovial cells. The chronic release of this antigenic stimulus triggers the body to produce rheumatoid factor (RF), an immunoglobulin (Ig) M molecule against autologous IgG. The pannus contributes to chronic synovitis since it produces collagenases and other proteolytic enzymes capable of destroying ligaments, tendons, cartilage and bone in the CS. These damages potentially result in ligamentous laxity and bone erosions, which may lead to subluxation and instability.Seronegative spondyloarthropathies [20] are a group of related disorders that cause inflammation and ossification ligamentous/tendinous insertion. Ankylosing spondylitis (AS), psoriatic arthropathy, reactive arthritis (Reiter's syndrome) are commonly seen.

Trauma[21, 22, 23, 24]: The CVJ is a very mobile transitional region of the vertebral column. This region's vulnerability to injury is particularly high because of the large lever-arm induced rostrally by the cranium and the relative freedom of movement of the craniocervical junction, which relies disproportionately on ligamentous structures rather than on intrinsic bony stability. Injuries disrupt the structural integrity and vital damages to spinal cord carries a high likelihood of death.

Tumors[25, 26, 27]: The neoplasms at CVJ arise from osseous or extensions from the soft tissue that surround the craniovertebral junction or they are neoplasms that arise from the neural structures contained within the bony anatomy. Osseous tumors includes Chordoma, chondrosarcoma, plasmacytoma, osteoblastoma, fibrous dysplasia, eosinophilic granuloma, metastatic tumor and giant cell tumor. Extra-axial lesions include meningioma, neurinomas, paragangliomas, glomus tumors and less frequent are dermoid, teratomas, neurenteric cysts and arachnoid cysts.

The complex osseous relationships of the CVJ with encompassed multiple neurovascular structures at the cervicomedullary junction a most challenging region for radiologic investigation. The commonest congenital CVJ anomalies were atlantoaxial dislocation, Arnold chiari malformation and less common are occipital assimilation and basilar invagination. The most common age group involved was 3rd decade with male predominance. This study shows that a variety of CVJ are associated with neural anomalies. ACM I and Syringomyelia are the commonest neural anomalies associated with bony CVJ anomalies. The commonest injuries involving the CVJ were odontoid fracture predominantly type II and the commonest combination of injury was C1 with odontoid fracture. Tuberculosis of craniovertebral junction was more common in adults predominantly involving the age group of 4th decade and above. Early diagnosis and treatment are important in preventing long-term neurological sequelae. CT and MRI with gadolinium contrast wherever applicable are the investigation of choice for diagnosis and planning the management.

VII. Conclusion

MRI is the imaging modality of choice for CVJ evaluation for its superior soft tissue characterization. Its multiplanar facility permits the better evaluation of the topographical relationships of structural lesions prior to surgery. Beam Harding artifacts from bone and air containing structures adjacent to brain are eliminated. 3-D reconstructed CT images are better for bony abnormalities at CVJ including lucent fracture lines, displacement of fractured fragments, dislocation, assimilation etc.

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