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Role of imaging in 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 and pre-treatment evaluation significantly affects prognosis and quality of life of patients. Aims: To localize, describe and characterize craniovertebral junction abnormalities, arrange frequently detected CVJ pathologic imaging findings and to emphasize clinical implications to improve our radiological report. Materials and methods: A Cross sectional study done for 24 months in 50 patients of all age groups with clinical suspicion of CVJ abnormalities. Results: In the acquired CVJ abnormality Trauma is the most common etiology(n=15) followed by Tuberculosis(n=4), Rheumatoid arthritis(n=3), Tumors(n=2). Most common age group involved was 2nd decade with male predominance. Most commonly presenting symptom is weakness of limbs followed by neck pain. Most common imaging abnormality is cervicomedullary junction compression and atlanto axial dislocation. ACM I and Syringomyelia are the commonest neural anomalies associated with bony CVJ anomalies. Most common congenital anomalies are atlanto axial dislocation followed by basilar invagination. Only one patient had syndromic association which was Klippel-feil syndrome. 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. Contrast enhancement and bone erosions are the most common findings seen in MRI Rheumatoid arthritis was the most common inflammatory pathology affecting the CVJ region. Formation of pannus and bone erosions are the most common findings seen in MRI. Conclusion: CT and MRI with gadolinium contrast wherever applicable are the investigation of choice for diagnosis and planning the management.

Keywords---Craniovertebral junction (CVJ), Computed Tomography Scan, Magnetic resonance imaging (MRI), Syringomyelia.

Introduction

The craniovertebral junction (CVJ) is a collective term that refers to the occiput (posterior Skull Base), atlas, axis and supporting ligaments. It encloses the soft tissue structures of the Cervico medullary junction (medulla, spinal cord, and lower cranial nerves). The CVJ abnormalities can be either due to bony or soft tissue abnormalities. They can be due to congenital and acquired causes. They are common in all age groups and almost equal in both sex groups. CVJ abnormalities constitute an important group of treatable neurologic 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 must be excluded. Multi-modality imaging of CVJ by using conventional radiography, high resolution CT (MDCT) and magnetic resonance (MR) imaging is integral to the detection, Classification and surgical planning.^{1,2} Our aim of the study is to localize, describe and characterize craniovertebral junction abnormalities, to detected CVJ pathologic imaging findings, to emphasize clinical implications to improve our radiological report.

Materials and Methods

It is Cross sectional study done for 24 months (October 2018 to October 2020) in 50 patients. Study is done in Department of Radiology, Osmania Medical College, Hyderabad.

Inclusion criteria: Patients of all age groups with clinical suspicion of CVJ abnormalities

Exclusion criteria: Patients with unstable general condition, claustrophobia, with MRI non compatible Implants (pace maker, orthopaedic implants etc.).

Imaging Technique

Plain radiograph of CVJ views of cervical spine with minimal magnification are taken by using Samsung machine in translateral view and AP- open mouth view.

CT Imaging Technique: All the patients are evaluated with 128 slice Multidetector Computed Tomography 46 (MAKE:HITACHI) (MODEL:SCENARIA) with 0.35 seconds rotation speed. Area from the lower end of the clivus to the inferior border of C2 vertebra was scanned, and the plane of scanning was kept parallel to the inferior border of the vertebral body of C2. The images were reconstructed in 0.5 mm thick slices and analyzed in bone window settings, length 300 Hounsfield and width 2500 Hounsfield. Post study reconstructions were done at 1 mm. Sagittal and coronal reconstructions were made wherever necessary. The magnification mode was commonly employed, and the scans were reviewed on a direct display console.

MRI imaging technique: All MRI examinations are performed with 1.5 MRI scanner (signa HDxt;GE Medical systems) keeping the patients neck in neutral position

Axial and sagittal T1- and T2-weighted images were obtained in each case. For axial imaging, the standard spin echo technique was used to produce T1-weighted images using TR = 300 to 600 milliseconds and TE = 12 to 15 milliseconds. T2-weighted images were obtained using TR = 3500 to 4000 milliseconds and TE = 110 to 120 milliseconds. The slice thickness for the axial imaging was 2 mm, with no interslice gap, and 3 mm for sagittal images. For sagittal imaging, the spin echo technique was used to produce T1- weighted images using TR = 250 to 400 milliseconds and TE = 12 to 14 milliseconds. T2- weighted images were acquired with TR = 3000 to 4000 milliseconds and TE = 110 to 115 milliseconds. Number of acquisitions ranged between 1 and 3 times. Coronal images were acquired if and when necessary, and T1 spin echo sequence were used. Image acquisition matrix was 256/256 or 512/512.

Results

This study was performed over 24 months from October 2018 to October 2020. A total of 50 patients were included in the study during this period. The diagnosis was based on clinical features and imaging. the study was approved by institutional ethics committee.

Table-1: Age distribution of CVJ anomalies

Age intervals of the patient in years	Number of patients		Total	Percentages
	Male	Female		
0-10	5	2	7	27
11-20	5	1	6	23
21-30	5	2	7	27
31-40	2	1	3	11.5
41-50	2	1	3	11.5
>50	-	-	-	-

Most common age group involved was 2nd decade.

Table-2: Sex and ethiological distribution of the study population

Sex	Number	Percentage
Male	31	62%
Female	19	38%
Etiological group		
Congenital anomalies	26	52%
CVJ trauma	15	30%
CVJ tuberculosis	4	8%
CVJ rheumatoid arthritis	3	6%
CVJ tumors	2	4%

Male predominance is observed in this study. About 48 patients had presented with motor weakness in the form ascending type of Spastic quadriparesis, 44 patients presented with neck pain, about 40 patients presented with sensory disturbances about 10 patients had recent significant head & neck trauma, about 36 patients presented with autonomic disturbances (bowel & bladder disturbance and erectile dysfunction) and about 6 patients presented with vertebro basilar insufficiency in the form of vertigo.

Table-3: Distribution of MR findings in CVJ anomalies and injuries

CVJ anomalies	Number	Percentage
Basilar invagination	8	30.7%
Platybasia	2	7.6%
Occipitalisation of atlas	8	30.7%
Atlanto-occipital dislocation	1	3.8%
Atlantoaxial dislocation(AAD)	12	46%
Hypoplastic clivus	1	3.8%
Chiari 1(cerebellar herniation)	6	23%
Chiari 2	2	7.6%
CMJ compression	16	61.5%
Syrinx	8	30.7%
Klippel feil syndrome	2	7.6%
Odontoid aplasia	1	3.8%
Os odontoideum	5	19%
Hypoplasia of atlas	5	19%
Hypoplasia of condyles	1	3.8%
CVJ injuries		
Odontoid fracture	10	66.6%
Fracture of atlas	2	13.3%
Fracture of axis	2	6.6%
Occipital condyle fracture	1	6.6%
Atlantoaxial dislocation	1	6.6%

Atlanto-occipital dislocation	1	6.6%
Cord compression	3	20%
Cord edema	3	20%
Absent flow void in vertebral artery	1	6.6%

ACM I and Syringomyelia are the commonest neural anomalies associated with bony CVJ anomalies. Most common congenital anomalies are atlanto axial dislocation followed by basilar invagination. Only one patient had syndromic association which was Klippel-feil syndrome.

Table-4: Combination of CVJ anomalies

CVJ anomalies	Number	Percentage
OA+AAD	05	19.2%
BI+OA	04	15.3%
OA+Hypoplasia of atlas	02	7.6%
AAD+OD	02	7.6%
BI+OA+AAD	02	7.6%
PB+OA	01	3.8%

The commonest injuries involving the CVJ were odontoid fracture predominantly type II and the commonest combination of injury was C1 with odontoid fracture.

Table-5: Age distribution in CVJ tuberculosis and CVJ rheumatoid arthritis

CVJ tuberculosis	Number	percentage
0-20	1	25%
21-40	1	25%
41-60	1	25%
>60	1	25%
CVJ rheumatoid arthritis		
0-20	0	0%
21-40	0	0%
41-60	1	33.3%
>60	2	66.6%

Tuberculosis of craniovertebral junction was more common in adults predominantly involving the age group of 4th decade and above. Contrast enhancement and bone erosions are the most common findings seen in MRI

Table-6: MR findings and CT findings in CVJ tuberculosis Rheumatoid arthritis

MR findings	Number of patients	percentage
Bone erosion	3	75%
Soft tissue component	3	75%

AAD	1	25%
Epidural component	1	25%
Contrast enhancement	3	75%
Transverse ligament breach	1	25%
Transverse ligament thickening	1	25%
CMJ compression	2	50%
Cord edema	1	25%
CT findings		
Bone erosion	3	75%
Soft tissue component	3	75%
AAD	1	25%
Epidural component	1	25%
Contrast enhancement	2	50%

Bone erosion, Soft tissue component involved common are the MRI and CT findings in the study.

Table-7: MR findings of CVJ rheumatoid arthritis

MR finding	Number of patients	Percentage
Bone erosion	3	100%
Pannus	3	100%
Ligament thickening	2	66.6%
AAS	2	66.6%
CMJ compression	3	100%

Rheumatoid arthritis was the most common inflammatory pathology affecting the CVJ region. Formation of pannus and bone erosions are the most common findings seen in MRI.

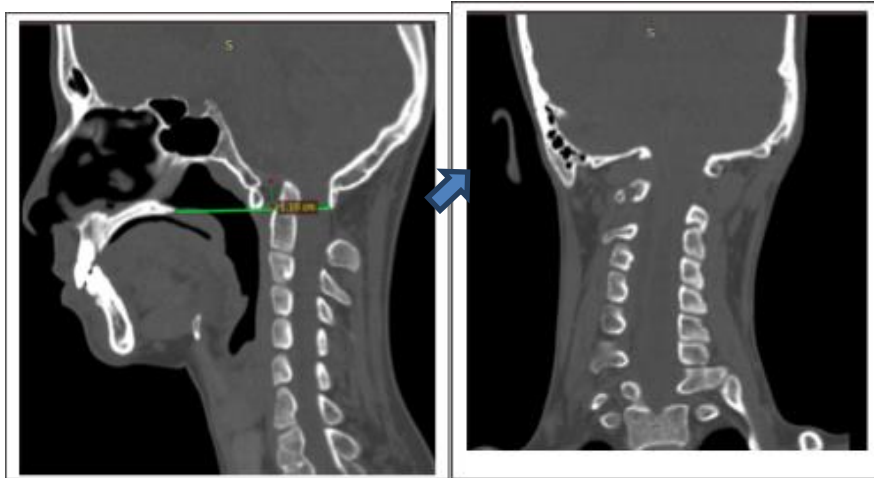
Illustrations

Figure-1: A sagittal image showing basilar invagination (11 mm above chamberlains line) associated with atlanto axial subluxation. B. coronal sections showing occipitalization posterior arch of atlas(arrow).

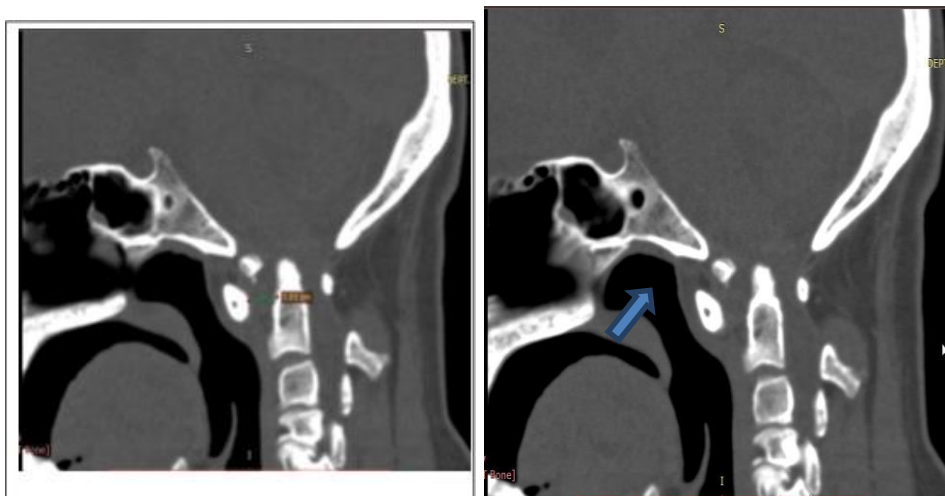


Figure-2: A. sagittal CT image showing increased predental interval measuring 1 cm, s/o atlantoaxial dislocation. B. there is associated accessory ossicle(arrow) s/o condyles tertius and flattened occipital condyle suggestive of platybasia.



Figure-3: A&B plain radiograph and sagittal CT image showing fused C2 and C3 vertebrae (red arrow) with basilar invagination suggestive of Klippel feil anomaly; C. showing complete assimilation of atlas (curved arrow); D. sagittal T2 weighted MR showing compression of CMJ with syrinx.

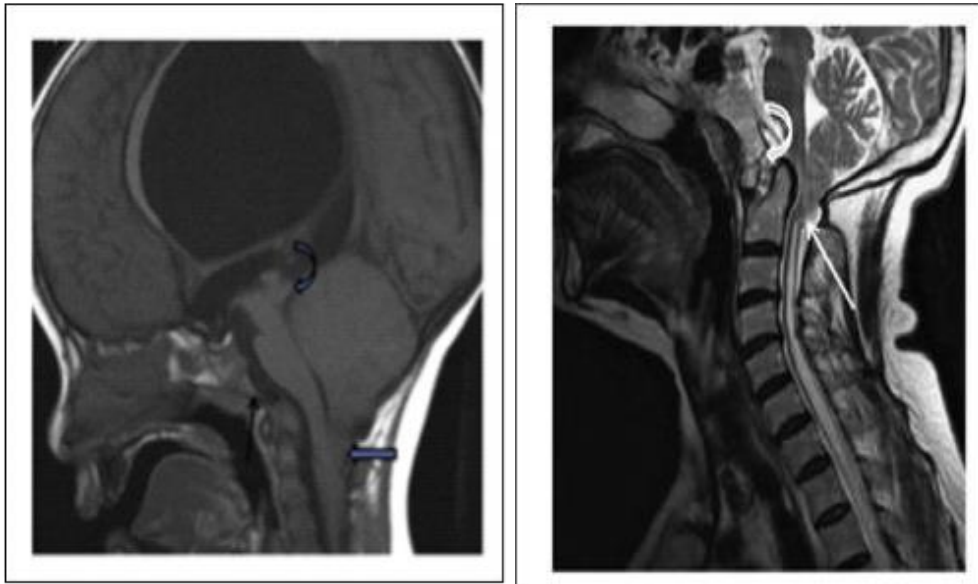


Figure-4: Chiari I malformation, mid sagittal T2 weighted image showing inferior displacement of the cerebellar tonsils with mild basilar invagination. compression of CMJ causing syringohydromyelia.

Right image: Chiari II malformation, sagittal T1 weighted image shows tectal beaking (curved arrow) with concavity of the clivus (arrow). The posterior fossa is small with inferior tonsillar herniation causing obstruction to the outlet foramina and obstructive hydrocephalus.

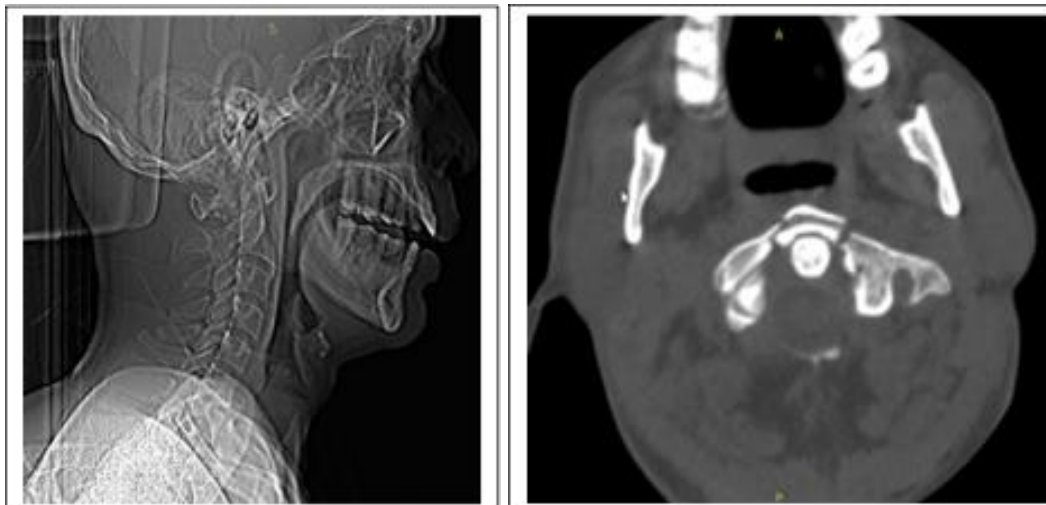


Figure-5: Topogram & axial CT image showing segmental fracture of anterior arch of atlas

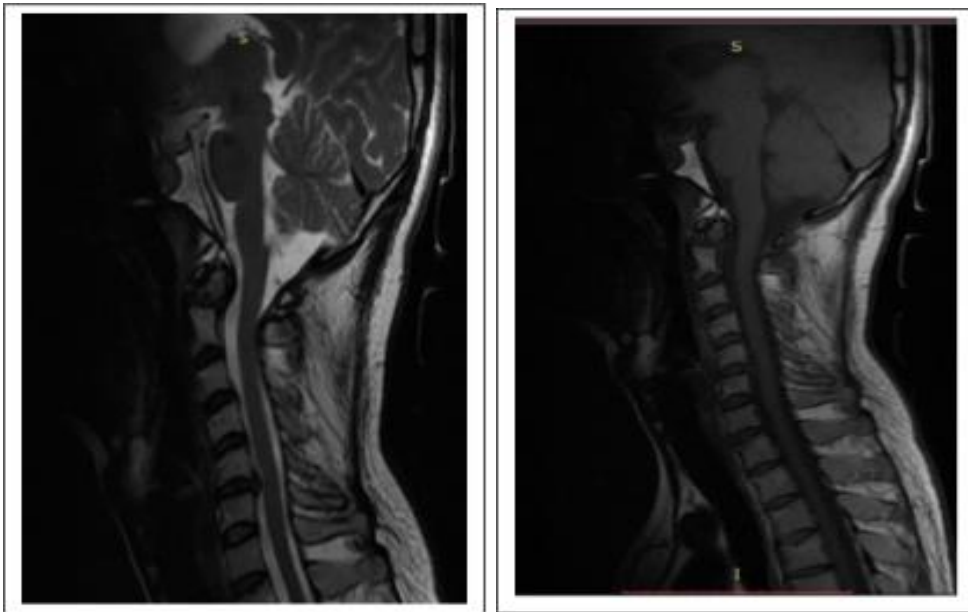


Figure-6: A&B. T1&T2 weighted sagittal images showing fracture at the base of odontoid process (type II) with retropulsion of fractured fragment causing indentation of anterior thecal sac.

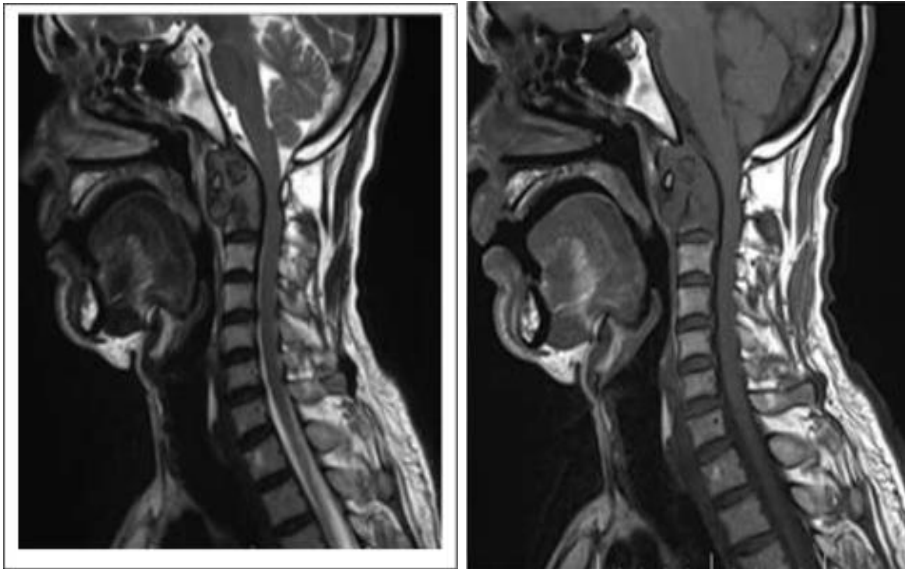


Figure-7: CVJ tuberculosis, 40-year-old patient with history of bilateral limb weakness. Sagittal T1 and T2 weighted images showing destruction of odontoid process of atlas and anterior arch of C1 with prevertebral abscess, secondary spinal canal stenosis.

Discussion

In this study, Developmental or congenital anomalies (52%) were the most common etiology group followed by traumatic (30%), infective (8%), inflammatory (6%), and neoplastic (4%). This distribution of Etiology was comparable with the study done by Rajshree et al. in this study of 62 patients, Developmental anomalies (53.2%) were the most common etiology group followed by Traumatic (16%), Degenerative (12.9%), Infective (6.45%), inflammatory and neoplastic (4.8% each). Another study of 189 cases by Kale and colleagues showed developmental cause as the most common followed by traumatic and tuberculosis in decreasing order.

In this study most common age group 11-20 years followed by 21- 30 years. In this study male to female ratio is 2:1(31 were males and 19 were females) which is comparable to study done by Ramen et al³ in which male to female ratio was 3:1. The most common presenting complaint is weakness of limbs followed by neck pain.

Total number of patients with congenital anomalies are 26, in which 18 were males and 8 were females. Majority of cases were in age group of 0-10 years (27%). This is consistent with the study done by ramen et al. where they reported 29% of congenital anomalies are in age group of 0-10 years. the commonest congenital CVJ anomalies are atlanto axial dislocation (46%), basilar invagination (31%), Arnold chiari malformation (30%), occipital assimilation (30%) were the common radiological features and often exist with each other in varying combinations. The most common combination identified is OA+AAD. other Less common congenital anomalies are os odontoideum, Klippel feil anomaly and platybasia. This is comparable with study done by Rajshree et al⁴ in which most common congenital anomalies are atlantoaxial dislocation (56%), basilar invagination (37%) and occipital assimilation (20%).

Arnold chiari malformations in this study total 8 patients had in which 6 cases were Type I and 2 cases were Type II. Syringomyelia was found in 6 patients and peg like tonsillar herniation was seen in 6 patients. 15 cases of CVJ trauma were reported in our study. Among those 11 were males and 4 were females. Majority of patients were between 30-40 years. The commonest injuries involving the CVJ were odontoid fracture predominantly Type II and the commonest combination of injury was C1 with odontoid fracture.

Table-8: Comparison with other studies

Imaging findings	Signore F et al ⁵	Ramen et al ⁴	Present study
Odontoid fractures	49%%	90%	66.6%
Fracture of atlas	23%	20%	13.3%
Fracture of axis	37%	10%	13.3%
Imaging findings	Lee et al ⁶	Dickman et al ⁷	Present study
C1 + odontoid fractures	32.5%	37.5%	20%
C1+ miscellaneous C2#	15%	28%	10%
C1+hangmans#	10%	12%	0

4 cases of CVJ tuberculosis were reported in this study. Commonest age group was 4th decade and above. Out of 4 cases erosion of dens was seen in 3 patients and osteolytic destruction of anterior arch of atlas was seen in 2 patients. pre and para vertebral abscess was seen in 3 patients. Epidural component was seen in 1 patient. AAD was seen in 1 patient.

Table-9: MR findings with other studies

MR Findings	Krishnan A et al. ⁸	Present study
Dens involvement	62%	75%
Prevertebral soft tissue	75%	75%
Epidural component	86%	25%
CMJ compression	41%	50%
MR findings	Bundschuh et al. ⁹	Present study
Bone erosion	100%	100%
Pannus formation	100%	100%
Atlantoaxial subluxation	50%	66.6%
CMJ compression	75%	66.6%

3 cases of rheumatoid arthritis involving CVJ were reported in this study. In our three patients Erosion of dens and enhancing soft tissue pannus along the dens was seen in all the 3 patients. Atlanto axial dislocation was seen in 2 patients. The most common location of pannus formation was retrodental space followed by predental space which nearly correlated with study done by Bundschuh et al.⁹

2 cases of CVJ Tumors were reported in this study. One case was meningioma which was anterior dural based and another was chordoma arising from clivus. This study shows tumors are the rare acquired CVJ abnormality which is similar to many other CVJ related studies.

Conclusion

CVJ is a most challenging region for radiologic investigation because of the complex osseous relationships of the CVJ with multiple neurovascular structures of the cervicomedullary junction. 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 patients without significant clinical symptoms, screening with plain radiographs is recommended. Knowledge of the craniocervical relationships based on plain radiographs is critical to diagnose cervical instabilities.

Review of these cases confirms that MRI is the imaging mode of choice for evaluation of the CVJ for a variety of reasons like its superior morphologic demonstration of soft tissue structures with multiplanar capabilities. The contrast between different tissues can be manipulated by altering the applied pulse sequence and thus better visualization and resolution of required tissue can be achieved.

Prior to surgery, topographical relationships of structural lesions is better evaluated by the multiplanar facility of MRI. However, a Multislice, Spiral or Helical, 3-D reconstructed CT image is as good and sometimes is better for clear-cut understanding of bony abnormalities at CVJ which can be extremely useful for surgical planning. Multimodality radiological evaluation is necessary for an accurate diagnosis as well as treatment planning.

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