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Research Article

STUDY OF CRANIO-VERTEBRAL JUNCTION ANOMALIES AND ITS ASSOCIATION WITH SOFT TISSUE ANOMALIES

Saumyaleen Roy., Nikhil Srivastava and Anup Singh*

Department of Medicine, IMS, BHU

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A B S T R A C T

Introduction- With the growing prevalence of cranio-vertebral anomalies in northern part of India a study was conducted to know the burden of cranio-vertebral anomalies, its associated clinical spectrum and radiological features in our geographical region.

Methodology- A cross-sectional study was conducted in north eastern parts of Uttar Pradesh at Sir sunderlal hospital, Banaras Hindu University, Varanasi. Patients with clinical features like short neck, low hair line, restricted neck movement associated with signs of high cervical cord myelopathy were included in the study. These patients were then evaluated by x-ray cervical spine along with magnetic resonance imaging (MRI) of cervical spine to look for bony and soft tissue anomalies.

Result- Forty three (43) patients were enrolled in study and all of them had some bony anomalies. Out of 43 patients, 42 patients had atlantoaxial dislocation (98%), 26 patients had platybasia (60%), 22 patients had basilar invagination (51%). Among soft tissue anomalies 10 patients had arnold -chiari malformation (23%) and 9 had syringomyelia.(21%)

Conclusion- Cranio-vertebral anomalies should be considered in patients presenting with short neck and restriction of neck motion along with symptoms and signs of high cervical cord myelopathy. The diagnosis can be easily made on plain radiographs in lateral and antero-posterior view of cervical spine and if a question of instability arises, lateral views in the flexed and extended position are obtained to gain an understanding of the biomechanics. Magnetic resonance imaging is the ideal tool in diagnosing and defining all bony anomalies and soft tissue anomalies except Atlanto-axial instability which is better evaluated on plain dynamic radiographs.

INTRODUCTION

The incidence of Cranio-vertebral junction anomalies (CVJ) in India is yet to be determined but it is more prevalent in the Indian subcontinent and more so from the northern part of the country (Goel,2009). The incidence of different types of abnormalities varies with the demographic environment and has ill-defined genetic factors. The craniovertebral junction (CVJ) comprises the occiput, atlas, and axis. Together, these structures enclose the cervicomedullary junction (CMJ) and the lower four cranial nerves. The signs and symptoms of these anomalies are diverse that include local radicular pain, high cervical cord compression, lower cranial nerves palsy and cerebellar involvement. The clinical manifestations are often delayed into the second and third decade because they are usually subtle and easily missed in children unless looked for specifically.

Corresponding author:* **Anup Singh
Department of Medicine, IMS, BHU

Physical findings are often unique and suggest the diagnosis, more so, when there is an associated syndromes, most commonly Arnold Chiari malformations and the Klippel feil anomaly. Classification of different bony and soft tissue anomalies are given in table 1.

Table 1 The Classification of Craniovertebral Junction Anomalies are

Major bony anomalies	Minor bony anomalies	Soft tissue anomalies
Basilar invagination	Clivus segmentation	Arnold-Chiari malformation
Atlantoaxial dislocation	Remnants around foramen magnum	Syringomyelia and Syringobulbia
Occipitalization	Abnormal occipto atlantal ligament	
Klippel-feil anomaly	Condylar hypoplasia	
Dens dysplasia	Failure of segmentation from occiput	
Platybasia	Atlanto axial fusion	
	Aplasia of atlas arches	
	Irregular atlantoaxial segmentation	
	Segmentation failure of C2-C3	
	Neural dysgenesis	

Different types of abnormalities with a complex pathological bony anatomy needs individual management, decisions tailored for that particular case. The present study was conducted to know the burden of cranio-vertebral anomalies, its associated clinical spectrum and radiological features in our geographical region.

MATERIALS AND METHODS

A cross-sectional study was conducted in Varanasi, eastern parts of Uttar Pradesh at Sir Sundarlal Hospital, Institute of Medical Sciences, Varanasi from June 2014 to June 2017.

Inclusion Criteria- All cases of craniovertebral anomalies with neurological deficit diagnosed clinically followed by plain radiography or through Magnetic Resonance Imaging (MRI) are included in the study

Exclusion Criteria

1. Any absolute contraindication for MRI such as ferromagnetic implants, pacemaker, aneurysm clips .
2. Patient who are unwilling for imaging.
3. Incomplete case records.

Patients were examined by history and examination regarding cervical pain and stiffness, weakness in limbs, sensory symptoms like paresthesias, any gait difficulty, lower cranial nerves (IX-XII) deficit or sphincteric disturbance like urinary frequency, hesitancy, retention or altered bowel habits were taken. Complaints of a short neck or torticollis were also taken into account during patient selection Digital X-ray **(**FCR capsula XL II, fujifilm**)**. Of the skull/brain to include the opisthion, basion, hard palate, and tuberculum sella was included in all the patients undergoing craniovertebral junction radiography. In selected patients, open mouth view through atlanto-axial joint was used. Known measurements and techniques were used to diagnose Atlanto-axial dislocation, Basilar invagination, and Platybasia.For Atlanto-axial dislocation, atlanto-odontoid distance was measured. In adults, a maximum distance of 3mm and in children 5mm (up to age 9 years) was considered as normal. The Basal angle and Boogards angle were used to diagnose Platybasia. Basal angle > 141° and Boogards angle >136° were taken as upper limit in all cases. Chamberlain's line, Mcgregor's line were used to diagnose Basilar invagination. Projection of dens 5mm above Chamberlain's line and 7mm above Mcgregor's line were taken as Basilar invagination.(Fig 1) Odontoid projection equal to or less than than one-third of its entire length above chamberlain's line was considered as normal.

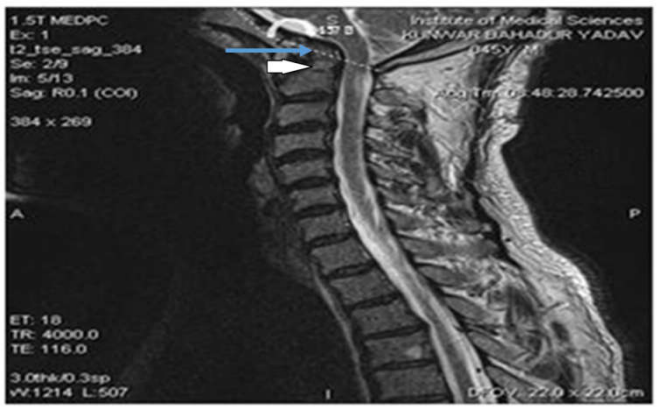

Fig 1 Platybasia in a patient on sagittal T2W MR image Note the co-existent Atlanto-axial dislocation (white arrow), and basilar invagination.(Blue arrow)

Atlanto-occipital joint axis angle was used to diagnose occipital condyle hypoplasia. It was abnormally obtuse (average 124°-127° in normal adults) in occipital condyle hypoplasia. MRI of cranio-vertebral junction including base of the skull was done on all patients who fulfilled the inclusion criteria. MR images were taken in neutral position (not dynamic). All examinations were performed on a 1.5 T, MRI system (Magnetom AVANTO, Siemens Medical Solutions, Erlangen, Germany).Arnold-Chiari malformation was diagnosed on Sagittal MR images of brain and cervical spine by measuring tonsillar herniation >5mm in the absence of any mass lesions(Fig 2). Any coexistent bony anomalies were evaluated in similar way that was used on plain radiographs. Syringomyelia was diagnosed as a hyperintense lesion in spinal cord parenchyma on T2W images.

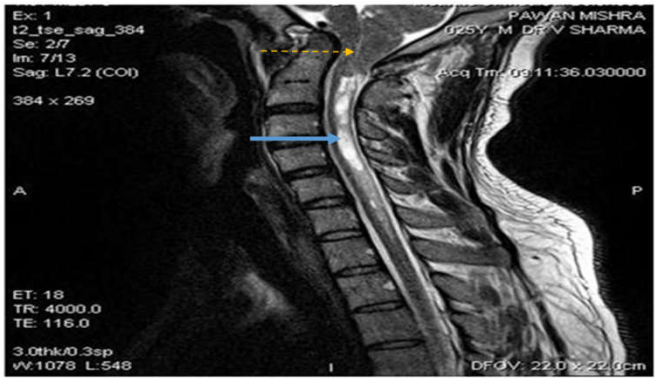

Fig 2 -T2-weighted sagittal MRI demonstrates syringomyelia (blue arrow) with a Chiari 1 malformation (yellow arrow)

It was further classified as Short segment syrinx (spanning less than 2 vertebral levels) and long segment (length >2 vertebral levels).

RESULTS

Forty three (43) patients were enrolled in the study period. Age and sex distribution of patients is shown in figure 3 and duration of illness at the time of presentation is shown in figure 4.

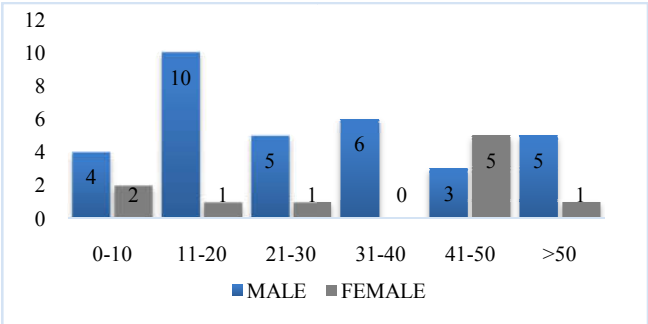

Fig 3 Age and Sex distribution in patients with Cranio-vertebral anomalies

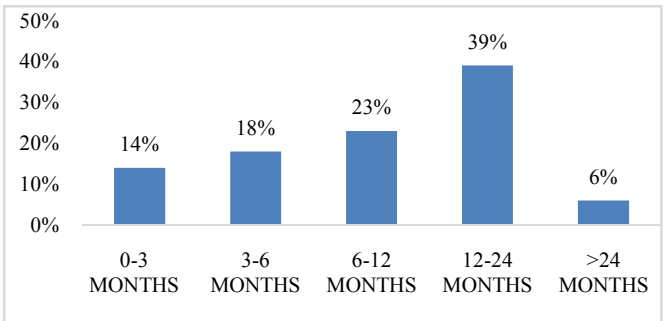

Fig 4 Duration of illness (in months) at the time of presentation

Motor disability and pyramidal tract signs were most common presenting complaints present in 100% of patients. Other presenting features are shown in table 2.

Table 2 Presenting symptoms and signs

Clinical symptoms	No of patients (%)
Motor disability and pyramidal tract signs	43 patients (100%)
Sensory symptoms:-	25 patients (57%)
Paresthesia	20 patient (46%)
Radicular pain (in form of shoulder pain)	5 patient (12%)
Bladder symptoms	4 patients (32%)
Bowel symptoms	1 patient (2%)
Cranial nerve involvement(IX,X,XI,XII)	11 patient (25%)
Cerebellar signs	12 patients (28 %),

Atlanto-axial dislocation was the commenest bony anomaly. Other bony and soft tissue anomalies are shown in table 3.

Table 3 Major type of bony and soft tissue anomalies

Serial no.	Types	Total patients
1	Atlanto-axial dislocation	42
2	Occipitalisation	15
3	Basilar invagination	22
4	Klippel Feil Syndrome	8
5	Platybasia	26
6	Dens Dysplasia	14
7	Tonsillar Ectopia	10
8	Syrinx	9

Among combination of different bony anomalies Atlanto-axial dislocation and Platybasia were most common which was present in 24(56%) of patients.(Table 4).

Table 4 Combination of bony anomalies in patients with Cranio- Vertebral Anomalies

SL. No.	Combination of anomalies	No. of patients	%
1	Atlanto-axial dislocation + Platybasia	24 patients	56
2	Atlanto-axial dislocation + Occipitalisation	15 patients	35
3	Atlanto-axial dislocation + Occipitalisation + Platybasia	11 patients	25
4	Atlanto-axial dislocation + Basilar invagination + Platybasia	16 patients	37
5	Atlanto-axial dislocation + Basilar invagination + Occipitalisation	11 patients	25

Combination of bony and soft tissue anomalies associated with Arnold Chiari malformations is shown in table 5.

Table 5 Association of bony and soft tissue anomalies in patients with Arnold- Chiari malformation

Serial no.	Anomalies	No of patients
1	Atlanto-axial dislocation	10
2	Platybasia	9
3	Basilar Invagination	8
4	Foramen Magnum Stenosis	0
5	Syrinx	7

DISCUSSION

The craniovertebral junction (CVJ) comprises the occiput, atlas, and axis, their articulations and various ligaments. Together, these structures enclose the cervicomedullary junction (CMJ) and the lower four cranial nerves. The craniovertebral junction (CVJ) is a complex transition between the skull and the upper cervical spine, and the brain and spinal cord, respectively. This is the most mobile of the upper cervical spine, especially in children. Thus, the CVJ is a funnel that is uniquely adapted to stability and motion. The unique bony configuration of the atlas and the axis vertebrae, as well as the articulations between the skull, the atlas and the axis vertebrae, allow for a variety of complex movements at the CVJ**.** Imaging plays a major role in identification, defining relation with adjacent structures and extent of cord damage and thus the prognosis. Cervical or occipital pain especially occurring in a younger group of patients raises suspicion of these anomalies. Arnold H. Menezes (2008) mention cervical or occipital pain in 85% of the patients and is in consistent with our findings. However patients mostly present with symptoms and signs corresponding to long tracts dysfunction and dominate over other involved structures like cranial nerve involvement or cerebellum. Sphincteric disturbance was uncommon amongst these patients even with severe myelopathy and profound weakness occurred in the form of urinary frequency and urgency that were mostly mild. Wadia (1967) and Menezes (2008) also found sphincteric involvement to be uncommon amongst patients with cranio-vertebral anomalies. Cause of relatively lower incidence of sphincter involvement in these patients remains unknown but may be hypothesized to be due to gradually progressive symptoms which enables the fibres supplying bladder and bowel to compensate for their vascular supply and remain intact until very severe cord compression occurs. In our study, only lower cranial nerves (i.e. IX- XII) were involved and there was no evidence of other nerve involvement, raised Intra cranial tension (ICT), papilledema, facial paralysis or hearing loss. Sharma *et al.*(1947) also did not found raised intracranial tension or papilledema in any patient. However, Menezes (2008) found VIII cranial nerve involvement to be the most common in his study. Cerebellar signs such as ataxia and nystagmus were infrequent in our study and were found only in association with Arnold- Chiari malformation. Wadia (1967) also found similar findings in their study.

Atlanto-axial dislocation was the most common bony anomaly present in almost all the patients except 1 patient (98%). This particularly high incidence may be related to our selection of patients; neurological deficit was prerequisite. Majority of the patients with completely reducible Atlanto –axial dislocation will present in first and second decade of life. This is consistent with study by Menezes AH (1995) who mentions maximum chance of reducibility of Atlanto –axial dislocation by 4-16 yrs. of age. Alternatively, all the patients with irreducible or fixed AAD usually present after second decade of life.

Basilar Invagination were present in 22 patients in our study. MRI was better than radiographs in detecting the length of tip of odontoid above chamberlain's line.

Occipitalisation was present in 15 patients (35%) There was variable occipitalisation of atlas either limited to the anterior arch, the posterior arch or the lateral masses or alternatively combination of these may also occur. Simultaneous C2-C3 fusion was found in our study in only 4% of the patients. However McRae(1953) found 70% patients of occipitalisation to be associated with C2-C3 fusion. Os odontoideum was present in 6 patients while ossiculum terminale persistence was found in 8 patients.

Platybasia was associated with basilar invagination in majority of the patients. MRI was optimal in assessing both these angles and is the main diagnostic imaging in evaluating platybasia. X-ray was inconclusive in majority of the patients due to poor visibility of bony landmarks and superimposition of osseous

structures. Smoker WRK(2008) also mention MRI and CT to be optimal for the accurate measurements of these angles.

Klippel – Feil anomaly was most commonly involving C5-C6 vertebrae followed by C6-C7 vertebrae. It was associated with Atlanto-Occipital fusion in majority of the patients. This might be due to our selection of patients (Neurological deficit was prerequisite). In addition, 2 patients showed spinal cord compression adjacent to the fused segments of vertebrae due to cervical canal stenosis. Similar observation was also mentioned by O'Donnel *et al* (2008) and Auerbach *et al* (2008) Soft tissue anomalies like Arnold-Chiari malformation and/or Syringomyelia were associated with various combination of bony anomalies in all the cases. Thus its important to look for bony anomalies in patients with Arnold- Chiari malformation. Goel (2012) mentioned that soft tissue anomalies occur secondary to bony anomalies, chiefly atlanto-axial instability which was consistent with our study. Also patients with Arnold – Chiari Malformation do not show changes in foramen magnum measurements suggesting that herniation occurs secondary to the reduced posterior fossa size. This is similar to what Amer *et al*(1997) observed.

Syringomyelia was seen most commonly in association with Arnold-Chiari malformation. However patients may develop syringomyelia even in the absence of tonsillar herniation. It was found in cervical area in all cases and was long segment in majority of patients. This is consistent with observation of Oldfield H *et al* (1994) who mentions that maximum systolic wave pressure impacts on upper cervical canal and thus the consequent syrinx formation.

CONCLUSION

The diagnosis of Cranio-vertebral anomalies should be considered in patients presenting with short neck and restriction of neck motion along with symptoms and signs of high cervical cord myelopathy. The diagnosis can be easily made on plain radiographs in lateral and antero-posterior view and if a question of instability arises, lateral views in the flexed and extended position are obtained to gain an understanding of the biomechanics. MRI is the ideal tool to use after plain radiographs are obtained. It shows excellent neural abnormalities and poor prognostic markers such as myelomalacia in addition to providing complimentary information on co-existing bony anomalies.

However routine MRI (done in neutral position) may underestimate the atlanto-axial instability, more so with the reducible type, due to the altering biomechanics on neck motions. Thus, dynamic MRI rather than conventional MRI will be a better modality in patients with instability. However, dynamic MRI is cumbersome to perform and its widespread unavailability in our setup limits its use. However, to assess the burden MRI to be superior to plain radiographs in diagnosing and defining all bony anomalies and soft tissue anomalies except Atlanto-axial instability which is better evaluated on plain dynamic radiographs.

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