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"CRANIO-VERTEBRAL JUNCTION ANOMALIES- SPECTRUM ON MAGNETIC RESONANCE IMAGING"

Medical Science

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ABSTRACT

OBJECTIVE: This prospective study was done in the Department of Radiodiagnosis, King George Medical University, Lucknow, over the period of one year from September 2007 to July 2008. The purpose of this study was to evaluate the spectrum of cranio-vertebral junction anomalies on Magnetic resonance imaging (MRI). **METHODS:** Fourty patients for whom MRI of cervical spine including cranio-vertebral junction was done for varying symptoms and showed imaging features of cranio-vertebral junction anomalies were selected for the study. Results were presented in numerical and percentage forms. **RESULTS:** There were 34 males and 6 female patients in the age range of 3-60 years. Maximum number of patients were in the age group between 11-20 years (18 patients-45%) followed by age group of 21-30 years (7 patients-17.5%). Most common presenting symptom was weakness of both upper & lower limbs seen in 16 patients (40 %) followed by neck pain & stiffness in 15 patients (37.5%) and sensory symptoms in 11 patients (27.5%). Weakness of lower limbs, lower cranial nerve dysfunction & bladder bowel symptoms were the least common presenting symptoms seen only in 1 patient (2.5%). Atlanto-axial instability was the most common abnormality present in 30 patients (75%). Occipitilisation of atlas was the second most common abnormality seen in 20 patients (50%) and basilar invagination; third most common abnormality seen in 14 patients (35%). Other less commonly found abnormalities were ossiculum terminale (in 6 patients-15%), platybasia (in 5 patients-12.5%), aplasia of atlas arches(in 3 patients-7.5%), os odontoideum(in 3 patients-7.5%), segmentation failure of C2-C3 (in 3 patients-7.5%), clivus segmentation (in 2 patients-5%). Atlanto-axial fusion(in 1 patient 2.5%) & hypoplasia of dens (in 1 patient 2.5%). Most common associated feature was syrinx formation found in 9 patients (22.5%). Out of 40 patients, 34 patient had developmental anomalies (85%) while 6 patients had acquired causes, including tubercular in 5 patients-12.5% & rheumatoid arthritis in 1 patient -2.5%. Myelopathic changes were found in 28 patients (70%) out of which motor symptoms were present in 25 patients (89.29%) .

KEYWORDS

Atlas, Axis, Craniovertebral junction anomalies, CVJ, MRI, Occiput

INTRODUCTION:

The cranio-vertebral junction (CVJ), lying between upper cervical spine and skull is the most complex structure of the axial skeleton **[1]**. It is a bridge between mobile cranium and relatively rigid spinal column. Anatomically, it consists of occiput, first (atlas) and second cervical (axis) vertebral segments, their connecting ligaments and articulations. All these structures, together protect the cervico-medullary junction. Although CVJ anomalies are defects of development, manifestation may not be present at birth. A detailed knowledge and understanding of bony anatomy, embryology and biomechanics of CVJ is important to be familiar with various anomalies associated with CVJ **[2]**. Magnetic resonance imaging (MRI) can be considered as a revolutionary modality to image the CVJ anomalies due to its excellent soft tissue resolution. In addition, it can also image associated anomalies of cervico-medullary junction along with bony abnormalities with a high degree of detail.

Here, we report usefulness of MRI to evaluate spectrum of various CVJ anomalies.

METHODS AND MATERIALS :

This prospective study was done in the Department of Radiodiagnosis, King George Medical University, Lucknow, over the period of one year from September 2007 to August 2008.

40 patients for whom MR imaging of cervical spine was done for symptoms of weakness of upper & lower limbs, tingling and numbness in upper limbs & neck pain and showing imaging features of cranio-vertebral junction anomalies, were selected for the study. Patients who were unco-operative, claustrophobic, having pacemaker/cochlear implants in situ and with history of significant cervical trauma were excluded from the study.

In all the patients selected for the study, detailed history was taken with respect to age, sex, occupation, socio-economic status & clinical profile. The findings of MRI in various CVJ abnormalities were then correlated with the demographic findings and clinical profile of patients.

MRI Examination:

MRI was performed on SIGNA EXCITE 1.5T GEMSOW (GE) MR scanner installed in Department of Radiodiagnosis, with patient in supine position with head first with holding deglutination, obtaining Trans-axial T2-GRE, T1-FSE, Coronal T2-Fr FSE, Sagittal T1-FSE, T2-FrFSE, T2-FrFSE fat suppression & Gadolinium enhanced fat suppression TIWI images in those where required.

Relevant craniometric lines which were taken for the study are as follows.(2)

RELEVANT LINES IN LATERAL VIEW

Sl. No.	Synonyms	Definition	Normal measurement	Implications
1	McRae's line (Foramen Magnum line)	Joins anterior and Posterior edges of Foramen magnum (basion to opisthion)	Tip of dens does not exceed this line	Odontoid process bisects the line in basilar invagination, When effective Sagittal diameter is <20mm, neurological symptoms occur (foramen magnum Stenosis
5.	Boogard's angle	Angle between McRae's line and line drawn from dorsum sellae to basion along the plane of clivus	122°	>135 ° in basilar invagination
8.	Atlanto-axial distance	Distance between anterior arch of C1& dens in neutral position	<3mm in adults & <4mm in children	Increased in atlanto-axial dislocation

RELEVANT LINES IN ANTERO-POSTERIOR VIEW

Sl. No.	Synonyms	Definition	Normal	Implications
1	Fishgold digastric line	Joins the fossae for digastric	Dens tip should not project	Corresponds to McRae's line on lateral view: may be oblique in unilateral

	(biventer line)	muscles on undersurface of skull (just medial to mastoid process)	above this line. Central axis of dens should be perpendicular to the line	condylar hypoplasia: oblique odontoid suggest paramedian abnormality
2.	Schmidt-fischer angle	Angle of axes of atlanto-occipital joints	<124°	Angle is wider in condylar hypoplasia

RESULTS:

- Maximum number of patients were in the age group between 11-20 years (18 patients-45%) followed by age group of 21-30 years (7 patients-17.5%).
- Among 40 patients, 34 were male and 6 female in 3-60 years of age range.
- Most common presenting symptom was weakness of both upper & lower limbs seen in 16 patients (40 %) followed by neck pain & stiffness in 15 patients (37.5%) and sensory symptoms in 11 patients (27.5%).
- Weakness of one sided upper & lower limb was found in 9 patients (22.5%).
- Neck deformity was found in 8 patients (20%) & fever was found in 7 patients (17.5%).
- Weakness of lower limbs, lower cranial nerve dysfunction & bladder bowel symptoms were least common presenting symptom seen only in 1 patient (2.5 %).
- Atlanto-axial instability was the most common abnormality found in 30 patients (75%).
- Occipitilisation of atlas was the second most common abnormality found in 20 patients (50%).
- Basilar invagination was the third most common abnormality found in 14 patients (35%).
- Other abnormalities found were ossiculum terminale (in 6 patients-15%), platybasia (in 5 patients-12.5%), aplasia of atlas arches(in 3 patients-7.5%), os odontoideum(in 3 patients-7.5%), segmentation failure of C2-C3(in 3 patients-7.5%) & clivus segmentation (in 2 patients-5%).
- Atlanto-axial fusion & hypoplasia of dens each was found in 1 patient (2.5%).
- Most common associated finding was syrinx formation found in 9 patients (22.5%).
- Chiari I malformation was seen in 5 patients (12.5%) .
- Other less common associated abnormalities were segmentation anomaly of other vertebral bodies in 2 patients (5%) intramedullary space occupying lesion & meningocele each in 1 patient (2.5%).
- Among 40 patients with combined craniovertebral junction anomalies, 34 patient were having developmental anomalies (85%) and rest of the 6 patients were having acquired causes (which included 5 patients -12.5 % tubercular & 1 patient -2.5% rheumatoid arthritis).
- Myelopathic changes were seen in 28 patients (70%) out of which motor symptoms were present in 25 patients (89.29%) .

DISCUSSION:

The cranio-vertebral junction (CVJ) comprises the occiput, atlas, and axis and is visible in MR imaging studies of the head & neck. MR imaging is well suited for evaluating the CVJ because of its wide availability, direct sagittal imaging capabilities; thus, it has renewed the understanding of CVJ anatomy and anomalies which is important for all radiologists**[3]**. There are numerous congenital anomalies of the CV Junction. These can be simple and clinically silent to complex with serious neurologic manifestations **[4].** MRI scan provides clear pictures of soft tissues and is the investigation of choice for spinal diseases **[3,9**]. In addition Dynamic MRI is extremely useful for evaluating CVJ abnormalities and, in particular, cord compression & cases of mobile atlanto-axial instability **[5]**.

CVJ is a transition site between mobile cranium and relatively rigid spinal column. It is also the site of the medullary-spinal junction. An understanding of the development of the cranio-cervical junction is essential to recognize the pathological abnormalities **[6]**.The exact incidence of CVJ anomaly is yet to be established in our country. In our study, this problem was found more common in age group of 11-20 years and more common among male than female population.

There are various craniometric lines, planes and angles which are used to define normal anatomical landmarks and abnormalites of CVJ namely McGregor line, McRae line, Chamberlain's line, Wackenheim clivus line, Welcher basal angle, clivus canal angle, height index of Klaus, Fishgold digastric line, atlantooccipital joint axis angle (Schmidt angle) and Fishgold bimastoid line(7).

SPECTRUM OF CRANIO-VERTEBRAL JUNCTION ANOMALY & CLINICAL PROFILE

In our study, there were 34 males and 6 female patients . Maximum number of patients were in the age group between 11-20 years (18 patients-45%) followed by age group of 21-30 years (7 patients-17.5 %). Compared to studies conducted by Jawalkar et al(8), Sankhe et al(7) and Rajshree U. et al(9), male to female ratio was higher in our study while results of age range were comparable.

Most common presenting symptom was weakness of both upper & lower limbs (40 %) followed by neck pain & stiffness (37.5%), this was in contrast to the study conducted by Sankhe et al(7), who had neck pain as the most common presenting symptom followed by weakness of limbs. In our study, the other presenting symptoms were weakness of one sided upper & lower limbs found in 22.5% and sensory symptoms in 27.5% patients.

Neck deformity was found in 8 patients (20%) & fever in 7 patients (17.5%). Lower cranial nerve dysfunction & bladder, bowel symptoms were found each only in 2.5% patients. Thus, the most common presenting symptom was weakness of both upper & lower limbs.

In our study, atlanto-axial instability was the most common abnormality found in 30 patients (75%). Atlanto-axial instability is termed when there is > 3 mm of distance between anterior arch of atlas and dens in adults & > 4 mm of distance in children in neutral position or > 3 mm of translation between atlas and axis during flexion & extension movement of spine.

Congenital atlanto-axial dislocation, in clinico-radiological practice was seen in 51.5-68% of cases of CVJ anomalies**(10)**, our results were comparable to this study. Atlanto-axial instability often produces symptoms by causing narrowing of the spinal canal leading to compression over cord and compressive myelopathy. Occipitilisation of atlas was the second most common abnormality found in 20 patients (50%). Basilar invagination was the third most common abnormality found in 14 patients (35%). These three anomalies presented in different combination along with other less common cranio-vertebral junction anomalies including ossiculum terminale (15%), platybasia (12.5%), aplasia of atlas arches, os odontoideum, segmentation failure of C2-C3, clivus segmentation etc.

Among 40 patients with combined craniovertebral junction abnormalities, cord compression with compressive myelopathic changes were seen in 28 patients (70%). Most common associated feature was syrinx formation found in 9 patients (22.5%). Chiari I malformation was found in 5 patients (12.5%). Other less common associated abnormalities were segmentation anomaly of other vertebral bodies in 2 patients (5%), intramedullary space occupying lesion & meningocele each in 1 patient (2.5%).

DEVELOPMENTAL VERSUS ACQUIRED CAUSES

Out of total 40 patients with combined cranio-vertebral junction anomalies, 34 patient were having developmental anomalies (85%) and rest of the patient were having acquired causes (which included 5 patients -12.5% tubercular & 1 patient -2.5% rheumatoid arthritis).

Thus, patients presenting with varying symptoms of CVJ anomalies were having more number of developmental causes than acquired and these results were comparable with the previous studies done by Rajshree et al (9) and Bhagwati et al(11).

CONCLUSION:

The anatomy of the cranio-vertebral junction, though complex, is well visualized on MR imaging(12). MRI is superior to conventional radiograph for demonstration of cervico-medullary junction thus making it an excellent tool for the demonstration of normal anatomy as well as various anomalies of CVJ, compressive myelopathic changes in the cord and details of surrounding soft tissue structures(13, 14).

Conflict Of Interest :- None

Figure 1. Atlanto-axial dislocation

Fig. 1-Mid Sagittal T2 Fat Suppresed Image- Showing Atlanto-axial Dislocation- Seen As Increased Distance Between Anterior Arch Of Atlas And Dens

Figure 2. Occipitilisation And Hypoplasia Of Atlas

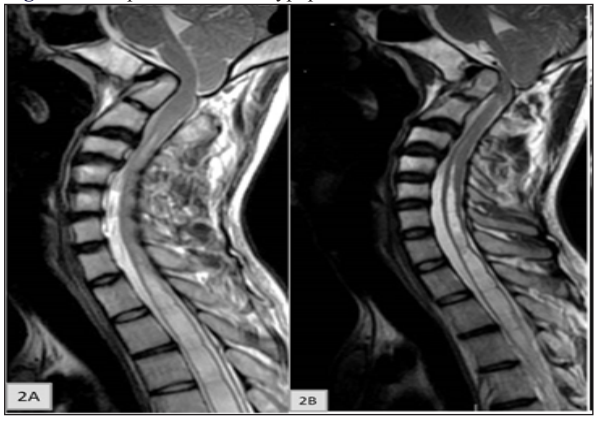

FIG. 2- Sag T2 FSE Image(A, B) showing occipitalisation of anterior arch of atlas with poor visualisation of posterior arch s/o hypoplasia along with basilar invagination & kinking of cervico-medullary junction. Downward displacement of cerebellar tonsils s/o associated Chiari 1 malformation and syrinx in cervical cord noted.

Figure 3. Basilar Invagination

Fig.3- Midsag T2 FSE Image –showing atlanto-axial dislocation with basilar invagination and compressive myelopathic changes in spinal cord seen as hyperintense signals within.

Figure 4. Ossiculum Terminale

FIG.4- Mid Sag T2 FSE Image - showing ossiculum terminale with atlanto-axial dislocation and compressive myelopathic changes in cervico-medullary junction.

Figure 5. Os odontoideum

Fig. 5- Mid Sag T2 FSE Image- showing os odontoideum seen as a separate bony part from the body of axis .

Figure 6. Os odontoideum

Fig. 6- Mid Sag T2 FSE image-showing os odontoideum along with atlanto-axial dislocation and body of C2 causing kinking of cervico-medullary junction & compressive myelopathic changes in cervical cord. Fracture of dens is a differential.

Figure 7. Clivus Segmentation

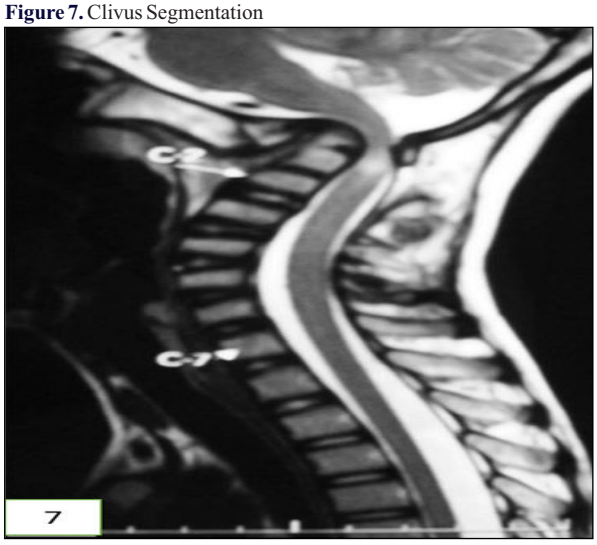

Fig. 7- Mid Sag T2 FSE-showing Clivus Segmentation Along With Occipitilization Of Anterior Arch Of Atlas, Basilar Invagination And Compressive Myelopathic Changes In Cervical Cord.

Figure 8. Platybasia

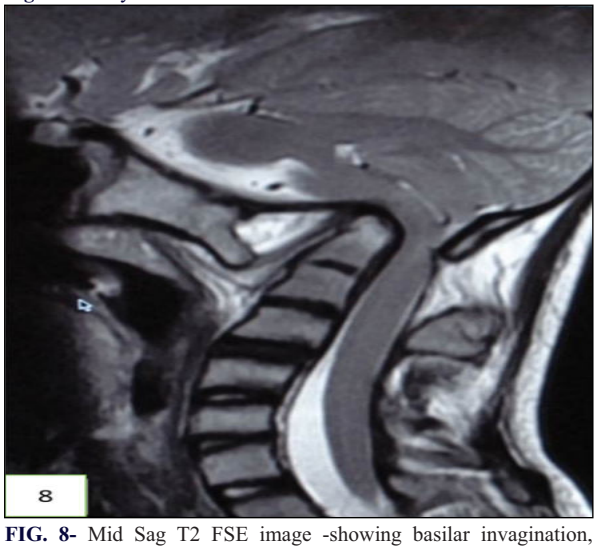

FIG. 8- Mid Sag T2 FSE image -showing basilar invagination, complete assimilation/occipitilization of atlas & compressive myelopathic changes in cervicomedullary junction.

Figure 9. CVJ Tuberculosis With Atlanto-axial Dislocation

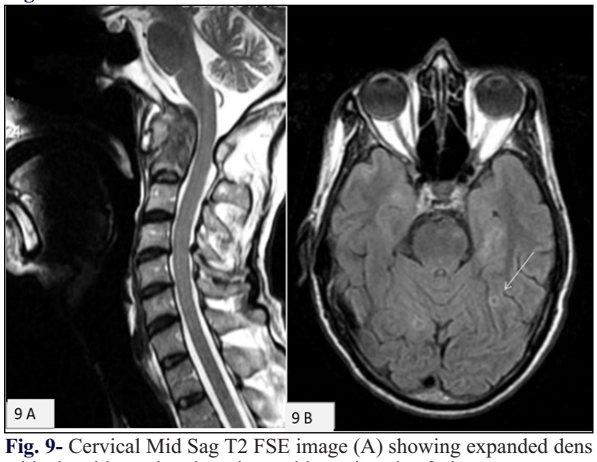

Fig. 9- Cervical Mid Sag T2 FSE image (A) showing expanded dens with signal intensity alterations with predental soft tissue component leading to atlanto-axial dislocation. Axial T2 FLAIR Brain(B) image- Multiple ring lesions in the b/l occpital lobes of same patient – tuberculomas.

Figure 10. Intra-medullary Space Occupying Lesion

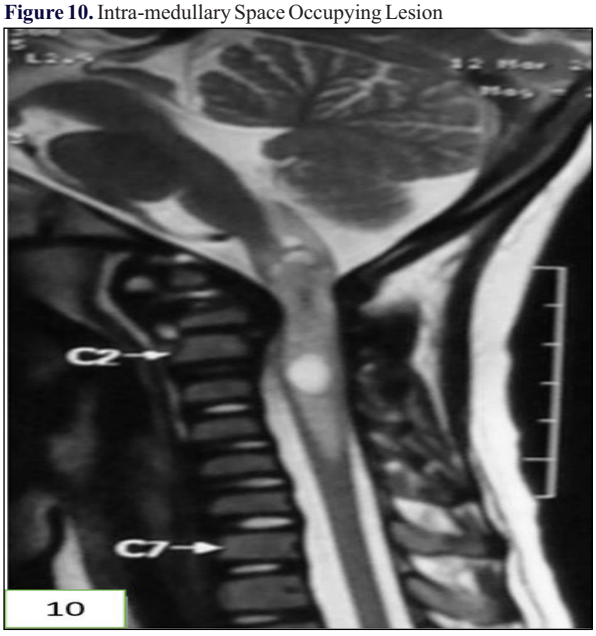

FIG. 10- Mid Sag T2 FSE image –showing intramedullary heterogenous signal intensity lesion in cervico-medullary region-likely neoplastic . Ossiculum terminale was also seen.

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