ISSN: 0974-2115
www.jchps.com Journal of Chemical and Pharmaceutical Sciences

A rare case with basilar invagination anomaly in craniocervical junction: role of CT & MRI

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ABSTRACT

In this paper we will present a 49 years old Iranian man with a totally different craniocervical junction. The case has referred with severe headache, as his chief complaint, to the emergency ward of Hazrat Rasoul hospital (Tehran, Iran) in March 2012. After admission and primary examinations, in order to find the cause of headache, the patient was referred to imaging department requesting brain CT scan. While, the case wasn't aware of his unusual cervical anatomy; after complementary examinations, i.e. MRI, we found an abnormal form of cervical spine junction with cranium and completely different craniocervical junction and chronic altered power ratio while the patient had no pathological symptoms.

KEY WORDS: Cervical spine, craniocervical junction, CT, MRI.

1. INTRODUCTION

The craniocervical junction (CCJ) is a functional unit composed of the occiput, atlas, and axis along with its neural components such as brainstem and spinal cord (Vender and McDonnell, 2001). The CCJ constancy is mostly based on robust compound ligamentous and bony constructions that are also liable for the majority of axial rotation and flexion-extension movements (Nessej, 2015). A common CCJ anomalies is basilar invagination (BI), in which a radiographic finding is signified by a prolapsed spine into the skull-base is identified when the tip of the odontoid process is situated at least 2 mm above the Chamberlain's line (Joaquim, 2014). Many osseous irregularities are related with BI, such as clivus and condyle hypoplasia, atlas integration and subaxial vertebral body fusion. Tonsillar herniation is defined as a related neural outcome that may cause neurological damage and can drive surgical management (Joaquim, 2014). Widely known reasons of BI include mechanical (force) or embryological dysgenesis, genetic abnormalities, and viral infections. The continuing slippage of the atlas over the axis secondary to this misalignment, a process similar to spondylolisthesis in the lumbosacral spine, may cause invagination of the odontoid process into the craniocervical cord (Goel, 2009).

The BI may be congenital, e.g. in the following conditions: osteogenesis imperfecta, Klippel-Feil syndrome, achondroplasia, Chiari malformations, cleidocranial dysostosis, and Schwartz-Jampel syndrome. Additionally, it could be acquired, e.g. in the following circumstances: rheumatoid arthritis, Paget's disease, traumatic injuries, hyperparathyroidism, and osteomalacia; which this condition is often associated with platybasia (Menezes, 1980).

According to Goel (2009), classification scale, the BI is founded on the presence or absence of clinical and radiological instability (Joaquim, 2014). This classification is important since it may assist surgeons to decide the best form of surgical treatment. Based on the Goel classification scale patients are classified in two groups:

Group A: The main characteristic of this group is the occurrence of a clear instability of the region manifested by the tip of the odontoid process spacing itself from the anterior arch of the atlas or the inferior end of the clivus. Diagnostic criteria including 1) atlantoaxial dislocation (generally with C1-C2 subluxation), and 2) tip of the odontoid process protruded into the foramen magnum (FM) and above the Chamberlain line, McRae line and Wackenheim's clival line. Patients may have also Chiari malformation with tonsillar herniation and syringomyelia (Joaquim, 2014).

Group B: There is no instability. The alignment of the anterior arch of the atlas, the odontoid process and the inferior aspect of the clivus remains normal. Probably, this condition is subordinate to a complete clivus hypoplasia (Joaquim, 2014).

Platybasia can be found in both groups, but it occurs most commonly in group B. Diagnostic criteria including 1) atlantoaxial relation are normal, 2) tip of the odontoid process is above the Chamberlain's line, and 3) tip of the odontoid is below the Wackenheim's clival line and the McRae's line. The distance between the ponto-medullary junction and the odontoid process is reduced markedly. Posterior cranial fossa volume could be evidently reduced in this group. Chiari I malformation is commonly found (Goel, 2009). Atlas (C1) vertebra is the principal cervical vertebra, which supports the cranium and helps in the transmission of weight to the axial skeleton. The atlas vertebra is the leading cervical vertebrae. It tolerates the straight mass of the skull, like the legendary Greek Titan Atlas had the sphere on his shoulders. The atlas vertebra encounters the occipital condyles, which margin the FM in the lower part of the occipital bone of the skull. This junction forms the atlanto-occipital joint, and is accountable for the main articulation between the spine and the skull. It is the only vertebra in the spine, which has no vertebral body. The atlas vertebra, in turn reposes on the axis vertebra, which is another cervical vertebra in the spine, with the articulation between these two vertebrae happening at lateral articular surfaces and a unique juncture between a concave facet (on the atlas) and an upward protruding structure on the axis called the odontoid process (dens). The axis (C2) vertebra is the sturdiest cervical vertebrae which is like an axle for spining of the atlas and head round the robust dens, which projects cranially from the superior surface of the body (Taccone, 1993).

Conventional radiography is an inexpensive and readily available means to evaluate the CCJ, although it offers relatively poor sensitivity and specificity in the detection of injury. X-ray film and routine computed tomography (CT) could result in the failure in the diagnosis due to their shortcomings in displaying delicate irregularity or no differentiation of the normal features of atlanto- axial joints. In comparison with radiography or routine CT, multi-detector CT scan (MDCT) offers significantly improved fracture detection. Additionally, MRI accurately depicts cord integrity and soft tissue injuries such as disk annulus, ligament, muscle, and joint capsule. MRI complements MDCT in the assessment of cervical spine injury has an important role in surgical planning and clinical prognostication (Muchow, 2008; Rostamzadeh, 2014).

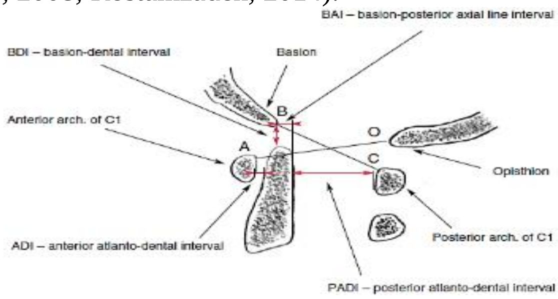

Figure.1. A schematic representation of anatomical position of baselines in the CCJ region.

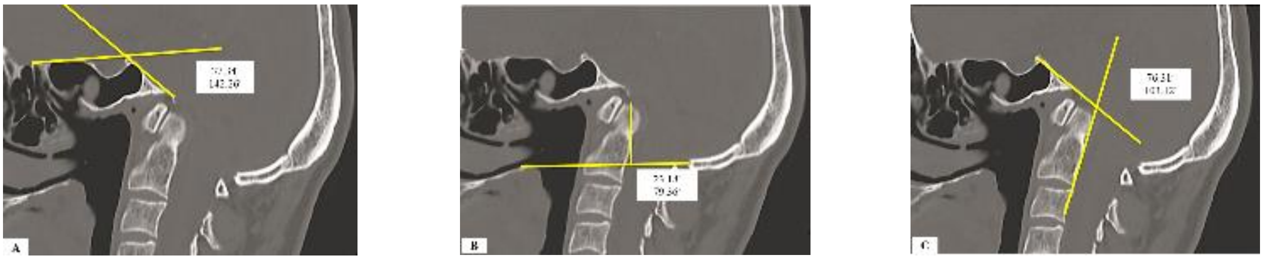

Figure. 2. A) The common baseline of cranio cervical junction (CCJ) region. Welcher-basal angle which was measured from a line extending from the body of sphenoid and another from the clivus. B) Chamberlain's line extending from the posterior edge of the hard palate to the opisthion with the tip from the odontoid process. C) The clivus-canal (craniovertebral) angle which measured from a line along the clivus crossing with a line along the posterior portion of the body of the C2. This angle varies between 150° and 180° depending on flexion/extension position and may represent spinal cord compression in situations of it being less than 150°.

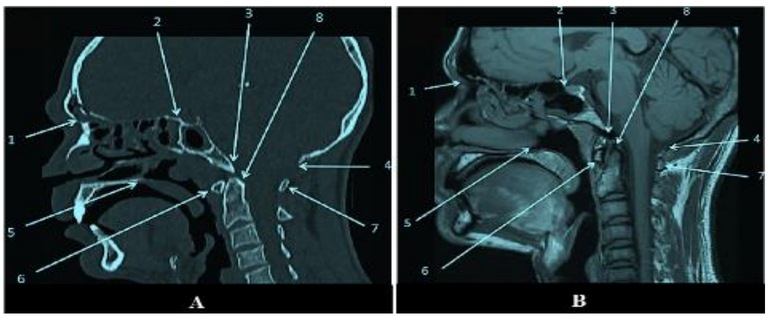

Figure 3. The anatomical landmarks in midsagittal plane of cranial base and cranio cervical junction (CCJ) region in CT (A) and MRI images (B). 1) nasion, 2) tuberculum sella, 3) basion, 4) opisthion, 5) posterior margin of hard palate, 6) anterior arch of atlas, 7) posterior arch of atlas, 8) odontoid process.

In the CCJ region there are some important virtual lines, angles and planes which are consider as reference points (figures. 1-3, table 1). These guidelines are using for assessment of anatomical abnormalities like boney-, ligamentous-, and joints disorders. They also are useful for management design in skull base surgeries and removing tumors of post cranial cavity and supra-tentorial tumors. The FM is an important landmark of the base of skull and is of particular interest to many fields of medicine. Variations of the form of FM have diagnostic, clinical and radiological importance. Furthermore, there exists some correlation between the shape of FM and ancestry of an individual. The dimensions of FM have clinical significance since the important structures that cross it can face density as in the cases of FM achondroplasia and FM brain herniation (Patel and Mehta, 2014).

Table.1. The most important anatomical lines, angles, and plans in the cranio cervical junction (CCJ) region.

Line/angle	Anatomic landmarks/relationship	Normal value
Wackenheim line	Posterior surface of clivus and dens	Tangent to the dens and behind it, may cross its posterior third
Chamberlain line	Hard palate to opisthion and dens	Dens protrudes <5 mm above it
McRae line (FM line)	Basion to opisthion and dens	Dens below line
McGregor line	Hard palate to basiocciput and dens	Dens protrudes <7 mm above it
Ranawat line/criterion	Distance in mid-dens coronal plane, between transverse axis of atlas and midpoint of C2 pedicle	>15 mm in males >13 mm in females
Klaus posterior fossa height index	Perpendicular distance between dens and tuberculum/internal occipital protuberance line	>30 mm
Welcher basal angle	Nasion to tuberculum line and tuberculum to basion line	<140°
Clivus-canal (craniovertebral) angle	Wackenheim line and posterior axial line	150° in flexion–180° in extension
Cervico-medullary angle	Measured on MRI, anterior medullary line and cervical spinal cord line	>135°
Basion-dental interval (BDI)	Distance of basion to dens tip	<12 mm
Basion-axial interval (BAI)	Perpendicular distance from basion to posterior axial line	<12 mm
Atlanto-dental interval (ADI)	Distance dens to anterior C1 arch	<3 mm in adults <5 mm in children
Condyle-C1 interval (occipito-atlantal joint gap)	Distance C1 lateral mass and occipital condyle	<2 mm adults <5 mm children
Clark Station	Atlantoaxial vertical relationship. Anterior atlas ring relative to axis height divided into equal vertical thirds	Ring of atlas opposite the upper third of axis

The frontal arch of the atlas bears a constant relationship to the dens. The predental space between these structures should be no wider than 3 mm in adults and 5 mm in children. The posterior arches of the atlas merge in the midline to form the posterior tubercle. This creates a dens arc that aligns with the spinolaminar line. In people in whom fusion has not occurred in the posterior arch of the atlas, this dens arc is absent and there is hypertrophy of the anterior arch. Alignment then depends on assessment of the anterior structures. In the past decades one of the most difficult areas to assess radiographically was the CCJ. Nowadays, numerous methods had been desired to allow one to make a clear diagnosis of occipito-atlantal subluxation or dislocation. Two of these will be mentioned only for their historical significance, since CT has rendered them obsolete as radiographic methods. However, they still are valid on sagittal CT reconstructed images (Daffner, 1996).

The reason that we presented this case is that the dens axis of the patient was abnormally located toward the brainstem while it had no effect on the vital signs of the case. This is a unique and rare condition for patient.

Case presentation: The presented case is a 49 years old Iranian man who referred to the emergency department of Hazrat Rasool hospital (Tehran, Iran) in March 2012. His chief complaint was severe headache, while he also had cervical pain, dysphagia, and upper limb paresthesias. When we faced with the case; in the first step, we referred him to perform a plain radiography of the CCJ region, i.e. lateral view with flexion and extension of cervical curvature. The X-rays were done with FFD of 170 cm and central point on the thyroid cartilage. The lateral view of the cervical X-ray showed BI as well as imitated movement of the cervical spine.

Moreover, cranial nerve function includes abducent and hypoglossal nerves which that are most commonly affected by craniocervical injuries were a part of examination of our patient.

After conventional radiography and physical examination, the case was referred to the imaging department for brain CT scan. A MDCT (Aquilion, 16 slice; Toshiba Medical Systems, Tokyo, Japan) was applied to perform the CT slices. In order to image acquisition, CT scan was done with no contrast media applying scanning protocol including 12-mm collimation with 1mm slice thickness, 24 cm field of view (FOV), 120 kVp, 300 mA, 512^{x}512 matrix size, 0.9-1 pitch, and 7.5 mm imaging increments. Axial images were reconstructed at 1 mm. Reforms in sagittal and coronal planes were acquired from the 1 mm axial reconstructions, reformatted to 3-mm thickness every 3 mm through the entire upper cervical vertebrae and base of occiput bone. The images obtained were independently evaluated by a neuroradiologist with a 10 year experiments in neuroimaging on the workstation which access to 3D (three-dimensional) reconstruction in an identical window parameter. The patient was not aware of his unusual cervical anatomy. In the serial images of the brain CT scan, we discovered an unusual form of cervical spine junction with cranium and completely different power ratio without any pathological symptom.

Images obtained through CT revealed that FM was semi-circular (roughly oval) shape in which sagittal diameter was 32.1 mm and transverse diameter was 27.4 mm.

The posterior atlantodental interval (PADI) is defined as space among the posterior surface of the dens and the anterior margin of the posterior arch of C1. The PADI is used to assess atlantoaxial subluxation. A minimum length of 14 mm is required to avoid cord compression. The PADI in our case was severely decreased (9 mm) resulting canal stenosis and cord compression. Additionally, the CT sagittal images illustrated basion–dens interval (BDI) was actually below zero and Chamberlain's line was become very steep.

An MRI scanning was also performed with a 1.5 Tesla MR Scanner (Avanto, Siemens Medical System Co., Germany), with a spine coil in flexion and extension position of cervical curvature. A T2-weighted fast spin echo sequence in the mid-sagittal image was obtained for measurement. The MRI with settings of repetition time/echo time 4000/120 ms, inversion time 160 ms, flip angle 90°, slice thickness 3.0 mm, inter-slice gap 0.1 mm, number of slices 15, FOV 240 × 240 mm^2, voxel size 0.8 × 0.5 × 1.0 mm^3, and echo train length 30. The MRI images show severe pressure effect of dens on medulla oblongata and same mild effect on lower part of cerebellar vermis. Moreover, anomaly there was a decreasing in the space and the volume of the fourth ventricle as well as in cerebrospinal fluid (CSF) circulation. Furthermore, severe syringohydromyelia was evident. The MRI images also demonstrated that the thickness of transverse ligament in the middle part was 1.2 mm, which was significantly less than the normal conditions (5-6 mm).

2. DISCUSSION

The spine is a "scaffold" for the erect human body and spinal cord that allows information to transit between the central nervous system and the peripheral movement executers (Hradil). The CCJ is an anatomically critical area where the brainstem, several cranial nerves, arteries and veins exist in a restricted space. There are several methods to describe the spatial relationship between the occiput, atlas, and axis. The most popular in the setting of trauma are the BDI and the basion-posterior axial line interval (BAI) (Bono, 2007). Originally described by Harris, the BDI and BAI have been carefully examined (Figure 2).

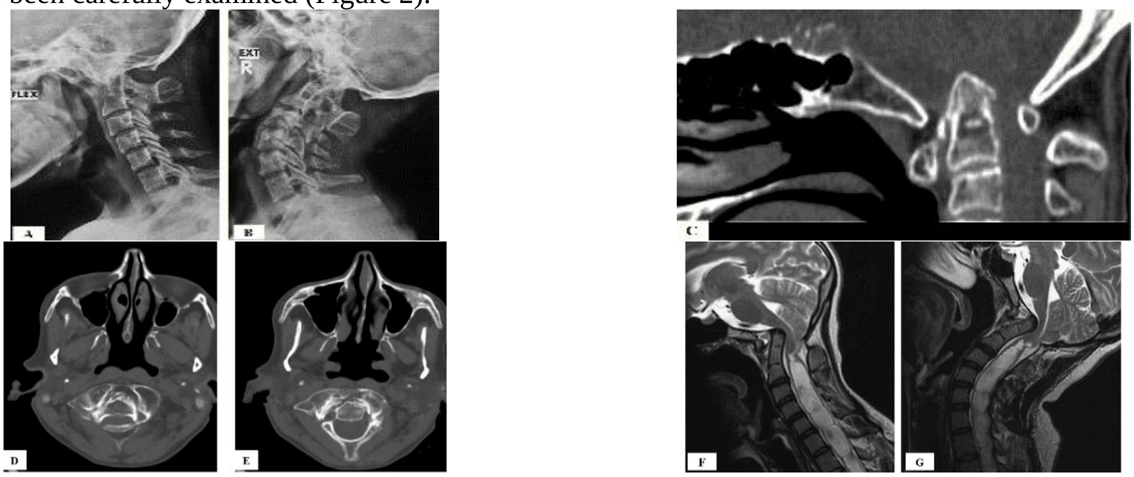

Figure.4. Lateral cervical radiograph images in flexion (A) and extension (B) position of our case shows basilar invagination (BI). Sagittal CT scan (C), and Axial CT scan (D and E) images in two different levels that show severely decreasing of PADI and steeping of Chamberlain line result in BI. The Sagittal T2W images of the patients in flexion (F) and extension (G) positions that shows severe pressure effect of dens to medulla oblongata and its packing compression.

In the first of the two companion studies, Harris. measured the BAI and BDI on lateral radiographs of 400 normal adults. The BAI and BDI did not exceed 12 mm in 98% and 95% of adults, respectively. The BDI and BAI have come to be known as Harris Measurements, and more descriptively as the Rule of Twelve. Management of inherited craniocervical irregularities requires a deep analysis of the neural and bony anatomy of the CCJ. Surgical treatment has a high rate of morbidity and mortality, especially in previously treated patients in nonreferral centers, although the benefits of surgery overweight the risks. Treatment should be individualized based on the patient anatomy, presence or absence of instability and surgeon's experience. Attention to the principles mentioned above has proven to be helpful in improving surgical results in the treatment of these patients (Joaquim, 2014).

As studies show McGregor and Chamberlain lines are measurable and evaluable only in the lateral view of the conventional cervical spine X-ray as well as in the sagittal view of CT scan. Furthermore, the important disadvantage regarding these lines is that for evaluation of BI the only landmark and reference is hard plate. Therefore, abnormalities and deformities of the regions such as face, mouth, larynx, pharynx, and nasal could affect on the accuracy and precision of the measurement and confirmation of the diagnostic when using these lines. In this study we have used cervico-medullary angle, as a reference point, in order to do final confirmation of BI; which was merely measurable through MRI. Thus, one can say that in comparison with conventional radiography, MRI is a valuable and helpful modality to find out and confirmation of BI. Respecting the Chamberlain's line, a diagnostic problem is that whether the examiner is noticed to the caudal surface of the FM edge or its cranial surface. The correct method is notice to the cranial/caudal edge of the FM. In neurosurgical approach, the transcondylar approach is frequently used to gain the lesions which are ventral to the brainstem and cervicomedullary junction. It was stated that considering the bony anatomy of the condylar area is significant in this method (Morera, 2010).

In our case the transverse diameter of the FM was slightly less than the normal cases and other patients with BI anomaly. This might be the reason for the symptoms mentioned earlier for our case. One main stabilizing ligament in the CCJ region is transverse ligament; which is the largest, the strongest, and the thickest craniocervical ligament. The transverse ligament goes posterior to the odontoid process of C-2 and attaches to the lateral tubercles of the atlas bilaterally. The transverse ligament upholds steadiness at the CCJ by locking the odontoid process anteriorly alongside the posterior aspect of the anterior arch of C-1, and it divides the ring of the atlas into two compartments: the anterior compartment houses the odontoid process, and the posterior compartment covers chiefly the spinal cord and spinal accessory nerves (Tubbs, 2011).

In the presented case the thickness of the transverse ligament is 0.8 mm, which is very thinner than normal cases. Therefore, the low thickness of this ligament might be the main reason for this anomaly in our case and extension of the dens axis toward the brainstem; because the craniocervical articulation is a very mobile transitional region of the vertebral column.

Since the presented case had dysphagia and upper limb paresthesias, our conclusion is that it could be due to the pressure of the dens axis on the medulla oblongata. The result of this pressure is destroying the spinal cord in the cervical region and involving nucleolus of the hypoglossal nerve (12^{th} nerve pair) and vagus nerve (10^{th} nerve pair). The reduction in the thickness of the spinal cord may affect on the nerve's root of the neck region as well as arm resulting upper limb paresthesias. Involving nucleolus of the medulla oblongata could be a reason for the case dysphagia. Furthermore, the patient also suffered from severe headache. This might be due to syringohydromyelia which could cause a defect on the CSF circulation in the brain ventricles and finally increases intra-cerebral pressure and headache (Rusbridge, 2006).

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