Editor-in-Chief : Atul Goel (INDIA)
Open Access HTML Format
For entire Editorial Board visit : http://www.jcvjs.com/editorialboard.asp
Journal of Craniovertebral Junction and Spine

Editorial

Is Chiari malformation nature's protective "air-bag"? Is its presence diagnostic of atlantoaxial instability?

Atul Goel

Department of Neurosurgery, K.E.M. Hospital and Seth G.S. Medical College, Parel, Mumbai, Maharashtra, India

Corresponding author: Dr. Atul Goel, Department of Neurosurgery, K.E.M. Hospital and Seth G.S. Medical College, Parel, Mumbai - 400 012, Maharashtra, India
E-mail: atulgoel62@hotmail.com

Journal of Craniovertebral Junction and Spine 2014, 5:25

Several authors have evaluated the subject of Chiari malformation since its first description by Cleland in 1883.[1] A number of hypothesis and speculations have been proposed as to the cause and effects of Chiari malformation.[2-6] A congenital anomaly is generally agreed to be the cause. Smaller posterior fossa volume that in unable to contain the cerebellar mass that herniates into the spinal canal has been the most accepted pathogenic phenomenon. Structural defects caused by genetic mutations, or lack of proper vitamins or nutrients in the maternal diet are also considered to be the probable cause. The cerebellar tonsils herniate into the spinal canal through the foramen magnum and appear to cause compression of the critical neural structures.

Frequently, the precise pathogenesis of Chiari malformation is unclear or not recognized. In some of such cases, Chiari malformation is associated with bone malformation and basilar invagination. In cases where no definite cause is identified or when there are associated bone anomalies, Chiari malformation is generally considered to be a component of the entire complex and a congenital anomaly. Basilar invagination is a frequent bony accompaniment and syringomyelia is a frequent soft tissue association. Associated syringomyelia is also considered to be a result of an error in embryogenesis. Syrinx formation is frequently implicated to blockage of the cerebrospinal fluid flow pathways at the level of foramen magnum by the herniated tonsils.

Access this article online	
Quick Response Code:	Website: www.jcvjs.com
	DOI: 10.4103/0974-8237.142302

Chiari malformation is rarely associated with tumors in the posterior cranial fossa,[7] hydrocephalus and meningomyelocele. It does seem that Chiari malformation is always secondary to an obvious or an unidentified pathology. Identification and treatment of the primary pathological cause rather than direct manipulation of the tonsils appears to be the solution.

We have evaluated the subject of craniovertebral junction for over 30 years.[8-12] During our study, we identified that atlantoaxial instability forms the primary point of instability at the craniovertebral junction.[8-12] Atlantoaxial joint is the center for movements, and occipitoatlantal joint is the center for stability of the most mobile and most stable joint-complex of the body at craniovertebral junction. The atlantoaxial joint is not only the center for mobility but is also a center of instability.

Atlantoaxial dislocation in cases with basilar invagination was considered to be irreducible or fixed until not long ago. Such an understanding resulted in treatment protocol that essentially focused on decompression of compressing bone elements either from anterior route by transoral surgery or by posterior route by foramen magnum bone and dural decompressive surgery. The identification that despite the fact that dynamic imaging does not reveal mobility of atlantoaxial instability, the atlantoaxial joint in such cases is abnormally or pathologically mobile and is potentially reducible by manual manipulations has revolutionized the treatment paradigm for such clinical anomalies.[10] The current treatment strategy for hitherto considered irreducible or fixed atlantoaxial dislocation is essentially focused on craniovertebral realignment and atlantoaxial stabilization.[13]

Atlantoaxial dislocation or instability has been essentially radiologically assessed and confirmed by the distancing of the odontoid process from the anterior arch of atlas. Our recent study on the subject has revealed that atlantoaxial instability can be present even when atlantodental interval is within the

range of normalcy. We re-classified atlantoaxial dislocation and basilar invagination on the basis of facetal alignment into three types.[14,15] Type A atlantoaxial facetal dislocation was when the facet of atlas was dislocated anterior to the facet of axis. Such a form of dislocation results in superior and posterior migration of the odontoid process and results in atlantoaxial dislocation and basilar invagination. There is an increase in atlantodental interval, and the odontoid process migrates posteriorly into the spinal canal. The process results in compression or compromise of the neural structures. Due to the early effects on the neural structures and the resultant neural deficits the clinical manifestations are rather acute or subacute in nature. In the lateral profile, the atlantoaxial facetal dislocation in such cases simulates lumbosacral listhesis.[16] In the acute form such instability results in atlantoaxial dislocation and in the more subacute or chronic form, it results in atlantoaxial dislocation and basilar invagination.

Type B facetal dislocation is when the facet of atlas is dislocated posterior to the facet of axis. In such a form of instability, the odontoid process does not migrate posteriorly into the spinal canal, and the atlantodental interval is not significantly increased or altered. Type C facetal dislocation is when there is no radiological misalignment of the facets and the instability is identified only on the basis of clinical understanding and intraoperative observations. The instability in such cases was labeled as central or axial. As the neural compromise is not significant or early in both Types B and Type C dislocation, the clinical manifestations are rather chronic in nature.[14,15]

In acute and chronic or longstanding atlantoaxial instability the natural defenses of the body seem to play a defining role to protect the neural structures from damage by the bones. In an acute state, there is a spasm of the neck muscles that restrict the movements. The muscles of the nape of the neck are under the spasm and restrict neck flexion movements that can be disastrous for neural structures. In the more chronic states that span over several years, the restriction of the neck movements become more fixed. In such cases, hyperextension of the neck is more easily possible, and flexion of the neck is restricted. Over the period, such chronic neck spasm results in shortening of the neck and wherever the potential or manifest compression is relatively unilateral it results in torticollis. Shortening of the neck is affected by reduction in the disc spaces rather than the reduction in height of the vertebral bodies. The reduction in the disc spaces frequently results in secondary osteophyte formation and radiological features of "spondylosis." Bone fusions and "Klippel Feil abnormalities" can be a result of such neck shortening. The bone fusions are more frequent at the craniovertebral junction and assimilation of the atlas and C2-3 bone fusions are relatively common forms of bone fusion. The frequently observed alteration in the cranial bones is platybasia and an overall reduction in the posterior cranial fossa volume.[8,17]

Chiari malformation and herniation of the cerebellar tonsils into the foramen magnum appears to the natural protective phenomenon in the face of atlantoaxial instability.[18] Chiari malformation appears to be Nature's air-bag phenomenon and the tonsils migrate into the spinal canal and are designed and positioned to prevent pinching of critical neural structures between bones. Syringomyelia is formed in an effort to neutralize cranial and spinal pressure and to support the bulk of Chiari malformation.[19] The very fact that there is a frequently observed atrophy of the cerebellar substance that is more prominent of the superior vermis suggests that an increased bulk of cerebellum in a smaller posterior cranial fossa may not be the cause of cerebellar herniation.[18,20] On the other hand, herniation of the tonsils in the spinal canal is a supremely designed and magically executed 'divine' phenomenon, an act of the Nature that is all protective and in no way pathological.

The very fact that atlantoaxial stabilization results in a reversal of most secondary alterations validates the hypothesis.[17] Increase in the neck size and torticollis and reversal of several other musculoskeletal changes including a reduction in the size of syrinx and reverse migration of the cerebellar tonsils occurs in the immediate or early postoperative phase.[18] We speculated that with the increase in length of the neck that is affected by an increase in the disc space height, there can be regression of the osteophytes, and there can even be potential reversal of bone fusions.[18] The return to normal of a number of so-called pathological entities by the atlantoaxial fixation and without direct manipulation suggests that the theory of embryological dysgenesis needs to be re-assessed.

Improvement of clinical symptoms following posterior fossa foramen magnum decompression simulates deflating of air-bag following a disaster.[18] However, such an operation has been frequently associated with recurrence of symptoms after a period. Presence of Chiari malformation can focus the attention of the clinician toward atlantoaxial instability. Treatment of the instability is the solution, and direct manipulation of tonsils or bone and dural decompression may be counter-productive.[18]

REFERENCES

1. Cleland. Contribution to the Study of Spina Bifida, encephalocele, and anencephalus. J Anat Physiol 1883;17:257-92.
2. Chiari H. UeberVerderbungen des Kleinhirns infolge von Hydrocephalie des Grossihirns. Dwochenschr 1891;17:1172-5.
3. Milhorat TH, Chou MW, Trinidad EM, Kula RW, Mandell M, Wolpert C, *et al.* Chiari I malformation redefined: Clinical and radiographic findings for 364 symptomatic patients. Neurosurgery 1999;44:1005-17.
4. Milhorat TH, Nishikawa M, Kula RW, Dlugacz YD. Mechanisms of cerebellar tonsil herniation in patients with Chiari malformations as guide to clinical management. Acta Neurochir (Wien) 2010;152:1117-27.
5. Smith BW, Strahle J, Bapuraj JR, Muraszko KM, Garton HJ, Maher CO. Distribution of cerebellar tonsil position: Implications for understanding Chiari malformation. J Neurosurg 2013;119:812-9.
6. Goel A, Achawal S. The surgical treatment of Chiari malformation association with atlantoaxial dislocation. Br J Neurosurg 1995;9:67-72.
7. Goel A, Cacciola F, Muzumdar DP. Trigeminal neurinoma associated with Chiari malformation and syringomyelia. J Clin Neurosci 2005;12:608-10.
8. Goel A, Bhatjiwale M, Desai K. Basilar invagination: A study based on 190 surgically treated patients. J Neurosurg 1998;88:962-8.
9. Goel A. Instability and basilar invagination. J Craniovertebr Junction Spine 2012;3:1-2.

10. Goel A. Treatment of basilar invagination by atlantoaxial joint distraction and direct lateral mass fixation. J Neurosurg Spine 2004;1:281-6.
11. Goel A, Desai KI, Muzumdar DP. Atlantoaxial fixation using plate and screw method: A report of 160 treated patients. Neurosurgery 2002;51:1351-6.
12. Goel A, Laheri VK. Plate and screw fixation for atlanto-axial dislocation. (Technical report). Acta Neurochir (Wien) 1994;129:47-53.
13. Goel A, Kulkarni AG, Sharma P. Reduction of fixed atlantoaxial dislocation in 24 cases: Technical note. J Neurosurg Spine 2005;2:505-9.
14. Goel A. Goel's classification of atlantoaxial "facetal" dislocation. J Craniovertebr Junction Spine 2014;5:3-8.
15. Goel A. Facetal alignment: Basis of an alternative Goel's classification of basilar invagination. J Craniovertebr Junction Spine 2014;5:59-64.
16. Kothari M, Goel A. Transatlantic odonto-occipital listhesis: The so-called basilar invagination. Neurol India 2007;55:6-7.
17. Goel A, Shah A. Reversal of longstanding musculoskeletal changes in basilar invagination after surgical decompression and stabilization. J Neurosurg Spine 2009;10:220-7.
18. Goel A. Chiari malformation — Is atlantoaxial instability the cause? Outcome analysis of 65 patients with Chiari malformation treated by atlantoaxial fixation. J Neurosurg Spine [In press].
19. Goel A. Is syringomyelia pathology or a natural protective phenomenon? J Postgrad Med 2001;47:87-8.
20. Goel A, Desai K, Bhatjiwale M, Muzumdar DP. Basilar invagination and Chiari malformation associated with cerebellar atrophy: Report of two treated cases. J Clin Neurosci 2002;9:194-6.

How to cite this article: Goel A. Is Chiari malformation nature's protective "air-bag"? Is its presence diagnostic of atlantoaxial instability?. J Craniovert Jun Spine 2014;5:107-9.