

## Suboccipital pain in Chiari formation: Its cause and significance

Pain in the suboccipital region, shoulders, and hands is a common presenting symptom related to Chiari formation. The pain characteristically exacerbates on coughing and sneezing and even laughing. In the series of 364 patients with Chiari formation, Milhorat *et al.* identified that 281 patients (81%) had suboccipital headache.<sup>[1]</sup> The pain was described as a heavy, crushing, or pressure-like sensation at the back of the head that radiated to the vertex and behind the eyes and inferiorly to the neck and shoulders. They described the headache had a pounding quality when it was severe, but it was nonthrobbing. The main feature of the headache that they described and that is generally observed with Chiari formation is the tendency for the headache to be accentuated by physical activity, Valsalva maneuvers, movements of the head, and sudden changes in posture. Various other studies on Chiari formation have identified headache occurring in 30%–90% of patients.<sup>[2-5]</sup> It is frequently difficult to differentiate migraine or stress/tension headaches from that related to Chiari formation. On the basis of severity of headache, the degree of tonsillar herniation, and presence (or absence) of syringomyelia, a Chiari Severity Index has been suggested.<sup>[6,7]</sup>

The basis of pathogenesis of Chiari formation and of the symptom of headache has been generally agreed to be relative increase of volume of the cerebellum and reduced volume of the posterior cranial fossa that houses it. Williams suggested that headaches that are exacerbated by cough are a result of craniospinal pressure dissociation.<sup>[7-9]</sup> On monitoring, pressure was identified to be higher in the ventricles than in the intrathecal space. Various morphological alterations that occur eventually lead to obstruction to the normal flow of cerebrospinal fluid (CSF) in the craniocervical junction. The herniating tonsils have been identified to have “valve-like” effect on the flow of CSF. Some authors have identified a role


of irritation of high cervical nerves as a cause of pain. Chronic irritation of basal dura in the vicinity of foramen magnum by herniating tonsils is considered to be an important factor that initiates and sustains the symptom of pain. The role of surgery is generally identified to decongest the craniovertebral junction, release the region of compression by herniating tonsils, and restore the flow of CSF.

The word tonsillar “herniation” has frequently been correlated to uncal “herniation.” The two otherwise diverse clinical situations evoke a sense of urgency of treatment considering the potential devastating neurological sequel related to uncal herniation. While uncal herniation is generally an acute clinical event, tonsillar herniation (Chiari formation) is characteristically a chronic formation. Some clinicians have identified symptom of headache related to Chiari formation an indication of the need of emergency treatment. In general, the presence of headache is related to other clinical symptoms and neurological and radiological findings to identify the need for and timing of surgical treatment.

In 2013, we identified atlantoaxial instability as the nodal point of pathogenesis of Chiari formation. Furthermore, it was speculated that Chiari formation or tonsillar herniation is not a pathological event but a natural protective introduction into the foramen magnum aimed to prevent compression of critical neural structures between bones in the event of potential or manifest atlantoaxial dislocation.<sup>[10,11]</sup> It was observed that there is atlantoaxial instability despite the fact that conventional radiological measures do not show any evidence of dislocation. There may not be any abnormal alteration in atlantodental interval or in facet alignment on static and dynamic imaging and there may not be any compression of dural or neural structures at the craniovertebral junction. Over the years of our experience, we now identify that the presence of Chiari formation is by itself a confirmatory evidence of

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

**How to cite this article:** Goel A. Suboccipital pain in Chiari formation: Its cause and significance. *J Craniovert Jun Spine* 2018;9:221-2.

Access this article online	
<b>Website:</b> <a href="http://www.jcvjs.com">www.jcvjs.com</a>	<b>Quick Response Code</b> 
<b>DOI:</b> <a href="https://doi.org/10.4103/jcvjs.JCVJS_117_18">10.4103/jcvjs.JCVJS_117_18</a>	

atlantoaxial instability. Direct observation of the atlantoaxial articulation and manual handling of bones of the region clarifies the issue of instability. We have labeled such atlantoaxial instability as “central” or axial’ atlantoaxial instability.<sup>[12]</sup> Our “highly” satisfactory clinical results following atlantoaxial stabilization aimed at arthrodesis for cases with Chiari formation with or without the presence of bone abnormalities at the craniovertebral junction is a confirmatory proof for such hypothesis. On the basis of this observation, the name Chiari “formation” is preferred over Chiari “malformation.”<sup>[13]</sup>

Chiari formation and syringomyelia are neural outcome of the presence of longstanding or chronic atlantoaxial instability. Basilar invagination, platybasia, Klippel–Feil abnormality, short neck, torticollis, and host of other neural and musculoskeletal alterations that are frequently associated with Chiari formation and syringomyelia are also an outcome of chronic atlantoaxial instability.<sup>[14,15]</sup> The potential of reversibility of all neural and musculoskeletal abnormalities following atlantoaxial fixation gives credence to such hypothesis.

We observe that the presence of suboccipital headache is an indication of atlantoaxial instability. The presence of unstable craniovertebral junction is indicated by the exacerbation of symptom on neck movements and on exercise. We observe that acts of coughing and sneezing involve sudden flexion of the neck, a movement that has the potential of critically jeopardizing the anatomical situation at the craniovertebral junction. The acute pain that is triggered seems to be a unique natural event that is an attempt to restrict acute or excessive neck movement. The spasm in the muscles of the nape of the neck and the pain in the region and in the hands seems to be a confirmatory evidence of the presence of atlantoaxial instability.

Our remarkable success in treating Chiari formation with atlantoaxial fixation and resolution of neurological symptoms and headache provides confirmatory evidence that such Chiari formation-related symptoms are a result of atlantoaxial instability. The symptoms can be present even in the absence of any major neural compression or distortion or even any direct radiological evidence of instability. Our earlier studies have discussed that it is not neural compression or deformation; it is subtle, potential,

or manifest instability-related microtrauma to the neural structures that is the cause of symptoms. It is now clear to us that there can be instability of the atlantoaxial joint without any radiological evidence. Understanding of this fact has the potential of revolutionizing the treatment of the clinical entity of Chiari formation.

#### ATUL GOEL

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

**Address for correspondence:** Prof. Atul Goel, Department of Neurosurgery, Seth G.S. Medical College and K.E.M Hospital, Parel, Mumbai - 400 012, Maharashtra, India. E-mail: atulgoel62@hotmail.com

#### REFERENCES

- 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.
- Grangeon L, Puy L, Gilard V, Hebant B, Langlois O, Derrey S, *et al.* Predictive factors of headache resolution after chiari type I malformation surgery. *World Neurosurg* 2018;110:e60-6.
- Elster AD, Chen MY. Chiari I malformations: Clinical and radiologic reappraisal. *Radiology* 1992;183:347-53.
- Nohria V, Oakes WJ. Chiari headaches. *Neurology* 1993;43:1272.
- Greenberg JK, Yarbrough CK, Radmanesh A, Godzik J, Yu M, Jeffe DB, *et al.* The chiari severity index: A preoperative grading system for chiari malformation type I. *Neurosurgery* 2015;76:279-85.
- Pisapia JM, Merkow MB, Brewington D, Henn RE, Sutton LN, Storm PB, *et al.* External validity of the chiari severity index and outcomes among pediatric chiari I patients treated with intra- or extra-dural decompression. *Childs Nerv Syst* 2017;33:313-20.
- Sansur CA, Heiss JD, DeVroom HL, Eskioglu E, Ennis R, Oldfield EH, *et al.* Pathophysiology of headache associated with cough in patients with chiari I malformation. *J Neurosurg* 2003;98:453-8.
- Williams B. Cerebrospinal fluid pressure changes in response to coughing. *Brain* 1976;99:331-46.
- Williams B. Cough headache due to craniospinal pressure dissociation. *Arch Neurol* 1980;37:226-30.
- Goel A. Is atlantoaxial instability the cause of chiari malformation? Outcome analysis of 65 patients treated by atlantoaxial fixation. *J Neurosurg Spine* 2015;22:116-27.
- Goel A. Is chiari malformation nature’s protective “air-bag”? Is its presence diagnostic of atlantoaxial instability? *J Craniovertebr Junction Spine* 2014;5:107-9.
- Goel A. Goel’s classification of atlantoaxial “facetal” dislocation. *J Craniovertebr Junction Spine* 2014;5:3-8.
- Goel A. Is chiari a “formation” or a “malformation?” *J Craniovertebr Junction Spine* 2017;8:1-2.
- Goel A, Shah A. Reversal of longstanding musculoskeletal changes in basilar invagination after surgical decompression and stabilization. *J Neurosurg Spine* 2009;10:220-7.
- Goel A. Basilar invagination, syringomyelia and chiari formation and their relationship with atlantoaxial instability. *Neurol India* 2018;66:940-2.