



CHIARI 1.5 MALFORMATION: A RARE CASE REPORT

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ABSTRACT A Chiari malformation is a structural abnormality at the back of the brain and skull. The Chiari 1.5 malformation is a rare entity and its true incidence is not known. Chiari 1.5 is defined as a tonsillar herniation within a Chiari I malformation with additional caudal descent of the brainstem through the foramen magnum. MRI is the best method for the diagnosis with sagittal T1 WI to assess tonsillar herniation. We represent a case of 31 year old female which was diagnosed with Chiari 1.5 malformation on MRI after she had minor head injury. Posterior fossa decompression surgery is done for relieving symptoms. Sometimes repeated surgeries are required.

KEYWORDS : Tonsillar herniation, Odontoid retroflexion, Chiari malformation, Basilar invagination.

CASE REPORT

A 31-year-old female came to neuro OPD with chief complaints of dizziness since 2 months. She had history of head injury. She did not have any bruise at the time of injury but she had episodes of dizziness several times a day since injury. No signs of raised intracranial tension, meningitis, or cranial nerve involvement were seen.

Magnetic resonance imaging (MRI) revealed herniation of the tonsils below the foramen magnum along with the brain stem herniation causing flattening of the medulla anteroposteriorly. This descent was measured in relation to the Mc Rae line drawn from the basion to the opisthion, that is, anterior margin to the posterior margin of foramen magnum. The AP diameter of the medulla oblongata was 8 mm, resulting in a flattened and elongated appearance in the midsagittal section.

Obex was seen to lie 23 mm below the foramen magnum. There was dorsal bump at the cervicomedullary junction (Figure 1).

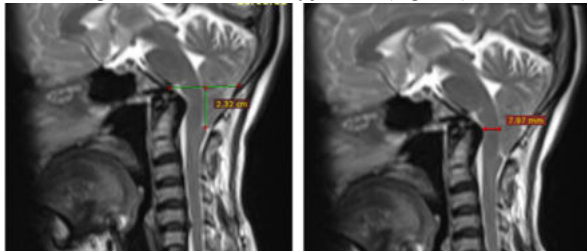


Figure 1: T2W sagittal image showing 23 mm of cerebellar tonsils herniation below the Mc Rae line and thinning of medulla oblongata with mid-sagittal antero-posterior diameter of ~8mm.

There was crowding at the foramen magnum with obliteration of subarachnoid space. Clivus-canal angle is 117° in our case which should be usually more than 150° (Figure 2).

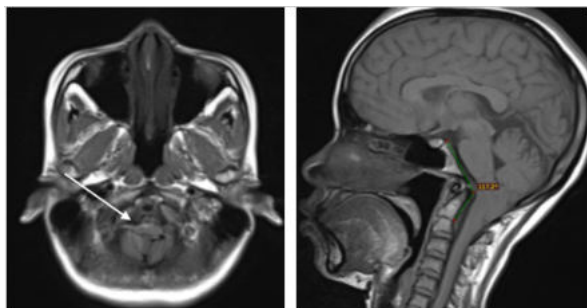


Figure 3: a) T1W Axial image showing crowding of foramen magnum b) T1W Sagittal image showing clivus-canal angle which measures 117° consistent with platybasia.

To evaluate the angulation of the odontoid process, we measured the

angle of retroflexion by drawing one horizontal line through a midpoint of the synchondrosis between the base and apex of the odontoid process and a second line through the midpoint of the synchondrosis and apex of the odontoid process on a midsagittal MR imaging and angle of retroversion, formed between the line drawn from the base of C-2 and its intersection with a line drawn from the odontoid tip. Angle of retroflexion of the odontoid process was 55.4° and angle of retroversion of the odontoid process was 58.3°. The odontoid process is retroflexed and superior tip of odontoid exceed 9 mm beyond Chamberlain line; which is typically considered abnormal and denotes basilar invagination. (Figure 3).

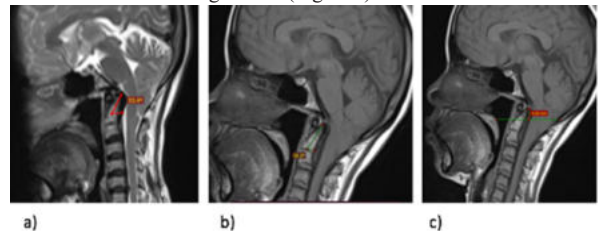


Figure 4: a) T2W sagittal image showing angle of retroflexion (55.4°) b) T1W sagittal image showing angle of retroversion (58.3°) c) T1W sagittal image showing tip of odontoid exceed 9 mm beyond Chamberlain line consistent with basilar invagination.

DISCUSSION

Chiari malformations are a group of defects associated with congenital caudal 'displacement' of the cerebellum and brainstem.

Traditionally the Chiari malformations include four separate anatomical entities, all of which involve the hindbrain. [1] Type I is defined as inferior displacement of the cerebellar tonsils through the foramen magnum into the cervical canal. A displacement of more than 5 mm below the foramen magnum after the age of 15 years is considered pathologic. Pointed configuration of cerebellar tonsils is typical.

Type II which is also known as the Arnold-Chiari malformation involves displacement of brainstem and lower cerebellum into the cervical spinal canal. The fourth ventricle is caudally displaced and extends below the foramen magnum. It is nearly always associated with lumbar myelomeningocele. Supratentorial anomalies are commonly seen. These include falx hypoplasia, hydrocephalus, callosal hypogenesis, fused enlarged massa intermedia, colpocephaly, abnormal gyral pattern, and Luckenschadel skull.

Chiari III malformation is characterized by herniation of posterior fossa contents in an occipital or high cervical encephalocele and other features of Chiari II malformation.

Type IV is described as cerebellar hypoplasia or aplasia.

It has been realized over the years that findings in some patients do not

exactly fit into these morphological entities. Recently, entities such as Chiari 0 and 1.5 have been described. [2] The essential difference between Chiari I and 1.5 is the presence of caudal descent of the brainstem in the latter in addition to tonsillar ectopia. [3] There is substantial clinical overlap between the two entities. However, younger age at presentation and more severe symptoms like bulbar signs are more common in Chiari 1.5. [4] The differentiation between the two entities is important for appropriate management as Chiari 1.5 patients are more likely to require extensive and complex surgeries as adjunct to decompression.

The imaging incidence of asymptomatic Chiari I malformation was determined to be 0.1-1%. Chiari 1.5 malformation is thought to be less common than the Chiari I malformation, although the exact incidence of Chiari 1.5 malformation is still unknown. [5]

Essential neuroimaging feature of Chiari 1.5 is the descent of obex and cerebellar tonsils below the foramen magnum. Syringohydromyelia is often present and tends to be persistent after posterior fossa decompression. Bone abnormalities frequently seen are basilar invagination, atlantooccipital fusion, scoliosis, retroflexed odontoid, abnormal clivus-canal angle. [6]

Established craniovertebral junction measurements state that a clivus-canal angle $< 150^\circ$ is abnormal regard less of patient positioning, and at angles $< 150^\circ$ there is a known increased risk of ventral cervical spinal cord compression. In our case the angle was decreased and measuring 117° . A decrease in the clivus-canal angle is a described feature of basilar invagination, a developmental finding associated with basi-occiput hypoplasia. It suggests a trend toward a shortened basi-occiput, which in conjunction with increased periodontoid tissue, may predispose this group to an increased risk of ventral cervical spinal cord compression in the absence of increased odontoid inclination. Chiari malformation patients have smaller odontoid retroflexion and retroversion angle, 55.4° and 58.3° respectively in our case. In recent studies normal angle of retroflexion in women was $78^\circ \pm 4.8^\circ$ and normal angle of retroversion was $70^\circ \pm 4^\circ$. [7]

The patient undergone decompressive posterior fossa craniotomy to relieve obstructive symptoms.

CONCLUSION

In conclusion, this article is an attempt to present a relatively lesser known entity in Chiari spectrum which is important to recognize because of differences in management and prognosis.

REFERENCES

1. Caldwell DL, Dubose CO, White TB. Chiari malformations. *Radiol Technol.* 2009; 80:340–54
2. Tubbs RS, Elton S, Grabb P, Dockery SE, Bartolucci AA, Oakes WJ. Analysis of the posterior fossa in children with the Chiari 0 malformation. *Neurosurgery.* 2001; 48:1050–5. [PubMed] [Google Scholar]
3. Tubbs RS, Iskandar BJ, Bartolucci AA, Oakes WJ. A critical analysis of the Chiari 1.5 malformation. *J Neurosurg.* 2004; 101:179–83.
4. Moore HE, Moore KR. Magnetic resonance imaging features of complex Chiari malformation variant of Chiari I malformation. *Pediatr Radiol.* 2014; 44:1403–11.
5. Chiari 1.5 Malformation : An Advanced Form of Chiari I Malformation; In-Kyeong Kim, Kyu-Chang Wang, In-One Kim, Byung-Kyu Cho *J Korean Neurosurg Soc.* 2010 Oct; 48(4): 375–379.
6. Brockmeyer DL. The complex Chiari: Issues and management strategies. *Neurol Sci.* 2011; 32: S345–7.
7. Besachio DA, Khaleel Z, Shah LM. Odontoid process inclination in normal adults and in an adult population with Chiari malformation Type I. *J Neurosurg Spine.* 2015; 23(6):701–706.