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Understanding Chiari 1.5 Malformation: A comprehensive overview

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Abstract

Chiari 1.5 malformation, a variant of Chiari malformation, presents unique radiological features and clinical implications. This review aims to summarize the radiological characteristics of Chiari 1.5 malformation. Magnetic resonance imaging (MRI) is the gold standard for diagnosing Chiari 1.5 malformation. Key radiological findings include herniation of the cerebellar tonsils below the foramen magnum, accompanied by herniation of the brainstem and fourth ventricle. The extent of tonsillar herniation, as measured by the distance from the basion to the tip of the herniated tonsils, is a crucial parameter for diagnosis and surgical planning. In addition to tonsillar herniation, Chiari 1.5 malformation often presents with associated abnormalities such as syringomyelia, hydrocephalus, and craniocervical junction anomalies. Syringomyelia, characterized by the formation of fluid-filled cavities within the spinal cord, is a common finding and may contribute to neurological symptoms such as pain, weakness, and sensory deficits.

Case Report

Our patient came with chief complaints of posterior cervical pain. On MRI imaging following findings were made note of: Herniation of cerebellar tonsils seen below the foramen magnum by ~7 mm with peg like appearance of the tonsils. Obex of the 4th ventricle is also seen extending just below the foramen magna. Brainstem appears stretched with distal medulla seen extending below the foramen magna. Dorsal medullary bump is present. Atlanto-occipital fusion seen along the fusion of the inferior clivus and the anterior arch of atlas. Odontoid process appears posteriorly oriented with mild basilar invagination and crowding at the level of the foramen magnum and compression of brain stem and cerebellar tonsils. Tip of the odontoid above the chamberlain line by ~ 9.0mm. No atlanto-axial dislocation. No platybasia (basal angle 132 degrees). Clivus angle is abnormal and measures 106-110 degrees.

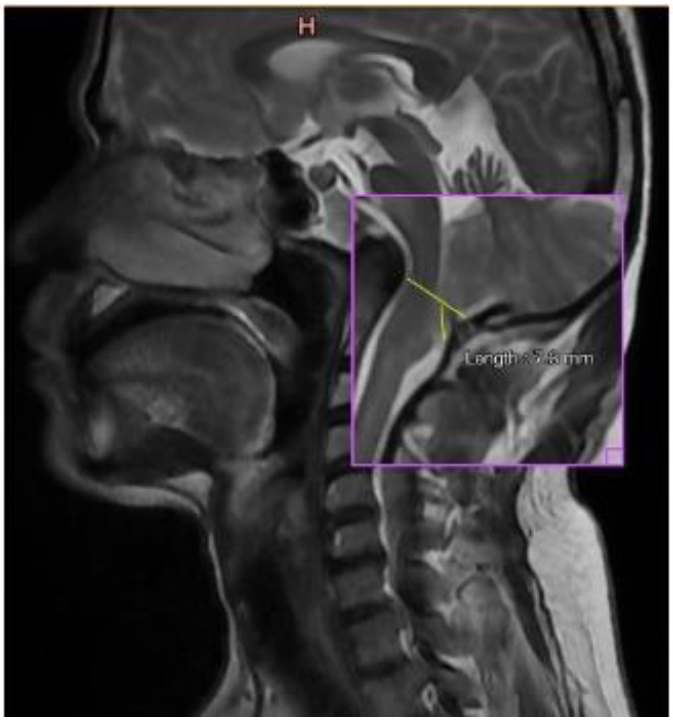

Figure 1: MRI T2 Sagittal image shows herniation of cerebellar tonsils below the foramen magnum by ~7 mm with peg like appearance of the tonsils.

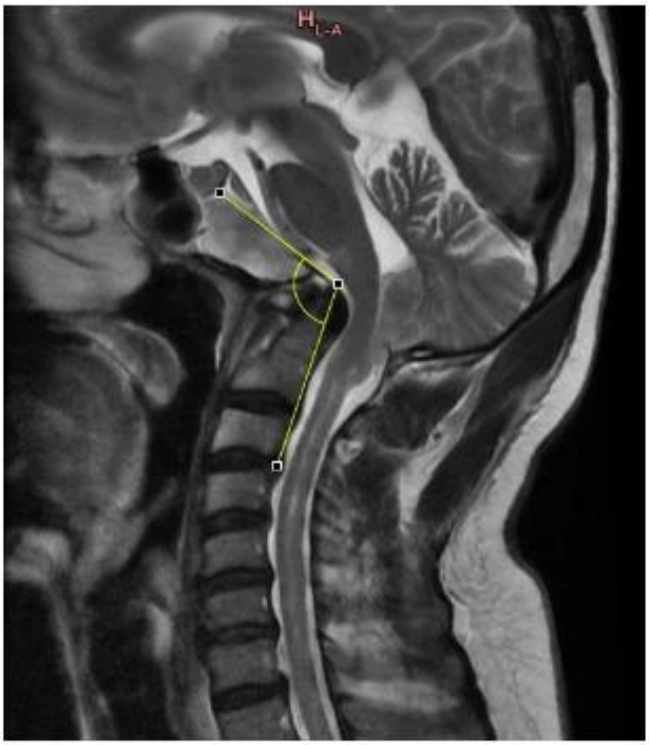

Figure 2: MRI T2 Sagittal image showing abnormal clivus angle (106 - 110 degrees)

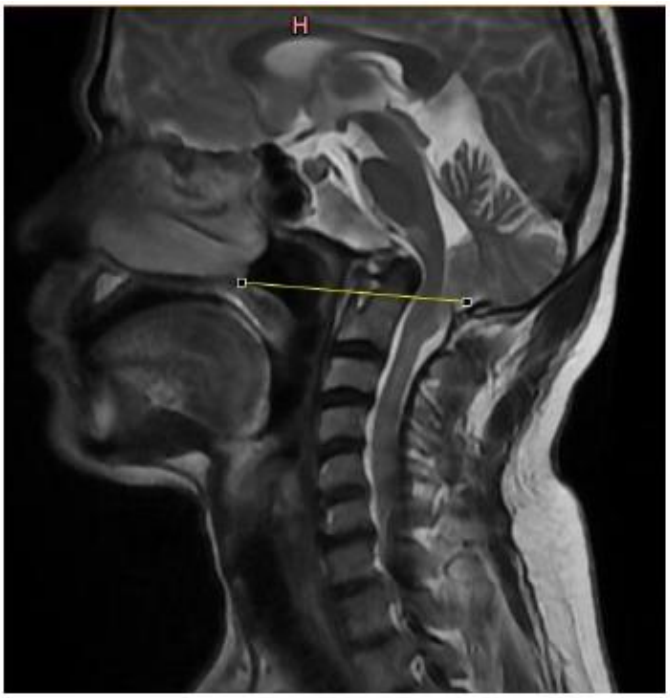

Figure 3: MRI T2 sagittal image shows obex of the 4th ventricle extending below the foramen magnum

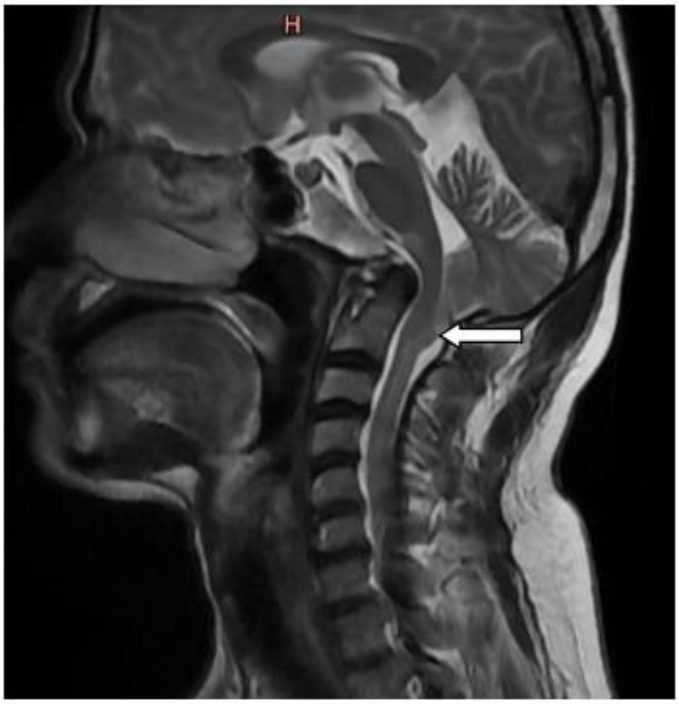

Figure 4: MRI T2 sagittal image shows that the brainstem is stretched. Dorsal medullary bump is present.

Discussion

Chiari malformations encompass four distinct anatomical variations, all involving the hindbrain.

Type 1 - Characterized by the downward displacement of cerebellar tonsils.

Type 2 - Invariably associated with myelodysplasia and entails the herniation of the medulla and vermis. Involvement above the tentorium cerebelli is commonly observed.

Type 3 - Typically defined by the herniation of posterior fossa contents through a low occipito-cervical bony defect.

Type 0 - Presents with syrinx formation without evident herniation of tonsils.

Type 1.5 - Distinguished by the caudal protrusion of the brainstem in addition to tonsillar herniation.

Type 4 - Defined as cerebellar hypoplasia or aplasia.

Type 5 - Represents Chiari 2 malformation alongside occipital or high cervical myelomeningocele.

Conclusion

A key neuro-imaging feature of Chiari 1.5 is the descent of the obex and cerebellar tonsils below the foramen magnum, often accompanied by persistent syringohydromyelia following posterior fossa decompression. Bone abnormalities commonly observed include basilar invagination, Atlanto-occipital fusion, and abnormal spinal curvature. An abnormal clivus-canal angle, indicating ventral brain compression, has been identified as a significant measurement for assessing the need for more complex surgical procedures in Chiari 1.5patients with a clivus-canal angle of <125°, particularly when combined with basilar invagination. The primary distinction between Chiari I and 1.5 is the presence of caudal descent of the brainstem in the latter, alongside tonsillar ectopia. While there is considerable clinical overlap between the two, Chiari 1.5 tends to manifest at a younger age and presents with more severe symptoms, including bulbar signs. Differentiating between these entities is crucial for appropriate management, as Chiari 1.5 patients often require more extensive and complex surgical interventions in addition to decompression.

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