Chiari 1.5: A lesser known entity

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Case description

A 14-year-old child presented with intermittent neck pain and gradually progressive weakness in right upper limb for 4 years. The weakness had started with right hand and gradually progressed to involve the forearm and arm. This was followed by lower limb weakness on the same side for about 6 months. There was no history of trauma, fever, or lumbar puncture. No signs of raised intracranial tension, meningitis, or cranial nerve involvement were seen.

Magnetic resonance imaging (MRI) revealed herniation of the tonsils 19.5 mm 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. Obex was seen to lie 12mm below the foramen magnum [Figure 1]. There was dorsal bump at the cervicomedullary junction [Figure 2]. There was crowding at the foramen magnum with obliteration of subarachnoid space [Figure 3]. Syrinx was present in the spinal cord from C2 to D2 level. Osseous abnormalities observed were retroflexion of the odontoid process and scoliosis of the cervicodorsal spine [Figure 4].

Discussion

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.

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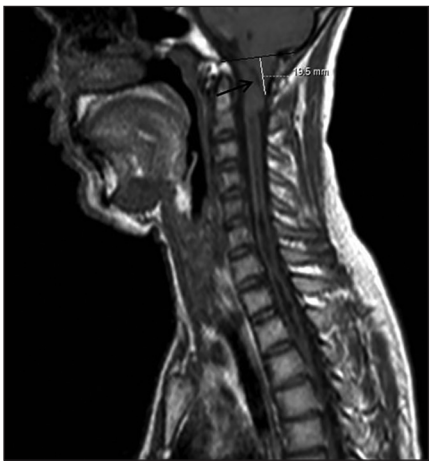

Figure 1: Midsagittal T1W MR image of the craniovertebral junction and cervicodorsal spine shows descent of obex (black arrow) and cerebellar tonsils inferior to Mc Rae line (black line). White line indicates the measurement of descent of cerebellar tonsils. T1W = T1 weighted, MR = magnetic resonance

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 myelomenigocele. 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

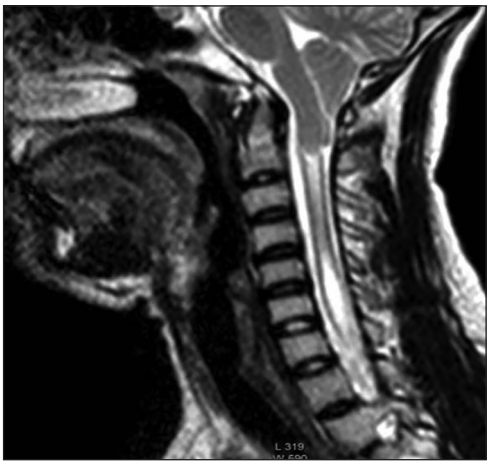

Figure 2: Midsagittal T2W MR image demonstrates dorsal bump at cervicomedullary junction with syringohydromyelia. T2W = T2 weighted

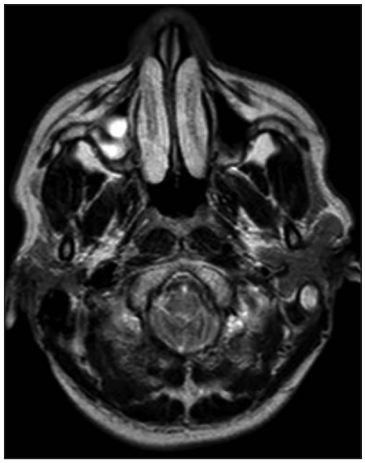

Figure 3: Axial T2W MR image shows crowding at foramen magnum due to descent of cerebellar tonsils

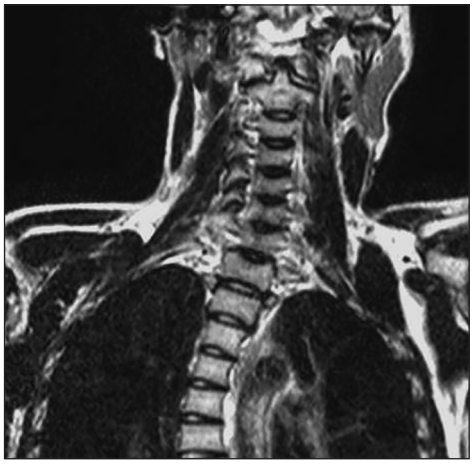

Figure 4: Coronal T2W MR image shows scoliosis of cervicodorsal spine convex towards right side

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.

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.[5] Abnormal clivus-canal angle is a measurement of ventral brain compression. It is formed by intersection of a line along the clivus with a line constructed along the posterior surface of axis body.[6] It has been found that Chiari 1.5 patients with clivus-canal angle of <125° together with basilar invagination require more complex surgery in addition to the standard decompression.[7]

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.

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Conflicts of interest
None declared.

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