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# A Rare Case of Chiari 1.5 Malformation with Frontal Encephalocele

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***Abstract:*** *Chiari 1.5 malformation or bulbar variant of Chiari I malformation is a rare congenital anomaly. It is a progression of chiari 1 malformation characterized by caudal descent of cerebellar tonsillar along with brainstem through foramen magnum causing syringomyelia and hydrocephalus. This is due to the discrepancy between the size and content of the posterior fossa which is called as crowded posterior fossa. Although chiari malformation with occipital encephalocele is common, we encountered a case of chiari 1.5 malformation with frontal encephalocele with syringomyelia and obstructive hydrocephalus.*

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## I. Introduction

Chiari 1.5 is a congenital condition characterised by anatomical defect in the base of the skull. It is associated with intracranial anomalies including intracranial cysts, complete/partial corpus callosal agenesis, inter-hemispheric lipomas, facial clefts. Today there are six recognized Chiari types, but type I is by far the most common. Chiari types I, II, and III involve varying degrees of herniation of rhombencephalic derivatives out of the posterior cranial fossa. Chiari 1.5 is the subject of our report and it may be noted that it is more of a clinical observation than an official type. Chiari IV malformation involves cerebellar hypoplasia or aplasia with no herniation of the hindbrain. Chiari Type 0 has recently been recognized as a form of Chiari where the herniation doesn't meet normal criteria but a syrinx is present and causes symptoms. Chiari I malformation has been defined as downward herniation of the cerebellar tonsils through the foramen magnum, whereas Chiari II malformation is associated with spina bifida and includes herniation of the vermis with corresponding descent of the rhombencephalon. Whereas Chiari III malformation is characterised by characterized by a small posterior fossa and a low occipital/high cervical encephalocele.

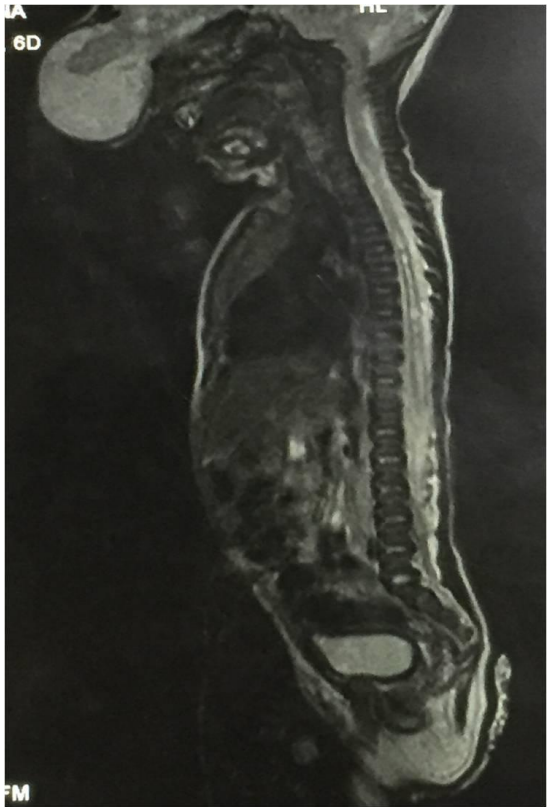
Chiari 1.5 malformation specifically refers to the tonsillar herniation seen in Chiari I malformation but with the addition of an elongated brainstem and fourth ventricle. Chiari 1.5 malformation is thought to be less common than the Chiari I malformation, although the exact incidence of Chiari 1.5 malformation is still unknown. Chiari malformation is usually associated with occipital encephalocele. But in the present case it is associated with frontal encephalocele.

## II. Material and Methods

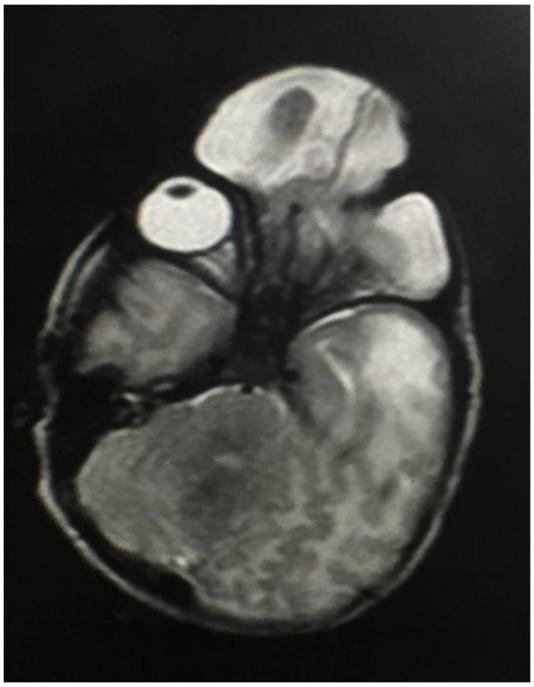
A six-day-old infant presented with two episodes of seizure and apneic spells was brought for further examination. Patient was afebrile. Images were obtained using 3 Tesla MRI. MRI sequences of T1W, T2W and T2Flair were studied and CSF flow study was done. CT axial and sagittal images were obtained to identify the defect in the skull and spinal column.

## III. Result

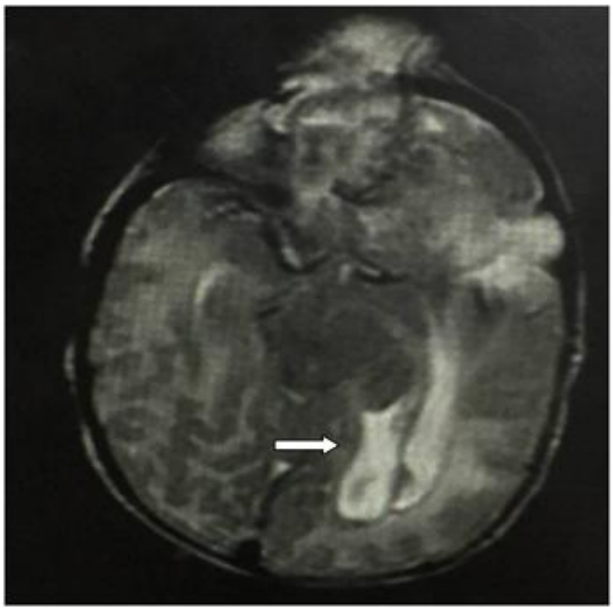
On CT Sagittal image frontal encephalocele with herniation of cerebellum and brain stem was observed. CT axial section showed crowding of posterior fossa with brainstem and cerebellar tonsil defect in the frontal skull bone with the herniation of meninges and brain tissue. Sagittal T2 weighted image revealed herniation of cerebellar tonsil and brain stem through the foramen magnum into the cervical spinal canal. Axial T2 image showed frontal encephalocele and obstructive hydrocephalus.



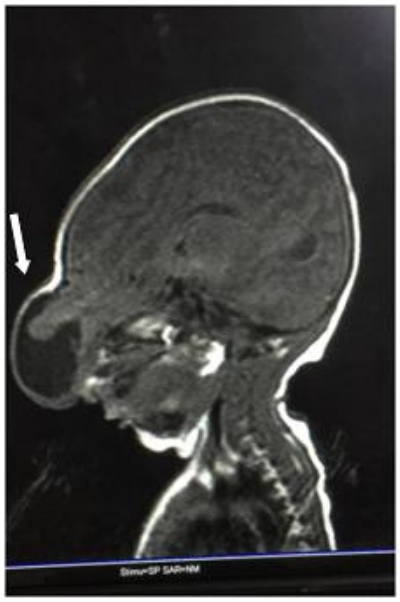

Sagittal T2 weighted image shows herniation of cerebellum and brain stem through the foramen magnum into the cervical spinal canal



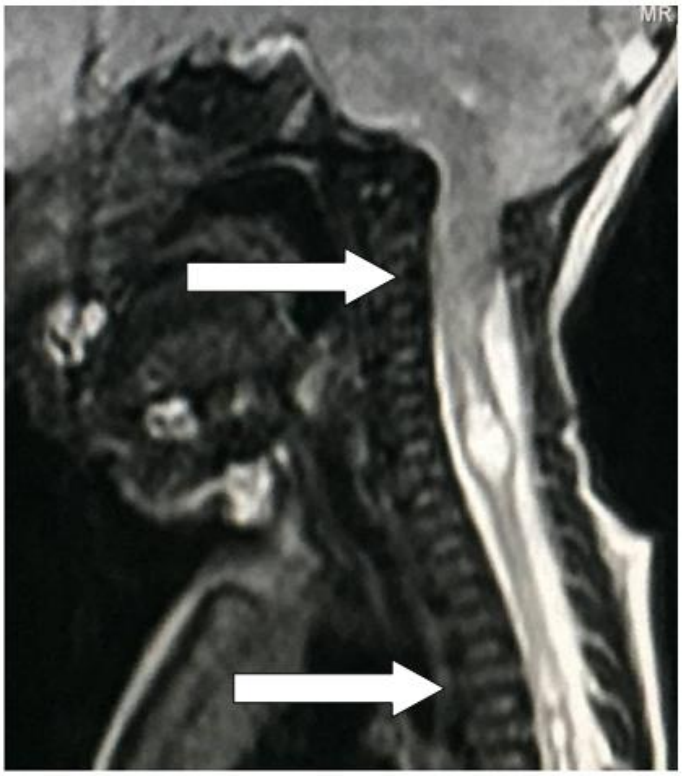
Axial T2 image shows frontal encephalocele



Axial T2 weighted image shows obstructive hydrocephalus



CT Sagittal image shows frontal encephalocele with herniation of cerebellum and brain stem




CT sagittal image shows herniation of cerebellum and brain stem into the cervical canal with syringomyelia

## IV. Discussion

The etiology of chiari 1.5 malformation is unknown. One of the theories proposed for pathogenesis of condition involves the presence of pressure difference between cranial and spinal subarachnoid space. Although the exact cause of Chiari is unknown, it is thought to be the result of abnormal fetal brain development during pregnancy. There is thought to be some genetic component to Chiari, since the defect can (rarely) be found in family members but no specific genetic cause has ever been identified. Some rare conditions that result in abnormal bone growth (craniosynostosis, skeletal dysplasia, achnodroplasia, etc.) can also present with similar symptoms and structural abnormalities.

Essential neuroimaging feature of chiari 1.5 is the descent of obex and cerebellar tonsil below the foramen magnum. Syringomyelia will often persist even after posterior decompression. Bone abnormalities frequently seen are basilar invagination, atlantooccipital fusion, scoliosis, retroflexed odontoid, abnormal clivus – canal angle. 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 standard decompression.

In our case we encountered a frontal encephalocele which is a rare finding (according to location - occipital encephalocele occurance observed in 75%, frontal in 15% and basal in10% of cases). Encephalocele occurs due to failure of surface ectoderm to separate from neuroectoderm, resulting in bone defect in cranial and facial bone, which allows herniation of meninges and brain tissue.

MRI is the imaging modality of choice, sagittal image is the best plain for assessing the presence of chiari 1.5 malformation. Axial images through the foramen magnum shows crowding of posterior fossa with cerebellar tonsil and brain stem.

In the case of advanced form of Chiari 1.5 malformation, sufficient posterior fossa decompression including tonsillectomy for normalizing CSF circulation around the foramen magnum is necessary. Sometimes repeated surgeries are required.

## V. Conclusion

Chiari 1.5 malformation with frontal encephalocele is an uncommon finding - 1 in 40,000 live births. The risk of a child inheriting a Chiari malformation from a parent is very small. Researchers are looking into which gene or genes may be responsible. It is possible that children born with a Type I or 1.5 Chiari malformation may have inherited a faulty gene or genes from a parent. Screening of the family members of a person with a Chiari malformation is not usually done. As many people with a Chiari malformation have no

symptoms and treatment is usually only required if symptoms are causing problems, screening is usually considered if family members have symptoms that suggest they may have a Chiari malformation and might also benefit from treatment. This article is an attempt to present a relatively lesser known entity in Chiari spectrum, analyze its conditions and manifestations and to make a strong case to the fraternity to broaden the scope of our research in this subject, which in our opinion is important due to the differences in its management and prognosis.

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