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An Unusual Case of Cerebellar Tonsillar Descent till C7 Vertebrae in a Case of Chiari Malformation with Meningomyelocele

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Abstract

Arnold–Chiari or Chiari malformations are a group of posterior fossa malformations that include the pons, medulla, and cerebellum. This group of disorders may consist of associated abnormalities like myelomeningocele and encephalocele. Chiari malformation has been classified majorly into four categories. Chiari I malformation has been defined as tonsillar descent more than 5 mm below the level of the foramen magnum. The maximum descent of tonsillar herniation reported is 27 mm below the foramen magnum around the C2 vertebrae. In this study, we report the case of a 4-month-old infant with swelling over the lumbosacral (LS) spine since birth with a gradually progressive increase in head size since the age of 2 months. The infant had occasional stridor with respiratory distress for 2 months. Magnetic resonance imaging (MRI) of the brain and spine revealed gross hydrocephalous with myelomeningocele at the L1–L5 level with associated Chiari malformation. The tonsillar descent was almost at the level of C7, which was unusual in his patient. The infant underwent a right ventriculoperitoneal shunt placement and myelomeningocele repair. It was decided that the malformation would be managed in the second sitting. The postoperative period was uneventful and the respiratory distress improved. Cases of tonsillar descent (Chiari I malformation) to the C7 vertebral level are very rare.

Keywords

- Chiari malformation
- myelomeningocele
- cerebellar tonsillar herniation

Introduction

Chiari malformations are a group of posterior fossa disorders with associated abnormalities of the pons, medulla, and cerebellum. This has been associated with spina bifida and especially myelomeningocele (MMC).[1,2] At the end of the 19th century, Chiari and Arnold described these complex hindbrain anomalies and their clinical and pathological significance.

In this modern era, Chiari malformation is diagnosed with an MRI of the brain and spine.[3] The condition is grossly divided into four types. Other types, like Chiari 0, Chiari 1.5, and Chiari 5 malformations, are also described in the literature. Chiari 1 malformation is described as the descent of the cerebellar tonsils more than 5 mm below the foramen magnum.[4] The maximum descent of the cerebellar tonsils described in the literature is 27 mm below the foramen magnum.[1] Chiari malformation in children presents with hydrocephalus and associated spinal dysraphism.

Chiari 1 malformation is the most common type of Chiari malformation and occurs in 0.5 to 3.5% of the general population with a slight female preponderance.[5]

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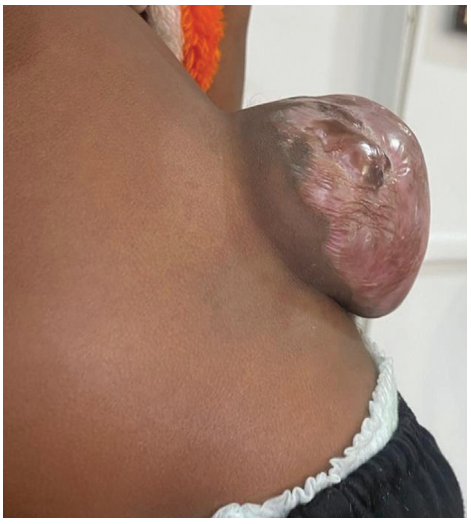

Fig. 1 Clinical image of the baby.

Case Summary

This is a case of a 4-month-old male term baby, the first child born out of a nonconsanguineous marriage. The mother had an uneventful pregnancy and the child was born by cesarean section. The intrauterine period of the infant was uneventful. The mother had taken prenatal iron and folic acid for 3 months in the second trimester. At birth, the baby was moving the upper limbs normally; however, the power in the lower limbs was weak. The baby had a swelling on the lower back of approximately 5×5 cm, which was fluctuant, irreducible, and nonpulsatile There was no cerebrospinal fluid (CSF) leak from the swelling. The infant gradually developed a progressive increase in the size of the head.

At presentation, the baby was active and alert, with occasional respiratory stridor and respiratory distress. The infant was having macrocephaly with frontal bossing (head circumference of 47 cm) and had a large 8×8 cm cystic, fluctuant, irreducible, nontransilluminant, nonpulsatile swelling in the lumbar region (►**Fig. 1**). On motor power examination, it was revealed that the baby had normal power in the bilateral upper limbs and reduced left lower limb (1/5 at the hip, 0/5 at the knee, and 0/5 at the ankle as per the Medical Research Council [MRC] grading) and right lower limb (3/5 in all the joints). MRI of the whole spine revealed a defect in the posterior elements of L1–L5 with herniation of the spinal cord into a large multiseptated CSF-filled sac of size $3.6 \times 8 \times 6.3$ cm containing neural elements (►**Fig. 2**). There was displacement of the cerebellar tonsils through the foramen magnum up to the level of C7 vertebrae alongside grossly dilated bilateral lateral ventricles (►**Fig. 3**).

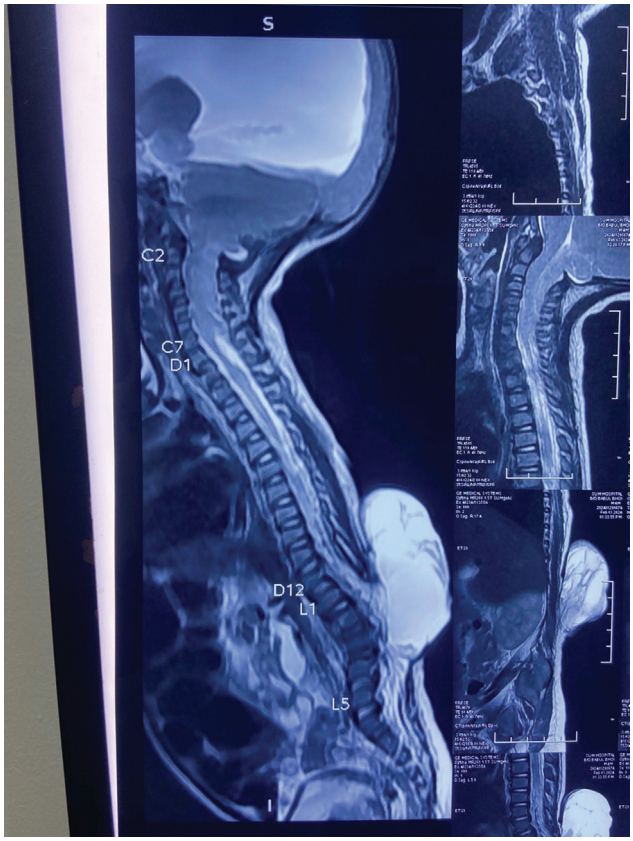

Fig. 2 Magnetic resonance imaging (MRI) of myelomeningocele.

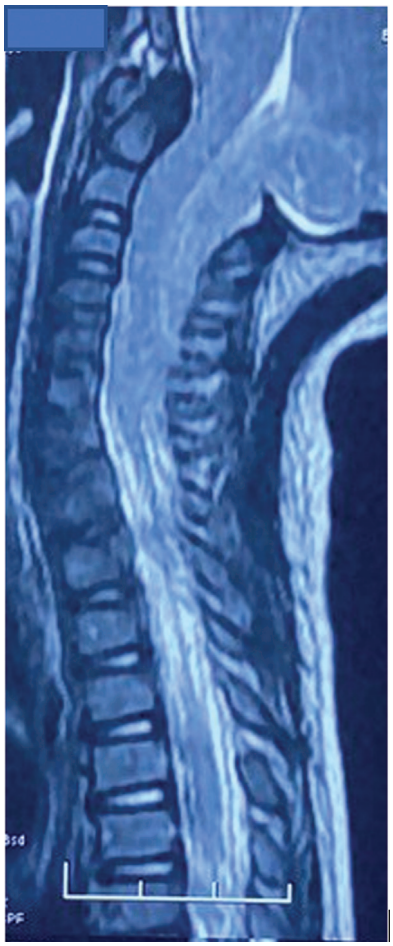

Fig. 3 Tonsillar descent till C7.

Further evaluation, including ultrasound of the abdomen and pelvis and 2D echocardiography, did not reveal any abnormalities. After adequate optimization, the patient underwent right-sided low-pressure ventriculoperitoneal shunt for hydrocephalus followed by meningomyelocele repair under general anesthesia. The post-op period was uneventful. The respiratory distress and stridor improved. The wound was healthy and the baby was discharged on postoperative day 5. It was decided that the malformation would be managed in the second sitting.

Discussion

There are many proposed theories like molecular, hydrodynamic, and mechanical, with different mechanisms that can result in Chiari malformation.[6] A small posterior fossa results in a small space for the cerebellum, which causes the descent of the tonsils through the foramen magnum. Severe cases lead to a descent of the brainstem and the fourth ventricle. The descent leads to obstruction and the development of hydrocephalus.[7] Chiari malformation is broadly classified into four types:

- Chiari I malformation is characterized by the descent of the cerebellar tonsils 5 mm below the foramen magnum.
- Chiari II malformation consists of brainstem and cerebellum herniation in addition to herniated cerebellar tonsils and vermis with an associated distal spinal dysraphism/MMC.
- Chiari III malformation involves herniation of the hindbrain (cerebellum with or without the brainstem) resulting in a low occipital or high cervical meningoencephalocele.
- Chiari IV malformation is a rare variant that demonstrates severe cerebellar hypoplasia, similar to primary cerebellar agenesis.[4]

Neurological symptoms in Chiari malformation are due to direct compression of the medulla and cervical cord or syringomyelia, which develops due to abnormal CSF dynamics at the craniovertebral junction.[8] Chiari malformation is diagnosed by MRI. Treatment considerations are surgical with suboccipital decompression with C1–C2 laminectomy with or without tonsillectomy.[9]

Conclusion

The unique aspect of our case was the exceptionally low descent of the cerebellar tonsils to the level of the C7 vertebral body, which is very rarely reported in the medical literature. Most of the previous studies have reported a maximum tonsillar descent of up to the C2 vertebral level.

Conflict of Interest
None declared.

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