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Case Report

A case report of a neonate with Arnold Chiari II malformation ☆

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ABSTRACT

Chiari malformations are structural defects in the cerebellum that are characterized by the downward displacement of one or both cerebellar tonsils through the foramen magnum. A case report of a female neonate with clinical features of Chiari II malformation is presented. The diagnosis of this condition was made through a combination of clinical and radiological features. In this case, the diagnosis was confirmed by a noncontrast computed tomography (CT) scan of the brain and multiplanar brain magnetic resonance imaging (MRI).

Introduction

Arnold Chiari Syndrome is an uncommon yet remarkable disorder. It's one of the most frequent malformations in the posterior fossa, Arnold Chiari syndrome, alternatively referred to as Arnold Chiari malformation (CM), was initially described in 1890 by Hans Chiari, an Austrian pathologist. This condition encompasses a collection of congenital abnormalities affecting the central nervous system (CNS), particularly the hindbrain and posterior cranial fossa and it is usually linked to myelomeningocele [1].

Arnold Chiari Malformation Type II (ACM II), also known simply as Chiari II malformation, is a complex and relatively rare neurological disorder. This condition, often diagnosed in infancy or early childhood, is characterized by structural defects in the cerebellum and brainstem, where part of the cerebellum and the brainstem protrude into the spinal canal [1].

Due to the relatively small posterior fossa, medulla oblongata and cerebellum are displaced down into the cervical spinal canal in addition to the pons and fourth ventricular lengthening. The findings of this syndrome are detected by medical imaging of the brain. Management of ACM depends on the severity of the condition and the patient's clinical features and surgical intervention is usually required [2,3].

The exact incidence of ACM II is challenging to ascertain, but it is closely associated with myelomeningocele, a form of spina bifida. This association is observed in about 90%-95% of the cases with myelomeningocele. The prevalence of spina bifida, and by extension ACM II, varies globally but is estimated

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to affect approximately 1 in every 1000 live births [1]. However, this rate can fluctuate based on factors such as geographic location, ethnicity, and the presence of folate fortification in food.

ACM II is typically diagnosed prenatally or shortly after birth. The neonatal period, defined as the first 28 days of life, is crucial for the diagnosis and management of ACM II. This early stage is when symptoms such as a high-pitched cry, swallowing difficulties, and respiratory problems are most prominent, often leading to the initial diagnosis [4]. In addition, the presence of associated conditions like myelomeningocele can prompt early screening and diagnosis in the neonatal period.

This case report aims to highlight the role of medical imaging in diagnosis of rare congenital anomalies of the brain.

Case report

A 15-day-old female neonate was born at 38 weeks and 2 days of gestation with a birth weight of 3.50 g, the delivery was a normal delivery, Apgar score was 4 at 1 minute and 6 at 5 minutes, admitted to NICU for hydrocephalus. The mother did not undergo a prenatal ultrasound scan and she had uncomplicated pregnancy. Mother denied taking folic acid supplementation before or during first trimester.

In this case report, we document a rare presentation of Arnold Chiari Malformation Type II in a neonate, delineating the intricate diagnostic journey and therapeutic interventions. Imaging studies played a pivotal role, revealing typical features consistent with the malformation, which prompted a tailored surgical strategy.

The patient was referred for MRI which confirmed the diagnosis of Chiari Il Malformation.

An axial and reformatted coronal and sagittal images showed an abnormal small posterior fossa with downward cerebellar tonsil and vermis herniation through the foramen magnum. CT images showed effacement of prepontine and cerebella-pontine angle cisterns with compression of the fourth ventricle and subsequent supratentorial hydrocephalus (Fig. 1).

Multiplanar MRI of the brain T1 and T2 weighted images showed supra-tentorial hydrocephalus likely related to fourth ventricle outlet obstruction (Fig. 2).

The report discusses the nuances of managing this complex condition, emphasizing a holistic approach that includes neurosurgical procedures, vigilant postoperative care, and a focus on supportive therapies aimed at optimizing developmental outcomes. The case report on Arnold Chiari Malformation Type II elaborates on a comprehensive intervention strategy for a neonate diagnosed with this rare condition. The intervention included a successful neurosurgical procedure to address the malformation, followed by meticulous postoperative care. The patient's management was marked by a collaborative approach involving continuous neurological assessment, physiotherapy, and occupational therapy. These interventions were integral in managing the immediate hydrocephalus concerns and facilitating the child's developmental progress.

On follow-up 6 months later, a noncontrast CT scan was done for the patient, there was no developed hydrocephalus and no significant interval changes, patient is now under continuous follow-up.

Discussion

Chiari Malformation, described by Hans Chiari in 1891, can have a variety of etiologies, mainly genetic and environmental [1]. There are 4 recognized types of Chiari Malformation, each distinct in its anatomical presentation and severity [5]. The common feature across all types is the displacement of brain structures into the spinal canal through the foramen magnum. Chiari Malformation Type I (CM-I) involves the herniation of cerebellar tonsils below the foramen magnum [2]. Chiari Malformation Type II (CM-II), which is the focus of this report, is more complex, involving the descent of cerebellar tonsils, vermis, fourth ventricle, and brainstem [6]. Chiari Malformation Type III (CM-III) is characterized by a high cervical or low occipital encephalocele, and Chiari Malformation Type IV (CM-IV) is a rare condition associated with an underdeveloped cerebellum [5]. Each type's prevalence and specific morphological features guide diagnosis and treatment approaches.

The diagnosis of CM-II is typically made postnatally, but with advances in prenatal screening, antenatal diagnosis is increasingly common. Symptoms occur in approximately 20% of children with CM-II, with presentation varying by age [7]. Neurogenic dysphagia, affecting 66% of infants, presents challenges such as poor feeding and aspiration pneumonia. Vocal cord paralysis, seen in 31% of cases, results in phonation changes, while impaired respiratory drive and stridor, both occurring in 38% of patients, can lead to significant respiratory complications [7].

A slight gender-based prevalence difference is noted in CM-II, with a higher occurrence in females [8]. This may be attributed to the complex interplay of genetics and environmental factors, such as maternal health and exposure to certain medications [6]. The diagnosis of CM-II can be established antenatally using ultrasonography, which is highly sensitive and considered 100% specific [9]. Early detection during the anomaly scan at 20 weeks is crucial for appropriate multidisciplinary counseling and management planning [10]. Classic imaging signs include the "lemon sign" and the "banana sign," which are indicative of neural tube defects and hindbrain herniation, respectively [6].

MRI is the gold standard for demonstrating Chiari II Malformation due to its superior soft tissue contrast and ability to visualize the neural structures and CSF flow [11]. In this case, although a CT scan confirmed the diagnosis, MRI would have provided a more detailed assessment of the malformation and its impact on CSF dynamics [11,12].

Tonsillar herniation in Chiari Malformations is measured by the extent of downward displacement of the cerebellar tonsils. MRI can accurately quantify this herniation, which is typically defined as greater than 5 mm below the foramen magnum for CM-I. For CM-II, the herniation is often more pronounced and involves additional structures, making the mea-

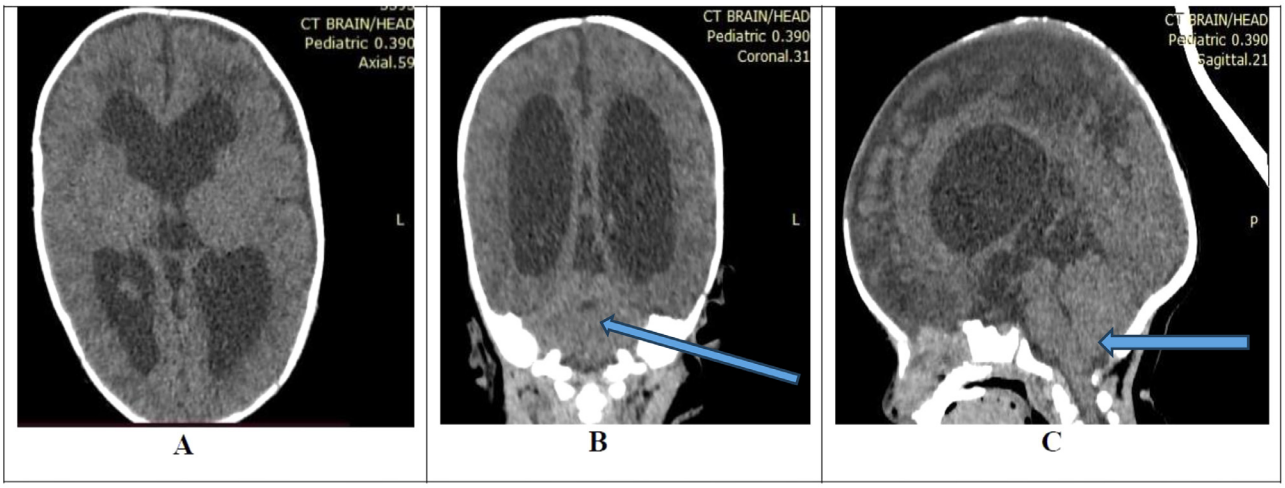

Fig. 1 – Demonstrate axial, coronal and sagittal multiplanar reconstruction of brain CT without contrast of the neonate showing abnormal small posterior fossa with downward cerebellar tonsil and vermis herniation through the foramen magnum. Effaced prepontine cistern and cerebellum-pontine angle cistern with attenuated fourth ventricle and subsequent supratentorial hydrocephalus.

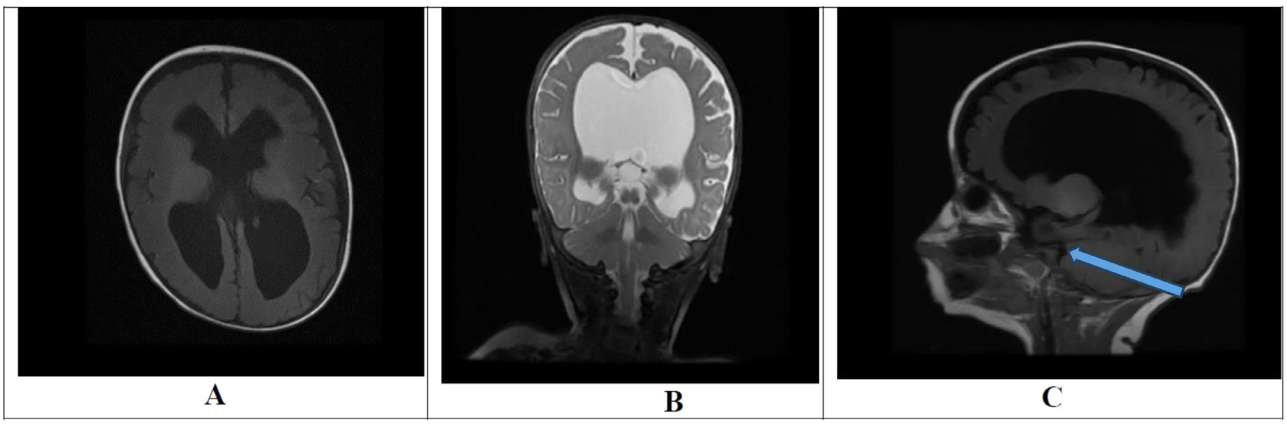

Fig. 2 – Axial T1, coronal T2, and sagittal T1 MRI shows supra-tentorial hydrocephalus likely related to fourth ventricle outlet obstruction. Findings are consistent with Arnold-Chiari Malformation type II.

surement criteria more complex and tailored to the individual patient's anatomy [4].

Treatment of CM-II is tailored to the malformation's extent and associated neurological impairments. Surgical intervention is frequently required to manage hydrocephalus, close neural tube defects, and decompress posterior fossa structures [13]. A multidisciplinary approach in spina bifida clinics ensures comprehensive care, with follow-up schedules adjusted according to the child's developmental stage [14].

Conclusion

Arnold Chiari Malformation Type II is a complex condition most prominently affecting neonates, especially those with concurrent myelomeningocele. Early diagnosis and intervention are critical for managing the condition and improving the quality of life for affected individuals. for accurate diagnosis the MRI and CT imaging is crucial.

Ongoing research is crucial for better understanding the etiology, improving diagnostic methods, and developing more effective treatment strategies.

Patient consent

Consent was obtained from the patient's parent.

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