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A Case Report on Arnold Chiari Type III: Constellation of Disorders, from Diagnosis to Treatment

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Arnold-Chiari type III is a malformation characterised by protrusion of the brainstem, cerebellum and lower part of the brain due to a cranial malformation. It is a rare congenital malformation in which the brain protrudes through a fissure in the skull. It occurs during pregnancy when the neural tube of the foetus does not close completely. The Chiari malformation type III, although described in the literature, remains rare, complicating our understanding of their common pathophysiological mechanisms and their management.

We report the case of a female infant, admitted for seizure with hypotonia and dysmophic syndrome. An MRI was performed showing encephalomeningocele leading for Arnold Chiari type III diagnosis.

The aim of this article is to understand and investigate the association between Arnold-Chiari type III malformation and encephalocele in order to develop more effective therapeutic strategies.

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Keywords: Chiari malformation type III; transfontanellar ultrasound; MRI; encephalocele; case report.

1. INTRODUCTION

The complex nature of Arnold-Chiari Type III requires careful recognition. It becomes important for all medical fields to have a holistic view of these neurological disorders. Arnold-Chiari Type III, also known as Chiari malformation Type III, has a very low occurrence in children. It is part of the spectrum of hindbrain herniation disorders. The hindbrain contents, or the so-called herniating parts, include parts of the mesencephalon, pons, medulla oblongata, cerebellum, and some of the posterior cranial fossa components and structures. At times, protrusion or herniation of hindbrain contents, meninges, and part of the skull is observed through defects in the cranium. Such a neural tube defect may result from many factors, including genetic, neurological, environmental, and teratological etiology. Most cases remain asymptomatic for a longer duration before presenting with typical hindbrain herniation symptoms (Marquez et al., 2023).

2. CASE PRESENTATION

A female patient born for a twin pregnancy, with a birth weight of 2300 g, hospitalised at H1 for hypotonia and dysmophic syndrome. An examination was carried out in the neonatal department: cardiac and abdominal ultrasound were normal. A transfontanellar ultrasound was performed, which showed an occipital encephalocele suggesting an Arnold Chiari type III malformation.

She has undergone surgery at the age of 5 months.

At the age of 18 months, she is hospitalised for management of multiple seizures repeated throughout the day, with apparent hypotonia and psychomotor impairment (can't hold the head up). The general examination on admission found: a hypotonic infant, apyrexia, absence of smooth pursuit movements with facial dysmorphia.

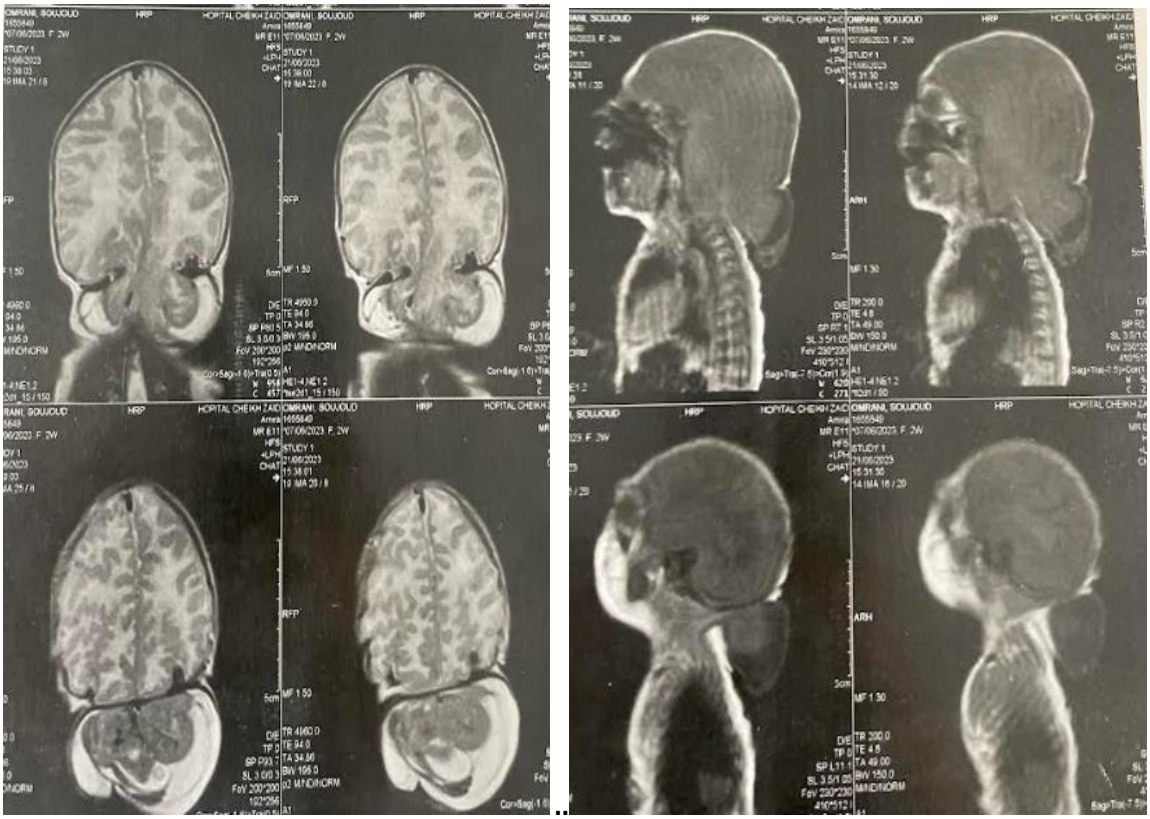

Fig. 1. Cerebral and medullar MRI of Arnold Chiari Type III malformation showing encephalocele and syringomyelia

Fig. 2. Clinical presentation of Arnold Chiari Type III showing cranial dysmorphia and absence of smooth pursuit movements

Weight and height and cranial circumference are less than two standard deviations. The eye examination found: an abolished pupillary reflex, with signs of blindness. The electroencephalography shows a diffuse slowing of background activity with multiple wave spikes. Cerebral MRI suggests Arnold-Chiari type III malformation with meningoencephalocele and syringomyelia. The patient was discharged with sodium valproate and prescribed psychomotor rehabilitation.

3. DISCUSSION

Arnold chiari Type III is defined by a severe ectopia of the cerebellum, the brainstem and IV ventricle coupled with an occipital or upper cervical encephalocele. This severe condition is very rare, accounting for less than 1% of Chiari malformations (Massimi et al., 2020; Caldarelli and Di Rocco 2010), varying from 0.65% to 4.4% in large series (Massimi et al., 2020; Cama et al., 1995; Isik et al., 2009). The estimated incidence of encephalocele is 1–2/5000 live births (about 75–90% of them being occipital) (Massimi et al., 2020; Furtado et al., 2009; Kiymaz et al., 2010). Encephaloceles are linked to embryologic vascular arachnoid between the 6th and 8th weeks of embryogenesis. 7-18% of encephalocele cases have severe mental and/or motor function disorders, depending on size, location, and brain tissue herniation. Mild intellectual prowess is seen compared to other abnormalities. Previous studies on head circumference and encephalocele are limited. (Markovic et al., 2020). Syringomyelia is a rare chronic progressive condition of various etiologies, characterized by the presence of intramedullary cavities filled with cerebrospinal fluid, and it is commonly related to Chiari malformation (Mekouar et al., 2012; Castillo et al., 1992).

All healthcare workers in the emergency, pediatric, and neurosurgical departments, as well as the general population, must have enough information about Arnold-Chiari type III for diagnosis and treatment. Clinical assessment, medical history, and neuroimaging are key tools for diagnosis. Ultrasound is used for antenatal diagnosis, while neonatal CT and pediatric MRI are used for postnatal diagnosis. Imaging findings correlate with functional impairment and symptomatic onset. Differential

diagnosis is important, considering associated cranial abnormalities. Accurate diagnosis is crucial for management, including determining intracranial volume and extent of bone defect (Khamis, 2024).

Imaging is critical for diagnosing these entities. MRI is the preferred modality. Other options include CT and ultrasound. Imaging identifies important anatomical features, aiding treatment decisions and prognosis. Radiological evaluation is key for accurate diagnosis. Modality choice depends on presentation history. Severe cases require MRI, while less typical cases can utilize MRI or ultrasound for supportive information (Pillai, et al., 2021).

There are advantages and disadvantages to each imaging modality. CT and MRI are scarce and expensive for diagnosing encephalocele. Ultrasound is the ideal protocol, with MRI also being used. MRI is the most common diagnostic method for encephalocele assessment. Non-enhanced MRI or ultrasound can be used initially, followed by area-specific neuroimaging. The best modality depends on the clinical setting and suspicion level. Lateral encephaloceles show as hypodense areas on a CT scan. Babies should be at safe weight and handled under anesthesia for CT or MRI in the neonatal period (Sefidbakht et al., 2020).

Several surgeries repair tissue layers to prevent herniation and hydrocephalus. Techniques include dural repair, skull and dural repair, skull base reconstruction, fluid shunting, and preparation for definitive repair. Initial treatment stabilizes Arnold-Chiari Type III and prevents complications. Neurosurgery and symptom-based CSF diversion are used. Antibiotics reduce CNS infection risk. Postoperative monitoring assesses neurological status. CSF infection is treated with antibiotics and leaks managed with drainage. Ultrasound and echocardiography check suture closure and cardiac complications. Long-term follow-up is required with multiple clinics (Nagy and Saleh 2021).

Low-grade Arnold-Chiari level 3 malformations with encephaloceles may not require surgery, and in some cases, just medical and physical therapy may improve outcomes. For this, it is essential to refer such cases to specialized centers. Translational neuroscience studies are doing researches to improve treatment and management in mid and long term (Jussila, 2021).

4. CONCLUSION

Arnold chiari type III malformation remains a very rare but very serious condition. Its diagnosis is relatively straightforward by clinical examination and imaging, especially with MRI. Management is multidisciplinary with a poor long-term prognosis.

ETHICAL APPROVAL

As per international standards or university standards written ethical approval has been collected and preserved by the author(s).

CONSENT

As per international standards, parental written consent has been collected and preserved by the author(s).

DISCLAIMER (ARTIFICIAL INTELLIGENCE)

Author(s) hereby declare that NO generative AI technologies such as Large Language Models (ChatGPT, COPILOT, etc.) and text-to-image generators have been used during the writing or editing of this manuscript.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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