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Case Report

Chiari type III malformation: Case report and review of literature [☆]

Younes Mekouar, M.D^{*}, Dalale Laoudiyi, M.D, Mohamed Reda Haboussi, M.D, Kamilia Chbani, M.D, Siham Salam, M.D, Lahcen Ouzidane, M.D

Department of Pediatric Radiology, Abderrahim Harouchi Children's Hospital, CHU Ibn Rochd, Casablanca, Morocco

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ABSTRACT

Chiari III (CM III) is the rarest of the Chiari malformations, characterized by high cervical or occipital encephalocele and osseous defects, associated with herniation of the posterior cerebral fossa contents through the foramen magnum.

We report the case of a female newborn, with a cervico-occipital mass, hypotonia and sharp osteotendinous reflexes in the lower limbs. An MRI was performed showing a low occipital encephalocele with caudal displacement of cerebellar tonsils.

Because of its high contrast resolution, MRI is more useful than CT, preoperatively, to assess the content of the encephalocele. MRI can identify the position of the brain stem and spinal cord, so that they are preserved during the surgical procedure.

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Introduction

Chiari malformations are complex hindbrain deformities first described and characterized by Hans Chiari in 1891 and result in a low position of the cerebellar tonsils within the upper part of the cervical spinal canal.

Chiari III (CM III) is the rarest of the Chiari malformations, characterized by high cervical or occipital encephalocele and osseous defects, associated with herniation of the posterior

cerebral fossa contents through the foramen magnum [1]. Type III has a worse prognosis than types I and II, characterized by a high mortality rate and severe neurological and developmental deficits in surviving patients [2].

Case report

We report the case of a female newborn, delivered by caesarean section following a full-term pregnancy of

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* Corresponding author. Y. Mekouar.

E-mail address: younes.mekouar@gmail.com (Y. Mekouar).

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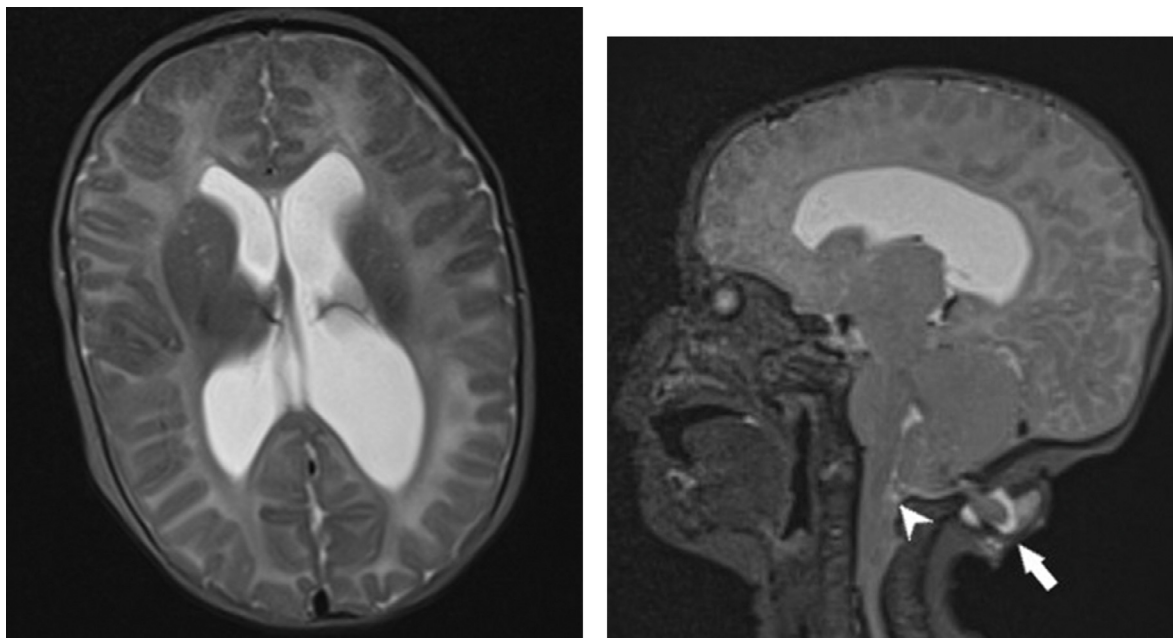


Fig. 1 – Axial and sagittal T2 weighted images showing a suboccipital meningoencephalocele (arrow) with tonsillar ptosis (head arrow) associated with moderate triventricular hydrocephalus in the context of an Arnold Chiari type III malformation

non-consanguineous parents. Morphological ultrasound performed at 22 weeks of amenorrhea showed ventriculomegaly with a posterior cervical cystic mass.

Examination of the newborn at birth found a cervico-occipital mass covered with dystrophic skin. The anterior fontanel was soft and small. The neurological examination showed marked hypotonia and sharp osteotendinous reflexes in the lower limbs.

An MRI was then performed showing a circular osseous defect measuring 10 mm in diameter, low occipital, above C1, with cerebrospinal fluid, meninges and cerebellar parenchyma with no obvious signal abnormality. There was moderate hydrocephalus with transependymal resorption in the frontal horns. The corpus callosum was present but atrophic. A caudal displacement of cerebellar tonsils is observed (Fig. 1). At the medullary region, a large fusiform syringomyelia cavity was found, extending from C6 to T10 (Fig. 2).

The neurosurgery team was consulted, which decided to operate because the brainstem was not embedded. The encephalocele was resected on day of life 20. Pathology revealed fragments of cerebellar tissue. The immediate postoperative course was uneventful but the patient failed to show signs of spontaneous breathing. The child is presently 1 month old and still requires intensive care.

Discussion

Pathogenesis

The etiology of CM III is mainly unknown. However, there is a theory that an incorrect neurulation during the ventricle extension causes an imperfect formation of the occipital area.

Later, there is a prolapse of the cerebellum and the brainstem. Another theory is that the ventricular system does not entirely descend due to abnormal neurulation. This abnormality caused a hypoplastic posterior fossa. Failure of the endochondral bone ossification is also a key factor for this condition [3].

Clinical presentation

The severity and prognosis of CMIII have been associated with the degree of brainstem embedded in the encephalocele. The malformation commonly presents as an occipital mass at birth, increasing in size as the child grows. Muzumdar et al. report a case of a newborn with 2 separate masses: an encephalocele and a myeloencephalocele [4].

Clinical findings include disordered eye movement such as titubation and downbeat nystagmus, sensory loss, weakness, ataxia, respiratory insufficiency, respiratory failure, amyotonia, hyperreflexia, dysphagia with secondary aspiration, spastic or decreased muscle tone, and inspiratory stridor [5].

The characteristics of the 2 main clinical presentations are described in (Table 1)

Imaging features

Ante-natal morphological ultrasound can reveal a cystic cervical mass, ventriculomegaly or microencephaly before birth. Although US is the modality of choice for evaluation of fetal anomalies it can be limited by maternal body habitus, fetal position, oligohydramnios, and ossification of bony structures. Recent studies have demonstrated that prenatal MRI is helpful in evaluating specific brain abnormalities, especially those involving the posterior fossa [7].

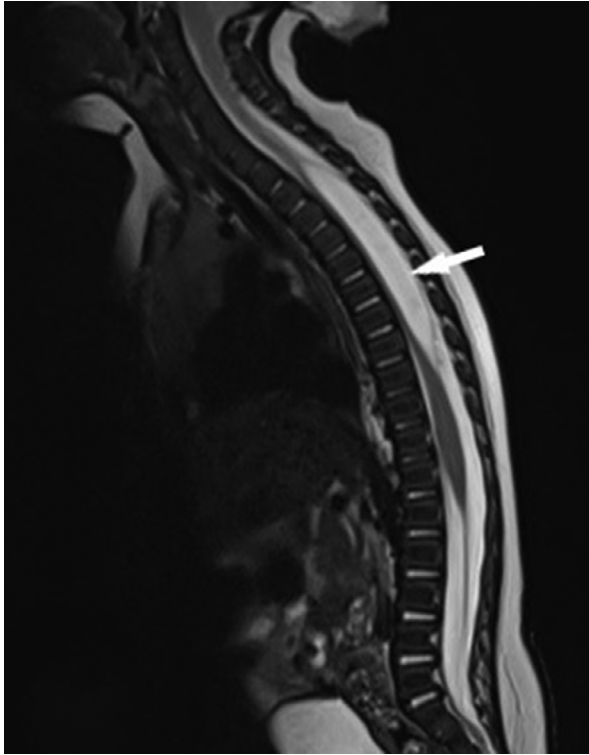


Fig. 2 – Sagittal T2 weighted image showing a fluid-filled cavity within the spinal cord : Syringomyelia (arrow)

Table 1 – Characteristics of the two main clinical presentations of CIII [6].

Encephalocele	Meningoencephalocele
Occipital bone defect	Cervical bone defect
Small posterior cranial fossa	Caudal displacement of the hindbrain
Hydrocephalus	Aberrant venous drainage
Syringomyelia	Respiratory failure
Abnormalities of the midbrain	Herniation

Postnatal MRI allows to evaluate the degree of herniated parenchyma embedded within the encephalocele, which is an important prognostic factor. The CT scan completes the work-up, better adapted to the analysis of the bone defect and all other associated bone malformations [7].

In CM III, an encephalocele or meningoencephalocele is associated with the anomalies found in CM II. The osseous defect is high cervical or suboccipital, sometimes extended to the parietal bone. The herniated tissue corresponds to cerebellar, occipital or brainstem parenchyma. It is very important in imaging to identify the position of the brain stem and spinal

cord, so that they are preserved during the surgical procedure. To avoid any possible complication during surgery, anomalies of the venous drainage (aberrant sinus) must be reported [8].

Syringomyelia is a rare chronic progressive condition of various etiologies, characterized by the presence of intramedullary cavities filled with cerebrospinal fluid. The most common etiology is Chiari malformation [8].

Conclusion

MC III is a rare malformation of the cervico-occipital joint with a poor prognosis. Because of these clinical and radiological features, it can be easily distinguished from Chiari types I and II as well as from isolated encephaloceles. Because of its high contrast resolution, MRI is more useful than CT, preoperatively, to assess the content of the encephalocele. It can indeed identify the presence of solid tissue and possible ventricular extension within the encephalocele. Preoperative identification of the brainstem and spinal cord is essential to preserve the patient's respiratory function.

Patient consent

Informed consent for patient information to be published in this article was obtained.

REFERENCES

- [1] Caldarelli M, Rea G, Cincu R, Rocco CD. Chiari type III malformation. *Child's Nerv Syst* 2002;18:207–10.
- [2] Isik N, Elmaci I, Silva G, Celik M, Kalelioglu M. Chiari malformation type III and results of surgery: a clinical study. *Pediatr Neurosurg* 2009;45:19–28.
- [3] Jeong DH, Kim CH, Kim MO, Chung H, Kim TH, Jung HY. Arnold-Chiari malformation type III with meningoencephalocele: a case report. *Ann Rehabil Med* 2014;38:401–4.
- [4] Muzumdar D, Gandhi S, Fattepurkar S, Goel A. Type III Chiari malformation presenting as intermittent respiratory stridor: a neurological image. *Pediatr Neurosurg* 2007;43:446–8.
- [5] Ivashchuk G, Loukas M, Blount JP, Tubbs R, Oakes WJ. Chiari III malformation: a comprehensive review of this enigmatic anomaly. *Childs Nerv Syst* 2015;11:2035–40.
- [6] Ortiz JF, Ruxmohan S, Alli A, Halan T, Alzamora IM, et al. Chiari malformation type III: a case report and review of literature. *Cureus* 2021;13(3):e14131.
- [7] Smith AB, Gupta N OC, Orit A GOA. Diagnosis of Chiari III malformation by second trimester fetal MRI with postnatal MRI and CT correlation. *Pediatr Radiol* 2007;37:1035–8.
- [8] Castillo M, Quencer RM, Dominguez R. Chiari III malformation: imaging features. *AJNR* 1992;13:107–13.