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Case Report

Arnold Chiari Type II: About a Case, Arnold Chiari II: Regarding a Case

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Abstract

Introduction: The type II of the Chiari malformations (I, II, III, IV) is the most frequent and important clinically due to its universal association to meningomyelocele (the most serious of the dysraphisms of the spinal column). It occurs when the spinal canal and column don´t close before birth, causing the spinal cord and its protective membrane to protrude through an opening and the presence of a sac on the back. It´s associated to hydrocephalus due to the blockage of the outlets of the IV ventricle or due to the narrowing of the aqueduct.

Objective: Presentation of a case assisted by the Cuban Medical Brigade in Bolivia, with symptoms suggestive of type II Chiari malformation. It´s an important rare disorder encountered in medical practice and are useful cases for study for Neonatologists.

Case Presentation: A male neonate product of a dystocic delivery by means of a Caesarian section due to a maternal cephalopelvic disproportion (CPD) is presented. The mother had only 2 prenatal consultations and lived in a far out rural area very distant from any health center. She had a 39 week term pregnancy, the baby weighed 3900 grams, and an Apgar 9/9. It drew to our attention the large cephalic circumference of 38 cm and a lumbar defect with a protruding sac where the meninges were observed associated with cerebrospinal fluid leakage. The defect is covered with a moist dressing with warm physiologic saline solution 0, 9%. Second choice intravenous antibiotic treatment is initiated and a cranial transfontanellar ultrasound is done.

Conclusion: When a neonate is seen with a lumbar defect, and a cephalic circumference above the 90 percentile the specialist must keep in mind the malformations of the Central Nervous System, especially of the neural tube and hydrocephalus. A harmless and immediate diagnostic procedure is a cranial transfontanellar ultrasound and consults whit pediatric neurosurgery.

Keywords: *Arnold Chiari/diagnosis; Newborn Hydrocephalus; Case Presentations*

Introduction

The first description of a congenital malformation of the brain and cerebellum with herniation of the spinal cord was made in 1883, by John Cleland, later in 1891 by Hans von Chiari and later, in 1894 Julius Arnold complete the description [1].

This malformation is a variable defect, in the formation of the brainstem, which is often associated with hydrocephalus. The most extreme form is herniation of structures of the lower portion of the cerebellum, known as cerebellar and brainstem tonsils through foramen magnum, so that parts of the brain typically reach the spinal canal by thickening and compressing it [2-4].

Type II is usually accompanied by a Myelomeningocele a form of spina bifida that occurs when the spinal canal and spine do not close before birth, causing the spinal cord and its protective membrane to protrude through a hole resembling a sack on his back [5,6]. It represents the most severe form of dysphemism of the spine, the protruding sac is composed of meninges and spinal cord, so it is accompanied by dysfunction of many organs and structures, not only of the central and peripheral nervous system, but also skeleton, skin and urinary tract [7].

Hydrocephalus occurs due to blockage of the outlet holes of the ventricle OR by tightness associated with the aqueduct, structures through which cerebrospinal fluid normally circulates. Arnold Chiari malformation may appear isolated, although it is often associated with other brain and spinal cord malformations such as syringomyelia and spina bifida [3,6].

Confirmation diagnosis is made by nuclear magnetic resonance imaging, which must include the entire spinal cord, in order to delimit the extent of syringomyelia, evaluate the structures of the posterior pit and the presence of hydrocephalus. Treatment is done by surgical decompression and several interventions are often needed, infants and children with myelomeningocele may need an operation to reposition the spinal cord and close the hole your back [7].

Hydrocephalus can be treated with a bypass system that drains excess fluid and relieves pressure inside the head. A strong tube surgically inserted into the head is connected to a flexible tube placed under the skin, where it can drain excess fluid into the chest or abdomen to be absorbed by the body [2].

Neural tube closure defects are a common condition in Pediatric Neurosurgery. An important goal during surgery is to create a barrier between the spinal canal and the outside, the technique most commonly used for this is the suture of the thoracolumbar fascia over the reconstructed dural sac [8].

Case Description

A male neonomine is admitted to the Neonatology service of the Roberto Galindo Terán Hospital of the Capital Cobija in Bolivia, born from a dystocic cesarean birth due to maternal cephalic deproportion (DCP), with only 2 consultations prenatal for proceedings from rural area far away from the health center, gestation time of 39 weeks, weight 3900 grams Apgar 9/9 clear amniotic fluid, placenta and normal cord VDRL non-reactive group O positive, which at birth attracts attention that presents increased DC: 38 cm and lumbar defect with sac protruding where meninges and cerebrospinal fluid are observed, covered with sterile dressing and physiological saline solution at 0.9% tibia and transferred to the neonatology service.

The physical examination observes eutrophic neonate with good coloration of skin and mucous membranes not apnea or cyanosis saturating 98 % with Fi02 of 0.21, CC 38 cm defect in lumbar region with tumor-looking sac where the meninges are observed, is placed in prone decubitus in thermal car to temperature 34°C. Channeled is initiated by inline antibiotic (Cefotaxime and Ampicillin) by risk of sepsis of the central nervous system, remains covered the defect in lumbosacral region, indicated study of blood chemistry and ultrasound (USD) renal and transfontanelle to rule out other malformations.

It stabilizes and moves to the III level, at the service of Neonatology in Peace to complete study and surgical treatment.

Complementary

- Hto: 0.50 fv
- LCN: 7,5 x 10/l

- Linf: 0,69
- Segm: 0,28
- Mon: 003
- Platelet Count: 306 x 10/l
- Blood sugar: 3.1 mmol/l
- PCR: negative
- USD Renal: kidneys of normal size and position with good parenchyma without telangiectasia, empty bladder. Normal adrenal glands
- USD Transfontanelle Ventricular Dilation (Obstructive Hydrocephalus).

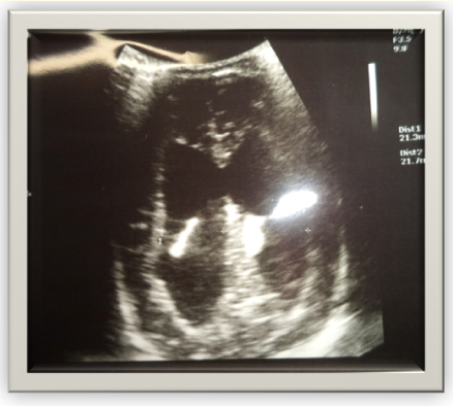

Figure 1

Coronal cut

- Right Occipital Horn: 21.3 mm.
- Left Occipital Horn: 21.7 mm with normal echogenicity choroid plexus floating inside.

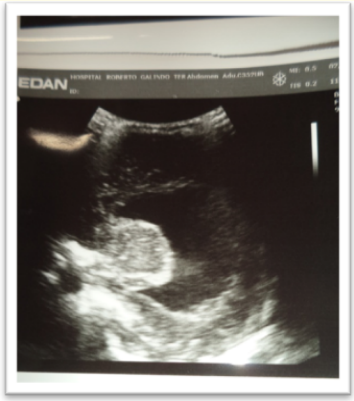

Figure 2

Sagittal cut

- Callous body present optical thalamus and normal caudate nucleus, heartbeat of present cerebral arteries.
- Side ventricle at 25 mm atrium level. Hydrocephalus.

Axial cut

- Right IVLH: 31/45 - 0.58.
- IVLH Left: 32/46 - 0.60.

Increased index side ventricles/Hemispheres.

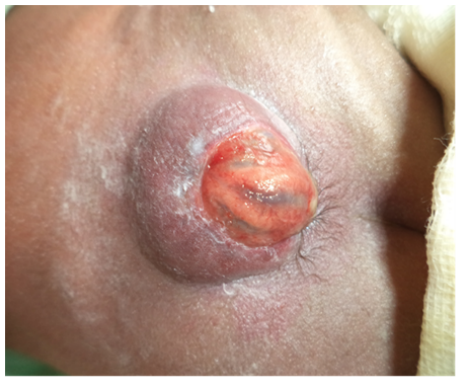

Figure 3: *Myelomeningocele Lumbosacro (herniary sac containing mofle and spinal membranes).*

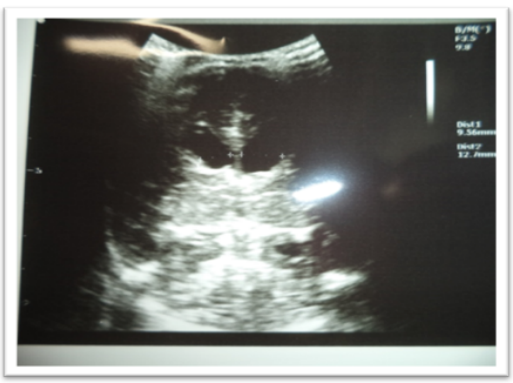

Figure 4: *Corte Coronal.*

Normal Interhemispheric caesura

- Right front horns 9.5 mm.
- Left Front horns: 12.7 mm.

Discussion

It is important to know the different types of Malformation of Arnold Chiari their form of presentation and clinical symptomatology to arrive at the diagnosis of certainty and timely treatment.

Four types of malformation are identified

1. **Type I:** Flow herniation of the cerebellar tonsils, greater than 5 mm, below the foramen magnum. It is not usually accompanied by descent of the brain stem or fourth ventricle or hydrocephalus; characteristically is associated with syringomyelia. It is the most common type in the child population and is usually only detected in adulthood, during a test aimed at diagnosing other diseases. All acquired or secondary Chiari malformations belong exclusively to Type I.
2. **Type II:** Is the so-called classic Chiari malformation (or Arnold-Chiari malformation itself); involves the protrusion of cerebellar structures and also of the brain stem through the foramen magnum; this complication can lead to this complication can lead to partial or total paralysis below myelomeningocele. Other types of intracranial alterations (tentative hypoplasia, craniolacunia, and Silvio aqueduct abnormalities) may be observed.
3. **Type III:** Is the most severe form of abnormality. The herniated cerebellum and brain stem are introduced into the cervical spinal canal, often accompanied by the fourth cerebral ventricle and compress the spinal cord, causing severe neurological symptoms.
4. **Type IV:** Involves an incomplete development of the structures of the cerebellum, a disease known as cerebellar hypoplasia [1].

The clinic of Arnold Chiari syndrome is highly variable expression, depending on the position, the degree of compression, the level of cell degeneration of the cerebellar tonsils and the presence or not of syringomyelia.

For the diagnosis, the background, prenatal ultrasound results, alpha fetoprotein, clinical symptomatology and imaging studies such as the simple x-ray showing the existence of craniolacunia (typical defects) should be evaluated rounded in the bones of the skull vault [9].

Transfontanellar skull ultrasound useful for the diagnosis of hydrocephalus, is harmless does not emit ionizing radiations and can be taken to the patient's bedside, facilitating immediate diagnosis of ventricular dilation and other malformations associated with the syndrome such as those of the urinary tract. Hydrocephalus is almost always of the non-communicating obstructive type and is due to compression of the structures of the posterior pit against the occipital hole, although sometimes there is an associated stenosis of the Silvio aqueduct [10].

Neural tube defects and hydrocephalus require surgical treatment and are a common condition in Pediatric Neurosurgery.

The prognosis is dependent on neurological commitment and associated malformations.

Conclusion

In the presence of a neonate with lumbar defect and cephalic circumference above 90 percentile the specialist should take into account the malformations of the central nervous system specifically of the neural tube and hydrocephalus, such as more innocuous and immediate diagnostic means performing Transfontanellar skull ultrasound and assessment with pediatric neurosurgery to define treatment.

Bibliography

1. Hernández Ruiz I and Soler Cano A. "Arnold Chiari Type I Malformation: presentation of a case". *Revista Médica Electrónica* 32.5 (2010).
2. Izquierdo Martínez M and Avellaneda Fernández A. "Rare Diseases: A Practical Approach". 51.12 (2004).
3. Ruiz de la Paz M., *et al*. "Mielomeningocele with malformation of Arnold Chiari type II: presentation of a case". *Mediciego* 15.2 (2009).

4. Hernández León L., etal. "Sacral agenesis associated with spinal dysmorphism and hydrocephalus". *Revista Cubana de Pediatría* 85.2 (2013): 265-272.

5. Martínez Sabater A. "Arnold-Chiari malformation: loss smile". *Index de Enfermería* 23.4 (2014): 256-259.

6. Camacaro L., *et al*. "Malformation of Arnold Chiari Type II Presentation of a case". *Revista Médica Electrónica Portales Medicos* (2014).

7. Rodríguez Loureiro JL., *et al*. "Myelomeningocele lumbosacro: case presentation". *Revista Cubana de Neurología y Neurocirugía* 2.1 (2012): 43-46.

8. Moya de Armas JM., *et al*. "Polypropylene mesh in the surgical closure of the infant's spina bifida". *Acta Médica del Centro* 6.1 (2012).

9. Sorolla Parker JP. "Craniofacial abnormalities". *Revista Médica Clínica Las Condes* 21.1 (2010): 5-15.

10. Martínez Sabater A and Sancho Cantus D. "Arnold-Chiari Malformation and Syringomyelia in primary care. About a case". *Medicina de Familia. SEMERGEN* 38.5 (2012): 331-334.

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