Arnold Chiari type III malformation. A case report

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Case Report

Pediatrics

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Background
Arnold-Chiari malformation is a rare disease, which consists of an anatomical alteration of the base of the skull, in which herniation of the cerebellum and brain stem occurs through the foramen magnum to the cervical canal. There are four types of presentation. Type I is the most common in adults; Pediatric forms of Chiari malformation (type II and type III) are present at birth. Type III: is a rare combination of small posterior fossa with occipital or high cervical encephalocele (1) Is presented the case of a guatemalan newborn male patient without referring personal prenatal or perinatal history, documented occipito-cervical protuberance at birth, which is why he was referred to neurosurgery department for treatment. After performing the first imaging studies and talking to the mother about the risks and complications of the treatment, she decides to reject them and take the patient home.

Keywords: Arnold-Chiari malformation.

The Arnold Chiari Chiari III malformation is the least common of the Chiari malformations, characterized by cervico-occipital encephalocele and bone defects, associated with herniation of the contents of the posterior cranial fossa through of the foramen magnum (2,3); and has a worse prognosis with a high mortality rate and severe neurological sequelae (2).

Case report

Is presented the case of a guatemalan newborn male patient without referring personal prenatal or perinatal history. The physical examination revealed an alert patient in regular general condition with a weight of 2800 grams, height of 48 cm, head circumference of 36 cm, temperature of 37 °C, respiratory rate of 30 per minute, heart rate of 128 per minute, arterial saturation oxygen at 85%, observing occipital encephalocele, the rest of the physical examination is normal., which is why he was referred to neurosurgery department for surgical treatment. It was performed a brain CT in simple and contrast phase with axial, coronal and sagittal sections, showing "lemon skull", absence of the cranial table in the occipito-parietal region, being more evident on the left side; through this defect is visualized the formation of an encephalocele, which causes retraction of the occipital horn of the corresponding lateral ventricle, thus conditioning the presence of colpocephaly. Likewise, herniation of the cerebellar tonsils is observed up to 0.5 cm below McRae's line. The findings described above being compatible with malformation of Arnold-Chiari type III.

Discussion

The Arnold Chiari Chiari III malformation is the least common of the Chiari malformations, accounting for approximately 1 to 4.5% of these malformations, characterized by cervico-occipital encephalocele and bone defects, associated with herniation of the contents of the posterior cranial fossa through of the foramen magnum (2,3) The etiology of Chiari malformation is thought to be secondary to an aberration in the dorsal induction process through which the embryonic neural tube is formed and closed (3). This process begins at the future craniocervical junction and then extends craniocaudal at approximately 3-5 weeks of gestation (3). Type III has a worse prognosis with a high mortality rate and severe neurological sequelae (2). The differential diagnosis to take into account is an isolated occipital encephalocele. Also some causes of an occipital encephalocele, such as Meckel Gruber syndrome, multerian duct and renal agenesis, cervicothoracic somite dysplasia and Walker-Warburg syndrome (3)

Conclusion

The Arnold Chiari Chiari III malformation is the least common of the Chiari malformations. The etiology of Chiari malformation is thought to be secondary to an aberration in the dorsal induction process through which the embryonic neural tube is formed and close and it has a worse prognosis with a high mortality rate and severe neurological sequelae. Imaging studies are essential for a proper diagnosis and decision making to offer the best treatment.

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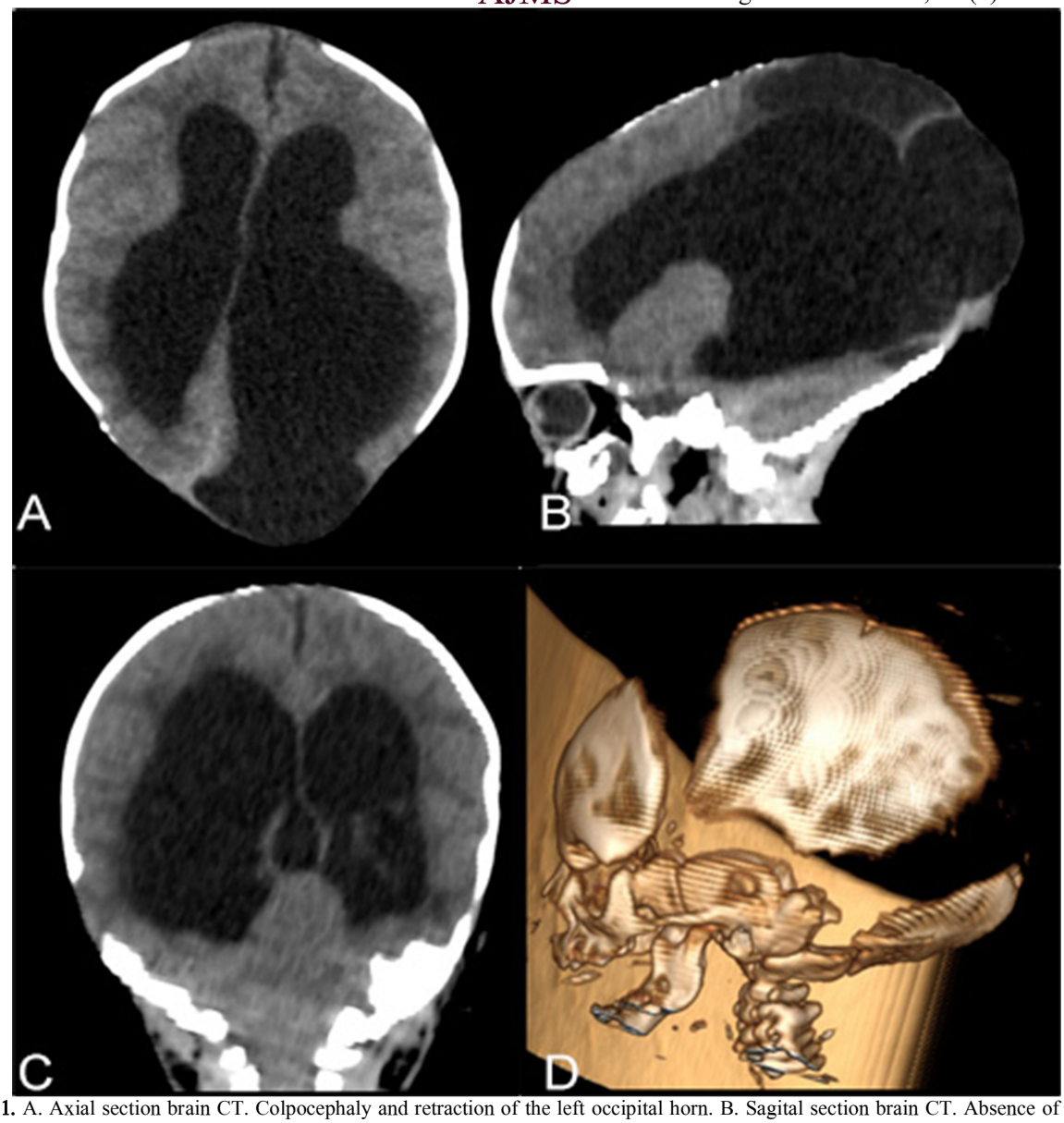

Figure 1. A. Axial section brain CT. Colpocephaly and retraction of the left occipital horn. B. Sagital section brain CT. Absence of the cranial table in the occipito-parietal region, being more evident on the left side; through this defect is visualized the formation of an encephalocele. C. Coronal section brain CT. Dilatation of the lateral ventricles and third ventricle. D. 3D reconstruction. ¨Lemon Skull¨ appereance due to the increase in the anteroposterior diameter with the absence of the cranial table in the occipito-parietal region.

Conflicts of interests

There was no conflict of interest during the study, and it was not funded by any organization.

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- To the memory of the patient because his case serves as a teaching through this publication.
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