www.jcimcr.org

JCIMCR OPEN ACCESS Journal of Clinical Images and Medical Case Reports

ISSN 2766-7820

Case Report

Open Access, Volume 5

Congenital hydrocephalus: A case report

J Fahri*; S Hani; Y Amime; S El Moussaoui; W Lahmini; M Bourrouss

Pediatric Emergency Department, Mohammed VI University Hospital, Morocco.

***Corresponding Author: J Fahri**

Pediatric Emergency Department, Mohammed VI University Hospital, Morocco.

Email: drjihanefahri@gmail.com

Received: Sep 12, 2024

Accepted: Oct 08, 2024

Published: Oct 15, 2024

Archived: www.jcimcr.org

DOI: www.doi.org/10.52768/2766-7820/3296

Abstract

Hydrocephalus is the accumulation of excessive quantities of cerebrospinal fluid, causing dilation of the cerebral ventricles and/or an increase in intracranial pressure. Manifestations may include an enlarged head, bulging fontanelle, irritability, lethargy, vomiting and convulsions. Diagnosis is based on ultrasonography in newborns and young children with a permeable fontanel, and on brain scans or MRI in older children. Treatment varies from observation to surgery, depending on the severity and progression of symptoms. We report in this article the case of a newborn admitted at H6 of life to our pediatric emergency department for hydrocephalus, in order to summarize the clinical features, complete treatment and prognosis of neonatal hydrocephalus.

Keywords: Neonatal hydrocephalus; Newborn; Treatment; Prognosis.

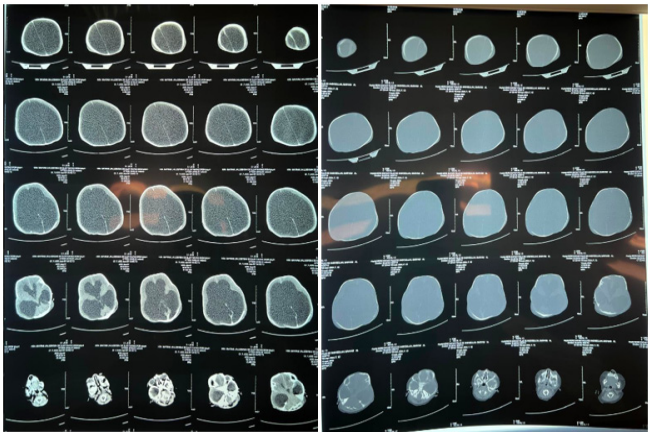
Introduction

Congenital hydrocephalus is characterized by ventricular distension secondary to an increase in cerebrospinal fluid volume, without prejudging its cause. The cranial perimeter is increased. This accumulation can lead to increased intracranial pressure and neurological complications. Causes of this condition include obstructions in the flow of CSF, cerebral malformations or prenatal infections. According to INSERM, the prevalence of congenital hydrocephalus is 46.5/100,000 at birth in 2022 in Europe, while its prevalence and incidence in many African countries, notably Morocco, are unknown. Diagnosis is based on cranial perimeter measurements and imaging studies, and the main treatment is often the placement of a shunt to drain excess fluid. Rigorous medical follow-up is crucial to optimize the newborn's chances of development and well-being.

Clinical observation

Female newborn admitted on the first day of life, from a 1st degree consanguineous marriage, to a 25-year-old mother with three gestites and three parities with no particular pathological history. The pregnancy was monofetal and poorly monitored, estimated at 38 SA + 5 days according to the date of the last menstrual period. Delivery took place in a peripheral hospital by caesarean section for bicatric uterus. The amniotic fluid was stained with a pea purée appearance. At birth, the Apgar score was 6, rising to 8 at 5th minutes. Clinical examination revealed major macrocrania with a head circumference of 54 cm, horizontal nystagmus, a sunset gaze and respiratory distress rated 3/10th according to Silverman's score, with no other abnormalities detectable at the limit of the clinical examination. The newborn was transferred to our unit at H6, initially hemodynamically and respiratorily stabilized. The infectious workup on admission was positive, with hyperleukocytosis at 26056/mm^3, predominantly neutrophilic polynuclear, and C-reactive protein at 106 mg/l. The initial chest X-ray showed an alveolar syndrome affecting both lung fields with thoracic distension. A brain scan revealed major active tri-ventricular hydrocephalus laminating the cortex with amygdala involvement. The newborn was started on dual antibiotic therapy with ceftriaxone 100 mg/kg/d combined with gentamycin 5 mg/kg/d. A neurosurgical opinion was sought; the newborn was scheduled for ventriculoperitoneal bypass.

Figure 1: Female newborn with major macrocrania.



Figures 2,3: Cerebral CT scan showing active tri-ventricular hydrocephalus with tonsillar involvement.

Discussion

Congenital Hydrocephalus (CH) is characterized by abnormal dilatation of the cerebral ventricles, and encompasses a diverse set of disorders present from birth [1]. This distension of the cerebral ventricles results from a dysfunction in the passage of cerebrospinal fluid, which fails to circulate properly from the choroid plexuses, where it is produced, to the arachnoid villi, where it should be absorbed [2]. Congenital hydrocephalus includes any form of primary hydrocephalus diagnosed before or after birth [3]. According to a recent systematic review and meta-analysis of epidemiological studies, the prevalence of congenital hydrocephalus varies considerably according to geographical region and type of congenital malformation registry [1]. The estimated worldwide prevalence of HC was 8.5 per 10,000 live births. A higher prevalence of HC was observed in Africa, Asia and South America than in other continents [4,5]. Similarly, a higher prevalence of HC was observed in low- and middle-income countries in Africa and South America than in high-income countries in Europe and North America (12.3 versus 7.9 per 10,000 births, respectively) [4,1]. A case-fatality rate of 25% has been reported for neonates with congenital hydrocephalus in the early neonatal period [6,7]. The pathogenesis of pediatric hydrocephalus is multifaceted, and its etiology differs according to a country's level of development. The pathogenesis of pediatric hydrocephalus is complex, and its etiology varies according to countries' level of development. In newborns, when hydrocephalus occurs without any obvious extrinsic cause, it is generally referred to as congenital hydrocephalus, as it is often present from birth. On the other hand, when hydrocephalus develops as a complication of another condition, such as hemorrhage, infection or neoplasm, it is called acquired or secondary hydrocephalus [8]. In developed countries, pediatric hydrocephalus is mainly caused by intraventricular hemorrhage linked to prematurity or congenital causes. In Africa, the post-infectious etiology varies between 7% and 60%, and is closely linked to the general level of public health in the country [9-11]. It is the main cause of hydrocephalus in newborns and infants, due to a higher incidence of neonatal sepsis. However, this predominance of post-infectious hydrocephalus is tending to diminish, especially in infants, with an etiological profile tending towards that of developed countries. Malformations are implicated in certain cases, notably those of the neural tube. Although these malformations are multifactorial in origin (genetic, environmental factors, etc.), it has been established that they are correlated with low folic acid (or vitamin B9) intakes. Despite the efforts made in the field of prevention through the generalization of extended vaccination programs, much remains to be done to raise public awareness. Better access to prenatal folic acid supplementation for women of childbearing age could further reduce the incidence of this pathology, which has serious neurological consequences. Although MRI and CT are the techniques of choice for the study of hydrocephalus [12,13], Trans-Fontanilla Ultrasound (TFE) also plays a crucial role in the diagnosis and characterization of brain lesions in newborns. It is often considered the first-line method for assessing at-risk newborns, sometimes being the only method required [14]. In Morocco, ETF is commonly used due to its superior availability compared with CT and MRI, particularly in remote rural areas. Being relatively easy to perform and effective in diagnosing hydrocephalus, ETF should be further disseminated in our health centers, with adequate training for nursing staff to facilitate its use. Popularizing its practice could have a positive impact on the time to diagnosis and therefore on the prognosis of hydrocephalus. Ventriculoperitoneal shunting (DVP) remains the gold standard treatment for hydrocephalus. Complication rates after two years generally vary between 1% and 50% [10-17]. In sub-Saharan Africa, complications range from 7% to 69%, mainly due to mechanical causes (11-54%) and infections (7-69%). DVP placement is often riskier than in the developed world, due to infectious complications and valve malfunction [25]. According to Choux et al. rigorous protocols and staff training in infection control measures during the preoperative, operative and postoperative periods can significantly reduce infectious complications [29]. VCE is currently used in a wide range of indications in the treatment of hydrocephalus. It is even used in post-meningitic hydrocephalus, which was long considered a relative contraindication, with success rates of between 59 and 77% [10-19]. Rigorous post-natal follow-up is essential to manage congenital hydrocephalus. By carefully monitoring shunt function, assessing neurological development, and providing support to families, it is possible to optimize outcomes and improve quality of life for affected children.

Conclusion

Congenital hydrocephalus is a relatively rare condition whose pathophysiology is well understood. Hydrocephalus can have serious consequences for the child in the medium and long term, as well as for the mother during childbirth.

Antenatal ultrasound helps to diagnose this pathology, and screening ultrasounds are recommended at 12, 22 and 32 weeks' gestation. Although these examinations are not compulsory, they do enable early detection of pathologies such as hydrocephalus, and provide a perspective on the evolution of the condition, enabling complementary examinations to be carried out and appropriate treatment to be proposed.

References

1. Isaacs AM, Riva-Cambrin J, Yavin D, Hockley A, Pringsheim TM, et al. Age-specific global epidemiology of hydrocephalus: Systematic review, metanalysis and global birth surveillance. PLoS One. 2018; 13(10): 0204926. 10.1371/journal.pone.0204926.

2. Rekate HL. Classification of hydrocephalus. In Cinalli G, Ozek MM, Sainte-Rose C (Eds.), Pediatric hydrocephalus Cham: Springer International Publishing. 2018; (1-17). 10.1007/978-3-319-31889-9_45-1.

3. Morota N. Prenatal hydrocephalus: Prenatal counseling, postnatal treatment, outcome. In Cinalli G, Ozek MM, Sainte-Rose C (Eds.), Pediatric hydrocephalus. Cham: Springer International Publishing. 2019; (1-19). 10.1007/978-3-319-31889-9_48-1.

4. Dewan MC, Rattani A, Mekary R, Glancz LJ, Yunusa I, et al. Global hydrocephalus epidemiology and incidence: Systematic review and meta-analysis. Journal of Neurosurgery. 2019; 130(4), 1065-1079. 10.3171/2017.10. JNS 17439.

5. Huang YH, Wu QJ, Chen YL, Jiang CZ, Gong TT, et al. Trends in the prevalence of congenital hydrocephalus in 14 cities in Liaoning province, China from 2006 to 2015 in a population-based birth defect registry from the Liaoning Women and Children's Health Hospital. Oncotarget. 2018; 9(18): 14472-14480. 10.18632/oncotarget.24239.

6. Rogers SC, Morris M. Infant mortality from spina bifida, congenital hydrocephalus, monstrosity, and congenital diseases of the cardiovascular system in England and Wales. Annals of Human Genetics. 1971; 34(3): 295-305.

7. Scala C, Familiari A, Pinas A, Papageorghiou AT, Bhide A, et al. Perinatal and long-term outcomes in fetuses diagnosed with isolated unilateral ventriculomegaly: Systematic review and meta-analysis. Ultrasound in Obstetrics & Gynecology: The Official Journal of the International Society of Ultrasound in Obstetrics and Gynecology. 2017; 49(4): 450-459.

8. Hannah Tully M, William Dobyns B. Infantile hydrocephalus: A review of epidemiology, classification and causes. EUR J Med Genet. 2014; 57(8): 359-68.

9. Peacock WJ, Currer TH. Hydrocephalus in childhood: a study of 440 cases. S Afr Med J. 1984; 66(9): 323-4.

10. Benjamin Warf C. Hydrocephalus in Uganda: the predominance of infectious origin and primary management with endoscopic third ventriculostomy. J Neurosurg. 2005; 102(1): 1-15.

11. Ba MC, Kpelao ES, Thioub M, Kouara M, Thiam AB, et al. Hydrocéphalie post-méningitique chez les nourrissons à Dakar (Post meningitis hydrocephalus in the infants in Dakar) Afr J Neurol Sci. 2012; 31: 8-15.

12. Dincer A, Ozek MM. Radiologic evaluation of pediatric hydrocephalus. Childs Nerv Syst. 2011; 27(10): 1543-62.

13. Pomschar A, Koerte I, Peraud A, Heinen F, Herber-Jonat S, et al. Hydrocephalus in childhood: Causes and imaging patterns. Radiology. 2012; 52(9): 813-20.

14. DjientcheuVde P, Nguefack S, Mouafo TO, Mbarnjuk AS, Yamgoue TY, et al. Hydrocephalus in toddlers: The place of shunts in sub-Sahara African countries. Childs Nerv Syst. 2011; 27(12): 2097-100.

15. Warf BC. Pediatric hydrocephalus in East Africa: Prevalence, causes, treatments, and strategies for the future. World Neurosurg. 2010; 73(4): 296-300.

16. Gathura E, Poenaru D, Bransford R, Albright AL. Outcomes of ventriculoperitoneal shunt insertion in Sub-Saharan Africa. J Neurosurg Pediatr. 2010; 6(4): 329-35.

17. Drake JM, Kestle JR, Tuli S. CSF shunts 50 years on-past, present and future. Childs Nerv Syst. 2000; 16(10-11): 800-4.

18. Schroeder HW. Success of endoscopic third ventriculostomy: What does reallymatter? World Neurosurg. 2012; 78(3-4): 233-4.

19. Figaji AA, Fieggen AG, Peter JC. Endoscopic third ventriculostomy in tuberculous meningitis. Childs Nerv Syst. 2003; 19(4): 217-25.