CASE REPORT

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A Case Report of a Dandy-Walker Syndrome in a girl baby

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Dandy-Walker anomaly is a rare congenital disorder. This syndrome is characterized by complete or partial agenesis of the cerebellar vermis, cystic dilatation of the fourth ventricle and enlargement of the posterior cavity with or without hydrocephalus. This neurological syndrome is more common in girls than in boys. The most important abnormality is the presence of a large dilatation of the fourth ventricle, which has been cysts and has been covered by a neuroglial vascular membrane. Hydrocephalus is not seen at birth, often occurs at 3 months, and is proven at 1 year in 80% of cases. In addition to hydrocephalus, pale, ataxia, abnormal breathing, seizures and prominent occiput may be present. Mental retardation is common due to brain abnormalities in these infants; a major fetal defect in this disorder is still discussed. Pre-birth ultrasonography may show hydrocephalus and ventricular enlargement. Postpartum diagnosis is by CT scan and MRI. In the case of progressive hydrocephalus, the treatment is surgical.

INTRODUCTION

Dandy Walker syndrome is a congenital intracranial deformity that includes a set of abnormalities of the posterior cranial cavity (1, 2). It develops at the 9th week of pregnancy at the time of the formation of the fourth cerebellum and ventricle (3); it is usually diagnosed after birth (4). The syndrome, first described and presented by Dandy and Blackfan in 1914, is characterized by the following tricusts: Posterior cavity enlargement by displacement of sinus transventive tenturium and turkula, cystic dilatation of the fourth ventricle and agenesis of the cerebellar vermis (6). The prevalence of Dandy Walker syndrome is 25,000 to 35,000 births (7). According to the results of studies that have been done so far, the disease is more common in girls than in boys (8). Although some reports have suggested that gene mutations are partly responsible for the development of the disease, the causative agent of the disease is still unclear (9). Dandy Walker syndrome is also a known cause of congenital hydrocephalus (10), 70-90% of patients have hydrophilic infections with enlarged lateral ventricles (11). Other infants also have other abnormalities including corpus callosum, occiput encephalocele, facial angioma, palatal cleft in the middle line, and cardiovascular and cystic kidney abnormalities (12, 21). The mortality rate is reported to be between 10% and 66% (13). Given the inadequate knowledge of the causes and the physiopathology of the syndrome, as well as the absence of a specific phenotype of this anomaly, each family with a history of certain genetic diseases requires more attention and more directed guidance (11). The purpose of this report is to help in understanding this rare disease, its manifestation and diagnosis through clinical signs and paraclinical methods. Familiarity with clinical symptoms and complications of this disease in different organs is one of the needs of the treatment staff. With the timely diagnosis, it might be possible to prevent the problem from occurring.

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CASE REPORT

This project was approved by the code of ethics of ajums IR.AJUMS.REC.1397.591 in student research committee of Ahvaz Jundishapur University of Medical Sciences. After taking the code of ethics and also the financial support of this university, we did their research project. The term infant, girl, weighing 4 kg, was the result of a selective caesarean section with Apgar 9/10, from a 29-year-old $G_3P_2Ab_1$ mother. Within a few minutes after birth, it has seizures, restlessness, tonic movements of hands, apnea, cyanosis, groin and bradycardia. The baby's seizure was controlled with two phenobarbital and phenytoin drugs. There was a murmur 4/6 in the baby's examination, chest retruction, Abnormal lung sounds (rally sound) and a newborn's hypotonia. The soft abdominal and organomalagy were absent, examination of other organs was normal. Due to hypotonia, the baby was fed by gavage. The baby was monitored for early diagnosis of brain structural disorder and seizer. The results of paraclinical tests were as follows: BS = 95, BC = Negative, PH = 7.3, Pco2 = 36, Po2 = 84, Hco3 = 20, Na = 134, K = 3.5, Ca = 8.5, WBC = 18000, Hb = 15.5 , PLT = 152000 Also, in the examination of mass effect CT scan was not seen in different regions of parenchyma, no signs of infarction, intracranial hemorrhage, hydrocephalus and middle line shifts, wide post fossa were observed, part of cerebellar vermicomy in the posterior region cerebellar hypoplasia and extensive cerebral involvement have been reported based on these symptoms of Dandy Walker syndrome.

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Image1 Brain CT scan in a girl baby with Dandy-Walker Syndrome

Dandy-Walker Syndrome in this girl baby is visible in Brain CT scan (Image 1).

The baby was discharged with a doctor's advice at the age of 15 days after controlling the seizure and training the mother to feed the baby by gavage. Eventually, the baby died at 7 months of age, due to lethargy and well-being. Parents' marriage is a kind of family (cousin, uncle's daughter). The baby's mother had a history of hypothyroidism during pregnancy; there was no problem in embryonic screening tests. The father of a 35-year-old child has no specific disease history. The first child was a girl with a female gender, who died at the age of 28 days with the diagnosis of congenital malformation of the cerebellum and seizure. The first child of the aunt was born of a family marriage with an abnormality of bowel obstruction. The second child of the unclean baby is a non-familial marriage with major malformations. The baby's uncle died at the age of 37 when he died of con's illness. Due to the fact that there have not been reports of such anomalies in the province of Khuzestan and also in the city of Dezful, we have decided to present this case report.

DISCUSSION

Dandy Walker syndrome is a rare, complex, and multifactorial disease (14). Although the syndrome has been seen as familial, it appears in most cases as a single occurrence (15). DWS may be due to chromosomal abnormalities or environmental factors (16). It has been reported in families that 3 children were involved in the syndrome in a family of them. Studies have shown that transmission of these rare familials, autosomal recessive or reversible X-linked. In the fetal period, the disease can be accompanied by spinal cord abnormalities (14). The prognosis of the disease depends on the severity of the syndrome and related anomalies. In people with DWS, rational growth varies. Some children have natural cognitive progress, while others, even when hydrocephalus is treated early, can never have rational natural development. Demographic studies showed that the deaths of children with Dandy Walker syndrome were very high (ten times more than the control group). However, CSF surgical evacuation techniques are beneficial and reduce mortality (17). The environmental factors associated with this disease include rubella, cytomegalovirus, toxoplasmosis or warfarin use in the first trimester. Maternal diabetes during pregnancy is also associated with increased risk of fetal death to the DWS. However, brain-related abnormalities associated with DWS often occur as a separate condition, and the cause is usually unknown. Also, when there is no chromosomal relation to the cause of the disease, the risk of family recurrence is relatively low and between 1% and 5% (18). It has also been seen that in 10 to 20% of people with DWS, signs and symptoms do not appear until the end of childhood or adulthood. The apparent symptoms of the illness may occur in different ways, and may be signs of increased intracranial pressure, such as irritability, vomiting, headache, and seizure. There may also be signs of a brain disorder, including motor instability, muscle imbalance, and impotence in the rapid movement of the eyes. A case report has revealed hyperactivity symptoms, stereotypic movements and enuresis at age 14 (19). Using evidence from morphological, metabolic and neuro-functional imaging, cerebellar involvement has been proven in various types of psychiatric illnesses such as schizophrenia, autism, depression and obsessive-compulsive disorder. The psychiatric manifestations of the Dandy Walker Complex (DWC) range from a range of psychosomatic symptoms to cognitive symptoms. In a study by Sinha et al., On a 25-year-old man with a 3-year history of psychiatric symptoms, simple magnetic resonance imaging of the brain, lower vermiculate hypoplasia associated with the fourth ventricular relationship with

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Sisterna magna without hydrocephalus showed a kind of dandy waker (20). The syndrome can be detected using advanced imaging techniques. Therefore, it is better to take routine pregnancy care, especially imaging measures, to be more sensitive to the birth of the baby with this abnormality.

REFERENCES

1. Chapman T, Mahalingam S, Ishak GE, Nixon JN, Siebert J, Dighe MK. Diagnostic imaging of posterior fossa anomalies in the fetus and neonate: part 2, posterior fossa disorders. Clinical imaging. 2015;39(2):167-75.
2. Zhang X, Gu Y, Sun X, Wang Y, Wang A. Dandy-Walker complex: a clinicopathologic study of 9 cases. Zhonghua bing li xue za zhi= Chinese journal of pathology. 2013;42(12):815-8.
3. Haddadi K, Zare A, Asadian L. Dandy-Walker Syndrome: A Review of New Diagnosis and Management in Children. Journal of Pediatrics Review. 2018;6(2):1-6.
4. Notaridis G, Ebbing K, Giannakopoulos P, Bouras C, Kövari E. Neuropathological analysis of an asymptomatic adult case with Dandy–Walker variant. Neuropathology and applied neurobiology. 2006;32(3):344-50.
5. Bokhari I, Rehman L, Hassan S, Hashim MS. Dandy-Walker malformation: a clinical and surgical outcome analysis. J Coll Physicians Surg Pak. 2015;25(6):431-3.
6. Pichiecchio A, Decio A, Di Perri C, Parazzini C, Rossi A, Signorini S. "Acquired" Dandy–Walker malformation and cerebellar hemorrhage: Usefulness of serial MRI. european journal of paediatric neurology. 2016;20(1):188-91.
7. Millen KJ, Gleeson JG. Cerebellar development and disease. Current opinion in neurobiology. 2008;18(1):12-9.
8. KhorsandZak H, Ehteshammanesh H, Mafinezhad Sh, Monemi A. A Rare Report of the Wolfram Syndrome Associated with the Dandy-Walker Syndrome. Journal of north Khorasan University of Medical Sciences. 2018;9(3):485-8.
9. Grinberg I, Northrup H, Ardinger H, Prasad C, Dobyns WB, Millen KJ. Heterozygous deletion of the linked genes ZIC1 and ZIC4 is involved in Dandy-Walker malformation. Nature genetics. 2004;36(10):1053.
10. Warf BC, Dewan M, Mugamba J. Management of Dandy-Walker complex–associated infant hydrocephalus by combined endoscopic third ventriculostomy and choroid plexus cauterization. Journal of Neurosurgery: Pediatrics. 2011;8(4):377-83.
11. Jurcă MC, Kozma K, Petcheşi CD, Bembea M, Pop OL, MuŢiu GA, Coroi MC, Jurcă AD, Dobjanschi L. Anatomic variants in Dandy–Walker complex. Romanian Journal of Morphology and Embryology. 2017;58(3):1051-5.
12. Khosravi N. Neonatal dandy-walker syndrome a case report. Tehran University Medical journal. 2002;9(28):39-44.
13. Forzano F, Mansour S, Ierullo A, Homfray T, Thilaganathan B. Posterior fossa malformation in fetuses: a report of 56 further cases and a review of the literature. Prenatal diagnosis. 2007;27(6):495-501.
14. Zeeshan QM HM, Hashmi SMM, Anwar MA. . Dandy-walker syndrome;clinical analysis and surgical outcome. Professional Med J. 2017; 24(1):47-50
15. Dandy-Walker Syndrome, DWS; Online Mendelian Inheritance in Man (OMIM).
16. Imataka G, Yamanouchi H, Arisaka O; Dandy-Walker syndrome and chromosomal abnormalities. Congenit Anom (Kyoto). 2007 Dec;47(4):113-8.
17. McClelland 3rd S, Ukwuoma OI, Lunos S, Okuyemi KS. The natural history of Dandy-Walker syndrome in the United States: A population-based analysis. Journal of neurosciences in rural practice. 2015;6(1):23.
18. Murray JC, Johnson JA, Bird TD. Dandy-Walker malformation: etiologic heterogeneity and empiric recurrence risks. Clinical genetics. 1985;28(4):272-83.
19. Prakash R, Singh LK, Kour J, Mishra B, Lakra A. Psychiatric comorbidities in Dandy-Walker variant disorder. The Journal of neuropsychiatry and clinical neurosciences. 2009;21(4):477-9.
20. Sinha P, Tarwani J, Kumar P, Garg A. Dandy–Walker variant with schizophrenia: Comorbidity or cerebellar cognitive affective syndrome? Indian journal of psychological medicine. 2017;39(2):188.
21. Nicholas Suraci, Partha Chatterjee. Meningitis Associated with Meningoencephalocele. Medical Science, 2016, 20(81), 164-167

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Dandy-Walker syndrome, vermis hypoplasia, hydrocephalus

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Conflict of interest
There are no conflicts of interest in this study.

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