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A rare case of dandy-walker syndrome with bilateral choanal atresia: a case report

Akram Hemmatipour[1], Maryam Nikbina[2], Zahra Safari Dehkohneh[3], Ali Hatami[4], Khadijeh Asadi Chahar Rah-e Gashin[5]

[1] Department of Nursing, Abadan University of Medical Sciences, Abadan, Iran
[2] Department of Midwifery, Shoushtar, Iran
[3] Student Research Committee, Shoushtar, Iran
[4] Department of Nursing, Shoushtar, Iran
[5] Department of Midwifery, Gachsaran Branch, Islamic Azad University, Gachsaran, Iran

SUMMARY

Introduction: Dandy-Walker syndrome (DWS) is a congenital neurological disorder that is characterized by a triad that includes complete or partial agenesis of the cerebellar vermis, torcular herophili, and so on. Choanal atresia is the obstruction or narrowing of the posterior nasal cavity. This disease may be congenital or acquired.

Methods: Considering the referral of the patient to Al-Hadi hospital in Shoushtar, the required data were collected through examination, tests, and documents in the medical records.

Results: The patient was a boy born at the 39th week of gestation via cesarean section because the ultrasound findings showed dilatation and enlargement of the fourth ventricle along with hydrocephalus. The patient was admitted with a diagnosis of DWS with bilateral choanal atresia, the absence of testis descent and the possibility of cardiac anomalies of hydrocephalus and macrocephaly with respiratory distress. The patient received Ampicillin, Dopamine, Fentanyl, and Midazolam based on the physician's order. After three days, the patient had a cardiac arrest, resuscitation was not successful, and the patient died eventually.

Conclusion: This syndrome can be diagnosed with advanced imaging techniques, so it is better to perfume routine pregnancy care, especially imaging measures, with more sensitivity to prevent the birth of babies with this abnormality.

Key words: dandy-walker syndrome, choanal atresia, hydrocephalus

Address for correspondence:

Khadijeh Asadi Chahar Rah-e Gashin, MSc Midwifery, Department of Midwifery, Gachsaran Branch, Islamic Azad University, Gachsaran, Iran, email: asadi11545@gmail.com

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INTRODUCTION

Dandy-Walker syndrome (DWS) is a rare congenital disorder with a prevalence of one case per 25,000-35,000 live births [1, 2]. This neurological syndrome is more common in girls than in boys [2, 3]. The syndrome is characterized by a triad that includes complete or partial agenesis of the cerebellar vermis [2, 4]. Cystic dilatation of the fourth ventricle, enlargement of the posterior fossa with an upward displacement of the tentorium, torcular herophili, and transverse dural sinuses are not always present. Although hydrocephalus is not diagnostic criteria for Dandy-Walker syndrome, its complications may increase intracranial pressure, and lead to gradual enlargement of the head, mental disability, and even death [1]. This syndrome is accountable for 4%-12% of all cases of congenital hydrocephalus [5]. The predisposing factors include environmental and genetic factors, contact with intrauterine teratogen, alcohol consumption, maternal infections (with viruses such as Rubella, Toxoplasmosis, and Cytomegalovirus) and diabetes [4-6]. Prenatal ultrasound can detect classic cases of this syndrome. Ultrasound may also be helpful in detecting hydrocephalus and enlargement of the ventricles. CT scan and MRI can be used to achieve a postnatal diagnosis, which shows the dilatation of the fourth ventricle and enlargement of the posterior fossa [7]. The outcomes of this syndrome are diverse and include natural or relatively natural development to disability and premature death [6]. Anomalies associated with this syndrome include polycystic kidney disease, cardiovascular defects, polydactyly, and cleft palate. According to studies, the prevalence of associated malformations was 50%-70% [5]. Chromosomal abnormalities are observed in 50% of patients with this syndrome, and most anomalies include trisomy 13 and 18, and triploidy. If Dandy-Walker syndrome is associated with single-gene defects, the risk of relapse in subsequent pregnancies will increase and will be higher than 5% [4].

There is no consensus on optimal surgical treatment for Dandy-Walker syndrome. The initial surgery on the posterior fossa and the removal of the membrane is a preferred method for managing this syndrome, especially in older children [4-8]. Other methods include ventriculoperitoneal shunt, shunt with y-connector, puncture of the lateral and third ventricles under ultrasound guidance and piercing the ventricle [8]. Khosravi reported the

case of a girl who was born at the 39th week of gestation based on LMP and 35th week based on ultrasound via cesarean section due to bradycardia. The patient was a firstborn, and the first- and fifth-minute Apgar scores were seven and nine, respectively. Sizes were below 10th percentile, and asymmetrical IUGR was detected. The examination of the organs was normal and the patient was normocephalic. In prenatal ultrasound, moderate hydrocephalus was found involving about 50% of brain ventricles and dominance of the third ventricle along with cystic dilatation of the posterior fossa and lack of cerebellar vermis. Postpartum CT scan revealed cerebellar vermis hypoplasia and cystic dilatation of the fourth ventricle, hydrocephalus, and developmental impairment in the grey matter and white matter, which led to Dandy-Walker syndrome. After consulting a neurosurgeon, the baby was discharged with a good general condition after 15 days of hospitalization due to a lack of hydrocephalus and the absence of intracranial pressure and was scheduled to be monitored regularly [3].

Rahmani et al. reported a case of a male fetus suffering from Dandy-Walker syndrome whose mother was a 38-year-old primiparous woman in her second pregnancy, who referred for fetal ultrasound. In the nomograms of the fetus and based on skull analysis, inferior vermian hypoplasia, as well as a suspicious relationship between the posterior fossa and the fourth ventricle, were observed, which suggested Dandy-Walker syndrome. A mother underwent a therapeutic abortion at a gestational age of 19 weeks and 2 days [1]. The aim of this study was to report a case of a newborn with Dandy-Walker syndrome.

CASE REPORT

The baby was a boy born on October 24, 2018 at Al-Hadi hospital in Shushtar. Based on ultrasound and LMP, the baby was born at the 39th week of gestation via cesarean section because the ultrasound findings showed dilatation and enlargement of the fourth ventricle along with hydrocephalus. The first- and fifth-minute Apgar scores were two and four, respectively. The baby experienced apnea immediately, but his breathing improved after resuscitation. The mother was a 24-year-old healthy woman and her baby was a secondborn. Mother was gravida 2 and had no history of abortion. The first child was a 4-year-old girl and had no genetic or hereditary disorder. The parents were healthy and had a close family relationship. The mother was healthy during pregnancy and did not take any kind of medication before pregnancy or during pregnancy. After birth, the baby was transferred to the NICU of Al-Hadi hospital after resuscitation and breathing through a bag valve mask. In the initial examination, the baby's weight was 1800 g, his height was 41 cm, and head circumference was 32 cm, all of which were below the 10th percentile. Therefore, the baby suffered from Intrauterine Growth Retardation (IUGR).

The baby's temperature was 36.5°C and the heart rate was about 40 bpm. Due to the lack of an increase in heart rate after giving oxygen through a bag valve mask, the patient was immediately intubated. The breathing with a bag valve mask was recorded at 60 to 80. In baby had systolic heart murmur in auscultation, and lung crackles could also be heard. The patient did not have organomegaly, but cardiomegaly was reported in a chest X-ray. The patient was admitted to the NICU with the diagnosis of Dandy-Walker syndrome with bilateral choanal atresia, the absence of testis descent and the possibility of cardiac anomalies of hydrocephalus and macrocephaly with respiratory distress. After the admission of the baby to the NICU, the patient was examined for septicemia. In the tests, sugar was 69 and calcium was 9.4 mg/dl. Other tests such as urea and serum electrolytes, blood culture, and CSF analysis were negative. During the period of hospitalization, the patient received Ampicillin, Gentamicin, Dopamine, Fentanyl, Ranitidine, and Midazolam along with 10% D/W saline based on the physician's order.

The patient had a cardiac arrest on October 27, 2018, resuscitation was performed for 30 seconds but was not successful, and the patient died eventually (Figure 1).

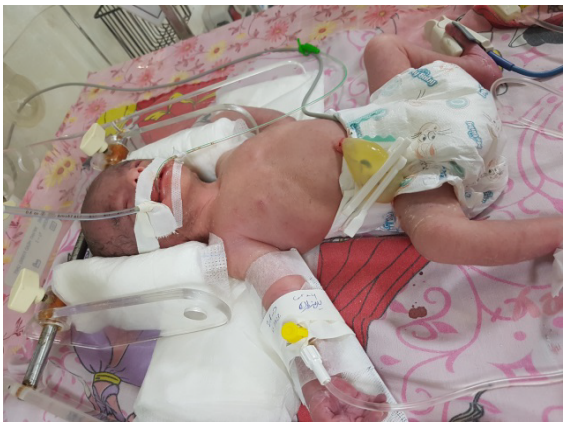

Fig. 1. Baby diagnosed with Dandy-Walker syndrome with bilateral choanal atresia

DISCUSSION

Dandy and Blackfan described Dandy-Walker syndrome in 1914 for the first time. The most common anomaly in this syndrome is dilatation of the fourth ventricle, which is in the form of a cyst and its roof is covered by neuro-glial-vascular membrane [3]. Almost 90% of these patients have hydrocephalus [9]. The patient of this study was diagnosed with dilatation and enlargement of the fourth ventricle along with hydrocephalus.

Ventriculoperitoneal shunt placement or Endoscopic Third Ventriculostomy (ETV) can be performed to treat hydrocephalus. In a study by Khoshnevisan et al. on ans eight-year-old girl with symptoms of Dandy-Walker syndrome accompanied by hydrocephalus, the patient underwent shunt placement went she was five, but she was treated with EVT due to increased pressure on the brain and since the symptoms of the patient did not stop, shunt revision was performed. The patient was discharged with partial recovery [10]. During 1997 to 2003, the Kids' Inpatient Database (KID) predicted the overall prevalence of Dandy-Walker syndrome to be 1.36 per 1,000 children [11]. This syndrome can be diagnosed before birth using fetal ultrasound at 18th week of pregnancy. This abnormality can also be diagnosed after birth using a CT scan and MRI if the fourth ventricular dilatation is observed and the posterior fossa cyst is enlarged [7]. In this study, Dandy-Walker syndrome was diagnosed using ultrasound in the eighth month of gestation.

The study of Maurits et al. demonstrated the effect of specialized multidisciplinary care on the treatment of a case of Complex Post-Traumatic Stress Disorder (C-PTSD) with psychotic disorder accompanied by severe neurological failure (Dandy-Walker syndrome and hydrocephalus) [12]. In a study by Salehi et al. on a female newborn, the patient suffered from Apert syndrome with Dandy-Walker deformity, Agenesis of the corpus Callosum (ACC) and pneumothorax [13]. In their study, Zack et al. reported a rare case of Wolfram syndrome associated with Dandy-Walker syndrome [2]. In some of the studies, it has showed that FOXC1 mutations are associated with Axenfeld-Rieger syndrome and Dandy-Walker syndrome, and these diseases can be highly associated [10-14]. In addition to Dandy-Walker syndrome, the patient of the present study suffered from bilateral choanal atresia, the absence of testis descent, cardiac anomalies, and lung crackles. Choanal atresia is the obstruction or narrowing of the posterior nasal cavity. This disease may be congenital or acquired [15]. This abnormality is diagnosed using the catheter 6 and its placement in the nose and the injection of contrast agent and performing cross-table lateral radiography [16]. Choanal atresia can be unilateral or bilateral; bilateral cases are more serious and require emergency treatment [15]. In bilateral choanal atresia, the baby shows signs of airway obstruction at birth. Symptoms are in the form of acute respiratory distress and cyanosis, which disappear when the baby cries and therefore is referred to as paradoxical cyanosis [3].

CONCLUSION

The baby in this study was diagnosed with respiratory distress and cyanosis due to bilateral choanal atresia and cardiac anomaly immediately after birth, and breathed through a bag valve mask, and was intubated after hospitalization.

DECLARATION

The authors state that they have no conflict of interest.

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