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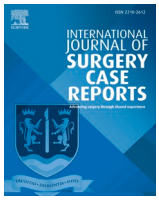

Case report

Dandy-Walker syndrome with bilateral choanal atresia: A case report

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ABSTRACT

Introduction: Dandy-Walker syndrome is a rare congenital brain malformation characterized by cerebellar vermis agenesis or hypoplasia, cystic dilatation of the fourth ventricle and a large posterior fossa causing upward displacement of tentorium and torcula. In this paper, we present a case of bilateral choanal atresia with Dandy-Walker Syndrome in a female newborn.

Case presentation: We present a case of a female patient who was born at 38th weeks of gestation via emergency cesarean section due to non-reassuring cardiotocography and abnormal antenatal ultrasounds findings. The imaging revealed the presence of Dandy-walker malformation. The patient presented with cyanosis and respiratory distress. Bedside flexible nasoendoscopy revealed bilateral choanal atresia which is confirmed by computed tomography of sinuses. Endoscopic bilateral choanal atresia repair was performed. On postoperative follow up, nasal endoscopy showed bilateral intact flap and patent neochoana.

Discussion and conclusion: Dandy-Walker syndrome is a congenital disorder that can be diagnosed prenatally. The syndrome is associated with multiple anomalies. However, there are few published reports of bilateral choanal atresia in Dandy-Walker Syndrome. Bilateral choanal atresia is considered a life-threatening condition in newborns that requires early surgical intervention.

1. Introduction

Dandy-Walker syndrome is a congenital brain malformation characterized by cerebellar vermis agenesis or hypoplasia, cystic dilatation of the fourth ventricle and a large posterior fossa causing upward displacement of tentorium and torcula [1]. Dandy-Walker syndrome was first described by Dandy and Blackfan in 1914 followed by Tagart and Walker in 1942 and was labeled as Dandy-Walker syndrome by Benda in 1954 [2,3]. The estimated prevalence of Dandy-Walker syndrome is approximately 1:30,000 live births with female preponderance [4]. The syndrome can be associated with other neurological anomalies that are not part of the criteria, such as agenesis of corpus callosum, holoprosencephaly, and occipital encephaloceles. Dandy-Walker syndrome is responsible for about 4–12% of congenital hydrocephalus cases [4]. Extra-cranial anomalies associated with Dandy-Walker syndrome, including cardiovascular anomalies, cleft palate, polydactyly and malformation in gastrointestinal or genitourinary system with incidence of 50–70% [5]. Prenatal diagnosis can possibly be done by using antenatal ultrasonography which can demonstrate the central nervous system abnormalities [5]. Imaging study of the brain can be used to confirm the postnatal diagnosis [5]. In addition, Dandy-Walker Syndrome can be rarely associated with bilateral choanal atresia. Bilateral choanal atresia is a serious airway condition in neonate that warrants early surgical intervention. This case report presented a case of Dandy-Walker syndrome with bilateral choanal atresia that was treated surgically.

This case has been reported in line with the SCARE 2020 criteria [6].

2. Case presentation

A female patient was born at 38th weeks of gestation via emergency cesarean section due to non-reassuring cardiotocography. Antenatal ultrasounds showed evidence of bilateral dilated ventricles and small choroid plexus. Fetal magnetic resonance imaging (MRI) revealed the presence of Dandy-walker malformation with intraventricular hemorrhage grade 1. The patient was born with weak cry, cyanosis and respiratory distress, Apgar score was six in the first minute and eight in the fifth minute. Suction catheter could not pass the nasal cavity during the resuscitation, so she was intubated and then admitted to neonatal

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intensive care unit (NICU). Parents are first degree cousins and healthy with unremarkable family history. On examination, the patient has micrognathia and right foot club. Echocardiography study was unremarkable for any cardiac anomalies.

Brain MRI was done in her first day of life showed features of Dandy-walker malformation associated with the left-sided intraventricular hemorrhage and enlarged supratentorial ventricular system without feature of acute hydrocephalus and encephaloceles. Three days post-delivery, the head circumference of the patient started to increase with some episodes of bradycardia. Brain computed tomography (CT) was done and reported interval enlargement of supratentorial ventricular system with sign of active hydrocephalus, interval development of right intraventricular hemorrhage and bilateral choanal atresia (Fig. 1). The patient underwent right frontal ventriculoperitoneal shunt insertion on the 8th day after birth.

Pediatric otolaryngology team was consulted due to the inability to pass nasogastric tube bilaterally and bilateral choanal atresia on brain CT. Sinus CT was done and confirmed the presence of bilateral mixed choanal atresia (Fig. 2). Endoscopic bilateral choanal atresia repair was performed on day 22 post-delivery. The procedure started by examining the nasal cavity which showed bilateral choanal atresia. The endoscopic choanal atresia repair was done by nasal septal cross-over flap technique utilizing otology set as described by Stamm AC [7]. This technique involves L shape incision on posterior septum about 0.8 cm anterior from atretic plate to raise superolateral base flap in the right side. In other side, inverted L shape incision with raise of inferolateral flap in the left side. The posterior septum, bilateral atretic plate and part of medial pterygoid were removed by combination of different instrument. The superolateral base flap used to cover roof of neochoane while inferolateral flap used to cover floor of neochoane (Fig. 3). Then bilateral nasal packing inserted, and pack use to advance more to neochoane to keep both flap in place. The patient was stable and tolerated the procedure very well and then shifted to the NICU intubated. Nasal packs were removed on day four postoperative, irrigation with normal saline and suction was done from both nostrils as well. The patient was started on mometasone and anti-reflux medication for 6 weeks. The patient was reviewed on day eleven postoperative for follow up and nasal debridement, showing improvement and nasal endoscopy showed bilateral intact flap and patent choana (Fig. 4).

3. Discussion

Dandy-Walker syndrome is a rare autosomal dominant inherited disorder of the central nervous system with a prevalence of one case per 30,000 live births [8,1]. Most of Dandy-Walker syndrome cases were reported in females. Similarly, the presented case was a 38-week gestation baby girl. Dandy-Walker syndrome can be prenatally diagnosed by ultrasound after the 18th week of gestation as the cerebellar vermis will be completely developed [9]. MRI can be used as well after the 20th week of gestation to evaluate any associated neurological anomalies [9]. Dandy-Walker syndrome in this case was diagnosed antenatally by ultrasound in week 38th gestation and was confirmed by fetal MRI.

Several predisposing factors contributed to the etiopathology of Dandy-Walker syndrome. Some cases may result from genetic factors such as chromosomal aneuploidy or Mendelian disorders [10]. Environmental factors, including alcohol, diabetes, and maternal infections

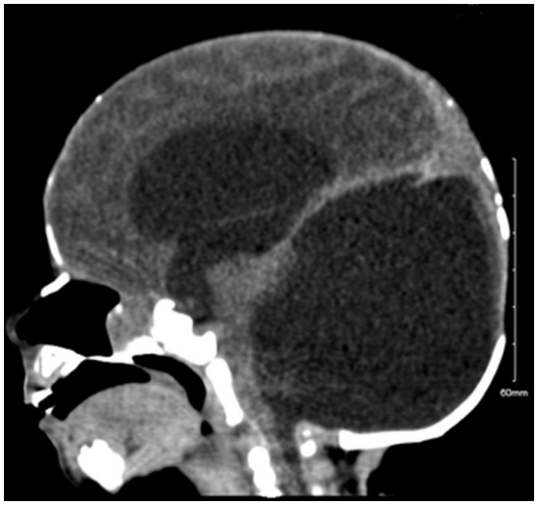

Fig. 1. A brain CT scan sagittal view showing an interval enlargement of the supratentorial ventricular system with signs of active hydrocephalus, interval development of right intraventricular hemorrhage and bilateral choanal atresia.

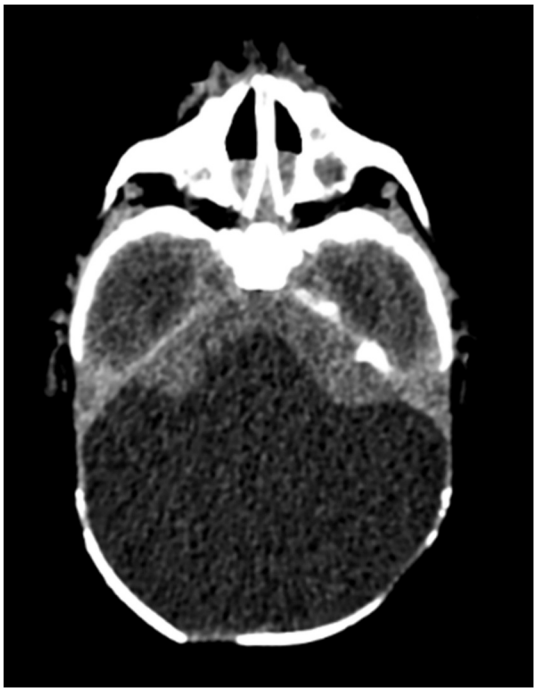

Fig. 2. A sinus CT scan axial view showing bilateral mixed choanal atresia.

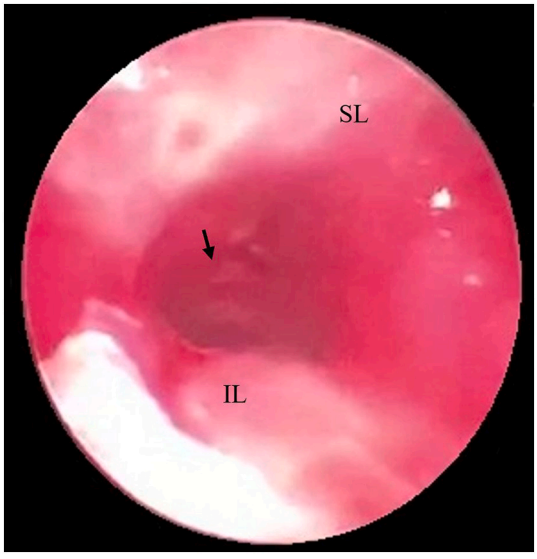

Fig. 3. Intraoperative endoscopic view of adequate neochoane opening (arrow) with superolateral flap (SL) and inferolateral flap (IL).

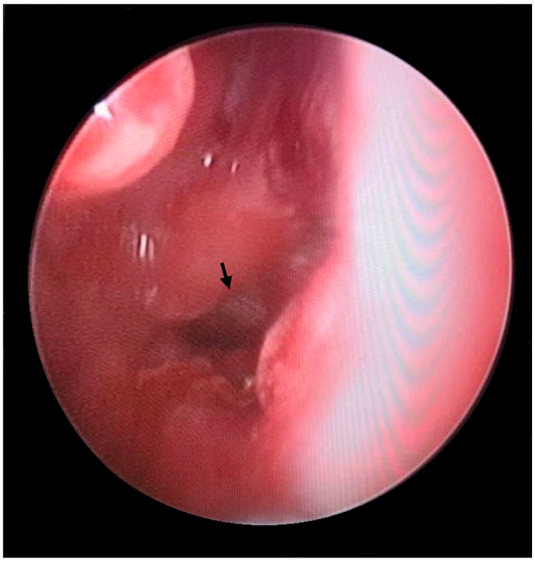

Fig. 4. Endoscopic view on day eleven postoperative after debridement showing intact superolateral and inferolateral flap with patent neo-choane (arrow).

[8]. The parents of our patient are healthy with no history of brain anomalies or similar presentation in the family.

Choanal atresia is a congenital malformation characterized by narrowing or obliteration of the nasal choana [11]. It is a relatively uncommon with an incidence of one case in 5000–7000 live births [12]. The nasal obstruction in choanal atresia can be either unilateral or bilateral [11]. Bilateral choanal atresia considered to be lethal in neonates and require immediate management, since they are obligate nasal breathers [11]. Choanal atresia is a common component of some congenital disorders such as CHARGE, Treacher Collins, Crouzon and Pfeiffer syndromes [13]. However, the presence of bilateral choanal atresia in Dandy-Walker syndrome is rarely reported and published in the literature. Hemmatipour reported a case of a male patient who was born at the 39th week of gestation via cesarean section because of the abnormal ultrasound findings. The patient was diagnosed with Dandy-Walker syndrome with bilateral choanal atresia and other associated anomalies. The patient admitted to NICU and eventually died due to cardiac arrest [14]. Our patient was born with signs of airway obstruction and CT confirmed bilateral choanal atresia. The patient underwent endoscopic bilateral choanal atresia repair successfully.

Cardiovascular anomalies, polycystic kidneys, cleft palate, and polydactyly are malformation frequently associated with Dandy-Walker syndrome. The presence of these extra-CNS anomalies is directly associated with fetal mortality [15]. In the presented case, the patient had an echocardiography study that showed no evidence of cardiac anomalies. Hydrocephalus has a major role in the neurological outcome of the disease and the progressive of the symptoms. Ventriculoperitoneal (VP), cystoperitoneal (CP) shunts and endoscopic third ventriculostomy (ETV) are surgical options used to treat hydrocephalus [16]. Controlling hydrocephalus with shunting has been found to improve mortality [17]. The patient in this case had active hydrocephalus which was controlled with right frontal ventriculoperitoneal shunt insertion.

4. Conclusion

Dandy-Walker syndrome is a rare congenital malformation that can be diagnosed prenatally. The syndrome is associated with multiple anomalies. However, there are few published reports of bilateral choanal atresia in Dandy-Walker Syndrome. Bilateral choanal atresia is considered a serious condition in newborns that require early surgical intervention.

Sources of funding

None.

Ethical approval

Not needed, as this case report does not require IRB approval.

Consent

Written informed consent was obtained from the patient's parent for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Research registration

Not applicable.

Guarantor

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CRediT authorship contribution statement

First author: Review of chart, literature review and drafting the case report.

Second author contributed to manuscript concept, critical reviewing and editing of the manuscript.

Senior author contributed to manuscript concept, critical reviewing and editing of the manuscript.

Declaration of competing interest

The authors have no conflict of interest related to this case.

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