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Case report

Dandy-Walker malformation associated with subarachnoid hemorrhage. A case report

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ABSTRACT

Introduction: Dandy-Walker malformation is a rare congenital brain defect characterized by vermian agenesia with cystic dilatation of the fourth ventricle, and posterior fossa enlargement. The etiology is still poorly understood but is presupposed to be multifactorial, infrequently caused by intracranial hemorrhage. We describe a case of male newborn known to have Dandy-Walker malformation associated with subarachnoid bleeding after the delivery, which is a quiet rare presentation only discussed in a few literatures before.

Case presentation: We present a rare case of a full-term male baby delivered vaginally, who was diagnosed with Dandy-Walker malformation during antenatal anomaly scan. At birth, the baby presented with a weak cry, cyanosis, respiratory distress and seizure. Post-delivery computed tomography scan revealed subarachnoid hemorrhage. In addition, a hydrocephalus was noted on the imaging and treated with ventriculoperitoneal shunt insertion with marked improvement of the posterior fossa cyst and the hydrocephalus as an outcome of early intervention.

Discussion: Few literature studies showed an association between intracranial bleeding during early fetal life and the development of Dandy-Walker malformation as it affects the posterior fossa components growth. However, our case highlights on an unusual presentation of the spontaneous subarachnoid hemorrhage after the delivery in a full-term baby diagnosed with Dandy-Walker malformation earlier.

Conclusion: This report highlights the importance of early recognition and implementing appropriate management of the hydrocephalus that associated with intracerebral bleeding to prevent the complications of high intracranial pressure plus brainstem herniation and achieve the best possible outcome.

1. Introduction

Dandy-Walker malformation (DWM) is a congenital brain abnormality characterized by cerebellar vermis hypoplasia, posterior fossa enlargement with lateral sinuses, tentorium, torcula displacement, and cystic dilatation of the fourth ventricle. In 1914, Dandy and Blackfan described this anomaly for the first time [1]. Up to 80 % of the patients will have a hydrocephalus as a complication of the malformation and as a consequence of the inadequate communication between the subarachnoid space and the enlarged fourth ventricle [2]. Numerous chromosomal abnormalities as well as teratogens exposure during the antenatal period have all been linked to the development of the DWM [3]. In children, the DWM frequently manifests as hydrocephalus, nausea, vomiting, intellectual impairment, and cerebellar signs [1]. The diagnosis of DWM is made by prenatal sonography; however, the definitive diagnosis is challenging before 18 weeks of gestation [4]. Treatment options and prognosis are based on the severity and existence of other abnormalities [5].

Several literature studies have described cerebellar hemorrhage and intraventricular hemorrhage in preterm neonates with DWM. In contrast, intracranial hemorrhage is rarely observed in full-term neonates who were delivered vaginally. Hence, we present a case of subarachnoid hemorrhage (SAH) in a term baby diagnosed antenatally as a Dandy-Walker malformation in the absence of any perinatal or postnatal external trauma. We reported this case according to the Surgical Case Reports (SCARE) guidelines [6].

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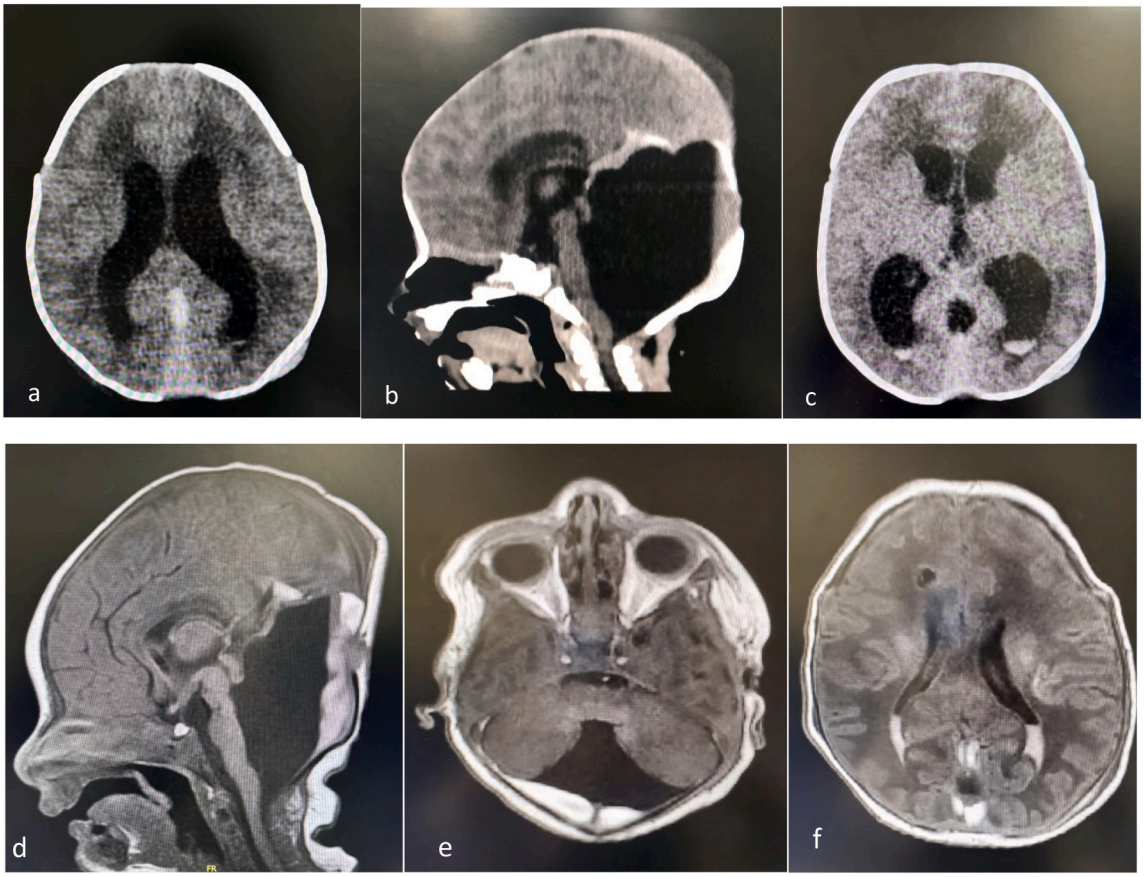

Fig. 1. A non-contrast CT study obtained on the 3rd day of life showed findings consistent with DWM. Hydrocephalus was noted in axial view (a). SAH in the dependent part of the large cystic lesion in the posterior fossa area in sagittal view (b). Intraventricular hemorrhage in the axial view (c). On the 7th day of life brain MRI revealed no signs of hydrocephalus. Sagittal view (d), Axial view (e) Fluid-fluid leveling representing acute hemorrhage involving occipital horns of lateral ventricles bilaterally (f).

2. Case report

A newborn male baby was delivered vaginally at the expected gestational age at Almadinah Almunawara in Saudi Arabia to young and healthy parents who are consanguineous cousins, and this baby is their second child, with an older healthy sibling. During the antenatal period of the second trimester, ultrasound revealed the presence of abnormal posterior fossa likely DWM with wide spacing of cerebellar hemispheres and agenesis of cerebellar vermis. At birth, the baby weighed 3.2 kg, with a weak cry, cyanosis, respiratory distress, and meconium-stained liquor. Apgar score was four in the first minute and he required a positive pressure ventilation for 1 min. After the admission to the neonatal intensive care unit (NICU), the baby developed one episode of seizure that lasted for seconds and resolved spontaneously involving the right side of the body. He was started on levetiracetam 20 mg/kg with a maintenance dose of 20 mg/Kg divided in 12 h. Furthermore, there were other abnormalities observed such as respiratory distress, retrognathia, retinal hemorrhage, narrow palpebral fissure, laryngomalacia, and mild tricuspid regurgitation. A neonatal head ultrasound performed on the second day of the baby's life confirmed the diagnosis of DWM.

On the 3rd day of life, the head circumference increased to 36 cm with enlargement and bulging of fontanelles. A non-contrast computed tomography (CT) showed a huge cystic lesion in the posterior fossa in communication with the fourth ventricle, high-riding tentorium, and torcula-lambdoid reversals, that correlate with DWM. In addition, there was evidence of SAH in the dependent part of the large posterior fossa cystic area, with active hydrocephalus. Moreover, a subdural hemorrhage and intraventricular hemorrhage involving the dependent part of occipital horns of lateral ventricles bilaterally were noted [Fig. 1].

Table 1
CSF findings on the 4th day after delivery consistent with subarachnoid hemorrhage.

Exam name	Normal	Findings
Appearance	Clear	Slightly cloudy
Color	Normal	Xanthrochromia
WBC	0–5 × 10^6/L	0
RBC	0–10 × 10^6/L	25,250 × 10^6/L
Monocyte	15–45 %	6 %
Lymphocyte	40–80 %	29 %

Four days after delivery, the baby underwent ventriculoscopy and during the observation by endoscope there was a hemorrhage, so an external ventricular drain (EVD) was inserted and joined to the general external ventricular drain system with pressure adjusted to the 10 cm level. Cerebrospinal fluid (CSF) sample were taken and showed a xanthrochromic appearance and a high red blood cell count of 25,250 × 106/L secondary to the presence of subarachnoid hemorrhage [Table 1].

Seven days post-delivery, a brain magnetic resonance imaging (MRI) confirmed the location of the EVD tip on the left side of the suprasellar cistern. No signs of hydrocephalus with no improvement in the intraventricular and subarachnoid hemorrhage [Fig. 1]. The baby underwent right ventriculoperitoneal (VP) shunt insertion on the 10th day of age and EVD removal after CSF was cleared. The non-contrast CT findings after the surgery revealed the ventricular catheter at the right frontal horn of the lateral ventricle, with a significant reduction of ventricular size and posterior fossa cyst which communicates with the fourth

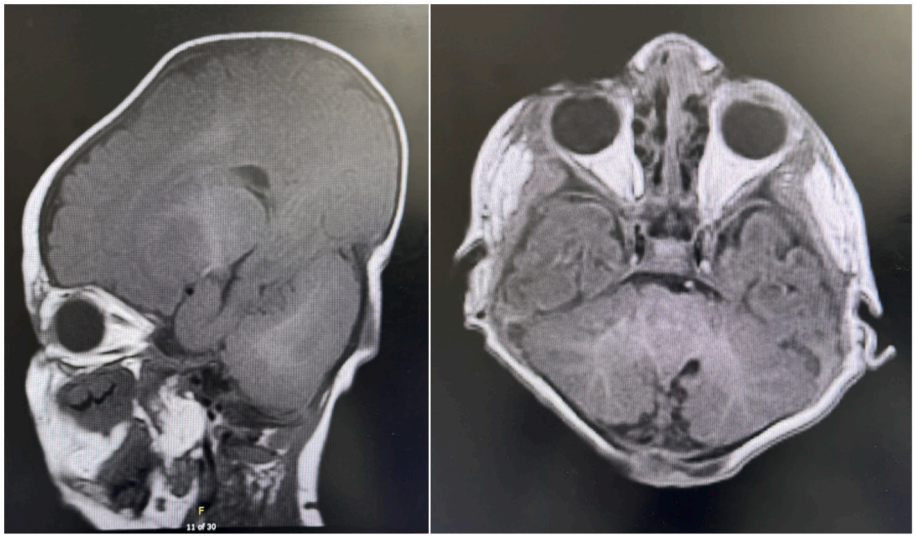

Fig. 2. A contrast MRI of the brain was done after four months of VP shunt insertion and revealed a significant decrease of the posterior fossa cyst plus absent signs of the hydrocephalus with adequate VP shunt functioning.

ventricle. Although, Marked resolution in both subarachnoid and intraventricular hemorrhage.

The patient was then transported to NICU in stable condition and reviewed four months postoperatively for follow-up and MRI of the brain with contrast was done showed a significant decrease of the posterior fossa cyst plus absent signs of the hydrocephalus with adequate VP shunt functioning [Fig. 2].

3. Discussion

DWM was hypothesized to be caused by Luschka and Magendie foramina atresia, as a result will lead to fourth ventricle dilatation and hypoplasia of cerebellar vermian. It's considered the most common posterior fossa malformation [7]. A small female predominance was reported in the prevalence which is contrary to our case as he is a male newborn [8]. The exact mechanisms of DWM are poorly understood, but its seen to be more associated with genetic defects, chromosomal abnormalities, and congenital infections as in fact all can contribute in the development of DWM. Similarly, our patient is a child of consanguineous cousins and this is supported by Recep Has et al. [9], where their data revealed a high prevalence of DWM among related parents.

The clinical presentation of DWM is commonly associated with structural abnormalities in the cerebellum, as well as the presence of hydrocephalus and intracranial hypertension [10]. As described, our patient had hydrocephalus that was observed since the 3rd day of life as the measured head circumference was increasing, moreover, the seizure development after the admission to NICU in the absence of metabolic causes is more related to the brain pathologies.

In our case, the patient was diagnosed with DWM through ultrasound during the second trimester anomaly scan, and confirmation of the diagnosis was made by neonatal head ultrasound scan after the delivery, similar to what was discussed in previous literatures. However, MRI is the method of choice for diagnosing DWM and distinguishing it from other central nervous system defects occupying the posterior fossa [11].

SAH occurs when the veins in the subarachnoid villi tear, leading to blood accumulation in the space surrounding the brain that can lead to hydrocephalus [12]. Intracranial hemorrhage in form subarachnoid, interventricular, sublegal hemorrhage are commonly observed in preterm newborn, who delivered by vacuum-assisted or forceps instruments which, is not supported by our described case as the patient is a full term baby and delivered through normal spontaneous vaginal delivery [13]. Few studies showed an association between the intracranial hemorrhage during early fetal life and the development of Dandy-Walker malformation as it affects the posterior fossa components growth, in contrast to our case who suffered from the subarachnoid and interventricular hemorrhage after the delivery where the brain almost completely well-developed [14,15].

Surgery is the standard treatment for our patient for his both active issues, DWM and subarachnoid hemorrhage. In the past, cyst fenestration and posterior fossa decompression were frequently used in the management of DWM, but they had limitations in restoring normal CSF circulation. As a result, VP and cystoperitoneal (CP) shunting have become extensively utilized [16]. One literature study, observed the reduction in ventricle size on postoperative images occurred more frequently in patients with a VP shunt, whereas a reduction in cyst size was more appreciable in patients with a CP shunt [17]. Our management approach for this patient, was divided into two stages: The first part was directed toward the temporary controller of the high ICP, and draining the bloody CSF by inserting an external ventricular drain. Once the CSF was crystal clear the hydrocephalus was managed by right ventriculoperitoneal shunt insertion.

4. Conclusion

DWM is a rare congenital abnormality that affects the development of the cerebellum and fourth ventricle. In an unusual case, we observed a rare association between DWM and non-traumatic postnatal subarachnoid and intraventricular hemorrhage. This report highlights the importance of early recognition and implementing appropriate management of the hydrocephalus that associated with intracerebral bleeding to prevent the complications of high intracranial pressure plus brainstem herniation and achieve the best possible outcome.

Ethical approval

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

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CRediT authorship contribution statement

First author: The author contributed in collecting the data of the

patient to prepare and write the case, drafting and editing of the manuscript.

Second author: The Paediatric physician responsible for this case, involved also in the review of the manuscript.

Third author: Main treating physician contributed in the supervising of the writing, critical reviewing, editing of the manuscript before submission.

Guarantor

Dr. Bassem Y Sheikh.

Registration of research studies

1. Name of the registry: None.
2. Unique identifying number or registration ID: None.
3. Hyperlink to your specific registration (must be publicly accessible and will be checked): None.

Consent

Written informed consent was obtained from the patients' legal guardian for publication of this case report and accompanying radiological images. A copy of the written consent is available for review by the Editorin-Chief of this journal on request.

Declaration of competing interest

There are no conflicts of interest.

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