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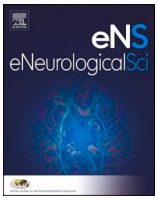

Case report

Dandy-Walker syndrome associated with a giant occipital meningocele: A case report and a literature review

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ABSTRACT

Background: Dandy-Walker malformation or syndrome is the most common posterior fossa malformation. It is commonly associated with other congenital anomalies such as cardiac defects; however, association with a giant occipital meningocele is extremely rare, as only around 34 cases have been described.
Case description: We report a case of a 2-month-old female infant who presented with a midline, gigantic mass in the back of the head. It was first discovered on a prenatal ultrasound. The mass measured about 15 × 5 cm, extending to the midback, not changing in size with crying, not attached to the back, and with a positive transillumination test. The diagnosis was confirmed after doing a brain computed tomography, which revealed hypoplasia of the vermis with an enlarged posterior fossa as well as cystic dilation of both ventricles with herniation through a bone defect.
Conclusion: Our case highlights a rare association between giant occipital meningocele and Dandy-Walker syndrome that is rarely discussed or reported in the medical literature. By reporting this extremely rare case of Dandy-Walker syndrome associated with a giant occipital meningocele, we hope to contribute to the creation of a database for future research so that a management protocol can be established for use by clinicians and neurosurgeons for better management of the condition.

1. Introduction

"Dandy-Walker malformation or syndrome is the most common posterior fossa malformation characterized by agenesis or hypoplasia of the vermis and cystic enlargement of the fourth ventricle, causing upward displacement of the tentorium and torcula (14)." According to the size of the meningocele sac, Talamonti et al. divided occipital meningocele into the following three categories in their study published in 2011: The diameters of the small, big, and giant varieties were as follows: up to 5 cm for the small, between 5 and 9 cm for the large, and over 9 cm for the giant (13). Dandy–Walker syndrome has an incidence of one in 30,000 births, and it is commonly associated with other congenital anomalies such as cardiac defects; however, association with a giant occipital meningocele is extremely rare, as only about 34 cases have been described (3). As a result, it was challenging to draw any clear conclusions about the behavior and management of the co-occurrence of these two conditions without more research. We report this extremely rare case of Dandy-Walker syndrome associated with a giant occipital meningocele. To the best of our knowledge, this is the first reported case in Iraq.

2. Case presentation

A 2-month-old female infant presented with a midline, gigantic mass in the back of the head. She had been delivered by cesarean section uneventfully as a fully term neonate of consanguineous parents, due to the presence of a cystic mass that had been shown by ultrasound prenatally. The mother received good prenatal care, and she regularly took folic acid and other supplements. No family history of a similar occurrence was noted. Two months after delivery, the patient was doing well and was presented to the neurosurgery department for a consultation. The physical and neurological examination revealed normal-range vital

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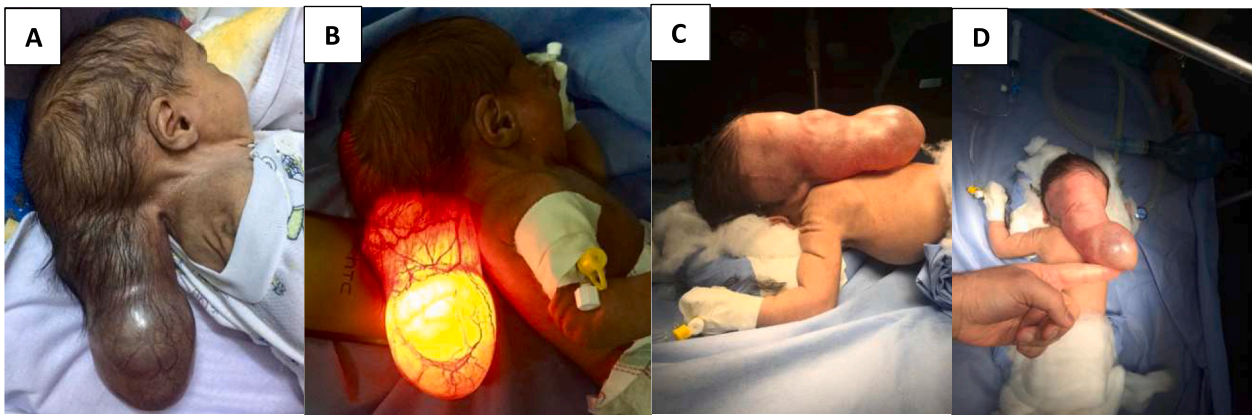

Fig. 1. : (A, B, C, D) showing multiple views of a large, completely skin-covered, irregular mass in the occipital region extending to the midback with positive transillumination as shown in (B).

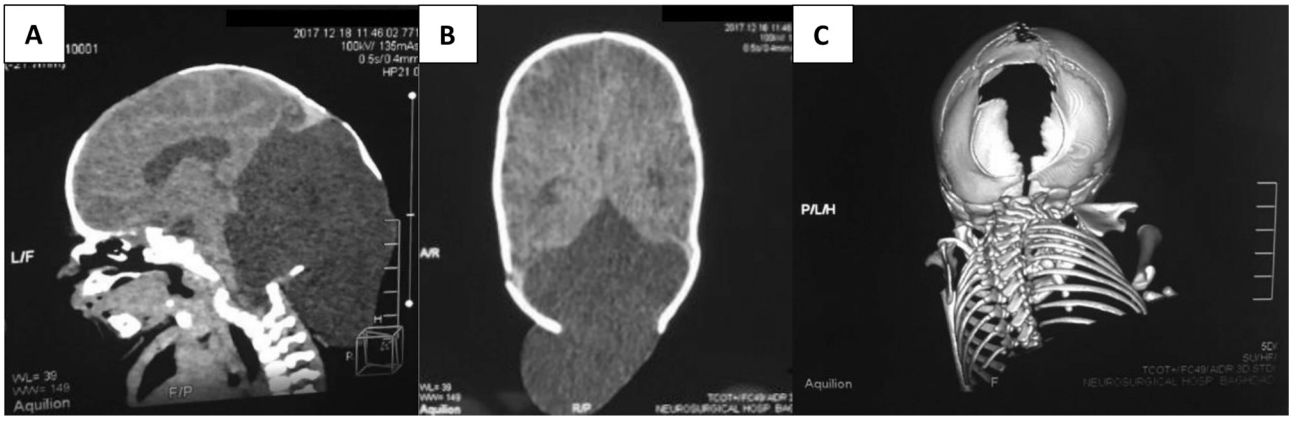

Fig. 2. : (A, B) sagittal and axial CT scan showing hypoplasia of the vermis with enlarged posterior fossa as cystic dilation of both ventricles with herniation through a bone defect (C) sagittal suture diastasis.

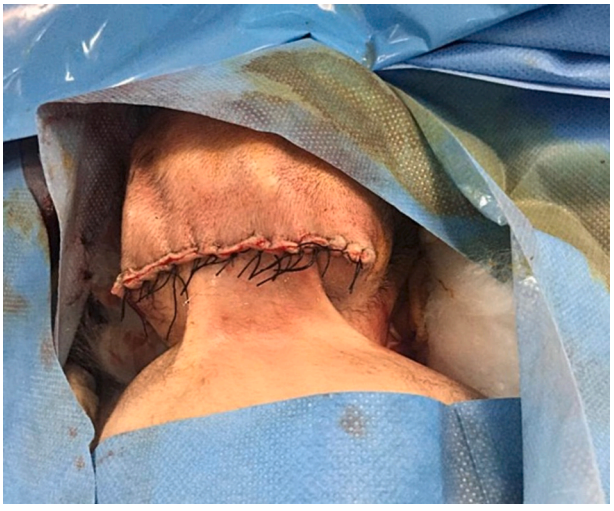

Fig. 3. : The occipital region of the head postoperatively after repair of the defect.

signs and no apparent neurological deficits. The presence of a giant, completely skin-covered, irregular cystic mass in the occipital region measured about 15 × 5 cm, extending to the midback, not changing in size with crying, and not attached to the back (as shown in Fig. 1). The transillumination test was positive (as shown in Fig. 1B). There was obvious microcephaly, as the head circumference was 30 cm with an open anterior fontanelle. After a brain computed tomography revealed hypoplasia of the vermis with an enlarged posterior fossa as well as cystic dilation of both ventricles with herniation through a bone defect (as shown in Fig. 2), the diagnosis of Dandy-Walker syndrome with a giant occipital meningocele was confirmed, and the decision was made to perform an elective surgery under general anesthesia to remove this excessive sac. The procedure includes a transverse incision, sac excision, and repair of the dura with a watertight dural closure using vicryl suture 4/0 (as shown in Fig. 3). 45 days later, the patient presented with hydrocephalus as a complication of the procedure and underwent another operation for a VP shunt insertion with medium-low pressure. The procedure was well tolerated by the infant; she did not require admission to a neonatal intensive care unit and was discharged home two days later, afebrile, and well-nourished on breast milk. Later on, after two years of regular follow-up visits, the signs of the cerebellum became obvious: nystagmus and tremors with epilepsy, and there were no signs of increased intracranial pressure.

3. Discussion

DWM is a congenital defect that affects the cerebellum and a few of its other structures, including the cerebellar vermis, fourth ventricle, and the posterior fossa (12). Meninges are herniated in cases of cranial meningocele. On the other hand, meninges and neural tissue can herniate into an encephalocele. Both of these birth defects fall under the term “cephalocele.” Meningocele typically appears around the sixth or

seventh week of intrauterine development, and its relationship to DWS is considered a rare occurrence (6). The indexed case was confirmed to have Dandy-Walker syndrome with a giant occipital meningocele. About 1 in every 30,000 live births is affected by DWM, with a small female predilection (9). Our case happens to be that of a female patient, which is consistent with what is illustrated in literature. On reviewing the literature, there is an age variation regarding the diagnosis of "Dandy Walker Syndrome," as it might present any time during the first year of life, as in a reported case in 2020 detailing a 10-month-old male with DWM who had a concomitant occipital cephalocele and presented with hydrocephalus and delayed developmental milestones (10). as the diagnosis was made as early as the baby's birth, while another case reported in 2021 revealed that an MRI of a female preterm newborn who was born at 36 weeks' gestation revealed linear soft tissue strands inside the lesion but no overt herniation of cerebellar or cerebral contents. Linked to the Dandy Walker deformity (7). There is also an extremely rare case report from 2002 of an adult Dandy-Walker syndrome presentation. Due to the patient's rapid onset of episodic vertigo and unilateral sensorineural deafness at age 75, a diagnosis was made. He was well for three more years before displaying the more typical adult-presenting symptoms of ataxia, abnormal gait, and cognitive impairment (2). In 1991, Bindal et al. (1) evaluated a personal series of 50 DWM cases and discovered eight cases related to OMC; despite finding only 11 more examples in the international literature, these authors estimated that this association would occur in 16% of DWM. A population-based analysis from a British survey (NorCAS) that compiles all major abnormalities in fetuses, stillbirths, and live-born children was published in 2006 by Long et al. (5). Among the posterior fossa abnormalities that occurred over the course of 18 years in a population of three million people, these investigators didn't cite any instances of DWM and OMC. Due to the widespread use of prenatal ultrasound, it's conceivable that many pregnancies ended after the diagnosis, underestimating the true prevalence of both diseases. However, a precise antenatal diagnosis may be achievable, but if missed, it can be diagnosed postnatally on a CT or plain MRI of the brain. Early treatment is crucial as it can prevent further neurological impairment (4). Clinical signs of high intracranial pressure, which is typically brought on by hydrocephalus or a posterior fossa cyst, are seen in the majority of patients. Due to a blockage in the normal cerebrospinal flow, excessive amounts of fluid accumulate in and around the brain, increasing intracranial pressure and head circumference, which ultimately results in neurological impairment. This is the cause of the association between hydrocephalus and Dandy-Walker malformation (4); however, in our case, it was evident only after surgical excision of the meningocele. Nearly all patients with DWM and OCM reported having hydrocephalus and shunting. In fact, there were several infants whose hydrocephalus was not initially visible but quickly developed following OMC closure (1,11). This demonstrates that hydrocephalus and OMC develop independently (8). However, it was hypothesized that the OMC might compensate for the elevated intracranial pressure (11).

4. Conclusion

Our case highlights a rare association between giant occipital meningocele and Dandy-Walker syndrome that is rarely discussed or reported in the medical literature. By reporting this extremely rare case of Dandy-Walker syndrome associated with a giant occipital meningocele, we hope to contribute to the creation of a database for future research so that a management protocol can be established for use by clinicians and neurosurgeons for better management of the condition.

Declaration of patient consent

Informed consent was obtained from the patient's parents to share the data for a scientific purpose.

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CRediT authorship contribution statement

Ahmed Dheyaa Al-Obaidi: Writing – original draft, Resources, Project administration, Investigation. **Ali Tarik Abdulwahid:** Supervision, Funding acquisition, Formal analysis, Data curation, Conceptualization. **Mustafa Najah Al-Obaidi:** Writing – original draft, Software, Conceptualization. **Abeer Mundher Ali:** Writing – review & editing, Writing – original draft, Funding acquisition. **Hashim Talib Hashim:** Writing – review & editing, Writing – original draft, Software, Project administration, Methodology, Investigation, Conceptualization.

Declaration of Competing Interest

There are no conflicts of interest.

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